Cardiovascular club I

11:00 AM

Thursday, February 21, 2019

1 QUALITY CARE ANALYSIS OF THE APPROPRIATENESS OF VETERANS AFFAIRS CARDIOLOGY CLINIC VISITS

A Motwani*, K Ayinapudi, T Singh, M Cassidy, AK Irimpen. *Tulane University, New Orleans, LA*

10.1136/jim-2018-000974.1

Purpose of study Southeast Louisiana Veterans Health Care System (SLVHCS) provides specialty care for New Orleans and Baton Rouge. In 2016 the New Orleans VA Cardiology Clinic had the highest number of cardiology clinic appointments per unit time across all specialties and VA practices nationally. In 2016 we determined that high visit density was due to more frequently spaced follow-up visits than necessary. In an effort to reduce unnecessary visits we implemented an algorithm of time to appropriate follow-up. This follow up study assesses efficacy of this algorithm.

Methods used A team consisting of 3 fellows, a clinic nurse, and a supervising physician was formed. We identified 220 patient encounters in the cardiology clinic during a one-week period from April 16–20, 2018 and stratified them by visit type and sub-specialty. Final chart review was done on 99 general cardiology visits seen by physicians. For each visit we identified the time of the prior and subsequent cardiology visit (1, 2, 3, 6, 9, 12, 24 months, >24 months or prn if discharged from clinic) and the reason for the follow up visit. This data was compared to our 2016 pre-algorithm data regarding appropriateness of follow up.

Summary of results 99 general cardiology visits with physicians were analyzed. 8% of patients were asked to follow up within a month compared to 21% on the previous analysis ($p=0.0082$). 26% patients were asked to follow up within 3 months compared to 47% before ($p=0.0016$). 22% were asked to follow up within 6 months compared to 17% and 11% were asked to follow up at 12 months compared to 4%. 2% of patients were discharged from the clinic. 47% of the visits occurred at closer follow-up than necessary.

Conclusions The instituted algorithm resulted in a decrease in unnecessary follow up visits at 1 month and 3 months making room for more appropriate visits. Regardless there were still visits occurring at higher frequency than necessary. Further interventions to decrease follow ups could include reiteration of criteria of follow up and discharge in clinics, continuing physician education and designation of team leaders in clinic groups.

2 SOCIODEMOGRAPHIC AND HEALTH CHARACTERISTICS PROFILE OF CONGESTIVE HEART FAILURE POPULATION: 30 DAY READMISSION AND MORTALITY AT THE VETERANS AFFAIRS CARIBBEAN HEALTHCARE SYSTEM: RETROSPECTIVE STUDY

J Mercado-Maldonado, C Rosales, S Vicenty, Z Ramos*. *VA Caribbean Healthcare System, San Juan, PR*

10.1136/jim-2018-000974.2

Abstract 2 Table 1 Demographics results

	Mean	SD
Age	82.5	8.1
BMI	26.3	6.5
Length of Stay	7.3 days	5.8 days

Abstract 2 Table 2 Comorbidities

	n	%
Male	390	99
Hispanics	384	97.7
Systolic Congestive Heart Failure	301	82.5
Hypertension	281	76.8
Type 2 Diabetes Mellitus	258	70.5
Coronary Artery Disease	245	66.9
Atrial fibrillation/flutter	211	57.7

Purpose of study Our purpose was to identify sociodemographic, comorbidities, medication use and other health variables, as well as to measure 30 day readmission incidence and mortality rates in the Congestive Heart Failure (CHF) population of the VA Caribbean Healthcare System. Also, we aimed to assess if a correlation existed between these variables, in order to target them and eventually decrease readmission rates.

Methods used We selected patients admitted to the VA Caribbean Health Care System with ICD-9 clinical diagnosis of Acute Decompensated Heart Failure from October 1st, 2012 to September 30, 2015. Over 1000 patient records were evaluated from the Computerized Patient Record System (CPRS) database. A total of 393 patients were selected and subdivided into comorbidities, laboratory values from day of readmission, medications, and 30 day mortality, among other variables.

Summary of results Preliminary results show that mean age was 82.3 years and 97% were Hispanic. Comorbidities included type 2 diabetes mellitus (70%), hypertension (76%), systolic CHF (82%), most of which belong to NYHA Class B-C, 66% had coronary artery disease, and anemia (60%). Only 40% of patients in the study were enrolled at CHF clinic, a program at VA Caribbean Healthcare System that provides follow up to patients with CHF. Mortality rate 30 days after readmission was 8%.

Conclusions By assessing a link between health and sociodemographic variables present in our patient population, we may be able to later target these, and eventually reduce 30 day readmission rates, morbidity and mortality.

3 ASPIRIN ALONE VERSUS DUAL ANTIPLATELET THERAPY FOLLOWING CORONARY ARTERY BYPASS SURGERY: COMPREHENSIVE PAIRWISE AND NETWORK META-ANALYSES OF RANDOMIZED CONTROLLED TRIALS

¹KG Hesterberg*, ¹PS Jagadish, ¹SB Latham, ^{1,2}R Shah. ¹UTHSC, Memphis, TN; ²VAMC, Memphis, TN

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Purpose of study Aspirin (ASA) therapy is class 1 indication following CABG to prevent graft occlusion. Several small-sized trials suggest that dual antiplatelet therapy (DAPT) including ASA

Abstract 3 Figure 1 A – Graft Occlusion; B – MACE; C – Major Bleeding; D – Graft Occlusion (Network Meta-analysis)

plus a p2y12 inhibitor could be more effective than ASA alone. However, the findings of individual trials have been scrutinized for small sample size and lack of statistical power. In addition, it is unclear whether some p2y12 inhibitors are more effective than others. Therefore, we performed standard pairwise meta-analyses and a Bayesian network meta-analysis with the largest sample to date by including trials from recent months.

Methods used In pairwise analyses, the pooled risk difference (RD) was estimated using a random model. For the network meta-analysis, we converted events rate to events/per 10 000 patient-days because follow-up duration varied across trials.

Summary of results Data from nine trials including 1677 patients were analyzed. Compared to ASA alone, DAPT decreased the absolute risk for graft occlusion by 7.4% (RD=−0.074; 95% CI, −0.132 to −0.017, $p=0.011$) and for MACE by 3.8% (figure 1B). It was also associated with a numerically higher risk of major bleeding, but without statistical significance (figure 1C). Results of the network meta-analysis was consistent with those from pairwise analyses (figure 1D). In addition, the rate of graft occlusion was not different between ASA +clopidogrel and ASA +ticagrelor (figure 1D).

Conclusions Following CABG, DAPT seems to be more effective at preventing graft occlusion compared to ASA alone. Clopidogrel- and ticagrelor-based DAPT seem to be equally effective for this indication.

4 RACE BASED DEMOGRAPHIC, ANTHROPOMETRIC AND CLINICAL CORRELATES OF NT-PROBNP AND MORTALITY

¹N Patel*, ¹O Gutierrez, ¹G Arora, ¹G Howard, ¹V Howard, ¹S Judd, ¹SD Prabhu, ¹E Levitan, ²M Cushman, ¹P Arora. ¹University of Alabama at Birmingham, Birmingham, AL; ²University of Vermont, Burlington, VT

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Purpose of study Population studies have shown that black race is a natriuretic peptide (NP) deficiency state. We sought to assess the effects of age, sex, body mass index (BMI) and estimated glomerular filtration rate (eGFR) on N-terminal-pro-B-type NP (NTproBNP) levels in white and black individuals, and determine whether the association of NT-proBNP levels with mortality differs across age, sex, BMI, and eGFR categories.

Methods used In a stratified random cohort from the REGARDS study, multivariable regression analyses were used to assess the aforementioned effects. Cox proportional hazard modeling was used to assess the modification in the relationship of NTproBNP levels with mortality by age, sex, BMI, and eGFR categories.

Models	Whites		Blacks	
	β Coefficient (SE)	Percentage difference (95% CI)	β Coefficient (SE)	Percentage difference (95% CI)
Age (per 10 years)	0.32 (0.03) ^a	38 (30 to 45)	0.29 (0.03) ^a	34 (22 to 43)
45-55 years	Referent		Referent	
55-65 years	0.23 (0.06) ^a	26 (11 to 42)	0.18 (0.08) ^a	20 (2 to 42)
65-75 years	0.67 (0.07) ^a	95 (72 to 125)	0.60 (0.09) ^a	82 (54 to 128)
>75 years	0.97 (0.08) ^a	164 (127 to 206)	0.83 (0.10) ^a	129 (90 to 177)
Female	0.27 (0.05) ^a	31 (20 to 43)	0.25 (0.06) ^a	28 (15 to 45)
Continuous BMI (1 kg/m ²)	-0.022 (0.01) ^a	-2.2 (-3.0 to -1.3)	-0.011 (0.005) ^a	-1.1 (-2.0 to -0.01)
BMI categories				
Normal (18.5 to 25.0 kg/m ²)	Referent		Referent	
Overweight (25.0 to 29.9 kg/m ²)	-0.12 (0.05) ^a	-11 (-21 to -2)	-0.15 (0.08) ^a	-14 (-25 to -1)
Obese 1 (30.0 to 34.9 kg/m ²)	-0.27 (0.07) ^a	-24 (-33 to -13)	-0.28 (0.09) ^a	-24 (-36 to -10)
Obese 2 (35.0 to 45.0 kg/m ²)	-0.37 (0.09) ^a	-31 (-43 to -17)	-0.27 (0.10) ^a	-24 (-37 to -7)
Continuous eGFR (per 10 mL/min/1.73 m ²)	-0.12 (0.02) ^a	-11 (-14 to -8)	-0.12 (0.02) ^a	-11 (-14 to -9)
eGFR categories (mL/min/1.73 m ²)				
≥90	Referent		Referent	
60 to 90	0.18 (0.06) ^a	20 (7 to 35)	0.11 (0.07)	12 (-2 to 26)
<60	0.61 (0.10) ^a	84 (57 to 122)	0.91 (0.11) ^a	148 (99 to 206)

A

B

Abstract 4 Figure 1 A – Demographic, Anthropometric and Clinical correlates of plasma NT-proBNP levels stratified by race: Multivariable Regression Results (Multivariable model adjusted for age (when sex, BMI, and eGFR are exposures), sex (when age, BMI, eGFR are exposure), body mass index (BMI) (when age, sex, eGFR are exposures), estimated glomerular filtration rate (eGFR) (when age, sex, BMI are exposures), exercise, smoking, alcohol, systolic blood pressure, antihypertensive medication, aspirin use, hyperlipidemia, diabetes, stroke, transient ischemic attack, atrial fibrillation, coronary artery disease, left ventricular hypertrophy, neighborhood socioeconomic status at census block level, and heart failure); Panel B – Baseline NT-proBNP and Mortality Stratified by Sex and Race. * $p<0.05$, ** $p<0.01$, # $p<0.001$.

Summary of results In 4106 (27,678 weighted) participants, every 10 year higher age was associated with 38% (95% CI: 30% to 45%) and 34% (95% CI: 22% to 43%) higher NTproBNP levels in whites and blacks, respectively. Female sex was associated with 27% (95% CI: 20% to 43%) higher NT-proBNP levels in whites and 28% (95% CI: 15% to 45%) higher in blacks. There was a significant linear inverse relationship between BMI and NTproBNP in whites and a nonlinear inverse relationship in blacks. Whites and blacks had a similar nonlinear inverse relationship between eGFR and NTproBNP (figure 1, Panel A). The risk of all-cause mortality predicted by NT-proBNP levels differed by sex in whites ($p=0.03$ for interaction), but not in blacks (figure 1, Panel B). **Conclusions** The association of age and sex with NTproBNP levels was similar in blacks and whites but the shape of the BMI relationship differed by race. NTproBNP level was a stronger predictor of mortality in white females than males.

5 COOPERATIVE ROLE OF MYELOPEROXIDASE AND NOX4 IN THE PATHOGENESIS OF ABDOMINAL AORTIC ANEURYSM

L Reid*, T Horimatsu, M Ogbi, S Li, D Weintraub, D Fulton, H Kim, N Weintraub. *Medical College of Georgia, Augusta, GA*

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Purpose of study Myeloperoxidase (MPO)-induced oxidative stress plays an important role in the pathogenesis of abdominal aortic aneurysm (AAA). However, mechanisms whereby MPO promotes AAA remain to be determined. Given that MPO utilizes hydrogen peroxide (H₂O₂) to generate a powerful oxidant HOCl, and Nox4 is a major producer of vascular H₂O₂, we hypothesized that MPO cooperates with Nox4 to promote vascular oxidative stress and AAA formation. **Methods used** Using two distinct animal models of AAA (AngII infusion and CaCl₂ application), we found that either MPO

or Nox4 knockout (KO) mice were protective against AAA formation. Interestingly, MPO accumulation in aortic wall was significantly reduced in Nox4 KO mice, raising possibility that Nox4 expressed in vascular wall may regulate uptake of MPO in the aorta. To examine whether MPO is being taken up into the blood vessel wall during AAA formation, abdominal aortas isolated from LDLR KO mice were incubated *in vitro* with saline or AngII (100 nM) for 12 hour, and then, exposed to MPO for 2 hour.

Summary of results Notably, *in vitro* AngII pretreatment significantly increased aortic MPO uptake (2.8 ± 0.4 fold) as compared to control. Infusion of AngII *in vivo* for 3 days (a time frame which precedes AAA formation) likewise enhanced aortic MPO uptake (1.8 ± 0.2 fold) with increased expression of Nox4 (4.7 ± 1.3 fold), suggesting that MPO uptake into the aorta is stimulated early during the development of AngII-induced AAA. More interestingly, aortas from Nox4 KO mice blocked the action of AngII pretreatment to promote aortic MPO uptake. Likewise, aortic Nox4 expression at 3 days after CaCl₂ treatment was also increased in abdominal (3.4 ± 0.7 fold), but not thoracic aorta, with concomitant increase of MPO uptake in abdominal (3.2 ± 0.2 fold), but not thoracic aorta.

Conclusions This study demonstrates that MPO uptake into vascular wall co-localizes with induction of Nox4, and Nox4 deficiency decreases vascular MPO uptake, implying a novel mechanistic linkage whereby MPO and Nox4 cooperate to promote vascular oxidative stress in AAA pathogenesis.

6 BLOCKADE OF CARDIAC HYPERTROPHY AND FIBROSIS BY TGF-BETA 1 RECEPTOR ANTAGONIST IN *NPR1* GENE-KNOCKOUT MICE

R Samivel, H Chen, U Subramanian, H Zhao, KN Pandey*. Tulane University Health Sciences Center, New Orleans, LA

10.1136/jim-2018-000974.6

Purpose of study The object of this study was to determine whether transforming growth factor-beta 1 receptor antagonist inhibits the cardiac hypertrophy and fibrosis in *Npr1* gene-knockout mice.

Methods used The *Npr1* null mutant (*Npr1*^{-/-}, 0-copy), heterozygous (*Npr1*^{+/-}, 1-copy), and wild-type (*Npr1*^{+/+}, 2-copy) mice were orally administered with transforming growth factor-beta 1 (TGF-β1) receptor antagonist GW788388 (1 mg/kg/day) for 28 days, the hearts were isolated, weighed, and used for quantification of hypertrophic and fibrotic markers using real time quantitative reverse transcription-polymerase chain reaction (qRT-PCR) and Western blot analyses. Together, systolic blood pressure (SBP), heart weight-to-body weight (HW/BW) ratio, and cardiac fibrosis were analyzed.

Summary of results The results of the present study demonstrate that HW/BW ratio was significantly higher in *Npr1* knockout mice than wild-type control mice. The *Npr1*^{-/-} mice showed significant induction of cardiac hypertrophy and fibrosis with markedly induced expression of hypertrophic marker collagen-1α (3.5-fold) and fibrotic markers monocyte chemoattractant protein (4-fold), connective tissue growth factor (CTGF, 5-fold), alpha-smooth muscle actin (α-SMA, 4-fold), and SMAD proteins (SMAD-2, 5-fold; SMAD-3, 3-fold) compared with wild-type control mice. In addition, the expression of phosphorylated extracellular-regulated kinase 1/2 (pERK1/2) proteins were also increased in *Npr1*^{-/-} mice.

On the other hand, *Npr1*^{-/-} mice treated with GW788388 showed significant inhibition of cardiac hypertrophy and fibrosis and reduced expression of fibrotic markers and SMAD proteins compared to vehicle-treated wild-type mice. In contrast, the expressions of pERK1/2 proteins were unaffected in drug-treated animals.

Conclusions The results of the present study suggest that development of cardiac hypertrophy and fibrosis in *Npr1*^{-/-} mice is regulated independently through TGF-β1-mediated SMAD-dependent pathways.

7 PUSHING THE JUNCTION GAPS IN PAIRED MYOCYTES TO EXTREME EFFICIENCY (SUPERCONDUCTIVITY) BY INTRACELLULAR ENALAPRIL (ANGIOTENSIN CONVERTING ENZYME INHIBITOR) WILL IMPROVE THE FUNCTION OF PAIRED HEART MYOCYTES

PI Altieri*, HL Banchs, W De Mello, N Escobales. University of Puerto Rico, Medical Sciences Campus, San Juan, Puerto Rico

10.1136/jim-2018-000974.7

Purpose of study The purpose of this study was to measure if superconductivity (S.C.) exists intracellularly and to find out, if Enalapril (E.) injected intracellularly in rapid sequence and increasing dose will produce a S.C. state.

Methods used S.C. is a concept of pushing materials to the extremes (Lonzarich) to get maximal efficiency. Applying this concept, experiments were done in intracellularly isolated paired myocytes measuring electrical conduction across junction gaps (G.I.) in controlled temperature and pressure state, after increasing the dose of E. (.25–1.25 ug/ml) injected in a single dose in a rapid way.

Summary of results The maximal increase in G.I. was 80% ($p < 0.005$) until a plateau was reached. We think a S.C. state was induced until a block of the renin angiotensin system was achieved or by blocking the quantum state through ionic or atom blocking effect or receptor blocking. The full mechanism will be discussed.

Conclusions This shows that a S.C. system is produced intracellularly improving G.I. and as a consequence improving left ventricular function by cell coupling. We speculate that mechanical energy is translated into chemical reactivity, improving cell function, still this is an enigma.

8 METABOLOMIC ANALYSIS OF LEFT VENTRICULAR REMODELING: THE BOGALUSA HEART STUDY

^{1,2}AC Razavi*, ^{1,2}C Fernandez, ¹X Mi, ¹J He, ^{1,2}L Bazzano, ¹J Nierenberg, ¹S Li, ¹TN Kelly. ¹Tulane University School of Public Health and Tropical Medicine, New Orleans, LA; ²Tulane University School of Medicine, New Orleans, LA

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Purpose of study Though left ventricular (LV) remodeling patterns are associated with an increased risk of future cardiovascular events compared to that of normal LV architecture, the metabolic determinants associated with left ventricular mass index (LVMI) and relative wall thickness (RWT) are unknown. We conducted a metabolome-wide association study to identify LV remodeling-related metabolites.

Methods used We examined 1,053 Bogalusa Heart Study adults (65.05% whites, 34.95% blacks). Echocardiographic measurements of LV diastolic diameter, LV posterior wall

Abstract 8 Table 1 Novel metabolites associated with left ventricular mass index

Pathway	Subpathway	Metabolite	Whites ¹		Blacks ¹	
			Beta (SE)	P-value	Beta (SE)	SE
Amino Acid	Glutamate Metabolism	Beta-Citrylglutamate	2.20 (0.50)	1.10E-05*	0.84 (0.92)	3.60E-01
	Histidine Metabolism	1-Methylimidazoleacetate	1.95 (0.47)	4.00E-05*	1.23 (0.80)	1.20E-01
	Histidine Metabolism	N-Acetylhistidine	2.73 (0.65)	3.00E-05*	1.55 (1.07)	1.50E-01
	Methionine, Cysteine, S-Adenosylmethionine, and Taurine Metabolism	N-Acetylmethionine	3.73 (0.81)	4.70E-06*	1.69 (1.17)	1.50E-01
Cofactor and Vitamin	Tocopherol Metabolism	Gamma-CEHC Glucuronide	0.82 (0.20)	4.10E-05*	0.08 (0.12)	5.20E-01
Lipid	Sphingolipid Metabolism	Sphingomyelin (d17:2/16:0, d18:2/15:0)	-4.20 (4.77)	1.70E-05*	-4.08 (2.36)	8.80E-02
Nucleotide	Purine Metabolism, Adenine Containing	N6-Succinyladenosine	1.64 (0.16)	8.90E-06*	0.48 (0.49)	3.20E-01
Xenobiotic	Benzoate Metabolism	O-Methylcatechol Sulfate	0.67 (0.37)	2.10E-05*	0.23 (0.21)	2.70E-01

*Statistically significant p-value (Bonferroni corrected p-value=4.16E-05). 1. Adjusted for age, sex, education, cigarette smoking, alcohol drinking, low density lipoprotein, high density lipoprotein, serum triglycerides, HbA1C, average systolic blood pressure, and body mass index.

diastolic thickness, and interventricular septum diastolic thickness were used to calculate left ventricular mass (LVM) and RWT for each individual. LVM was indexed to height^{2.7} to calculate LVMi. Untargeted metabolomic analysis of fasting serum samples was conducted. Multivariable-adjusted linear regression models were employed to assess the relationship of metabolites with RWT and LVMi.

Summary of results Following quality control, 1202 metabolites were tested for association with RWT and LVMi. After Bonferroni correction, 8 total metabolites, derived from amino acid, cofactor, lipid, nucleotide, and xenobiotic pathways, were significantly associated with LVMi in whites with consistent effect direction in blacks (table 1). No significant associations were found between metabolites and RWT.

Conclusions This analysis identified novel metabolites associated with LVMi, suggesting that serum metabolites may occupy a critical role in LV remodeling.

medical management. Three months follow up MRI revealed reduction in size of tumor and pituitary bleed.

Discussion Myxedema coma and pituitary apoplexy are well known life threatening endocrine emergencies. Coincidence of these entities is exceedingly rare. Myxedema coma occurring as a result of pituitary lesion is much less seen and occurs as a result of severe hypothyroidism. Pituitary apoplexy generally presents with sudden onset of excruciating headache, nausea, visual disturbances. However, our patient had atypical presentation with no classic apoplectic symptoms but did have biochemical evidence of hormonal deficiencies and clinical manifestation of severely reduced thyroid function. Life threatening bradycardia and hypotension could be a part of early syndromic presentation.

Conclusion Our case emphasizes that early recognition of myxedema coma and its cardiac complication is important in improving survival outcomes. Establishing early diagnosis is necessary as mortality is high ranging from 20%–60%.

9 AN UNUSUAL CASE OF MYXEDEMA COMA PRESENTING AS SEVERE BRADYCARDIA AND HYPOTENSION SECONDARY TO PITUITARY APOPLEXY

S Bhogal*, T Bhandari, K Mawa, T Paul. *East Tennessee State University, Johnson City, TN*

10.1136/jim-2018-000974.9

Case report A 63 year old male presented to our hospital with severe bradycardia, hypothermia and hypotension. On presentation, his temperature was 85.4 C, blood pressure (BP) of 80/61 mm Hg, respiratory rate of 18/min and heart rate of 40/min. On physical examination, he was slow to respond but arousable. EKG showed severe bradycardia, right bundle branch block and prolonged QTc interval of 521 msec. Keeping high index of suspicion for myxedema coma based on his clinical presentation, he was started on levothyroxine and hydrocortisone. External warming with blankets was provided. Initial labs revealed TSH 0.172 µIU/ml, free T4 0.41 ng/dl and cortisol of 4.5 ug/dl. Six hour after initiation of treatment, there was resolution of bradycardia, prolonged QTc and hypotension with HR of 68/min, QTc of 464 msec and BP of 104/62 mm Hg, respectively. Emergent MRI revealed suprasellar mass measuring 2.5×2.2×2.0 cm, abutting and uplifting the optic chiasma, compatible with hemorrhagic pituitary microadenoma. Surgery was offered but patient opted for

Adult Clinical Case Symposium

12:00 PM

Thursday, February 21, 2019

10 CALCIFIC LONGUS COLLI TENDINITIS: A RARE CASE OF NECK PAIN

A Singh*, A Gural, K Cutts. *Texas Tech Univ HSC Amarillo, Amarillo, TX*

10.1136/jim-2018-000974.10

Introduction The differential diagnosis of acute neck pain presenting with dysphagia is wide and not limited to retropharyngeal abscess, trauma and soft tissue swelling. One rare and underdiagnosed entity is Calcific Longus colli tendinitis, which is acute neck pain associated with torticollis, dysphagia and odyndophagia.

Case report A 30-year-old female presented with acute onset of severe neck pain which progressed over 3–4 days. The pain was described as sharp, non-radiating, aggravated by neck movement without relieving factors. She developed dysphagia to both solids and liquids progressing to odyndophagia and torticollis in a couple of days. Besides subjective fever ROS was

negative. She was treated for a dental abscess a month ago with clindamycin for 10 day. She had poor dentition and limited neck movement in all directions with paraspinous tenderness. WBC of 13.5 K. CT scan without contrast showed prevertebral fluid collection at C2-C5. Vancomycin and ciprofloxacin were initiated for possible prevertebral abscess. MRI with contrast showed calcific deposits at anterior to C2-C3 with prevertebral edema. NSAIDs and steroids were started, and she was discharged after resolution of her symptoms.

Discussion Calcific tendinitis of the longus colli (CTLTC) is a rare clinical presentation of neck pain in young patients in their 3rd to 6th decade. This is an underdiagnosed entity with very little data on its incidence and prevalence. Calcium hydroxyapatite crystals deposits in the longus colli muscle is hypothesized to rupture and cause local inflammation with fluid collection in the prevertebral/retropharyngeal space. The contributing factors for calcium deposition could be previous trauma, URI or repetitive stress. This classically presents with triad of neck pain and stiffness, torticollis and odynophagia (with occasional dysphagia). Radiographic findings are used to confirm the diagnosis. CTLTC is self-limiting with expected spontaneous resolution over a 1-to-2-week period. Treatment involves symptomatic support with analgesia, anti-inflammatory medicine and avoidance of aggravating neck movements. Surgical incision and drainage should be avoided.

11 AN UNCOMMON CAUSE OF OTORRHEA

S Sanders*, LS Engel, J Hart. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.11

Introduction While otorrhea most commonly is the result of infection, it is important to consider other diagnoses if conventional therapies are unsuccessful in resolving symptoms. Differential diagnosis includes otitis media, otitis externa, trauma, foreign body, neoplasm, granulation tissue.

Case A 63 year old man with poorly-controlled type 2 diabetes presented to primary care clinic with a complaint of left-sided otalgia for five days. He denied facial numbness, dysphagia, tinnitus, vertigo, fever. Cerumen impaction was found on exam, so the patient was given instruction for irrigation followed by neomycin-polymyxin B sulfates-hydrocortisone otic drops to treat presumed otitis externa. The ear pain improved, but he reported development of otorrhea after several months. The patient was referred to ENT, where exam demonstrated a tender posterior external auditory canal mass, thought to be a furuncle. Cultures grew candida parapsilosis. He was prescribed systemic antibiotics and antifungals without improvement. Computed tomography of his orbits/face showed an enhancing focus in left external auditory canal extending into parotid consistent with mass vs. infection. Despite initial resistance, the patient agreed to biopsy and was diagnosed with adenoid cystic carcinoma. He underwent left lateral temporal bone resection, superficial parotidectomy, neck dissection, and significant reconstructive surgery. After adjuvant radiation, PET scan shows resolution of neoplasm and surveillance scans without recurrence.

Discussion Ear canal cancer is an uncommon cause of otorrhea and may be indistinguishable from otitis externa in early stages. However, lack of response to conventional treatment should prompt further investigation and expert consultation. Salivary gland cancers make up only 1% of all cancer diagnoses, but otorrhea is not an uncommon presenting symptom.

This patient's stage IVa cancer with local invasion into the ear canal is associated with >50% 5 year mortality, but aggressive treatment was available due to persistent investigation into his presenting complaint.

12 HYPOKALEMIC THYROTOXIC PERIODIC PARALYSIS IN TWO YOUNG ADULTS

C Stallings*, C Foster, A Wynn, A Diaz-Thomas. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.12

Case report Periodic paralysis is an uncommon condition which may be either familial secondary to a genetic channelopathy or acquired in the setting of a hypokalemic, hyperthyroid state. Both etiologies can lead to acute weakness difficult to distinguish from more prevalent conditions. This diagnosis is more commonly encountered in men (95%) than women with a predilection for those of Asian descent. We present two unusual cases of hypokalemic, thyrotoxic periodic paralysis (TPP), both in young Caucasian patients with underlying developmental disorders. In these cases, the presentation was difficulty with ambulation: acute weakness for a seventeen-year-old female and subacute weakness for a twenty-four-year-old male. Both cases had potassium levels that were very low. The seventeen-year-old female had an abnormal EKG with type 3 Brugada pattern with broad T waves. Both cases had hypothyrotropinemia with hyperthyroxinemia. There were no typical findings of Graves' disease on physical examination for either patient. The history was incomplete given their degree of developmental delay. Repletion of the potassium and treatment of the underlying thyrotoxicosis resolved the paralysis. The weakness associated with TPP usually resolves with correction of the hypokalemia. Potassium should be administered slowly in these patients as there is a high risk for rebound hyperkalemia. Maintaining a euthyroid state can prevent most future attacks. TPP should be considered in all cases of acute weakness and hypokalemia with or without clinical hyperthyroidism. These cases reinforce the importance of a thorough history and physical exam in a patient with a developmental disorder who may not be able to identify specific symptoms until in extremis.

13 LEVAMISOLE INDUCED NECROSIS SYNDROME

GD Bedanie*, L Dobbe, M Elmassry, M Phy. *Texas Tech University Health Science Center, Lubbock, TX*

10.1136/jim-2018-000974.13

Introduction Levamisole induced necrosis syndrome (LINES) is rarely documented in medical practice. Prevalence is not well established as it is not common knowledge among physicians. Levamisole is an anti-helminthic and immunomodulatory agent, pulled from the US in 1999 due to reports of agranulocytosis and vasculitis. It remains available worldwide and in the majority of the US black market used for cocaine adulteration. By 2009, the US Drug Enforcement Administration estimated 70% of the cocaine seized contained levamisole. Levamisole appears to have noradrenergic and dopaminergic receptor affinity, and partially metabolized into an amphetamine-like compound. It presumably acting as a bulking agent and effect enhancer. The first case report was published in 2009.

Case summary A 25-year-old woman with a history of crack cocaine abuse with multiple admissions over the last 6 years for recurrent non-healing, necrotic wounds. Her wounds began to appear several months after she started using crack cocaine in 2012. Different treatments were unsuccessful. The patient reported wound closure when stopping cocaine. She presented with new wounds on upper chest and lower extremities. On examination vital signs were normal, cachectic appearing patient and multiple necrotic wounds ranging from 3–14 cm on lower extremities. Laboratory studies revealed positive anti-neutrophilic cytoplasmic antibody (ANCA), anti-myeloperoxidase antibody (MPO) and anti-proteinase-3 antibody (PR3). Patient was treated with topical clobetasol, wound care and cessation counseling.

Conclusion Levamisole induced necrosis syndrome is not commonly seen in medical practice. A high index of suspicion is very important in crack cocaine abusing patients with non-healing necrotizing and recurrent wound. Delays in diagnosis can lead to complications, higher costs, unnecessary and aggressive treatment modalities. MPO, PR3 and ANCA are often positive. The simultaneous presence of positive MPO and PR3, with typical cutaneous findings, is sensitive and specific for LINES. Successful longterm therapy include cocaine use cessation, counseling and skin grafting for extensive necrosis.

14 THOUGHTS OF THE GRAVE AFTER TREATING GRAVES

LN Bailey*, RD Smalligan. UAB-Huntsville, Huntsville, AL

10.1136/jim-2018-000974.14

Case report An 18-year-old male with a hx of Graves disease presented 4 wks after radioactive iodine ablation. His methimazole was stopped per protocol 2 days before the ablation and resumed shortly thereafter. About 2 weeks later he developed what was thought to be a URI with rhinorrhea and cough that progressed to include fever, tachycardia, hypertension and clammy skin. Meds on admission included methimazole 10 mg BID and atenolol 50 mg QHS. PE: alert, BP 148/81, P 125, R 28, sats 99% on RA, HEENT – eyes nl with no lid lag or proptosis, neck with mild thyromegaly, no nodules, warmth, or erythema; lungs clear, heart RRR without murmurs, neuro exam nonfocal. While in the ED he admitted to recent suicidal ideation. Labs showed WBC 4 k, 55 s, 29 L, Hgb 14, free T3 and free T4 higher than measurable levels and TSH below detectable limits. The patient was admitted on telemetry, his methimazole was increased to 10 mg TID and propranolol 30 mg TID was initiated. Over a two day hospital stay his vitals normalized and he had no further suicidal thoughts.

Discussion Graves disease is the most common cause of hyperthyroidism in adolescents and young adults. Treatment options include pharmacotherapy, usually with methimazole, radioactive iodine ablation therapy or surgery. The advantage of radioactive iodine therapy is its ability to destroy the hyperactive thyroid tissue without the invasiveness of surgery; it is also less expensive. Most patients develop hypothyroidism after radioablation and are then able to be managed with simple, well-tolerated, inexpensive levothyroxine which has minimal side effects. A small subset of patients, however, can develop rebound hyperthyroidism. This can occur weeks to months after ablative therapy and effectively managed with increased methimazole as was done in our case. Thyrotoxicosis has been observed following trauma, non-thyroidal surgery, and infection. Although our patient's hyperthyroid state was

most likely one of the uncommon post-ablative reactions, his recent history of URI-like symptoms does suggest a possible alternative etiology: post-viral hyperthyroidism. Physicians must consider hyperthyroidism (even paradoxical as in this case) when new psychiatric symptoms, including suicidal ideation occur. Fortunately the suicidal ideation and other sxs usually resolve once the condition is treated.

15 LEIOMYOMATOSIS PERITONEALIS DISSEMINATA WITH ENDOMETRIOSIS

^{1,2}A Reddy*, ¹J Kaur, ¹J Neill. ¹University of Mississippi Medical Center, Jackson, MS; ²University of South Alabama, Mobile, AL

10.1136/jim-2018-000974.15

Introduction Leiomyomatosis peritonealis disseminata (LPD) is a rare benign disorder characterized as numerous smooth muscle nodules in the peritoneal cavity. LPD is difficult to diagnose by clinical evaluation due to unknown etiology and incidental findings. To date, less than 200 cases have been reported and one other case of LPD with endometriosis and ascites has been documented. LPD commonly occurs in reproductive age and rarely seen in postmenopausal women. We present LPD in a postmenopausal woman with endometriosis.

Case report A 54-year-old female presented with pain, nausea, vomiting and weight loss. Physical exam demonstrated a palpable mass arising from adnexa. CT scan showed: 6.4 cm large multi-loculated heterogeneous mass in the right adnexa concerning for ovarian carcinoma, large 17 cm fibroid uterus, extensive peritoneal carcinomatosis, a small volume of ascites and numerous hypo-dense metastatic liver lesions. CT guided biopsy of peritoneal carcinomatosis of omentum revealed spindle cell neoplasm immunophenotypically consistent with leiomyoma and stained positive for desmin, caldesmon, smooth muscle actin, and Ki-67. Microscopy sections of the omental mass showed proliferation of poorly circumscribed spindle cells arranged as nodules in short interlacing and haphazard arrangements in omentum. No cytological atypia was seen in the spindle cell proliferation. No mitotic activity identified. Sections of posterior bladder mass showed endometriosis with an adenomyoma pattern of smooth muscle hypertrophy. Immuno-histochemistry confirmed strong positive reactivity in the spindle cells to desmin and WT1. Findings consistent with LPD.

Discussion LDP is not considered in the differential diagnosis of multiple peritoneal nodules because of its low incidence and unfamiliarity among clinicians. The treatment for this benign condition is conservative because in most cases malignant transformation is rare and tumors regress. For conclusive diagnosis of LPD, radiological imaging proves challenging, thus direct sampling is required to exclude any malignancies. LPD is problematic to identify clinically, so diagnosis is dependent on pathological and surgical results, nevertheless, the prognosis is often good.

16 AN UNCOMMON DERMATITIS IN THE PRIMARY CARE SETTING

J Bassett*, M Kleinman. University of Tennessee Health Science Center, Memphis, TN

10.1136/jim-2018-000974.16

Case report A 43-year-old male with past medical history of Factor V Leiden and two previous deep vein thromboses (DVTs) requiring anticoagulation presented to an academic

Abstract 16 Figure 1 Pinpoint lesions forming linear streaks on the lower extremity

resident practice with acute onset of rash. It started two days prior on his feet and lower extremities and spread to the upper extremities and trunk. He denied pruritus or pain, tick bites, new skin products or detergents. His only medication was rivaroxaban. No family members were affected. His physical exam was normal, aside from the rash on his extremities and trunk. It was petechial with erythematous, pinpoint lesions in linear groups. There were no purpura or signs of bleeding. Despite his well appearance, his rash was concerning for serious illnesses, including thrombocytopenia, platelet dysfunction or idiopathic thrombocytopenic purpura. His labs were unremarkable, and once they resulted, further history was taken. He dined out three days prior to onset and ate shiitake mushrooms, which we believe caused his rash. Over time, the rash developed more streaks with mild pruritus and ultimately resolved in a week without treatment. This case describes the presentation of a rare toxic-mediated dermatitis that results from the consumption or handling of raw or undercooked shiitake mushrooms (*Lentinula edodes*), which are commonly used in Asian cuisine. Given the characteristic appearance of this rash, early clinical diagnosis can limit additional unwarranted diagnostics as in our case.

17 SYSTEMIC LUPUS ERYTHEMATOSUS PRESENTING AS ACUTE SYSTOLIC HEART FAILURE

K Bano*, R Parikh, G Krishnan, M Konda, LG Johnson. *UAMS, Little Rock, AR*

10.1136/jim-2018-000974.17

Case report Systemic lupus erythematosus (SLE) is a chronic inflammatory illness, with a myriad of clinical presentations.

Cardiac involvement in SLE is frequent, with pericarditis being the most common manifestation. However, myocarditis with subsequent cardiomyopathy is rare.

A 19 year old caucasian male with no significant past medical history, presented with complaints of dyspnea on exertion, weakness, 40 lbs weight loss over 1 month, pedal edema, orthopnea, chills, non-productive cough, high grade fevers for 1 week and a non pruritic rash on both cheeks. He denied recent viral illness, sick contacts or travel outside the US. On presentation, he was tachycardic, normotensive and hypoxemic requiring supplemental oxygen. On examination, he had pedal edema and rales on lung auscultation. Labs revealed pancytopenia with WBC 2200/uL, hemoglobin 9 g/dL, platelet 93,000/uL, troponin 0.21 ng/ml, BNP 623. Peripheral blood smear showed normochromic anemia without schistocytes. ECG showed T-wave inversions in infero-lateral leads. Trans-thoracic echocardiogram showed an LV EF of 15%–20%, mild to moderate mitral regurgitation and a small pericardial effusion. Further work up revealed positive ANA screen, ANA titer 1:640, Anti DS DNA positive: 226 IU/mL, low C3 and C4 levels, negative anti-phospholipid antibodies. Diagnosis of SLE was made based on 4/11 ACR criteria positive for SLE. Acute systolic heart failure was deemed secondary to Lupus Myocarditis after other common causes of myocarditis were ruled out. Treatment was initiated with plaquenil and high dose methylprednisolone. Repeat echocardiogram performed 3 days later showed an improvement in LV EF to 30% and decrease in size of the pericardial effusion with improvement in pancytopenia.

Myocarditis and systolic dysfunction secondary to SLE are relatively rare causes of new onset heart failure. These may easily be overlooked as a potential cause of systolic heart failure in a young patient. Delay in diagnosis and initiation of treatment may lead to progression to overt heart failure and cardiogenic shock, with reported cases of sudden death secondary to complications. Prompt recognition and initiation of treatment with high dose glucocorticoids, immunosuppressive agents or IVIG can be life saving.

18 STRONGYLOIDIASIS INFECTION IN AN IMMUNOCOMPROMISED HOST

C Ashangari*, S Siddiqui, S Prakash, B Khandheria. *Texas Tech Univ HSC Amarillo, Amarillo, TX*

10.1136/jim-2018-000974.18

Introduction Strongyloidiasis has been recognized as one of the life-threatening parasitic infections in the immunocompromised patients. We report an intestinal infection case of *Strongyloides stercoralis* in a 53-year-old man.

Case A 53 years old African American male from Sudan was referred to the GI clinic from the PCP due to rectal bleeding since few weeks and anemia. He was noted to have a hemoglobin of 6.6 and received 2 units of PRBCs. PMH is significant for myelodysplastic syndrome and myelofibrosis. His medications were Hydrea and Jakafi which were discontinued recently. Physical exam was unremarkable except for splenomegaly. Colonic biopsy showed Inflammatory pseudopolyp in the cecum, ulceration and granulation tissue with eosinophilic inflammation in transverse colon, hyperplastic polyp with eosinophilic inflammation in rectal polyp associated with parasitic worms morphologically consistent with *Strongyloides*

stercoralis. There was no evidence of dysplasia and malignancy. Patient was started on Ivermectin after which he was doing better. Repeat labs in 6 weeks showed a hemoglobin of 10.4 and hematocrit of 32.

Discussion Most of the reported cases of strongyloidiasis worldwide involved immunocompromised hosts including AIDS, leukemia, lymphoma, solid organ transplant recipient, long term corticosteroid usage and chronic pulmonary disease. In this report, the patient too, is categorized as immunosuppressed individual by having myelodysplastic syndrome and myelofibrosis. In relation to that, nematode transmits its molting signals by means of molting hormones (ecdysteroids). In healthy adult, level of ecdysteroid-like substances is generally negligible, but in immunocompromised host, it could be reversed. Prognosis tends to be grave in this population and delay in diagnosing strongyloidiasis frequently results in death, despite vigorous treatment.

19

A RARE CASE OF IDIOPATHIC SPONTANEOUS RETROPERITONEAL HEMORRHAGE

J Ruiz*, W Kogler, C Canha, M Ganji. *UF Health Jacksonville, Jacksonville, FL*

10.1136/jim-2018-000974.19

Case 41 year-old male with no past medical history presents with sudden onset severe right flank and abdominal pain since 2 hour priors to presentation. Pain is constant, 10/10, located in right lower quadrant and radiates to his right flank and back. Denies any trauma, fever or chills, dysuria, hematuria, polyuria, diarrhea, constipation, hematochezia, dizziness, chest pain or SOB. Computed tomography (CT) abdomen and pelvis revealed right perinephric hematoma and right retroperitoneal hemorrhage with evidence of active arterial extravasation. Interventional radiology consulted and underwent angiography which revealed active contrast extravasation arising from a distal intraparenchymal branches in the inferior pole. Patient underwent embolization and repeat angiogram demonstrated resolution of active contrast extravasation. He was discharged four days after embolization since hemoglobin remain stable and no active bleeding noticeable.

Discussion Spontaneous Retroperitoneal Hemorrhage (SRH) is a very rare clinical entity. There are very few reported cases in the literature. The overall incidence of SRH is unknown. The most common cause of SRH is neoplasm, most commonly renal cell carcinoma and angiomyolipomas. Other common causes include anticoagulation with warfarin, heparin, and clopidogrel, aneurysm, vasculitis and in extremely rare cases, idiopathic. Our patient falls in the idiopathic SRH category which as previously mentioned is an exceedingly rare finding. Our patient was a 42 year old male, had no history of malignancy, no prior surgeries, trauma or invasive procedures, and was not on anticoagulation. These patients usually present with nonspecific symptoms such as acute flank pain, signs of hematuria and abdominal pain. Definitive diagnosis can be achieved with non-contrast CT. Stable patients can be managed expectantly by fluid resuscitation with packed red blood cells and fresh frozen plasma. Historically these patients have been managed surgically, however interventional radiology with intra-arterial embolization is now the treatment of choice for these patients. SRH is a

very rare, very dangerous clinical presentation, and it is important that it be included in the differential for patients with similar presentations as a delay in diagnosis could be devastating.

Neonatal Case Report Symposium

12:00 PM

Thursday, February 21, 2019

20

NEWBORN IVC THROMBUS WITH SUSPECTED PROTEIN C DEFICIENCY

SE Hudson*^{1,2}. ¹UAB (University of Alabama at Birmingham), Birmingham, AL; ²Children's of Alabama, Birmingham, AL

10.1136/jim-2018-000974.20

Case report A Caucasian newborn male born at 38 weeks gestation via SVD to a 27 yo G1P0 female with APGARs of 8/9 developed increased work of breathing requiring supplemental oxygen hours after birth. Sepsis work up showed acidosis and hypoxia on blood gas and normal initial chest radiograph and blood cultures. Upon transfer to UAB RNICU, the child had developed gray skin color with mottling, absent femoral pulses, hypotension, acidemia with lactate >24 mmol/L, and en route had received boluses with multiple resuscitative fluids and prostaglandins. A bedside echocardiogram showed a thrombus in the inferior vena cava (with blood flow peripherally around the clot), very limited left ventricular function, and possible left coronary thrombus. Other labs performed after transfer to Children's of Alabama CVICU included a troponin of >50 ng/mL, ALT of 270 U/L, AST of 1195 U/L, INR of 2.4, and severe liver failure. Despite multiple efforts, the infant developed sepsis, anuria, and multiple life-threatening conditions and care was withdrawn several days later via parental consent.

Pathology: Autopsy revealed an extensive left ventricular myocardial infarct with ventricular dysfunction and failure resulting from an organized occlusive left main coronary artery thrombus. Other factors included a focal acute pneumonia in the left lower lobe, multiple renal infarcts with congestion, hemorrhage and acute oliguric failure due to renal vein thrombi, and hypoxic-ischemic encephalopathy with infarction and encephalomalacia. A Hereditary Thrombophilia Panel (Invitae) revealed a PROC gene variant associated with protein C deficiency. Protein C activity was 12% (normal 70%–180%), protein S was 30% (normal 70%–150%), and Anti-thrombin III activity was 30% (normal 39%–87%).

Background: Protein C is a vitamin K-dependent glycoprotein that functions as a natural anticoagulant. This case presented with an IVC thrombus (relatively uncommon) and also coronary artery, renal, and cerebral thromboses which have been reported elsewhere.

Recommendation: Coagulopathic genetic testing is recommended for any individuals with family members affected by similar events, as those with PROC genetic defects have an increased risk of venous thromboembolic disorders in later life.

21 NEONATE WITH CARDIOFACIOCUTANEOUS SYNDROME

S Peravali*, KA Willis, AJ Talati, P Zaveri. *The University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.21

Case report Cardiofaciocutaneous syndrome (CFC) is a rare genetic disorder with only a few hundred cases reported worldwide. It is an autosomal dominant condition due to mutation in genes in the RAS-MAPK signaling pathway, with distinctive facial features, cardiac malformations, skin lesions, growth retardation and cognitive delay. Most of the patients reported in literature have survived beyond the early childhood years with no reports of death in infancy. Here we present a neonate with a severe phenotype of CFC syndrome who died at 2 months of age.

Baby boy born at 37 weeks gestation presented on day 1 of life with progressively worsening respiratory distress requiring mechanical ventilation. ECHO showed ASD, right ventricular hypertrophy, supraventricular pulmonary stenosis, bicuspid aortic valve and significant pulmonary hypertension. In the 1st week he developed hyperkeratotic scaly skin lesions over the trunk, extremities, non-pitting edema and bilateral chylothorax. At 4 weeks he developed multifocal atrial arrhythmias with poor response to medications. Whole exome sequencing was positive for a *de novo* mutation in the MAP2K2 gene, consistent with a diagnosis of CFC. His respiratory and cardiac status continued to worsen with respiratory failure, uncontrolled arrhythmias, hypotension, metabolic acidosis and acute renal failure. Due to progressive clinical deterioration and absence of definitive treatment for his condition, parents decided to discontinue life-sustaining measures.

Abstract 21 Figure 1

Abstract 21 Figure 2

22 EXTREMELY-PREATURE INFANT WITH ATYPICAL PHYSICAL CHARACTERISTICS: BECKWITH-WIEDEMANN SYNDROME

M Martinez*, F Bahadori-Esfahani MD. *LSU Shreveport, Bossier City, LA*

10.1136/jim-2018-000974.22

Introduction Beckwith-Wiedemann syndrome (BWS) is a rare pediatric disease of overgrowth and presents with an array of physical characteristics. Classic findings include macrosomia, macroglossia, hypoglycemia, hemihyperplasia, omphalocele and or other abdominal wall defects. We present a case of an extremely premature infant with in-utero diagnosis of an omphalocele, further physical and genetic testing confirmed the diagnosis of BWS.

Case Premature infant born at 25 weeks and 6 days via urgent c-section secondary to premature rupture of membranes and chorioamnionitis was admitted to the Neonatal Intensive Care Unit. Prenatal history significant for poor prenatal care and perinatal ultrasound consistent with protruding abdominal wall mass. At the time of delivery resuscitation was required secondary to prematurity, he was intubated and dosed with surfactant. Initial physical exam significant for large for gestational age at 96.6% for weight (1.230 kg) with normal head circumference at 81% (25.5 cm), and 4 cm omphalocele. As infant continued to grow he was noted to have macroglossia. A renal ultrasound obtained secondary to oliguria on day of life 34 noted bilateral prominent hydronephrosis. On day of life 48 he developed feeding intolerance and an Upper GI study was performed and significant for small bowel atresia. At this time due to omphalocele, macroglossia, small bowel atresia and bilateral hydronephrosis a genetic testing was obtained and confirm diagnosis of BWS. Infant underwent multiple surgical procedures for correction of omphalocele and bowel atresia. He was discharge on G-tube feeds and on room air.

Discussion Beckwith-Wiedemann syndrome is a genetic disorder presenting with multiple physical and metabolic findings. Many of the patients are diagnosed later in life when physical

abnormalities become prominent. Literature review has few documented cases on neonatal diagnosis of BWS with no documentation of extremely premature infant diagnosis. Prenatal ultrasound can help guide in-utero diagnosis when polyhydramnios, wall defects and other organ anomalies are visualized. Our patient, unfortunately born at 25 weeks gestation and poor prenatal care ultrasound findings were limited. The typical characteristics were initially masked in our patient due to his prematurity.

23 A RARE CASE OF ISOLATED BILATERAL HEMIMELIA

FI Sukhera*, B Drabu, EG Szyld. *The University of Oklahoma, Oklahoma City, OK*

10.1136/jim-2018-000974.23

Case report Bilateral lower extremity deformities are rare, however cases have been reported of longitudinal and transverse deficiencies. Majority of cases reported involve the absence or defect of either tibia or fibula, total absence of both tibia and fibula is extremely rare. The most common cause of such defects is amniotic bands, however sporadic cases have also been reported.

Here we present a case of a neonate who was born in a tertiary care hospital to a 25 year old G5P1031 with a history of anxiety, depression and class III obesity. On routine prenatal ultrasound, fetus was noted to have bilateral hemimelia with no other obvious congenital anomalies, amniocentesis was offered but parents declined. Patient was born at term via normal vaginal delivery, exam was notable for bilateral hemimelia. Plain radiography of lower extremities confirmed bilateral loss of tibia and fibula with essentially unremarkable femurs bilaterally. Genetics team was consulted who agreed that hemimelia was not syndromic or due to amniotic bands. During hospital stay infant remained stable on room air and was discharged from the hospital within 72 hours with plans to follow up outpatient with pediatric orthopedics, cardiology and genetics.

Bilateral hemimelia is rare condition and physicians should be aware that isolated cases may occur. Due to lack of substantial literature and guidelines for recommended postnatal screening requirements it can become challenging for pediatricians to prioritize which tests should be performed prior to discharge. It is thus crucial to have subspecialty involvement prenatally, as early psychology, genetics and orthopedics consults can to help identify a strong plan prior to discharge and thus ease parental anxiety regarding this rather difficult diagnosis. It is our goal to make physicians aware that not all cases are syndromic and if appropriate workup is initiated as soon as prenatal diagnosis is made then we can avoid prolonged hospital stays.

24 A NEONATE WITH WOLF-HIRSCHHORN SYNDROME AND PARTIAL PATAU SYNDROME

AR Wheeler*, J Philips. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.24

Case report This term male was born via C-section due to NRFHT. Prenatal echo revealed possible double outlet right ventricle or tetralogy of Fallot. Cytogenetics via amniocentesis confirmed unbalanced 4 p deletion and partial trisomy 13q.

After delivery, brief PEEP and blow-by oxygen was required and he was transferred to the NICU on room air. Physical exam showed micrognathia, hypertelorism, low set ears, broad nasal bridge, continuous systolic murmur, 2 vessel-cord, rocker bottom feet, and hypertonia bilaterally in the upper extremities. Echo revealed an interrupted aortic arch, large posterior VSD, hypoplastic left ventricular outflow tract, hypoplastic and bicuspid aortic valve, large PDA, large pulmonary valve with dysplastic and thickened leaflets, moderate pulmonary insufficiency, and large secundum ASD. He developed increasing respiratory distress and was placed on a ventilator and PGE1. He continued to deteriorate and his parents withdrew care on DOL 2.

Discussion Wolf-Hirschhorn is caused by a deletion in chromosome 4 and its severity is determined by the extent of the deletion. The characteristics include micrognathia, downturned mouth, poorly formed ears, short philtrum, broad and flat nasal bridge, prominent glabella, delayed growth, intellectual disabilities, feeding difficulty, seizures, cleft lip and/or palate, cardiac and skeletal abnormalities. Trisomy 13 is caused by translocation or *de novo* errors in division. Characteristics include micro/anophthalmia, cleft lip, cleft palate, polydactyly, holoprosencephaly, intellectual disability, simian crease, and cardiac defects including VSD, ASD, PDA, and DORV. Craniofacial, CNS, limb, and genital abnormalities can also be seen. Most children with these syndromes are not due to inheritance but rather a *de novo* occurrence or translocation. Management includes surgical correction, symptomatic, and supportive treatment. It is likely that in the setting of an infant with both trisomy 13 and Wolf-Hirschhorn that the associated features would be more severe.

Conclusion Early diagnosis, family discussion, and detecting the degree of abnormalities in these syndromes is necessary to facilitate prognosis and extent of elective interventions.

25 TETRALOGY OF FALLOT AND SITUS INVERSUS IN CLINICALLY WELL APPEARING NEONATE

¹R Kiani*, ²JY Choi, ³M Howell, ³S Yang. ¹Tulane Triple Board, New Orleans, LA; ²Tulane School of Medicine, New Orleans, LA; ³Tulane Pediatrics, New Orleans, LA

10.1136/jim-2018-000974.25

Case report Tetralogy of Fallot (ToF) is the most common type of congenital cyanotic heart disease, occurring in 1:3600 live births. The etiology appears to be multifactorial, however there is familial recurrence risk of 3%. Dextrocardia is a laterality defect with an incidence of 1:12 000 where the heart is on the right side of the chest, and situs inversus is a congenital condition where major visceral organs are opposite of their normal position (situs solitus). While, there is a 90% incidence of congenital heart defects in patients with dextroversion (dextrocardia with situs solitus), the association of situs inversus totalis with congenital heart defects is rare (less than 5%–10% of cases). Reports of the association of ToF with situs inversus totalis are rare.

Our case is a 40 6/7 week female newborn born via vaginal delivery to an 18 year old primigravid Hispanic mother. APGARs were 9 and 9 at 1 and 5 min respectively. Birth weight was 3339 g. The baby went to newborn nursery with an uneventful course until day of discharge, day of life 3, when a heart murmur was noted, with PMI greater on the right side.

Chest x-ray and electrocardiography confirmed dextrocardia, ToF and closed ductus arteriosus. She was transferred to the NICU and pediatric cardiology consulted. Abdominal ultrasound revealed situs inversus of liver, spleen, gallbladder, inferior vena cava and aorta while small bowel follow-through confirmed right-sided stomach emptying into left-sided duodenum – consistent with situs inversus totalis. Microarray showed duplication at Xq12 with OPHN1 gene located in the region noting a copy number variant from general population – significance of which is unclear. Chromosomal analysis revealed 46XX. She continued to remain clinically stable, and on day of life 7 was discharged home with outpatient cardiology and genetics follow-up.

Our case is unique given the rarity of the combination of ToF and situs inversus totalis. Unlike many cases in literature, our patient was diagnosed in the neonatal period and remained clinically asymptomatic. Thus, it is likely that her clinical course shall remain no different from any other patient with ToF.

26 AN INFANT WITH ZELLWEGER SYNDROME

A Ricci*, J Phillips. *University Of Alabama At Birmingham, Irondale, AL*

10.1136/jim-2018-000974.26

Case report A former 38 wk female with hypotonia and dysmorphic facial features was transferred with concern for Zellweger Syndrome (ZS). She was intubated secondary to central hypoventilation and required prolonged mechanical ventilation. She had frequent desaturations concerning for seizures and was diagnosed with multiple subclinical seizures on long term EEG. She was treated with Keppra, Vimpat, and phenobarbital. She developed hemochezia most likely secondary to liver disease related to ZS. Labs revealed transaminitis. PE showed dysmorphic facial features, microcephaly, single palmar crease, and hypotonia. Head ultrasound (US) revealed grade II IVH with mild ventriculomegaly and bilateral subependymal cysts. Renal US revealed bilateral hydronephrosis and renal cysts. Mother's previous child had died at 8 months from complications of ZS with confirmed pathogenic variants in the gene PEX6. A normal plasma acylcarnitine profile, increased very long chain fatty acids, low red blood cell plasmalogen content and pipecolic acid confirmed ZS. **Discussion** Zellweger spectrum disorders are a heterogeneous group of autosomal recessive disorders characterized by a defect in peroxisome formation and are caused by mutations in one of the 13 PEX genes. ZS patients accumulate very long chain fatty acids, phytanic and pristanic acid, C27 bile acid intermediates and pipecolic acid in plasma. They also have a deficiency of plasmalogens in erythrocytes. ZS occurs in about 1 in 50 000 births. Clinically, ZS can be highly heterogeneous with core features of liver dysfunction, developmental delay with other neurological abnormalities, adrenocortical dysfunction, and hearing and vision impairment. Patients usually develop life-threatening complications during the first year of life. Diagnosis is made by detection of very long chain fatty acids. Additional tests on blood and urine samples to detect other substances associated with peroxisome metabolism may be performed. Genetic testing is also available. Treatment includes cholic acid to aid with bile acid synthesis dysfunction.

Conclusion Early recognition and diagnosis of this syndrome could help with early intervention to improve developmental outcomes of infants with Zellweger Syndrome and to establish a true frequency in the general population.

27 CONGENITAL HEPATOBLASTOMA: A CASE REPORT

C Cojocar*, E Patel, A Uber, M Eklund, W Southgate. *Medical University of South Carolina, Charleston, SC*

10.1136/jim-2018-000974.27

Case report A female infant of dichorionic diamniotic twin gestation was born at 37 weeks by cesarean section. Pregnancy and delivery were uncomplicated. Presentation at birth was significant for abdominal distention and respiratory distress. A large abdominal mass was palpated in the right upper quadrant and extended past midline. Initial labs demonstrated thrombocytopenia, anemia, hyperbilirubinemia, transaminitis and elevated alpha fetoprotein. An abdominal ultrasound showed a 10×8×7 cm heterogeneous mass arising from the right hepatic lobe with some vascularity. Based on those findings, she was presumed to have infantile hepatic hemangioendothelioma. An abdominal MRI was obtained to attempt to provide a definitive diagnosis but results were inconclusive and a liver biopsy was recommended. Pathology confirmed a diagnosis of mixed embryonal/fetal hepatoblastoma (HBL), stage III. Further imaging did not demonstrate metastasis.

Chemotherapy was initiated with Cisplatin, 5-Fluorouracil (FU), Vincristine, and Doxorubicin. The tumor initially responded, decreasing in size to 9×6 cm. Complications of treatment included neutropenic colitis, renal failure requiring dialysis and hyperbilirubinemia which required discontinuation of initial chemotherapy. A salvage regimen consisting of Oxaliplatin and 5-FU further reduced her tumor burden. However, she remained critically ill with persistent respiratory failure, worsening acidosis, increasing vasopressor support, and dialysis dependence. Parents made the decision to withdraw care at 40 days of life.

HBL is the most common malignant liver tumor of childhood but rarely presents in the neonatal period. Congenital HBL has a favorable outcome with a 3 year overall survival of 86%. We draw attention to the rarity of this condition and the difficulty in definitive diagnosis in the neonatal period. We also highlight the therapeutic challenges that were encountered due to the significant comorbidities leading to limited chemotherapy options. Despite the reported favorable outcome and evidence of chemo sensitive tumor, our patient had a poor outcome due to adverse effects from treatment.

28 A CASE OF PARTIAL TRISOMY OF CHROMOSOME 1 WITH PERSISTENT HYPOGLYCEMIA AND RESPIRATORY DISTRESS

¹S Gijnjupalli*, ²A Simonin, ³E Bassig, ²A Safarulla. *¹Medical College of Georgia at Augusta University, Augusta, GA; ²Augusta University, Augusta, GA*

10.1136/jim-2018-000974.28

Case report We present a case of partial trisomy 1 who was transferred to our NICU for persistent hypoglycemia and respiratory distress.

Abstract 28 Figure 1 Physical features: A. Flat nasal bridge, B. Overlapping toes and C. Low set ears. Parent/Guardian consent obtained.

33 week delivery for non-reassuring fetal heart tones. Unremarkable pregnancy.

Metabolic labs and genetic work up sent for hypotonia. Metabolic work up was unremarkable and tone normalized. Cytogenetic analysis revealed a duplication of a segment of 1q21 to 1q32 resulting a partial trisomy of this region. Hypoglycemia noted at a month of age with blood sugar in 20–30 mg/dl. Cortisol, insulin and growth hormone levels were normal. Started on diazoxide per recommendation from pediatric endocrinology. This was continued for a period of 10 days and gradually weaned off given stable sugars. Dysmorphic features include flat nasal bridge, high arched palate, low set ears, retrognathia, overlapping toes bilaterally. Hypothyroidism and a large ostium secundum atrial septal defect noted. Given the degree of retrognathia and glossoptosis and inability to be weaned from positive pressure ventilation, patient received a tracheostomy. Also received a Gtube due to poor oromotor strength and to protect airway.

Partial trisomy 1 with persistent hypoglycemia requiring diazoxide therapy and significant respiratory distress necessitating tracheostomy has not been described prior.

29 PROPIONIC ACIDEMIA IN A NEONATE

K Miller*, J Philips. *University of Alabama, Birmingham, AL*

10.1136/jim-2018-000974.29

Case report A 3 day-old female was transferred for further workup and management of hyperammonemia and metabolic acidosis. On arrival, physical exam revealed tachypnea with intercostal retractions and nasal flaring, and lethargy with responsiveness to stimuli. She had normal cardiac and abdominal exams, and her head exam revealed a soft and flat anterior fontanelle. Initial lab work at presentation revealed a pH of 7.2, a bicarbonate of 3 umol/L, and an ammonia level of 431 umol/L. The patient's acidosis was corrected with sodium bicarbonate, and she received maintenance IV fluids and antibiotics while sepsis work-up, genetics testing, and metabolic testing were pending. Her tests revealed elevated levels of propionylcarnitine among various amino acids, and she was found to have a homozygous pathogenic variant of *PCCB* confirming a diagnosis of propionic acidemia.

Discussion Propionic acidemia (PA) is an autosomal recessive disease that results in an inability to catabolize certain amino acids, fatty acids, and cholesterol among other substrates. PA is caused by deficiency of the enzyme propionyl-CoA carboxylase due to a genetic mutation, most commonly of *PCCA* or *PCCB*, and the disease affects approximately 1/100,000 people in the US. Patients generally present within the first few days of life with metabolic decompensation, but rarely patients present in childhood with waxing and waning symptoms. In the neonatal presentation, these patients generally feed and act normally until a few days of life, when the infant starts having poor feeding, lethargy, and hypotonia. When ill, patients are at risk of metabolic decompensation and death. Diagnosis is suspected by urine and plasma organic acid measurement, plasma acylcarnitine analysis, or newborn screening. A definitive diagnosis can be established by confirmation of *PCCA* or *PCCB* mutations or by demonstration of deficient activity of propionyl-CoA carboxylase. Management includes having a protein-restricted diet and prescribing biotin and carnitine acid medications. Patients need regular cardiac exams due to concern for QTc prolongation and arrhythmias, and depending on difficulty of feeding, gastrostomy tube may be indicated. The prognosis can range

from normal cognitive development to significant neurodevelopmental deficits to death.

Pediatric Clinical Case Symposium

12:00 PM

Thursday, February 21, 2019

30 ULTRA LATE GROUP B STREPTOCOCCUS MENINGOENCEPHALITIS & BACTEREMIA

AS Marshall, K Gutermuth*, M Kong. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.30

Introduction Group B streptococcus (GBS) is a common cause of bacterial infection in neonates and infants requiring hospitalization and antibiotic therapy. While early-onset (<6 DOL) and late onset (6–89 DOL) infection are fairly common, infections in children older than 3 mo are much rarer. Ultra-late disease (infection presenting >90 DOL) is mostly seen in premature infants (most often <28 wga) than their peers with early or late disease.

Case A 5moF presented to the ED with lethargy, poor oral intake, and fever for 2 days. She was an ex-37 wga twin born via C-section for breech presentation and no ICU stay. Maternal serologies were significant for Group B Streptococcus treated appropriately. Medical history included non-operative hip dysplasia, and reflux managed with Ranitidine. Upon arrival, she was found to be tachycardic and hypotensive despite fluid resuscitation. She had poor perfusion and ultimately was intubated due to fluid refractory septic shock and admitted to the Pediatric Intensive Care Unit. Antibiotics were empirically started and her blood cultures grew gram-positive cocci in clusters 7 hours after collection, as did her cerebral spinal fluid culture. Final culture data from admission revealed *Streptococcus agalactiae* Serogroup B meningococcal infection with bacteremia sensitive to Penicillins. Her course was complicated by disseminated intravascular coagulation, and generalized tonic-clonic seizures related to her meningococcal infection without abscess or empyema. Ultimately, she improved, and she was discharged on anti-epileptics after completing a 3 week course of Ampicillin. Complement and further work-up revealed her to be immunonormal.

Discussion GBS is a well-known cause of meningitis in children younger than 3 months old. Ultra-late GBS disease most often occurs in preterm infants. Ultra-late infection was more commonly seen in children with an abnormal immune system, most commonly a complement deficiency, and in a few cases was the antecedent infection leading to a diagnosis of HIV. This case represents a rare clinical presentation of ultra-late GBS in an immunonormal child and highlights the need to consider coverage for this pathogen even in otherwise healthy older infants who present with septic shock.

31 AN ATYPICAL PRESENTATION OF INTUSSUSCEPTION

C Lares*, B Dillard, MN Frascogna. *University of Mississippi Medical Center, Jackson, MS*

10.1136/jim-2018-000974.31

Case report Psychiatric complaints are increasingly common presentations to the Pediatric Emergency Department (PED),

particularly in adolescents. We present a case that highlight the importance of ruling out medical causes of psychiatric symptoms.

A 12 year old male presented to the PED with severe agitation after a sudden onset of abdominal pain. He was asymptomatic prior to onset of symptoms. On arrival the patient was extremely combative, uncooperative and was unable to communicate or describe his symptoms to the point that he required physical and chemical restraint in order to be assessed. Physical exam was significant for tachypnea, elevated blood pressure, dilated reactive pupils and profuse sweating. Abdominal examination showed a non-distended abdomen with bowel sounds and no appreciable tenderness or mass. His neurological exam was significant for altered mental status with significant agitation. After sedation with olanzapine he remained drowsy with no complaints of abdominal pain with serial abdominal exams. Laboratory workup revealed leukocytosis (WBC 16000) and mild elevation of anion GAP of 17. His glucose, electrolytes, liver function tests and lactate were all normal and he had a negative toxicology screen. Abdominal xray showed moderate amount of stools and non-obstructive pattern. Received an enema which resulted in passage of a normal stool. Given his persistent altered mental status and initial complaint of abdominal pain, a CT Abdomen/Pelvis with IV contrast was ultimately obtained revealing a small bowel-small bowel intussusception. Pediatric Surgery emergently performed an exploratory laparotomy finding an incarcerated intussusception of ileum requiring resection with primary anastomosis.

Sudden onset of psychiatric symptoms, including severe agitation, presenting with abnormal vital signs should raise concern for an underlying emergent medical cause and prompt further workup.

32 NON-TYPE B HAEMOPHILUS INFLUENZAE: CASES FOR EXPANDING HAEMOPHILUS INFLUENZAE VACCINATION

T Miller, J Putman*, K Mather, K Martin. University of Oklahoma School of Community Medicine – Tulsa, Tulsa, OK

10.1136/jim-2018-000974.32

Case report After the introduction of *Haemophilus influenzae* serotype b (Hib) vaccination in the mid-1980's, there was a significant decrease in the rate of infections caused by Hib. Because of this decrease, multiple other serotypes of *H. influenzae* have become an important cause of invasive disease in the pediatric population. In the last 10 years, there have been 21 reported cases of *H. influenzae* meningitis in Oklahoma. Based on 2017 National data from the Centers for Disease Control and Prevention (CDC), there were 25 reported cases of invasive *H. influenzae* type B compared to 151 cases of invasive non-typeb disease. We present 2 cases of non-typeB *H. influenzae* meningitis in Tulsa, OK.

The first case is a previously healthy 11 month old male who presented to an outside hospital with complaint of one day of fever, vomiting, irritability, and cough. Microbiologic testing identified *H. influenzae* serotype f in blood and cerebral spinal fluid (CSF) cultures. The patient was treated with a four week course of ceftriaxone, but his course was complicated by a cranial empyema requiring a craniotomy. He continued with inpatient rehabilitation after discharge.

The second patient was a previously healthy 14 month old male that presented with a one day history of altered mental

status, fever and vomiting. Microbiologic testing identified *H. influenzae* serotype a in the CSF culture. This patient was treated with a four week course of ceftriaxone and recovered without any further obvious sequelae.

Prior to the introduction of Hib vaccination in the United States, *H. influenzae* was the most common cause of invasive bacterial infection in children causing significant morbidity and mortality. Since the initiation of the immunization series, there has been a drastic decrease in the rates of invasive Hib. Unfortunately, other serotypes of *H. influenzae* are now noted to cause invasive disease in a similar but delayed fashion. The CDC recommends that all cases of invasive disease caused by *H. influenzae* isolate be serotyped and reported to the appropriate state agency to evaluate the changes and trends in serotype distribution. Ultimately, the goal for continued surveillance is to evaluate the need for development of vaccinations against other non-type B *H. influenzae* serotypes.

33 ALL THAT SPINS IS NOT VERTIGO

P Redmond, N Freeman*, B Dillard, MN Frascogna. University of Mississippi Medical Center, Jackson, MS

10.1136/jim-2018-000974.33

Case report Chest pain and dizziness are common complaints seen in the pediatric emergency department (PED) with many causes being benign in nature. Good medical history taking can help tease out more serious causes of these complaints that may warrant further workup. The following case demonstrates how a thorough history obtained in the emergency department unearthed a potentially detrimental anomaly had it been left undiagnosed.

An 11 year old male presented to the PED with complaints of dizziness for five days that worsened with lying flat and exertion. He was worked up for infectious causes at a local clinic and ultimately diagnosed with vertigo and a heart murmur. He was sent home with a prescription for meclizine and referred for an outpatient pediatric cardiology evaluation. His mother made the decision to bring him to the PED due to continued bouts of dizziness. The patient's complaints at home had all been related to recurrent episodes of dizziness; however, after thorough questioning in the emergency department, he disclosed to the physician that he had also been having chest pain on exertion. There was no known family history of sudden cardiac death or congenital heart disease. The physical exam was only notable for a II/VI systolic murmur. An electrocardiogram was obtained and was notable for right ventricular hypertrophy. An echocardiogram obtained had findings suspicious for an anomalous left coronary artery arising from the right aortic cusp. He was admitted for further evaluation of the suspected vascular anomaly. While admitted, a CT angiogram showed a vessel arising from the right coronary cusp taking an interarterial course. An exercise stress test was negative for ischemia, but he did have chest pain after four minutes of exertion. He underwent cardiac catheterization, which confirmed that the left coronary artery arose from the right sinus and coursed between the two great vessels. He underwent operative repair and recovered fully without any complication.

Anomalous origin of the coronary artery is a rare defect that carries a significant risk of sudden cardiac death. Due to the detailed history obtained in the emergency department,

the previously unreported complaint of chest pain was revealed. This additional history prompted further work up and eventually the diagnosis.

34 FROM CONSTIPATION TO HYDRONEPHROSIS, AN INCIDENTAL FINDING OF UNDIAGNOSED POSTERIOR URETHRAL VALVE IN A TODDLER

S Gonzalez*, E Klepper, S Vick. *Our Lady of the Lake Children's Hospital, Baton Rouge, LA*

10.1136/jim-2018-000974.34

Case report Posterior urethral valves are a congenital lower urinary tract outflow obstruction affecting males with incidence of 1 in 8000. Prenatal ultrasounds are important in order to identify neonates with hydronephrosis. There are rare cases in which the diagnosis is not made until later in life. Those patients present with urinary dysfunction, including overflow incontinence or voiding difficulties. We present a case of a 2 year old male who presented for constipation, found to have a severe acute kidney injury secondary to undiagnosed posterior urethral valves.

A 2 year old male with a past medical history of constipation presented to the emergency department (ED) for one day of abdominal distention, pain and constipation. In the ED, he was noted to have elevated blood pressure in the 99th percentile for age. An abdominal X ray revealed significant stool burden. A complete metabolic panel was remarkable for a severe acute kidney injury with BUN of 58 and creatinine of 3.11. A renal ultrasound noted severe bilateral hydronephrosis. Nephrology recommended amlodipine for the hypertension. Urology was consulted to assist with evaluation for an obstructive process. A voiding cysto-urethrogram revealed grade 5 vesicoureteral reflux on the left and grade 1 on the right. He was taken to the operating room and found to have type 3 urethral valves which were resected. Repeat labs demonstrated improvement of his acute kidney injury with creatinine of 0.84 post operatively.

This case demonstrates a late presentation of posterior urethral valves masked as abdominal distention thought to be secondary to constipation. Specific symptoms that can lead to the presumptive diagnosis include dribbling and poor urine stream, urinary tract infections and less frequently, hypertension. Ultimately, it is important to identify and treat these patients as quickly as possible, since studies have documented detrimental effects on renal function when presenting after 1 year of age with a higher creatinine level. Upon further questioning, following admission, it was discovered that the patient was indeed having dribbling and poor urinary stream. This highlights the importance of obtaining a detailed history and review of symptoms in light of the potential for anchoring bias.

35 FLU RELATED NECROTIZING ENCEPHALITIS

M Adams*, W Sasser, N Rockwell. *UAB, Birmingham, AL*

10.1136/jim-2018-000974.35

Case report A 9 year old previously well male presented as transfer to our ED with 4 days of fevers, headache, abdominal pain, and myalgia. Four days prior he was seen at a local urgent care and diagnosed with Influenza B and provided

Tamiflu of which he only tolerated two doses in the subsequent days. On the morning of presentation he was found by mother with right eye deviation, confusion and difficulty walking. He was brought to outside ED where initial work up was mostly reassuring. CBC, CMP, blood gas, CRP/ESR, ammonia, and UA all within normal limits with exception of mild thrombocytopenia. LP revealed CSF with WBC 5, RBC 28, Protein 384, Glu 54. CT head with initial report of no acute intracranial process.

In our ED, he was found to be responsive to painful stimuli, had persistent right eye deviation, and hypertonicity (without clonus). He developed generalized tonic-clonic activity for which he was given Ativan and later a Keppra load. He remained with GCS of 4 with decerebrate posturing to painful stimuli and was intubated and transferred to PICU. Further review of initial head CT revealed diffuse cerebral edema with areas of hypodensity. Head MRI was performed which revealed diffuse abnormality of thalamus, bilateral hippocampi, brainstem, and left upper cervical cord with associated punctate hemorrhages concerning for a viral related necrotizing encephalitis.

He was kept on a neuroprotective algorithm and he was started on IVIG, steroids (Dexamethasone), therapeutic hypothermia, and high dose Tamiflu. Broad-spectrum antibiotics were given until blood, CSF, and HSV studies were negative. All infectious etiologies remained negative other than Influenza B. He had slow but steady improvement with extubation on day 13 of hospitalization and discharge on day 38 with noted mild right spastic hemiparesis but otherwise able to ambulate with walker and perform all ADLs without assistance.

Acute Necrotizing Encephalitis (ANE) is a rare and often fatal condition that is typically viral related. While there are many different viruses associated with ANE, the most common are Influenza (A and B), Parainfluenza, Varicella, HSV, and Enterovirus. It differs from other encephalitis forms in that it usually has an absent pleocytosis, normal ammonia, and often elevated CSF protein. This case demonstrates a rare but serious complication to a common viral illness, influenza.

36 STERNAL PSEUDOTUMOR OF CHILDHOOD

¹JS Lim*, ¹JS Chawla, ¹G Hescok, ²J Congeni. ¹Louisiana State University, New Orleans, LA; ²LCMC, New Orleans, LA

10.1136/jim-2018-000974.36

Case report Sternal pseudotumor of childhood is a rare, self-limited inflammatory process affecting young children, and case reports describing this topic are rare. It is neither neoplastic nor infectious in nature and often benefits from a 'wait and see' approach, sparing invasive and often unnecessary malignancy work-ups. More documentation of such cases in the literature would provide more knowledge of this benign disease process and potentially reduce costly and invasive work-ups.

Our patient is a healthy 13-month-old male who presented with two days of an acutely developing firm, immobile anterior chest wall mass just lateral to the sternum. He was otherwise well appearing and without other symptoms. Ultrasound and CT of the chest showed an ill-defined soft tissue mass. MRI of the chest revealed a dumbbell-shaped soft tissue mass in the pre and post-sternal area consistent with sternal pseudotumor of childhood. Biopsy of mass revealed inflammatory

changes without evidence of neoplastic or infectious processes. The patient was discharged with close outpatient follow-up.

This case demonstrates the classic presentation of sternal pseudotumor of childhood supported by the dumbbell-shaped pre and post-sternal soft tissue swelling seen on MRI and lack of neoplastic or infectious pathology on biopsy. In the absence of other abnormalities on history or physical exam, a rapidly growing sternal mass in a young, otherwise healthy, child should raise suspicion for such pathology. The current reports on this benign mass in young children consistently document the benefit of a 'wait and see' approach as it typically resolves without intervention. However, the incidence of sternal pseudotumor of childhood is unknown due to the limited number of case reports available in the literature. Additional case reports are needed to better define and increase awareness among physicians of this disease process, as well as encourage a 'wait and see' approach in these specific patients, potentially reducing unnecessary, costly and invasive work-ups in this patient population.

37 IT'S JUST CHRONIC SINUSITIS

C Pribble, P Noor*, A Tanios. *Saint Louis University, St. Louis, MO*

10.1136/jim-2018-000974.37

Case report A previously healthy 9 year old Caucasian female presents with a 7 week history of bilateral nasal congestion, frontal headache and purulent rhinorrhea. Her symptoms started after returning from Tennessee, where she swam frequently in a hotel pool. Despite multiple completed antibiotic regimens, symptoms progressively worsened. She presented to our hospital with new onset diplopia for the past week, along with development of partial ageusia and facial pain. On admission, physical exam was unremarkable except for right maxillary tenderness with overlying erythema and swelling, right nasal congestion, decreased hearing in the right ear and inability to abduct the right eye. Facial CT revealed maxillary sinus congestion, possible neoplastic or infectious mass, and temporal bone involvement. Ophthalmology diagnosed an isolated cranial nerve VI palsy. ENT recommended face and brain MRI, which showed solid enhancing tumor involving the entire nasopharynx extending anteriorly into the posterior aspect of nasal cavity and inferiorly into oropharynx. Patient underwent endoscopic nasal biopsy, which showed rhabdomyosarcoma.

Acute rhinosinusitis accounts for approximately 1.6 million office visits per year, or 0.6% of visits for patients 0–20 years. Typical symptoms include facial pain, rhinorrhea, congestion and headache. Refractory bacterial sinusitis often requires additional beta-lactamase inhibitors or MRSA-coverage, which can prolong treatment. Rhabdomyosarcoma accounts for approximately 4% of pediatric cancers, and is frequently found in the head and neck. Presenting symptoms of head and neck rhabdomyosarcoma can mimic sinusitis, and include facial swelling, neuropathy and hearing loss, as well as increasing risk of infection. Of note, the sixth cranial nerve is particularly vulnerable.

This case shows how similarly chronic sinusitis and nasopharyngeal tumors may present, and the importance of evaluating for neurologic changes. Symptoms of sinusitis, such as headache or congestion, may mask the signs of a underlying mass. To further complicate the differentiation, tumors can cause obstruction and increase the risk of infection. The

development of neurological changes should raise concern for underlying malignancy. CT and MRI were helpful in differentiating neoplastic mass from infectious mass, with biopsy providing final diagnosis.

38 THE SPICE OF DEATH: A CASE OF CARDIAC ARREST AFTER SYNTHETIC CANNABINOID USE

¹RW Steele*, ^{1,2}J Moran, ²A Patton, ^{2,3}C Kokes, ¹L James, ¹E Storm, ¹S Schexnayder. ¹University of Arkansas for Medical Sciences, Little Rock, AR; ²PinPoint Testing, LLC, Little Rock, AR; ³Arkansas State Crime Laboratory, Little Rock, AR

10.1136/jim-2018-000974.38

Case report A 16-year-old previously healthy male was found unresponsive in a bathroom after family heard him fall. He was known to be smoking K2 at the time. He was estimated to be down for approximately 40 min prior to EMS arrival, who found him apneic and pulseless, and began CPR. Cardiac monitor showed asystole. He received multiple doses of epinephrine while enroute to the referring hospital. After arrival there, he was intubated and given additional epinephrine with return of spontaneous circulation (ROSC). In the ED, he experienced two more episodes of cardiac arrest and received additional epinephrine, amiodarone, and atropine prior to sustained ROSC. A dopamine infusion was begun for hypotension. Arterial blood gas was also obtained and showed severe respiratory acidosis with pH 6.56, pCO₂ 153 mm Hg, and pO₂ 61 mm Hg. ECG showed sinus tachycardia, short PR interval, and premature atrial contractions. Troponin I was 0.04 ng/ml. He was transferred to the PICU at the referral center, where he was noted to have a GCS of 3, respiratory failure, metabolic and respiratory acidosis, acute renal failure, acute hepatic injury, and cardiogenic shock. At the time of his admission, his physical exam showed no cranial nerve reflexes, and fixed and dilated pupils. He was removed from life support at family request and died. Initial urine toxicology screen was negative, but comprehensive toxicology testing detected the presence of a K2 metabolite of 5F-ADB (78.9 ng/ml). The parent drug 5F-ADB was not detected, indicating a relatively short half-life.

39 HOPSCOTCHING NEONATE: IS IT TOO EARLY TO PLAY THIS GAME?

W Chen*, MS Evangelista, GH Degaffe. *MGovern Medical School at UTHealth, Houston, TX*

10.1136/jim-2018-000974.39

Case report A 5 day old term infant was admitted to our hospital due to persistent posturing of the right hip. Family noticed asymmetric movement of the lower extremities soon after birth. Radiograph of the affected region showed shallow and dysplastic right acetabulum with subluxed femoral head concerning for hip dysplasia. Infant was instructed to see the orthopedic surgeon after discharge from the nursery. Due to persistent flexion and abduction of the right hip on follow up visit, she was admitted for a full evaluation. The history on admission was consistent with an infant who was otherwise doing well without fever, swelling or redness at the affected joint. Her exam revealed a flexion and abduction of the right hip joint at rest. Manipulation of the right lower extremity caused the infant to cry and attempt to withdraw the limb.

Laboratory values obtained on admission were normal except for a mild elevation of C reactive protein. MRI of the hip showed a fluid collection. Patient subsequently underwent right hip irrigation and debridement. Empiric antibiotics were initiated. Culture of synovial fluid grew *Staphylococcus hominis*. She continued to improve on a 3 week course of intravenous antibiotics, followed by a short oral antibiotic regimen. Follow up imaging failed to show prior noted hip dysplasia.

Discussion Septic arthritis in healthy newborns is rare. We present a case of culture proven right hip septic arthritis in a term neonate with persistent flexion and abduction of the right hip. The lack of systemic symptoms and initial imaging findings of right hip dysplasia in our patient made the differential diagnosis challenging. Pseudoparalysis on exam and abnormal inflammatory markers, notably, C-reactive protein, are strong indicators that warrant further work up for septic arthritis. Advanced imaging is often required for neonates with septic arthritis to rule out osteomyelitis. Culture negative infection is not uncommon in this population. Treatment options are similar for neonates compared to their older counterparts with septic arthritis. Delayed diagnosis and treatment is associated with significant sequelae to the affected joint. Pediatricians should keep septic arthritis on the differential when neonates present with asymmetric range of motion of the extremities on newborn exams.

Case Reports in Cardiovascular Medicine

2:00 PM

Thursday, February 21, 2019

40 LAMBL'S EXCRESCENCES; AN ENIGMA OF DIAGNOSTIC CARDIOLOGY

H Amin, M Jilani*, P Pitroda, D Villarreal. *SUNY Upstate Medical University, Syracuse, NY*

10.1136/jim-2018-000974.40

Introduction Cardioembolism accounts for roughly one fifth of all ischemic strokes. A rare etiology encountered in patients with cardioembolic stroke are Lambl's excrescences (LEs) which are filiform extensions that form along the lines of valve closure due to age-related degenerative process. LEs are primarily diagnosed through TEE where they are visualized as mobile filamentous echo densities on aortic, mitral and prosthetic valves. These valvular strands can potentially result in thromboembolic complications.

Case report A 48 y.o. male presented with a complaint of vertigo and vomiting. His medical history was significant for similar episodes of dizziness. His physical exam was negative for focal neurologic findings other than bilateral horizontal nystagmus. Brain MRI showed multiple hypo densities in the left and right cerebellum, left thalamic and left medial occipital region, indicative of an embolic process. Due to symptom onset >4.5 hours, thrombolysis was not attempted and he was started on aspirin and statin. TSH and hypercoagulability work up was unremarkable. CT angiogram head and neck was negative for arterial occlusion, stenosis, or aneurysm. EKG was negative for arrhythmias. TEE with bubble study was then performed which showed a single echo density on the ventricular side of aortic valve consistent with LE. A literature

search for guidelines on LEs management was done and after considering all evidence, we decided to treat the patient with anticoagulation until a definitive cause of stroke was determined.

Discussion Literature regarding management of LEs in patients with and without stroke is currently limited and standard guidelines regarding the management of LEs have not been established. A number of authors resorted to surgical removal of the excrescences at the first episode of stroke. Others opted for anticoagulation or antiplatelet therapy with the initial occurrence of stroke and reserved surgical management only for a recurrent stroke. Both proclaimed treatment strategies were associated with favorable outcomes at 3 months to 1 year follow up.

Conclusion We conclude that LEs should be considered in the differential of cardio embolic stroke after excluding other etiologies. Anticoagulation should be initiated until another definitive cause of stroke is found and/or treated.

41 DANGEROUS BLIND LOVE: LEFT EYE CENTRAL RETINAL ARTERY OCCLUSION BY KOCURIA ROSEA ENDOCARDITIS IN AN IMMUNOCOMPETENT MALE

NL Rabelo-Pagan*, K Soto-Moreno, Y Rodriguez-Rodriguez, G Galindez-De Jesus, P Vargas. *University of Puerto Rico Medical Sciences Campus, Las Piedras*

10.1136/jim-2018-000974.41

Case report *Kocuria rosea* bacteria is a commensal organism which is found on the skin. Bacterial infections with *Kocuria* species are present in clinical settings of malignancies, and immunosuppression. Reported cases of *Kocuria* infections in immunocompetent individuals as bacteremia and endocarditis. A 53yo male patient with no medical history. He presented with acute complete left eye vision loss. Ophthalmology evaluation diagnosed OS central retinal artery occlusion. Head CT revealed an early subacute intraparenchymal hemorrhage within the left occipital lobe, with smaller remote infarcts. Findings very suggestive of thromboembolic cause. Blood cultures reported the presence of *Kocuria rosea*, a rare Gram-positive cocci, coagulase negative, actinobacteria, atypical organism finding in endocarditis. Patient was started on IV Vancomycin and Gentamicin therapy. Transthoracic echocardiogram revealed severe mitral valve regurgitation, as well as a large mobile vegetation attached to the posterior mitral valve leaflet. Further visualization by transesophageal echocardiogram visualized vegetation was no longer present. Three days after transesophageal echocardiogram the patient presented with acute right sided hemiparesis and neurological decline. Head CT demonstrated an acute left ischemic infarct of moderate size. Patient was transferred to CCU for stabilization. Once stable, patient had a mechanical mitral valve replacement without complications. Was later discharged home after IV ABX completion and, achievement of anticoagulation levels for PT/OT. This case demonstrates one of the most feared complications of infective endocarditis; septic cardioembolization, even with adequate treatment. Drawing importance to significance of isolation and clinical presence of *Kocuria* species in humans as infectious agent. Due to its high similarity, shared physical properties to other Gram-positive cocci it can be misdiagnosed, categorized as a contaminant. *Kocuria* species should be considered a source of infection, especially in inpatients.

42 ENDOPTHALMITIS AS PRESENTING FEATURE IN A PATIENT WITH UNDERLYING INFECTIVE ENDOCARDITIS

GD Bedanie*, N Adhikari, M Rahman, M Zitun. *Texas Tech University Health Science Center, Lubbock, TX*

10.1136/jim-2018-000974.42

Introduction Endophthalmitis is a rare bacterial or fungal ocular infection involving the vitreous and/or aqueous humors. It is a vision-threatening condition which requires emergency management. Most cases are due to external inoculation of organisms related with trauma, corneal injury or eye surgery. The commonest endogenous risk factor is the presence of infective endocarditis. We are presenting a case of patient presented with acute onset of sharp right eye pain, decreased vision in the setting of mitral valve bacterial infective endocarditis.

Case summary A 51-year-old male with type 2 diabetes presented to emergency center with acute onset of sharp pain, photophobia and decreased vision in the right eye for 2 days duration. Patient denies history of eye trauma or recent surgery. Examination by ophthalmologist revealed redness of the right eye with decreased movement and absent reaction to light. Pan uveitis was considered and he was sent home with oral and topical steroids. 5 days later, patient was back due to worsening of eye symptoms. Vitreous samples were taken by ophthalmology and patient was given intra-vitreous as well as systemic antibiotics. MRI of head showed right endophthalmitis. A culture of specimen from the vitreous and anterior chamber, as well as blood culture grew methicillin sensitive staphylococcus aureus. Transthoracic echocardiogram revealed small mobile echogenic structure on mitral valve. Acute bacterial infective endocarditis and endogenous endophthalmitis were diagnosed. Patient was treated with IV nafcillin for 6 weeks and IV steroid for endogenous endophthalmitis. Since patient didn't improve with medical management, he underwent right eye enucleation due to excessive pain and risk of sympathetic ophthalmia.

Conclusion Endogenous endophthalmitis is rare but devastating disease that is commonly misdiagnosed. The majority of patients have an underlying predisposing infection, most commonly infective endocarditis. Urgent treatment is vital to preserve vision; however, in most cases, the visual prognosis is poor leading to blindness in the affected eye.

43 PREMATURE CORONARY ARTERY DISEASE

CH Harris*, K Ellard, LS Engel, J Martinez. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.43

Introduction Premature Coronary Artery Disease is defined clinically as coronary artery disease (CAD) before age 45 in males and age 55 in females. Although, similar risk factors and management exist for premature CAD compared to CAD diagnosed later in life, premature CAD patients report angina less frequently. Patients with premature CAD also have a higher prevalence of normal coronaries or single vessel disease on angiography. Although CAD usually presents after the fifth decade of life, we present a case of premature CAD in a 26 year old man diagnosed with 3-Vessel Coronary Artery Disease.

Case A 26 year old man with no significant past medical history presented to the Emergency Department after onset of intermittent, substernal chest pain lasting minutes while at

rest. He had similar pain and palpitations for the preceding three months associated with physical exertion at work and relieved with rest. The patient was not obese, denied any significant family history of coronary artery disease, and never smoked. His blood pressure was 210/115 mmHg at presentation. Physical exam and bedside echocardiogram were unremarkable. His initial blood work demonstrated a markedly elevated cholesterol of 332 mg/dL and LDL of 276 mg/dL. EKG demonstrated normal sinus rhythm, lacking ST segment or T wave abnormalities to suggest ischemia. Initial troponin was elevated at 0.45 ng/mL and eventually peaked at 3.12 ng/mL. A diagnosis of NSTEMI was made. Coronary angiography revealed 3 vessel CAD requiring subsequent coronary artery bypass graft surgery.

Discussion This is a rare case of severe 3-vessel premature CAD. Although premature coronary artery disease is uncommon, this case highlights that manifestation of coronary disease in premature CAD often presents as rapid progression to myocardial infarction. Recognizing risk factors (family history, dyslipidemia, metabolic abnormalities, and blood pressure) for premature CAD in young patients as well as need for interventions is important for improving prognosis and modifying effects of this disease for overall patient outcome.

44 AN UNUSUAL CASE OF YOUNG MALE WITH SILENT WIDOW MAKER

¹T Bhandari*, ¹S Bhogal, ¹K Mawa, ²H Saini, ¹P Sankhyani, ¹A Mahajan, ¹T Paul. *¹East Tennessee State University, Johnson City, TN; ²Gian Sagar Medical College, Rajpura, India*

10.1136/jim-2018-000974.44

Case report A 34 yo M with past medical history of hypertension presented to the ER with chest pain which was atypical in nature since 1 week. Pain was described as left sided, episodic, intermittent, exertional lasting 5 min with associated shortness of breath. Patient had presented to ER 1 week ago at the time of 1st onset and was noted to have normal EKG and normal troponins after which he was discharged, subsequently he presented 1 week later due to progressive worsening and recurring episodes. Vitals in ER: BP: 146/101 mm Hg, pulse 60/min, RR: 18/min, Temp: 98.3 F. His physical examination was benign except that he was noted to have sinus bradycardia. EKG showed no significant ST-T wave changes. 1st Set of troponins was 0.12. Triple CTA was ordered emergently considering his pain that showed findings concerning for acute coronary syndrome with 100% stenosis. ECHO showed normal EF: 55%-60%. Coronary Angiography showed proximal to mid LAD total occlusion, with collaterals from RCA to distal LAD, after which PCI to proximal mid LAD with Drug eluting Stent was performed. Patient was started on optimal medical medical therapy with risk factor reduction.

Discussion Coronary artery disease (CAD) is more common in elderly and middle age people with significant risk factors, but premature CAD in younger population is unusual in the presence of limited risk factors. Combination of smoking, obesity, dyslipidemia, positive family history and undiagnosed hypertension is enough to cause premature CAD in young patients, which should be differential in atypical presentations. These patient may not have characteristic EKG changes or Cardiac enzyme elevations. It is beneficial to identify even a single risk factor and advise risk reduction early in age.

Conclusion Our case highlights the importance of being vigilant about cardiac pathology in young patients despite atypical presentation. Aggressive risk factor modifications should be considered in these patients.

45 ABSTRACT WITHDRAWN

46 A CASE OF PERICARDITIS AND CARDIAC TAMPONADE

S Werner*, ME Ziada, H Phillips, A McCormick, LS Engel, J Amoss. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.45

Introduction Cardiac tamponade is characterized by an accumulation of pericardial fluid causing compression of cardiac chambers, potentially leading to decreased cardiac output and cardiogenic shock.

Case A 52 year-old man with a history of type 2 diabetes mellitus, chronic diastolic heart failure and tobacco abuse presented to the Emergency Department with gradual worsening dyspnea and chest discomfort for 1 day. Vital signs were unremarkable. Physical exam was significant for a pericardial rub. PR depressions were appreciated on EKG. Computed tomographic scan of the chest revealed a large pericardial effusion. The patient was admitted for pericarditis with pericardial effusion. Echocardiogram showed a moderate sized pericardial effusion without hemodynamic compromise. He was treated with non-steroidal anti-inflammatory medication and colchicine; he demonstrated clinical improvement and was discharged. The patient returned to the hospital five days later for progressively worsening dyspnea with exertion and chest discomfort. Exam was significant for tachycardia, pulsus paradoxus and elevated jugular venous pressure. EKG was unchanged from prior studies. Echocardiogram showed an increase in the effusion and excessive motion of the interventricular septum. The patient was taken for elective pericardiocentesis, where 1200 cc of cloudy serous pericardial fluid was drained. The fluid was significant for red blood cell predominance and was concerning for exudative etiology. Cultures did not grow. Cytology was negative for malignancy. Renal function and blood urea nitrogen were normal. The patient improved significantly and was discharged with further outpatient autoimmune workup.

Discussion This patient had physical exam findings consistent with pericarditis initially, as well as subsequent pulsus paradoxus and elevated jugular venous pressure on repeat presentation. Echocardiography can assist in diagnosis with visualization of cardiac chamber collapse. Management should focus on maintaining hemodynamic stability and removal of pericardial fluid.

47 ACUTE CARDIAC TAMPONADE SECONDARY TO METASTATIC UROTHELIAL CARCINOMA

B Dao*, Z Huston. *Bassett Medical Center, Cooperstown, NY*

10.1136/jim-2018-000974.46

Introduction Cardiac metastasis occurs in 10% of patients with urothelial carcinoma. Only six cases have been described to cause

cardiac tamponade, a rare complication that can result in rapid hemodynamic collapse and requires prompt diagnosis and intervention. Our case presents a rare cause of cardiac tamponade and illustrates the complexities of diagnosis and management.

Case A 58 year old male with a history of urothelial cancer presented with a one day history of worsening dyspnea on exertion, chest pain, dry heaves, and left lower extremity edema. Patient had tachypnea, tachycardia, and hypotension. CT angiography thorax was negative for pulmonary embolism, but demonstrated moderate pericardial effusion. Physical exam revealed pulsus paradoxus and Kussmaul's sign, but EKG did not demonstrate electrical alternans. Bedside transthoracic echocardiogram (TTE) demonstrated right ventricle collapse and right atrial inversion consistent with cardiac tamponade. Emergent pericardiocentesis was performed, producing 800 ml of serosanguinous fluid, and pericardial drain was placed. Pathology of fluid was positive for urothelial carcinoma. Over six days, the drain produced an output of 2.8 L. After multidisciplinary discussion with cardiothoracic surgery, cardiology, and oncology the pericardial drain was removed and TTE prior to discharge showed no reaccumulation. Patient was started on pembrolizumab as an outpatient. Two months follow-up TTE revealed no evidence of recurrent pericardial effusion.

Discussion The nonspecific presentation of cardiac tamponade casts a broad differential, thus making diagnosis difficult. However, clinical outcomes without intervention can be catastrophic. Furthermore, establishing the etiology of tamponade has great implications on management. The majority of previously documented cases utilized pericardial window and systemic chemotherapy to prevent reaccumulation. However, our case demonstrates successful prevention of fluid reaccumulation with prolonged catheter drainage and immunotherapy. In patients with active urothelial carcinoma presenting with dyspnea, malignant pericardial effusion should be in the differential and prolonged catheter drainage should be considered as a successful alternative to a more invasive pericardial window.

48 AN UNCOMMON CAUSE OF CARDIAC TAMPONADE

JR Rivas*, A Richardson, M Morales Hernandez, F Franchi. *University of Florida, Jacksonville, FL*

10.1136/jim-2018-000974.47

Case presentation A 69 years old female with a history of hypertension and hyperlipidemia came to the emergency room with a complaint of worsening shortness of breath, nonproductive cough, chest pain, poor appetite, and unintentional weight loss. She was treated with antibiotics before without improvement. On physical exam, she was tachypneic, tachycardic and hypotensive with distention of the jugular vein, and distant heart sounds. Initial workup showed leukocytosis, lactic acidosis, acute kidney injury, and transaminitis. Electrocardiogram showed normal sinus rhythm without electrical alternans. Computed tomography of the chest revealed a large pericardial effusion and right upper lung mass measuring 2.2 cm. Echocardiography confirms significant circumferential pericardial effusion with right atrium and ventricle free wall collapse consistent with cardiac tamponade (CT). She underwent emergent pericardial window with 800 ml of serosanguineous fluid removed and pericardial biopsy that reported metastatic adenocarcinoma favoring lung primary.

The patient was started Alectinib for Stage IV lung adenocarcinoma.

Discussion Cardiac metastasis is not uncommon in patients with advanced cancer but CT complicating malignant pericardial effusion is a rare presentation of any malignancy. The prevalence of combined metastasis to the pericardium and heart ranges from 0.1% to 21%. Primary lung cancer is the most common cause. The increase in pericardial fluid is due to obstruction of lymphatic and venous drainage of the heart, direct mediastinal invasion, and hematogenous spread. The acute treatment for this emergency involves the prompt removal of the pericardial fluid by pericardiocentesis. Intrapericardial administration of sclerosing agents can be considered as a non-surgical option.

Conclusion Malignant cardiac tamponade from non-small cell lung cancer (NSCLC) is an uncommon clinical entity associated with poor prognosis. Longer term survival is possible in some patients after successful systemic therapy such as molecular targeted therapy. Further research is warranted to guide optimal management and help clinicians prognosticate this understudied emergency in NSCLC.

49 SYMPTOMATIC DYSPNEA FROM MINOXIDIL INDUCED PLEUROPERICARDIAL EFFUSION

M Nayyar*, V Duong, KT Weber. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.48

Case report An 85-year-old male with resistant hypertension placed on minoxidil 6 months prior by his primary care provider (PCP) was evaluated for several month history of dyspnea and bilateral lower extremity edema. Age appropriate screening and laboratory work up was pursued by his PCP, and referral for transthoracic echocardiogram (TTE) for investigation of his symptoms. TTE revealed a large circumferential pericardial effusion with mild diastolic collapse of the RV prompting immediate cardiology assessment in the outpatient echo lab. Examination was significant for a hemodynamically stable with 7 cm of JVD and bilateral pitting edema to his knees with no further significant findings. Laboratory work up was only remarkable for stable normocytic anemia. Given his symptoms and TTE findings, he was then taken back to the operating room for a pericardial window draining 1100 mL of straw colored fluid, a left thoracotomy with drainage of 400 mL of straw colored left pleural effusion, and placement of left pleural chest tubes. Further diagnostic evaluation of the pericardial fluid revealed absence of neoplastic cells, lack of leukocytes and bacterial or fungal growth, negative adenosine with normal protein and glucose. Pericardial biopsy also showed normal tissue. After 1500 mL of pleural fluid was

Abstract 49 Figure 1

drained, his chest tubes were removed and he was discharged with complete resolution of his symptoms.

Minoxidil is an agent of choice in patients with resistant hypertension on several classes of anti-hypertensives. Pericardial effusion and tamponade is rare with minoxidil use but has been reported to be lethal. The complaints of dyspnea, JVD and bilateral edema prompting TTE investigation resulting in likely life saving intervention in our patient. It is prudent to consider tamponade in patients on minoxidil when presenting with these symptoms. Figure 1 reveals large sized effusion with right ventricular mid diastolic collapse representing incipient tamponade.

Moving Your Clinical Case Presentation into a Published Manuscript

2:45 PM

Thursday, February 21, 2019

50 SALMONELLA BRAIN ABSCESS IN AN INFANT

JL Fouth*, E Reade. *University of Tennessee, Chattanooga, TN*

10.1136/jim-2018-000974.49

Introduction *Salmonella spp.* are classically associated with gastroenteritis and bacteremia in specific hosts. We report a case of *Salmonella* causing a brain abscess in an immunocompetent pediatric patient.

Case report A 7 month-old male presented with irritability, refusal to bear weight, and bulging fontanelle. Emergent magnetic resonance imaging (MRI) showed a right-sided fluid filled structure associated with uncal herniation and hydrocephalus (figure 1). Emergent craniotomy was performed and

Abstract 50 Figure 1

demonstrated copious purulent material. Repeat drainage was required due to significant fluid reaccumulation. Fluid cultures grew *Salmonella Enteritidis*. Review of medical records revealed a history of increased head circumference between ages two and four months. He was admitted at 5 months old with several days of diarrhea and fevers. Stool culture was negative during that admission. Patient received six total weeks of ceftriaxone. The patient was discharged after 26 days to an inpatient rehabilitation facility with left-sided hemiparesis.

Discussion Brain abscesses can be secondary to bacteremia, an infected hematoma, or traumatic injury. *Salmonella* brain abscesses are rare in the United States. Our review of the literature showed only one other pediatric case report of *Salmonella* brain abscess. The indolent nature of our patient's presentation was similarly reported in the other case. The child's parents were employed at a chicken processing plant, a known reservoir of *Salmonella*.

51 GETTING HIGH ON LOPERAMIDE

M Konda*, G Krishnan, R Parikh, K Bano, K Defore. *UAMS, Little Rock, AR*

10.1136/jim-2018-000974.50

Case report Loperamide is an over-the-counter inexpensive antidiarrheal medication that is a potent intestinal u-opioid receptor agonist. It was thought to have little misuse potential as it does not readily cross the blood-brain barrier when taken at recommended dosing of maximum 16 mg/day. However, when taken in large amounts, it can cross the blood-brain barrier and cause euphoria or 'high'. We present an atypical case of acute opioid withdrawal caused by naltrexone in a patient who was abusing loperamide.

A 73-year-old-man with history of opioid abuse was transferred to our facility from an outpatient detox center for seizure like activity and explosive diarrhea after he took naltrexone. He denied any recent opioid use. Shortly after he took his first dose of naltrexone, he developed generalized shivering of his body lasting about 10 s with no loss of consciousness. He also complained of abdominal cramps with vomiting and several episodes of diarrhea. His urine drug screen obtained before he was started on naltrexone was negative for opioids or any other drugs. His other work-up including complete blood count, basic metabolic panel, liver function test, ECG were normal. The patient later confessed to taking about 80 pills (160 mg) of loperamide a day to 'get high' for the past month after he found about it on the internet. The patient's presentation was consistent with acute opioid withdrawal precipitated by naltrexone. He was admitted overnight for monitoring due to association of loperamide with arrhythmias. He, however, continued to have several episodes of diarrhea and was discharged to an outpatient substance abuse program.

As prescription opioids have become less accessible, the use of loperamide as an opioid alternative is on the rise as it is readily available. Loperamide toxicity can not only lead to life threatening ventricular arrhythmias and cardiac arrest but there is also a possibility for withdrawal. Health care professionals need to be aware of the potential for loperamide misuse. A thorough history of use of over-the-counter medications should be obtained in patients with potential for substance abuse.

Adolescent Medicine and Pediatrics

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

52 DO NEONATES WITH MASTITIS OR OMPHALITIS OFTEN HAVE INVASIVE DISEASE?

K Baldwin*, NP Shah, CM Pruitt. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.51

Purpose of study Mastitis and omphalitis are rare but potentially serious infections in the neonatal population. Young infants with these infections often undergo extensive testing (including lumbar puncture) with parenteral antibiotics and hospitalization. The objective of our study is to describe the proportion of infants with bacteremia, meningitis, or other manifestations of invasive infection. We hypothesize that the proportion of infants with invasive disease is low.

Methods used Retrospective cohort of infants 0–90 days old seen in our tertiary pediatric emergency department (2007–2016) with ICD-9 or 10 codes consistent with mastitis or omphalitis. Predefined clinical criteria for both diagnoses were confirmed by chart review, with exclusion of benign diagnoses (e.g. umbilical granuloma, nonspecific cord inflammation). Severity of condition was defined as growth of pathogenic bacteria in blood or cerebrospinal fluid (CSF), necrotizing soft tissue infection or other adverse outcome. Demographic, clinical and microbiologic data were collected using a standardized form. Descriptive statistics are presented. Our study is part of a large multicenter study of neonatal mastitis and omphalitis.

Summary of results Of 161 patients screened, 68 met inclusion criteria. We present preliminary data of 21 infants (10 mastitis, 11 omphalitis). All but one infant (20/21, 95%) were admitted to the hospital. Only one infant had invasive infection, which manifested as necrotizing soft tissue infection. No infant had a blood or CSF culture positive for pathogenic bacteria.

Conclusions Neonates with mastitis or omphalitis have low rates of invasive infection. Larger studies are needed to determine clinical factors associated with invasive infection. Absence of these factors could lead to outpatient management for a select population of infants with mastitis or omphalitis.

53 ATTENTION-DEFICIT/HYPERACTIVITY DISORDER, ARE RESIDENTS FOLLOWING GUIDELINES?

¹K Bradham*, ¹R Trimm, ²AG Darden. ¹University of South Alabama, Mobile, AL; ²University of Oklahoma, Oklahoma City, OK

10.1136/jim-2018-000974.52

Purpose of study Attention-Deficit/Hyperactivity Disorder (ADHD) is the most common pediatric neurobehavioral disorder. Although the American Academy of Pediatrics (AAP) have published guidelines about the diagnosis, evaluation, and treatment of ADHD, it is not clear if resident education prepares trainees to apply the guidelines to their practice. The

supervising faculty at the University of South Alabama and senior residents express concerns that the care and management of patients with ADHD does not follow the AAP guidelines. This study allowed for significant and meaningful evaluation of gaps in residents' knowledge in an effort to work toward more efficient education of residents and quality of patient care in the area of ADHD.

Methods used Retrospective chart review of 50 charts of children ages 4–18 years seen April–June 2015. Charts were selected if (1) the child was age 4–18 years, (2) the ICD 9 diagnosis code was related to ADHD (314.00 or 314.01), and (3) if the pediatric resident completed the chart. A single reviewer audited and analyzed the charts using a checklist based on the AAP practice guidelines for clinical diagnosis, evaluation, and treatment of ADHD.

Summary of results Findings indicate that a majority of patient encounters were follow up visits (96%), of which 19% had medication adjustment(s). Symptoms of inattention (19%) and hyperactivity/impulsivity (21%) were addressed at the follow up visits. Academic performance (56%) was the most commonly addressed behavioral component. Co-morbidities were documented 20% of the time. At least one component of the medications side effect profile was mentioned (92%). Many physical examination measurements such as height (66%), weight (100%), blood pressure (96%) were addressed.

Conclusions A majority of the patient encounters were for follow up visits including medication management. Documentation of the discussion of side effects from stimulant medication was a relative strength of residents. Documentation of core symptoms of ADHD was a relative weakness of the residents. The chart review revealed that documentation rates of co-morbidities in this study were below rates documented in the literature. An educational intervention will be developed to improve residents' documentation of core symptoms and co-morbidities of ADHD.

54 DOES POINT OF CARE ULTRASOUND IMPACT OUTCOMES IN PEDIATRIC SKIN AND SOFT TISSUE INFECTIONS?

M Byrd*, NP Shah, CM Pruitt. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.53

Purpose of study In spite of published data on point of care ultrasound (POCUS) for the diagnosis of skin and soft tissue infection (SSTI), there is limited evidence on the impact of POCUS on patient outcomes. Our purpose was to determine if the use of POCUS contributes to improved outcomes for SSTI.

Methods used This is preliminary analysis of an ongoing prospective cohort study in our tertiary children's emergency department. Patients 2 months – 19 years were approached for enrollment if they presented for possible SSTI. Immunocompromised patients, those already on antibiotics, and patients requiring surgical consultation or inpatient admission were excluded. Use of POCUS was at the discretion of the treating provider. Medical charts were reviewed to obtain demographic and clinical information including medical history, examination findings, POCUS

findings, diagnosis and treatment. One-week phone follow-up was performed to query outcomes. Adverse outcomes were defined as a subsequent change in antibiotic regimen, incision and drainage (I and D), or hospitalization. Patients lost to follow up were excluded from the final analysis. Data are presented descriptively, with Chi-square Test used to compare categorical variables, with $p < 0.05$ considered statistically significant.

Summary of results Of 388 patients screened, 230 met inclusion criteria, with POCUS utilized in 107 (46.5%). Median age for both groups was 5.0 years, and 54.8% were female. Abscess diagnosis did not differ between the two groups [POCUS 77/107 (72.0%) vs. no POCUS 97/123 (78.9%), $p = 0.22$]. Adverse outcomes were infrequent, occurring in 34 (14.8%) of the cohort, with subsequent I and D (19/230, 8.3%) being the most common. Adverse outcomes occurred in 15/107 (14.0%) patients with POCUS vs. 19/123 (15.4%) with no POCUS ($p = 0.76$).

Conclusions Our interim findings do not suggest that the use of POCUS impacts patient outcomes for children with SSTI. Larger studies are needed to determine if POCUS leads to improved outcomes for pediatric SSTI.

55 RESPIRATORY SIGNS OR SYMPTOMS AMONG YOUNG INFANTS WITH INVASIVE BACTERIAL INFECTIONS: A MULTICENTER STUDY

B Cawthon*, NP Shah, CM Pruitt. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.54

Purpose of study Young infants are at high risk for invasive bacterial infection (IBI) (defined as bacteremia or bacterial meningitis). Clinicians often imply, if signs or symptoms of respiratory tract infection (RTI) are present, that IBI is less likely, though it is unknown how many young infants with IBI have concomitant RTI signs or symptoms. The purpose of our study is to determine, among young infants with IBI, 1) the proportion who have RTI signs or symptoms, and 2) clinical factors associated with RTI signs or symptoms.

Methods used This is a secondary analysis of an 11-center, 5 year retrospective cohort of infants ≤ 60 days with IBI (defined by growth of an *a priori* pathogen in blood or cerebrospinal fluid culture). In addition to demographic, clinical, and outcomes data, signs or symptoms of RTI were abstracted, as well as viral respiratory test and chest roentgenogram results. Univariate associations were tested by χ^2 , and logistic regression was utilized to calculate odds ratios (OR) with 95% confidence intervals (CI).

Summary of results Of the 397 infants with IBI, 134 (33.8%) had RTI signs or symptoms. Rates of viral respiratory tests (27.7%) and chest roentgenograms (33.5%) were low: 14 (3.5%) infants had a positive viral tests, and 2 had radiographic pneumonia. Infants with RTI signs or symptoms were more likely to be premature (24.6% vs. 11.8%, $p = 0.001$), have a complex chronic condition (19.4% vs. 10.6%, $p = 0.016$), and have a 30 day adverse outcome (38.1% vs. 20.5%, $p < 0.0001$). In the logistic regression model, prematurity (OR 1.87, 95% CI 1.05 to 3.33, $p = 0.03$) and 30 day adverse outcomes (OR 1.96, 95% CI 1.24 to 3.18, $p = 0.007$) were associated with RTI signs or symptoms.

Conclusions In young infants with IBI, signs or symptoms of RTI are not uncommon. Surprisingly, infants with RTI signs or symptoms were more likely to have adverse outcomes. In contrast to existing literature, signs or symptoms of RTI, even with a positive viral test, might not reassure the clinician against the presence of IBI. Prospective comparison studies are needed to corroborate these findings.

56

YOUNG & NOT HUNGRY: DISORDERED EATING IN A 10 YO FEMALE

EN Chiccarelli*, A Glinky, R McPeters, T Kingdon. *LSU, New Orleans, LA*

10.1136/jim-2018-000974.55

Case report We detail a case of disordered eating in a 10 yo female. Initially, the patient presented to gastroenterology clinic with 6 months of postprandial periumbilical pain and weight loss. She received an extensive workup to include abdominal ultrasound and plain film, upper and lower endoscopy, and laboratory studies which were normal. She was then admitted to the floor for a functional pain test, which was positive. Following the functional pain test, she fell asleep and was noted to be bradycardic to the 40 s. On chart review, her weight loss at that point was severe with a decline from the 35th to the 22nd percentile. She remained admitted for medical stabilization and correction of malnutrition. Ultimately due to poor oral intake, the patient required nasogastric tube feedings. On insertion of the tube following administration of an anxiolytic medication, the patient became tearful and was fearful of gaining weight with tube feedings. On retrospective history at the time, family noted increasingly restricted diet at home since the abdominal pain began and was mostly eating vegetables at the time of admission. This patient ultimately fit clinical criteria for anorexia nervosa. When weight and bradycardia stabilized, the feeding tube was removed, and she began eating by mouth. She continued to have functional abdominal pain which was not responsive to medications. Ultimately, she was discharged to intensive outpatient care and family therapy. This case highlights the increasing incidence of eating disorders among prepubertal patients. Clinicians should include disordered eating in the differential diagnosis of rapid weight loss, and bear in mind that such patients may not be forthcoming with their fears, anxieties, and eating habits.

57

INTRODUCTION OF SPIROMETRY USE IN A GENERAL PEDIATRICS ASTHMA CLINIC

HE Cox*, RM Keilman, N Webster, J Whitehead, K Garcia. *University of Tennessee Chattanooga, Chattanooga, TN*

10.1136/jim-2018-000974.56

Purpose of study Asthma affects approximately 1 40 000 children in Tennessee and is the third leading cause of hospitalization. The AAP recommends spirometry testing initially at the diagnosis of asthma and annually. As a part of the state-wide Tennessee AAP initiative, we conducted a quality improvement project to review spirometry use in annual asthma patient visits and then began measures to increase its utilization.

Spirometry Utilization

Abstract 57 Figure 1

Methods used Initial efforts focused on staff education. Resident staff reviewed a 30 min lecture on spirometry while nursing staff attended a spirometry lecture. Following didactic sessions, educational materials were provided to all staff. Spirometry was first introduced using a single asthma patient per day model during the month of December 2017. In January 2018, clinic protocol fully implemented spirometry use in asthma patients. Nursing reviewed scheduled patients the day prior to identify the need for spirometry, then on the day of visit, the patient received spirometry. A retrospective chart review was completed every 3 months to ascertain spirometry utilization.

Summary of results 83% of pediatric residents and 100% of nursing staff completed training for spirometry testing. Prior to implementation, no asthma patients routinely had spirometry performed. After implementation, spirometry use increased to 23.1% within 3 months. Six months after implementation, spirometry was performed in 53.6% of asthma patients. By April of 2018, our clinic was at the state average for spirometry utilization, and surpassed state average by July, 2018 (figure 1).

Conclusions Spirometry education is feasible and can be successfully implemented in the general pediatric clinic. Ongoing educational efforts through didactic session and electronic updates have kept spirometry use >50% which was our initial goal.

58

TEXT REMINDERS TO IMPROVE INFANT VACCINATION IN A RESIDENT-RUN CLINIC

^{1,2}WC Hancock, ^{1,2}R Hunt, ^{1,2}L Kokoska, ^{1,2}B Bagga, ^{1,2}J Yaun. ¹University of Tennessee Health Science Center Memphis, Memphis, TN; ²Le Bonheur Children's Hospital, Memphis, TN

10.1136/jim-2018-000974.57

Purpose of study Immunizations are a cornerstone of pediatric preventive care. Patient reminder systems are an inexpensive and automated way to improve on time vaccination rates. We utilized a previously described effective text reminder system developed by colleagues at the University of Kentucky to alert parents that their child is due for vaccines. We adopted this system in our resident-run clinic (RRC) in an effort to improve the rate of on time immunizations in our infant patient population. This is an interim analysis of the project.

Methods used Infants one month of age and younger seen at the University of Tennessee RRC were eligible. This clinic

cares for a large underserved, urban population in Memphis, Tennessee. 28 infant-parent dyads were consented for participation. They were each given an informational handout and instructed on how to enroll in the program. Parents then received automated text reminders for vaccines based on their child's birthdate. Bimonthly chart reviews were conducted to record dates of immunization administration for each infant. Administration dates that were ± 1 month from the scheduled date were considered on time. Using a random number generator, 28 infants from 2017 whom did not receive the intervention were evaluated for on time immunizations to provide baseline data.

Summary of results Baseline data for the non-intervention group showed that 89%, 61%, and 57% of them received their immunizations on time at 2, 4, and 6 months, respectively. Infants in the intervention group received 86% and 71% of their 2 and 4 month interventions on time. These infants are still completing their 6 month immunizations.

Conclusions On time immunization rates were already very high at 2 months of age for both groups, while data for 6 month immunizations is pending. At 4 months, we saw a 10% increase in on time vaccination rates compared to the non-intervention group, although the results were not statistically significant ($p=0.4$). The difference may become larger and statistically significant as more children are enrolled. Alternatively, this methodology may not be an efficient reminder for our patient population. Ongoing enrollment is underway to evaluate if this is indeed an effective method of improving our infant vaccination rates.

59 FITZ-HUGH-CURTIS SYNDROME, AN UNEXPECTED DIAGNOSIS IN AN EARLY ADOLESCENT PATIENT

N Hidalgo Corral*. *University of South Alabama, Spanish Fort, AL*

10.1136/jim-2018-000974.58

Case report Previously healthy 14 year old female presented with 2 day history of low grade fever, non-bloody, non-bilious emesis and severe right flank and epigastric pain. She was empirically started on Ceftriaxone for suspected UTI. Pain continued and abdominal CT with contrast revealed a 3.2 cm left adnexal mass and peripheral irregular enhancement of the liver. Diagnostic laparoscopy was performed and no ovarian mass was visualized, but a fibrinous material covering the liver was evidenced. Core needle liver biopsies were obtained and found negative. Liver superficial peel reported mesothelial cells. Patient developed a malar rash and hypertension requiring Lisinopril and amlodipine. Laboratory studies demonstrated coombs positive warm antibody anemia, mild thrombocytopenia, lymphopenia, antiphospholipid antibodies and low C4. Methylprednisolone was added for suspected Systemic Lupus Erythematosus (SLE) with improvement of hematologic parameters. Pelvic examination demonstrated, thin cervical mucus. Gonorrhea amplified probe from cervix was positive. Patient was treated for Fitz-Hugh-Curtis Syndrome completing 10 days of Ceftriaxone and Azithromycin with progressive clinical improvement. Patient denied sexual activity or sexual abuse.

Fitz-Hugh-Curtis Syndrome is the acute inflammation of the liver capsule or perihepatitis associated with *Neisseria gonorrhoea* or *Chlamydia trachomatis*. Usually presents in a

sexually active female with right sided abdominal pain, fever and vaginal discharge. It can mimic multiple pathologies. Visualization of fibrinous, patchy exudate described as violin strings adhesions on the surface of the liver is reported. Low complement is associated with disseminated gonococcal infections.

Fitz-Hugh-Curtis Syndrome should be considered in the differential diagnosis of early adolescents females who present with abdominal pain even if there is a negative history of sexual activity. Early adolescent might not report sexual activity and a high index of suspicion should be exercised in this age group to avoid missing the diagnosis and delaying treatment. Some studies have shown a predisposition to have disseminated gonococcal infection in patients with low complement like patients with SLE.

60 ABNORMAL UTERINE BLEEDING, WHAT A HEADACHE

SR Induru*, S Hill, N Durant. *University of Alabama, Birmingham, AL*

10.1136/jim-2018-000974.59

Presentation A 15 year old girl with abnormal uterine bleeding (AUB) for 2 years presents with AUB, headaches, and galactorrhea. Oral contraceptives and medroxyprogesterone acetate have been unable to manage her AUB, described as heavy vaginal bleeding with clots and cramping. She also noted new onset leakage of cloudy white discharge from both breasts for 1 week and denied manipulation of breasts, pain, trauma, or bloody discharge. Patient is sexually active, denies illicit drug use, and is currently incarcerated. Remainder of history is significant for cold intolerance and worsening daily headaches without aura or visual changes, photophobia, phonophobia, or anosmia. Her exam is significant for white discharge from the nipples bilaterally but benign neurologic and fundoscopic exams.

Management Differential diagnosis for galactorrhea with associated menstrual abnormalities and headaches included pituitary adenoma, hypothalamic causes such as stress, side effects from medications such as medroxyprogesterone, systemic diseases such as systemic lupus erythematosus or thyroid disease, and pregnancy. Labs included prolactin 13 ng/ml (range 3–24 ng/ml), elevated Factor VIII, but otherwise normal von Willebrand panel, renal panel, thyroid function, LH, FSH, GH, and pregnancy tests. Pelvic ultrasound (US) revealed an 'elongated, cystic structure associated with the right ovary [that] could represent a so-called daughter cyst.' MRI head with and without contrast revealed 'subtle enhancement in the right pituitary measuring 0.5 cm $\times$ 0.5 cm.' Patient was referred to pediatric adolescent gynecology, hematology, and endocrinology.

Discussion Due to incarceration, further evaluation by specialty teams was delayed. Pelvic US results were most likely due to a prior STI. MRI mass reviewed to be an incidentaloma, not a prolactinoma as those have prolactin levels greater than 100 ng/ml and are usually associated with oligomenorrhea. Multidisciplinary discussion after her work-up revealed stress as the likely cause of her symptoms. This case illustrates the importance of having real-time multidisciplinary discussions on patients with multiple vague complaints and complex social situations prior to an extensive work up to provide expedient, cost effective care.

61 COMMUNICATION IS KEY: PATIENT, CAREGIVER, AND PHYSICIAN PERSPECTIVES ON TRANSITION

S Leung*, M Walgrave, S Mennito. *Medical University of South Carolina, Charleston, SC*

10.1136/jim-2018-000974.60

Purpose of study A transition of care (TOC) process from pediatric-to-adult medicine ensures that adolescents receive ongoing care into adulthood, a time of high risk for preventable mortality and morbidity. Most transition research excludes healthy teens, who account for 75% of pediatrics. This research explores patient, caregiver, and physician perspectives on TOC for healthy adolescents and how TOC can be improved.

Methods used Using a semi-structured interview guide, two researchers conducted key informant interviews with adolescents ages 12–18 y and their caregivers seen in Pediatric Primary Care Clinic. Primary care doctors who work in Pediatrics, Internal Medicine, and Family Medicine were also interviewed. Three researchers coded the data via descriptive qualitative analysis, identified themes based on code frequency, and generated a communication model.

Summary of results Twelve adolescents, 10 caregivers, and 36 physicians were interviewed. Prior research has focused on the systematic steps of transition. This study reveals the critical role of communication in those steps. The generated model establishes a progression of communication responsibilities for physicians and patients. Educating physicians about patient-centered communication for TOC with appropriate system supports is essential. Physicians must take ownership of their patients' TOC as well as improve communication and rapport building to increase patient knowledge on TOC and autonomy. Thus, patients can be better informed decision makers and adhere to continuity of care resulting in improved patient health outcomes.

Abstract 61 Figure 1

Conclusions An effective TOC process is dependent on strong, consistent communication. Educating physicians about effective communication in TOC is the start to helping patients establish continuity of care. Effective communication bridges physicians and patients, resulting in better patient-centered health outcomes. Interventions centered on TOC communication can be identified and improve care for all adolescents.

62 FACTORS ASSOCIATED WITH FALSE POSITIVE SEPSIS SCREENS IN THE PEDIATRIC EMERGENCY DEPARTMENT

E Li*, NP Shah, CM Pruitt. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.61

Purpose of study While recognition bundles are recommended for pediatric sepsis, validated criteria for such screens are

Abstract 62 Table 1 Sepsis Recognition Bundle Criteria

Screening Criteria, n (%)	False Positives (n=53)	Possible Sepsis (n=47)	P*
High-risk condition	26 (49%)	37 (79%)	0.002
Temperature abnormal	40 (76%)	32 (68%)	0.41
Heart rate elevated	46 (87%)	37 (79%)	0.28
Respirations elevated	35 (66%)	26 (55%)	0.27
Systolic BP low	3 (5.7%)	4 (8.5%)	0.58
Skin abnormal	2 (3.8%)	7 (15%)	0.052
Capillary refill abnormal	1 (1.9%)	8 (17.0%)	0.008
Pulse abnormal	0	5 (10.6%)	0.015
Mental status abnormal	3 (5.7%)	13 (27.7%)	0.003

*univariate (χ^2)

lacking. False positive screens can engender 'alarm fatigue' and lack of urgency for patients who need timely therapies. The purpose of our study was to evaluate which elements of our emergency department (ED) sepsis screening tool are associated with false positive alerts.

Methods used Retrospective cohort of ED patients with positive screens for sepsis (May-September 2017). The tool employs age-specific vital signs, examination findings, and pre-existing high-risk conditions. False positive sepsis alerts were defined by patients lacking any of the following: ≥ 2 isotonic fluid boluses; blood culture and parenteral antibiotics; or inpatient admission. Medical records were reviewed to abstract demographic and clinical information. We report descriptive statistics, utilizing χ^2 to compare categorical variables. Multivariate logistic regression was used to determine criteria associated with false positive screens.

Summary of results 100 subjects have been preliminarily analyzed, with 53 (53%) false positive sepsis screens. Univariate comparisons are displayed in Table 1. In the regression model, the only criterion significantly associated with false positive screens was lack of a high-risk preexisting condition ($p=0.034$).

Conclusions Our ED recognition bundle lacks specificity for pediatric sepsis. Presence of a high-risk condition can help clinicians more strongly consider sepsis for patients with positive screens. Prospective studies are needed to derive and validate screening criteria for pediatric sepsis.

63 AN ACCOUNT OF ORAL MUCOSAL LESIONS

E Landry, MN Frascogna, L Munk*. *University of Mississippi Medical Center, Jackson, MS*

10.1136/jim-2018-000974.62

Case report Mucocutaneous eruptions are common in the pediatric population. We present a well-described but uncommon syndrome with a unique presentation.

16 year old male with no past medical history presented with oral lesions, fevers, cough, and weight loss. Two weeks prior to presentation, he was seen by his physician for cough and fever. He was started on Amoxicillin for suspected pneumonia. Four days later, he went to his local ER and was prescribed Azithromycin for possible pneumonia. His fevers resolved, but he developed several painful lesions on his buccal mucosa and began to experience red and watery eyes. The lesions spread to involve his posterior

pharynx, fluid filled and hemorrhagic bullae formed. He was unable to eat or drink due to pain, resulting in a 20 lb weight loss over 2 weeks. His initial laboratory workup revealed a leukocytosis of 12.5. A chest x-ray was obtained, showing no focal consolidation. His presentation was concerning for the development of Stevens Johnson Syndrome vs Toxic Epidermal Necrolysis vs Drug Reaction. He was admitted to the hospital for IV hydration and workup. Pediatric Dermatology and ENT were consulted. HIV, herpes simplex, varicella zoster, Epstein Barr virus and ANA were all unrevealing. Given his respiratory symptoms along with mucocutaneous lesions, mycoplasma studies were obtained. Mycoplasma IgG and IgM by IFA returned positive. While hospitalized, he developed a migratory erythematous dermatopathy consistent with Mycoplasma induced mucositis and rash. Supportive care was continued throughout his stay. At discharge, his lesions were improving and he was able to maintain oral hydration.

In a pediatric patient presenting with respiratory symptoms with mucocutaneous eruptions, the more uncommon presentation of mycoplasma induced rash and mucositis should be considered. This syndrome is most prominent in young males. It is characterized by prominent mucosal lesions with less severe skin findings. Classic diagnostic criteria includes detachment of <10% of body surface area, involvement of at least 2 mucosal sites, with or without skin findings, and evidence of atypical pneumonia. Treatment involves antibiotics and supportive care including hydration, pain control and mucosal care. Immunosuppression with systemic corticosteroids and IVIG has been used in severe cases.

64 FAILURE TO THRIVE AS A HEALTH INDICATOR OF UNSAFE NEIGHBORHOODS

¹A Paracha*, ¹E Hemming, ¹A Hendrix, ²R Munoz, ³A Merrill, ²S Schaefer, ¹J Hayes.
¹University of Oklahoma, Oklahoma City, OK; ²University of Oklahoma, Tulsa, OK; ³Legal Aid of Oklahoma, Tulsa, OK

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Purpose of study This study explored the relationship between poor neighborhood safety, as indicated by poor housing and rate of violent crimes such as homicide and rape, and incidences of failure to thrive (FTT), an indicator of poor growth and development due to undernutrition, in pediatric patients living in Tulsa, OK in 2013–2014.

Methods used Environmental variables used as predictors of FTT were poor housing conditions, as captured by housing code violations, and neighborhood safety, as captured by the incidences of homicide and rape per census tract. Housing and crime data were collected from the Tulsa Health Department and Tulsa Police Department respectively for the Tulsa metro area. FTT diagnoses were collected from the University of Oklahoma Health Sciences Center electronic medical records serving the same area. 225 census tracts and 2 calendar years of data were included in analysis. 2013 was used as the calibration sample for the prediction model while 2014 was used as a validation sample. Hierarchical regression was used to analyze the data.

Summary of results Results from the calibration sample in step 1 of the hierarchical regression model indicated that housing complaints were a significant predictor of FTT cases in a particular census tract ($R_{adj}^2=0.063$; $p<0.01$). In step 2, after controlling for poor housing conditions, the results indicated

that incidences of homicide and rape were also distinct and significant predictors of FTT cases in a particular census tract ($\Delta R^2=0.065$; $p<0.001$). Results from the validation sample supported the overall stability of the model, with housing complaints once again serving as a significant predictor of FTT cases ($R_{adj}^2=0.043$; $p<0.01$), while incidences of homicide and rape continued to serve as unique predictors of FTT cases in a census tract after controlling for housing conditions ($\Delta R^2=0.046$; $p<0.001$).

Conclusions Our research suggests the importance of interventions at the community level that improve both housing quality and neighborhood safety in order to significantly reduce the incidence of FTT diagnoses in the pediatric population for a given community.

65 THE ENTIRE OFFICE IS IMPORTANT FOR EFFECTIVE ADOLESCENT VACCINATION

¹J Roberts*, ¹E Dawley, ²C Garbe, ²M Cooper, ¹E O'Brien, ²C Madden, ²PM Darden.
¹Medical University of South Carolina, Charleston, SC; ²Oklahoma University Health Sciences Center, Oklahoma, OK

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Purpose of study Vaccine delivery rates for Quadrivalent Meningococcal Conjugate Vaccine (MenACWY), Tetanus, Diphtheria, Acellular Pertussis Vaccine (Tdap) and Human Papillomavirus (HPV) vaccine vary. The purpose of this study was to compare the attitudes and practices of parents of adolescents, front office staff, nurses and clinicians for each vaccine.

Methods used Self-completed surveys about MenACWY, Tdap and HPV vaccines. Questions focused on the perceived importance of each vaccine as well as strength of recommendation. Both front office staff and parents of 11–17 year olds were asked about receipt of these vaccines. Front office staff responded about vaccine-related questions parents directed to them.

Summary of results Twenty practices in two practice-based research networks in OK and SC contributed 311 parent, 108 front office staff, and 196 nurse/clinician surveys. Parents reported clinicians recommend each vaccine, 58%, 69%, 68% for MenACWY, Tdap, HPV respectively ($p=0.01$). Parents reported lower proportions of children received MenACWY and HPV vaccine v. Tdap (54%, 51% vs 72%, $p<0.0001$). Parents perceived MenACWY and Tdap as being very important for the health of their child compared to HPV vaccine (66% and 67% v. 58% v, $p=0.04$). Parents perceived practices place equal importance on all three vaccines (78%, 78%, 76%). Front office staff reported more questions from parents about HPV vaccine than MenACWY and Tdap before the visit (54% vs 25%, 21%; $p<0.01$) and after the visit (26% vs 8%, 9%; $p<0.01$). Nurses and clinicians reported giving a strong recommendation to fewer adolescents for HPV vaccine than for MenACWY and Tdap (76% vs 88% and 88% respectively; $p<0.05$). Nurses and clinicians placed less personal importance on the HPV vaccine compared to the MenACWY and Tdap (87% vs 94%, 93%; $p<0.05$), and the importance the office places on HPV vaccine (85% vs 92%, 92%; $p<0.05$).

Conclusions Parents and nurses/clinicians perceived the HPV vaccine as being less important than MenACWY and Tdap. Recommendations for each vaccine are not consistent. Parents are leaving the visit with questions about HPV vaccine. Complete office agreement on the importance of HPV vaccine and getting information to parents should be a priority.

66

RATES OF EPINEPHRINE ADMINISTRATION FOR TREATMENT OF PEDIATRIC ANAPHYLAXIS IN A LARGE METROPOLITAN EMERGENCY MEDICAL SERVICES SYSTEM

¹N Smith*, ¹M Anderson, ²M Cox, ¹C Knoles. ¹University of Oklahoma Health Sciences Center, Oklahoma City, OK; ²MCD/OMD OKC and Tulsa EMS, Oklahoma City, OK

10.1136/jim-2018-000974.65

Purpose of study Anaphylaxis is a life-threatening emergency requiring immediate recognition and treatment to avoid potentially devastating effects including death. Studies to date show a deficit in the ability or willingness of Emergency Medical Services (EMS) providers to correctly identify anaphylaxis and administer epinephrine prior to arrival at the emergency department. The purpose of this study is to evaluate the EMS system in Oklahoma City and Tulsa to determine the rate of appropriate epinephrine administration in the pediatric population.

Methods used A retrospective chart review was performed using EMS records for patients from 0 to 18 years of age who were transported to the Emergency Department by EMS between July 1 2013 and March 1 2018. A total of 410 pediatric patients were identified with the documented impression of 'Allergies' or 'Drug Allergy'. The charts were reviewed for history, physical exam findings and interventions. An algorithm based on the World Allergy Organization criteria was used to evaluate each case and patients were categorized based on the indication for epinephrine administration and the subsequent administration of epinephrine.

Summary of results Of the 410 records reviewed, a total of 64 patients were found to meet the criteria for epinephrine administration. Of those, 13 patients (20.31%) were given epinephrine while 51 patients (79.69%) were not given epinephrine. An additional 6 patients were given epinephrine without meeting WAO criteria, making a total of 19 patients that were given epinephrine, with 68.42% of those doses being indicated. Of note, 239 of the patients reviewed were found to fit criteria for a mild allergic reaction with no treatment indicated or given. Further, 101 of the patients had received epinephrine prior to EMS arrival and assessment.

Conclusions This retrospective chart review confirms the findings of previous similar studies showing that there is a deficiency in the appropriate use of epinephrine by EMS Providers. Plans are in process to further investigate the cause for this deficit. Furthermore, steps are being taken to both educate EMS personnel and adjust protocols to increase the rate of administration of epinephrine to patients who would benefit.

67

MENTAL HEALTH BELIEFS AND REFERRAL PRACTICES AMONG PEDIATRIC PRIMARY CARE RESIDENTS

S Wee*, R Hanson, E Wallis, F Pastrana, E Fanguy. Medical University of South Carolina, Charleston, SC

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Purpose of study While children exposed to potentially traumatic events face increased risk for adverse physical and mental health concerns, many are neither identified nor referred for trauma-focused interventions. Pediatricians are often a

child's only contact with a health professional, offering an opportunity to be a bridge to mental health treatment services for vulnerable youth. It is unclear whether pediatricians have the knowledge to screen and appropriately refer youth for mental health services. This study examined relations among pediatric primary care residents' beliefs and practices related to mental health and referrals.

Methods used Pediatric primary care residents ($n=49$) completed a survey examining:

- number of referrals in the past six months for mental health, trauma-specific, and abuse/neglect concerns; and
- beliefs about mental health services.

Beliefs were assessed with a modified 17-item version of the Physician Belief Scale (PBS), with responses rated on a 5-point Likert scale. A sum score was computed from the PBS items. Analyses included chi-square tests, univariate Analysis of Variance, and Spearman's correlation.

Summary of results 55.1% of participants reported making 1–5 general mental health referrals in the past 6 months, and 34.6% made ≥ 6 referrals. 61.2% reported making 1–5 trauma specific referrals, while 36.7% made no referrals. 73.5% of residents made 1–5 referrals for abuse and neglect concerns, and 24.5% made no referrals. No significant difference was found among referral practices across residency years. The mean sum score for the PBS was 36.85 ($SD=6.41$, $Range=24-51$). No significant differences in beliefs were found across residency years. Trauma-specific referrals were positively associated with general mental health referrals ($r_s=0.42$, $n=49$, $p=0.002$). No other significant relations were found between number of referrals and mental health beliefs across residency years.

Conclusions This study suggests residents who make general mental health referrals are more likely to refer for trauma-specific concerns; beliefs about mental health may not be associated with referral practices; and beliefs about mental health appear to be stable across residency years.

Adult Case Reports/Aging/Geriatrics

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

68

SPONTANEOUS ILIOPSOAS MUSCLE HEMATOMA AND DEEP VEIN THROMBOSIS IN PATIENT ON LOW DOSE ASPIRIN

M Abdou*, MA Alsharif, M Naguib. Texas Tech Univ HSC Amarillo, Amarillo, TX

10.1136/jim-2018-000974.67

Case report Iliopsoas muscle hematoma is a rare condition often associated with large bleeding and can be life threatening. It is usually traumatic but can occur spontaneously during coagulopathy due to hemophilia or anticoagulation.

We report a case of spontaneous iliopsoas muscle hematoma in a patient only on low dose aspirin and occasional NSAIDs.

80 year-old man with hypertension, diabetes, and abdominal aortic aneurysm repair presented with 3 week-history of right hip pain and 3 days history of right leg swelling interfering with weight bearing. Radiographs showed only mild degenerative changes and venous doppler of the lower extremity showed no thrombosis. He had no trauma. Medications were aspirin 81 mg, metoprolol, simvastatin, metformin and occasional NSAIDs. Range of motion of the right hip was restricted with pain and 3+ pitting edema of the right leg was noted. Vitals signs were normal. Hemoglobin was 8.2 g/dL (baseline 11.2), BUN 38 mg/dL, serum creatinine 1.4 mg/dL, platelets and INR were normal. CT scan of the right hip showed a 10 cm hematoma in the right iliopsoas muscle. CT angiogram of the aorta ruled out vascular connection to the hematoma. The spontaneous hematoma had no identifiable etiology and was presumed to be related to the use of low dose aspirin with occasional NSAIDs. Percutaneous drainage yielded 800 ml of culture negative blood. A month later, he returned with right deep venous thrombosis identified on ultrasound due to compromised venous return by the hematoma. Anticoagulation was not pursued, and inferior vena cava (IVC) filter was placed.

In conclusion, iliopsoas hematoma should be included in the differential diagnosis of acute flank or hip pain in persons not on anticoagulation. Also, the condition can produce ipsilateral lower leg venous thrombosis. Since anticoagulation is contraindicated due to spontaneous bleeding, IVC filter placement seems appropriate to avoid thromboembolic complications.

69 DO NOT NEGLECT A NECK PAIN

C Ashangari*, MK Islam, A Taha, R Bharadwaj, B Khandheria. *Texas Tech Univ HSC Amarillo, Amarillo, TX*

10.1136/jim-2018-000974.68

Background For centuries, the diagnosis and treatment of deep neck space infections have challenged physicians and surgeons. The complexity and the deep location of this region make diagnosis and treatment of infections in this area difficult. These infections remain an important health problem with significant risks of morbidity and mortality.

Abstract 69 Figure 1 Tagged WBC scan

Case 71 year old male with PMH of CAD, DM presented with generalized weakness, neck pain and body ache for 3 weeks. Vitals were normal and there was tenderness in the neck on physical exam. Labs showed high WBC. Blood cultures were positive for gram-positive cocci. He was started on antibiotics. TTE was suspicious for possible aortic valve thickening which did not exclude vegetation. TEE showed thickening and suspicious lesion in the aortic valve, and vegetation cannot be ruled out completely. Infective endocarditis management was continued. Tagged WBC scan showed large accumulation of white blood cells seen in anterior left chest, extending from region of clavicle to his sternum. The next day, repeat CT neck soft tissue was done because of increased pain which showed intramuscular abscess involving sternocleidomastoid and possible extension into the left sternoclavicular joint. IR guided abscess drained 6 mL of pus. Blood, abscess fluid both grew MSSA. With abscess drainage and IV antibiotic, his symptoms improved significantly and did well with physical therapy.

Discussion Direct extension of infection through fascial planes may involve deep neck spaces. Vital organs are at risk of being involved even leading to death. Early diagnosis and medical management can be lifesaving.

70 PULMONARY MANIFESTATIONS OF RENAL CELL CARCINOMA

C Ashangari*, MK Islam, A Taha, MA Alsharif, P Tumula, B Khandheria. *Texas Tech Univ HSC Amarillo, Amarillo, TX*

10.1136/jim-2018-000974.69

Background Pulmonary embolism (PE) is a rare cause of respiratory failure in patients with cancer. Such an embolism may occur at any stage of the patient's illness and indeed may be the first presentation of an occult carcinoma. We present a case of PE that can rarely present as the initial manifestation of renal cell carcinoma diagnosed on CT scan.

Case 66 year old male with PMH of HTN and HLD presented to the ER with dyspnea and chest pain. He was transferred from an outside facility for further evaluation and care after he got TPA due to acute pulmonary embolism diagnosed on CT angio scan. He denied blood in the stool, hematuria, hematemesis. Patient complained of weight loss and tiredness for the last 2 months. He denied any PCP visit for several years. He chews tobacco. His oxygen saturation was 92% on 2 L of oxygen. Physical examination revealed tachycardia, mild tachypnea. Hemoglobin was 10.4. UA showed 3–5 RBC. EKG showed slight S1, Q3 but no obvious T inversion in III. There were multiple pulmonary nodules on CXR involving both lung field, suggestive of metastatic lung disease. CTA showed a massive thrombus occluding the right main pulmonary artery and multiple pulmonary nodules throughout both lung fields measuring upto 2 cm in diameter. Echocardiography showed slight dilatation of the right ventricle with mild tricuspid regurgitation. CT abdomen/pelvis showed right renal mass with renal vein and IVC involvement. Doppler ultrasonography of the lower limbs did not reveal evidence of DVT. His symptoms improved gradually over the next 72 hour. IR guided lung biopsy showed metastatic renal cell carcinoma. Patient was discharged on lovenox, pazopanib and follow up CTA.

Repeat CTA after 3 months showed no PE and the patient is doing well.

Discussion The diagnosis of pulmonary tumor embolism is difficult to make before death, but it is a not uncommon finding at autopsy, where an incidence ranging between 1% to 26% has been reported in patients with known malignant tumors. Thus renal cell carcinoma should be considered in the differential diagnosis when symptoms occur without any apparent source. A high index of suspicion can lead to an early diagnosis of renal cell carcinoma and can assist in instituting an early intervention improving overall prognosis.

71 METFORMIN ASSOCIATED LACTIC ACIDOSIS

S McClelland*, Q Palmer, J Frazier, S Beland. *University of Arkansas for Medical Sciences, Little Rock, AR*

10.1136/jim-2018-000974.70

Case report 83-year-old AAF with HTN, uterine cancer, diabetes, CAD, heart failure, and CVA presented to the ED with worsening shortness of breath, lower extremity swelling, and diarrhea. Patient's shortness of breath worsened over one month and furosemide dose was increased and metoprolol was initiated. Patient's metformin had also been increased two weeks prior to presentation for tighter blood glucose control while receiving carboplatin with steroids for newly diagnosed uterine cancer. After increasing metformin, patient had 5–10 loose stools daily. Admission labs H/H: 9.4/29.1, CO₂: 9, BUN/Cr: 50/2.3, Na: 128, Glucose: 225, BNP>4800, troponin 0.09, lactate: 6.8, and ABG: 7.42/18/110. ECG showed sinus tach with LVH. Physical exam significant for dyspnea, crackles to bilateral lung bases, and 1 +bilateral lower extremity edema. Metformin was discontinued and DM was controlled with SSI while inpatient. Within 12 hours of admission, lactic acidosis and diarrhea resolved, and shortness of breath improved with IV lasix drip. AKI improved during hospitalization with creatinine returning to 1.5 at discharge.

In summary, this case report highlights the importance of identifying patients at risk of developing metformin associated lactic acidosis (MALA) and the appropriate management. MALA is a rare, but potentially fatal complication with a mortality rate ranging from 10% to 45%. Patients with predisposing risk factors, such as, renal dysfunction, decompensated heart failure, hypoxemia, alcoholism, cirrhosis, contrast exposure, sepsis and shock, are more likely to develop MALA. MALA pathogenesis is attributed to impairment of metformin clearance, tissue oxygenation, and impaired lactate metabolism. Gastrointestinal side effects are common with initiation and increased doses of metformin. Gradual up titration of metformin and close clinical and laboratory monitoring may have decreased the likelihood of developing MALA. Management of MALA includes supportive care, discontinuing metformin, and treatment of underlying conditions. However, many patients also require hemodialysis to correct metabolic acidosis and eliminate lactate and metformin. Despite the rarity of metformin associated lactic acidosis, it is imperative for clinicians to

recognize predisposing risk factors and take appropriate action.

72 VISUAL HALLUCINATION WITH INTACT COGNITION: A RARE CASE OF CHARLES BONNET SYNDROME

¹P Poudel*, ²S Brangman. ¹Suny upstate medical university, Syracuse, NY; ²Suny Upstate, Syracuse, NY

10.1136/jim-2018-000974.71

Introduction Visual hallucinations are very confusing experiences for patients. When they occur in the setting of visual impairment and intact cognition, after ruling out other potential causes, it is known as Charles Bonnet Syndrome (CBS). We present one such case.

Case An 86-year-old woman presented with a complaint of seeing abnormal objects such as dump trucks in her driveway and her ex-husband in her house. She had no significant findings on physical examination. CBC, CMP, urinalysis, TSH and vitamin B12 levels were normal. CT head was unremarkable. Ophthalmic examination showed diabetic macular edema with macular degeneration of both eyes. Without any history of psychotic symptoms in the past, her symptoms couldn't be attributed to any psychiatric disorder. She scored 4/5 on mini cognitive assessment and had the good memory of all her past activities. A presumptive diagnosis of CBS was made. The patient was reassured of the benign nature of the hallucinations and eventually discharged home.

Discussion CBS was first described by Charles Bonnet in 1760 after his grandfather started experiencing visual hallucinations. It is due to deafferentation of the visual association cortex after the damage to the neurons caused by lesion along the visual pathway. The decreased sensory input stimulates intracerebral perception and causes spontaneous neuronal discharge manifesting in the form of hallucinations.

Conclusion Although CBS was first described in 1760, it is still under-recognized, patients are often referred for inappropriate psychiatric care. There is no effective treatment and management lies on assuring sanity to the patients.

REFERENCE

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Abstract 72 Figure 1 Fluorescein angiography showing ARMD

J Surla*, J Dillon, K Hoppens, M Bernton, B McConaughy, K Zamora, LS Engel, J Martinez. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.72

Introduction Methanol is a highly toxic with poisoning occurring through cutaneous, gastrointestinal, or inhalational exposure. Ingestion of even a small amount of methanol can lead to development of profound high anion gap metabolic acidosis and ocular toxicity.

Case A 37 year old woman with a history of post-traumatic stress disorder, Bipolar disorder, alcohol and inhalant abuse was brought to the Emergency Department by police for evaluation of loss of consciousness followed by facial trauma after inhaling a dust cleaner containing difluoroethane. She denied any other drug or alcohol use. She reported having diarrhea after ingesting an unknown substance at a party 2 days prior to presentation. She otherwise denied nausea, vomiting, abdominal pain, drowsiness, or vision change. On presentation, she was afebrile, HR 100 bpm, BP 127/74 mmHg, RR 18/min, SpO₂ 96% on RA. Physical exam was remarkable for diffuse swelling and ecchymosis of her left eye. Her exam was otherwise unremarkable. Initial laboratory findings were significant for an elevated serum methanol level of 94 mg/dL. Serum ethanol and urine toxicology screen were negative. BMP: Na 142 mmol/L, K 3.3 mmol/L, Cl 113 mmol/L, HCO₃ 19 mmol/L, BUN 9.0 mg/dl, Cr 0.66 mg/dl, Glu 109 mg/dl, AG 10. ABG: pH 7.35, pCO₂ 38, pO₂ 76 mmHg, bicarbonate 21 mmol/L. The measured and calculated serum osmolality were 286 mOsm/Kg and 293 mOsm/Kg, respectively. Per toxicology recommendation, the patient was started on fomepizole. Serum methanol decreased to undetectable levels within 24 hours without HD.

Discussion Our patient, surprisingly, did not demonstrate the stigmata of methanol toxicity in the presence of a severely elevated serum methanol level. She had no visual or neurologic changes. She did not have a high osmolal gap or high anion gap metabolic acidosis, as would be expected. Interestingly, the patient's methanol level decreased to undetectable levels within 24 hours. This is unusual, particularly since previous studies have shown that the plasma half-life of methanol during fomepizole treatment is greater than 48 hours.

Allergy/Immunology/Rheumatology

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

K Ellard*, BL Boudreaux, M Bravo, LS Engel, R McCarron. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.73

Introduction Systemic lupus erythematosus is a chronic autoimmune illness that can affect multiple organs. Neurological

symptoms include cognitive dysfunction, delirium, seizures, and headache.

Case A 21-year-old woman was brought to the hospital following two witnessed seizures. At the time of presentation, she had a leftward gaze with general tonic clonic seizures for which she received multiple rounds of lorazepam. She continued to thrash around in bed and required intubation and propofol. On physical exam, she had dilated pupils at 5 mm that were minimally reactive to light. Neurological exam was limited due to her sedation however no focal deficits were noted. She had severe anasarca. Her initial labs included a creatinine of 2.0 mg/dl, an elevated potassium of 6.0 mEq/L, a CPK of 370 ng/ml, and a lactic acid of 2.5 mg/dl. Her toxicology screen was positive for cannabinoids. Her urinalysis had 600 mg of protein. Neurology ordered levetiracetam. Magnetic Resonance imaging of her brain demonstrated bilateral parietal occipital and posterior frontal areas of abnormal signal involving the gray matter and adjacent subarachnoid spaces consistent with PRES syndrome. She was positive for ANA and anti-DNA with low C3/C4, consistent with systemic lupus erythematosus. Cerebrospinal fluid studies were unremarkable. She required intermittent dialysis for her lupus nephritis, and renal biopsy was consistent with necrotizing crescentic glomerulonephritis. PRES syndrome pathology was thought to be secondary to lupus cerebritis. Her condition improved with prednisone, mycophenolate, and hydroxychloroquine.

Discussion The patient described above had the typical labs (positive ANA/anti-DNA and low C3/C4) as well as clinical findings (kidney dysfunction, neurological involvement) that led to a diagnosis of systemic lupus erythematosus. Her MRI findings were consistent with lupus cerebritis. Renal biopsy helped confirm the diagnosis of lupus. Treatment of lupus focuses on decreasing disease activity and minimizing organ damage and frequently includes hydroxychloroquine with/without a course of glucocorticoids. SLE has a variable clinical course ranging from benign illness to organ failure, and death, so it is critical to make the diagnosis and begin treatment early in order to help prevent organ damage.

J Kennard*. *University of Mississippi Medical Center, Brandon, MS*

10.1136/jim-2018-000974.74

Introduction Antisynthetase syndrome is a rare immune-mediated disorder that is seen in patients with myositis who have antibodies against amino-acyl t-RNA synthetases. Here we present a case of a patient who was treated for recurrent multifocal pneumonia but was later diagnosed with antisynthetase syndrome.

Case report A 41-year-old female presented to the emergency department after a fall. She reported increasing muscle weakness and shortness of breath over the past several weeks. Prior to this presentation, she had been admitted with similar symptoms twice in the past four months. During the previous admissions, imaging was consistent with multifocal pneumonia and she was treated with antibiotics and steroids.

Physical exam was significant for hypoxia, coarse rales over the lung bases and 3/5 muscle strength in the proximal

muscles of the upper and lower extremities. Dermatologic exam was benign. Computed tomography of the chest revealed diffuse bilateral ground glass opacities and focal consolidations. Human immunodeficiency virus, mycobacterium tuberculosis, fungal antigens, and broncheolar lavage cultures returned negative. Anti-nuclear antigen (ANA) was negative; however, extractable nuclear antigen panel returned positive for anti-Jo-1 antibody. Creatinine kinase (CK) and aldolase levels were elevated at 3450 and 106.4 U/L, respectively. Magnetic resonance showed edema and enhancement of the muscles of the pelvis and femurs. Muscle biopsy revealed inflammatory infiltrates extensively throughout consistent with polymyositis. She was started on treatment for interstitial lung disease (ILD) and inflammatory myositis with steroids, intravenous immunoglobulin and mycophenolate mofetil. The patient responded with improvement in dyspnea and weakness. CK and aldolase improved to 170 and 5.9 U/L respectively at follow-up four weeks later.

Discussion Features of antisynthetase syndrome can include myositis, ILD, non-erosive arthritis, and mechanic's hands. Myositis and ILD are the most common presenting features and in up to 20% of cases, ILD may be the initial presentation. This patient's initial symptoms were dyspnea and cough later followed by rapidly progressing muscle weakness. In a patient presenting with dyspnea and recurrent pneumonia, antisynthetase syndrome should be on the list of differential diagnoses.

76 AN UNEXPECTED PRESENTATION AT AN ATYPICAL AGE. A CASE OF HENOC SCHONLEIN PURPURA

S Khanal*, P Ghimire, D Subedi. *SUNY Upstate Medical University, Syracuse, NY*

10.1136/jim-2018-000974.75

Case report We present an 81 year old female with medical history of hypertension, presenting with acute onset left sided hemiparesis, facial droop and blurry vision. On arrival, airway and vitals were stable and a stroke code was initiated. CT scan of head and MR angiogram of head and neck were negative for obvious intracranial hemorrhage, ischemia, significant stenosis or plaque buildup. Per neurology, reperfusion was not indicated.

On further questioning, she gave history of abdominal pain and diffuse rash throughout her torso and extremities for the past 6 months. She described her abdominal pain as diffuse, nonspecific, without bleeding or change in bowel habits. Exam showed left sided weakness and petechial and purpuric rash diffusely. Lab work revealed kidney injury with a creatinine of 3.7. Blood counts including platelets and coagulation studies were normal; urinalysis revealed erythrocytes, leukocytes, trace protein without leukocyte esterase, nitrites or casts. Rheumatologic studies including Anti Nuclear Cytoplasmic Antibody, Cryoglobulin, Rheumatoid Factor, Anti Nuclear Antibody, protein electrophoresis and complement level were within normal limits. Erythrocyte Sedimentation rate (ESR) and C-Reactive Protein (CRP) were noted to be elevated. Infectious work up including HIV and viral hepatitis was negative. CT of the

abdomen showed diffuse colitis and stool study was negative for infection.

Rheumatology recommended a skin biopsy and initiating the patient on high dose IV steroids in the context of negative scans, diffuse petechial rash and elevated ESR and CRP. Skin biopsy subsequently revealed leukocytoclastic vasculitis in small vessels, predominantly venules, with IgA deposition consistent with IgA vasculitis. Over the next few days, she showed significant clinical improvement with resolving hemiparesis, improving pain and clearing of her rash. She was discharged on tapering dose steroids and is currently following up with rheumatology.

IgA vasculitis or Henoch-Schönlein purpura is a common systemic vasculitis with up to 90% cases found in children with typical tetrad of arthritis/arthralgia, abdominal pain, renal disease and palpable purpura. This was an atypical presentation with strong neurological involvement in an adult, not attributable to any atherosclerotic disease.

77 A RARE CASE OF IGG4 NEPHRITIS

J Maldonado*, DS Rees, M Touchy, MV Naljayyan, EA Aguilar, S Barry, F Yazdi, RB Varelzdis, SA Morse, E Reisin. *LSUHSC-NO, New Orleans, LA*

10.1136/jim-2018-000974.76

Case A 42 yo. black male with a past medical history of asthma and sarcoid presented to clinic with chest/abdominal pain. He was having diarrhea 10–15 times daily for the last few months. He reported dyspnea. He had nightly fevers with sweats, intermittent purpuric rash on his thighs and forearms. On exam showed palpable lymph nodes in his neck and abdominal pain. Labs results: FeNa <1%, urine protein/creatinine 1900 mg/g, c3 50 mg/d, c4 8 mg/dL, creatinine 3.32 mg/dL, troponin 0.02 ng/ml. No dysplasia or malignancy on colonoscopy/endoscopy. Fat pad biopsy with Congo red stain was negative for amyloid. Right cervical lymph node biopsy showed reactive lymphoid hyperplasia with polytypic plasmacytosis and increased IgG4-positive plasma cells. Chest CT showed interval development of small bilateral pleural effusions with associated subsegmental atelectatic changes and enlarged bilateral hilar lymph nodes. Abdomen CT showed questionable thickening of the sigmoid colon and mild thickening of the terminal ileum. Kidney biopsy showed diffuse interstitial fibrosis and plasma cell rich interstitial inflammatory infiltrate of which the majority of the plasma cells were IgG4 positive consistent with IgG4 related interstitial nephritis. The patient was treated with prednisone and Rituximab and weaned off daily prednisone over the next 3 months. One month later, creatinine decreased to 1.42 mg/dL, GFR increased from 24 to 70 ml/min, urine protein/creatinine ratio decreased to 140 mg/g, and complements levels normalized.

Conclusion The patient presented with an acute kidney injury with a significant, nephrotic range proteinuria. Biopsy showed an IgG4 related interstitial nephritis that responded well to treatment with prednisone and rituximab. This is a rare case of a glomerulonephritis had typically been described in individual cases and case series without large scale studies regarding optimal therapy. Physicians should be aware of this as

cause of interstitial nephritis for appropriate diagnosis and treatment.

78

A RARE OCCURRENCE OF BEHCET'S SYNDROME: A CASE REPORT

J Watts*. *LSUHSC School of Medicine, New Orleans, LA*

10.1136/jim-2018-000974.77

Case report We present the case of a 28-year-old African American man who presented to the UMC Clinic with myalgia and skin lesions. He previously presented to Tulane ophthalmology with bilateral anterior uveitis 1 year prior. His symptoms initially started 2 years ago with myalgias on his forearms and legs, which left hyperpigmented spots. Due to the muscle pain in his physically demanding occupation, the patient needed to change to a less intense job. The patient had painful skin ulcers in his mouth and on his genitals. A significant improvement and rapid resolution of the ulcers after taking colchicine led to the strong indication of Behcet's disease. A biopsy of the patient's recurrent genital ulcers demonstrated an acanthotic epidermis and a dermis with a dense infiltrate of lymphocytes and neutrophils. All of these findings were consistent with Behcet's disease in our patient.

Behcet's syndrome is a rare multi-system disease found in certain ethnic groups and in geographic regions. More commonly found along the historical Silk Road in the Mediterranean and western Asia extending east to Japan, Behcet's prevalence ranges from 80 to 370 cases per 1 00 000 in Turkey and 13.5 to 20 cases per 1 00 000 in Japan and China to 1 in 3 00 000 in the United States and United Kingdom.

The occurrence of Behcet's syndrome is uncommon in the United States and in African Americans. The initial findings of anterior uveitis 2 years prior was found as an isolated inflammation. Even still, the painful ulcers led to a differential diagnosis of polyarteritis nodosum, erythema nodosum, and even rare tumid lupus erythematosus, but not Behcet's disease. We present the rare occurrence of the disease to highlight the possible missed diagnosis of the disease in a non-typical Behcet's patient.

Cardiovascular

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

79

MANAGEMENT OF HYPERTENSION (HTN) IN AN ACADEMIC FAMILY PRACTICE SETTING

^{1,2}EA Aguilar*, ¹S Barry, ²A Acharya, ²B Arun, ²S Majeed, ²M Nguyen, ²J Oschwald, ²F Perrin, ²M Usman, ²J Campbell, ²M Coleman, ²E James, ²T Carlson, ²J Hudson.
¹LSUHSC, New Orleans, LA; ²LSU, Kenner, LA

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Purpose of study HTN is a risk factor for CVA and stroke. Target BP are SBP $\leq$ 140 mmHg and DBP $\leq$ 90 mmHg for general

Abstract 79 Table 2 Distribution of drugs among the controlled and non-controlled

Meds	Controlled		Non-controlled	
	n	%	n	%
ACEi	30	32	16	23
ARBs	8	9	7	10
Beta Blockers	18	19	12	17
Diuretic	45	48	45	63
Ca channel blockers	28	30	26	37

Abstract 79 Table 1 Number of patients and mean blood pressure in each group

	All	Controlled	Non-controlled
N	164	93	71
SBP	138 $\pm$ 15	128 $\pm$ 8	152 $\pm$ 11
DBP	84 $\pm$ 10	79 $\pm$ 9	89 $\pm$ 8
Heart Rate	83 $\pm$ 14	82 $\pm$ 14	84 $\pm$ 15

population without DM. The SPRINT trial showed that patients at high risk for CVA, without DM, SBP <120 mm Hg, as compared to SBP <140 mm Hg, resulted in lower rates of CVAs and death from any cause. Treatment means a 20% to 25% reduction in CVA. The purpose was to identify patients diagnosed with HTN and their pharmacological treatment.

Methods used After obtaining IRB approval, a cross-sectional retrospective chart review of 164 adults diagnosed with hypertension and treated at LSU Family Medicine was divided into controlled SBP <140 mmHg (n=93) and non-controlled SBP $\geq$ 140 mmHg (n=71) groups.

Summary of results The mean BP was SBP=138 $\pm$ 15 DBP=84 $\pm$ 10, with the controlled group SBP=128 $\pm$ 8 DBP=79 $\pm$ 9 and the non-controlled group SBP=152 $\pm$ 11 DBP=89 $\pm$ 8 (p<0.0001). 32% of controlled were treated with ACEi and only 23% of the non-controlled. Other medications were used to a lesser extent. Charts 1&2.

Conclusions Groups were treated per guidelines and did not differ significantly in age, comorbidities, laboratory values, BMI, height, and other demographic factors. ACE inhibitors were used in a greater percentage of controlled vs. uncontrolled patients. No or only a very small differences in frequency were noted for other drugs prescribed. Many questions are unanswered, including patient compliance, which may play a role in the percentage of controlled vs uncontrolled.

80

OUTCOMES OF PERCUTANEOUS ANTEGRADE INTRALUMINAL CORONARY INTERVENTION OF CHRONIC TOTAL OCCLUSION WITH REMOTE SURGICAL BACK-UP

O Akinseye*, C Nwagbara, S Haji, S Koshy, UN Ibebuogu, RN Khouzam, N Garg. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.79

Purpose of study Percutaneous coronary intervention (PCI) of chronic total occlusion (CTO) is not favored in facilities without on-site surgical back-up (Class III). We reviewed the

outcomes of patients who had CTO intervention with remote surgical back up in our institution.

Methods used All patients who underwent attempted antegrade intraluminal PCI of a native coronary artery CTO from January 2013 to July 2017 were analyzed.

Summary of results A total of 20 cases (18 patients, 58.1 ± 7.0 years, 70% males) were identified. Mean amount of contrast and fluoroscopy time was 306.7 ± 125.7 cc and 46.1 ± 21.1 min respectively. J-CTO score was 2.33 ± 1.2 in unsuccessful versus 1.53 ± 1.2 in successful cases. Procedure was successful in 85% (17/20). Total stent length was 43.4 ± 20.9 mm and the mean stent diameter was 3.1 ± 0.4 mm, with 82% requiring only 2 stents or less. Mean guidewire used per case was 2.5 ± 1.1 . TIMI 3 flow was achieved in 16/20 (80%) cases. There were 2 non-flow limiting coronary dissection and 1 wire perforation with no adverse outcome. Two patients had post-PCI myocardial infarction (MI). At 30 days, no reported adverse event post-procedure. At a mean follow up of 19.5 ± 13.7 months there was no cardiac death, MI, target vessel revascularization or stroke. There were 4 re-hospitalizations (20%) for angina requiring repeat coronary angiography in 3 cases – 2 without intervention, and 1 referred for CABG.

Conclusions Careful attempt at antegrade intraluminal CTO intervention done at a center without a surgical back up is feasible in selected patients, and with a reasonable safety profile and success rate.

81 LEFT ATRIAL INTRAMYOCARDIAL HEMATOMA FROM BLUNT TRAUMA MIMICKING LEFT ATRIAL MYXOMA

O Akinseye*, M Nayyar, MK Teshome, T Fan. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.80

Introduction Intramyocardial myocardial hematoma are rarely clinically apparent after a blunt chest trauma. We report a 27 year old male Afghanistan Veteran who had been blown up by an improvised explosive device six years prior to presentation with chest pain, dyspnea and syncope.

Case report A 27 year old Caucasian male with history of obstructive sleep apnea presented with chest pain and dyspnea on exertion for about a year but with recent progression. He also reported a recent episode of syncope while driving. He gave a history of blunt trauma from explosion of an improvised bomb six years ago while serving in Afghanistan. Laboratory investigation showed minimally elevated troponin of 0.97 ng/mL but electrocardiogram was only remarkable for nonspecific ST segment and t wave changes. Patient was started on dual antiplatelet therapy and heparin infusion for non-ST elevation myocardial infarction. Cardiac catheterization revealed normal coronaries. Trans-thoracic echocardiogram revealed a large left atrial (LA) mass measuring 4.2×6.1 cm consistent with atrial myxoma that fills $\frac{3}{4}$ of the LA and obstructs flow through the mitral valve. Cardiothoracic surgery was consulted and LA mass excision was planned. Exploration of the LA cavity revealed two masses. The dominant mass was $6 \text{ cm} \times 6 \text{ cm}$ on the superior side of the right pulmonary veins. A second mass of about 4 cm diameter was on the inferior side of the right pulmonary veins. A posterior LA wall dissection was noted. An incision was made into the atrial endocardium and the mass was found to be an endocardial organized hematoma occupying a pseudo-aneurysm of the LA. The

hematoma was evacuated and a reconstruction of the LA wall dissection was performed. The large neck of the pseudo-aneurysm on the free wall was left open. The patient did well and was discharged to home on the seventh post-operative day. He completed 6 months of anticoagulation with warfarin and has continued to do well on follow up.

Discussion/Conclusion Ruptures of the LA can occur after blunt trauma. These patients usually present acutely with tamponade symptoms. A LA pseudo-aneurysm is a rare injury that should be considered in patients with a history of blunt trauma who present with chest pain.

82 AN EASILY MISSED BUT CRITICAL DIAGNOSIS; SPONTANEOUS CORONARY ARTERY DISSECTION

N Almaddah*, D Dishmon, K Najib, F Ashraf, RN Khouzam. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.81

Case report A 55-year-old African American female with past medical history of hypertension, presented with a severe sudden onset pressure like chest pain. A 12 lead electrocardiogram revealed >2 mm ST segment elevations in leads V2-V5. Emergency coronary angiography revealed a long segment of diffuse smooth narrowing of the mid left anterior coronary descending artery (LAD). Because of the classic angiographic appearance diagnosis of SCAD was made. However, in view of ongoing severe chest pain not responding to medical therapy, the decision was made to proceed with percutaneous coronary intervention (PCI). She had successful stenting of the LAD with TIMI 3 flow regained. The patient was discharged home with plan to get image screening for fibromuscular dysplasia (FMD).

Discussion First line therapy for majority of patients with SCAD is conservative management and in the hospital monitoring for 3–5 days. Observational data showed that SCAD lesions heal in about 70%–97% of patients. Systemic

Abstract 82 Figure 1

anticoagulation theoretically may worsen the intramural hemorrhage.

PCI can be harmful as there is a high chance of the wire to go in to the false lumen and further complicating the case therefore you should always have a surgical backup before attempting PCI in SCAD.

SCAD association with multifocal FMD has been well reported. The prevalence of concomitant FMD in extracoronary arteries range from 25%–86%. Image screening is recommended in all SCAD patients.

83 ELECTROPHYSIOLOGIC CHANGES PRODUCED BY INTRAVENOUS ENALAPRIL IN THE HUMAN HEART-IMPORTANCE IN HYPERTENSION AND ARRHYTHMIAS

PI Altieri*, R González, W De Mello, HL Banchs. *University of Puerto Rico, Medical Sciences Campus, San Juan, Puerto Rico*

10.1136/jim-2018-000974.82

Purpose of study Intravenous Enalapril (E.) improves left ventricular function and reduces arrhythmias. The purpose of this study is to discuss our data, how these changes occurs.

Methods used A group of patients who received intravenous E. was studied.

Summary of results The mechanisms involved in these changes are:

1. Reduces regional wall stress
2. Reduces neurohormonal activation after an acute ischemic episode.
3. Reduces left ventricular remodeling.
4. Improves wall motion index.
5. Increase intraventricular conduction velocity.
6. Improves diastolic dysfunction.

Conclusions All above mechanisms are based on improvement of junction gap function by an increase of coupling of myocytes and inhibiting the mitogenesis effect of angiotensin II in smooth muscle cells. All above mechanisms will be discussed with scientific data.

84 CAN THE GRACE SCORE PREDICT THE LONG TERM OUTCOME OF PATIENTS WITH DISEASE OF THE LEFT ANTERIOR DESCENDING CORONARY ARTERY?

JK Bissett*. *CAVHS/UAMS, Little Rock, AR*

10.1136/jim-2018-000974.83

Purpose of study Risk stratification in Patients with coronary disease facilitates selection of patients for diagnostic studies, treatment, and followup. The GRACE score contains predictors such as age, renal function, ECG changes and elevated biomarkers., and may identify patients with increased mortality. The purpose of this study was to examine the 10 year predictive ability of the Grace score in a study of patients with documented disease of the left anterior descending.

Methods used 52 patients without coronary bypass grafting, interventricular conduction defect, or paced rhythm were referred for evaluation of coronary artery disease. The GRACE score, TIMI risk score, and the Charlson comorbidity index were calculated and the patients were treated with medical therapy and revascularization if indicated. The survival

was then compared for each risk score by Kaplan-Meier estimations and Cox regression for all-cause mortality.

Summary of results

1. 16 patients (31% expired at a mean interval of 2946.06 ± 2025.99 days. 36 patients (69%) survived for with a mean duration of 3885.47 ± 1948.73 days
2. A GRACE score ≥ 100 predicted reduced survival by the Kaplan-Meier estimate with log rank $p=0.015$, and hazard ratio 2.40. Cox regression with GRACE score ≥ 100 also predicted long-term all-cause mortality $p=0.019$.
3. Kaplan-Meier survival comparison using TIMI scores of ≥ 5 and less than 5 were not significant by the log-rank test $p=0.49$, or Cox regression 0.49
4. Kaplan-Meier comparison using the comorbidity risk index based on ≥ 5 were not significant by the log-rank test $p=0.34$, or c\ Cox regression 0.47.
5. Adding the TMII and comorbidity sciores improved trhe correlation with survival over each test separately, but did not acheive statistical significance Kaplan_Meier $p=0.13$, Cox regression $p=0.14$ with this patient group.

Conclusions

1. The GRACE score using clinical information, ECG findings and laboratory data predicts all-cause mortality in patients with disease of the left anterior descending with a hazard ratio of 2.48 at long term followup to 3885 days.
2. The TIMI risk score and comorbidity index were not predictive of mortality in this patient group.
3. Additional studies are required to understand how these variables might be used to improve diagnosis and treatment.

85 HEART RATE RESPONSE TO REGADENOSON DURING MYOCARDIAL PERFUSION IMAGING IN PATIENTS WITH TYPE 2 MYOCARDIAL INFARCTION

CM Colon*, R Marshall, C Roth, A Farag, AE Iskandrian, FG Hage. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.84

Purpose of study Patients with type 2 myocardial infarction (T2MI), supply-demand mismatch, are at high risk of mortality and future cardiovascular events. We have previously demonstrated that a blunted heart rare response (HRR) to regadenoson is associated with increased risk of cardiovascular events in patients undergoing myocardial perfusion imaging (MPI) but has not been investigated in T2MI.

Abstract 85 Figure 1 Death/MI/Late CR

Methods used We retrospectively studied T2MI patients (troponin I levels >99th percentile) who underwent vasodilator gated SPECT MPI within 3 months of T2MI at a single tertiary care institution between January and December 2013. HRR was calculated as the percentage increase in heart rate and categorized as <10%, 10%–30%, and >30%. The primary outcome was a composite of death, myocardial infarction or coronary revascularization (CR). The secondary outcome was all-cause mortality.

Summary of results We studied 227 patients with T2MI (62 ±14 years, 57% men, peak troponin 0.2 ng/ml, 58% abnormal MPI). The median HRR was 28% [14–39]. The proportion of patients with a HRR <10%, 10%–30% and >30% were 16%, 46%, and 38%, respectively. During a median follow-up of 20 months, 66% experienced the primary outcome (39% death, 42% MI, 5% CR). A decreasing HRR was associated with increased risk of events (figure 1). Compared to patients with a normal HRR (>30%), those with a blunted HRR (<10%) were associated with a hazard ratio of 2.3 (95% CI 1.4 to 3.6, $p=0.001$) for the primary outcome and 2.5 (95% CI 1.4 to 4.4, $p=0.003$) for all-cause mortality.

Conclusions A blunted HRR to regadenoson is associated with increased risk of cardiovascular events and all-cause mortality in patients with T2MI. This novel prognostic marker provides important risk stratification in this high-risk cohort.

86 ATHEROSCLEROTIC RENAL ARTERY STENOSIS: WHERE ARE WE NOW? A REVIEW OF THE LITERATURE

KG Hesterberg*, MK Teshome, RN Khouzam. *UTHSC, Memphis, TN*

10.1136/jim-2018-000974.85

Purpose of study Atherosclerotic renal artery stenosis (ARAS) is a not uncommon finding in the elderly population, with increased prevalence in patients with coronary artery disease or peripheral vascular disease. There has been much debate about optimal management and which patients benefit from revascularization. Large trials were published; however, the results have been criticized for low sample sizes, lack of clinical end points and methodological issues. As such, we undertook a review of current literature to provide further guidance.

Methods used A literature search was conducted to identify relevant trials, meta-analyses and review articles.

Summary of results Randomized controlled trials (RCTs), have not shown a benefit of renal artery stenting. However, the studies had various inclusion and exclusion criteria, limiting their generalizability. Meta-analysis of available trials suggests there is a benefit of stent placement with ARAS, with decreased diastolic blood pressure and number of antihypertensive medications. Additionally, observational studies and case series suggest patients with malignant hypertension, flash pulmonary edema and stenosis improve with revascularization.

Conclusions Current RCTs results do not clarify what patient benefits from renal artery stenting. These studies are difficult to interpret. Trials are on-going to evaluate this topic further. At this time, a reasonable approach is to reserve revascularization for significant bilateral renal artery stenosis, in the setting of malignant hypertension, declining renal function or flash pulmonary edema.

87 PREDICTABLE DELIVERY OF ANTIPLATELET AGENTS AFTER STENT PLACEMENT IN PERCUTANEOUS CORONARY INTERVENTION

¹I Ifedili*, ²D Puperi, ¹UN Ibebuogu. ¹University of Tennessee Health Science Center, Memphis, TN; ²University of Texas at Austin, Austin, TX

10.1136/jim-2018-000974.86

Purpose of study Dual antiplatelet therapy (DAPT) is required after an acute coronary syndrome (ACS), especially if a percutaneous coronary interventions (PCI) is done with placement of a stent. Current guidelines depend completely on patient compliance to intake of these medications after hospital discharge, leading to disastrous consequences such as recurrent ACS or in stent thrombosis when patients are not able to consistently take these oral medications. We propose various designs of implantable devices which would ensure consistent therapeutic systemic antiplatelet agents levels independent of patient compliance and which can be modified if systemic antiplatelet therapy is not needed anymore. This is the first study on implanted systemic delivery of antiplatelet agents to the best of our knowledge.

Methods used A review of literature on devices for systemic drug delivery was done. We chose subdermal and subcutaneous implantation of device rather than other measures including the use of pumps because this will be entirely patient independent and can be easily removed if complications arise. We then looked at various antiplatelet drugs that can be incorporated into the devices to release a predictable steady state of active drug into the systemic circulation to perform its antiplatelet function. We put forward various antiplatelet drug eluting biopolymer designs and appropriate sites of implantation.

Summary of results The use of a peripherally implantable device that releases active antiplatelet agents at a constant level will reduce the rate of complications arising from subtherapeutic systemic dose of antiplatelet agents by ensuring consistent delivery even when patients oral intake is affected, or they forget to take their medications. Implantation in a medically accessible site also helps if there becomes a need for device to be removed due to complications such as significant bleeding.

Conclusions An implantable device will ensure therapeutic levels of antiplatelet agents leading to less occurrence of repeat ACS or in stent thrombosis even when patients in an outpatient environment cannot guarantee consistent oral intake of medications after ACS or coronary stent placement.

88 HOLE IN THE HEART: MANIFESTING AS PLATYPNEA ORTHODEXIA SYNDROME (POS)

P Poudel*. *Suny Upstate Medical University, Syracuse, NY*

10.1136/jim-2018-000974.87

Introduction Platypnea is a rare condition seen in patent foramen ovale (PFO), pulmonary arteriovenous (AV) malformation, hepatopulmonary syndrome. Here we report a case of platypnea which was mismanaged as heart failure but later on, it was found that he had platypnea-orthodeoxia syndrome (POS) which resolved with successful close of PFO.

Case A 63-year-old male was recently discharged after he was treated for heart failure. A week later, he started having shortness of breath again. Physical examination findings were

normal. There were no significant findings in laboratory markers. Chest Xray showed right-sided pleural effusion. Echocardiography (ECHO) showed normal ejection fraction with mild diastolic dysfunction. He was given diuretics given the history of diastolic heart failure. He was doing fine when his oxygen saturation started dropping again. CT angiography was negative for pulmonary embolism. On further evaluation, it was found that there was postural variation in the drop of saturation. Differential diagnosis of platypnea was sorted out. Pulmonary AV malformation mostly seen in cirrhosis can manifest as platypnea. Right upper quadrant ultrasound was unremarkable for cirrhosis. Echo with bubble studies showed atrial septal defect consistent with PFO. He underwent surgical repair of PFO and his postural variations of desaturations resolved. The patient was given a diagnosis of platypnea-orthodeoxia syndrome secondary to PFO.

Discussion POS is an uncommon condition of positional dyspnea and hypoxemia. Since Burchell et al described this syndrome half-century ago, no more than 50 cases have been reported in the literature. It is believed that the upright position stretches the shape of the interatrial communication which increases blood flow from right atrium to left atrium. Diuretics can worsen this condition because a decrease in preload is known to increase interatrial communication and subsequent shunt. The main modality of treatment is repair of PFO.

Conclusion Not all the cases with dyspnea and diastolic dysfunction have heart failure. We should be cautious in giving diuretics in patients with platypnea because it can worsen the symptom.

89 IS EARLY INVASIVE MANAGEMENT AS ST ELEVATION MYOCARDIAL INFARCTION WARRANTED IN DE WINTER'S SIGN? A PEAK INTO THE WIDOW-MAKER

J Raja*, RN Khouzam. UTHSC, Memphis, TN

10.1136/jim-2018-000974.88

Purpose of study This review aims to address the important question of mode and urgency of intervention, on detection of de Winter's sign.

Methods used A systematic review of published clinical research articles from PUBMED data source was performed. Manuscripts selected for review had inclusion criteria of clinical research studies, addressing concepts of de Winter's sign, cardiac catheterization, thrombolysis, and delayed treatment.

Summary of results The original article by de Winter describes this sign in 30 of 1532 patients with anterior myocardial infarction. It describes significant myocardial loss despite successful procedures. This cautions towards the importance of early recognition of the sign and prompt intervention to save muscle tissue. The retrospective analysis by Verouden et al. looked at 1890 patients; who underwent primary percutaneous coronary intervention (PCI) of the LAD, and found 35 with this pattern. The analysis found that, compared to patients with anterior myocardial infarction and ST-segment elevation, patients presenting with this sign were younger, commonly males, and frequently had hypercholesterolemia. Ever since the original article, case presentations around the world had been reported. Shergill et al. describe a case treated with streptokinase thrombolysis, contradictory to the standard of care in Non ST Elevation Myocardial Infarction

with resolution of EKG changes. The similarity in all these reports were the prompt recognition of the sign, early cardiac catheterization revealing complete or near-complete occlusion of the LAD, and successful resolution of the EKG changes after stenting.

Conclusions de Winter's sign, although rare (~2%), should be recognized, as it reveals underlying severe coronary artery pathology, frequently involving the LAD which is associated with a high rate of mortality, if not recognized promptly. This systematic review emphasizes awareness and strong consideration of early activation of the cath lab with PCI; which may yield better treatment outcomes. The evidence suggests that de Winter's sign, presenting with ST depression, should be treated as STEMI equivalent, with prompt recognition and early intervention.

90 DEVELOPMENT OF A SHORT-TERM WARFARIN CONTROL MEASURE AMONG PATIENTS WITH NON-VALVULAR ATRIAL FIBRILLATION

¹L Shi*, ¹S Liu, ²Y Zhao, ¹AK Irmpen. ¹Tulane University, New Orleans, LA; ²Xavier University of Louisiana, New Orleans, LA

10.1136/jim-2018-000974.89

Purpose of study Overall Time in therapeutic range (TTR) is considered as gold standard of long-term warfarin control for non-valvular atrial fibrillation (NVAF). However, TTR calculations are complex and not readily available in warfarin management. This study aimed to examine short-term measures of warfarin control and compare them with overall TTR in predicting clinical outcomes.

Methods used Using the US Veteran Affairs database (1/1/2005–12/31/2015), NVAF patients who newly initiated warfarin and were treated ≥ 12 months were selected. Short-term measures within 3/6 months from index date included percentage of international normalized ratio (INR) in therapeutic range (PINRR), number of INR tests (NINR). Clinical outcomes were stroke/systemic embolism (SE), major bleeding, and all-cause mortality. Receiver Operating Characteristics (ROC) curves and area under curve (AUC) for each proxy along with dichotomous cutoffs were estimated in predicting overall TTR <65%. Multivariate Cox proportional hazard models using alternative proxy measures and overall TTR <65% were performed to rank model fitting by Akaike information criterion (AIC).

Summary of results Among 63,816 NVAF patients (mean age (SD): 71.1 (9.6) years, 99% male) with median 3.1 follow-up years (IQR: 3.5), 56% of patients had an overall TTR <65%. PINRR and NINR within 3/6 months were moderately correlated to overall TTR <65%. Composite measures based on

Abstract 90 Table 1

	Stroke/SE	Major Bleeding	Total Mortality
Composite Measure3m [aHR (95% CI)]			
H3 vs. L3	2.57 (2.11–3.14)	2.15 (1.88–2.45)	1.18 (1.10–1.26)
M3 vs. L3	1.66 (1.38–1.98)	1.43 (1.27–1.60)	1.04 (0.99–1.09)
Composite Measure6m [aHR (95% CI)]			
H6 vs. L6	2.24 (1.85–2.72)	2.29 (2.01–2.62)	1.28 (1.20–1.36)
M6 vs. L6	1.47 (1.23–1.76)	1.52 (1.35–1.72)	1.04 (1.00–1.10)

NINR and PINRR cutoffs were used to define 3 month/6 month risk groups: high ($H_{3/6}$), moderate ($M_{3/6}$), and low ($L_{3/6}$). Associations of composite measures within 3/6 months with clinical outcomes are presented below.

Conclusions More than half of the population had low TTR control. A composite measure including number of INR and INR in therapeutic range at month 3 and 6 may predict clinical outcomes in patients receiving warfarin.

91 AORTICO-LEFT ATRIAL FISTULA – A RARE COMPLICATION OF INFECTIVE ENDOCARDITIS

A Tambe*, R Carhart, M Badrinath. *SUNY Upstate Medical University, Syracuse, NY*

10.1136/jim-2018-000974.90

Introduction Aorto-cavitary Fistula (ACF) is a rare complication in infective endocarditis (IE) causing rapid clinical deterioration due to right heart failure. We describe a 31 year old male with mitral valve IE which was later complicated by the development of an aortic-left atrium fistula causing cardiogenic shock and death.

Case A 31 year old male presented with fever, chills and weakness. His history included intravenous drug use and bioprosthetic mitral valve replacement 3 years ago. On presentation he was tachycardic, hypotensive and hypoxic. A systolic murmur in the mitral area was heard on exam. Pertinent laboratory findings were Lactic acid 2.9, creatinine 2.12, Troponin T 0.27. Electrocardiogram showed ST elevation in the inferolateral leads. Chest radiograph was unremarkable. Transesophageal echocardiogram (TEE) revealed left ventricular ejection fraction of 40%–45% and bioprosthetic mitral valve with a 2 cm vegetation (figure 1 – left). Oxacillin was started along with vasopressors and intravenous fluids for septic shock. After septic shock resolved, patient refused mitral valve surgery.

Four weeks later, patient developed acute respiratory failure secondary to pulmonary edema requiring intubation. Repeat ECHO showed severely impaired right ventricular systolic function. The vegetation was 0.8×1.4 cm in size and a fistula was seen between aortic root and left atrium (figure 1 – below).

Discussion ACF is a rare and serious complication of IE that occurs in almost 1.6% of cases. ACF occurs due to extensive local destruction of tissue. TEE and transthoracic echocardiography (TTE) can detect fistulas in 53% and 97% cases respectively. Fistula formation causes sudden hemodynamic deterioration from sudden right heart failure. Prevalence of heart failure with left-sided infective endocarditis ranges from

Abstract 91 Figure 1 Left – vegetation, Right – Fistula

19% to 44%, and is a class I recommendation for valvular surgery. Despite aggressive surgical measures, the mortality rate remains high.

92 WITHDRAWN

93 DEEP T WAVE INVERSIONS IN PRECORDIAL LEADS, NOT ALWAYS CARDIAC ISCHMIA

F Ashraf*, N Almaddah, K Najib, RN Khouzam. *UTHSC, Memphis, TN*

10.1136/jim-2018-000974.91

Case report A 28-year-old man with history of bradycardia, gastroparesis, dysphagia and a feeding tube with severe protein calorie malnutrition was brought in to the hospital from clinic due to bradycardia. He denied any chest pain, but did admit to being very tired. Heart rate on arrival was 42 beats/min, blood pressure was 96/50 mm HG, temperature could not be recorded. He was cachectic on exam, no major cardiac abnormalities were noted though.

Laboratory work up was remarkable for alkaline phosphatase 760 U/L, AST 475 U/L, ALT 636 U/L, GGT 310 U/L, troponin was negative. Electrocardiogram (EKG) showed massive T wave inversions in V1-V3, triggering concern for cardiac ischemia (Wellens sign). Cardiology was then consulted.

Considering patients lack of chest pain and three negative troponin, he was not treated for cardiac ischemia. A temperature was finally recorded and was very low at 34-degree C. The patient was then re-warmed, with improvement in t wave changes gradually. Echocardiogram was also done which showed ejection fraction of 55% and a trace pericardial effusion. Two days later, patient had a normal temperature, and T wave changes had resolved completely.

Discussion Hypothermia is known to cause a number of EKG changes, which include Osborne Waves, bradycardia, QT prolongation and ventricular ectopy. Deep T waves inversions however, are not classically associated with hypothermia. Infact, deep T wave inversions in precordial leads (Wellens sign) is highly specific for critical stenosis of the left anterior descending artery (LAD). Patients may be pain free by the time the EKG is taken and have normally or minimally elevated cardiac enzymes; however, they are at extremely high risk for extensive anterior wall MI within the next few days to weeks. This triggered an alert in the patient described above. However, hypothermia was the culprit in this situation, and rewarming resulted in resolution of changes.

A review of literature shows deep t waves inversions are extremely rare in hypothermia. We are able to find only 2 cases describing T wave inversions in hypothermia, and none of them had the magnitude to trigger concern for cardiac ischemia (Wellens sign). The case also highlights importance of co-relating EKG changes with your patients symptoms.

F Ashraf*, N Almaddah, TM Fan. *UTHSC, Memphis, TN*

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Case report 69 year old man with history of Heart failure with Reduced Ejection Fraction (HFrEF), atrial fibrillation (AF), chronic kidney disease came in with dyspnea on exertion. Patient had an echocardiogram done 2 months ago which showed left ventricular ejection fraction (LVEF) of 30%–35%, E/A: 0.86, E velocity of 97 cm/sec, A velocity 113.0 cm/sec, suggestive of grade 2 diastolic dysfunction (DD). A repeat echocardiogram was done now which revealed E/A ratio of 2.8, A velocity decreased to 43.0 cm/sec, suggestive of grade 3 DD. A review of the 2 echocardiograms revealed patient was in atrial fibrillation 2 months, ago while he was in normal sinus rhythm at the time of the current echocardiogram.

An echocardiogram was repeated in a weeks' time, which now showed E/A ratio of 1.7, A velocity was up to 86.8 cm/sec, indicative of grade 2 DD again. Patient was still in sinus rhythm at that time.

Discussion E/A ratio is key component of grading DD on echocardiogram. In the case above, conversion of atrial fibrillation to sinus rhythm resulted in a temporary increase in E/A ratio, most likely due to atrial stunning and hence decrease in A velocity. As the atrium recovered, in about 1 weeks' time, A velocity increased again, and E/A ratio dropped again to a level of grade 2 DD. This phenomenon if ignored, can result in patients being labelled incorrectly for DD after planned or spontaneous cardioversion for AF.

Abstract 94 Figure 1

MJ Clanahan*, F Ashraf, RN Khouzam. *The University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.93

Case report Malingering is a deliberate falsification or fabrication of psychiatric or physical ailments with ulterior motive for personal benefit. It shares commonalities with factitious disorder, but is not generally considered a psychiatric illness. Malingering causes massive strain on our health system,

including healthcare personnel time, insurance funding, and medical resources. This is prevalent in the field of Cardiology as it can be difficult to distinguish falsified from real chest pain in the presence of several risk factors. Herein, we report a particularly egregious case of a patient presenting with typical cardiac chest pain and was found to have two cardiac catheterizations in the previous two weeks at outside hospitals. A careful search of existing scheduled drug databases revealed underlying motive. Because the patient was readily willing to undergo several invasive cardiac catheterizations (in short sequence) for secondary gain, this suggests some overlap with factitious disorder. Furthermore, it is important to note the incredible burden of cost on the United States health system, which has been estimated to be as high as \$180 billion per year. This extraordinary case highlights the importance of thorough individual investigation and healthy skepticism in the context of symptoms and histories that pique our interest as health professionals.

NA Halapin*, WS Rohn, K Mentore, LS Engel, J Martinez. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.94

Case report A 65-year-old African American woman with history of atrial fibrillation, status post pulmonary vein isolation ablation (PVIA) fifteen years prior, presented with two days of intermittent palpitations, dyspnea, and diaphoresis. She had been on flecainide and apixaban for a number of years following her ablation procedure. Rhythm strips during an episode of palpitations revealed atrial flutter with RVR. She was treated with 5 mg IV metoprolol succinate and then started on 25 mg oral metoprolol succinate every six hours, which achieved rate control and enabled clearer analysis of the rhythm strip which demonstrated a clear saw tooth pattern of p-waves, indicating Atrial Flutter with 2:1 conduction. An electrophysiology study identified typical atrial flutter involving the cava tricuspid isthmus. Radiofrequency ablation (RFA) was used to interrupt the tract, resulting in restoration of sinus rhythm during the remainder of the patient's hospitalization and she was discharged on her flecainide and apixaban.

Discussion Atrial flutter is most often be the result of a macro-reentry current in the right atrium induced by another arrhythmia or may be induced by medications commonly used for rhythm control in atrial fibrillation. Atrial flutter may also be related to prior cardiac procedures or existing structural heart damage that can cause an atypical type of circuit in the right atrium or less frequently in the left atrium. Given our patient's history of ablation for atrial fibrillation 15 years prior to presentation, as well as chronic use of flecainide, there was a distinct possibility that she had an atypical circuit in the right or left atrium. This macro-reentry circuit can lead to a more regular conduction through the ventricles as compared to atrial fibrillation, which is why rate control is more difficult, and is not an optimal long-term treatment for atrial flutter. The definitive treatment of typical atrial flutter is ablation, which has a significantly higher rate of success when compared to atrial fibrillation. Ablation treatment of atrial flutter can lead to the onset of atrial fibrillation despite the history of PVIA.

97 MASSIVE LEFT VENTRICLE THROMBUS WITH THROMBOEMBOLIC PHENOMENON

M Haltom*, N Almaddah, I Ifedili, R Lam, RN Khouzam, R Askari, A Holden, UN Ibebuogu. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.95

Case report A 35-year-old African-American male with a history of alcohol dependence and cocaine use presented with severe abdominal pain. Computed tomography of the abdomen and pelvis with contrast found a large LV thrombus with bilateral renal infarcts. Heparin drip was initiated, and patient was admitted. Electrocardiogram demonstrated sinus bradycardia. A transthoracic echocardiogram (TTE) demonstrated a LV ejection fraction of 20%–25%, severe global hypokinesis and an LV thrombus measuring 7.1×2.7 cm. Patient underwent coronary angiogram, showing angiographically normal coronary arteries. Cardiothoracic Surgery was consulted for potential thrombectomy. Due to the large size of the thrombus, patient underwent aortotomy with thrombectomy.

Discussion Cardiac emboli are a rare but significant cause for abdominal pain in the emergency department. Management of LV thrombus formation in patients with reduced ejection fraction should include a multi-disciplinary team and shared decision making with the patient due to the high morbidity and mortality risk of all treatment options. Further research is needed to address management in patients with massive thrombi that have high risk of thromboembolic phenomenon if treated with only anticoagulation.

Abstract 97 Figure 1

98 TETRALOGY OF FALLOT – PUSHING THROUGH DESPITE LONG ODDS

F Han*, MR McMullan, WF Campbell. *University of Mississippi, Jackson, MS*

10.1136/jim-2018-000974.96

Case report We describe a 26 year old with Tetralogy of Fallot, valvular pulmonary stenosis, and infundibular stenosis who

was repaired with an RVOT reconstruction, VSD patch repair, and a pulmonary homograft. This patient is the first patient to our knowledge, who has survived acute free pulmonary insufficiency as an adult which was necessitated by an infected conduit. He experienced multiple recurrences of RVOT obstruction, as well as pulmonary valve insufficiency. As a result, he has required multiple palliative surgeries and transcatheter pulmonary valve replacements. He received biventricular ICD placement in 2011 for ventricular tachycardia. He also developed acute on chronic renal failure, with dialysis dependence, one year prior to presentation. This was thought to be related to a hypoperfusion injury related to his cardiomyopathy. He later on developed MRSA bacteremia, pulmonary abscesses, and endocarditis of his ICD and pulmonary valve. He was admitted for removal of his infected ICD leads, removal of his vegetation, excision of his entire conduit, and recovery on ECMO with free pulmonary insufficiency. He returned to the OR two days later for chest closure, ECMO decannulation, and placement of a subcutaneous ICD. He awaited sterilization of blood cultures prior to pulmonary valve replacement.

He was admitted for elective transcatheter pulmonary valve replacement. He coded multiple times due to VT and VF, both in the catheterization lab and post-admission. He had cardiogenic shock with ejection fractions as low as 5%, requiring high degrees of vasopressor support. He also needed CRRT. He experienced shock liver and severe pulmonary edema. Due to significant abdominal ascites, he was given a peritoneal dialysis catheter but, this became infected. After a two month hospitalization, the patient survived and at his most recent clinic visit, is recovering well from a cardiac standpoint, and has been following along with his baseline dialysis regimen. He has regained much of his original body weight and is able to do regularly scheduled physical therapy. This case overall serves to illustrate his remarkable recovery through severe pulmonary valve insufficiency and later successful replacement.

99 DOG RELATED TRAGEDY TRIGGERING BROKEN HEART SYNDROME

M Ibrahim*, M Jbara, V Ramu. *ETSU, Johnson City, TN*

10.1136/jim-2018-000974.97

Case report A 91-year-old female patient presented to our hospital with what was initially thought to be a heart attack from coronary artery disease, but turned out to be a severe cardiac sequel with broken heart syndrome (Takatsubo cardiomyopathy) due to bereavement after her pet's death. She presented with clinical features suggestive of anterior ST-elevation myocardial infarction, but heart catheterization showed no angiographic evidence of obstructive coronary artery disease (>50% luminal stenosis) or acute plaque rupture. Left ventriculogram showed significant apical hypokinesis which is the pathognomonic finding for broken heart syndrome. Emotional stress such as a relative's death or financial or health catastrophe have been reported to result in broken heart syndrome, but to have it after a companion pet death is a rare occasion we wanted to report.

Introduction Takatsubo cardiomyopathy represents 1 to 2 percent of acute coronary syndrome cases, it is uncommon syndrome with a presentation that mimics acute myocardial

infarction. It presents with chest pain. EKG can show ST-elevation, diffuse T-wave inversion or ventricular arrhythmias. Diagnosis of Takatsubo syndrome depends on these criteria:

1. Transient apical and/or midventricular left ventricular wall motion abnormalities,
2. Absence of obstructive epicardial CAD or angiographic evidence of acute plaque rupture,
3. New EKG abnormalities such as ST-segment elevation and/or diffuse T-wave inversions.

Case presentation A 91-year-old Caucasian woman with hypertension, CAD presented to the emergency with severe chest pain that radiated to the left shoulder, associated with nausea, vomiting and lower limb weakness. A few days prior to the emergency department visit her beloved puppy was brutally attacked by two larger dogs while she witnessed the whole event. Vital signs showed heart rate of 75 beats per minutes; blood pressure 142/66 with SpO₂ of 90% on non-rebreather mask. Chest examination showed normal cardiac sounds, without murmurs. No peripheral edema or cyanosis. The EKG showed ST segment elevation in the anterior chest leads. Troponins were elevated to 0.49. Left heart ventriculogram showed the presence of apical hypokinesis consistent with Takotsubo cardiomyopathy and an ejection fraction of 30%–35%.

100

MANAGEMENT DILEMMA FOR A PATIENT IN NEED OF CORONARY ARTERY BYPASS GRAFT AND HEPATIC TRANSPLANT WHERE ONE INDICATION MAKES THE OTHER TREATMENT A HIGH RISK SURGERY

I Ifedili*, S Salem, EA Christian, R Askari, RN Khouzam. *University of Tennessee Health Science Center, Memphis, TN*

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Case report A 62 year old African American male with history of coronary artery disease (CAD) with severe 3 vessel disease with in-stent restenosis of his proximal left anterior descending artery (LAD), mid left circumflex artery (LCx), and severe diffuse disease of right coronary artery, decompensated chronic liver disease (DCLD) from hepatitis c cirrhosis (status post curative hepatitis treatment) with ascites, splenomegaly, and thrombocytopenia, history of cerebrovascular accident, hypertension, chronic non occlusive portal thrombosis, presented with nausea, vomiting, chest pain and elevated troponin from known DCLD and CAD. Patient with recent multiple admissions for hepatic and/or cardiac complications and could not undergo coronary artery bypass graft (CABG) due to high perioperative mortality from DCLD at same time could not undergo liver transplantation due to significant CAD. Initial option was to refer him for non-intervention measures with a dim prognosis, but patient was upbeat with wanted all measures to be done to improve his independence. A multidisciplinary team decision was taken for patient to undergo percutaneous coronary intervention with placement of bare-metal stent to his proximal LAD and mid LCx. He was optimized and had procedure without complications with plan for liver transplant followed by CABG and close follow-up to modify his antiplatelet regimen

Abstract 100 Figure 1

when his platelet count returns to a value of less than 50 000.

101

HIGH-OUTPUT HEART FAILURE SECONDARY TO UNCORRECTED POPLITEAL ARTERIOVENOUS FISTULA

PS Jagadish*, QM Mirza, PR Higgins, TM Fan, KB Ramanathan. *University of Tennessee Health Science Center, Memphis, TN*

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Background High-output heart failure is characterized by elevated cardiac output and low systemic vascular resistance (SVR). Common etiologies include liver disease, obesity and, arteriovenous fistulas (AVF). This case describes a patient who developed severe pulmonary hypertension and high-output heart failure following traumatic AVF.

Case A 63 year-old male with hepatitis C, cirrhosis, and chronic atrial fibrillation presents to the pulmonary clinic for worsening exertional dyspnea. Vitals are normal. He has bilateral lower extremity edema (left >right). Chest x-ray shows cardiomegaly with clear lung fields. LVEF on echocardiogram is 55%–60% with severe right atrial and ventricular dilation and paradoxical septal motion. Estimated RV systolic pressure is 60 mmHg. Lower extremity venous Doppler ultrasound shows an AVF between the left popliteal artery and vein. Confirmatory CT angiogram reveals dilation of the left iliac, femoral, and popliteal veins (figure 1). The patient recalls a nail gun injury to his left leg 7 years ago.

On right heart catheterization, pulmonary artery pressure is elevated at 71/22 mmHg (mean 44 mmHg), pulmonary vascular resistance is 1.8 Wood units, and SVR is low at 453 dyn $\times$ s/cm⁵. Cardiac output and index are high, at 11.7 L/min and 4.84 L/min/m², respectively.

After interdisciplinary discussion between the Pulmonary and Cardiology services, the patient was referred to vascular surgery. He underwent successful ligation of the popliteal AVF with immediate improvement in dyspnea. At 3 month follow-up, both his exercise tolerance and lower extremity swelling had dramatically improved. Six-month echocardiographic follow-up is pending.

Conclusion Traumatic AVF is an often overlooked cause of pulmonary hypertension, especially in the presence of high-output heart failure.

Abstract 101 Figure 1

102 ASYMPTOMATIC CELIAC ARTERY ANEURYSM IN A PATIENT WITH CONGENITAL BICUSPID AORTIC VALVE

¹V Kohli*, ²D Das. ¹East Tennessee State University, Johnson City, TN; ²Quillen College of Medicine, ETSU, Johnson City, TN

10.1136/jim-2018-000974.100

Case report The incidence of celiac artery aneurysm (CAA) is 0.005%–0.01%. Only around 200 cases have been reported since 1745. Quite often the diagnosis is made after the rupture of an aneurysm, which has high mortality. We report a clinical presentation of asymptomatic CAA with a history of a congenital bicuspid aortic valve (BAV).

A 67-year-old female presented for routine outpatient follow up. She denied any complaints of abdominal and back pain, hematochezia or melena. There was no history of smoking or drinking alcohol. Family history was negative for connective tissue disorder or vascular aneurysms. Past medical history included hypertension, congenital BAV complicated with ascending aortic arch aneurysm status post ascending and transverse aortic graft without aortic valve replacement done 7 years back. An incidental finding of CAA with the largest dimension measuring at 9 mm was found that time. Her BP was 155/79, HR 69, RR 12, temp 98.0 and BMI was 28.66. Physical examination was remarkable for loud S2, grade 2/6 systolic murmur, regular rate, and rhythm. Carotid bruits were absent.

Most recently her CTA chest again showed CAA with an increase in the size measuring at 15 mm. Currently, no intervention was planned as the size of the CAA is small. The patient remains hypertensive as she is non-complaint to her anti-hypertensive medication atenolol.

The important etiology for CAA includes infection, congenital causes, atherosclerosis, fibromuscular dysplasia, and polyarteritis nodosa. In our patient, it is likely from either atherosclerosis due to her age and hypertension. But, since she has a congenital BAV, a possible association can be considered.

Due to the lower incidence of CAA, the diagnosis is difficult to make. The less familiar diagnosis is also a challenge for further management. Current guidelines suggest medical management for an aneurysm that is <20 mm as we see in our patient. CAA is usually seen in association with abdominal aneurysms or with splanchnic artery aneurysms at 18% and 50%, respectively. The association with ascending aortic aneurysm is usually rare and with a congenital bicuspid aortic valve is never reported.

103 HYPERTROPHIC CARDIOMYOPATHY: IMPORTANCE OF CONTRASTED ECHOCARDIOGRAPHY FOR LEFT VENTRICULAR APICAL ANEURYSM AND THROMBUS

JO Kolawole*. University of Mississippi Medical Center, Jackson, MS

10.1136/jim-2018-000974.101

Introduction Left ventricular apical aneurysms in the setting of hypertrophic cardiomyopathy (HCM) has been noted to confer a 3-fold increase in HCM related events and a 2-fold increase in thromboembolic events. Identifying this subset early with an appropriate imaging modality may prevent HCM related events.

Case description 73-year-old African American male with HCM diagnosed at age 59 presented to an outside facility for chest pain. He had an implantable cardioverter defibrillator placed for episodes of non-sustained ventricular tachycardia at the outside facility and was placed on medical therapy. The patient was also referred as an outpatient for alcohol septal ablation therapy. His past medical history includes type 2 diabetes mellitus. He was a former 50-pack-year cigarette smoker with no known family history of HCM or sudden cardiac death. Examination at clinic follow up was significant for grade III/VI systolic murmur with atrial sensed ventricularly paced rhythm on electrocardiogram. Non-contrast echocardiogram showed severe concentric left ventricular hypertrophy with asymmetrically hypertrophied antero-septum and evidence of LVOT obstruction, peak gradient of 63 mmHg and overall normal left systolic function. Septal reduction therapy was not indicated at the time given his diffuse nature of severe hypertrophy and his good functional capacity with no obvious symptoms. He had regular clinic follow up with general cardiology and electrophysiology and continued to do well. However, he subsequently presented to the clinic after a hospital admission for transient unilateral weakness and vision loss concerning for stroke and had negative brain imaging. Contrast echocardiogram obtained at that visit showed apical thinning and an apical left ventricular (LV) thrombus. He was subsequently anticoagulated without further neurological sequela. There was thrombus resolution at subsequent imaging.

Conclusion A high index of suspicion is needed for detection of LV apical aneurysm and apical thrombus in HCM. Contrast echocardiography or cardiac magnetic resonance should be considered as complications may be preventable with appropriate therapies.

104 WELLENS SYNDROME: A CASE REPORT

AE Martinez*, C Canha, J Ruiz. University of Florida Health Jacksonville, Jacksonville, FL

10.1136/jim-2018-000974.102

Case report A 45 year-old male with a past medical history of hypertension, tobacco, and cocaine abuse presented to the hospital for new onset chest pain of 2 days duration. The pain was described as intermittent substernal pressure, 7 out of 10 in severity, nonradiating, and nonexertional. The episodes would last approximately 5 min. No associated symptoms. The patient denied prior episodes of chest pain. Admitted to using cocaine one day prior to presentation.

No abnormal physical exam findings. Vital signs: blood pressure 150/98 mmHg and heart rate 67 bpm. Given the patient's clinical presentation, serial cardiac enzymes and electrocardiogram (ECG) were obtained. Cardiac enzymes were negative and the ECG revealed normal sinus rhythm, normal axis, biphasic T-waves in leads V1 to V3, and a deeply inverted T wave in V4. This pattern was recognized as Wellens Syndrome and the patient was taken for cardiac catheterization. He was found to have severe stenosis of the proximal left anterior descending artery (LAD) and one drug-eluting stent was deployed. The patient was discharged with atorvastatin, aspirin, and prasugrel with no complications.

Discussion Wellens syndrome was first described by Dr. Wellens, a Dutch Cardiologist, in the in the early 1980's through the observations of patients with unstable angina in which they noted that up to 75% of the patients with Wellens sign would go on to develop anterior wall myocardial infarction's (MI) within weeks without treatment. Wellens syndrome is an evolving waveform noticeable through ECG showing deeply inverted or biphasic T waves in leads V2-V3 which indicates critical proximal LAD artery stenosis. Definitive treatment typically involves percutaneous coronary intervention to relieve the occlusion.

Conclusion Wellens syndrome is becoming an increasingly prominent finding in patients with acute coronary syndrome. Physicians may be at risk of overlooking this critical finding due to lack of positive cardiac enzymes, commonly painless presentation, and lack of the prototypical ST segment elevation seen in acute myocardial infarction. This ECG pattern serves as a warning sign for critical stenosis of the LAD and gives physicians an indicator for possible impending acute myocardial ischemia.

105

DRUG-RELATED COMPLETE ATRIOVENTRICULAR BLOCKADE CAUSED BY DILTIAZEM

¹R McPeters*, ¹TV Joshi, ^{1,2}M Mathai, ¹LS Engel, ¹J Martinez. ¹LSU Health Sciences Center, New Orleans, LA; ²University of Medicine and Health Sciences-St Kitts, Basseterre, Saint Kitts and Nevis

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Introduction Calcium channel blockers (CCB) have been linked to atrioventricular (AV) block. AV block is a delay or interruption in transmission of impulse from the atria to the ventricles due to impairment in the conduction system. Diltiazem, a non-dihydropyridine CCB commonly used to treat various cardiac conditions, is associated with various types of heart block. We present a patient with symptomatic bradycardia with two separate types of AV block secondary to Diltiazem therapy.

Case An 80 y/o man with type 2 diabetes and hypertension on diltiazem presented with exertional dyspnea and chest discomfort. Labs were significant for Hgb of 12.7 g/dl and creatinine of 1.5 mg/dl. Troponin, BNP, and chest x-ray were unremarkable. Initial EKG showed 1st degree AV heart block with T-wave inversions and a ventricular rate of 61 BPM. EKG taken 1 hour later showed AV dissociation, 3rd degree complete heart block, and a ventricular rate of 44 BPM. A temporary venous pacer was placed. The patient returned to NSR with normal heart rate and his pacer was

removed. He was discharged and advised to follow up with cardiology for placement of a loop-recorder. During the patient's follow up, he remained in NSR and 1st degree AV block. Diltiazem was the causative agent for his complete heart block.

Discussion Diltiazem toxicity can result in heart block in patients on chronic therapy, concomitant chronic renal failure (CRF), elderly patients with CRF due to higher elimination half-lives, electrolyte disturbances and with underlying AV conduction disease. 56% of patients whose AV block resolved after discontinuing the offending drug experienced recurrence. True drug-induced AV block occurs in only 15% of patients who have 2nd/3rd-degree AV block. Beta-blockers and digoxin are more likely to cause AV block than Diltiazem. Our patient's advanced age and underlying conduction abnormality led to diltiazem toxicity resulting in a complete heart block. There are no specific treatment guidelines for heart block induced by AV nodal blockers. Transvenous pacing, removal of the causative drug, supportive care and identification of the underlying conduction disease are mainstays of therapy.

106

IMMUNOSUPPRESSION OF THE RETICULOCYTES

FP Mercado Cabrera*. University of Puerto Rico, San Juan, Puerto Rico

10.1136/jim-2018-000974.104

Background Precise diagnosis of the cause of anemia requires a multimodality approach and has important implications for treatment in patients with unclear etiology.

Case 52 years old female with arterial hypertension, type 2 diabetes mellitus, non-ischemic cardiomyopathy status post uncomplicated orthotopic heart transplant 4 months ago who developed progressive shortness of breath for about a week from mild limitation of daily activities to shortness of breath at rest.

Found with normocytic normochromic anemia at 5 g/dL at the emergency room. Physical exam and anemia workup within normal limits. Medications included tacrolimus, mycophenolate, cyclosporine, prednisone, trimethoprim/sulfamethoxazole (TMP/SMX), valganciclovir, diltiazem, pantoprazole, atorvastatin, insulin, and multivitamins. Bone marrow biopsy showed pure red blood cell aplasia of unknown etiology. Hematologic and infectious evaluation prior to transplant was unremarkable and was repeated once anemia was found with same results. It was determined that mycophenolate had the highest myelosuppression potential from all the possible causes and it was discontinued. Patient was transfused packed red blood cells and with improvement of anemia to 10 g/dL, sustained with the aid of supplemental iron only. Follow up cardiac biopsy at 10 days was normal (1R) with no signs of graft disease present.

Decision making Differential diagnosis of acquired red blood cell aplasia is broad and it is difficult to determine the definitive culprit without removing possible offending agents. It is of paramount importance to take a systematic approach discontinuing first the most likely culprit which would also not require major changes in patient's quality of life. TMP/SMX was also high in the differential diagnosis but changing antibiotic prophylaxis will require a burdensome regimen that

requires frequent administration. Fortunately, we had an adequate response with the discontinuation of the mycophenolate and follow up biopsy was normal.

Conclusion Post transplant pure red blood cell aplasia is rare and a cause of great concern specially since complete bone marrow suppression can occur. Determining the most likely cause is usually challenging and requires a systematic multimodal approach.

107 A CASE OF A KILLER CORONARY VASOSPASM

K Najib*, KG Hesterberg, N Almaddah, MK Teshome, RN Khouzam. *UTHSC Memphis, Memphis, TN*

10.1136/jim-2018-000974.105

Case report A 52-year-old female with history of hypertension was admitted after ventricular fibrillation arrest. Urgent cardiac catheterization revealed severe diffuse spasm that resolved with intracoronary nitroglycerin and peripheral bicarbonate for significant acidosis. Initial laboratory studies revealed hypokalemia, lactic and metabolic acidosis, hypomagnesemia, and elevated troponin of 2.5 ng/mL. Hospital course was remarkable for ventricular tachyarrhythmia. Lidocaine and amiodarone infusions were started followed by norepinephrine and phenylephrine infusions for shock. Repeat cardiac catheterization showed severe diffuse vasospastic angina relieved with intracoronary nitroglycerin. Left ventriculogram showed ejection fraction of 25% to 30% and distal anterior, inferior and apical akinesis. Impella ventricular support device was placed. The patient's condition deteriorated into multiorgan failure and increased pressure support requirements, unfortunately leading to her demise.

Discussion Coronary artery vasospasm manifests as angina, myocardial infarction, ventricular tachyarrhythmia or sudden cardiac death. This case represents a challenging management dilemma as the patient experienced ventricular tachyarrhythmia secondary to diffuse coronary vasospasm and was unable to tolerate first-line therapy with calcium channel blockers or nitrates. She required pressor support that increased sympathetic surge, promoting resistance to antiarrhythmic therapy and exacerbating coronary vasospasm. Other management strategies to consider include procainamide, which can slow ventricular tachyarrhythmias even if it fails to terminate them. General anesthesia can reduce the sympathetic surge. Veno-arterial extracorporeal

Abstract 107 Figure 1 Pre and post intracoronary nitroglycerin

membrane oxygenation provides acute support in the setting of cardiogenic shock and arrest.

108 THROMBUS IN TRANSIT WITH MYXOMA PHYSIOLOGY

A Nanda*, J Raja, RN Khouzam. *University of Tennessee, Memphis, TN*

10.1136/jim-2018-000974.106

Case report 73 year old male presented with syncope. Trans-thoracic and transesophageal echocardiogram (TEE) showed an echodensity in the right atrium (figure 1 B, E), crossing over to the LA (figure 1 A, D) through a PFO, prolapsing across the mitral valve (MV) (figure 1 C, F), mimicking myxoma physiology. CT chest showed bilateral pulmonary emboli with right sided cardiac strain. Venous duplex showed right distal femoral vein and popliteal vein thrombus. Patient received heparin anticoagulation and was referred for thrombectomy. Preoperative TEE revealed complete resolution of the thrombus. Surgery was cancelled. He was treated with warfarin anticoagulation. PFO closure was not pursued. No systemic or pulmonary embolic events noted in follow up.

Discussion Right sided thrombus load with an undiagnosed patent foramen ovale (PFO) occurs more commonly than clinically detected. This is a unique case of thrombus in transit mimicking the 'ball in valve mechanism' typical of left atrial (LA) myxoma physiology, wherein paradoxical systemic embolism is prevented with therapeutic long term anticoagulation. There was no systemic embolization, with complete resolution on stringent therapeutic anticoagulation.

Conclusion This case highlights the risk of large burden VTE to the systemic circulation in the setting of undiagnosed PFO. It illustrates that despite the hemodynamic and end-organ risk, PFO closure is not indicated without a trial of persistently therapeutic anticoagulation.

Abstract 108 Figure 1

109

UTERINE INTRAVENOUS LEIOMYOMATOSIS WITH FEMORAL VEIN, INTRACAVAL, INTRACARDIAC AND PULMONARY ARTERY EXTENSION

M Nayyar*, O Akinseye, M Shahreyar, C Nwagbara, KT Weber. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.107

Case report A 54-year-old female patient with a recent diagnosis of cardiac thrombi and pulmonary embolism of unknown etiology (s/p failed thrombolytic therapy currently on edoxaban) presented with continued dyspnea and pleuritic chest pain. Presenting physical exam and laboratory data was unremarkable. Repeat transthoracic echocardiogram revealed a large right atrial (RA) thrombus, now with extension into the IVC. Upon cardiothoracic surgery consultation and opening of her RA, a single mass was found to be emanating from the IVC, through the right heart chambers and into the right pulmonary artery (PA). A right PA counter incision was made and an intact 38.2×2.8 cm mass was removed (figure 1). Pathology revealed a benign, vascular, smooth muscle mass with estrogen and progesterone receptor positivity favoring uterine intravenous leiomyomatosis (IL).

Abstract 109 Figure 1 Infiltration into IVC (red arrow), right atrium (yellow star) and ventricle (yellow arrow) and gross specimen (38.2 × 2.8 cm).

IL with intracardiac and pulmonary extension may be misdiagnosed because it is rare, with definite diagnosis requiring histopathological analysis. A high index of suspicion is required in middle-aged women with RA mass. Complete surgical removal is associated with no risk of recurrence. Several hormonal therapies such as aromatase inhibitors and GnRH may inhibit the growth of incompletely resected tumors.

110

BIATRIAL THROMBI IN THE SETTING OF ATRIAL FIBRILLATION

C Nwagbara*. *University of Tennessee health science center, Memphis, TN*

10.1136/jim-2018-000974.108

Case report History: An 84 year old man with paroxysmal atrial fibrillation (not on anticoagulation), end stage renal disease on peritoneal dialysis presented with dyspnea and confusion of a day duration.

On examination, he was tachycardic, hypotensive and disoriented.

Investigation: CT chest showed multiple filling defects in both atria and the left atrial appendage. Transthoracic echocardiogram did not show intracardiac thrombi. Transesophageal echocardiogram revealed large thrombi in the left atrium and left atrial appendage and a small thrombus in the right atrium.

Hospital course: The patient was intubated for acute respiratory failure and hemodynamic support was provided with vasopressors. Parenteral anticoagulation was instituted for intracardiac thrombi. A coagulopathy work up was performed with no positive findings. He remained encephalopathic. Brain imaging was unremarkable. His family decided to institute comfort measures only.

Discussion Atrial fibrillation predisposes to atrial thrombi due to ineffective atrial contraction. Thrombi most commonly found in the atrial appendage are likely due to lower velocity in comparison to the rest of the atrial cavity. Biatrial thromboses have also been reported in the setting of hypertrophic cardiomyopathy. It is unclear if the thrombi originate from the appendage and then migrate into the atrial cavity. This has not been studied in current literature.

Abstract 110 Figure 1

Transesophageal echocardiography is vital in diagnosing intraatrial thrombosis. The atrial thrombi were not seen on our patient's transthoracic echocardiography and the transesophageal study was prompted by filling defects seen CT chest. Adequate visualization of the atria should be performed in these patients prior to any procedures that can result in embolization, in the right clinical setting. Multimodality imaging should be employed when needed.

111

MULTIMODALITY IMAGING TO IDENTIFY ATRIAL MYXOMA

OB Obafemi*, JD Pollard, ME Hall. *University of Mississippi Medical Center, Jackson, MS*

10.1136/jim-2018-000974.109

Case description 61-year-old man history of type two diabetes mellitus, hypertension, hyperlipidemia and hyperthyroidism presented with a left thalamic stroke resulting in right sided

vision loss, dysarthria and left sided weakness. Patient was at home in his usual state of health, when he developed sudden right eye vision loss with an inability to speak. On evaluation at his local hospital, CT head w/o contrast significant for chronic lacunar infarcts. MRI brain was performed revealed an acute left thalamic infarct. For evaluation of a cardiac source of stroke, the patient had a TTE evident for a 2×2 cm mass in the left atrium. Patient was then transferred to our institution for further evaluation. TEE was ordered to better characterize the left atrial mass, which showed a left atrial 2.7×1.5 cm mass attached to the atrial septum that appeared more gelatinous and was suggestive of a left atrial myxoma. However, thrombus and other cardiac tumor could not be excluded. Cardiac MRI was suggested for further characterization. Tissue characterization would be important and useful for optimal management (surgical or medical) of the left atrial mass. Cardiac MRI was evident for a large mobile mass attached to the interatrial septum most suggestive of left atrial myxoma. Using delayed enhancement imaging and inversion time imaging the mass was also not thought to be a thrombus. Patient subsequently underwent excision of the left atrial mass with patch closure of resulting atrial septal defect. Pathology showed a large, soft, gelatinous mass and confirmed myxoma.

Discussion Primary cardiac tumors are rare with a lifetime incidence of 0.001%–0.03%, with approximately 75% being benign and nearly 50% of these being myxomas¹. As it relates to stroke, myxomas in particular are a rare cause and account for less than 1% of ischemic strokes³. TTE and TEE are essential imaging modalities to assess atrial masses, however both TTE and TEE provide limited tissue characterization and confident distinction between thrombi, benign and malignant tumors is often not possible². This distinction is particularly important for the management of left atrial masses.

112 PERI-PROCEDURAL ANTIBIOTIC PROPHYLAXIS IN VENTRICULAR SEPTAL DEFECT: A CASE STUDY TO RE-VISIT GUIDELINES

I Pour-Ghaz*, N Garg, S Salem, A Nanda, RN Khouzam. *UTHSC, Memphis, TN*

10.1136/jim-2018-000974.110

Background Ventricular septal defects (VSD) are the most common cause of congenital heart disease and most of small VSDs close without intervention. VSDs are associated with high velocity jet flow from left ventricular (LV) to right ventricular (RV) and when peri-membranous, can direct flow towards the right ventricular outflow tract (RVOT) and pulmonary valve (PV). Current American Heart Association/American College of Cardiology do not recommend prophylactic antibiotics for infectious endocarditis (IE) in acyanotic (AC) congenital hemodynamically insignificant VSDs.

Case presentation A 23-year-old woman with congenital small-uncomplicated VSD presented to the Emergency Department 37 days post-partum with fever, chills, fatigue, grade III/IV harsh diastolic murmur and *streptococcus viridans* positive blood cultures. Transthoracic and transesophageal echocardiogram showed a large vegetation on the PV, a medium vegetation on the aortic valve (AV), and a 3 mm small restrictive peri-membranous VSD. VSD flow jet was directed at the PV. Post treatment of bacteremia, she developed a new congestive

heart failure with LV systolic dysfunction and ejection fraction of 30%. VSD was closed and at two years TTE showed complete healing of the PV/AV.

Discussion In this case, presence of an unrepaired small VSD was the most likely predisposing factor in development of IE in a patient did not receive antibiotics prior to delivery. VSD was causing a left to right shunt with a velocity of 5 m/sec directed at the RVOT and PV. Jet flow likely caused denudation and seeding on the PV and it is likely that this turbulent flow in the LVOT caused IE of the AV as well.

Conclusion Incidence of bacteremia following vaginal delivery is not known. Congenital heart disease (CHD) has been shown to be a risk factor for IE post-partum. Currently antibiotic prophylaxis is not recommended for deliveries in those with AC congenital hemodynamically insignificant VSDs. This may be appropriate for low risk CHD, however, there are no data on the effectiveness of prophylaxis for IE in VSD cases after normal delivery. We propose that some defects can predispose patients to a greater risk of peri-procedural IE and prophylaxis maybe appropriate.

113 A CASE OF ABDOMINAL PAIN WITH AN UNEXPECTED AORTIC SURPRISE

¹LV Rodriguez-Torres*, ²M Milan Esteve. ¹Hospital Menonita Cayey, Anasco, Puerto Rico; ²San Juan City Hospital, San Juan, Puerto Rico

10.1136/jim-2018-000974.111

Case report Periaortitis is a rare disease, with an estimated incidence of 0.1–1.3 cases/100,000 persons per year. It is caused by fibro-inflammatory tissue surrounding the abdominal aorta, most commonly the infrarenal portion of the abdominal aorta. It can be seen as Idiopathic retroperitoneal fibrosis, inflammatory abdominal aortic aneurysm and perianeurysmal retroperitoneal fibrosis. It has been postulated that an exaggerated inflammatory response to advanced atherosclerosis is the main pathogenetic process, which this patient presents.

We present a 71 y/o non-smoker Puerto Rican female with past medical history of non-insulin dependent diabetes mellitus, hypertension, nephrolithiasis and hypothyroidism who came to the Emergency Room complaining of severe hypogastric abdominal pain, back pain, nausea, vomits, dysuria, tenesmus and general malaise for 2 weeks. Partially treated with antibiotics for a Urinary Tract Infection (UTI) for 5 days without improvement. Physical exam remarkable for hypogastric tenderness as well as bilateral costovertebral angle tenderness. Laboratories demonstrated an elevated ESR: 35 mm/hr; CRP: 0.9 mg/dl. Urinalysis showed pyuria, bacteriuria and positive nitrites. Renal panel, liver panel, pancreatic enzymes all normal. CT abdomen-pelvis showed bilateral perinephric stranding, calcified atherosclerosis of the abdominal aorta and branches with focal stranding on infrarenal abdominal aorta compatible with Periaortitis (early retroperitoneal fibrosis). Final urine culture and blood culture came back negative. Work up for vasculitis, autoimmune disease and malignancy were requested, but only positive for Neuronal Nuclear Antibody titer and Antinuclear Antibody with a ratio 1:320. Patient was started on broad spectrum antibiotics for UTI. Systemic steroids were provided due to the diagnosis of Periaortitis. After completing 7 days of intravenous antibiotics, she was discharge home with oral steroids. It is important to recognize Periaortitis early in its

course in order to prevent severe secondary complications. In this patient all work up for malignancy, vasculitis and auto-immune disease was normal. Most likely cause of findings was advance atherosclerosis which if diagnosed on time, can be life-saving.

114 ENDOCARDITIS: TO TEE OR NOT TO TEE?

H Shi*, F Simeone. *Tulane University Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.112

Case report The Eustachian valve is an embryologic remnant of the IVC valve that can form as a thickened fold of endocardium in the right atrium in adults. Eustachian valve endocarditis (EVE) is extremely rare and has only been described 30 times in the literature. A 45-year-old man with a history of IV drug use presented with shortness of breath. CT revealed septic emboli to his brain, lungs, kidneys, and spleen. Blood cultures grew MRSA and candida albicans. A TTE demonstrated mild mitral and tricuspid regurgitation. TEE revealed a large pedunculated and poly-lobulated 35 mm ×14 mm Eustachian valve vegetation in the right atrium (Figure: red arrow) along with a 23 mm ×9 mm vegetation on the left coronary cusp of the aortic valve (Figure: blue arrow). Persistent bacteremia prompted initiation of daptomycin, ceftaroline, and fluconazole. The patient suffered septic shock, requiring mechanical ventilation, vasopressors, and CRRT. Surgery was consulted for valve replacement but patient was poor operative candidate due to high risk of reseeding and poor prognostic factors (cerebral emboli). The patient remains in critical condition in the ICU. The fetal Eustachian valve assists blood flow from the IVC through the patent foramen ovale into the left atrium. This valve is vestigial and rarely can be associated with tumors, thrombi, and endocarditis. EVE has been most commonly been associated with *Staphylococcus aureus* and IV drug use. It is hypothesized that in IV drug users, increased right side expression of fibronectin and fibrinogen bind microorganisms more readily. The initial diagnostic modality for suspected EVE is TTE. However, this study can occasionally miss the Eustachian valve. TEE can view the Eustachian valve best with midesophageal four chamber and bicaval views. While the initial TTE here only showed trace MR and TR, multi-planar TEE revealed very large vegetations on two valves. Studies have shown that TEE should be performed more liberally with a lower threshold for suspected clinical endocarditis, especially in IV drug users. Our case highlights diagnosis and treatment of a large Eustachian valve vegetation with a concomitant coronary cusp aortic valve vegetation previously not described in the literature. This case also adds to the clinical repertoire suggesting the need of a TEE over a TTE for definitive diagnosis.

115 MYOCARDIAL INFARCTION IN A YOUNG FEMALE WITH SUSPECTED NORMAL CORONARY ARTERIES MANIFESTING AS ACUTE STROKE

¹L Sun*, ¹D Villarreal, ^{1,2}D Landsberg. ¹SUNY Upstate University Hospital, Clinton, NY; ²Crouse Hospital, Syracuse, NY

10.1136/jim-2018-000974.113

Case report A 28 year old female with type 2 diabetes presented with one week history of dizziness. CT head showed a

cerebellar infarct with local mass effect. EKG demonstrated right bundle branch block with ST elevations in anterolateral leads with reciprocal changes in inferior leads and Q waves suggestive of a recent anterior infarct. Transthoracic echo showed mid to apical dyskinesis and multiple large thrombi within the left ventricle (LV), largest measuring 3×2 cm; LVEF 25%–30%. Coronary angiography revealed significant organizing clot burden in the proximal and mid left anterior descending artery (LAD) with no evidence of any atherosclerotic disease. Remaining coronary arteries were normal. Patient underwent transaortic LV thrombectomy and median sternotomy for single-vessel bypass grafting with left internal mammary artery to LAD.

Discussion Myocardial infarction with angiographically normal coronary arteries (MINCA) typically occur in those younger than 50, accounting for 8% of all myocardial infarction (MI). Proposed mechanisms of MINCA include aortic dissection, hypercoagulable states, cocaine abuse, endothelial dysfunction and coronary vasospasm. Aside from diabetes, there is no evidence to suggest our patient had any prior history of heart disease, illicit drug use, or family history of early coronary disease or coagulopathies. Hypercoagulable work up thus far has revealed absence of antiphospholipid syndrome and normal protein C and S activity. Although it is not possible to definitively determine the cause of our patient's thrombotic MI, it is likely that a primary coronary event resulted in LAD thrombi which led to an acute MI with systolic dysfunction and subsequent propagation of LV thrombus formation. Given significant clot burden in LAD and absence of risk factors as mentioned above, it is suspected that the primary inciting event was a spontaneous coronary artery dissection (SCAD).

Conclusion This is a unique case depicting a young female presenting with dizziness found to have a cerebellar stroke secondary to embolization from significant LV thrombi as a result of SCAD or MINCA. It is important to determine the underlying etiology and pathophysiology of the ischemic event for appropriate long term management and prevention.

116 MODERATOR BAND ENDOCARDITIS CAUSED BY METHICILLIN RESISTANT STAPHYLOCOCCUS AUREUS

MK Teshome*, AA Asfaw, K Hesterberg, T Woods. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.114

Case report A 50-year-old woman with history of hypertension and history of illicit drug use presented to the emergency department with 3 days of shortness of breath, altered mentation, hypotension and multiple end organ damage. Four blood cultures were positive for MRSA. Initial Transthoracic echocardiogram (TTE) was negative. Chest x-ray and Computer Tomography of chest showed multiple nodular opacities with cavitation. Transesophageal echocardiogram (TEE) confirmed a diagnosis of right ventricular mural endocarditis attached to the moderator band extending to the right ventricular outflow tract. There was mild to moderate tricuspid valve regurgitation, but no valvular vegetation. Two sets of blood culture were negative before the patient was discharged to complete 6 weeks course of intravenous antibiotics.

Abstract 116 Figure 1

Abstract 116 Figure 2

Discussion Mural endocarditis is a rare form of infective endocarditis, which has been described mostly in patients with underlying predisposing factors. Staphylococci are among the commonest etiologies. Early and meticulous performance of TEE is advised when TTE is negative and the clinical suspicion of infective endocarditis is high.

117 RECURRENT CORONARY ARTERY THROMBOSIS IN A YOUNG PATIENT

MK Teshome*, AA Asfaw, RN Khouzam. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.115

Case report A 28 yo. woman with SLE on treatment presented to our hospital with anteroapical STEMI. Left heart

Abstract 117 Figure 1

catheterization (LHC) showed 99% thrombotic occlusion of the proximal left anterior descending coronary artery (LAD), which was treated successfully with drug eluting stent (DES). In the subsequent 18 months she had several recurrent thrombotic occlusions of the LAD and its branches while compliant to dual antiplatelet therapy and anticoagulant. Subsequently, after consecutive two episodes of STEMI 10 days apart due to thrombotic total occlusion of the LAD, she received one vessel CABG (LIMA to LAD). After these recurrent episodes, she developed heart failure with reduced ejection fraction (20%) and moderate sized left ventricular thrombus identified by transthoracic echocardiogram.

Pertinent laboratory results Positive Anti DS DNA antibody; Negative Anti β_2 glycoprotein IgG, Anticardiolipin IgG and IgM antibodies-; Elevated C-RP; Normal C3 and C4 levels.

Discussion SLE is a systemic disease with varied cardiovascular manifestations as pericardial, Valvular, myocardial and coronary artery involvement. Generally, coronary artery involvement is in form of accelerated and premature atherosclerosis which doesn't appear to differ from general population in pattern and extent of disease. Recurrent thrombosis with myocardial infarction tends to occur in 0.5% to 0.6% of those who have positive APLA. The recurrent, isolated coronary artery thrombotic occlusion in our patient suggests that SLE patients are at higher risk of not only accelerated atherosclerosis but thrombotic occlusion of coronary arteries even in the absence of APLAs.

118 STAPHYLOCOCCUS LUGDUNENSIS ENDOCARDITIS WITH TRICUSPID VALVE INVOLVEMENT

W Vutthikraivit*, M Ansari. *Texas Tech University Health Sciences Center, Lubbock, TX*

10.1136/jim-2018-000974.116

Case report *Staphylococcus lugdunensis*, is a rare but potentially aggressive coagulase-negative staphylococci. It can cause a wide variety of infections including endocarditis which has an acute and severe clinical course, similar to that caused by *S aureus*.

Case description A 31-year-old female with one automatic implantable cardioverter defibrillator (AICD) and one OPTIMIZER pulse generator presented to the hospital with complaints of fever, periodic sweat, anorexia, and worsening exertional dyspnea for 6 weeks. She had a past medical history of peripartum cardiomyopathy in 2005 that led to the placement of the CIEDs. Vital signs revealed fever, tachycardia, and

tachypnea. Physical examination revealed a grade 3 holosystolic murmur at the left lower sternal border and apex, otherwise unremarkable. A complete blood count revealed leukocytosis with neutrophil predominate. A CT angiogram showed bilateral subsegmental pulmonary embolisms (PEs). The initial diagnosis was sepsis with non-massive PEs, and the patient was empirically started on vancomycin, piperacillin-tazobactam, and anticoagulation. Two sets of blood cultures (BCx) drawn on the admission and the 4th hospital day grew *S. lugdunensis*. Infective endocarditis was confirmed with a 3.0×2.3 cm mass sits on the atrial aspect of the tricuspid valve found by transesophageal echocardiogram (TEE). Antibiotics were changed to nafcillin and gentamicin. BCx remained positive despite of adequate treatment. Repeated TEE on the 15th hospital day revealed a large mobile echogenic structure 4.5 cm ×2.5 cm attached to the tricuspid valve and leads. Because of poor treatment response, persistent large vegetation, multiple PEs, and the presence of a virulent pathogen, tricuspid valvular replacement with two CIEDs removal surgery was performed on the 17th hospital day. Her postoperative course was uneventful and repeat BCx were negative. Patient was discharged on day 26 with gentamicin, nafcillin, and rifampin regimen.

Discussion *S. lugdunensis* endocarditis is associated with significant morbidity and mortality. Once a patient with CIED placement is known to have *S. lugdunensis* endocarditis, the early surgical intervention may be the management of choice and should not be delayed.

Endocrinology and Metabolism

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

119 POORER ENDOGENOUS C-PEPTIDE RESPONSE IN INSULIN TREATED ADOLESCENTS WITH TYPE 2 DIABETES

¹MM Barr*, ²A Ashraf. ¹UAB School of Medicine, Birmingham, AL; ²University of Alabama at Birmingham, Birmingham, AL

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Purpose of study This study aims to discover if there were differences in glucose handling during a mixed meal tolerance test (MMTT) between adolescents with Type 2 Diabetes (T2DM) who were on insulin regimens vs those not using insulin.

Methods used Cross sectional and observational study. Data from initial study investigating the effect of Vitamin D on glucose and c-peptide release during MMTT (Clinical trial number NCT01325987). Statistical analysis used SAS version 9.4 (SAS Institute, Cary, NC, USA).

Summary of results Comparing baseline values between patients on insulin treatment and those who are not, insulin treated patients had higher fasting glucose, 2 hour glucose, and incremental glucose area under the curve (AUC). There was also a significant difference in HbA1C, with the insulin treated group having a higher HbA1C on average. Comparing patients by HbA1C $\geq 8\%$ to $<8\%$ the average 2 hour glucose and incremental glucose AUC were still significantly higher for $\geq 8\%$ group. There was no significant difference in overall

native insulin release, but insulin treated patients tended to have lower late phase endogenous c-peptide response compared to those with higher HbA1C.

Conclusions It remains unclear if the dysregulation in glucose control in more severe cases of pediatric T2DM is caused purely by a more severe disease state – as indicated with a higher HbA1C – or if insulin treatment contributes to accelerated deterioration in endogenous c-peptide production. Although glucose control during an MMTT was not different between insulin treated patients and those with higher HbA1C, the late phase native insulin release during MMTT was lower in the insulin treated group.

120 ACUTE ABDOMINAL PAIN: AN UNUSUAL PRESENTATION OF VAN WYK-GRUMBACH SYNDROME

¹J Carter*, ²L Berry, ^{1,3}D Jelley, ^{1,3}L Stuemky. ¹University of Oklahoma School of Community Medicine, Tulsa, OK; ²The University of Texas Rio Grande Valley School of Medicine, Edinburg, TX; ³The Children's Hospital at Saint Francis, Tulsa, OK

10.1136/jim-2018-000974.118

Background Van Wyk-Grumbach syndrome, or hypothyroidism with precocious puberty and pituitary enlargement secondary to hormonal overlap, is a rare condition which typically presents with signs of precocious puberty or vaginal bleeding.

Case presentation The patient is a 5 year old female with chief complaint of severe acute abdominal pain. Initial exam revealed hoarse voice, dry skin, and growth failure. Abdominal imaging revealed large bilateral ovarian cysts. Subsequent evaluation revealed a delayed bone age, which prompted further investigation including a free T4 level below assay limits, elevated TSH level of 916 uIU/mL, and significant pituitary enlargement on MRI. A unifying diagnosis of hormonal overlap, also known as Van Wyk-Grumbach syndrome, was made. The patient's signs and symptoms rapidly improved after the initiation of thyroid hormone replacement therapy. Her linear growth increased significantly and her abdominal pain resolved and ovarian cysts decreased. At nine month follow-up, she had resolution of pituitary enlargement on repeat MRI.

Conclusion This case emphasizes the importance of maintaining a broad differential diagnosis when investigating common complaints such as abdominal pain. A thorough evaluation is required to detect rare diagnoses and atypical presentations.

Abstract 120 Figure 1

121

DO PLACENTAL α 2-ADRENOCEPTORS PLAY A ROLE IN PREECLAMPSIA DEVELOPMENT?¹RP Chow*, ²J Zhao, ²TM Curtis, ¹T Lyons, ¹J Yu. ¹Medical University of South Carolina, Charleston, SC; ²Queen's University of Belfast, Belfast, UK

10.1136/jim-2018-000974.119

Purpose of study Preeclampsia (PE), characterized by *de novo* hypertension and proteinuria during pregnancy, remains a clinical challenge. Its pathogenesis includes excessive placental release of the anti-angiogenic factor soluble fms-like tyrosine kinase-1 (sFlt1); therefore, mechanisms that regulate sFlt1 expression are of interest. The human placenta expresses α 2-adrenoceptors (A2ARs), and levels are elevated in PE. A2ARs have also been implicated in embryonic placental development in a mouse model, and may act by decreasing sFlt1 levels. We aimed to:

1. confirm the presence of A2ARs in two commonly used human trophoblast cell lines;
2. determine if PE-relevant pathological milieus modulate A2ARs expression; and
3. explore the possible role of A2ARs in sFlt1 regulation.

Methods used Cultured human BeWo and HTR-8/SVneo trophoblasts were treated with dimethylxaloylglycine (DMOG, a hypoxia mimetic, 10 μ M) vs. control, or 'heavily oxidized, glycated' low-density lipoprotein (an oxidative stress stimulus) vs. native LDL (both 50 μ g/ml) for 24 hour. A2AR subtypes (2a, 2b and 2 c) were detected by immunostaining. Additionally, cells were treated with the A2AR agonists clonidine (10–100 μ M) and medetomidine (5–50 μ M), and the A2AR antagonist yohimbine (10–100 μ M). sFlt1 mRNA expression (RT-PCR) and protein secretion (ELISA) were measured. All experiments were repeated three times independently.

Summary of results All three A2AR subtypes were detectable by immunostaining in both trophoblast cell types. DMOG and oxidative stresses appeared to affect the expression and/or cellular distribution of the 2b and 2 c subtypes. Functional studies using pharmacological agents did not support a consistent effect of A2ARs in regulation of sFlt1.

Conclusions Human trophoblast cells express all three subtypes of A2ARs, and they appear to be modulated by hypoxia and oxidative stresses, two key pathological conditions associated with preeclampsia. However, functional studies suggest limited role of A2ARs in regulation of trophoblast expression or release of sFlt1. Whether these receptors are involved in other pathogenic mechanisms of PE warrants further investigations.

122

DIABETES MANAGEMENT IN THE VISION-IMPAIRED INDIVIDUAL

K Ellard*, M Bravo, S Lillis, T deSilva, M Reinoso, LS Engel, J Martinez. LSU Health Sciences Center, New Orleans, LA

10.1136/jim-2018-000974.120

Case report A 67-year-old man with Type 2 Diabetes, blindness, and hypertension presented with a three month history of dizziness. He reported episodic hypotension the lasted a few months as well as increased thirst and urinary frequency. He was adherent to his outpatient medication regimen that included linagliptin for treatment of his diabetes. He reported that he was independent at home but due to his vision impairment, received assistance in preparation of his weekly

medications. Due to visual impairment, he had difficulty checking his serum glycemic control through finger sticks. On physical exam the patient was orthostatic upon standing and had dry mucous membranes. The remainder of his physical exam was unremarkable. Initial laboratory studies were significant for a blood glucose of 745 mg/dl (70–100 mg/dl), serum osmolality of 307 mmol/kg (275–295 mmol/kg), and an anion gap of 17. The patient was diagnosed with diabetic ketoacidosis and was treated with IV hydration, IV insulin, and electrolyte management. Prior to discharge, his HgA1c was noted to be 15.5%. Initiation of an outpatient insulin regimen was considered. However, due to concerns regarding the limitations of his visual impairment on his ability to both monitor his home blood sugars and dose his insulin, he was discharge on an oral regimen consisting of metformin, linagliptin and glimepiride.

Discussion A large percent of patients with type 2 diabetes develop eye disease within 20 years of their diagnosis of diabetes. Retinopathy can result in significant impairments in quality of life. As the number of patients with diabetes mellitus is anticipated to rise over the next decade, there will also be an increasing need for better eye-care. Routine eye examinations are imperative as early recognition of the ophthalmologic complications of diabetes can help delay the progression of visual impairment. Once vision impairment develops, many changes are irreversible. With proper education, resources, and support for medication adherence, patients with vision impairment can achieve adequate blood glucose control.

123

RAPID ONSET OF EUGLYCEMIC DIABETIC KETOACIDOSIS DUE TO CANAGLIFLOZIN IN A CASE OF NEWLY DIAGNOSED TYPE 2 DIABETES MELLITUS

B Gyawali*, F Martinez, C Martinez-Balzano. SUNY Upstate Medical University, Syracuse, NY

10.1136/jim-2018-000974.121

Case report Sodium-glucose co-transporter 2 inhibitors (SGLT2i) are new agents used in the treatment of type 2 diabetes mellitus (T2DM). Canagliflozin was the first in this class of drugs and is often prescribed to younger patients with fewer co-existing illnesses. However, SGLT2i may be associated with twice the risk of euglycemic diabetic ketoacidosis (EDKA) compared to other agents¹. This case report seeks to raise risk awareness of EDKA in this population.

A 26-year-old man, an otherwise healthy man, presented to his primary care physician with polyuria, polydipsia and weight loss with intermittent abdominal pain. He was diagnosed with T2DM and prescribed with metformin and canagliflozin. A week later, he presented with nausea and intractable vomiting. He was found to have anion gap metabolic acidosis with a normal lactate level. He was admitted to the intensive care unit where additional laboratories revealed leukocytosis of 21,800/uL, bicarbonate of <5 mmol/L, and the anion gap of 32 mEq/L. His serum glucose was 112 mg/dL and beta-hydroxybutyrate was 12.27 mmol/L. Urinalysis was positive for ketones and negative for infection. Blood cultures were negative for bacterial growth. He was diagnosed with and treated for EDKA. Canagliflozin was discontinued, and he was given intravenous fluids with dextrose and regular insulin. His acidosis resolved after 48 hours. Glycemic control was achieved with basal and bolus insulin regimen. Evaluation for type 1 diabetes mellitus was negative, including insulin, islet cell, glutamic acid decarboxylase, and zinc transporter-8 autoantibodies.

Sustained reduction in HbA1c, potential benefits on hypertension reduction and weight loss has led to the increased use of SGLT2i as secondary agents in the treatment of T2DM. In this sense, canagliflozin has been associated with the largest effect on HbA1c, with near 1% reduction². It has been suggested that the urinary glucose excretion induced by SGLT2i, leads to decreased insulin and increased glucagon secretions, which in turn cause break down of fatty acids and overproduction of ketone bodies; thus, precipitating EDKA³. Clinicians should be aware of this potential complication.

124

LATE ONSET CONGENITAL ADRENAL HYPERPLASIA OR OVARIAN MASS?

¹S Hari Gopal*, ^{1,2}R Shridharani. ¹University of Tennessee College of Medicine at Chattanooga, Chattanooga, TN; ²University of Tennessee College of Medicine, Chattanooga, TN

10.1136/jim-2018-000974.122

Introduction Elevated 17-hydroxyprogesterone (17-OHP) is commonly associated with congenital adrenal hyperplasia (CAH). Benign steroid cell tumors are a rare sex cord tumor constituting 0.1% of ovarian tumors. In the pediatric population, these tumors have been reported to present with elevated testosterone levels with normal adrenal androgens, while in the adult population adrenal androgens are elevated with normal DHEAS levels.

Clinical case We report a 16 year-old female referred by Gynecology with findings of an elevated testosterone of 169.5 ng/dL, 17-hydroxyprogesterone (17-OHP) of 1873 ng/dL, and a normal DHEAS of 223 ug/dL with a 3.5 cm solid, uniformly enhancing mass in her right ovary on MRI. Her history revealed irregular menses since menarche at age 12 years and hirsutism 3.5 years prior to presentation. She was noted to have facial hair, thinning of scalp hair, and clitoromegaly on her initial exam.

Her initial ACTH stimulation test was notable for elevated 17 OHP and androstenedione with a normal DHEAS. The patient was started on hydrocortisone 15 mg/m²/day to treat possible late onset CAH. At her 1 month follow up, her labs revealed failure of suppression of the adrenal androgens with a testosterone of 150 mg/dL, 17-OHP of 585 ng/dL, and DHEAS of 43 ug/dL. The patient subsequently underwent right salpingo-oophorectomy. Pathology showed a 3.5 cm benign steroid cell tumor. A repeat ACTH stimulation test, 2 months off hydrocortisone, and following the removal of her ovarian mass, showed normal androgen levels.

Discussion Signs of virilization in a pediatric patient include hirsutism, clitoromegaly, and irregular menses. 17-OHP, testosterone, androstenedione, and DHEAS should be obtained in the initial evaluation. In the setting of an elevated 17-OHP, patient should be evaluated for late onset CAH. DHEAS mainly derives from the adrenal gland and in the adult population has been shown to be normal in cases of benign ovarian steroid cell tumors. In a pediatric patient with an elevated stimulated 17-OHP with normal DHEAS and a poor response to steroid treatment, an evaluation for an ovarian source should be considered.

125

HYPOCALCEMIA IN THE SETTING OF PRIMARY HYPOPARATHYROIDISM

CH Harris*, R Garikipati, LS Engel, J Amoss. LSU Health Sciences Center, New Orleans, LA

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Introduction Hypocalcemia is a common laboratory finding that can often be attributed to other electrolyte/vitamin abnormalities (hypomagnesemia, vitamin D deficiency). Hypocalcemia as a manifestation of hypoparathyroidism, is frequently due to surgical sequelae (thyroidectomy/parathyroidectomy) and clinical symptoms are not always apparent. We present a case of Primary Hypoparathyroidism presenting as symptomatic hypocalcemia.

Case A 40 year old African American man with Diabetes Mellitus Type 2 presented for left eye swelling following a fall. He additionally complains of recurrent falls associated with intermittent dizziness, nausea, and generalized muscle weakness over a two week period of time. Physical exam, upon arrival, was significant for somnolence, left periorbital hematoma associated with fall, and positive Chvostek's sign with percussion of the facial nerve. Initial labs were remarkable for markedly reduced corrected calcium of 5.1 mg/dL, inappropriately low intact PTH 18 pg/mL, elevated phosphorus of 5.9 mg/dL, magnesium of 1.7 mg/dL, decreased urine calcium <2, low levels of 25-hydroxyvitamin D at 5.4 ng/mL, and normal Cr of 1.0. EKG was significant for prolonged QTc without arrhythmia. Ultrasound of thyroid/neck revealed a right superior parathyroid gland 3.2×2.1×2.3 cm in size consistent with a parathyroid adenoma. The patient received IV calcium in addition to vitamin D and magnesium replacement with eventual resolution of hypocalcemia as well as presenting symptoms.

Discussion Parathyroid adenomas are often responsible for primary hyperparathyroidism. However, this is an interesting case of a parathyroid adenoma manifesting as hypoparathyroidism. When significant hypocalcemia is detected, search for underlying etiology and delivery of appropriate treatment is important for prevention of serious complications such as life-threatening arrhythmias, seizures, and tetany.

126

TRAUMA RELATED ORCHIECTOMY

D James*, A Dokun. University of TN Health Science Center, Memphis, TN

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Introduction Although rare, trauma is an unfortunate cause of testicular injury. If the degree of damage is great or if repair fails, orchiectomy is performed. This poses significant risk for testosterone (T) deficiency and development of symptomatic hypogonadism. Fortunately, residual testicular tissue may compensate and protect against androgen insufficiency.

Case A 42-year-old male without any known medical history was admitted to the trauma unit after sustaining a shotgun wound to the right thigh and scrotum. As a result, he underwent right orchiectomy and debridement of the left testicle. The patient retained approximately 50% of his left testicle. T levels, four days after the incident, revealed total and free T of 36 ng/dL (reference, 264–916) and 1.4 pg/mL (reference, 6.8–21.5), respectively with sex hormone binding globulin of 19.7 nmol/L (reference, 16.5–55.9). Corresponding follicle stimulating hormone (FSH) and luteinizing hormone (LH) were 5.88 mIU/mL (reference, 1.4–18.1) and 17.31 mIU/mL (reference, 1.5–9.3), respectively.

Management Initially asymptomatic, the patient was managed by watchful waiting as prompt initiation of T replacement can suppress endogenous T production. By six-week follow-up, despite increases of total and free T to 132 ng/dL and 3.4 pg/mL, respectively, the patient reported debilitating fatigue and

hot flashes. Biweekly T supplementation was introduced at that time. Three months later, there was complete resolution of hypogonadal symptoms despite intermittent adherence to therapy. Total T was 192 ng/dL with last dose of T being 25 days prior. The patient's wife, who accompanied him to the clinic, raised concerns of ill temperament and aggression in her husband's behavior. Given changes in demeanor, symptom resolution, and urological evaluation indicating low risk for left testicle atrophy, the dosage of T cypionate was halved. At this lower dose, the adverse effects to his conduct vanished. Clinic follow-up has been arranged with plans to wean T as tolerated.

This demonstrates the remarkable compensatory abilities of the endocrine system. Despite failing initial expectant management, it is hopeful that exogenous T will evidently be unnecessary as the patient's remaining organ is given the proper milieu to foster recovery.

127 OSTEOPOROSIS REVERSAL WITH VITAMIN D MONOTHERAPY

C Carrasco*, S Jaroudi, B Bates, N Adhikari, A Peiris. *Texas Tech University Health Science Center, Lubbock, TX*

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Introduction Osteoporosis is characterized by deficient bone mineral density (BMD) evidenced by dual energy x-ray absorptiometry (DEXA). The altered skeletal microarchitecture in osteoporosis leads to increased bone fragility and fractures. Initial therapy of choice is often oral calcium with vitamin D3 supplementation and bisphosphonates. We report a patient who demonstrated improved BMD with a high dose of vitamin D monotherapy.

Case description A 57-year-old female diagnosed with osteopenia (T-score in the lumbar spine of -1.6) during a screening DEXA presented for routine care. Patient was prescribed calcium and vitamin D3 supplements but admitted noncompliance when she returned 3 years later with a left wrist fracture. A repeat DEXA showed a lumbar spine T-score of -2.5 confirming osteoporosis. Secondary causes of osteoporosis were excluded. Bisphosphonate therapy was offered however patient declined due to concerns with potential side effects. Her vitamin D level at this stage was 36 ng/mL. Treatment was initiated with 4000 IU of vitamin D3 and 1500 mg of oral calcium daily. Over the course of a few months, her vitamin D supplementation was increased to 5000 IU daily then to 6000 IU daily. Repeat vitamin D values were 45 ng/mL and 52 ng/mL over a period of 6 and 12 months respectively. Patient reported she had not been compliant with calcium nor had she taken any other medications for osteoporosis except for vitamin D3. Five years after initial presentation the DEXA showed improvement with a lumbar spine T-score of -2.2.

Discussion This case shows an unusual improvement in osteoporosis with treatment based on intensive vitamin D therapy alone. The regression of osteoporosis to osteopenia, which correlated with vitamin D level increase, may suggest that calcium absorption is enhanced when vitamin D levels are in the 50–60 ng/mL range.

Conclusion The current practice of calcium supplementation and bisphosphonates administration with only minimum vitamin D replacement in patients with osteoporosis needs to be re-examined. Aggressive vitamin D replacement in osteoporotic patients could potentially help to reverse the disease process.

128 THE WRATH OF CALCIUM

MK Islam, S Abdelrazig, J Kimbugwe*. *Texas Tech Univ HSC Amarillo, Amarillo, TX*

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Introduction Hypercalcemia causes several effects on the gastrointestinal tract; dysphagia is one of the rare presentations among them.^{1 2} We are displaying a case of severe hypercalcemia presenting as dysphagia which improved after normalizing serum calcium.

Case An 83 year old female presented with 2 months of dysphagia for solids and liquidss, fatigue and constipation. Denied history of odynophagia, dysphonia, dysarthria, halitosis, neck swelling. On exam she was alert, moist mucous membranes, no neck masses. Examination of the oropharynx, chest, abdomen, cardiovascular and central nervous system was normal. lab revealed Hgb 13 g/dl, WBC 5.6, platelet 199, sodium 137 mEq/L, potassium 3.6 mEq/L, chloride 99 mEq/L, bicarbonate 28 mEq/L, creatinine 3.6 mg/dl, calcium 14 mg/dl, albumin 3.2 g/dl, corrected serum calcium 14.6 mg/dl, phosphate 3.4 mg/dl, intact PTH 395 pg/mL, 25hydroxy vitamin D 54.5 ng/mL, 1,25dihydroxy vitamin D 25 ng/ml (normal 19.9–79.3), PTHrP <2.0 pmol/L (normal <2.0), urine calcium/creatinine clearance ratio 0.04. Normal thyroid function, CXR and Barium swallow. Sestamibi scan showed no parathyroid adenoma. She was treated with IV fluids, calcitonin for 48 hours and cinacalcet afterward. She reported marked improvement of dysphagia once calcium level was normalized.

Discussion Dysphagia is the least reported effect of hypercalcemia on the gastrointestinal tract. Imaging ruled out mechanical causes and improvement in swallowing proved that hypercalcemia was the cause of dysphagia. Hypercalcemia causes depression of neuronal activity.³ Reduced neuronal activity results in reduced contractility of the smooth muscle in the lower 1/3 of the esophagus.

Conclusion Measuring serum calcium should be one of the first steps in the workup for dysphagia.

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129 ASSESSING VARIATION IN SOLUBLE ALPHA KLOTRO CONCENTRATIONS WITH RESPECT TO DIABETIC STATE AND THE ORAL GLUCOSE TOLERANCE TEST

AE Mikhael*, F Stentz. *University of Tennessee Health Science Center, Nashville, TN*

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Purpose of study To assess alpha Klotho concentration with respect to transition from a healthy to a Type 2 Diabetic state, and to note any correlations between glucose, insulin, and alpha Klotho levels during the OGTT.

Methods used

- Healthy subjects underwent an OGTT at the onset and conclusion of six months. Baseline and end of study samples from eight healthy subjects who developed diabetes and eight

Abstract 129 Figure 1 Average Soluble Alpha Klotho Concentrations of Healthy Subjects Followed for 6 Months

Abstract 129 Table 1

Patient	OGTT Glucose Difference of Time 0 min to 120 min	OGTT Klotho Difference of Time 0 min to 120 min	OGTT Insulin Difference of Time 0 to 120 min
1	10	9	30.7
2	-5	-13.1	5.7
3	47	-49.3	55.4
4	84	69.5	180.6

who did not develop diabetes during this period were assayed for alpha klotho levels via ELISA.

- OGTT plasma samples were obtained from four prediabetics followed for six months. OGTT samples from timepoint 0, 60, and 120 min were assayed for alpha klotho via ELISA. Insulin values were obtained using an Immulite Immunoassay, and glucose levels were obtained using a glucose analyzer.

Summary of results The average alpha klotho concentrations of healthy subjects who progressed to T2DM was significantly lower (p value=0.03) than for those who did not progress to T2DM. Levels of alpha klotho were observed to positively covary with glucose ($R^2=0.83$) and insulin levels ($R^2=0.74$) during the OGTT at both baseline and EOS.

Conclusions Poor glycemic control correlates with lower alpha klotho levels. As the kidney is a main site of synthesis for klotho, lower alpha klotho levels in recently diagnosed diabetics may indicate quite early stages of renal damage.

Our results suggest that alpha klotho levels vary during the OGTT. This indicates that there is a relationship between these three variables, and that this relationship can be noted during the duration of the OGTT.

130 BRUGADA PATTERN IN SEVERE HYPOTHYROIDISMS

M Morales Hernandez*, M Omar, JM Hernandez, JR Rivas, E Sottlie. *University of Florida College of Medicine, Jacksonville, FL*

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Case report A 18 year old Asian female with hypothyroidism and poor adherence to thyroid hormone replacement presented to the emergency room complaining of chills, malaise and dyspnea on exertion for 4 months. Denied syncopal episodes or family history of sudden cardiac death. She was hypotensive, febrile, tachycardic, with physical exam remarkable for crackles in left lower lung. Chest radiography revealed left lower lobe consolidation. Electrocardiogram

(ECG) showed sinus tachycardia with coved ST segment elevations in V1 and V2 consistent with Brugada type 1 pattern and a QTc of 605 ms. Complete cell count, electrolytes, and cardiac enzymes were within normal limits. Thyroid stimulating hormone returned as 124.5 mIU/L with a free T4 of <0.10 ng/dL and free T3 of -0.7 ng/dL. Echocardiogram demonstrated a small pericardial effusion and preserved ejection fraction. She was started on broad spectrum antibiotics, antipyretics, and was loaded with 200 mcg of intravenous levothyroxine. The following morning the patient remained febrile and repeat ECG showed resolution of ST segment changes and prolonged QTc interval. Patient was discharged in stable condition after she was transitioned to oral antibiotics and oral levothyroxine.

Brugada syndrome is an inherited disorder characterized by ST elevation in right precordial leads, right bundle branch pattern and high incidence for sudden cardiac death. Patients that present with similar ECG changes as Brugada syndrome but are asymptomatic are considered to have Brugada pattern. Worldwide prevalence of Brugada pattern is 6.1 per 1000 subjects with highest prevalence reported in Thailand and lowest in the United States. Brugada pattern is commonly related to fever, increased autonomic tone, and cocaine or psychotropic drug use. We present a case of Brugada pattern in a patient with severe hypothyroidism without myxedema coma. ECG changes resolved after intravenous levothyroxine despite persistence of febrile episodes. Thyroid hormone is known to affect the action potential duration and repolarization currents in cardiac myocytes; the exact mechanism remains unclear.

The underlying mechanism on how hypothyroidism allows for Brugada pattern is not elucidated. Prompt management of severe hypothyroidism is warranted.

131 USING TELEMEDICINE TO IMPROVE OUR CARE OF TYPE 1 DIABETES

GB Nelson*, K Sumpter, E McCoy. *University of Tennessee Health Science Center, Memphis, TN*

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Purpose of study Type 1 Diabetes (T1D) is an autoimmune condition that results in loss of pancreatic beta cells, insulinopenia and, without adequate insulin administration, can lead to life-threatening diabetic ketoacidosis (DKA). Responding to high blood sugars and presence of ketones with appropriate increase of insulin is important to avoid DKA and hospitalization. In order to decrease rates of DKA and admissions our institution provides instructions for patients to follow in case of hyperglycemia and/or ketones. We also provide access to a 24/7 call center that can assist with these instructions or connect to a physician. To improve this process, we must examine how our patients are using these resources. The purpose of this study is to examine the characteristics of how and when patients use the call center in order to adjust our patient care appropriately. We hypothesize that the majority of patients admitted for DKA do not call the transfer center within 3 days prior to admission.

Methods used This is a retrospective review, conducted from February 2018 until August 2018, using both admission and call center data. Patients admitted at diagnosis of diabetes were excluded. Call center logs were cross-checked for calls from the patient within 3 days prior to admission date

(referred to as a 'pre-admission call'). Calls greater than 3 days prior or any time after admission were not counted as a pre-admission call.

Summary of results Of patients admitted for DKA, 10% had called with blood sugar related questions within the 2 weeks prior to admission. Only 7% called within 3 days prior to admission. Of note, 40% of these patients called within 2 hours of admission. Pre-admission calls 2 weeks and 3 days prior to admission accounted for 1.1% and 0.6% of all transfer center calls respectively. Calls related to blood sugar questions in general accounted for ~60% of all calls.

Conclusions The percentage of patients admitted for DKA who called with a blood sugar related question within 3 days prior to admission was 7%. Nearly half of these patients called immediately prior presenting to the ER, likely too late for effective home management. Determining specific characteristics of patients who call and do not call will help target interventions to improve our patient care.

132

METABOLIC CHANGES AFTER ONE YEAR OF BARIATRIC SURGERY IN OBESE HISPANIC PATIENTS

H Nuñez, PI Altieri, A Suárez, HL Banchs. *University of Puerto Rico, Medical Sciences Campus, San Juan, Puerto Rico*

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Purpose of study Bariatric surgery (B.S.) is a very useful method in the management of morbid obesity. The report of Clalit-Reseach Institute in Telaviv showed in a span of 4.5 years a mortality in the surgical group of patients (P.) 1.3% compared to 2.3% in the non-surgical P. Our purpose is to compare the metabolic changes (M.C.) between the 2 types of B.S., sleeve (S.) and Roux-N-Y (R.).

Methods used A total of 34 P. who underwent S. and 102 P. who underwent R. were compared.

Summary of results The comorbidities were the same and the mortality 1.5%. The pre-op values in B.M.I., total cholesterol (C.), LDL, HDL and F.B.S. were the same in both groups. In the R. group a significant reduction of LDL (14%) vs. the S. group (7.62%) was observed, $p < 0.0093$. The HDL was increased, R. (11.73%) in S. (23%) $p < 0.9319$. The C. was reduced, R. (10.49%), S. (1.80%) $p < 0.096$. The F.B.S. showed a reduction, R. (11.42%), S. (13.76%) $p < 0.9916$. The B.M.I. showed a reduction in R. of (34.04%), and in S. (30.08%) $p < 0.9315$. In comparison between the 2 groups, no differences were found in B.M.I., T.G., F.B.S and HDL.

Conclusions The P. with R. has greater benefits in losing weight, although B.M.I. changes were the same, but complications in R. surgery were greater. Follow up for 5 years have shown the same reduction in the metabolic parameters. Both groups showed an increase of C-reactive protein of 20%. Due to these findings we prefer S. over R. surgery. A longer follow up of more than 5 years will clarify if these changes persist longer.

133

METASTATIC NEUROENDOCRINE CARCINOID TUMOR TO LEFT ORBIT

^{1,2}A Reddy*, ¹J Kaur, ²WP Daley. ¹University of South Alabama, Mobile, AL; ²University of Mississippi Medical Center, Jackson, MS

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Introduction Carcinoid tumors, low-grade neuroendocrine tumors, arise from the enterochromaffin cells, predominantly originating in the gastrointestinal or bronchia tracts. With the potential of malignancy relatively low, the rate of metastases for carcinoid tumors is about 50%–75% in patients with the most common sites being liver, bone, and lymph nodes. Even rarer is carcinoid tumor metastasis to the orbit which accounts for only 4% to 5%. Here we present a case of metastatic neuroendocrine tumor to the left orbit.

Case report A 66-year-old male with a history of neuroendocrine carcinoid since 1 year presented with abnormal left eye movements. Computer tomography angiography revealed persistent and slightly increased inferior oblique insertion presenting as an enlargement. Left orbitotomy and inferior oblique biopsy were performed. The biopsy of the left inferior oblique muscle revealed a metastatic neuroendocrine carcinoma, grade 2 to 3. Appropriately controlled immunohistochemistry showed the tumor cells stained positive for CD56, synaptophysin, and chromogranin. No mitoses were identified. The Ki-67 proliferation index was estimated as 4%–5%. The tumor extended to the biopsy tissue edges. The histopathological findings were diagnostic of a metastatic neuroendocrine tumor to the left orbit.

Discussion Neuroendocrine tumor metastasis to orbital structures is an uncommon event. The most common presenting symptoms of orbital carcinoid tumors are swelling, inflammation, and restriction of ocular mobility from a mass. Upon literature review, there were only a few reported cases of metastatic orbital carcinoid tumors which presented as orbital inflammation and restricted movements. This case report highlights a rare presentation of the metastatic neuroendocrine tumor as an orbital mass. The clinician and pathologist during diagnosis and analysis should be able to make a distinction between an orbital outlet obstruction, an orbital infection, or the rarer systemic carcinoid syndrome.

134

ANGIOEDEMA OR GLUE SNIFFER'S RASH

DS Rees*, M Touchy, J Maldonado, MV Naljayyan, EA Aguilar, S Barry, F Yazdi, RB Varelzdis, SA Morse, E Reisin. *LSUHSC, New Orleans, LA*

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Description 46 year old male with past medical history of bipolar disorder, deep vein thrombosis, hyperlipidemia, pulmonary embolism, hypertension, and prior suicide attempt presented with chief complaint of swelling of lips and face that began approximately 8 hours prior to arrival. Patient reported shortness of breath, difficulty swallowing, headache, nausea, vomiting, constipation, decreased urination, and dark urine. He is on lisinopril. He also reports that he has been redoing the roof of an RV and has been exposed to the chemicals involved in that process. On physical examination, blood pressure 102/75 mmHg, heart rate 81bpm, temperature 97.3F, respiratory rate 18 BPM, SpO2 96%. Notable for swelling of lips and around the eyes but otherwise unremarkable.

CBC WNL Na 136 mEq/L, K 3.5 mEq/L, Cl 99 mEq/L, bicarbonate 21 mEq/L, anion gap 16 mEq/L, BUN 42 mg/dL, creatinine 5.58 mg/dL, blood glucose of 150 mg/dL, calcium 11.1 mg/dL, phosphorus 5.4 mg/dL, magnesium 1.6 mg/dL, CPK 644 U/L, lactate 1.2 mmol/L, total bilirubin 1.7 mg/dL. Urinalysis with 2+protein, 1+ketones, 3+blood, 2+bilirubin, 2 RBCs, 1 WBC, FENa 0.3%, myoglobin 40 ng/ml, urine

osmolality 662. Renal US was unremarkable except for left lower pole renal cyst. Urine toxicology was positive for tetrahydrocannabinol (THC) and amphetamines.

His acute kidney injury resolved with IV fluids. This could simply be a case of acute kidney injury stage 3 from volume depletion due to nausea and vomiting, but it is interesting that he had exposure to an unknown chemicals. Some RV resurfacing chemicals contain volatile solvents like toluene.

Conclusion Toluene can cause toxicity by being ingested, inhaled, or absorbed across the skin. Symptoms include CNS disturbances, GI disturbances, electrolyte imbalances, rhabdomyolysis, and hepatorenal toxicity. Toluene is a common substance of abuse and is frequently inhaled or 'huffed.' This causes exposure of the chemical around the nose, mouth, and eyes and can cause irritation or allergic reaction referred to as a 'glue sniffer's rash.'

135 LIPID ABNORMALITIES AND METABOLIC SYNDROME IN PEDIATRIC PATIENTS WHO RECEIVED HEMATOPOIETIC STEM CELL TRANSPLANTS

¹TH Richardson*, ^{1,2}A Ashraf. ¹UABSOM, Birmingham, AL; ²Children's Hospital of Alabama, Birmingham, AL

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Purpose of study Adults who received hematopoietic stem cell transplants as children have higher rates of cardiovascular disease and the metabolic syndrome. Little is known about the progression of lipid abnormalities and the development of metabolic syndrome while the patients are young. This study assessed the prevalence of the metabolic syndrome after hematopoietic stem cell transplant in the pediatric population focusing on changes in lipid profiles.

Methods used This is a retrospective study of 52 patients at Children's of Alabama who received hematopoietic stem cell transplants. Systolic and diastolic blood pressure, liver function, thyroid function, hemoglobin A1c, total cholesterol, HDL, LDL, and triglycerides were compared prior to transplant, 2 years, 5 years and 10 years after transplant. Patients were excluded if they did not have adequate: anthropomorphic data (e.g. height and weight), demographic data, clinical information (i.e. physicians' notes), and laboratory values (n=114).

Summary of results There was no significant difference in average total cholesterol, HDL cholesterol, triglycerides, liver function, systolic blood pressure, or diastolic blood pressure between the time intervals. The percentage of patients who met 3 of 5 criteria for metabolic syndrome at 2, 5, and 10 years post-transplant were 17%, 21%, and 14%. 10 years after transplant, 54% of patients had elevated total cholesterol, 43% had elevated triglycerides, 39% had low HDL, 14% had a BMI in the 95th percentile, and 7% had elevated blood pressure. 2 years after transplant, 75% of patients had at least one abnormal lipid value. 5 years after transplant, 63% of patients had at least one lipid abnormality. 10 years after transplant, 71% of patients had at least one lipid abnormality.

Conclusions There was no statistically significant difference in lipid profiles, liver function tests, or blood pressure between the different time points. The percentages of patients who met 3 out of the 5 criteria for metabolic syndrome at 2, 5, and 10 year post-transplant was higher than the normal, non-

obese pediatric population. There was also a significant percentage of patients who had at least one lipid abnormality.

136 SURGICAL PREPARATION FOR THYROIDECTOMY IN AN UNUSUAL CASE OF GRAVES DISEASE

Z Riemenschneider*, M George, M Marin. University of Oklahoma Health Sciences Center, Oklahoma City, OK

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Case report Our patient is an 18 year-old female with Graves disease who presented after three days of bilateral upper extremity myalgia, bone pain, intermittent rash, bilateral hand paresthesia, and weakness. On exam she appeared to be in obvious pain and was tremulous, but exhibited no other signs of thyrotoxicosis. She had thyromegaly, 1/5 strength in both upper extremities, and tenderness to palpation of the humeri bilaterally. Initial lab work showed elevated AST and ALT, low TSH, and a free T4 of 4.6 ng/dL. Rheumatology work-up revealed a mildly elevated ANA (titer of 1:40) and negative anti-neutrophil cytoplasmic antibodies.

Her symptoms were attributed to adverse effects of methimazole which she started one month prior. Methimazole was discontinued and she was started on propranolol 30 mg q6 and prednisone 20 mg q12. After three days off methimazole her pain and weakness improved significantly and her liver function tests trended down. Because of her adverse reaction to methimazole, propylthiouracil was contraindicated as an alternative therapy. Additionally, she was deemed a poor candidate for radioiodine ablation as her free T4 was greater than three times the normal range at its peak value of 5.4 ng/dL. She instead underwent total thyroidectomy. To prepare for surgery, she was treated with oral potassium iodide over the course of 10 days in conjunction with beta blockade and a corticosteroid. On this regimen we were able to reduce her free T4 to 2.44 ng/dL prior to surgery. The operation was successful in removing her thyroid gland with only minimal resultant hypocalcemia.

Discussion Arthralgia, myalgia, rash, and paresthesia are well known adverse effects of thionamide use. This case is unique because of our patient's bone pain and weakness which, to our knowledge, have not commonly been described as adverse reactions to methimazole. Additionally, our case highlights an infrequently utilized method for rapid pre-operative preparation prior to thyroidectomy with oral potassium iodide, beta-blockers, and corticosteroids. A review of the literature reveals that the data for this method is still limited, but our case represents an additional piece of evidence in support of this rapid preparation regimen.

137 A CLASSIC PRESENTATION OF AN EXCEEDINGLY RARE CONDITION: MYXEDEMA COMA

P Staiano*. UF Health Jacksonville, Jacksonville, FL

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Case report Myxedema Coma is an extremely rare and life threatening presentation of hypothyroidism. The classic signs of are hypothermia, bradycardia, hyponatremia, encephalopathy, hypoglycemia, and hypotension. When suspected, treatment must be started immediately due to the high mortality

rate. Discussed here is a case of a man who was found down, unresponsive, with no identifying information.

A 66 year old male presented to the hospital after being found unresponsive. He was hypotensive, severely hypothermic, bradycardic, and obtunded requiring mechanical ventilation. On physical exam he was disheveled with diffuse anasarca and periorbital edema, expiratory wheezes, and rhonchorous breath sounds. He was started on intravenous levothyroxine, stress dose steroids, and empiric antibiotics. Pertinent labs included TSH 90, T3 0.70, T4 0.10, glucose 50, and sodium 127. Respiratory viral panel was positive for Rhinovirus, and endotracheal tube aspirate grew *Haemophilus influenzae*. Vasopressors were used briefly due to persistent hypotension. Stress dose steroids were discontinued once adrenal insufficiency was ruled out. The patient steadily improved, making a full recovery.

Myxedema Coma has a high mortality rate (25%–60%), even when treated promptly, thus survival is largely dependent on time to treatment with levothyroxine and steroids. Initial dose of levothyroxine is 200–400 mcg intravenously followed by 50–100 mcg daily. The lower dose (200 mcg) is used for patients with known heart disease. Start hydrocortisone 100 mg every 8 hours until adrenal insufficiency is ruled out. Draw T4, TSH, cortisol, with or without a cosyntropin stress test after treatment is started, and repeat free T4 and T3 1–2 days later to monitor response. When transitioning to oral, start with 75% of the required intravenous dose.

Supportive treatment involves passive rewarming, mechanical ventilation, and vasopressors as needed. Caution should be used with active rewarming due to vasodilation and potential for worsening hypotension.

Myxedema Coma is an extremely rare, life threatening event with a high mortality rate even when treated. When suspected, treatment must be started immediately prior to laboratory results. In this case, the classic signs of myxedema coma were evident at presentation, and appropriate treatment was started early with a good outcome.

138

THYROTOXIC PERIODIC PARALYSIS IN A YOUNG PATIENT PRESENTING WITH SUDDEN ONSET BILATERAL LOWER EXTREMITY PARALYSIS AND HYPOKALEMIA

MN Vinan Vega*, S Thavaraputta, RM Medrano, B Mantilla, SK Koshy. *Texas Tech University Health Science Center, Lubbock, TX*

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Case report Thyrotoxic periodic paralysis (TPP) is a rare disorder with intermittent episodes of muscle weakness that can result in sudden onset paralysis, often associated with severe hypokalemia.¹ Its incidence is 0.1%–0.2% in hyperthyroid patients in North America.²

Hypokalemia results from the rapid transcellular shift of potassium by thyroid hormone induced beta adrenergic sensitization of Na⁺/K⁺–ATPase rather than total body depletion.³ We present a case of TPP with immediate resolution of paralysis after treatment with methimazole and potassium supplementation.

A 34-year-old male presented with sudden onset of lower extremity flaccid paralysis with no associated pain or paresthesia and sparing of upper extremities and cranial nerves. Patient had history of Grave's disease (with elevated levels of thyroid stimulating immunoglobulin and anti-thyroid peroxidase antibody)

diagnosed four years ago and multiple hospitalization for similar episodes of flaccid paralysis precipitated by non-compliance with taking methimazole. Physical examination showed elevated blood pressure (147/90 mmHg), bilateral enlargement of thyroid without palpable nodules, 1/5 motor strength and hyporeflexia in bilateral lower extremities. Sensation was intact. Laboratory findings revealed hypokalemia (2.8 mmol/L), suppressed thyroid stimulating hormone (<0.01 microIU/mL) and elevated Free T4 (3.86 ng/dL). Paralysis resolved within after starting methimazole and propranolol and he was discharged the next day with complete resolution of symptoms.

TPP is an uncommon cause of periodic paralysis, however it is important to recognize this in patients with thyrotoxicosis presenting with flaccid paralysis. Though the paralysis is secondary to associated hypokalemia, the electrolyte imbalance gets easily corrected with aggressive treatment of thyrotoxicosis without a need for chronic potassium supplementation. Failure to recognize this condition may result in inadvertent excessive potassium supplementation and subsequent hyperkalemia and arrhythmias.⁴

139

PERINATAL INTERKINGDOM MICROBIAL COMMUNITY COMPOSITION HAS LONG-TERM HOST METABOLIC CONSEQUENCES

KA Willis*, CK Gomes, AJ Talati, JF Pierre. *The University of Tennessee Health Science Center, Memphis, TN*

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Purpose of study During early life, intestinal bacterial community composition influences long-term metabolic outcomes, but the role of fungi is completely unexplored. We tested if maternal perinatal antibiotic exposure alters fungal community composition associated with long-term metabolic consequences.

Methods used We randomized pregnant C57BL/6J mice to penicillin or cefoperazone (PCN and CPZ, 500 mg/L) in drinking water or control water on gestational day 10 until post-natal day 15. At 3 and 4 weeks of life, EchoMRI was used to quantify body composition. Stool contents were analyzed by 16S rRNA and ITS sequencing for interkingdom microbial community and predictive functional metagenomics.

Summary of results In female offspring, fat mass increased after both antibiotics, and lean mass decreased after CPZ compared with controls (ANOVA $p < 0.0001$). In male offspring, CPZ increased fat mass, without changes in lean mass (ANOVA $p < 0.001$). After PCN, the bacterial order *Bacilli* decreased while *Clostridia* expanded, and the fungal order *Eurotiales* decreased while *Dothideales* expanded (ANOSIM

Abstract 139 Figure 1 Microbial community composition

$p=0.01$). After CPZ, the bacterial order *Alphaproteobacteria* was nearly eliminated, *Bacteroidia* was reduced, and *Clostridia* expanded markedly. CPZ led to a similar but less extensive expansion of the fungal order *Dothideales* compared to PCN (ANOSIM $p=0.01$). Bacterial metagenomic pathways related to carbohydrate metabolism increased with antibiotic exposure, while ribosome and DNA repair pathways decreased.

Conclusions Commensal fungal and bacterial community composition in offspring are altered by maternal antibiotic exposure and associated with long-term host metabolic effects.

140

CASE SERIES: DEFICIENT THYROID TRANSPORT TO THE BRAIN EXPLAINS ENIGMATIC DEVELOPMENTAL DELAYS

¹M Yousif*, ¹R Raza, ¹G Purcarin, ^{1,2}K Wierenga, ¹M George. ¹University of Oklahoma Health Science Center, Oklahoma City, OK; ²Mayo Clinic Florida, Jacksonville, FL

10.1136/jim-2018-000974.138

Case report Thyroid hormone plays a vital role in neurodevelopment, metabolism, growth, and many other critical processes of the human body. This is primarily completed via the coordination of the hypothalamic-pituitary-thyroid axis, which results in the production of thyroid hormones, and the thyroid hormone transporters via which the thyroid hormone is transported intracellularly. The monocarboxylate transporter 8 (MCT8) is an important protein for the regulation of local thyroid hormone activity in the brain and thus for normal brain development, and is encoded on the X chromosome. Loss of function mutations on this gene results in a defect in the MCT8 transporter that leads to a hypothyroid state of the brain. Male subjects with this mutation display a severe intellectual and motor disability disorder known as Allan-Herndon-Dudley syndrome (AHDS). The hallmark of AHDS include: intellectual disability, global developmental delay, hypotonia, spastic or dystonic quadriplegia, dysphagia and seizures. Elevated T3 levels, low T4 and rT3 concentrations, in combination with a normal or slightly elevated thyroid stimulating hormone (TSH) are the biochemical hallmarks of AHDS.

Male patients affected by AHDS present with non-specific hypotonia and developmental delay early in life, with the other clinical symptoms often presenting at somewhat older age. The non-specific presentation may contribute to the length of the diagnostic odyssey. Herein we report the experience with Allan-Herndon-Dudley syndrome in our mid-level academic center. We collected data on 7 patients (from 4 unique families), and report the diagnostic, biochemical, genetic and clinical profiles to further define this unusual clinical and biochemical phenotype.

Gastroenterology

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

141

AN UNEXPECTED OUTCOME

M Antoine*, B Sharma. *Suny upstate, Syracuse, NY*

10.1136/jim-2018-000974.139

Introduction Hepatic encephalopathy (HE) is a spectrum of neuropsychiatric abnormalities caused by liver disease. A small fraction of patients can become refractory to conventional medical management and liver transplantation has generally been the only treatment option, however, there are emerging data on alternative therapies. We present a case of a patient with refractory HE who exhibited an inadvertent improvement in cognition after a procedure intended for treatment of splenic artery hemorrhage.

Case A 57-year-old female with cirrhosis secondary to Hepatitis C and multiple hospitalizations for HE was being treated aggressively with rifaximin, lactulose, and zinc for recurrent HE without a clear precipitating factor in spite of medical compliance. Neurologic etiology was ruled out. Her MELD (Model For End-Stage Liver Disease) score was 14. Pending transplantation evaluation, a Computed Topography (CT) of her abdomen was ordered to rule out large porto-systemic shunt as the cause of the persistent HE and it did show a large spleno-renal shunt and splenic artery aneurysm. She was arranged for Balloon-occluded Retrograde Transvenous Obliteration (BRTO) as rescue therapy to transplantation but developed a hemodynamically significant bleed and was admitted to the Intensive Care Unit. The bleed was caused by rupture of the splenic artery aneurysm. She was severely encephalopathic with physical examination showing sustained somnolence and asterixis despite aggressive medical therapy. Coil embolization of the splenic artery was performed which stopped the bleeding. She was stabilized and discharged from the hospital. Weeks after discharge, patient showed unexpected marked improvement of mental status and cognitive function.

Discussion The splenic artery embolization that was performed for this life-threatening bleeding seems to have inadvertently improved her encephalopathy. From literature review, though limited, partial splenic artery embolization (PSE) has in fact been performed for refractory hepatic encephalopathy. In this patient, the embolization decreased splenic blood flow, splenic venous pressure and portal venous pressure leading to improved portal hemodynamics. Treatment options for refractory hepatic encephalopathy are at this time limited and PSE is a promising option. Further research should continue to evaluate its efficacy and complications.

142

ISCHEMIC COLITIS FROM DEXTROMETHORPHAN OVERDOSE

M Badrinath*, U Masood, S John, A Khan. *SUNY Upstate, Syracuse, NY*

10.1136/jim-2018-000974.140

Introduction Toxicity of dextromethorphan (DXM) which is a commonly used anti-tussive, usually presents with neurological symptoms and no vascular complications. We describe ischemic colitis (IC) in a young male after DXM overdose.

Case A 27 year old male presented with abdominal pain and multiple episodes of watery stools mixed with bright red blood, four hours after consuming four bottles of DXM. He did not have cardiovascular risk factors, hypercoagulable conditions or vascular anomalies. Vitals were normal, physical exam revealed diffuse abdominal tenderness and bright red blood mixed with stool on rectal exam. Laboratory findings revealed leukocytosis of 16200/uL and normal lactic acid. Urine drug toxicology was positive for DXM. Computed tomography (CT) of the abdomen/pelvis revealed thickened

Abstract 142 Figure 1

colon (figure 1). Stool studies for infectious etiology were negative. Colonoscopy revealed inflamed and congested mucosa. Biopsy showed focal necrosis of colonic mucosa consistent with IC. Patient recovered with supportive care and was discharged a week later.

Discussion IC is due to reduced blood supply to colon causing an ischemic injury, usually affecting the watershed areas with an incidence of 4.5 to 44 cases/100,000 person-year. Common causes are arterial occlusion, venous thrombosis or hypoperfusion of the mesenteric vasculature. DXM is an opioid related to codeine that exhibits phencyclidine-like effects at large doses by blocking the excitatory NMDA receptors with mild alpha-1 activity. Neurological symptoms due to toxicity are seen 4 hours after ingestion. While therapeutic doses of DXM do not cause significant vasoconstrictive effects, overdose may cause a colonic hypoperfusion state from significant alpha-1 receptor agonism. To our knowledge, this is the first case of IC secondary to DXM overdose. CT abdomen showed no gross vascular anomaly which could have precipitated this event. Naranjo probability scale revealed DXM to be a probable cause of IC in our patient.

143 NO SHIGA, SHERLOCK: NON-SHIGA-TOXIN INDUCED HEMOLYTIC UREMIC SYNDROME

L Buchanan*, F Pedigo, M Horn. *University of Mississippi Medical Center, Jackson, MS*

10.1136/jim-2018-000974.141

Case report Hemolytic Uremic Syndrome (HUS) is a well-documented sequela of infection with Shiga-toxin producing organisms such as *Escherichia coli*, particularly strain O157:H7. HUS precipitated by non-Shiga-toxin producing organisms is more poorly understood. Here, HUS secondary to *Campylobacter jejuni* is described.

A 26-year-old female with no medical history presented with two days of bloody diarrhea associated with abdominal pain, nausea, and vomiting that began after consuming chicken stir-fry. Supportive measures, including intravenous fluids, were initiated. Advanced imaging revealed pancolitis. Stool cultures resulted positive for *Campylobacter jejuni*. Over the following days, her serum creatinine doubled, platelets decreased from 187,000 to 40,000, and hemoglobin fell from 12 to 9.8. Hemolysis labs were positive, and a review of her peripheral blood smear revealed schistocytes. Thrombocytopenic Purpura and other causes of HUS were excluded. Shiga-toxin returned negative, while ADAMTS13, ANA, and complement levels were all unremarkable. Thus, diarrheal HUS secondary to *Campylobacter* was confirmed.

Because HUS is a potentially fatal condition, prompt diagnosis and management are imperative. The CDC estimates that *Campylobacter* infection affects over 1.3 million people in

Abstract 143 Figure 1 Schistocytes on peripheral smear

the United States yearly. Due to the prevalence of this organism, practitioners should be acutely aware of this rare, but potentially deadly, sequelae from this common infection.

144 BUDD CHIARI: A CASE DEMONSTRATING IMPORTANCE OF THROMBOCYTOSIS WORKUP AND DISCONTINUATION OF ORAL CONTRACEPTIVES

V Chat*, S Rassameehiran. *Medical College of Georgia, Augusta, GA*

10.1136/jim-2018-000974.142

Background Budd-Chiari syndrome (BCS) or hepatic venous outflow tract obstruction is a rare entity among vascular disorders of the liver and has a highly variable presentation. Most patients with BCS are female with an average age of 39 years. We report a case of a young female with longstanding BCS and emphasize the importance of early recognition of a hypercoagulable state and early discontinuation of pro-coagulant medications.

Case report A 29-year-old woman with a 5 year history of thrombocytosis presented with mild intermittent upper abdominal pain of one-month duration. Her only medication was the oral contraceptive pill (OCP), which she had been taking for 13 years. She denied personal or family history of chronic liver disease, deep vein thrombosis, or pulmonary embolism. Physical examination was remarkable for jaundice and palpable left liver lobe at the epigastrium without stigmata of chronic liver disease. Initial laboratory revealed Hgb 15.9 g/dL, WBC 10.7 K/ μ L, platelet 428 K/ μ L, TB 2.7 mg/dL, total protein 7.3 g/dL, albumin 3.9 g/dL, alkaline phosphatase 200 IU/L, AST 59, IU/L, ALT 83 IU/L. MRI of the abdomen with and without contrast revealed heterogeneous enhancement of the liver, enlarged caudate lobe, macroregenerative nodules, diminutive hepatic veins, and small ascites. Hypercoagulable work-up showed positive JAK2 mutation. The

diagnoses of longstanding BCS and polycythemia vera were made. She underwent hepatic venography and transjugular intrahepatic portosystemic shunt (TIPS) placement, which resulted in resolution of portal hypertension (HVPG was 22 mmHg prior to TIPS placement and PSG was 7 mmHg after the procedure). She continued on anticoagulant therapy after the procedure.

Conclusion BCS is a rare condition with a high mortality rate if untreated. Up to 87% of patients with BCS have at least one underlying risk factor for thrombosis, the most common being a myeloproliferative disorder. In this case, our patient was predisposed due to polycythemia vera and OCP use. Primary care providers should re-evaluate the use of OCP and offer different methods of contraception when thrombocytosis is present.

145 PLESIOMONAS SHIGELLOIDES INFECTION: A RARE CAUSE OF CROHN'S EXACERBATION

M Ibrahim*, N Nair, J Phemister, J McKinney. *ETSU, Johnson City, TN*

10.1136/jim-2018-000974.143

Introduction Exacerbation of Crohn's disease can be caused by various triggers, infection being one of the major causes. Pathogens commonly implicated include *Shigella*, *Salmonella* spp, *Campylobacter jejuni* and *Clostridium difficile*. We present the rare case of exacerbation of Crohn's secondary to infection with *Plesiomonas shigelloides*, a bacterium that is now emerging as a cause of enteric disease in humans following the consumption of raw seafood.

Case report A 31-year-old male patient with Crohn's disease presented to the hospital with abdominal pain, vomiting and diarrhea of 1 week duration following a trip to Florida. Diarrhea was non-bloody and watery. Pain was in the right lower quadrant, non-radiating, intermittent in nature. Physical examination revealed hyperactive bowel sounds and tenderness in the right lower quadrant without guarding or rigidity. Computed Tomography (CT) scan of the abdomen showed thickening and narrowing of the distal ileum and moderate grade strictures representing acute on chronic sequela of distal ileal Crohn's disease. He was started on steroids and was empirically treated with Zosyn. Stools analysis was positive for *Plesiomonas shigelloides* and Zosyn was changed to Ciprofloxacin. Patient's diarrhea and pain improved significantly and he was discharged with tapering dose of oral prednisone.

Discussion *Plesiomonas shigelloides* is a facultative anaerobic gram-negative bacillus of Enterobacteriaceae group. It usually causes diarrheal illness when there is a co-infection with another pathogen, which reflects its status as a 'fellow traveler' with known pathogens, rather than being the causative agent itself. However, rarely, certain strains are capable of causing infection by themselves, like in our case. Though it usually causes a self-limited diarrheal illness, it can present with bacteremia, cellulitis and peritonitis in susceptible hosts. Management is supportive, unless patient is immunocompromised, elderly or has an extra-intestinal manifestation, which warrants antibiotic therapy and hospitalization. Clinicians should keep in mind the possibility of infection with this unusual organism in patients presenting with symptoms of traveler's diarrhea.

146 CHOLEDOCHOLITHIASIS: AN INCIDENTAL FINDING DURING ENDOSCOPIC ULTRASOUND GUIDED LIVER BIOPSY IN POST-TRANSPLANT PATIENTS

S Jadeja*, M Malespin. *UF Health Jacksonville, Jacksonville, FL*

10.1136/jim-2018-000974.144

Case report A 55-year-old male with a history of chronic kidney disease and cryptogenic cirrhosis status post orthotopic liver transplant in 2006 (on tacrolimus) presented to the Gastroenterology clinic for routine follow up. Patient denied symptoms of decompensated liver disease, but did endorse intermittent abdominal pain. Fibroscan demonstrated severe steatosis and high probability of cirrhosis. Endoscopic ultrasound guided liver biopsy (EUS-LB) showed nonalcoholic steatohepatitis (NASH) without cirrhosis and no evidence of rejection. Findings of choledocholithiasis (two stones measuring up to 3 mm in the common bile duct) were present and the patient was scheduled for endoscopic retrograde cholangiopancreatography (ERCP) for stone removal.

Liver biopsy remains the gold standard for the evaluation and management of liver diseases. It plays an essential role in diagnosing cholestatic diseases, drug-induced liver injury, liver disorders after transplant, as well as other hepatopathies. EUS-LB has shown to provide excellent diagnostic yield with lower complication rates when compared to percutaneous and transjugular approaches. This case demonstrates an additional utility of EUS-LB as it offers the advantage of same-session liver biopsy and diagnostic biliary evaluation in post-transplant patients. The finding of choledocholithiasis refers to the presence of gallstones within the common bile duct. Patients typically present with nausea, vomiting and abdominal pain while some patients can be asymptomatic. Complications of choledocholithiasis include pancreatitis, cholangitis, and secondary biliary cirrhosis. Hence, removal of stones is recommended following diagnosis. The traditional methods of liver biopsy would have undoubtedly missed this finding in our patient, potentially leading to further complications and procedures in the future.

EUS-LB is a useful, reliable, and safe approach for the evaluation and management of liver diseases. Its high diagnostic yield, low complication rate, and multiple diagnostic applications make this an excellent choice for the assessment of post-transplant patients. We presented a case in which choledocholithiasis was an incidental finding during EUS-LB prompting appropriate intervention with ERCP.

147 ACUTE EPSTEIN-BARR VIRUS HEPATITIS WITH CHOLESTASIS

H Kenninger*, NG Verne. *Tulane University Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.145

Case report A 27-year-old woman presented with a five-day history of subjective fevers and associated pruritis for the preceding 24 hours.

Temperature on arrival was 99.8F, HR 134bpm, blood pressure 138/77 mmHg, respiratory rate 20 breaths/min. Scleral icterus was present. No abdominal pain was appreciated. Labs returned with WBC 9.5 with 18.5% reactive lymphocytes on differential, Total Bilirubin 6.9, Direct Bilirubin 4.5, AST 166

(<35), ALT 185 (30–65), and Alkaline Phosphatase 131 (40–120). Abdominal Ultrasound revealed an enlarged echogenic liver with a 2.1×1.3×1.3 cm lymph node but no ductal dilatation was identified. Pruritus improved with ursodiol. She was discharged follow up in GI clinic the next day.

Four days later, the patient returned to ED with ongoing subjective fevers, malaise, dark urine, and elevated LFTs. Temperature was 99.6F, heart rate was 93 and she was again normotensive. Petechiae were noted on the posterior pharynx. No scleral icterus, hepatomegaly, or lymphadenopathy was appreciated. Lab work returned with WBC 21.0 with 5.8% reactive lymphocytes, Total Bilirubin 3.4, AST 338, ALT 408, Alkaline phosphatase 194. Anti-Smooth muscle antibody was positive at 34. EBV IgM and IgG were also positive. Liver biopsy returned with cholestatic pattern. Acute EBV infection and Autoimmune hepatitis (AIH) were considerations. Final pathology report was most consistent with EBV. The patient was discharged to home with a diagnosis of Acute EBV hepatitis with a cholestatic pattern and close follow-up was arranged.

Discussion Epstein-Barr Virus (EBV) is a common human herpes virus with a prevalence of close to 90% in the world population. EBV infection generally presents with a constellation of non-specific symptoms such as malaise, fevers, nausea, and lymphadenopathy. Up to 80% of patients with acute EBV infection will have an asymptomatic, self-limited rise in transaminases during the course of their infection. Less than 5% of patients with EBV infection have icterus and cholestasis. EBV induced hepatitis must be considered on the differential in the patient who presents with clinical signs of acute hepatitis, cholestasis, and only mildly elevated hepatic transaminases. The exact mechanism that produces cholestasis in EBV infection is unclear but is thought to be primarily immune-mediated.

148 NODULAR GASTRIC ANTRAL VASCULAR ECTASIA

P Kudravalli*, S Saleem, S Rawlins. *SUNY Upstate Medical University, Syracuse, NY*

10.1136/jim-2018-000974.146

Introduction Gastric antral vascular ectasia (GAVE) is the underlying cause for 4% of non-variceal upper GI bleeding. Nodular GAVE and gastric hyperplastic polyps have similar appearance on upper GI endoscopy (EGD) as well as histology which could delay specific targeted therapy. We herein, through this case would like to highlight that high clinical suspicion is required to diagnose nodular GAVE.

Case A 70-year-old male with a past medical history significant for CAD s/p DES on plavix, mechanical AVR on warfarin, iron deficiency anemia was admitted for the evaluation of fatigue and melena for a month. Physical examination was positive for black stool. The only significant lab was a drop in hemoglobin/hematocrit (H g/dl/H%) of 10/32 to 4/12.5 and INR of 1.58.

Fibrosure was suggestive of F4 cirrhosis. EGD showed nodules in the gastric antrum which were presumptively treated as GAVE with argon plasma coagulation (APC). Surgical pathological showed reactive gastropathy and gastric polyps. Review of the past histology suggested that because of the overlap in the histopathological features of hyperplastic polyps and GAVE, they were misinterpreted as hyperplastic polyp rather than nodular GAVE in 2015.

Abstract 148 Figure 1

Discussion GAVE can be classified endoscopically as punctate, striped, nodular or polypoidal form. The light microscopic findings considered specific to GAVE do not differentiate GAVE from hyperplastic gastric polyp. The first line of treatment for GAVE is endoscopic ablation with Nd:YAG-laser or APC.

Conclusion Oftentimes, there is a delay in the diagnosis and treatment of nodular GAVE as the histopathological appearance could be similar to gastric polyps. Misdiagnosis of nodular GAVE can delay targeted therapy and have fatal outcomes.

149 AUTOIMMUNE HEMOLYTIC ANEMIA IN THE SETTING OF ULCERATIVE COLITIS FLARE; A RARE ASSOCIATION

D Markabawi*, H Dharia, M Lee. *SUNY Upstate, Syracuse, NY*

10.1136/jim-2018-000974.147

Introduction Ulcerative colitis is a chronic inflammatory disease limited to the mucosal layer of the colon that almost always involves the rectum. It is commonly associated with extra-intestinal manifestations. Autoimmune hemolytic anemia is a disease caused by autoantibodies against RBCs leading to destruction of antibody-coated RBCs. The association between AIHA and UC is rare and has been seldom reported.

Case A 29 year old man with a 10 year history of ulcerative colitis, presented to the ER with pallor and exertional dyspnea 2 weeks after starting Mesalamine for a recent UC flare. Labs revealed severe anemia with a Hgb of 5.8 g/dl and Hct of 17.7, MCV of 120 fl, RDW of 24.8 and a reticulocyte index of 6.57. Total bilirubin was elevated at 3.7 mg/dl. Direct bilirubin was 0.5 mg/dl, LDH was 832 U/L and Haptoglobin was <15 mg/dl. Ferritin, iron studies, folate and vitamin B12 levels were all within normal limits. Hemolytic anemia was suspected. Coombs test was positive for anti IgG, negative for anti C3 and elution was positive with all cells. This was consistent with warm AIHA due to autoantibodies. Workup for secondary causes of AIHA was negative (ANA, RF, IgM to EBV and mycoplasma, CT thorax/abdomen/pelvis to screen for lymphoproliferative disorder). Colonoscopy showed mild colitis. Pathology showed colitis without granuloma formation. His AIHA was felt to be associated with UC flare. He was started on Prednisone at a dose of 120 mg daily. He was discharged after 2 days.

Steroids were tapered off over 3 months. Repeat hemoglobin at 6 months was 15.3 g/dl.

Discussion The prevalence of AIHA in patients with UC was reported to be 150/100000. Studies found that AIHA was more common in patients with UC involving the colon. There is little consensus regarding treatment of AIHA associated with UC. High dose systemic steroids is the first step. Cases refractory to steroids require better control of UC and stronger immunosuppression. Surgery appears to control AIHA in cases of UC refractory to medical therapy and requiring total procto-colectomy. In cases of hemolysis where active UC is completely ruled out, treatment of primary AIHA is appropriate. Rituximab and even splenectomy can be attempted.

150

SPLENIC INFARCTION AS A RARE MANIFESTATION OF CELIAC DISEASE

AA Masud*, R Chandra. *Texas Tech University, Amarillo, TX*

10.1136/jim-2018-000974.148

Case report A 55 year old Caucasian man came with chief complaint of progressive fatigue, shortness of breath and left upper quadrant pain for 6 months. There was no history of hematemesis, melena or urinary bleeding. He had no past medical or surgical history. He had a family history of anemia in father and sister. He was a smoker but did not use alcohol or IV drugs. He had weight loss in last six months. On examination: vitals signs were stable, no fever. He was pale. He had a normal cardiac, lung and abdomen exam. No hepatosplenomegaly and no lymphadenopathy. No skin rash.

Labs showed a microcytic anemia with Hb of 8, MCV 76, and platelets 5 00 000. Peripheral smear showed iron deficiency. Inflammatory marker CRP was elevated. Though iron deficient his ferritin was elevated as an acute phase reactant. CT abdomen done for abdominal pain revealed wedge shaped splenic infarcts, no significant lymphadenopathy or masses.

Investigation of splenic thrombosis: Hb electrophoresis was negative for any hemoglobinopathy. Alpha thalassemia was also ruled out by gene test. ECHO was negative for any clots or vegetations. ANA was negative.

Upper GI endoscopy showed duodenal scalloping of celiac disease, biopsy of this confirmed celiac disease. Colonoscopy showed no malignancy. Anti Gliadin antibodies were 10 fold elevated. Homocysteine level was elevated at 17. Thrombocytosis was from iron deficiency. The bone marrow had no stainable iron, and had no lymphoproliferative disorder. Flow cytometry was normal.

A diagnosis of celiac disease causing splenic infarcts was made.

Discussion Splenic infarction in celiac disease is multifactorial in etiology. Low grade reactive thrombocytosis of iron deficiency alone would not typically cause splenic thrombosis. An additional prothrombotic factor is inflammatory state produced by antigliadin antibodies Hyperhomocysteinemia from concomitant folate and B12 deficiency of malabsorption also contributes.

Literature has a few case reports of celiac disease induced thrombosis in unusual regions like the cerebral venous sinuses, hepatic veins, retinal veins. Protein S deficiency and development of antiphospholipid antibodies in celiac disease have also been described in some cases.

151

A 52-YEAR-OLD MAN WITH KLEBSIELLA PYOGENIC LIVER ABSCESS AND METASTATIC PLEURAL EMPYEMA

N Merza*, D Lin, W Hafeez, T Naguib, M Saadaldin. *Texas Tech Univ HSC, Amarillo, TX*

10.1136/jim-2018-000974.149

Case report Klebsiella liver abscesses are most often seen in middle-aged, diabetic men living in Southeast Asia and are notable for metastatic complications such as endophthalmitis and meningitis. Metastatic pleural empyema from liver abscesses is typically due to amoebic liver abscesses as opposed to E.coli or polymicrobial sources. There have been few cases of Klebsiella liver abscesses in the United States, although the number is rising. We report a rare case of a 52-year-old Caucasian male with no history of diabetes mellitus admitted for Klebsiella liver abscess with metastatic pleural empyema.

A 52-year-old nondiabetic Caucasian male with history of laparoscopic cholecystectomy and no recent travel history presented with constant, right upper quadrant abdominal pain radiating to the back for the past 3 months. He was afebrile with no leukocytosis. One month prior to his presentation, a CT-Abdomen showed a complex fluid collection posterior to the right hepatic lobe that measured 3×2.5 cm, which was increased to 5×3.1×4.8 cm on imaging on admission.

He had a CT-guided drainage 5 days later, a day after the empiric antibiotics started, and culture grew *K. pneumoniae*. The patient began exhibiting dyspnea with decreased breath sounds on the right lung base associated with fever and leukocytosis of 16 000. Chest x-ray (CXR) showed complete white-out of the right lower lobe of the lung, suggesting a large right pleural effusion. The patient underwent CT-guided pleural fluid drainage with pigtail chest tube placement. Pleural fluid analysis showed a lactate dehydrogenase of 2628 and a total protein of 4.7 g/dL consistent with exudate. The culture of the pleural fluid also grew *K. pneumoniae*. A course of Zosyn was initiated for a total of 4 weeks from the day of the abscess drain placement. Repeat CXR showed scalloping at the right lung apex with loculated component to the pleural empyema. The patient exhibited symptomatic improvement with significant drainage of pleural fluid. He was seen in clinic approximately 6 weeks after his initial admission with significant symptomatic improvement. He was placed on oral ciprofloxacin and long term follow-up with a repeat imaging.

152

METASTATIC GOBLET CELL ADENOCARCINOMA PRESENTING AS RECURRENT PARTIAL SMALL BOWEL OBSTRUCTION

¹NP Nair*, ¹T Aasen, ²M Zayko, ¹L Schmidt, ²R Velilla. ¹East Tennessee State University, Johnson City, TN; ²ETSU, Johnson City, TN

10.1136/jim-2018-000974.150

Case report Goblet cell carcinoid tumor of the appendix (GCTA) is a rare appendiceal neoplasm, displaying features of neuroendocrine tumor and adenocarcinoma. The common clinical presentations are acute appendicitis (22.5%), asymptomatic (5.4%), generalized abdominal pain (5.15%) and appendiceal mass (3.09%). GTCA presenting as small bowel obstruction is very rare, with very few cases reported so far.

A 77 year-old male presented with nausea, vomiting and constipation for two days. He was previously admitted with

similar complaints and conservatively managed for a partial small bowel obstruction (SBO) diagnosed on abdominal X-ray. He denied having abdominal surgeries, family history of colon cancer or inflammatory bowel disease. Physical exam showed abdominal distention and hyperactive bowel sounds. Abdominal CT scan showed partial SBO at the terminal ileum. Fluid passed from ileum to colon, thus conservative management was utilized. Once symptoms improved, colonoscopy was performed which showed a terminal ileal stricture 7 cm from the ileocecal valve with no abnormalities on the luminal aspect. The patient subsequently underwent exploratory laparotomy that revealed adhesions between distal small bowel and an adjacent ill-defined 4.5 cm mass, and multiple peritoneal implants. The right colon and involved small bowel were resected. Microscopic examination of the mass revealed a metastatic, moderately differentiated goblet cell adenocarcinoma (Grade 2), positive for neuroendocrine immunohistochemical stains.

GCTA is an infiltrative, malignant tumor that accounts for less than 5% of primary appendiceal tumors. It typically involves the entire appendix but may arise and spread from an effaced appendix like in our case, further contributing to diagnostic difficulty. Once GCTAs extend beyond the appendix, they are classified as adenocarcinomas, and are graded and staged as such. GCTA metastasizes in 15%–30% of cases, compared to 2%–5% of classic appendiceal carcinoids. It is imperative to consider the possibility of GCTA in patients with SBO, especially those with a transition point in the terminal ileum. A thorough evaluation of the appendix is essential for early detection and prevention of metastatic disease.

153 SYMMERS' ITCH

C Oglesby*, H Catherine, B Spieler, LS Engel. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.151

Introduction Schistosomiasis is an endemic snail-borne parasitic disease associated with exposure to freshwater in various parts of the world including Africa, Southeast Asia and South America. This disease can present as an acute infection, manifesting with pruritus, fever, angioedema, arthralgias, dry cough, diarrhea and headache or as an indolent infection with resultant periportal fibrosis, granulomatous inflammation in the bladder, and inflammation of the bowel. The pathology associated with chronic infection of the liver is known as 'Symmers' pipestem fibrosis', which leads to significant portal hypertension.

Case A 52 year old Brazilian man presented with painless rectal bleeding of 5 months duration. On initial history, the patient reported no previous medical history and he was not taking any medications. On further interview, he inferred an isolated infection with schistosomiasis 20 years prior that was treated with a medication that he could not identify. On admission, he was found to be pancytopenic with elevated INR and liver enzymes. Triple phase contrast computed tomography of his abdomen revealed hepatosplenomegaly with portal hypertension. He was found to have MELD-Na 17.7 and Childs-Pugh Class B. Endoscopy revealed several polyps with no gross parasites. Subsequent testing for autoimmune and parasitic causes revealed that the patient had schistosomiasis. At the time of hospitalization, the cause of his insidious cirrhosis remained unclear (treatment failure or re-infection).

Discussion Symmers' periportal fibrosis represents longstanding schistosomal infection secondary to *Schistosoma mansoni* that progresses to cirrhosis if left untreated. In earlier stages of cirrhosis, treatment is praziquantel or other anti-parasitic regimens. In its advanced stages, the cirrhotic liver damage is irreversible and is associated with multiple life-threatening processes such as variceal bleeding, spontaneous bacterial peritonitis, hepatic encephalopathy and secondary organ failure. These complications are mitigated by vaccinations and medications to reduce the burden of disease such as diuretics, beta-blockers, and antibiotics. Failure to respond to therapy necessitates interventional measures to reduce portal hypertension and ultimately liver transplantation.

154 KINK IN THE HOSE

C Oglesby*, H Catherine, LS Engel, D Raines. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.152

Introduction Pancreatitis is an acute inflammatory process that is caused most commonly by gallstones or alcohol. Less common etiologies of pancreatitis include hypertriglyceridemia, malignancy, instrumentation via cannulation during ERCP, anatomical variants such as pancreatic divisum or presence of an ansa-loop. General management is aimed at supportive care with intravenous fluids, pain management, and early enteral feedings.

Case A 39-year-old African American man with a history of pancreatitis secondary to alcohol use and pancreatic pseudocyst came to the hospital with epigastric pain that was similar to previous episodes but now he had the sudden development of ascites over a course of two days. He reported continued abstinence from alcohol. A paracentesis was performed and revealed a SAAG <1.1, a WBC of 750 cells/mm³ and an unremarkable cytology result. The cause of the acute ascites was thought to be due to an episode of pancreatitis that led to ductal disruption and persistent leak of pancreatic enzymes into his abdomen. He was scheduled for ERCP for major pancreatic duct stent placement, but this was complicated by the anatomical variant of the patient's pancreatic duct, known as an ansa-loop. After multiple unsuccessful attempts at cannulation, the patient eventually had a stent placed in the major papilla to alleviate increased resistance leading to a retrograde flow of pancreatic enzymatic fluid. There have since been no complications or recurrence of pancreatic ascites.

Discussion Pancreatic ascites is a rare cause of ascites, identified by evaluation of the ascitic fluid which reveals a SAAG <1.1 and clinical as well as laboratory evidence to suggest this as a source. The leak of pancreatic enzymatic fluid is caused by a ductal disruption in the major or minor pancreatic duct, or rupture of a pancreatic pseudocyst. Treatment involves symptomatic relief of ascites via paracentesis with consideration of antibiotics to cover for spontaneous bacterial peritonitis. To prevent recurrence, an ERCP is performed with placement of stent in the major pancreatic duct to relieve retrograde pressure. This procedure can be complicated further if there is an anatomical variant, such as an ansa-loop, in which case, the minor pancreatic duct will need to be cannulated and stented.

155

CURIOUS CASE OF FAP

C Oglesby*, S Veerisetty, G LaBier, LS Engel, D Raines. *LSU Health Sciences Center, New Orleans, LA*

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Introduction Familial adenomatous polyposis (FAP) is an autosomal dominant condition that carries an increased risk of colon cancer. Patients are typically asymptomatic at presentation and are on average in their fourth decade of life at age of diagnosis. Polyps in the colon are in the hundreds to thousands in number with extracolonic manifestations found in the stomach, as fundic gland polyps, and duodenal adenomas with predilection for periampullary lesions.

Case A 36 year old African American woman presented to the Emergency Department with one week of cramping abdominal pain, constipation, and right flank pain. She was previously evaluated at an urgent care, at which time, she was given omeprazole and had a right upper quadrant ultrasound. The results of the ultrasound were still pending at time of presentation. On admission she was found to have circumferential thickening at the hepatic flexure and multiple hypodense lesions in the liver and lung concerning for metastatic disease. Although the patient's family history was negative for colorectal malignancy, she was unaware of her paternal heritage. She subsequently underwent a colonoscopy to evaluate for malignancy and obstructive disease. She was found to have numerous polyps throughout her colon without obstruction.

Discussion Familial adenomatous polyposis, caused by germline mutations of the adenomatous polyposis coli (APC) gene is highly associated with the development of colorectal cancer by the age of 40 if not caught early and treated appropriately. Treatment includes colectomy with yearly upper endoscopies using the Spigelman classification for duodenal adenomatous polyps to risk stratify patients for balloon assisted enteroscopy. Furthermore, first degree relatives of patients with FAP should be screened for this disease via endoscopy and genetic testing given its autosomal dominance pattern.

156

A RARE MANIFESTATION OF CMV HEPATITIS IN AN IMMUNOCOMPETENT HOST: REPORT OF A CASE

S Parajulee*, N Adhikari, GD Bedanie. *Texas Tech University Health Sciences Center, Lubbock, TX*

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Case report Cytomegalovirus (CMV) is a common virus in the herpesvirus family (Herpesviridae). Cytomegalovirus, also called human herpesvirus 5 (HHV-5), alters innate and adaptive immunity in the host and interferes with the antigenic T cell surveillance and immune function. CMV infection is usually asymptomatic and self-limiting illness in an immunocompetent host. It usually causes more serious infections in the immunocompromised host, although the severity varies with the degree of immunosuppression. Symptoms of CMV may include fever, night sweats, fatigue, swollen glands, muscle and joint pains, blurred vision, and personality change. It can cause up to 7% of cases of mononucleosis syndrome and has shown to manifest symptoms that are similar to those for Epstein-Barr virus-induced mononucleosis. CMV can present

in immunocompetent individuals, but it usually presents with this mononucleosis with no other complications. It is less common to have liver involvement in persons who are immunocompetent. We report a case of 23-year-old immunocompetent male in Lubbock, Texas who presented with fever for two weeks, associated with hepatosplenomegaly and maculopapular rash associated with amoxicillin use with a significantly delayed elevation in transaminases who was found to have CMV infection with hepatitis. He presented initially with fever of unknown source and after a week of hospital care he began to have an increase in his transaminases, but the patient was completely asymptomatic. The patient clinically improved with supportive treatment. Our report signifies the importance of consideration of CMV infection as a cause of unexplained fever in a young adult associated with hepatitis.

157

IS THE RIGHT SIDE ALL RIGHT? – A MUST ASK BEFORE TRANSJUGULAR INTRAHEPATIC PORTOSYSTEMIC SHUNT

R Parikh, S Kovelamudi*, G Krishnan, K Bano, M Konda, D Raghavan. *UAMS, Little Rock, AR*

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Case report Transjugular intrahepatic portosystemic shunt (TIPS) is now a widely accepted modality of treatment in management of patients with complications of cirrhosis and portal hypertension. We present an unfortunate case of a patient who developed right-sided heart failure, a rare but fatal complication of TIPS.

A 68 year old man with cirrhosis (alcohol and chronic hepatitis C) and prior TIPS, hypertension and atrial fibrillation presented with coffee ground emesis, melena and atrial fibrillation with rapid ventricular response. He was not on anticoagulants because of recurrent variceal bleeds. Endoscopy showed one large varix in distal esophagus that was banded, severe portal hypertensive gastropathy and several varices in the rectum. Duplex ultrasound showed decreased flow through TIPS. He underwent recanalization of occluded TIPS and subsequently developed acute pulmonary edema, right heart failure, cardiogenic shock and non-oliguric renal failure requiring intubation and multiple vasopressors. TIPS was emergently downsized with consequent improvement in his hemodynamics. Right heart catheterization showed severe pulmonary hypertension. Retrospectively, we found that patient had ongoing cough and exertional dyspnea for a few years. Review of prior imaging revealed evidence of severe interstitial lung disease with honeycombing, predominantly in upper lobes, which had progressed significantly. Differential diagnoses were usual interstitial pneumonia (UIP)/idiopathic pulmonary fibrosis (IPF) or hypersensitivity pneumonitis (he worked as a spray painter). He was diuresed aggressively, also given a trial of prednisone and sildenafil. After an extended stay in the hospital, his respiratory status failed to improve. He was transitioned to hospice care and passed away.

TIPS is known to worsen the hyperdynamic circulatory state in cirrhotics. Presence of moderate to severe pulmonary hypertension is a contraindication to TIPS. Model for End-Stage Liver Disease (MELD) score can be used to predict post-TIPS outcome. We aim to reinforce the need for strict pre-procedural screening. Studies to assess the incidence and risk factors of this complication are needed.

158 A JAUNDICED 86 YEAR OLD MAN WITH 40 POUND WEIGHT LOSS

R Patel*, R Freedman, A Zirulnik, J House. *University of Florida College of Medicine Jacksonville, Jacksonville, FL*

10.1136/jim-2018-000974.156

Introduction We report a case of a large hepatic cyst that presented in an elderly patient with diffuse jaundice, early satiety, and profound weight loss to highlight an uncommon differential diagnosis when considering progressive jaundice and ominous weight loss in patients.

Case An 86-year-old retired mechanic presented with jaundice, abdominal distention, generalized pruritus and a 40 pound weight loss over the last 6 months that he attributes to early satiety. He also reported dark colored urine for the past week and regular bowel movements with normal colored stool. On physical exam, this patient appeared to be a healthy 86 year old African-American male with generalized yellowing of his skin, eyes, and mucus membranes. Laboratory results showed an AST/ALT of 243 and 256 IU/L respectively. His total bilirubin was 7.0 mg/dL with a direct amount of 5.9 mg/dL. Alkaline phosphatase was markedly elevated at 1,425 IU/L. He was hepatitis A, B, C negative. An initial right upper quadrant ultrasound found two well defined, anechoic hepatic cysts with the dominant cyst measuring roughly 17 cm in diameter. There was also dilation of the distal common bile duct of approximately 1.1 cm. A follow-up CT abdomen confirmed the hepatic cysts nearly occupying the entire right lobe of the liver. Interventional radiology was consulted and recommended a therapeutic percutaneous hepatic cyst drainage of the dominant cyst which ultimately drained 1.7 liters of serous fluid with an accordion drain left in place.

Discussion While most hepatic cysts are benign, our case demonstrated, large hepatic simple cysts are more likely to be symptomatic or produce complications. The common complications include biliary obstruction, as demonstrated by our patient, as well as spontaneous rupture, infection, hemorrhage, or torsion around the base of a pedunculated lesion.

Conclusion The prognosis of our patient is excellent with exception of the risk of recurrence. What initially presented clinically as a forbidding list of deadly diseases with a poor prognosis, such as pancreatic adenocarcinoma or biliary carcinoma, ultimately turned into a benign diagnosis with near complete resolution of symptoms seemingly overnight.

159 METASTATIC NEUROENDOCRINE CARCINOMA OF THE COLON IN A YOUNG WOMAN

M Plummer*, M Parsi. *Tulane University Health Sciences Center, New Orleans, LA*

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Case report A previously healthy 30 year-old woman presented to the emergency department with a 3 week history of nausea, vomiting, abdominal distention and maroon colored stools. Her family history was significant for colon cancer in her paternal uncle, diagnosed at the age of 59. A CT of the abdomen showed a mass in the transverse colon with innumerable liver lesions suggestive of metastasis.

Colonoscopy showed multiple polyps throughout the colon, some covering more than 75% of the circumferential surface. Surgery was consulted and she underwent a right hemicolectomy. The resected specimen had a 6.6 cm small cell neuroendocrine carcinoma (NEC) with lymphovascular invasion arising from a tubulovillous adenoma in the hepatic flexure. It also had a 3.5 cm intramucosal adenocarcinoma in the transverse colon arising from a tubulovillous adenoma along with multiple other tubular adenomas. Regional lymph nodes were negative for malignancy, but multiple tumor deposits were noted in the subserosa and mesentery. Outpatient oncology follow-up was arranged to discuss palliative treatment options.

Neuroendocrine carcinoma of the colon is rare, accounting for less than 1% of colon cancer cases. The median age of diagnosis is 70 years. NECs of the colon are extremely aggressive with more than 60% of cases metastatic at diagnosis, most commonly to the liver. Based on retrospective data, tumor resection has not been shown to improve outcome. Chemotherapy, however, has been found to increase median survival time. Regardless of the stage of disease at diagnosis, prognosis is poor with an estimated, combined 5 year survival rate of 8%. The patient-specific details make this case uniquely different. First, the patient's age stands apart and raises suspicion for an inherited predisposition for malignancy given the numerous tubular adenomas and presence of a second primary colon cancer. Second, her small cell NEC was found to arise from a tubulovillous adenoma, a rare phenomenon. One retrospective study of 100 patients with NECs noted that adenomatous polyps were intimately related with tumor formation in 30% of cases. Lastly, the presence of a synchronous, second primary malignancy in patients with NEC is extremely rare. In summary, NEC of the colon is a rare, aggressive malignancy with poor prognosis despite therapy.

160 NITROFURANTOIN INDUCED LIVER INJURY: A RARE COMPLICATION OF A COMMONLY PRESCRIBED MEDICATION

D Radadiya*, N Nair, J Phemister, J McKinney. *East Tennessee State University, Johnson City, TN*

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Introduction Drug-induced liver injury (DILI) can occur from a wide range of medications. Nitrofurantoin, a common antibiotic prescribed for urinary tract infections (UTI), can rarely cause DILI, leading to serious sequelae such as acute liver failure. The presentation can be indistinguishable from autoimmune hepatitis. Here we present a case report of the patient presenting with new onset jaundice following treatment with nitrofurantoin.

Case description An 82-year-old female with the history of recurrent UTI presented with new onset jaundice and pruritus for 1 week. She denied prior liver disease, blood transfusions, alcohol use, recent travel or sick contacts. Surgical history was significant for remote cholecystectomy. Medications included alendronate, allopurinol, amlodipine, and nitrofurantoin that was started a week ago for UTI. Physical exam was unremarkable except for generalized jaundice and icteric sclera. No ascites or encephalopathy was observed.

Laboratory tests revealed AST 53 IU/L, ALT 33 IU/L, ALP 367 IU/L, total bilirubin 19.9 mg/dL, direct bilirubin 7.7 mg/dL, albumin 3.0 g/dL, and INR 1.3. Viral hepatitis serology and secondary hepatitis markers were negative. Ultrasound of the liver showed coarse echotexture without any obvious etiology. MRCP showed no common bile duct dilation. After ruling out all other causes, thorough history was re-taken. The patient reported similar reaction with nitrofurantoin about 7 years ago. Nitrofurantoin was immediately stopped and UTI was treated with Ertapenem. Over time, liver enzymes trended down slowly. The patient was asymptomatic 3 months following discharge with complete normalization of liver enzymes.

Discussion Nitrofurantoin, a frequently prescribed antibiotic for the treatment of uncomplicated UTI, can rarely cause DILI. Overall prognosis is variable, mainly influenced by the stage of disease at diagnosis. Therefore, clinicians need to be aware of this adverse effect of Nitrofurantoin since early recognition and prompt withdrawal of the drug halts inflammation and prevents progression to end-stage disease.

161 BEWARE OF THE J-STOMACH: IMPLICATIONS FOR MISINTERPRETATION OF A GASTRIC EMPTYING SCINTIGRAPHY

¹S Reddy*, ²R Bou Said, ²J Diaz, ²R McCallum. ¹TUHS, Paul L. Foster School of Medicine, El Paso, TX; ²TUHS, El Paso, TX

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Case report The purpose of this case report is to raise awareness of the J-shaped stomach, an entity that can alter the interpretation of radionuclide gastric emptying (GE) studies and effect clinical decision-making.

A 46 year old female was referred to our motility center for abdominal pain, distention, nausea, and intermittent vomiting after meals. She had inconsistent GE results from previous medical centers, but the working diagnosis was gastroparesis. Therefore, our goal was to attain the status of her GE before considering any therapies.

The standard egg-beater meal could not be tolerated, so the patient ingested 8 FL oz of nutritional Ensure Supplement (250 calories) labeled with T99m-Sulfur colloid. Anterior and posterior images of the stomach were obtained in a standing position at 30 min, and then hourly over 4 hours with the geometric mean being calculated. Based on initial ROI calculations, early rapid GE indicated dumping syndrome. However, after re-examination of the images, the ROIs were altered after images of isotope ascending from a vertically elongated stomach were identified. The repeat calculations revealed moderate gastroparesis: 23% retained at 4 hours (normal <10%). An Upper GI series confirmed the diagnosis of a J-shaped stomach, with the gastric body located in the pelvic cavity, and the antrum ascending to join the duodenum.

When a patient's past history includes inconsistent GE interpretations, this raises suspicion for the entity, the J-shaped stomach warranting a more detailed re-examination of the scintigraphic images, and consideration for an Upper GI series. The prevalence of a J-shaped stomach is unknown, but awareness of its existence is necessary as it can significantly alter the diagnosis obtained by a scintigraphic GE study and hence treatment decisions.

162 AN UNUSUAL CAUSE OF RIGHT LOWER QUADRANT PAIN IN A YOUNG FEMALE PATIENT

C Rosero*, J Goldstein. ETSU, Johnson City, TN

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Case report Acute diverticulitis is the inflammation of colonic mucosal outpouchings commonly observed in the adult population. Right-sided involvement is an unusual presentation and typically affects Asian or African American patients. We present a case of acute right-sided diverticulitis mimicking acute appendicitis in a young female patient.

A 22-year-old otherwise healthy young woman presented to the emergency department (ED) complaining of worsening right lower quadrant pain that started the day before admission. Accompanied by nausea, emesis, anorexia and fever, no diarrhea or constipation. Last menstrual period, two weeks before onset. On physical exam, her vital signs were stable. There was significant tenderness to palpation in the right lower quadrant and flank. No rebound or McBurney's point tenderness and decreased bowel sounds. Otherwise unremarkable.

Laboratory workup revealed only leukocytosis (16.4 K/uL) and urine pregnancy test was negative. Abdominal computed tomography with intravenous contrast described focal thickening and diverticula involving the right hepatic flexure, concerning for acute diverticulitis. Additionally, a grossly normal sized appendix surrounded by inflammatory fluid not related to acute appendicitis was reported.

The patient was started on intravenous broad-spectrum antibiotics and fluids, pain control and oral intake restriction. Her abdominal pain gradually decreased and was able to tolerate an oral diet. She was discharged home with a scheduled outpatient Gastroenterology appointment for a colonoscopy.

Acute diverticulitis is a disease commonly observed in middle-aged people with risk factors including low fiber diet, sedentary lifestyle and obesity and usually involves the sigmoid colon. Right colonic involvement has been occasionally reported in people of African American and Asian race. Our patient's age and overall health status did not suggest the possibility of diverticulitis as the etiology of her symptoms. This case presents right-sided diverticulitis as an unexpected cause of right lower quadrant pain in a healthy young woman. It illustrates a potential differential diagnosis to be considered in order to avoid unnecessary treatments or potential complications.

163 GLYCOGENIC HEPATOPATHY, A RARE COMPLICATION OF TYPE 1 DIABETES MELLITUS

B Sharma*, M Antoine, M Shah, S John. SUNY Upstate Medical University, Syracuse, NY

10.1136/jim-2018-000974.161

Introduction Glycogenic hepatopathy (GH) is excessive intrahepatic glycogen accumulation that manifests as hepatomegaly with transient elevation in liver transaminases. It is a rare complication of long-standing poorly controlled type 1 Diabetes Mellitus (DM1). GH is often seen in patients who present with frequent diabetic ketoacidosis (DKA) as these patients are usually hyperglycemic and treated with high doses of insulin leading to excessive storage of glycogen in their liver.

Case A 19-year-old female with history of poorly controlled DM1 and frequent admissions for DKA presented with abdominal pain, nausea, vomiting and hepatomegaly. Labs were significant for serum bicarbonate 9, anion gap (AG) 38, beta-hydroxybutyrate (BHB) 5.2, lactic acid (LA) 3.3 and glycosylated hemoglobin (HbA1c) 12.1%. Liver function was significant for ALT 29, AST 53, ALP 166 with normal bilirubin and prothrombin time. She was treated with insulin infusions and IV fluids for DKA, however, her AG continued to be elevated despite normal BHB. LA was rechecked and came back at 8.8 mmol/L. CT abdomen showed hepatomegaly. Work up for hepatitis A, B, C, CMV, EBV, HIV, autoimmune hepatitis, hemochromatosis, Wilson's disease and celiac disease were negative. She underwent a liver biopsy which showed diffuse swelling of hepatocytes and marked increase of intracytoplasmic glycogen consistent with GH. She was counseled on diabetic diet and proper insulin use to achieve strict glycemic control and was discharged home with a close follow up.

Discussion GH usually presents with abdominal pain, nausea, vomiting and hepatomegaly. Lab work up often shows elevated liver transaminases, elevated HbA1c which reflects poor long-term glycemic control and sometimes elevated serum lactate. Infectious, autoimmune and metabolic causes of liver injury are ruled out first. Imaging including ultrasound and CT is usually nonspecific showing hepatomegaly. GH is often confused with non-alcoholic fatty liver disease as it can present similarly and appear similar on imaging. The gold standard to confirm the diagnosis is liver biopsy, however, it is invasive and leads to GH to be underdiagnosed. Treatment involves tight glycemic control which usually results in reversal of the disease. Pancreatic transplantation has been reported in severe cases.

164 MESALAMINE-INDUCED LIVER INJURY, A RARE PRESENTATION

B Sharma*, M Antoine, M Shah, H Hegazy. *SUNY Upstate Medical University, Syracuse, NY*

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Introduction Mesalamine is a 5-aminosalicylate used for inflammatory bowel disease flare ups and maintenance. It has an anti-inflammatory effect that it achieves by inhibiting leukotriene production leading to lower levels of Interleukin-1 and tumor necrosis factor alpha. Common side effects include nausea, vomiting and diarrhea. Hepatotoxicity has been reported, however it is rare with incidence of clinical liver disease at 3.2 cases per million prescriptions.

Case A 54-year-old female with history of Crohn's colitis diagnosed 4 weeks prior to presentation for which she was started on mesalamine 2 weeks later, presented with painless jaundice. On exam, she was clinically jaundiced. Labs were significant for elevated alkaline phosphatase (ALP) 1803, total bilirubin (TB) 12 with direct fraction (DB) 11.1. ALT and AST were also mildly elevated at 151 and 169, respectively. Albumin was 2 and INR was within normal limits. Work up for viral hepatitis was negative. Autoimmune panel was negative including anti-nuclear antibody, anti-smooth muscle antibody, anti-liver kidney microsomal antibody, anti-mitochondrial antibody and normal IgG levels. Acetaminophen and salicylate levels were negative. Abdominal ultrasound and MRCP were negative for any liver or biliary pathology. Reviewing her previous records before starting the

mesalamine showed normal liver function and given her negative work up it was believed that her presentation was secondary to mesalamine. Mesalamine was stopped on admission. Her liver function peaked on 3rd day of discontinuing mesalamine with ALP 2168, TB/DB 14.8/12.8 and ALT/AST 151/210 then started trending down. Unfortunately, on her 6th day of hospitalization, she developed a cardiac arrest of unclear etiology and expired.

Discussion Mesalamine hepatotoxicity is rare and can present as asymptomatic elevation in liver enzymes, hypersensitivity reactions or as idiosyncratic hepatocellular or cholestatic injury. The latter usually happens 1–6 months after starting therapy and the mechanism behind it is unclear. Clinicians should have high index of suspicion in patients developing hepatotoxicity after starting mesalamine as discontinuing the medication can result in improvement of liver function in a rapid manner. Further research is needed to better understand the mechanism behind such adverse reactions.

165 TURMERIC SUPPLEMENT-INDUCED HEPATOTOXICITY

FK Suhail*, U Masood, F Liu, S John. *SUNY Upstate Medical University, Syracuse, NY*

10.1136/jim-2018-000974.163

Case report A 61 year old female with benign polycystic liver disease presented with fatigue and polyarthralgias for 1 week. Review of medications did not reveal any obvious hepatotoxic drugs. Physical examination was notable for right upper quadrant pain. Laboratory findings were notable for aspartate aminotransferase of 1553 U/L, alanine aminotransferase of 2607 U/L, alkaline phosphatase of 246 U/L and total bilirubin of 1.6 U/L with a direct component of 1 U/L. Hepatic synthetic function was intact. Infectious causes were ruled out. Autoimmune work up yielded positive antinuclear antibody (1:250) with normal anti-smooth muscle antibody and serum IgG levels. Imaging was negative for portal or hepatic vein thrombosis with no evidence of biliary ductal pathology. Acetaminophen level was negative but patient reported taking turmeric supplements for 5 months. A liver biopsy showed panlobular hepatitis with early parenchymal collapse suggestive of a morphologic counterpart of acute hepatitis. The plasma cells did not outnumber the other inflammatory cells enough to favor a diagnosis of autoimmune hepatitis. Patient was thought to have DILI from turmeric pills which were discontinued. She was discharged home on prednisone. Her liver function tests (LFTs) normalized after 3 weeks and the prednisone was tapered off.

DILI accounts for 10% of cases of acute hepatitis. Most patients are asymptomatic and detected incidentally. Severe cases can lead to acute liver failure and chronic DILI can lead to fibrosis/cirrhosis. DILI is usually associated with prescribed medications but herbal supplements have increasingly been implicated and not adequately studied. The temporal association of liver injury, normalization of LFTs upon withdrawal of turmeric pills and treatment with prednisone suggested turmeric as the likely causative agent. Extensive workup performed to rule out other causes of hepatitis solidifies this conclusion. It should be noted that herbal supplements are not regulated by the Food and Drug Administration and may contain additives that could be hepatotoxic. In conclusion, this case should serve as a reminder to clinicians to keep herbal medications in the differential for DILI.

166

EXTRAHEPATIC HEPATOCELLULAR CARCINOMA METASTASIS MASQUERADING AS A FOREHEAD HEMATOMA

A Thomas*, R Patel, N Davison, C Isache. *University of Florida Health, Jacksonville, FL*

10.1136/jim-2018-000974.164

Case report A 77 year old female with history of cervical canal stenosis with cord compression presented for ambulatory neurosurgery but was sent to the ED for shortness of breath and significant abdominal distension. In the ED, abdominal imaging revealed moderate ascites and cirrhosis with multifocal hyperattenuating liver lesions, porta hepatitis, and mesenteric and anterior mediastinal lymphadenopathy highly suspicious for metastatic hepatocellular carcinoma. The patient denied history of alcohol abuse and was negative for hepatitis. Serum alpha fetoprotein was elevated at 97.9 ng/ml. Analysis of the ascites fluid revealed transudative fluid without malignant cells. The patient was also noted to have a soft tissue mass on her forehead that had been present for 4 months. She originally noted the soft tissue mass after bumping her head on a cupboard and thought it was a hematoma that persisted. CT head revealed a 3.1 cm mass that eroded through the frontal calvarium. A core needle biopsy of the mass stained positive for Hep par 1 and polyclonal CEA, which was highly suggestive of metastatic hepatocellular carcinoma. She was discharged home to follow with Oncology for possible palliative chemotherapy.

Hepatocellular carcinoma (HCC) is the most common type of primary liver cancer and is second in rank for cancer mortality worldwide. Cirrhosis secondary to hepatitis B and C are the leading risk factors for developing HCC but it has also been linked to obesity, diabetes mellitus, and non-alcoholic fatty liver disease (NAFLD). HCC carries a high mortality, with a 5 year survival rate of less than 12%. Extrahepatic metastases occur in 30%–50% of HCC cases, with the most common sites of metastasis being the lung, regional lymph nodes, and bone. Skull and cutaneous metastases are rare, especially for first presentation of HCC, but it is important to hold high clinical suspicion of an underlying primary malignancy, as early diagnosis may lead to improved quality of life with early treatment or palliation.

167

LEIOMYOMA IN SIGMOID COLON

AG Tikue*, GD Bedanie, A Rakvit. *Texas Tech University, Lubbock, TX*

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Introduction Colonic leiomyoma is rarely seen in clinical practice. It arises from muscularis mucosa, muscularis propria or vascular smooth muscles.¹ Based on literature reviews, most leiomyomas in gastrointestinal tract are found in stomach and small intestine. It is rare to find leiomyoma in colon. Colonic leiomyoma accounts only 3% of gastrointestinal leiomyomas.²

Case summary A 54-year-old woman who presented with chronic lower crampy abdominal pain. She denied vomiting, hematemesis, melena, weight loss and fever. Physical examination and laboratory examination were essentially normal. Colonoscopy showed 10 mm sessile polyp in sigmoid colon. Polypectomy was performed. Pathology showed colonic mucosa overlying well differentiated smooth muscle cells,

arising from the muscularis mucosa and arranged in fascicles. It was positive for smooth muscle actin immunohistochemical stain, supporting the diagnosis of leiomyoma. CD117 highlighted mast cells, but was negative in the smooth muscle cells.

Discussion Leiomyoma in sigmoid colon is a rare finding. Colonic Leiomyomas are generally asymptomatic. They are usually incidentally found during routine colonoscopy. However, leiomyoma can cause symptoms such as abdominal pain, intestinal obstruction, hemorrhage and bowel perforation depending on the location, size and direction of tumor growth. Leiomyoma is traditionally treated with surgical resection. But in recent years, colonic leiomyoma can be resected endoscopically, due to the development of endoscopic techniques and new devices.

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168

INTRAHEPATIC CHOLESTATIC INJURY WITH INTRAVENOUS THIRD GENERATION CEPHALOSPORIN

¹S Bijlani, ²RM Medrano, ¹M Heinrich, ²MN Vinan Vega*, ²S Koshy. ¹Texas Tech University Health Science Center, Lubbock, TX; ²TTUHSC, Lubbock, TX

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Case report Drug induced hepatotoxicity (DIH) presents similarly to many acute and chronic liver diseases. Variants include cholestatic, hepatitis, or mixed. Cholestasis results in jaundice and varying degrees of elevation of direct bilirubin (DB), alkaline phosphatase (ALP) and gamma-glutamyl transpeptidase (GGT). We present a rare case of intrahepatic cholestasis related to DIH from ceftriaxone the background of chronic alcohol abuse.

A 57 year old white male with a history of chronic heavy alcohol, admitted in septic shock, secondary to urosepsis, and initially treatment with 1 dose of vancomycin and a 3 day course of piperacillin-tazobactam. Blood and urine cultures, after were positive for *E. coli*, with adequate susceptibility to ceftriaxone, and hence piperacillin-Tazobactam was replaced with IV Ceftriaxone on the 4th day.

Total bilirubin (TB) increased from 0.9 mg/dl to 5.3 mg/dl within a span of 24 hours with continued rise a maximum value of 14.8 mg/dl on day 6 with a predominant of DB >10 mg/dl. Concomitantly the ALT and AST, that were 54 IU/L and 69 IU/L at baseline, peaked to 121 IU/L and 125 IU/L respectively on day 6. ALP increased from a baseline value of 191 IU/L to 415 IU/L with elevated GGT. There was no extrahepatic biliary obstruction with Magnetic Resonance (MR)Cholangiopancreatography showing no dilatation of common bile duct with minimal pericholecystic fluid. INR was 1.5 and serum albumin was 2 gm/dl at baseline suggesting likely chronic alcoholic liver disease. On discontinuation of Ceftriaxone on 6th day, the TB, AST, ALT and ALP steadily decreased to baseline value over a time period of 2 weeks. Though parenteral Ceftriaxone can cause

biliary sludge, intrahepatic cholestasis is rare. This is likely a result of dose independent idiosyncratic immune-allergic response that is associated with fever, rash and eosinophilia. In most cases, the recovery is within a few weeks. In summary, the recognition of DIH as in Ceftriaxone induced intrahepatic cholestatic injury requires a high index of suspicion due to many conditions masquerading its clinical presentation. Immediate removal of the offending agent is imperative to the resolution of liver injury before irreversible complications ensue.

169

NOT YOUR AVERAGE CASE OF MONONUCLEOSIS

C White*, M Hess. *SUNY Upstate Medical University, East Syracuse, NY*

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Case report Acute pancreatitis is a common cause for hospital admissions in the United States. While gallstones and alcohol use are well known precipitants, less common etiologies such as infections are often overlooked. Specifically, viral causes may be missed if the characteristic signs of the infection are overshadowed by the final diagnosis. We present a case of acute pancreatitis caused by the Epstein Barr virus (EBV).

A 59 year old previously healthy male presented to the hospital with a week of generalized fatigue, myalgias, subjective fevers, as well as one day of sharp epigastric pain associated with nausea. He had recently cycled in wooded areas in the Northeast United States and had been prescribed Doxycycline by his primary provider the previous day given concern for Lyme disease. History was also notable for a close contact with a respiratory tract infection and no recent alcohol consumption. With worsening symptoms, the patient sought urgent medical attention.

Labs on presentation showed an elevated lipase 590 U/L, ALT 192 U/L, AST 75 U/L, ALP 165 U/L, total bilirubin 0.6 mg/dL, and white blood cell count of 17 000. Platelets were decreased at 149 000 and triglycerides were normal at 63 mg/dL. Computed tomography of the abdomen showed an edematous pancreas with surrounding stranding as well as hepatosplenomegaly. Ultrasound of the gallbladder revealed no stones or sludge. The patient was therefore diagnosed with acute pancreatitis. Autoimmune work-up was unremarkable and serologies were negative for Babesia, Ehrlichia, Lyme, VZV, HSV, Coxsackie, Leptospira, as well as hepatitis A, B and C. EBV PCR subsequently returned positive indicating an acute infection. The patient clinically improved with conservative therapy.

EBV has rarely been associated with acute pancreatitis and even then has only been reported in patients less than 25 years of age. While primary EBV infections are often subclinical, others present with the classic symptoms of fever, fatigue, lymphadenopathy, and pharyngitis. Atypical presentations of EBV, such as this case, highlight the ability of this virus to infect multiple organ systems. Since approximately 15%–25% of acute pancreatitis cases are caused by unknown etiologies, expanding the work-up to include a broad infectious differential could prove to be fruitful in further defining causes of this disease.

Health Care Research, Quality Improvement and Patient Safety

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

170

INNOVATIVE APPROACH TO CANCER SCREENING HEALTH FAIRS: EDUCATION, AWARENESS, AND SCREENING WITH AN INTERDISCIPLINARY APPROACH @ TEXAS TECH HSC

¹A Bharadwaj, ²C Ashangari*, ²H Tewari, ²R Bharadwaj. ¹Amarillo High School, Amarillo, TX; ²Texas Tech HSC-Amarillo, Amarillo, TX

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Purpose of study Cancer screening remains an effective means to reach underserved and rural population. Majority of such events have limited cost and time effectiveness due to fewer types of screenings offered, depth of education, and interdisciplinary participation. At the Texas Tech Health Science Center, we have created a unique approach to overcome these deficiencies through a comprehensive approach.

Methods used We collaborated with multiple subspecialties in an interdisciplinary manner to incorporate multiple cancer screenings including colorectal, cervical, HPV vaccine, breast, skin, lung, Hepatitis C and smoking cessation. Participants were guided with the help of volunteers to go through every booth. Each activity booth provided education followed by 'Ask the Doc' session before proceeding for the screening. This way every individual who came to the event received education about every type of cancer that was offered. Multiple students, faculty, and caregivers worked in an interdisciplinary fashion from the SOM, SOP, and SON.

Summary of results Each of the 140 participants (36.46% Hispanics and 34.88% Caucasians, and 65% female) received education about every cancer type. Sixty students, 8 faculties, and 10 caregivers participated in the activity. Majority of the participants were between age 50 and 71 years (36.9%) and 67.4% were first-timers. Self-perceived importance for cancer screening was the most common reason (50.39%) to attend the event. Participants perceived an improvement in their knowledge by a score of 2.9 on the scale of 10 ($p < 0.0001$). Participants were equally interested in the different types of cancers. The most common event information source was family/friends (21.7%) followed by Facebook (20.16%) and TV/Radio (17.05%). Interestingly, Facebook was the most common source amongst females (25.30%) as compared to males (10.87%), whereas family/friends were the most common source for males (34.78%), as compared to females (14.46%).

Conclusions Integrating multiple cancer screenings and education in an interdisciplinary manner is an excellent approach to best utilize available resources for maximum community benefit with time and cost-effectiveness at an educational institution.

171

HIGH RISK OPIOID PRESCRIBING AND DISPENSING TO CHILDREN 0–18 YEARS OLD

W Basco*, J Zhang, P Mauldin, S Ball, K Simpson. *Medical University of South Carolina, Charleston, SC*

10.1136/jim-2018-000974.169

Purpose of study Previous studies demonstrated declines in overall opioid prescribing and dispensing for children, but less detail is known about high risk prescribing. This study evaluated three CDC-defined state level opioid outcomes that we considered ‘high risk:’

- the percentage of children receiving ≥ 90 morphine mg equivalents/day;
- the rate of patients receiving opioid prescriptions from multiple providers; and
- the percentage of extended-release opioid prescriptions dispensed to opioid-naïve children (CDC state-level opioid indicators #23, 24, and 25).

Methods used Using 2010–2017 South Carolina (SC) Prescription Drug Monitoring Program (PDMP) data, we identified dispensed prescriptions for opioid analgesic preparations to children 0–18 years old, excluding cough and cold opioid preparations, tramadol, and propoxyphene. We calculated the frequency of high-risk prescribing. Measures #23 and 25 are expressed as percentages, while measure #24 is an expression of opioid prescriptions per population. The SC Dept of Health and Environmental Control and the Institutional Review Board of MUSC approved this study.

Summary of results Dispensing of opioid analgesics to children 0–18 decreased 32% overall from 80/1,000 children/year in 2010 to 54/1,000 children/year in 2017. Each high-risk measure declined over time. The percentage of subjects receiving ≥ 90 morphine mg equivalents/day (Measure #23) declined from 3.8% in Q1 of 2010 to 2.9% in Q4 of 2017, a 24% decline ($p < 0.01$). For Measure #25, the percentage of subjects who received an extended-release opioid who were opioid naïve was 65.9% in Q1 of 2010, peaked at 81.8% in Q1 of 2015, and declined back to 64.6% by Q4 of 2017. Receipt of opioids from multiple providers (Measure #24) peaked at 9.59/million children/6 months in 2014, declining to 0.85/million children/6 months by 2017.

Conclusions Among children 0–18, receipt of ≥ 90 morphine mg equivalents/day and receipt of opioids from multiple providers declined during the years 2010–2017. However, fully 2/3 of the children who received extended-release opioid analgesics were opioid naïve and should not have received extended-release products intended for opioid-tolerant individuals. In addition to reducing opioid prescribing for children overall, efforts are needed to specifically reduce high risk prescribing.

172

PAIN MANAGEMENT IN PEDIATRIC PATIENTS WITH SICKLE CELL ANEMIA PRESENTING TO THE EMERGENCY DEPARTMENT: A QUALITY IMPROVEMENT PROJECT

S Blakemore*, NP Shah, J Lebensburger, K Monroe. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.170

Purpose of study About 98% children with Sickle Cell Anemia (SCA) suffer from pain secondary to vaso-occlusive crisis.

Delays in pain management in the emergency department (ED) can reduce the overall quality of care. The National Heart, Lung and Blood Institute 2014 guidelines on SCA pain management recommend that the first dose of analgesic medication be given within 30 min of triage and that subsequent doses be given every 15–30 min as needed. Currently our institution takes an average of one hour to administer the first analgesic and there is much variation in the time between subsequent doses. There is supporting evidence for decreased time to analgesia in sickle cell pain if Intranasal Fentanyl (INF) is used. We initiated a Quality Improvement project to address delays in pain management at our institution. The project aims are to improve the time to first dose as well as decrease time between doses, to decrease emergency department length of stay, and decrease ED bounce backs secondary to pain crisis.

Methods used Using a multidisciplinary team based approach involving hematologists, pharmacists, emergency department physicians and nurses, and the hospital IT department, a sickle cell pain pathway was formulated. We used the Model for Improvement Iterative plan-do-study-act (PDSA) cycle method. The first PDSA cycle involved assessing for acceptability of INF amongst sickle cell patients and families presenting in pain crisis. The second cycle involved the implementation of an updated sickle cell pain order set in our hospital's electronic medical system. This order set went live October 2, 2018 and allows the physician to order initial and subsequent doses of INF and IV Opioids to decrease the time between doses.

Summary of results About 75% of families approached accepted INF as an initial medication for pain crisis. The remaining 25% did not have sufficient information about the medication or did not like intranasal medications. We will be obtaining monthly reports to collect data to ascertain if the order set helped improve the time to analgesic administration.

Conclusions INF was found to be reasonably acceptable method of pain management in SCA patients/parents. Results of order set implementation are awaited to show evidence for effectiveness.

173

PATIENT-RELATED RISK FACTORS ASSOCIATED WITH KNEE ARTHROSCOPY

¹CJ Bloise*, ¹B Fong, ¹T Veale, ¹J Poche, ¹K Jeffers, ²C Leonardi, ¹V Dasa. ¹Louisiana State University Health Sciences Center School of Medicine, New Orleans, LA; ²Louisiana State University Health Sciences Center School of Public Health, New Orleans, LA

10.1136/jim-2018-000974.171

Purpose of study Arthroscopic knee surgery (AKS) is one of the most common orthopaedic procedures; however, few studies have examined risk factors associated with it. We sought to investigate whether patient factors are related to patient-reported outcomes prior to and following knee arthroscopy.

Methods used This was a retrospective chart review of all patients who underwent AKS from 2012 to 2017 by a single surgeon at a university-based orthopedic practice. Data collected included age, sex, BMI, race, Charlson Comorbidity Index (CCI), insurance status, and Oxford Knee Score (OKS). The OKS was obtained at the preoperative visit and at approximately 14, 42, 90, and 180 days after AKS. Multivariate analyses with OKS scores as repeated measures, and age, sex, BMI, race, CCI, and insurance status as fixed effects

were conducted as well as analyses in which preoperative OKS scores were treated as fixed effects.

Summary of results Of 318 patients who underwent AKS during the study period, 228 had complete OKS data, of whom the average age and BMI were 56 years (SD=12) and 33 kg/m² (SD=7), respectively. Patients were 64.5% female, 47.6% Caucasian, 42.5% African American, 41.2% with private insurance, 30.4% with Medicare, and 28.4% with Medicaid. Lower (worse) preoperative OKS scores were significantly associated with increasing BMI (LR -0.26, $p=0.001$) and patients with Medicaid compared to those with private insurance (-5.6, SE 1.5, $p=0.002$). Medicaid patients also had significantly lower post-surgery OKS scores compared to Medicare (-6.0, SE 1.9, $p=0.005$) and tended to be lower than privately insured patients (-3.8, SE 1.8, $p=0.09$). Age, sex, race, and CCI were not significantly associated with pre- or post-operative OKS scores.

Conclusions This study found that patient insurance status was linked to knee arthroscopy outcomes with Medicaid patients being at a greater risk for worse outcomes compared to those with Medicare and private insurance. Further investigation is warranted to ascertain the cause of these disparities and whether they can be minimized.

174

MEDICAL STUDENTS' EXPERIENCE AS INTERPRETERS IN THE CLINICAL SETTING

¹S Dahotre*, ²J Sundaresan, ¹E Omoruyi. ¹McGovern Medical School, Houston, TX; ²University of Texas Medical Branch, Galveston, TX

10.1136/jim-2018-000974.172

Purpose of study To identify how frequently medical students were asked to interpret in a healthcare setting and explore issues related to the use of non-professional interpreters.

Methods used An online survey was distributed to 3rd and 4th year medical students at McGovern Medical School and UTMB to identify bilingual students who served as interpreters in the clinical setting and assess how this role impacted their overall clinical experience.

Summary of results Of the 216 students who completed the survey, 53% (114/216) were found to be fluent in a language other than English. Of these students, 89% (102/114) reported that they were asked to interpret while in the clinical setting, most commonly (75%, 85/113) because their team preferred not to wait for a certified interpreter. Most bilingual students (71%, 91/129) stated they never felt pressured to interpret by an authority figure, but 19% responded that they have 'occasionally,' 'often,' or 'always' been in a situation in which they felt uncomfortable interpreting for the patient. The most cited reasons for discomfort included insufficient medical vocabulary to adequately relay information or a high-stakes clinical setting (obtaining informed consent). Of the bilingual students, only 85% felt that being fluent in another language served as an advantage to them while in the clinical setting. In addition, 39% (76/195) of students felt 'unprepared' or 'very unprepared' to provide care for Limited English Proficiency patients in the future.

Conclusions AAMC Guidelines state an ad hoc interpreter should only be used when a trained medical interpreter is unavailable. Studies show that use of professional interpreters results in a significantly lower likelihood of errors of

potential consequence than use of ad hoc interpreters. However, the convenience of utilizing medical students as interpreters seems to outweigh the clear benefits of a trained interpreter. Measures should be taken to identify bilingual students who are willing to serve as interpreters for LEP patients, and schools should institute guidelines outlining situations in which use of an ad hoc interpreter is acceptable. Additionally, more research should be done to determine whether introduction of a medical Spanish course would be beneficial for better preparing students for treatment of LEP patients in the future.

175

DEVELOPMENT OF PLAIN LANGUAGE BIOBANKING PAMPHLET IN ENGLISH AND SPANISH

M Doucet*, E Friday, C Arnold, M Vazquez, T Davis. LSUHSC-Shreveport, Shreveport, LA

10.1136/jim-2018-000974.173

Purpose of study Address disparities of low income and minority patients' awareness, understanding and recruitment in biorepositories by developing patient-centered pamphlets that are literacy and culturally appropriate.

Methods used Reviewed biobanking materials used nationally. Interviewed clinical research assistants who consent patients for biobanking to identify issues patients were concerned about. Conducted focus groups with target patients and the medical centers' community advisory board to elicit input on terms to use to explain biobanking in simple terms and strategies to disseminate the information. Developed iterative drafts with biobanking and health literacy experts to create accurate, understandable and actionable pamphlet.

Assessed reading level with Flesh Kincaid and used the Patient Education Materials Assessment tool (PEMAT) to score understandability and actionability. Translated materials into Spanish with help of fellow Latino Medical Student Association members. Edited iteratively based on input from Hispanic provider's and patients to improve Spanish expressions and cultural appropriateness. Conducted two focus groups of Hispanic patients to determine understanding, acceptance and intention to participate.

Summary of results The final pamphlets were a one-page tri-fold with colored pictures that illustrated key points, white space, brief bullet paragraphs and use of headers asking questions. The pamphlets were written on a 6th grade reading level. The PMAT indicated a score of 88% in understandability and 80% in actionability. The Hispanic patient focus groups revealed patients had never heard of biobanking and none had ever been asked to participate in medical research or biobanking. Patients indicated the biobanking brochure was informative and helpful. All said they were open to participating if asked but wanted to be asked by their physician, especially one who was language concordant.

Conclusions Diverse participation in biobanks is necessary to identify how specific drugs and treatments are most useful for different groups. Patients underrepresented in clinical trials and biobanking found the pamphlets helpful. Future efforts should involve patients and clinicians in the development of biobanking and medical research materials. Brief patient-centered pamphlets have the potential to improve the consenting process.

176

IMPLEMENTATION OF POSTPARTUM DEPRESSION (PPD) SCREENING AT THE OU LATINO CLINIC

B Dusenberry*, M Dunlap, E Douville. *Oklahoma University Health Sciences Center, Oklahoma City, OK*

10.1136/jim-2018-000974.174

Purpose of study The most recent data from the CDC estimates 11.5% of mothers suffer from PPD. In Oklahoma, rates are even higher at 14.9%. Postpartum depression effects not only the mother but the family as a whole. PPD can lead to poor nutrition and health of the child, reduced breastfeeding, impaired bonding with the child and impaired relationship with the mother's partner. The purpose of this project is to 1) increase screening of mothers of newborns for postpartum depression and provide those who screen positive with resources and 2) increase provider awareness and knowledge of postpartum depression and the local resources available.

Methods used Residents and faculty members completed an anonymous online survey about their current screening practices and knowledge of postpartum depression. A short teaching session on postpartum depression as well as teaching on the local resources available to our families was presented during the residents' continuity clinic. Maternal depression screening was started in clinic and data on screening will be reviewed weekly. A post survey will be collected in a few months. Descriptive statistics with simple frequency counts were determined.

Summary of results 55 residents and faculty completed the pre intervention survey (72.7% were residents and 27.3% were faculty). 18.2% of respondents state they screen for PPD, 45.5% said they sometimes screen, and 36.3% said they did not screen. Only 34.5% of respondents stated they had resources to give to mothers with PPD. And while respondents rated the importance of PPD at 9 on a 1 to 10 scale, when asked about their knowledge of PPD the average score was 4 out of 10, indicating low baseline knowledge. Baseline data showed no maternal depression screens with PHQ-9 were completed. Weekly data collection on screens preformed is being collected.

Conclusions The pre intervention surveys clearly show that our residents and faculty agree that PPD is a major issue but also shows that we do not have the knowledge or resources to adequately address this issue. The teaching session addressed how to formally screen for PPD, information on PPD, and community resources available for mothers. A post survey will further evaluate the progress of our intervention on providers and weekly data collection on the increase in our screening rates.

177

PATIENT CHARACTERISTICS FOR NO-SHOW ENDOSCOPY APPOINTMENTS AT A LARGE UNIVERSITY MEDICAL CENTER

A Goel*, N George, S Garg, S Inamdar, B Tharian. *University of Arkansas for Medical Sciences, Little Rock, AR*

10.1136/jim-2018-000974.175

Purpose of study Patient no-show for scheduled endoscopy procedures is a widespread healthcare problem. It results in treatment delays and financial losses. We reviewed the patients who did not show up for their scheduled endoscopy appointments at our center to identify key characteristics of this population.

Methods used This was an IRB-approved retrospective study from April 2017 to October 2017. All patients who were scheduled for gastrointestinal endoscopy procedures and did not show up for their appointment were included in the study. Demographics, social characteristics and appointment waiting time of these patients were obtained through review of medical charts.

Summary of results During the study period, 495 patients who were scheduled to undergo gastrointestinal endoscopy procedures did not show up for their appointment. The mean age of this population was 55.05 ± 12.51 years. 166 (33.54%) patients were married, 168 (33.94%) were single, and 161 (32.52%) were legally separated, divorced or widowed. A large majority of these patients were either unemployed (256, 51.72%) or disabled 126 (25.45%), while only 113 (22.83%) patients had employment. Similarly, current (188, 37.98%) and former smokers (149, 30.1%) formed the majority and only 158 (31.92%) were never smokers. Only 57 (11.52%) patients had a previous history of no-show and the rest (438, 88.48%) did not show up for their appointment for the first time. Patients with a waiting time <4 weeks missed fewer (65, 13.13%) as compared to patients with waiting times of 4–12 weeks (115, 23.23%), 12–24 weeks (119, 24.04%), and >24 weeks (196, 39.60%).

Conclusions Most of the patients who did not show up for their endoscopy appointment did not have a partner, were either unemployed or disabled, were current or former smokers, or had a long wait time to endoscopy appointment. Further studies are needed to identify factors that predict no-shows. This will help design interventions to reduce the no-show rate and improve healthcare delivery to patients.

178

MANAGEMENT OF PRENATALLY DIAGNOSED ACHONDROPLASIA

¹C Gooch*, ²A Hurst, ²J Blount, ²MH Halsey. ¹UAB, Birmingham, AL; ²University of Alabama at Birmingham, Birmingham, AL

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Case report Many common genetic conditions, such as Down syndrome and 22q11.2 deletion syndrome, have guidelines for managed care. Achondroplasia, the most common skeletal dysplasia, also has published guidelines, which include a focus on preventing foramen magnum compression and sleep apnea, both central and obstructive, as these are the main causes of morbidity in achondroplasia. These guidelines target pediatric management and do not address recommendations in the neonatal period for the relatively new occurrence of prenatally diagnosed achondroplasia. There has been little research in neonatal achondroplasia management, thus there is little data-driven recommendations for how best to care for these infants. The medical community agrees that these children need comprehensive physical exams to assess neurological status and polysomnography. However, the timing (pre-symptomatic vs post-symptomatic) of the tests and correct method of imaging (CT vs MRI) for accomplishing these general recommendations has not been researched. This presentation will detail one hospital's multi-disciplinary approach to creating and implementing our own neonatal achondroplasia protocol and our ongoing research in this area. The presentation will begin with a case presentation describing the situation that launched our protocol. It will describe the multi-disciplinary

input into our protocol and its implementation at our hospital. It will end with a discussion of the results we have thus far and with presentation of our ongoing research projects in neonatal achondroplasia across four departments.

179 A QUALITY IMPROVEMENT PROJECT TO IMPROVE ASTHMA MANAGEMENT IN A LATINO CLINIC

D Hahn*, M Cooper, R Blucker, M Dunlap. *University of Oklahoma Health Science Center, Oklahoma City, OK*

10.1136/jim-2018-000974.177

Purpose of study Asthma is the most common chronic disease of childhood. It has a significant impact on a child's health and health care utilization. When asthma is controlled with routine care and education, patients are less likely to utilize the emergency department, urgent care center, or be hospitalized for asthma related illness. At the OU Latino Clinic, our parents are mainly Spanish-speaking, but the majority of patient information is in English. In particular, the Asthma Action Plan (AAP) in our Centricity Electronic Medical Record (EMR) is only in English; therefore, it is difficult to provide appropriate asthma education that includes an AAP to our patients with asthma. The purpose of this QI project is to develop a system in which we can provide and document AAPs in both English and Spanish, so we can better provide appropriate asthma education to our patient population.

Methods used We have developed an Asthma Action Plan in Spanish and it has been incorporated into our EMR. We are reviewing numbers of AAPs given in our Latino Clinic (Phase I), and then will track numbers of visits for asthma in both the clinic and in the emergency department (Phase II). Currently, data is being pulled weekly from our EMR to look at all patients with a diagnosis of asthma that visit the clinic for a sick, return, or well visit and whether they had an asthma action plan printed at that visit and/or in the last year. By the end of Phase II, we hope to see a decrease in sick visits and a decrease in ED visits for asthma in our patients.

Summary of results Twenty-seven patients with asthma had visits in week one with 6 of them being well child checks (WCC) and 21 being return or sick visits; 11.1% of those patients had an AAP printed at that visit and 59.3% in the last year. In week two, 29 patients with asthma were seen, 6 were WCCs and 23 were sick or return visits; 24.1% received AAPs at that visit and 65.5% in the last year. In week three, 28 patients were seen, 3 were WCCs and 25 were return or sick visits; 17.8% received AAPs at that visit and 67.8% in the last year.

Conclusions If asthma education is to be effective, it must be understood by the patient and family. We expect to see an improvement in health care utilization as a result of our simple intervention of providing written information in their native language.

180 PEDIATRIC IBUPROFEN INGESTION: DO WE NEED TO BE CONCERNED?

¹S Hanback*, ¹AW Cohen, ²B Whitworth, ²A Slattery, ¹MH Nichols. ¹*University of Alabama at Birmingham, Birmingham, AL*; ²*Children's of Alabama, Birmingham, AL*

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Purpose of study Upon reviewing Regional Poison Control Center (RPCC), Children's of Alabama's data, it was revealed

that ibuprofen is involved in 4% of all poisoning exposures and 7.2% of all pharmaceutical poisoning exposures in the pediatric population. There is a dearth of recent studies in the literature regarding the effects and outcomes of pediatric patients with these ingestions. The objective of this study was to evaluate the epidemiology of pediatric ibuprofen exposures reported to the RPCC by age, intent, amount ingested, clinical effects, and outcome.

Our study elucidates current trends of ibuprofen ingestions in Alabama as reported to the RPCC

Methods used A retrospective review of greater than 4000 ingestions over a six-year period involving ibuprofen exposures reported to Regional Poison Control Center was performed. Inclusion criteria were: patients with ibuprofen ingestions, 0–18 years of age, reported to our RPCC from 2012–2017. Cases excluded were those with unknown outcomes and co-ingestants. Outcome measures were *no effect*, *mild effect*, *moderate effect*, and *major effect* as defined by The American Association of Poison Control Centers. This study was approved by UAB IRB.

Summary of results In total, 4420 cases were reviewed. Among those cases, outcomes were 2549 *no effect*, 1 *major effect*, 68 *moderate effect*, and 130 *minor effect*. Of cases with symptoms, the most frequent intent was suspected suicide attempts. GI symptoms (129) were the most common symptoms seen, followed by cardiovascular (33) and neurological (29). The single case which resulted in a major effect experienced metabolic acidosis, after ingesting 20 grams (370 mg/kg) of ibuprofen. Of the categories with clinical effects, approximately 97% of children under 6 years old had *no effect* where as only 64% of children in the 13–18 age range had *no effect*.

Conclusions Ibuprofen remains a relatively safe over the counter analgesic for use in the pediatric population. In children less than 6 years of age, inadvertent exposures rarely result in poor outcomes

181 WIDE PROVIDER VARIATION IN COST FOR THYROIDECTOMY: POTENTIAL BENEFITS OF STANDARDIZING PRACTICE?

B Herring*, S Jang, Z Aburjania, H Chen. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.179

Introduction Identifying provider variation in surgical costs could control rising healthcare expenditure and deliver cost-effective care. While these efforts have mostly focused on complex and expensive operations, provider-level variation in costs of thyroidectomy has not been well examined.

Methods We retrospectively evaluated 989 consecutive total thyroidectomies performed by 14 surgeons at our institution between September 2011 and July 2016. Data were extracted from the McKesson Business Insight program. Total length of stay and cost were evaluated using the Mann-Whitney U and the Kruskal-Wallis tests. Categorical variables were evaluated with chi-square.

Summary of results Median patient age was 48 years (range 8–90), 81% were females, 64% were Caucasians, and 77% were outpatients. The number of thyroidectomies performed by the 14 surgeons ranged from 4 to 635 (mean=71). The median costs per provider varied widely from \$4,390.94 to \$16,754.15 ($p<0.001$). The mean length of stay was $1.2d\pm 8.2$ with wide

Abstract 181 Figure 1 O/E v. time

variation among providers (0d to 5.5d). Providers whose hospital cost exceeded the institutional mean demonstrated a significantly higher ICU admission rate, anesthesia fees, and lab costs ($p < 0.001$).

Conclusions We found substantial variation in hospital cost among providers for thyroidectomy despite practicing in the same academic institution, with some surgeons spending 4x more for the same operation. Implementing institutional standards of practice could reduce variation and the costs of surgical care.

182 EFFECTIVENESS OF A NEONATAL LACTATION NURSE IN PREVENTING HYPERNATREMIC DEHYDRATION IN A RURAL KENYAN HOSPITAL

¹M Howard*, ¹A Sorrentino, ²A Shirk. ¹University of Alabama at Birmingham, Huntsville, AL; ²Kijabe Hospital, Kijabe, Kenya

10.1136/jim-2018-000974.180

Purpose of study Hyponatremic dehydration in neonates is a potentially life-threatening condition often leading to a variety of adverse neurologic and vascular sequelae. It is one of the most common reasons for hospitalization in the neonatal period in East Africa, and is often caused by breastfeeding difficulties. In October 2015, a full-time lactation nurse was added to staff at Kijabe Hospital in Kenya, in efforts to identify babies at risk of developing hyponatremia due to breastfeeding difficulties.

Our main objective is to determine the impact of hiring a full time neonatal nurse to foster lactation and examine babies born in Kijabe Hospital. Specifically, to determine how this affected the rate of hyponatremic dehydration and related outcomes.

Methods used A database of neonates admitted to Kijabe Hospital revealed 228 newborns (<28 days old) admitted to Kijabe Hospital with hyponatremia between October 1, 2014 and December 31, 2016. Chart review was then used to compare records of the neonates with hyponatremia between two groups: those born in Kijabe before the hiring of the full-time lactation nurse (October 1, 2015) and those born after. For each newborn: admission information, symptoms, laboratory findings, comorbidities, treatments, complications, and mortality were recorded.

Summary of results The average serum sodium concentration was higher in the group of neonates born before the full-time nurse was hired than the group after ($p = 0.0064$). Creatinine levels on admission were significantly lower in the group after the nurse was hired compared to before ($p = 0.0043$); significance remained when only maternal lactation issues were

included ($p = 0.0149$). There was no significant difference in days for hyponatremia to correct between the groups.

Conclusions Hiring a full-time lactation nurse at Kijabe Hospital was associated with decreased sodium and decreased creatinine upon admission. This indicates that hiring a lactation nurse is beneficial and may decrease the severity of illness associated with hyponatremic dehydration.

183 OVERUSE OF ANA TESTING IN AN ACADEMIC MEDICAL CENTRE

A Igwe*, Case Western Reserve, Cleveland, OH

10.1136/jim-2018-000974.181

Purpose of study The American college of rheumatology advises that repeated anti-nuclear antibody testing after a previously positive ANA is not indicated. Once an ANA is positive ACR recommendation, is not to re-check, especially in established rheumatology patients (Juvenile idiopathic arthritis, systemic lupus), as it is of no clinical utility (1–3).

Aim To determine how many patients with a positive ANA and have subsequently undergone repeated ANA testing despite prior positive ANA.

Methods used Electronic medical records system (epic) at the Metrohealth System were examined using statistical search software. Specifically we looked at ANA tests ordered from Jan 2004 -> April 2018. Data analyzed using R-Studio. We analyzed the only positive ANA/positive titer (as per lab reference) via immunofluorescence.

Summary of results 1120 patients had positive ANAs. 1043 out of 1120 (93.12%) underwent subsequent ANA re-testing despite having a one or more previously positive ANA. 642 of the 1120 (57.32%) were re-tested and had subsequent negative ANAs.

Conclusions Results demonstrate that there is frequent re-testing of positive ANA which in turn raises concerns about the wasteful use of these tests and unnecessary increase of health care expenditures. Furthermore, subsequent negative ANA in patients who had a previously positive ANA further raises the concern regarding its clinical utility.

184 STORAGE OF MEDICATIONS: A SURVEY OF FAMILIES IN A PEDIATRIC EMERGENCY DEPARTMENT

A Ivy*, S Mohr, A Webb, K Monroe, MH Nichols. UAB, Birmingham, AL

10.1136/jim-2018-000974.182

Purpose of study The national opioid problem has reached crisis levels in recent years. Alabama has the highest per capita prescribing rate of opioids in the country. Previous data suggests that opioids are often stored unsafely, and children are at high risk of exposure to these potentially dangerous medications. We designed our study to evaluate the perceptions and practices of caregivers related to prescription medication storage as well as to provide education regarding safe medication storage and the opioid crisis.

Methods used Following IRB approval, caregivers of patients in the Pediatric ED were asked to participate in a survey about their medication storage practices and beliefs, with a focus on opioid medications. Data was collected via an online survey tool documenting demographic information along with

knowledge and behaviors related to medication storage. A brief instruction about the opioid crisis and safe storage was provided verbally along with an opioid safety brochure.

Summary of results 233 families, accounting for 511 children, participated; three families declined. Only 14% of caregivers reported storing their prescribed medications in a locked or latched place. Most believed their child or children's friends could not easily access their prescribed medications (82%). Most respondents who did not keep their medications in a locked or latched place had never thought about it (47%). 33% of respondents were unaware of the opioid crisis. 87% of caregivers said they would use a medication lock box if given one.

Conclusions Many caregivers are not aware of the opioid crisis. Many caregivers do not keep medications, including opioid containing medications, locked up, but most store them 'out of reach.' About half of caregivers stated they had 'never thought about' locking up medications. Most parents do not think their children or their children's friends could gain access to medications in their home. Most parents would use a lock box if one was given to them. This opens the door for further study, education, and interventions.

185 IMPROVING RESIDENTS' CONFIDENCE IN LEADING PEDIATRIC CODES

J John*, P Klingborg. *University of Tennessee-Chattanooga, Chattanooga, TN*

10.1136/jim-2018-000974.183

Purpose of study Pediatric residents have an important role as first responders during pediatric emergencies in Children's hospitals. Even though a multi-disciplinary team is involved during the code, pediatric residents are sometimes in charge of leading the code. Based on exit surveys from 2013–2015 graduating residents, only 32% strongly agreed that they can independently resuscitate a patient. The goal of our study was to improve residents' confidence level in leading pediatric codes.

Methods used A pre-study survey investigated the number of real and mock codes residents participated in and rated their confidence level on a scale of 1–10. Based on this data, investigators recognized a need for resident-focused mock codes in addition to the unannounced multidisciplinary mock codes. These sessions were limited to 6 pediatric residents. 2 code scenarios were conducted using high fidelity patient simulator. Initially, investigators arranged 3 mock code sessions. During our subsequent PDSA cycle, this increased to 6 sessions. 96% of residents participated in at least one of these sessions and 52% of residents participated in 2 during the second PDSA cycle. Resident confidence was then investigated with a validated, scaled post-study survey.

Summary of results Our pre-survey showed that 100% of PGY1 and PGY2 residents and 78% of PGY3 residents have only done ≤ 2 mock codes. Only 22% of PGY3 residents participated in 3–4 mock codes. 87% of PGY1 residents rated a confidence level of 2 (not confident); 75% of PGY2 residents rated 4–6 (mild confidence). Post survey showed that 57% of PGY1% and 71% of PGY2 residents participated in ≥ 3 –4 mock codes and 86% of PGY3 residents participated in ≥ 5 –6 mock codes. Resident confidence level ratings showed 75% of PGY1 now rated 4–8 (mild confidence to confident) and 86% of PGY2 rated 6–7 (confident). On 2018 ACGME exit survey, 100% of PGY3 pediatric residents agreed and 50% strongly agreed that they can independently resuscitate a patient.

Conclusions Pediatric residents lead a crucial role in many children's hospitals, often being the first responders in pediatric codes. According to ACGME senior exit survey results, many pediatric residents do not feel confident in this role. Our study demonstrates implementation of resident-focused mock codes increases confidence ratings and supports use of high fidelity simulation in resident education.

186 IMPACT OF THE ADULT CONGENITAL HEART DISEASE CLINIC ON PATIENT SURVIVAL IN MISSISSIPPI

S Kiparizoska*, C Richards, K Windham, F Han, WF Campbell, ME Hall, B Kogon, MR McMullan. *University of Mississippi Medical Center, Jackson, MS*

10.1136/jim-2018-000974.184

Purpose of study The University of Mississippi Medical Center (UMMC) opened Mississippi's first Adult Congenital Heart Disease (ACHD) clinic in January 2014. There is no published data assessing the rates of survival of MS ACHD patients. The aim of this work is to assess the impact of the ACHD clinic on patient survival in the last 5 years.

Methods used We retrospectively analyzed electronic health data from UMMC's ACHD patients (n=3863) and included the following: diagnosis name, age, sex, and race in our search from 2013–2017. Diagnosis name was further classified by anatomy complexity and an outcome of an all cause death was assessed. Death rates were calculated by dividing the total amount of all cause deaths of patients with CHD at UMMC by the total number of patients at UMMC who have a CHD diagnosis.

Summary of results Death rates of patients with a current or repaired CHD dropped from 6.6% in 2013 to 1.1% at the end of 2017. In the entire population, 60% of patients were female and 40% were male and 54% of total patients identified as white and 42% as black. There was no trends identified between death rates and age, sex, or race.

The death rate for those patients with complex or unspecified anatomy appeared to remain stable over the study period, while those with moderate anatomy appeared to increase. Most of the reduction in death rates appeared to occur in the simple anatomic subgroup (figure 1).

Conclusions The death rate has continued to decrease since the opening of the ACHD clinic supporting the model of specialized medical care for ACHD patients. Further longitudinal assessment is needed to determine the overall effect of this care on the aging ACHD population in MS.

Abstract 186 Figure 1

187

RATES AND RISK FACTORS FOR THE HEALTHCARE UTILIZATION OF THE NEONATAL INTENSIVE CARE UNIT GRADUATES WITHIN THE FIRST TWO YEARS OF LIFE

M Rydzewska*, J Sun, M Kong, Y Feygin, S Duncan. *University of Louisville, Louisville, KY*

10.1136/jim-2018-000974.185

Purpose of study To assess the perinatal and neonatal characteristics of infants admitted to neonatal intensive care units in Kentucky and describe the risk factors and trends in re-hospitalization and healthcare utilization of the infants discharged from the NICU during the first two years of life.

Methods used This is a retrospective cohort study using administrative data set on the infants born alive between January 1st 2012 and December 31st 2015 and enrolled in Kentucky Medicaid. Study population was identified by the presence of Current Procedural Terminology (CPT) codes 99468, 99477, 99469, 99748, 99479, 99 480 during the first week of life. The admission and discharge diagnoses (ICD-9 CM and ICD-10-CM codes- up to 4 per infant) were classified into clinically significant categories.

Summary of results We identified a cohort of 6338 infants whose perinatal and neonatal characteristics are included in table 1. Our data thus far show that the most common reasons for NICU admissions are respiratory distress (not RDS) (58%), low birth weight (50%), prematurity (49%), respiratory distress syndrome (29%), neonatal infection (16%), jaundice (16%), neonatal drug withdraw (13%), cardiac and circulatory disorders (13%) and hypoglycemia (12%).

Abstract 187 Table 1 Study population characteristics (n=6338)

Characteristics	N	%
Gender: Male	3567	56.5
Gender: Female	2771	43.7
GA:	533	8.4
GA: 29–36 weeks	2570	40.5
GA: >37 weeks	446	7
BW < 999 gm	331	5.2
BW: 1000–2449 gm	2778	43.8
BW > 2500 gm	390	6.2
Gestation: Single	1742	27.5
Gestation: Multiple	326	5.1
Mode of delivery: CS	1395	22.0
Mode of delivery: Vaginal	866	13.7
Race: White	2643	41.7
Race: Black	337	5.3
Race: Other	1745	27.5

Conclusions We are currently examining the trends in re-hospitalization and healthcare utilization for this cohort.

188

INCREASING THE SUPPLY OF BREASTFEEDING KNOWLEDGE-MIDPOINT ASSESSMENT

K McCoy*, N Mandlik. *UTHSCSA, San Antonio, TX*

10.1136/jim-2018-000974.186

Purpose of study Many mothers consider their pediatrician to be a good source of breastfeeding education and support,

however they do not receive adequate training in breastfeeding. Some sources report an average of only 3 hours annually. A designated breastfeeding curriculum increases both pediatrician comfort level and exclusive breastfeeding rates. Until our intervention, our pediatric residency program did not have an in-depth breastfeeding education program for all residents.

Methods used Based on successes described in prior studies, and the AAP's Breastfeeding Curriculum, we created an integrated educational curriculum and implemented with our resident class of 2020. The curriculum includes readings, small group discussions, didactics, and hands-on experience shadowing a lactation consultant. We used the AAP's evaluation tool that includes Likert scale items about clinical skills opportunities, confidence in support for a breastfeeding mother, and assesses relevant knowledge. A limited mid-point assessment of skills and confidence was collected halfway through the intern year.

These mid-point surveys were compared to study subject baselines, to year-end class of 2019, and the graduating class of 2017 using Mann-Whitney U test. We also administered a curriculum evaluation itemizing rotations completed to that point and open-ended feedback. A validated infant feeding attitude scale was added to pre-and post-assessment as well. Descriptive analysis and unpaired t-test will be performed on final results.

Summary of results An increase in intervention group scores for clinical experience and confidence at midpoint of intern year was statistically significant compared to baseline. They were also equivalent to class of 2017 and 2019 scores, indicating an increase during the intern year alone when compared with the entire 3 year training period. All interns who worked with a lactation consultant in the newborn nursery noted it was the most helpful part of the curriculum.

Conclusions The implemented curriculum increased both resident opportunity for clinical skills and confidence in assisting breastfeeding mothers. Based on these results and end of year assessment, we will continue implementation of our curriculum.

189

CRC SCREENING IN RURAL COMMUNITY CLINICS: SUSTAINABILITY IN ANNUAL SCREENINGS

P Murphy*, T Davis, C Arnold. *LSUHSC-Shreveport, Shreveport, LA*

10.1136/jim-2018-000974.187

Purpose of study To assess the effectiveness of two health literacy informed telephone follow-up strategies to improve annual repeat screening with Fecal Immunochemical Test (FIT) in rural community clinics.

Methods used A two-arm randomized controlled trial implemented in 4 rural community clinics. Clinics reported CRC baseline screening rates of 3% to 5%. Eligible patients, age 50–75, were recruited. The RA recommended screening and gave brief literacy and culturally appropriate education using a pamphlet (4th grade level), the FIT kit, simplified instructions (3rd grade level) and a demonstration of how to use it. At 4 and 8 weeks patients who had not returned their kit received either 1) a personal follow-up call (PC) from a central prevention coordinator using motivational interviewing skills and reminding them to complete FIT kits; or 2) an

automated follow-up call (AC) using plain language and motivational messages encouraging patients to complete and mail the FIT. In years 2 and 3, rather than give the information in person, patient education and FIT kits were mailed to participants with follow-up calls following the same procedure as Year 1.

Summary of results 620 patients were enrolled: 311 in AC arm and 309 in PC arm; 66% were African-American, 55% women; 40% had limited literacy. During Year 1, 69% completed screening in AC arm versus 67% in PC arm, with 43% and 44% respectively needing a follow-up reminder call. In Year 2, percentage screened decreased: 40% screened in AC arm and 37% in PC arm. Number of patients that needed at least one follow-up reminder call increased to 74%. Among those called, 19% in the AC arm completed the kit versus 15% in the PC arm. To date in Year 3, 33% screened in AC and 34% in PC with 85% and 79% respectively needing a follow-up call.

Conclusions Personal communication with simplified instructions and follow-up reminders influenced completion of all patients, particularly those with limited literacy. The less costly and time consuming automated call was equally effective as a personal call. Screening rates in Years 2 and 3 declined. Screening sustainability suggests personal contact/relationships and accountability by the patient and provider strategies facilitate annual screenings. CRC screening with FIT is only effective when completed annually.

190 HEPATITIS C VIRUS SCREENING IN FEDERALLY QUALIFIED HEALTH CENTERS IN RURAL APPALACHIA

FS Olanrewaju*, SW McKenzie, P Vanhook, A Falodun. *East Tennessee State University, Johnson City, TN*

10.1136/jim-2018-000974.188

Purpose of study The prevalence of Hepatitis C Virus (HCV) in the US is estimated at 2.7 to 3.9 cases per 1 00 000 with 19 659 deaths in 2014. It is the most common blood-borne infection, with a higher age-adjusted mortality rate than Hepatitis B Virus or Human Immunodeficiency Virus. Without treatment, nearly 1.1 million people will die from HCV by 2060. About 34 000 new cases of HCV were reported in 41 states in the US in 2015. The incidence of HCV in Tennessee was 13 023 per 1 00 000.

This is a descriptive study to ascertain the HCV prevalence and usefulness of screening in medical outreach settings (MO) compared to indigent healthcare clinics (IHC) in northeast Tennessee.

Methods used Between April 2017–May 2018, routine, opt-out HCV testing was performed in 3 IHC and 3 MO sites in the Tri-Cities, TN region. During screening, demographic information was collected and the de-identified data were analyzed.

Summary of results A total of 120 persons were screened for HCV. Among these, 16 (13.33%) were HCV- antibody positive. Of all patients screened, 67 (55.8%) were born between 1945–1965. 50% (8) of HCV-antibody positive patients were from this group. The frequency of males and females screened were 46.7% and 53.3% respectively, with a higher proportion of males (56.25%; $p=0.4092$) found to be positive for HCV. Non-Hispanic whites and African Americans made up 87.5% and 10.8% respectively of all clients screened; 93.75% ($p<0.0003$) of the positive cases were ascribed to

non-Hispanic whites. Screening occurred in six testing locations, 3 each for MO events and IHCs. A total of 15 (93.75%; $p<0.0003$) HCV-antibody positive cases were found in the IHCs compared to 1 (6.25%) found in a MO event.

Conclusions Screening demonstrated HCV antibody prevalence of 13.33% among clients tested in 3 indigent care clinics and 3 medical outreach events the in Tri-Cities region. Most HCV-antibody positive persons were non-Hispanic whites (93.75%) in the indigent care clinics. This analysis shows the higher yield of targeted screening at indigent healthcare facilities. Targeted HCV screening is critical in the era of direct-acting antiviral agents.

191 PRICE TRANSPARENCY CAN DECREASE COST OF SEIZURE MANAGEMENT AMONG PEDIATRIC RESIDENTS

R Parlar-Chun*, J Malek. *MGovern Medical School, Houston, TX*

10.1136/jim-2018-000974.189

Purpose of study The United States leads the world in health-care costs. While this problem is multifactorial, clinicians have responsibility in providing cost-efficient patient care. The ACGME expects residents to incorporate considerations of cost awareness in patient care, yet cost is not always transparent. The purpose of this study is to evaluate the effect of price transparency on management cost among pediatric residents.

Methods used 56 Pediatric residents were presented five scenarios of patients with seizures (febrile-complex in duration/complex in focality/simple, nonfebrile- generalized/focal) and were asked to choose labs, microbiology, imaging, and medications from a table. They were also asked if they would admit the patient and if they would prescribe abortive medications. Confidence in their management choices were assessed. After completion of the scenarios, residents were given the same scenarios but with the prices of the management selections available. Prices were obtained from a hospital chargemaster. Primary outcome was mean difference in costs after price transparency which was also stratified by PGY status. Secondary outcomes included price awareness effect on confidence, decision to admit, and decision to prescribe abortive medications. Paired t-test and Wilcoxon signed rank test were used as appropriate.

Summary of results All five scenarios had significant reduction in cost of management after price transparency ranging from an average difference of \$503 to \$1638. There was no significant effect on confidence or prescription of abortive medication. There were mixed results in effect on admission. There were mixed results among PGY status, with a trend towards no effect in PGY1s and towards significance among PGY2 and PGY3.

Abstract 191 Table 1 Price transparency effect on average cost of management

Scenario	Pre	Post	Difference (95% CI)	P value
1	\$1596	\$1093	\$503 (123.5, 881.6)	0.01
2	\$5092	\$3682	\$1410 (616.3, 2204.6)	<0.001
3	\$7047	\$5408	\$1638 (698.1, 2579)	0.001
4	\$5847	\$4483	\$1364 (536.7, 2190.9)	0.002
5	\$7200	\$6187	\$1013 (407.9, 1619)	0.001

Conclusions Price transparency may be an effective way to decrease cost of management among pediatric residents. Further investigation in a generalized, real world population, and an evaluation of balancing measures is warranted.

192 THE PHYSICIAN'S ROLE IN CARE SEAT SAFETY

¹SB Phillips*, ²A Manhas. ¹University of Alabama, Northport, AL; ²University of Alabama, Tuscaloosa, AL

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Purpose of study Accidents are the number one cause of death in the United States for those ages 0–19. Motor vehicle accidents make up a significant portion of those accidents. The AAP recommends that children ride rear facing until they are no longer able to ride rear facing due to weight or height limitations. Since motor vehicle accidents are a leading cause of death and riding rear facing is the safest position while in a vehicle, it is important that parents are notified of the American Academy of Pediatric's recommendations.

Methods used A questionnaire was given to 106 caregivers of children under 2 years of age during their visit with their pediatrician or family medicine physician. The survey evaluated if their child was using a rear facing car seat and if a doctor had recommended they use a rear facing car seat. Parents were given a handout from the Centers for Disease Control and Prevention with illustrations and recommendations of car seat positions.

Summary of results Overall, 90% of caregivers reported that their child rode in a rear facing car seat. When respondents were asked if their doctor told them their child should ride rear facing, 83% reported they had been told. When asked if they thought riding rear facing was safer than riding forward facing, 92.5% agreed that rear facing is the safest way to ride.

When asked who else other than a physician had told them to use a rear facing car seat, the overwhelming response was a family member or friend. In fact, 74% of respondents cited family or friends had told them to place their child in a rear facing car seat.

Conclusions New guidelines have emerged from the American Academy of Pediatrics since this study's initiation that recommend children stay rear facing as long as weight and height allow. It appears that most children are riding rear facing and most parents feel that children should ride rear facing. However, there is room for improvement on conveying this information to parents during their clinic visit. Also with the majority of respondents citing family or friends as their source of information about car seat safety, there is an opportunity to discuss pediatric car seat safety with adults during their clinic visits as well.

193 INCREASING COMFORT WITH SENSORY PROCESSING DIFFICULTIES IN PRE-HOSPITAL SETTING: PRE-POST STUDY OF EDUCATION AND SENSORY TOOLS IN EMERGENCY MEDICAL SERVICES PROVIDERS

¹NP Shah*, ²K Hert, ¹A Klasner. ¹University of Alabama at Birmingham, Birmingham, AL; ²Alabama Department of Public Health, Montgomery, AL

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Purpose of study Interfacing with patients with sensory processing difficulties is challenging to health care providers and even more problematic for Emergency Medical Services (EMS) personnel in the acute care setting. Sensory training might be an effective non-pharmacologic method to deal with these patient populations. The purpose of this study was to evaluate if an educational session and placement sensory tools would improve the comfort of EMS providers in prehospital setting.

Methods used EMS providers from two agencies in the Alabama Gulf EMS System were selected for this study. Pre-education questionnaires were administered to EMS providers to assess their frequency and comfort level in taking care of these patients. Educational session included video presentation of various topics related to sensory processing difficulties and education on sensory tools. Post-education questionnaires were administered to EMS providers 3 months post educational session to assess the use of sensory tools and their comfort in patient care. Comfort level was assessed on a Likert scale of 1–10 with 1 being not comfortable at all and 10 being extremely comfortable. We performed descriptive statistics and non-parametric Wilcoxon signed rank test to compare medians.

Summary of results Total of 177/225 (78.6%) EMS providers completed the pre-education questionnaire. In the pre-education period, 159 (89.8%) of the EMS providers transported patients with sensory processing difficulties. Pre-education median comfort level was 7.5 (range: 1–10). At post-survey 135/177 (76.3%) EMS providers received educational training. 37 (27.4%) used the sensory tools within the prior 3 months. Post –education median comfort level was 8 (range: 3–10). Pre and post median comfort levels were significantly different ($p=0.006$).

Conclusions Sensory training can be an effective method for EMS providers to increase comfort in taking care of patients with sensory difficulties. Further research with larger sample size is needed to confirm/refute these findings.

194 PEDIATRIC RESIDENT RATE OF BURNOUT: A 3-YEAR TREND

N Sharma*, T Mabe, A Hendrix, K Mather. University of Oklahoma School of Community Medicine, Broken Arrow, OK

10.1136/jim-2018-000974.192

Purpose of study Our study seeks to trend the rate of burnout in Categorical Pediatrics Residency Programs and compare the rate of burnout at our institution to nationwide results.

Methods used With the help of the Association of Pediatric Program Directors Longitudinal Education Assessment Research Network (APPD LEARN), the Pediatric Residency Burnout Resilience Study Consortium 2018 Annual Study included 61 training programs in the United States. Our institution has participated in the 2016–2018 studies. The Maslach Burnout Inventory (MBI) was utilized to determine burnout while an additional survey assessed the utilization of a previous site-specific intervention.

Summary of results Average nationwide burnout among pediatric residents was 54% in 2016 ($n=1410$), 54% in 2017 ($n=1753$), and 52% in 2018 ($n=1867$). The average rate of pediatric resident burnout at our institution was 47% in 2016 ($n=17$), 63% in 2017 ($n=16$), and 56% in 2018

(n=18). Burnout in the 2016 Program Year (PGY) 1 was 29% with an increase of 21 percentage points in 2017 PGY2 (total 50%), and a decrease of 7 percentage points in 2018 PGY3 (total 33%). Additionally, 47% of respondents (n=15) across all PGY indicated that they participated in gratitude journaling, a site-specific intervention. Survey response rate for our program in 2018 was 65% compared to the nationwide rate of 62%.

Conclusions The burnout rate at our institution decreased more than the national average from 2017–2018. The 2016 PGY1 class was monitored as they experienced burnout through three years of residency with the highest rate of burnout occurring in their PGY2. The decrease in burnout during their PGY3 could be due to interventions for the past academic year such as changes in inpatient coverage, protecting weekends off, wellness programs, and gratitude journaling. Our institution will continue previous interventions and implement the following intervention for the next academic year: promoting counseling services, arranging for our Employee Assistance Program to contact all residents to schedule a voluntary appointment, and guarantee from the program to protect appointment time from rotation duties.

195 PROVIDER CONFIDENCE WITH AN ELECTRONIC HEALTH RECORD: AN EDUCATIONAL INTERVENTION

¹TD Smith*, ²R Trimm. ¹University of South Alabama Children's and Women's, Mobile, AL; ²University of South Alabama, Mobile, AL

10.1136/jim-2018-000974.193

Purpose of study To assess the efficacy of clinical location-specific PowerPoint guides developed to increase providers' confidence with a newly implemented Electronic Health Record (EHR) at the University of South Alabama (USA). Initial training for the EHR was in the form of online modules. Previous research has looked at teaching an EHR system to medical students. Others have looked at ways to improve providers' use of EHR long after initial implementation. To our knowledge, evaluating ways to improve use of EHR as an extended part of implementation has not been studied.

Methods used Guides were created and disseminated to healthcare providers. Providers were then asked via email to complete an online survey assessing competence in using the EHR before and after reviewing the guide with a Likert scale self-assessment. After six months for completion, responses were analyzed. Providers were required to state that they had viewed the guide. Surveys that did not include pre- or post-survey confidence on any one question were considered incomplete. Excluding incomplete surveys, the remainder were analyzed as both continuous and dichotomous data using the paired T test with unequal variance. For continuous assessment, answers were assigned a value from 1 to 5 with neutral=3. For dichotomous assessment, answers deemed 'confident' (somewhat/very confident) were assigned a value of 1; answers deemed 'not confident' (very uncomfortable – neutral) were assigned a value of 0. These values were averaged and assessed for USA C and W in total.

Summary of results Of 34 surveys, 21 were incomplete and excluded. 1 respondent had not viewed the guide. The remainder failed to answer 1 or more questions on the survey. Of 13 complete and valid surveys, 12 participants stated that they would be comfortable teaching others to use the EHR. There was a significant increase in confidence for USA C and W overall after introduction of the EHR guides when analyzed as dichotomous responses, $t(7) = 2.36$, $p=0.000004$ and as continuous responses, $t(11) = 2.2$, $p=0$.

Conclusions Clinical location-specific PowerPoint guides as used for increasing providers' confidence with using a new EHR were proven efficacious at USA C and W.

196 INTENSIVE CARE UNIT TELEMEDICINE IMPROVING MORTALITY

¹MB Whitlow*, ¹E Wallace, ²D McCormack, ¹JE Johnson, ¹W Stigler. ¹University of Alabama Birmingham, Birmingham, AL; ²Vaughan Regional Medical Center, Selma, AL

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Purpose of study Rural hospitals play an essential role in the care of many patients. The rural communities in Alabama are particularly vital, and can struggle with lack of resources. Upon presentation of a critically ill patient, rural providers must either transfer the patient to a higher acuity setting or take the responsibility for caring for these individuals without critical care oversight. In July of 2018, a tele-critical care program was initiated in the intensive care unit at Vaughan Regional Medical Center in Selma, Alabama. Here we show the preliminary results of implementation of tele-critical care in a rural hospital in Alabama.

Methods used In order to evaluate efficacy, the observed to expected mortality ratio was calculated as well as the risk adjusted mortality numbers were collected from January 2016 to August 2018 and presented in control chart format.

Summary of results The average O/E mortality from January to May 2018 was 1.08. Following the enactment of the telemedicine ICU program the average O/E was 0.83. Additionally, the risk adjusted mortality showed a reduction in mortality from July 2018 to August 2018 compared to the expected mortality over that same time period.

Abstract 196 Figure 1

Conclusions Preliminary results show a trend in improved O/E ratio with the implementation of tele-critical care and thus indicating the importance of tele-critical care in improving healthcare in rural regions.

Hematology and Oncology

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

197

HEMOPHAGOCYTIC LYMPHOHISTOCYTOSIS INDUCED BY POSTPARTUM

M Abdelmisseh*, Kern Medical, Bakersfield, CA

10.1136/jim-2018-000974.195

Case report Hemophagocytic Lymphohistiocytosis is a rare potentially fatal immunopathology resulting in overproduction of cytokines and unregulated proliferation of macrophages leading to multiorgan injury. HLH is either triggered by primary genetic disorder, in the pediatric population, or by secondary triggers such as malignancy, rheumatologic illness or viral infection.

We present a rare case of HLH precipitated by pregnancy and postpartum in a 28-year-old healthy female with 2 previous uncomplicated pregnancies.

28-year-old female G3P3 who was 22 days postpartum presented with pyrexia, persistent for 1 month, and dyspnea. Patient's laboratory was significant for anemia, thrombocytopenia, transaminitis with hepatic dysfunction and hepatosplenomegaly on CT of the abdomen. A sepsis workup did not identify any source of infection and the patient's symptoms and hepatic dysfunction continued to deteriorate despite adequate antibiotic therapy. Further laboratory workup suggested the possibility of Hemophagocytic lymphohistiocytosis. The diagnosed was appropriate as the patient met 5 diagnostic criteria out of 8: elevated ferritin (25074 ng/dL), significantly elevated IL-2 receptor (16455), fever, hepatosplenomegaly, anemia, and thrombocytopenia.

The workup for hematologic malignancy was ruled out by normal bone marrow biopsy; all rheumatologic workup was negative as well, and finally, all infectious disease workup was negative.

The patient was then treated successfully with corticosteroid and cyclosporin. Ferritin and IL-2 receptor trended down to normal values post-treatment. The liver function recovered as well.

This case is unique because the patient didn't have any of the previously well documented secondary triggers of hemophagocytic lymphohistiocytosis, which created difficulties and challenges to make the proper diagnosis and management of HLH induced by pregnancy and postpartum. Also, there is not vast literature on the associated of HLH with pregnancy as there were only 14 case reports of HLH during pregnancy.

198

THE LINK BETWEEN LYNCH SYNDROME AND HEREDITARY RENAL CELL CARCINOMA

I Alwine*, JC Henegan. University of Mississippi, Jackson, MS

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Case report Lynch syndrome is an autosomal dominant inherited cancer predisposition syndrome due to germline mutations in the DNA mismatch repair genes *MLH1*, *MSH2*, *MSH6* and

PMS2. Mutation carriers are at markedly increased risk of cancers of the colon, endometrium, ovary, ureter, renal pelvis and skin. However this is not a complete list of malignancies associated with Lynch syndrome. We propose that *MSH6*-variant Lynch syndrome should routinely be grouped with hereditary renal cell carcinoma syndromes as well.

A 62-year-old man with hypertension, diabetes mellitus, and a paternal family history of Lynch Syndrome (diagnosed in the setting of malignant melanoma with a pathogenic variant in *MSH6*) presents with a telangiectatic papule and beads of pigmentation on his left central parietal scalp. A shave biopsy and subsequent wide local excision reveals invasive superficial spreading malignant melanoma, clinical stage IA. A few months later the patient presents to the emergency department for abdominal pain and imaging reveals a right upper pole renal mass. The patient underwent a right radical nephrectomy with pathology consistent with a right renal cell carcinoma, chromophobe variant. He subsequently underwent germline that found he inherited the deleterious *MSH6* variant.

Lynch syndrome is associated with a 50%–80% lifetime risk of colorectal cancer, which is primarily associated with the pathogenic variants in *MLH1* and *MSH2*. In our case a patient with the pathogenic variant in *MSH6* the presenting malignancies were melanoma and renal cell carcinoma, chromophobe variant. Mutational carriers of *MSH6* typically present later in life, carry a smaller risk of colorectal cancer than patients with other Lynch syndrome variants, and have a greater risk for extracolonic malignancies. Although the frequency of renal cell carcinoma or melanoma *MSH6*-variant Lynch Syndrome is not defined, a report from Memorial Sloan Kettering noted 0.6% of cases of renal cell carcinoma and 0.6% of cases of melanoma contained *MSH6* mutations. More research is needed into the mechanisms associating pathogenic variants in mismatch repair genes with renal cell carcinoma.

199

UNGODLY ACT OF JANUS KINASE

N Amilineni*, F Tawadros, K Chakraborty. ETSU, Johnson City, TN

10.1136/jim-2018-000974.197

Case report Essential thrombocythemia (ET) is a chronic myeloproliferative neoplasm (MPN) characterized by excessive clonal platelet production, increasing risk of both thrombosis and hemorrhage. Incidence is between 1 to 2.5 new cases per 100,000, where a single gain of function point mutation in the tyrosine kinase Janus kinase 2 (*JAK2*) gene in exons 14 or 12 has been identified in up to 65 percent of those with ET, distinguishing ET from reactive thrombocytosis. We present an uncommon etiology of cerebrovascular accident (CVA) in adults lacking conventional risk factors.

A 67-year-old male with a past medical history of prior CVA, recurrent deep vein thrombosis (DVT) and bilateral pulmonary emboli (PE), was admitted with left arm weakness. MRI brain demonstrated an acute infarct involving the right frontal lobe, proving to be his second stroke. Cerebral perfusion CT demonstrated a severe partial thrombosis of the right supraclinoid Internal Carotid Artery (ICA).

As the patient lacked relative cardiovascular risk factors, further hypercoagulable workup was ordered. Results were negative for prothrombin gene mutation, anti-cardiolipin

antibodies, lupus anti-coagulant, protein C and S antigen, Factor V Leiden mutation, anti-thrombin 3 and hyper-homocysteinemia to address both arterial and venous events. He was later found to have the JAK2 V617F gene mutation, and bone marrow biopsy confirmed hypercellular marrow consistent with JAK2 +MPN. He was started on cytoreductive therapy for his new diagnosis of ET.

This case highlights the importance of considering JAK2 +MPN as an etiology of unexplained arterial and venous thrombosis in adults with no risk factors. At the time of ET diagnosis, between 9 to 22 percent of patients have already suffered one of the many well-known thrombotic complications—including CVA, myocardial infarction, DVT and PE. Patients often present with multiple vascular complications and carry a diagnosis of refractoriness to anticoagulation. As the median age of ET diagnosis is only 50, earlier testing for JAK2 mutation in this patient population will establish a diagnosis and allow for earlier initiation of appropriate treatment with cytoreduction.

200 DURABLE COMPLETE RESPONSE WITH NIVOLUMAB IN METASTATIC MERKEL CELL CARCINOMA

E Andres*, F Tawadros, S Oad, D Jaishankar. *ETSU, Johnson City, TN*

10.1136/jim-2018-000974.198

Case report Merkel-cell carcinoma (MCC) is a rare, aggressive neuroendocrine tumor of the skin linked to UV light exposure and the merkel-cell polyoma virus. MCC is difficult to treat and often recurs with subsequent poor prognosis. We present a case of an elderly patient with recurrent MCC with a rare right renal metastasis, who had complete response to the PD-1 inhibitor, nivolumab. An 81 year old male presented with a diagnosis of recurrent, metastatic right preauricular MCC. He was treated with surgical excision, parotidectomy and modified radical neck dissection with two positive nodes. Eleven months after adjuvant radiotherapy he was found to have metastatic MCC to the left neck and underwent resection with modified radical left neck dissection. MCC recurred over the right temple 2 months later. Staging studies revealed a 5.4 cm ×6.8 cm right renal mass on PET and MRI was suggestive of metastatic MCC. Due to rarity of kidney involvement a CT urogram was performed and imaging was most consistent with metastatic disease. Urology opted not to biopsy this lesion. Patient initiated immunotherapy with nivolumab 240 mg. He demonstrated excellent dermatological response after two cycles. Interim PET scan showed resolution of nodules over right side of face and neck and shrinking right kidney mass. PET scan following 12th cycle showed no evidence of disease. Patient has completed nineteen cycles with excellent tolerance. He is currently 14 months out from Stage IV diagnosis with no clinical or radiographic evidence of disease. Until recently the mainstay of treatment for metastatic merkel cell carcinoma was multiple agent chemotherapy with cyclophosphamide, doxorubicin and vincristine with poor responses. Avelumab (a PD-L1 inhibitor) tested in patients with chemotherapy-refractory MCC had response rate of 31.7%, with 82% of respondents having ongoing responses at a median follow-up of 10.4 months and was thus approved by FDA. Nghiem et al have demonstrated the efficacy of pembrolizumab as first line treatment in metastatic MCC in phase II trial. This case

provides further support for using first line immunotherapy in metastatic merkel cell carcinoma with a documented durable complete response of 1 year.

201 IMPLICATIONS OF MEAN PLATELET VOLUME IN HEALTH AND DISEASE: A LARGE POPULATION STUDY ON DATA FROM NATIONAL HEALTH AND NUTRITION EXAMINATION SURVEY

S Ball, M Arevalo*, S Wongsangsak, J Dennis, K Nugent. *Texas Tech University Health Sciences Center, Lubbock, TX*

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Purpose of study Mean platelet volume (MPV) is a measure of the platelet size and is an indicator of platelet activation. We conducted a large population study with National Health and Nutrition Examination Survey (NHANES) data to understand the relationship of MPV with health and disease in humans.

Methods used The NHANES is a cross-sectional survey of the non-institutionalized United States adult population, administered every 2 years by the Centers for Disease Control and Prevention. Participants answer a questionnaire, receive a physical examination, and undergo laboratory tests. Values of MPV were collected over a 6 year period (2011–2016). Weighted 10th and 90th percentiles were calculated, and logistic regression was used to predict likelihood [Odds ratio (OR)] of being in categories with MPV <10th percentile or >90th percentile. Statistical analysis was performed using the Stata/SE 15.1.

Summary of results In our study including 17 969 individuals, the mean MPV was 8.40 [SD=0.92] femtoliter (fL), 10th percentile being 7.3 fL and 90th percentile 9.6 fL. Black race and obesity were associated with lower odds of MPV <10th percentile. Male participants, individuals aged 45–64 years, and participants with a recent (last 12 months) hospital-stay were more likely to have an MPV <10th percentile. Obese individuals, Blacks and Mexican Americans had higher odds of having MPV >90th percentile. Individuals with a diagnosis of emphysema had significantly higher adjusted Odds [OR 1.92, 95% CI: 1.11 to 3.31; p=0.021] of having MPV <10th percentile. Individuals with cancer were less likely to have MPV >90th percentile [OR 0.74, 95% CI: 0.55 to 0.99, p=0.042]. A diagnosis of coronary artery disease, congestive heart failure, asthma, and chronic obstructive pulmonary disease were not found to have significant associations with MPV.

Conclusions Our study showed that obese individuals are more likely to have higher MPV. Individuals with emphysema had higher odds of having MPV <10th percentile and those with cancer were less likely to have MPV >90th percentile. Future studies should help determine the utility of MPV in diagnosis and prognosis of clinical conditions.

202 A RARE CASE OF REVERSIBLE PANCYTOPENIA WITH HEMOLYSIS

M Arevalo*, S Ball, S Wongsangsak, A Rakvit. *Texas Tech University Health Sciences Center, Lubbock, TX*

10.1136/jim-2018-000974.200

Introduction Pernicious anemia is a rare autoimmune disease with a 0.1% prevalence in the general population. It is the

primary cause of vitamin B12 deficiency and responsible for 20%–50% of cases. The wide spectrum of clinical manifestations can pose a diagnostic challenge. The most common hematologic sign is anemia, but it can also cause thrombocytopenia, neutropenia, pancytopenia, hemolysis, and pseudothrombotic microangiopathy.

Case report A 45-year-old Hispanic woman presented to the emergency room complaining of hematemesis. She reported a persistent headache for 2 weeks and use of ibuprofen. Laboratory findings were significant for a hemoglobin of 5.3 g/dL, mean corpuscular volume of 114.7 fL, Platelets of 22 K/UL and mild leukopenia. Vitamin B12 was low at 150 pg/mL, Folate was normal. The peripheral blood smear showed hypersegmented neutrophils, macrocytosis, giant platelets, and numerous nucleated red blood cells. An unconjugated hyperbilirubinemia with high LDH (2233 U/L) was suggestive of intravascular hemolysis. She underwent upper endoscopy for hematemesis, which showed diffuse atrophic gastritis. Pernicious anemia was confirmed with positive intrinsic factor antibodies. The patient showed marked improvement in blood counts on cyanocobalamin replacement, and after 3 days of daily subcutaneous and 2 doses of weekly intramuscular cyanocobalamin, all her blood counts normalized.

Discussion Pernicious anemia can rarely present as pancytopenia. Platelet count in this case was unusually low for vitamin B12 deficiency. Upper gastrointestinal bleeding as a presentation is also not typical. The patient had intravascular hemolysis due to ineffective erythropoiesis. Treatment of pernicious anemia is lifelong parenteral replacement with cyanocobalamin. Hematologic manifestations have an excellent response to vitamin B12 however if present and left untreated, neurological manifestations may progress and lead to irreversible sequelae despite B12 replacement. There is an increased risk for all type of gastric tumors in these patients. However, guidelines of endoscopic surveillance are not well established.

Conclusion Reversible systemic causes should be carefully ruled out in a patient with pancytopenia before proceeding to evaluation for primary bone marrow failure syndromes.

203

A CASE SERIES OF SMALL CELL CARCINOMA OF THE BLADDER

KC Arthur*, JC Henegan, S Elkins. *University of Mississippi Medical Center, Jackson, MS*

10.1136/jim-2018-000974.201

Case report Small cell carcinoma of the bladder (SCCB) is a rare and aggressive neuroendocrine bladder cancer. Clinically and histologically, SSCB is like small cell carcinoma of the lung. Optimal treatment for early stage disease is unknown, although cisplatin/etoposide is often first line therapy. We hypothesize that cisplatin and etoposide alone is inadequate initial treatment for SSCB based on three cases of SSCB diagnosed at our institution between January 2017 and August 2018.

Case 1 A 47-year-old female smoker presented with flank pain, and imaging showed a posterior bladder mass. Pathology was mixed small cell and squamous cell. She was Stage IV (cT4a cN1 cM1) at diagnosis. She received 4 cycles of cisplatin/etoposide and progressed on this initial regimen. She

then progressed on all later lines of therapy (including doxorubicin, vincristine, and cyclophosphamide as well as ipilimumab/nivolumab) and passed away less than one year from diagnosis.

Case 2 A 59-year-old male former smoker presented with gross hematuria. Work up revealed a large, invasive bladder tumor. Pathology was mixed small cell and large cell neuroendocrine carcinoma and high grade papillary urothelial carcinoma. He was Stage IIIa (cT3 cN0 cM0) at diagnosis. He received 4 cycles of cisplatin/etoposide and progressed on this regimen with the development of lung metastasis. He is now on treatment with ipilimumab/nivolumab.

Case 3 A 57-year-old male former smoker presented with abdominal pain and hematuria. Pathology from TURBT showed neuroendocrine carcinoma, small cell type with a small component of invasive urothelial carcinoma. He was Stage IVa (T4aN2M0) at diagnosis. He was treated with cisplatin/etoposide and had concurrent whole pelvic radiation.

Discussion Our personal experience with the three cases of SCCB described in this case series has led us to hypothesize that chemotherapy with cisplatin and etoposide is inadequate initial treatment for this uncommon subset of bladder cancer. We propose that a complementary therapy be incorporated into initial therapy such as radiation or, as recently published for small cell carcinoma of the lung, immunotherapy.

204

EXCEPTIONAL RESPONSE TO PACLITAXEL IN PATIENT WITH HEAVILY PRETREATED METASTATIC BLADDER CARCINOMA

G Bader*, JC Henegan. *University of Mississippi Medical Center, Ridgeland, MS*

10.1136/jim-2018-000974.202

Introduction Bladder cancer is the 9th most common malignancy worldwide. Transitional cell carcinoma presents 90% of cases. Patients with locally advanced or metastatic disease will ultimately progress on first and second lines of therapy. Responses to third line or later are limited. Identifying responders to later lines of therapy is important in order to determine treatment selections for future patients.

Case report A 72 y old man was diagnosed with cT2N0M0 muscle invasive bladder carcinoma for which he received neoadjuvant treatment with gemcitabine/cisplatin followed by a cystoprostatectomy (ypT3b ypN0). He relapsed with 1.2 cm nodule in left lung upper lobe, 1 cm nodule in right upper lobe and 2.7 cm right paratracheal node 12, 21 and 31 months later respectively. The first 2 relapses were treated with radiation. The third relapse was managed with gemcitabine/carboplatin since his hearing loss precluded use of cisplatin. After 3 cycles his disease progressed with evidence of left lung upper lobe 4 mm nodule and his treatment was changed to atezolizumab. After 6 cycles his disease continued to progress with evidence of new pulmonary and osseous metastases and he was transitioned to weekly paclitaxel. After 17 weeks of Paclitaxel monotherapy CT and bone scans showed no evidence of disease. Treatment was held. He relapsed 5 months later.

Discussion Most patients with progressive, advanced urothelial carcinoma have poor performance status, impaired renal

function, advanced age and/or significant comorbidities. Thus, effective and tolerable salvage treatment is needed. The limited literature regarding the effectiveness of paclitaxel monotherapy following failure of a platinum based regimen and immunotherapy indicates a low response rate (although good disease control) with low toxicity profile and no kidney injury. Factors that determine response to paclitaxel are unclear. Most patients who previously responded to paclitaxel in palliative setting had already shown some response to initial platinum based treatment.

Conclusion Paclitaxel is a management option for frail patients with previously treated advanced urothelial carcinoma. Future work is needed to determine factors associated with a response to paclitaxel in third line or later treatments.

205 A CAT IN HAT- PATIENT WITH EMPHYSEMATOUS PYELONEPHRITIS AND PERSISTENT LEUKOCYTOSIS FOUND TO HAVE CHRONIC MYELOMONOCYTIC LEUKEMIA (CMML)

G Bedanie*, P Ratanasrimetha, A Sultan, KZ Thein, N DCunha, L Tijani. *Department of Internal Medicine, Texas Tech University Health Sciences Center, Lubbock, TX*

10.1136/jim-2018-000974.203

Introduction CMML is a rare myeloid neoplasm affecting about 1000 patients in US each year. Patient with leukocytosis presenting with infection might have an underlying leukemia that can be masked by the presence of infection. We are presenting a case of CMML stage 0 progressing directly to stage 2.

Case summary A 78-year old woman presented with severe sepsis from right emphysematous pyelonephritis, ended up getting nephrectomy and had multiple admissions later for persistently elevated white blood cell count. On first admission, patient was sick and was in sepsis. Laboratory studies revealed WBC 14 K, Hgb 8.1 mg/dl and platelets 80 K. Peripheral smear showed predominant hyper-segmented neutrophils, left shift and monocytosis. Hematology recommended to continue treatment for infection. Following IV antibiotics treatment, leukocytosis resolved, patient improved and was discharged home. One month later on follow up, WBC was found to be high (36 K), Hgb 9.2 mg/dl and platelets 191 K. Patient was re-admitted and treated with IV antibiotics for possible occult infection. MRI of abdomen/pelvis did not show any collection. Despite prolonged and multiple time treatment, leukocytosis persisted over 2 months. Hematology was re-consulted for further evaluation. On review of labs, patient had persistent leukocytosis with lymphocytosis and monocytosis (WBC 38 K, Hgb 7 gm/dl, PLT, 140 K, ANC 25372, AMC 8000). Patient continued to deny symptoms of infection. Bone marrow biopsy showed CMML with monocytic differentiation and 69% blasts. Cytogenetic analysis revealed 46 XX del [12,p12,16]. Patient was diagnosed with CMML stage 2 and started on Azacitidine with favorable response.

Conclusion CMML is a rare variety of myeloid leukemia; monocytosis is a distinguishing feature. Most common cause of leukocytosis are infection, inflammation or leukemia. CMML is a subtype of leukemia that shows features of myelodysplastic syndrome/myeloproliferative neoplasm diagnosed by bone marrow biopsy. Treatment is chemotherapy.

206 INTRAPARENCHYMAL SCHWANNOMA MIMICKING MALIGNANCY

S Bhama*. *University of Mississippi Medical Center, Jackson, MS*

10.1136/jim-2018-000974.204

Case report Cellular schwannomas are a variety of benign nerve sheath tumors that arise from cranial nerves, spinal roots, or peripheral nerves and comprise 8% of primary brain tumors in adults. Intracranial schwannomas not arising from cranial nerves are exceedingly rare, accounting for less than 1% and are often mistaken for malignant neoplasms.

68 year old Caucasian male with a history of hypertension presented with left lower extremity weakness, gait instability, urinary incontinence and short term memory loss. Magnetic resonance imaging of the brain showed multiple enhancing parafalcine masses, the largest measuring 3.2 cm with mass effect, extensive vasogenic edema and leptomeningeal enhancement. Imaging was concerning for metastatic disease vs a multifocal glioma. Computed tomography of the chest, abdomen and pelvis did not reveal a primary tumor. Cerebrospinal fluid cytology was negative for malignancy. Stereotactic needle biopsy was unsuccessful due to inability to penetrate the firm, rubbery mass and pathology showed scattered atypical astrocytes. Craniotomy and open subtotal resection revealed a pearly white mass with significant calcifications, encasing both pericallosal arteries. Initial pathology showed a spindle cell neoplasm and transitional meningioma with concern for a sarcoma. A second opinion was consistent with a cellular schwannoma with diffuse S100 positivity, but negative for GFAP, SOX10 and EMA.

Intracerebral schwannomas typically demonstrate calcifications, peritumoral edema and cysts. Histologically, cellular schwannomas demonstrate compact spindle cell proliferation (Antoni A architecture) alternating with loose myxoid areas (Antoni B) with rare Verocay bodies. Tumor cells demonstrate nuclear pleomorphism and hyperchromasia, erroneously suggesting malignancy. Since location is variable, differential diagnosis includes malignant peripheral nerve sheath tumor (MPNST), gliomas, meningiomas and melanoma. Compared to its malignant counterparts which require aggressive treatment including chemotherapy, radiation and surgical resection, cellular schwannomas are slow growing benign tumors which can be cured with complete surgical excision. Since such few cases have been reported, more studies are necessary to determine the best course of treatment when a gross total resection is not possible.

207 SONIDEGIB IN NEVOID BASAL CELL CARCINOMA SYNDROME

C Canha*, B Bajric, AE Martinez, F Rana. *University of Florida, Jacksonville, FL*

10.1136/jim-2018-000974.205

Case report A 52-year-old Caucasian male presented with multiple basal cell carcinomas on his face, back, chest, and arms (15 lesions). The patient explained a history of multiple basal cell carcinomas with onset in early adulthood that were managed over the years by plastic surgery with periodic simple surgical excision and Mohs surgery. He referred a prior diagnosis of nevoid basal cell carcinoma syndrome. No other

substantial past medical or family history. Considering the elevated number of lesions and thus impractical surgical therapeutic approach, decision to start Sonidegib was made. The patient was started on Sonidegib 200 mg daily and followed periodically. Initially, monthly and afterwards on a three-month basis. He tolerated treatment with no major adverse effects. Over the course of six months, the patient experienced near complete response with resolution of most basal cell carcinomas and the remaining lesions showed a significant decrease in tumor size.

Discussion Nevroid basal cell carcinoma syndrome, is a rare autosomal dominant neurocutaneous disease characterized by developmental anomalies such as palmar pits and rib anomalies, and tumorigenesis such as medulloblastoma and basal cell carcinoma. It is caused by a mutation of PTCH1 (protein patched homolog) gene, this PTCH gene is an oncosuppressor gene involved in the Sonic Hedgehog Homolog signaling pathway and is crucial in embryonic development, cell division and tumorigenesis. Sonidegib and Vismodegib are hedgehog pathway inhibitors that are promising as future therapeutic options for this syndrome. However, further investigations are necessary to determine whether they have the potential to truly cure individual basal cell carcinomas in nevroid basal cell carcinoma syndrome and to clarify optimal treatment regimen for these patients.

Conclusion The patient tolerated a course of six months with Sonidegib with near complete resolution of all previous basal cell carcinomas and no evidence of skin cancer recurrence. Because of the effectiveness of surgery for the treatment of basal cell carcinoma, experience with systemic chemotherapy is limited. This rare case provides insight on systemic therapy options for basal cell carcinomas when surgery and topical options are not a viable option.

208 A RARE EVOLUTION OF MYELOID MALIGNANCIES

C Capra*, S Elkins. University of Mississippi Medical Center, Jackson, MS

10.1136/jim-2018-000974.206

Case report A seventy-four Caucasian male was referred to Hematology for concern of acute leukemia. He had previously been diagnosed with Essential Thrombocythemia (ET) after presenting with an asymptomatic thrombocytosis three years ago. Bone marrow evaluation prior to therapy was hypercellular (60%) with moderately increased numbers of atypical megakaryocytes. Calreticulin (CALR) gene was found mutated and he was treated with Hydroxyurea for two years until he developed thrombocytopenia. Bone marrow evaluation at this time was suggestive of myelodysplasia with a hypercellular marrow at 45% with normal numbers of microcytic, hypolobulated megakaryocytes, decreased erythropoiesis, and less than 2% blasts. He exhibited a complex karyotype that included deletions of chromosomes 17 and 5, with fluorescence *in situ* hybridization (FISH) confirming a deletion of 5q. He was now diagnosed with myelodysplastic/myeloproliferative neoplasm (MDS/MPN), unclassifiable, and started on Azacitadine with Hydroxyurea. This was continued until July, 2018, when he developed worsening thrombocytopenia with detectable peripheral blasts. Bone marrow evaluation was now markedly hypercellular (>90%) with sheets of large, atypical blasts with occasional blebs. Flow cytometry

revealed 77% blasts expressing CD41a and CD71, confirming evolution into acute megakaryoblastic leukemia. FISH now demonstrated a RUNX1 rearrangement and a deletion of TP53. He received induction chemotherapy with liposomal Daunorubicin-Cytarabine.

Previous case series report the incidence of transformation of ET into AML between 3%–7%, and these patients typically have a more aggressive disease due to frequent deletions in the short arm of chromosome 17 corresponding to a deleted TP53 gene. Standard induction chemotherapy with goal of allogeneic transplant should be sought in eligible patients. Liposomal Daunorubicin-Cytarabine was approved in August, 2017, for the treatment of therapy-related AML and AML with myelodysplastic-related changes. Our patient fell into the latter category, and to our knowledge, is the first reported patient to undergo this specific evolution and treatment.

209 PANNING FOR PRECIOUS METALS: A CASE OF SEVERE PANCYTOPENIA DUE TO COPPER DEFICIENCY

B Case*, S Elkins. University of Mississippi Medical Center, Jackson, MS

10.1136/jim-2018-000974.207

Case report We present a 23-year-old female with a medical history significant for Roux-en-Y procedure four years prior to admission that was complicated post-operatively by enterocutaneous fistula formation, small bowel resection, and infection. She presented to our hospital for worsening abdominal pain and bilateral lower extremity edema, as well as reopening of the enterocutaneous fistula. Admission labs were pertinent for a white count of 0.6 with an ANC 400. Hemoglobin was 6.4, hematocrit was 20.2, and platelet count was 39 000. Other notable labs included an MCV of 108, RDW of 18, and reticulocyte count of 2.2%. Iron studies were normal. Fibrinogen and LDH were normal, as well as INR and aPTT. However, haptoglobin was <10. Albumin was 1.2.

The initial differential diagnosis included primary marrow failure syndromes, nutritional deficiencies, paroxysmal nocturnal hemoglobinuria, or hematologic malignancy. Bone marrow biopsy showed a 30% cellular marrow with trilinear hematopoiesis and no increased blasts. Flow cytometry showered no detectable aberrant marker expression. PNH panel was negative. Vitamin B12 was 949 and folate was 5.5. Copper levels were found to be undetectable and felt to be the most likely cause of pancytopenia. Copper was supplemented in her daily TPN with 1.5 times the daily recommended amount. All cell lines slowly started to improve prior the patient's discharge from the hospital.

Copper deficiency is an uncommon cause of pancytopenia, but much more common in patients who have a history of malabsorptive disorders or prior gastric surgeries. The mechanism of copper deficiency causing cytopenias is multifactorial, including abnormal iron metabolism and mobilization from liver stores, as well inhibition of differentiation and self-renewal of CD34 hematopoietic progenitor cells. The bone marrow can sometimes show vacuoles in myeloid precursors, decrease in granulocyte precursors, and ring sideroblasts, which can mimic MDS. Copper deficiency is an important consideration in patient with cytopenias following gastric surgery.

210

LONG TIME RESPONSE WITH ADO-TRASTUZUMAB EMTANSINE (T-DM1) IN A RECURRENT METASTATIC BREAST CANCER

S Manthri, S Singal, B Youssef, K Chakraborty*. *ETSU, Johnson City, TN*

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Case report Breast cancer (BC) is the most common cancer in women worldwide. The 5-year survival of patients with metastatic disease is estimated at 23%. Ado-trastuzumab emtansine (T-DM1) is a HER2-antibody drug conjugate which incorporates the HER2 targeted actions of trastuzumab with the microtubule inhibitor emtansine, resulting in cell cycle arrest and apoptosis. Currently T-DM1 was approved for the treatment of HER2-positive pre-treated metastatic BC. A 52-year-old female was diagnosed with Triple Positive Stage IIIC multifocal invasive ductal carcinoma of left breast. After completing neoadjuvant chemotherapy based on data from the Neo-sphere trial with dual HER2 blockade, she underwent bilateral mastectomy. Final pathology showed partial response. Post operatively, due to poor wound healing adjuvant treatments were delayed. She received 4 cycles of adjuvant chemotherapy with dd adriamycin, cytoxan and adjuvant radiation therapy. She was started on Q21 days trastuzumab following completion of adjuvant chemotherapy, and adjuvant endocrine therapy was initiated after completion of radiation therapy. Imaging done due to incomplete response to neoadjuvant treatment, showed liver lesions and liver biopsy confirmed recurrence. She was started on T-DM1, endocrine therapy with anastrozole was continued. She was initially receiving T-DM1 Q21 days, subsequently due to grade 1 fatigue and thrombocytopenia it was changed to Q28 days cycle. She is currently status post 37 cycles (20 cycles of Q21 days and 17 cycles of Q28 days). T-DM1 was approved for treatment (single-agent) of HER2-positive, metastatic BC based on phase III data from the EMILIA trial which randomized HER2-positive (HER2+), unresectable, locally advanced or metastatic BC patients. T-DM1 had 9.6 months median PFS vs 6.4 months with lapatinib + capecitabine. Another phase III trial, TH3RESA study, compared T-DM1 to the treatment of the physician's choice in a patient who progressed on two or more anti-HER2 treatments. Median PFS in the T-DM1 arm was nearly double that of patients in the control arm (6.2 vs 3.3 months). Herein, we present a case of a woman with recurrent triple positive metastatic BC with a lengthy progression-free survival on T-DM1 chemotherapy.

211

LIMBIC ENCEPHALITIS AS PRESENTATION OF HODGKIN LYMPHOMA IN A PEDIATRIC PATIENT: THE OPHELIA SYNDROME

¹VA Cruz Flores*, ¹D Miller, ¹P Prasad, ²M Stark, ¹G Tuttrup, ¹D Yu, ¹L Yu. ¹LSU Health Sciences Center, New Orleans, LA; ²Children's Hospital of New Orleans, New Orleans, LA

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Case report A 15 year old female presented with fever and altered mental status, leading to intubation and mechanical ventilation. She was diagnosed with an aseptic meningitis after CSF analysis. Brain MRI was normal. She was treated empirically with antivirals, and made a full recovery in three days. She returned three days later with altered mental status. Brain MRI showed limbic encephalitis. Workup

for autoimmune encephalitis and paraneoplastic syndrome was negative. Despite treatment with steroids and IVIG, she continued to deteriorate neurologically. Prior to plasmapheresis, a central line for plasma exchange was placed, and a post-operative chest X-ray showed a mediastinal mass. Thoracoscopic resection revealed it to be classical nodular sclerosing Hodgkin Lymphoma. After two cycles of chemotherapy, she was in remission of her Hodgkin Lymphoma, and had made a near-complete neurological recovery, with only some remnants of memory loss. Ophelia syndrome is the association of Hodgkin Lymphoma with limbic encephalitis as a paraneoplastic neurological syndrome. There is few literature available about this syndrome, with only 11 cases reported between adult and pediatric patients. Its pathophysiological mechanism is unknown, but is believed to be an autoimmune response to Hodgkin Lymphoma. Antibodies to the metabotropic glutamate receptor 5 (mGluR5), a receptor found mainly in the hippocampus and responsible for behavioral learning and behavior, has been associated with Ophelia syndrome. Literature through published case reports suggest these patients can have a good prognosis if treated promptly, with resolution or near-resolution of neurological symptoms. It is important to recognize the possibility of a paraneoplastic syndrome in a patient with limbic encephalitis, and its association with Hodgkin Lymphoma.

212

WOLF IN SHEEP'S CLOTHING: A CASE OF PROSTATE ADENOCARCINOMA TRANSFORMATION TO SMALL CELL CARCINOMA

C Cutrer*, E Dyer, E Long, K Wilkinson, JC Henegan. *University of Mississippi Medical Center, Jackson, MS*

10.1136/jim-2018-000974.210

Case report A 59 year old Caucasian male with metastatic castrate-resistant prostate cancer (mcrpc) presented to the Emergency Department with worsening weakness and pain, primarily in his chest and back. Additional symptoms included shortness of breath and hematuria. Physical exam was unremarkable. Prostate-Specific Antigen (PSA) was 94.3 ng/mL, which was increased from 12 ng/mL eight weeks prior.

Regarding his prostate cancer, he was diagnosed with *de novo* widespread metastatic castrate-sensitive prostate cancer twenty-two months prior to this admission. At that time He was treated with androgen deprivation therapy (he had not pursued docetaxel chemotherapy due to the need to have bilateral percutaneous nephrostomy tubes in place for urinary tract obstruction) and progressed to mcrpc 14 months prior to this admission. Since the transition to mcrpc he had been on enzalutamide 160 mg daily.

During this admission, computed tomography (CT) showed progression of his cancer including new pulmonary nodules, mediastinal and bilateral hilar lymphadenopathy, and liver lesions as well as progression of his osseous disease. Due to the rapid progression of disease without a proportional rise in PSA, there was concern for dedifferentiation of adenocarcinoma to a small cell type. A subsequent liver biopsy showed poorly differentiated carcinoma with neuroendocrine phenotype, based on strong expression of CD56 and patchy expression of neuron-specific enolase, with negative expression of prostate-specific markers. Prior to changing his

treatment, the patient had an acute change in cognition. CT imaging showed hemorrhagic brain metastases. Ultimately, the patient was discharged to inpatient hospice and expired shortly thereafter.

213

CURIOUS CASE OF NEUROENDOCRINE TUMOR WITH NEGATIVE 5-HYDROXYINDOLEACETIC ACID

M Dogra*, LM Perera, G Ghelani. *SUNY Upstate, Syracuse, NY*

10.1136/jim-2018-000974.211

Case report Gastropancreatic neuroendocrine tumor (NET) is a rare slow-growing cancer. The rarity, non-specific symptomatology and inaccessibility enable these tumors to be overlooked, resulting in delayed and erroneous diagnoses. We present a case of NET which presented as chronic diarrhea and pancreatic mass with negative 5-hydroxyindoleacetic acid (HIAA).

A 40 year old male presented with chronic diarrhea and leg cramps. Lab workup showed hypokalemia (1.8 mmol/L), hypophosphatemia (0.2 mg/dl), hyperchloremic metabolic acidosis (pH- 7.15) and acute kidney injury (creatinine-3.58 mg/dl). Stool osmolal gap was 18 suggesting secretory diarrhea. Stool studies were negative for *Clostridium difficile*, ova, parasites, cryptosporidium, microsporidia, cyclospora, rotavirus antigen, IgA, Tissue transglutaminase IgG, IgA, and anti-gliadin IgG, IgA. Stool cultures were sterile. Urine 5-HIAA and Chromogranin levels were normal. CT abdomen showed left upper quadrant (LUQ) mass arising from the pancreatic tail.

Despite negative HIAA, octreotide scan was done which confirmed increased uptake in the LUQ corresponding with the mass on CT. Endoscopic ultrasound and biopsy confirmed well-differentiated NET of the pancreas. The patient was started on octreotide, niacin and loperamide with improvement in symptoms. He underwent successful surgical resection of the tumor.

Chronic diarrhea can be caused by multiple conditions including irritable bowel syndrome, inflammatory bowel disease, malabsorption syndromes, chronic infections and rarely NET. The investigative strategy is delineated according to the age, comorbidities and clues obtained from the history and physical exam. Stool osmotic gap helps by differentiating between secretory and osmotic diarrhea. Further testing for secretory diarrhea should aim at ruling out infections and structural abnormalities. NET should be investigated after common causes are ruled out. Urine 5-HIAA and chromogranin are generally elevated in NET, however, some patients can be symptomatic with normal or low levels due to fluctuations in levels. Further testing including imaging and biopsy should still be considered if clinical suspicion is high. Our case aims to sensitize physicians to consider NET as a differential in secretory diarrhea, even in patients with negative 5-HIAA levels.

214

NOT SO POETIC CASE OF ABDOMINAL DISTENSION AND DYSPNEA

K Ellard*, M Bravo, LS Engel, S Sanne. *LSU Health Sciences Center, New Orleans, LA*

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Introduction POEMS syndrome is an acronym describing patients who present with the group of symptoms including polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, and skin changes. The etiology of POEMS remains unresolved.

Case A 44 year old man who had been diagnosed with ascites and found to have abdominal lymphadenopathy 6 months prior presented to the hospital with dyspnea, diffuse joint pain, peripheral neuropathy and abdominal swelling and lower extremity edema despite initiation of furosemide. He was tachycardic (pulse 108 bpm) on presentation. On physical exam he appeared chronically ill with a distended, non-tender abdomen without a detectable fluid wave. He had decreased bowel sounds, splenomegaly and bilateral pitting edema to the ankles. His presenting labs were significant for CO₂ of 21 mmol/L, WBCs 12.7 10^3 /uL Hemoglobin 12.3 gm/dL, hematocrit 36.3%, platelets 816 10^3 /uL. Paracentesis on admission was not consistent with spontaneous bacterial peritonitis. Echocardiogram suggested the presence of pulmonary hypertension. The patient underwent a multidisciplinary evaluation for his ascites with SAAG >1.1, thrombocytosis, weight loss, lymphadenopathy, organomegaly, and an osteosclerotic lesion of T10, noted on imaging. Vascular endothelial growth factor was elevated at 264 pg/mL. Kappa light chain and lambda light chain were both elevated at 99.5 mg/L and 92.6 mg/L, respectively. TSH was 12.33 uIU/mL, FT4 was 0.52 ng/dL, and cortisol stimulation test result was 8.4 mcg/dL. Bone marrow biopsy results demonstrated increased plasma cells that rimmed lymphoid aggregates with lambda predominance suggestive of POEMS syndrome.

Discussion The diagnosis of POEMS syndrome requires the presence of two mandatory criteria (polyneuropathy plus monoclonal plasma cell disorder), plus at least one major criteria (osteosclerotic bone lesions, Castleman disease, or elevated serum or plasma vascular endothelial growth factor levels), along with at least one of the six minor criteria (organomegaly, skin changes, endocrinopathy, thrombocytosis, papilledema, extravascular volume overload). The treatment course is multidisciplinary and depends on the severity of disease, but frequently includes chemotherapy, steroids, and treatment of the endocrinopathy.

215

COMPARISON OF DEEP SEDATION REGIMENS FOR CHILDREN UNDERGOING ELECTIVE LUMBAR PUNCTURE

D Fitzgerald*, A Dalabih, S Schexnayder. *University of Arkansas for Medical Sciences, Little Rock, AR*

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Purpose of study Regimens for children undergoing sedation for painful procedures vary considerably by provider and institution. We sought to understand differences in two different regimens used by providers at our institution.

Methods used Review of Sedation Team quality database at Arkansas Children's Hospital. Deidentified patients were selected who underwent lumbar puncture, most of whom also received intrathecal chemotherapy. Patients who received propofol alone were compared with those receiving propofol and fentanyl, based on the choice of the sedation physician.

Summary of results 284 patients who received deep sedation for lumbar punctures (270 with intrathecal chemotherapy) were identified. 59 received propofol alone (P) and 225 received a combination of propofol and fentanyl (PF). 66% and 63% in the P and PF groups were male, respectively. The patients receiving P were similar in age to those receiving PF (P, 10.3 years vs. PF, 9.8 years). Weights (kg)/BMI were similar, 39.8/19.5 for P, and 38.2/19.4 for PF. Patients receiving propofol alone received higher doses of propofol (3.84 mg/kg P, 3.17 mg/kg PF). Fentanyl doses were 0.96 mcg/kg in the PF group. Procedure duration was almost identical in the two groups (16.5 vs. 16.6 min respectively). Major complications were rare and not different between groups (1.7% P vs 4% PF, 95% CI, -5.2 to 6.0). One P patient experienced vomiting and apnea requiring manual ventilation, and nine PF patients experienced the following complications: apnea requiring ventilation, 6 (2.6%); hypoxia requiring increased supplemental oxygen, 2 (0.9%); and hypotension requiring volume administration 1, (0.4%). Recovery times were not statistically different: 17 min and 15 min for the P and PF groups, respectively ($p=0.25$).

Conclusions Both P and PF are safe and effective deep sedation regimens at the doses given for children undergoing elective lumbar puncture. Small opioid doses did not increase complications nor recovery times. Serious complications are rare, but their occurrence reinforces the need for properly trained and credentialed teams with appropriate equipment.

216 FILLED WITH FLUID; A CASE OF CHYLOUS EFFUSIONS RELATED TO MALIGNANT RECURRENCE

¹V Fonseca-Ferrer*, ²D Vera-Santiago, ¹C Rivera-Franceschini, ¹WD Marrero, ¹HR Cintrón-Colón. ¹San Juan City Hospital, Toa Alta; ²Damas Hospital, Ponce, Puerto Rico

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Case report Chylothorax with simultaneous chyloperitoneum has been linked to malignancies but rarely seen in cervical adenocarcinoma. This is the case of a 45-year-old female with medical history of cervical cancer, status post chemoradiation that presented to the Emergency Room with complaints of dyspnea, abdominal distention, bilateral leg edema and fatigue for the past 3 months associated with anorexia and weight loss. Patient denied any symptoms of cough, chest pain, fever, chills, diarrhea, dysuria or recent travel. Chest XR demonstrated blunting of both costovertebral angles and cardiophrenic angles compatible with bibasilar pleural effusions. Chest CT showed evidence of a collapsed left lung due to large left pleural effusion but no pulmonary embolism identified. Thoracentesis of 1.4 liters of turbid orange fluid was done. Pleural/Serum protein ratio was 1.9 (>0.5); Pleural/Serum LDH: 1.52 (>0.6); Pleural Fluid LDH: 392 (>200). Triglyceride and cholesterol levels were 400 mg/dl and 99 mg/dl respectively; consistent with a chylous effusion. Cytology revealed malignant adenocarcinoma. Due to concomitant ascites, patient underwent therapeutic paracentesis, removing 800 ml of milky, salmon-colored fluid. Serum-Ascites Albumin Gradient was 0.3 (<1.1) with total ascites protein of 3.7. Total ascitic fluid cholesterol and triglycerides of 91 mg/dl and 596 mg/dl, respectively. Findings were consistent with chylous ascites. Cytology positive for adenocarcinoma. These confirmed the diagnosis of

chylothorax and chyloperitoneum secondary to metastatic cervical adenocarcinoma. Finally, talc pleurodesis was done with symptomatic improvement. This atypical presentation of both chylothorax and chyloperitoneum is commonly seen in other malignancies such as lymphoproliferative disorders. However, it is an extremely rare occurrence in cervical adenocarcinoma. As a take home message, not all patients presenting with simultaneous ascites and pleural effusions are related to hepatic dysfunction for which further workup needs to be done. To our knowledge, only 2 cases have been reported in literature with chylous effusions related to cervical adenocarcinoma.

217 TREATMENT OF THERAPY-RELATED AML WITH LIVER INVOLVEMENT

¹N Jadhav, ¹A Fuentes*, ²S Moss, ²JJ Schmieg, ¹S Hebert. ¹LSUHSC, Houma, LA; ²Tulane, New Orleans, LA

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Case report Liver involvement in Acute Myeloid Leukemia (AML) is rare and often associated with a poor prognosis. It most commonly presents with a cholestatic pattern of liver failure. However, we report a case of therapy-related AML (t-AML) with liver involvement presenting with a mixed pattern of liver test abnormalities, making selection of a safe and effective treatment course challenging.

A 62-year-old Caucasian female presented with weakness, fever, and icterus for 2 weeks. She was treated with 5-fluorouracil, cisplatin and concurrent radiation for anal SCC 18 months prior. She was found to have pancytopenia and liver failure with elevated AST, ALT, and total bilirubin to 2983 u/L, 3391 u/L, 12 mg/dL, respectively. Marrow biopsy (BM bx) noted 90% blasts with monocytic differentiation, consistent with M5 AML. FISH analysis showed a KMT2A/MLLT3 rearrangement from a t(9;11) translocation. Liver biopsy revealed diffuse leukemic involvement. She was treated with hydroxyurea 2 g BID $\times$ 3 days and methylprednisolone 2 mg/kg daily followed by taper for her liver injury. Unable to receive anthracycline due to her liver function, she was induced with dose reduced cytarabine (50 mg/m²). BM bx on day 21 showed 60% cellularity with $<1\%$ blasts and liver bx on day 25 was without leukemic involvement, indicating remission. Her liver enzymes and bilirubin normalized. Consolidation with liposomal daunorubicin/cytarabine, potentially followed by allogeneic HSCT was planned.

The presentation of AML with acute liver failure is rarely reported and is associated with a high mortality rate. While KMT2A/MLLT3 rearrangements are associated with monocytic t-AML, they often occur 1–2 years after use of topoisomerase inhibitors. 5-FU is rarely reported to lead to t-AML, and only a few reported cases of cisplatin-based regimens without etoposide have been associated with MLL rearranged t-AML.

Induction with standard dose, single-agent cytarabine can offer complete response rates up to 35% in AML. AML with monocytic differentiation is highly associated with organ infiltration. As illustrated in this case, when liver failure is directly due to leukemic infiltrate, careful selection of systemic therapy with appropriate dose reductions can allow for improvement of liver function and treatment of AML with a complete response.

218 A RARE AND UNUSUAL T-CELL LEUKEMIA

GD Gibson*, C Capra, S Bhama, C Bigelow. *University of Mississippi Medical Center, Jackson, MS*

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Case report Adult T-cell lymphoma/leukemia (ATLL) is an aggressive lymphoproliferative malignancy of mature T cells caused by the retrovirus human T-cell lymphotropic virus type 1 (HTLV-1). ATLL is a rare malignancy in the United States accounting for only 0.2% of all lymphoma cases. A 35-year-old African American female with asthma, dyslipidemia, and hypothyroidism presented with unintentional weight loss, anorexia, lethargy, diffuse weakness, abdominal pain, and altered mental status. On presentation, she was tachycardic, lethargic, and confused with a significant delay in answering questions and following commands. Objective data revealed a leukocytosis of $67.1 \times 10^9/L$ with 36% lymphocytes, hypercalcemia measured at 22.0 mg/dL, as well as elevation in transaminases, alkaline phosphatase, lactate dehydrogenase, uric acid, and phosphorus. Peripheral smear identified a subset of atypical lymphoid cells with scant cytoplasm and lobulated nuclei described as 'flower-like' in appearance. Initial treatment with calcitonin, bisphosphonate, and continuous renal replacement therapy produced an improvement in her mental status and resolved the hypercalcemia. Imaging with contrast-enhanced computed tomography scan showed mildly enlarged bilateral axillary and inguinal lymph nodes. Histopathology of bone marrow showed hypercellularity with approximately 10% consisting of neoplastic cells showing CD4 and CD25 by flow cytometry. Peripheral blood polymerase chain reaction confirmed exposure to HTLV-1. Acute variant of ATLL was diagnosed based upon marked lymphocytosis along with hypercalcemia, hyperuricemia, and elevated serum LDH. Patient was started on a chemotherapy regimen of Vinblastine, Cyclophosphamide, Doxorubicin, Prednisone, Carmustine, Etoposide and Carboplatin, a derivation of phase III Japan Clinical Oncology Group (JCOG) Study 9801. There are four clinical subtypes of ATLL: acute, lymphoma, chronic, and smoldering which differ individually in features and prognosis. Acute and lymphoma subtypes are often clinically more aggressive with survival measured in months. Prognosis is often dismal despite aggressive chemotherapy and therefore clinical trials should be encouraged. JCOG 9801 is one of the largest trials to date for ATLL, with high response rates and noted goal of proceeding to allogeneic stem cell transplant.

219 RETROPERITONEAL HEMATOMA, COAGULOPATHY, AND RENAL FAILURE FOLLOWING A HOSPITAL FALL WHILE UNDERGOING TREATMENT FOR SEPSIS. A RARE PRESENTATION OF ACUTE MYELOID LEUKEMIA

C Gregg*, A Burke, AV Varma, LS Engel, S Walvekar. *LSU Health Sciences Center, New Orleans, LA*

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Case report A 65-year-old man with a history of hyperlipidemia and renal calculi was otherwise healthy until three weeks prior to admission, when he developed a fever hours after a routine screening colonoscopy. He was treated with ciprofloxacin but continued to have fever and was admitted to an outside hospital for presumed sepsis secondary to pneumonia.

He improved clinically on broad spectrum antibiotics although he developed an acute kidney injury during that hospitalization. Upon discharge, he had a mechanical fall while leaving the hospital, and presented again the following day with large left-sided subcapsular and retroperitoneal hematomas, which required blood products. He developed an exudative, bloody pleural effusion and his renal function continued to worsen, requiring initiation of hemodialysis. Anemia evaluation showed mild depression in Factor VII level (40%) and slightly prolonged PT, which corrected with a mixing study of normal plasma. He did not improve with Factor VII infusion, and he was transferred to our hospital for evaluation of coagulopathy. Upon presentation to our hospital, he had thrombocytopenia, anemia, and leukocytosis without blast cells. Further hematologic studies were abnormal but not suggestive of a particular etiology. The patient underwent supportive care with blood product transfusions, drainage of recurrent pleural effusions, and hemodialysis. Bone marrow biopsy revealed acute myeloid leukemia (AML) and he was transferred for induction chemotherapy and stem cell transplantation.

Discussion AML accounts for just over one percent of adult cancer deaths in the United States, but it is the most common acute leukemia in adults. AML can often present with abnormalities in one or all cell lines, and it results from the malignant clonal expansion of precursors to hematopoietic cells. Unexplained coagulopathy and infection may be the first presenting signs of acute leukemia or myelodysplasia. Hematologic malignancy should be considered in patients with new bleeding diathesis or unexpected susceptibility to infection.

220 EXTREMELY HIGH PROCALCITONIN: INFECTION OR NOT INFECTION? ... A CASE OF NEUROENDOCRINE TUMOR!

MK Islam*, R Chandra. *Texas Tech University HSC Amarillo, Amarillo, TX*

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Learning objective To recognize neuroendocrine tumor induced procalcitonin production as a noninfectious cause of extremely elevated procalcitonin levels (a scenario that can be mistaken as sepsis).

Case Patient is a 72 year old lady with metastatic non-small cell lung carcinoma of neuroendocrine origin. She had brain and liver metastasis and received chemoradiation therapy. She presented with generalized weakness, poor oral intake and hypotension. Denied any fever, cough or dysuria. Examination was noted for hypotension, normal cardiac and lung exam. She had mild suprapubic tenderness. Labs showed WBC 17000, Hgb 10.1, normal renal and liver function. Her procalcitonin level was very high at 133.1 mcg/L. Urinalysis showed UTI from ESB E Coli sensitive to levofloxacin and meropenem. She was managed as urosepsis with IV meropenem and fluids. Her hypotension resolved quickly with fluid and her repeat urine cultures were negative. Her blood cultures were throughout negative in 4 sets. However her leukocytosis persisted despite IV antibiotics for 2 weeks. Her procalcitonin also remained elevated at 132 mcg/L. Due to persistent elevation of procalcitonin after 2 weeks of antibiotic without any ongoing clinical evidence of infection, work up for tumor induced procalcitonin production was done. The calcitonin levels were drawn and came back as 32150 pg/ml, which is 6000 times normal.

Discussion Procalcitonin level above 10 mcg/L is almost exclusively due to severe bacterial sepsis. Our case highlights an exception to this. Neuroendocrine tumors which are positive for TTF-1 (Thyroid Transcription Factor -1) stain can produce calcitonin which is a byproduct of procalcitonin. Checking calcitonin levels in patients with ongoing leukocytosis and elevated procalcitonin levels without clinical evidence of sepsis may help in shortening antibiotic courses in patients with neuroendocrine tumors. This is an area of future study in infectious disease.

Conclusion This case highlights neuroendocrine tumor as a rare cause of extremely high levels of procalcitonin in humans. Though the patient was treated as urosepsis at the onset, the elevated levels of procalcitonin levels were due to tumor production in this case. Prolonged antibiotic treatment did not diminish the procalcitonin levels.

221 ACUTE MYELOID LEUKEMIA EVERYWHERE EXCEPT THE BONE MARROW: A CASE OF MYELOID SARCOMA WITH MULTI-ORGAN INVOLVEMENT

SM Jeong*, S Elkins, C Bigelow. *University of Mississippi Medical Center, Jackson, MS*

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Introduction Myeloid sarcoma (MS) is an extramedullary tumor of immature myeloid cells that is reported in 2.5%–9.1% of patients with acute myeloid leukemia (AML) and usually occurs concomitantly or following onset of systemic leukemia. Isolated MS, which occurs in the setting of a normal bone marrow biopsy, is a rarer phenomenon with an incidence of 2 cases per million in adults. Common sites of occurrence include the soft tissue, bone, and lymph nodes. Here, we report a case of isolated MS with involvement in the kidney, lungs, bone, spleen, liver, and lymph nodes.

Case report A 53-year-old Caucasian male presented to the ER with abdominal pain and a 15 lb weight loss. CT imaging revealed pulmonary nodules, a 3.9 cm right renal mass, diffuse lymphadenopathy, and a lytic lesion within the left iliac wing. Complete blood count was significant only for microcytic anemia. An excisional left supraclavicular lymph node biopsy was initially concerning for metastatic renal cell carcinoma, however further immunohistochemistry studies revealed myeloid sarcoma.

Repeat CT imaging one month later showed progression of widespread lymphadenopathy, including a lymph node mass encasing the trachea, hypodensities in the liver and spleen, and a 1.4 cm adrenal nodule. A pre-treatment bone marrow biopsy was negative for leukemia. The patient underwent an AML induction regimen unique to our facility comprised of daunorubicin, cytarabine, etoposide, and vincristine. As the patient did not have marrow involvement, CT imaging took the place of a day 14 marrow and revealed minimal change from the start of therapy. Due to his lack of response, a repeat excisional lymph node biopsy was taken from his left neck mass and revealed persistent MS. Unfortunately, the patient had multiple complications during his hospital stay and ultimately decided to pursue hospice care.

Discussion Isolated MS is a rare entity with much of the literature restricted to case reports and small retrospective series. Our patient had evidence of multi-organ involvement including the lung and kidney, two of the lesser common sites. Isolated tumors can arise anywhere in the body, making the diagnosis challenging and requiring a high index of suspicion.

222 NOT YOUR TYPICAL ADOLESCENT ABDOMINAL PAIN

C Johnson*, C Parrish, S DeLeon, HC Allen. *University of Oklahoma Health Sciences Center, Oklahoma City, OK*

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Case report Our patient is a 17 year old female with history of anxiety/depression who presented with abdominal pain and decreased appetite for 1 month. Abdominal pain was intermittent in nature and would last for days, then self-resolve. She presented to our emergency department after 10 days of worsening abdominal pain. On arrival, patient experienced a generalized tonic clonic seizure that resolved with Ativan. Initial exam showed diffuse abdominal tenderness with active bowels sounds and generalized weakness predominantly in her lower extremities. Initial lab work showed hyponatremia, hypomagnesium and dehydration. Patient continued to have generalized seizures due to hypertension with systolic pressures as high as 180 mmHg. CT head showed concern for edema and diffusion restriction. Brain MRI performed demonstrating posterior reversible encephalopathy syndrome. Extensive abdominal work-up, including ultrasound, CT, and stool and urine studies were all negative. Patient was admitted to PICU and was started on Doxycycline for concerns of a tick borne illness. Vitals during admission showed tachycardia and hypertension. Renal ultrasound and urine VMAs were performed to rule out pheochromocytoma; both were normal.

Given her constellation of symptoms, there was a high suspicion for Acute Intermittent Porphyria (AIP). A quantitative urine porphobilinogen was elevated, suggesting a diagnosis of AIP. It is believed that her initial abdominal pain and subsequent decreased appetite before hospital admission led to a starvation state that triggered an AIP attack. AIP specialists were consulted, and hemin was given while inpatient with improvement of all symptoms.

AIP is an autosomal dominant error of heme metabolism. This disorder typically presents in adulthood, most often in women of reproductive age. There are few reported pediatric cases, primarily in females which present soon after menarche. Most common symptoms in children are seizures, abdominal pain, hyponatremia, dehydration, and several psychiatric diagnoses. Elevated urine porphobilinogen is a common lab finding seen with genetic testing showing splice-site mutation IVS4-1G>A in the HMBS gene confirming the diagnosis of AIP. Although a rare diagnosis, it is important to consider AIP when evaluating a patient for intermittent abdominal pain.

223 CHALLENGES IN DIAGNOSING TYPE 2M VON WILLEBRAND DISEASE: THE ROLE OF GENETIC TESTING

R Ksayer*, K Zakharia, M Janbain. *Tulane, New Orleans, LA*

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Case report Von Willebrand Disease (VWD) diagnosis currently requires determining the antigen(Ag)and activity levels of VW protein. Available activity testing include Ristocetin Cofactor activity (RCo) and GPIbM that evaluate VW/platelet interaction;and collagen binding test(CB) that assesses VW/collagen interaction.

Most practices rely on including only one activity testing in their initial evaluation(generally RCo).Some experts recommend

the upfront use of one more test (CB, less frequently GPIbM). Genetic evaluation is required to confirm diagnosis after several non-conclusive Ag/Activity tests.

We report the case of a 28 year old lady, with a history of heavy menstrual bleed and frequent epistaxis. Her family history was positive with a mother and identical twin sister having the same symptoms. She presented for evaluation after the birth of her niece with a spontaneous intracranial bleed. The initial workup showed VWFAg 81.6%, VWF:RCO 41% (RCO:Ag ratio 0.5) with normal VWF multimers. Platelet aggregation studies were normal and factor VIII 100%. On repeated testing values were: VWFAg 67%, VWF:RCO 31% (RCO:Ag ratio 0.46), VWF:CB 68% (CB:Ag ratio 1), and GPIbM 71%. GPIbM did not confirm the low VWF:RCO level. With her strong personal and family history of bleeding, a genetic testing was sent and showed a missense mutation (Ser1731Thr) in exon 30.

This mutation in A3 domain, of autosomal dominant inheritance, is associated with type 2M VWD. Unlike more common mutations associated with type 2 M that occur in A1 domain, with consequent decreased VW-dependent platelet adhesion; this mutation in A3 domain leads to abnormal collagen binding with normal VW/platelet interaction. Previously described cases with this mutation, had decreased VWF:CB and normal RCo:Ag ratio (>0.7). Our patient had normal VWF:CB and low RCo:Ag ratio with normal GPIbM test. Genetic testing was crucial to confirm her diagnosis of VWD.

This is the first case describing an association between this mutation and an isolated decreased RCo activity. In addition, this case highlights the importance of upgrading the rank of genetic testing in diagnosing VWD. Available laboratory assays used in routine evaluation of patients with suspected VWD lack sensitivity and specificity with high inter assay and inter laboratory variability.

224

PANCREATIC ADENOCARCINOMA- THE GREAT MIMICKER?

A Kunnumpurath*, N Mehta, RT Varghese, R Desikan, DG Jacob. *White River Medical Center, Batesville, AR*

10.1136/jim-2018-000974.222

Case report A 79 year-old female was admitted for decreased urine output over 72 hours. She had diffuse abdominal pain without analgesic relief. Over the past month, she had decreased appetite, intermittent nausea and vomiting without weight change. On exam, she was hemodynamically stable without a palpable abdominal mass or lymphadenopathy. Labs were significant for mild leukocytosis (11.6 k/ul), elevated BUN (64 mg/dl), and elevated Creatinine (8.2 mg/d). UA showed 0–2 RBC and was negative for infection. A foley was placed without urine. Renal ultrasound was positive for a possible mass obstructing the right kidney. CT Abdomen showed bilateral perinephric inflammatory changes. Urology performed a retrograde urethrogram and bilateral stent placement, but had difficulty in stenting the right ureter. MRI Abdomen showed an ill-defined lesion within the retroperitoneal region obstructing the second portion of duodenum, proximal right ureter and IVC with an additional two hepatic lesions. BUN and creatinine rose to 94 mg/dl and 12.8 mg/dl respectively and hemodialysis

(HD) was initiated. A nephrostomy tube was placed in the right renal pelvis draining 1200 mL of urine within 24 hours. HD was discontinued. CT-guided liver biopsy was consistent with features of carcinoid, but due to significant expression of GATA-3, a more substantial biopsy was recommended. Chromogranin A was elevated at 631 ng/ml (25–140). An EGD was unable to obtain a biopsy as the scope could not be advanced beyond the obstruction. Exploratory laparotomy was performed. The tumor was unable to be resected. A palliative gastrojejunostomy to relieve the obstruction was performed with biopsies from the mesenteric wall, duodenum and liver. Pathology confirmed a primary pancreatobiliary adenocarcinoma. CEA 1130 ng/ml (0–3) and CA 19–9 6915 u/ml (<34) were elevated. Patient started palliative chemotherapy (Gemcitabine and Abraxane) and died shortly thereafter.

Pancreatic cancer constitutes ~3% of all cancers and those presenting as AKI are rare. Our patient presented with oliguric AKI due to the tumor obstructing Right Kidney. Multiple radiologic modalities were unable to identify the primary and the pancreas appeared to be normal making the diagnosis challenging. Diagnosis of pancreatic cancer can be difficult, more so when it has a rare atypical presentation like AKI.

225

A TALE OF TWO PRIMARY METASTATIC CARCINOMAS

G LaBier*, A Fuentes, TM Reske, LS Engel. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.223

Introduction The incidence of multiple primary cancers although rare, may be increasing due to better diagnostic techniques, prolonged life span and increased long term survival of cancer patients.

Case A 51-year-old man with a history of Diabetes Mellitus Type II, Hyperlipidemia, Chronic Obstructive Pulmonary Disease, and Tobacco Abuse presented to his primary care physician with increasing abdominal pain, bloating, and dark stools. The patient underwent colonoscopy which revealed a friable 4 cm mass spanning 50%–75% of the lumen circumferentially in the rectosigmoid junction. Biopsy demonstrated a moderately differentiated adenocarcinoma. Subsequent CT-imaging revealed five separate rim-enhancing liver lesions. Multiple bilateral pulmonary nodules were also identified. The patient began palliative treatment with a FOLFIRI chemotherapy regimen. Repeat CT imaging after Cycle 9 revealed an interval decrease in size of his largest liver lesion and resolution of all focal solid pulmonary nodules. Approximately 4 months later, routine imaging noted interval increase in size of his metastatic liver lesions. The patient was determined to have progression of disease and he was transitioned to a FOLFOX +BEV and a repeat liver biopsy was obtained. Pathology revealed metastatic squamous cell carcinoma consistent with a pulmonary primary cancer. Further molecular analysis showed high expression of PD-L1 (50%), ROS1 negative, EGFR negative, BRAF negative, and ALK negative.

Discussion The incidence of synchronous colorectal and lung cancer is relatively rare. Multi-centric cancers can be classified by organ involvement and histology types. Furthermore, those malignancies that are observed at the same time or within 6 months are termed as synchronous multiple primary cancers, and those cancers that develop at more than a

6 month interval are termed as metachronous multiple primary cancers. Our patient can be classified as synchronous group II which includes multiple primary cancers that originate from different tissues. Because of the rare incidence, misdiagnosis and delay in treatment can occur. When synchronous colorectal and lung cancer are misdiagnosed as colorectal cancer with pulmonary metastasis, adjuvant chemotherapy for colorectal cancer may worsen patient outcomes.

226 ANY PLATELETS ARE BETTER THAN NO PLATELETS

G LaBier*, A Fuentes, LS Engel, N Masri. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.224

Introduction Immune thrombocytopenia (ITP) is a condition in which autoantibodies develop against platelet antigens leading to destruction of circulating platelets and platelet underproduction.

Case An 87 year-old woman with Diabetes Mellitus Type II, Heart Failure, Hypertension, Stage IV Chronic Kidney Disease, and Mild Alzheimer's Dementia presented with bilateral lower extremity bruising and petechia. She was found to have an isolated thrombocytopenia with a platelet count of 5 K/uL. The patient was deemed to be a poor surgical candidate for splenectomy and thus received intravenous steroids and two doses of 1 mg/kg IVIG with subsequent improvement in her platelet count to 128 K/uL. She was discharged on a course of oral steroids. She represented to the hospital 10 days later with worsening bruising in her chest and upper extremities and was found to have a platelet count of 24 K/uL. She again received intravenous steroids and IVIG without improvement in her platelet count. The platelet count subsequently trended down to 1 K/uL and remained less than 10 K/uL for two weeks. She was successfully treated with 4 doses of rituximab. Additionally eltrombopag, a novel thrombopoietin activator, was started concurrently after the first dose of rituximab. She was seen in hematology clinic 1 month after initiating the eltrombopag and her platelet count improved to 104 K/uL.

Discussion The diagnosis of ITP is based on clinical and laboratory data. Refractory ITP occurs when there is no response to conventional therapy or when disease reoccurs after splenectomy. While splenectomy produces long-term platelet response rates in up to 70% of patients, individuals who are not candidates for this procedure require medical management. Treatment is based on a three tiered approach with all options in a given tier being utilized before advancing to a subsequent tier. The quality of evidence and efficacy is greatest with tier 1 options. Tier 1 consists of low dose corticosteroids, rituximab, and thrombopoietin receptor agonists. Tier 2 regimens include vinca alkaloids, dapsone, danazol, cyclophosphamide, antimetabolites, and cyclosporine A. Tier 3 consists of novel agents mainly utilized in small clinical trials. Fortunately, our patient responded to a combination of therapies given in tier 1.

227 USTEKINUMAB-ASSOCIATED TTP IN A JEHOVAH'S WITNESS PATIENT TREATED WITHOUT PLASMA EXCHANGE

E Long*, S Elkins, M Fryoux. *University of Mississippi, Jackson, MS*

10.1136/jim-2018-000974.225

Case report Thrombotic Thrombocytopenic Purpura (TTP) is a rare hematologic condition characterized by thrombocytopenia and microangiopathic hemolytic anemia (MAHA). Prior to the development of plasma exchange (PLEX) as a treatment, TTP was fatal in almost 90% of cases. With PLEX, survival may be 90%. Unfortunately, Jehovah's Witness (JW) patients cannot undergo PLEX, as it requires the use of plasma, a blood product which is forbidden by their religion. Consequently, treatment of TTP in JW patients must rely on other methods. With an estimated 8.3 million JW worldwide, and an incidence of TTP of 2 cases per million persons per year, TTP in a JW patient is exceedingly rare. We present a case of TTP in a JW treated without PLEX.

A 40-year-old female presented with a three day history of subjective fever and abdominal pain. She had a hemolytic anemia with H/H of 8.7 and 26.1; a platelet count of 12; an elevated LDH, and an undetectable haptoglobin. Peripheral smear showed 3-4 schistocytes per high-power field and thrombocytopenia. Her history was notable for Crohn's disease, for which she was receiving ustekinumab. Patient was a JW.

Patient was diagnosed with TTP, given MAHA and thrombocytopenia. Told that treatment consisted of PLEX with plasma replacement, the patient stated that she could not accept plasma. She could accept plasmapheresis with albumin as a replacement. Given her thrombocytopenia and inability to receive platelets, placing a line for pheresis was risky. She was given IV Solumedrol, and IV Rituximab, and sent to the MICU.

Unfortunately, the patient became obtunded. CT showed no intracranial bleed. She then had a PEA arrest and died. ADAMTS-13 level returned as <1, with a positive inhibitor.

TTP in a JW patient who chooses not to receive PLEX is exceedingly rare. A literature review found 8 cases of successful treatment of TTP in JW without PLEX. Recombinant factor VIII containing ADAMTS-13 has been used, as well as Rituximab and steroids, and Vincristine.

Ustekinumab is a monoclonal antibody against interleukin 12 and 23 used for treatment of Crohn's disease. Reports exist of ustekinumab-induced TTP. In patients who cannot undergo PLEX for religious reasons, use of ustekinumab should be avoided.

228 UNDERSTANDING PAIN EXPERIENCE AMONG TERMINALLY ILL CANCER PATIENTS IN NORTH PALESTINE A CROSS-SECTIONAL DESCRIPTIVE STUDY

¹HM Mallah*, ²R Musa, ²N Bani Fadl, ²S Musmar, ¹S Ball, ¹K Nugent. ¹TTUHSC, Lubbock, TX; ²AnNajah National University, Nablus, Palestine

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Purpose of study Chronic pain is common among terminally ill cancer patients, and it strongly affects their quality of life. Understanding the pain experience and its proper assessment guide its management. In our study, we aimed to evaluate the severity of cancer-related pain and the adequacy of prescribed analgesics in the terminally ill cancer patients in North Palestine.

Methods used A cross-sectional, non-interventional, descriptive study was conducted in North Palestine on 77 terminally ill cancer patients who met the pre-specified inclusion criteria.

Pain experience was evaluated using the Brief Pain Inventory-Short Form (BPI-SF). Social and health profile data were gathered by a data collection sheet through face to face interviews. The pain management index (PMI) was calculated for patients to determine the adequacy of pain management interventions, according to the World Health Organization (WHO) pain ladder recommendations. The relationships between adequacy of pain management and patients' socioeconomic and clinical factors were analyzed by the covariance method. Data were tested by Chi-square and Fisher exact tests.

Summary of results Fifty-nine of the 77 patients (76.6%) studied reported moderate to severe pain. According to the PMI, only 64.9% of the patients received adequate pain management. Thirty-five patients (45%) wished to have additional treatment or an increase in the dose of the pain medications prescribed to them. Although males and females reported similar pain severities, females in the study population were more likely to be inadequately treated ($p=0.027$). Pain severity was significantly different in patients who received healthcare services once or more in last month, as compared to those who did not have access to healthcare ($p=0.024$).

Conclusions Many terminally ill cancer patients were found to suffer from considerable pain and received inadequate treatment in our study. It's recommended that the BPI-SF is used to evaluate pain severity among cancer patients and the adequacy of their management. We also emphasize that physicians shouldn't underestimate patients' complaints of pain and that analgesics should be prescribed equitably without discrimination in regards to gender and socio-economic status.

229 WHEN CURE IS IN SIGHT, TROLLING OF CANCER RESUMES

A Mansurov*, H Rehman, R Zafar, D Jaishankar, K Chakraborty. *East Tennessee State University, Johnson City, TN*

10.1136/jim-2018-000974.227

Case report Breast Cancer is the most common malignancy in women worldwide, with 2 million new cases each year. Though early stages are curable, late metastatic spread is always a possibility. Meta-analysis has revealed that even after 5 years of adjuvant endocrine therapy, recurrences occur with risks ranging from 10% to 41% depending on the tumor size, grade and regional lymph nodes involvement¹. The common sites of distant recurrence have been the bones, liver, lungs and the brain; however, other locations have also been involved with lesser frequency.

A 68 year old female developed weight loss and early satiety. Eight years ago, she was diagnosed with stage IIB hormone receptor positive, HER2 non-amplified invasive Lobular carcinoma. After modified radical mastectomy with axillary lymph node dissection, she received 4 cycles of Doxorubicin and Cyclophosphamide followed by 4 cycles of Docetaxel and radiation therapy. She was on Tamoxifen for 3 years and was switched to aromatase inhibitor for better benefit due to menopause.

Biopsy of a Fundus Mass discovered during endoscopy revealed invasive poorly differentiated carcinoma. No other malignancy was noted in the stomach body or antrum biopsies. Immunohistochemical stains revealed hormone receptor positivity, HER2 amplification and GATA3 positivity. A direct

comparison to previous breast pathology specimen demonstrated nearly identical morphology, supporting the interpretation of the fundus mass to be a late metastatic recurrence of invasive lobular carcinoma. CancerTYPE ID[®] also showed 96% probability of breast adenocarcinoma.

As patient's daughter was recently diagnosed with Breast Cancer, BRCA mutation analysis was performed. Patient is BRCA2 positive. Now she is receiving double Her2 blockade therapy with excellent clinical response.

Recurrence of the breast cancer confined only to the stomach and change in HER2 amplification 8 years after definitive therapy is striking. Thus, with previous history of breast malignancies, the potential for recurrence must always be entertained, regardless of how unlikely the location of recurrence may appear to be.

230 PANCYTOPENIA – A RARE MANIFESTATION OF ISOLATED FOLATE DEFICIENCY IN PREGNANCY

S Manthri*, H Rehman, E Patterson, E Spradling. *ETSU, Johnson City, TN*

10.1136/jim-2018-000974.228

Case report New-onset pancytopenia in second trimester of pregnancy is challenging particularly as the management and outcome of pregnancy are entirely dependent on the underlying etiology. Severe folate deficiency can cause megaloblastic anemia and pancytopenia. We report a case of 24-year-old woman (gravida 6, para 5) who had pre-term spontaneous vaginal delivery at 30 week gestation presented with intractable nausea and vomiting was found to have severe pancytopenia. Complete blood count revealed; hemoglobin of 8.9 g/dL with a mean cellular volume of 81fL, platelets $42 \times 10^9/L$ and neutrophils $1.1 \times 10^9/L$. Her last recorded hematological profile 4 months ago was within normal limits. Reticulocyte count was 0.5% and renal function, liver function tests and coagulation screen were normal. A blood film showed normochromic anemia and no significant red cell fragmentation. Leucopenia and thrombocytopenia were seen with no immature precursors. Ferritin was normal and vitamin B₁₂ level was 288 pg/mL, however serum folate was subtherapeutic at 1.0 ng/mL. An autoimmune screen was unremarkable. An urgent bone marrow (BM) was carried out to exclude a primary hematological disorder. The aspirate showed granulocytic precursors with enlarged metamyelocytes/bands and abnormal nuclear folding. Left shifted erythroid precursors and megakaryocytes with hyperlobated nuclei were seen. However, no leukemic process was found on microscopy, immunophenotyping and flow cytometry evaluation. Karyotype analysis revealed normal XX karyotype. She was started on folic acid supplementation and on day 9 of hospitalization, her counts normalized. Folate deficiency is rare in the Western world and its incidence in pregnancy has been reported to be as low as 0.2%. Folate deficiency if left untreated could progress to severe pancytopenia. Despite the increased folate requirements associated with pregnancy, serum levels are not routinely measured. Hyperemesis during pregnancy and multiparity are recognized as risk factors prompting investigation. Folic acid deficiency causing severe BM failure is an infrequent but possible clinical scenario in pregnancy which can be reversible following prompt diagnosis and treatment.

231 NON PRODUCTIVE COUGH AS PREAMBLE OF RESPIRATORY FAILURE IN A PATIENT WITH HYPEREOSINOPHILIC SYNDROME

¹WD Marrero*, ¹L Piñeiro, ¹HR Cintrón-Colón, ¹V Fonseca-Ferrer, ²R Fernandez. ¹San Juan City Hospital, Villalba; ²San Juan City Hospital, San Juan, PR

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Case report Hypereosinophilic syndrome (HES) is a rare disorder characterized by peripheral blood eosinophilia of $1.5 \times 10^9/L$ or higher and evidence of end organ manifestations attributable to the eosinophilia and not otherwise explained in the clinical setting. HES has an unknown prevalence, most often affects people age 20 through 50. Few patients with prolonged eosinophilia develop organ dysfunction that characterizes HES.

This is the case of a healthy 52 year old man without history of significant systemic illness who presents with non-productive cough, daily febrile episodes and left upper extremity pain for a period 2 months. On physical examination, lungs are clear upon chest auscultation and left sided axillary pain is elicited upon arm abduction. No lymphadenopathy, masses, or organomegaly were detected. Initial blood work revealed severe leukocytosis of eosinophilic predominance (WBC, $40.62 \times 10^3/\mu L$; eosinophil count $23.56 \times 10^3/\mu L$) as well as an elevated sedimentation rate (SED RATE, 112 mm/hr). Chest CT without contrast reported changes in the left axillary vein with associated inflammatory changes in the left axillary lymph nodes. No suspicious supraclavicular, mediastinal, hilar or axillary lymphadenopathy. Further investigation for the cause of eosinophilia and leukocytosis was pursued. Known causes of eosinophilia (parasites, immunodeficiency and malignant disease) were not identified. Patient developed respiratory failure and multiple-organ dysfunction.

HES is a rare heterogeneous disease, any organ system is vulnerable to get affected. The symptoms are common in many other medical problems, making an initial diagnosis more difficult. Sometimes no underlying cause can be found for an observed eosinophilia, even after an extensive diagnostic evaluation. Although HES is rare, our case illustrates the importance of considering diagnosis in patients with significant eosinophilia and have a high index of suspicion in the background of persistent peripheral eosinophilia; after exclusion of secondary causes. Early diagnosis and determination of the specific subtype, followed by appropriate treatment may prevent irreversible organ damage.

232 A RARE CASE OF METASTASIS

D Matrana*, TV Joshi, S Holloway, LS Engel, S Sanne, F Lopez. LSU Health Sciences Center, New Orleans, LA

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Case report A 51 year-old man with squamous cell carcinoma of the oropharynx (T4N2cMx SCC, p16 positive) who recently completed chemoradiation presented to the Emergency Department with worsening leg pain and raised lesions on his legs, arms, back and chest that had progressed over a month. He was most concerned with a larger lesion on his left leg. On Exam he had a 8.2×3.4 cm subdermal lesion on his left leg and other skin lesion of various stages on his body. The patient also had lymphadenopathy in his cervical chain as well as his axilla, bilaterally. He was scanned

radiologically for metastatic evaluation and underwent biopsies of the large left leg lesion and a lesion on his back. Radiologic results were concerning for metastatic disease to his adrenal glands, bilaterally; heart; lungs; skin of chest; left lower extremity; parietal lobes bilaterally; and right occipital lobe. Left lower extremity biopsy of mass revealed moderate to poorly differentiated squamous cell carcinoma that was p16 positive and his back biopsy resulted as squamous cell carcinoma.

Discussion Squamous cell carcinoma of the oropharynx often presents initially as a neck mass, however other common presentations include dysphagia, odynophagia, otalgia or sore throat. In 2018 alone, there are more than 17 000 new cases with the predominant cause being HPV. HPV type 16 causes approximately 60% of oropharyngeal cancers; other leading causes consist of tobacco and alcohol. The initial diagnostic work-up for squamous cell carcinoma includes CT with contrast of primary location and neck; biopsy/FNA of lesion; HPV 16 IHC testing. Chest CT, FDG-PET, EUA with endoscopy occur on as needed basis. As with most malignancies, treatment is based upon staging, p16 positive stage 1 consists of radiotherapy (RT) with possible chemoradiation after RT. Those who have N2 or above, despite their tumor, receive systemic therapy as well as radiotherapy. In regard to prognosis, many factors play a role including presence of HPV infection, cancer stage and smoking history. Five-year survival for patients with oropharyngeal cancer is approximately 65%. Of note, distant metastasis as seen in this case is rare, occurring in approximately 15%–20% of all patients.

233 UNUSUAL PRESENTATION OF ATYPICAL HEMOLYTIC UREMIC SYNDROME IN THE SETTING OF RECURRENT PANCREATITIS IN A PEDIATRIC PATIENT

^{1,2}A McKernan*, ^{1,2}A Bauchat, ^{1,2}D Aviles, ^{1,2}P Prasad. ¹LSU Health Sciences Center, New Orleans, LA; ²Children's Hospital of New Orleans, New Orleans, LA

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Case report Atypical Hemolytic Uremic Syndrome (aHUS) is estimated to affect 3 per 1 million children while acquired thrombotic thrombocytopenic purpura (TTP) affects 1 per 10 million children every year. If untreated, studies have shown TTP to have a mortality rate >90%. These life-threatening conditions are classified as microangiopathic hemolytic anemias (MAHA) and present in similar manners with hemolytic anemia, thrombocytopenia, and renal impairment. We present a case of a 5-year-old male with recurrent pancreatitis, initially diagnosed with TTP but found to have aHUS.

A 5-year-old male with two previous episodes of pancreatitis presented to the emergency department with one day of abdominal pain. Initial blood work showed an elevated Amylase and Lipase. An abdominal CT was concerning for pancreatitis that was subsequently confirmed with MRCP; he was treated with fluid resuscitation and pain control. On day two of hospitalization, patient developed fatigue, jaundice and pallor. A non-immune hemolytic anemia process was confirmed with an acute drop in hemoglobin, schistocytes on peripheral smear, an indirect hyperbilirubinemia, elevated lactate dehydrogenase and negative Coombs testing. He developed an acute kidney injury with gross hematuria and thrombocytopenia with little improvement following repeated platelet and blood transfusions. Clinical picture was

consistent for MAHA with suspicion for TTP. He was empirically treated with serial plasmapheresis and steroids while ADAMTS13 levels were pending. His clinical picture and laboratory abnormalities gradually improved with treatment, and ADAMTS13 levels drawn prior to therapy were normal (97%). Genetic testing was sent for aHUS markers and revealed a pathogenic variant of CD46 (MHP) gene mutation consistent with aHUS.

This case demonstrates an unusual presentation of aHUS in association with pancreatitis. It stresses the importance of a multi-disciplinary approach and initiating potentially life-saving treatment while definitive diagnostic testing is pending. Maintaining a broadened differential and adapting medical therapy based on clinical changes is imperative.

234 ATYPICAL CAUSE OF ATYPICAL HUS: A RARE CASE OF ATYPICAL HUS CAUSED BY DGKE MUTATION

NB Patel*, D Hansen, C Milner. *University of Mississippi Medical Center, Madison, MS*

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Case report Hemolytic uremic syndrome (HUS) is a serious hematological disorder characterized by hemolytic anemia, acute renal failure, and thrombocytopenia. Atypical HUS is associated with one or more genetic mutations that lead to uncontrolled complement activation. We present a rare case of atypical HUS secondary to a DGKE mutation.

This is a 37-year-old white female with history of End Stage Renal Disease (ESRD) status post deceased-donor renal transplant (DDRT) in 1994 who presented with cough and shortness of breath. Patient was treated for post-influenza bacterial pneumonia. A few days after admission, she developed worsening hemolytic anemia, thrombocytopenia, and acute encephalopathy.

Patient began plasma exchange for suspected thrombotic thrombocytopenic purpura (TTP). After two days of plasma exchange, patient's mental status returned to baseline and her hemolytic anemia and thrombocytopenia improved but did not normalize. ADAMTS13 enzyme activity was then found to be within normal limits. With normal ADAMTS13 enzyme activity and absence of an inhibitor, TTP was ruled out. There was no concern for a drug-induced thrombotic microangiopathy as her immunosuppression had been discontinued. Extensive evaluation revealed a heterozygous DGKE mutation thus given the underlying genetic mutation and clinical concern for aHUS, patient was started on eculizumab. She remains on hemodialysis and every 2 week eculizumab with resolution of thrombocytopenia and plans for a potential repeat renal transplant.

Atypical HUS caused by DGKE mutation is a pediatric disorder and it very rarely manifests in adults. DGKE is an enzyme which functions to promote thrombosis via activation of protein kinases. Most cases of DGKE occur in the homozygous setting and are inherited in an autosomal recessive fashion. Our patient had a heterozygous DGKE gene mutation. The treatment of atypical HUS due to DGKE mutation is not well established. Some case reports have demonstrated clinical benefit with plasma exchange and/or eculizumab while others have not. Further investigation is required to better understand the biology of a heterozygous DGKE mutation and definitive therapy especially given cost prohibition with long-term use of eculizumab.

235 A CUTANEOUS COINCIDENCE: MELANOMA AND LYMPHOMA IN A COLLISION TUMOUR

LM Perera*, M Badrinath, K Gada, M Dogra, A Jurdi. *SUNY Upstate, Syracuse, NY*

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Case report The simultaneous occurrence of two different tumors is uncommon. A collision tumor, single entity involving two distinct types of cancer, is even rarer. Here, we report a unique event of melanoma and mantle cell lymphoma presenting together in a cutaneous collision.

Our case involves a 61 year old male with no past medical history who presented with skin lesion progressing in size for one year associated with fatigue and unintentional weight loss. Physical exam was significant for 1 cm wide irregular lesion over base of neck with palpable submandibular and supraclavicular lymphadenopathy. Lab work, including CBC with differential, was normal. Punch biopsy revealed melanoma with Breslow thickness of 0.92 mm. Wide surgical excision showed pathology consistent with superficial spreading type of malignant melanoma, Breslow thickness of 1.2 cm, with infiltration of CD 20+lymphoid cells with irregular nuclear contours, scant cytoplasm and prominent nucleoli. A sentinel lymph node biopsy of the left supraclavicular lymph node showed effacement of nodular architecture by sheets of B-cells positive for CD20, CD5 and BCL2. FISH studies showed presence of t(11;14) (q13;q32) between *IGH* and *CCND1* genes consistent with mantle cell lymphoma. Bone marrow biopsy confirmed extensive involvement with mantle cell lymphoma. PET scan showed diffuse lymphadenopathy involving cervical, mediastinal, hilar, iliac and inguinal chains. Patient was started on R hyper-CVAD therapy with Rituxan, Cyclophosphamide, Vincristine, Doxorubicin and Dexamethasone alternating with high dose Methotrexate and Cytarabine for 6 total cycles of chemotherapy. Repeat PET scan after chemotherapy showed no lymphadenopathy or abnormal metabolic uptake. Repeat bone marrow biopsy showed low normocellular marrow with left shift and macrocytic anemia but otherwise no evidence of mantle cell lymphoma. Given complete remission, autologous stem cell transplant was performed and patient is doing well post-transplant.

Studies show increased association between melanoma and lymphoproliferative disorders, with non-Hodgkin's lymphoma being the most common. The presence of a collision tumour is a rare phenomenon, and to our knowledge, this is the first report of melanoma and mantle cell lymphoma presenting synchronously in a cutaneous collision tumor.

236 INTRADURAL SPINAL METASTATIC NEUROENDOCRINE TUMOR: A CASE REPORT

P Ratanasrimetha*, A Sultan, K Thein, K Nugent, R Tran, L Tijani, F Hardwicke. *Texas Tech University Health Sciences Center, Lubbock, TX*

10.1136/jim-2018-000974.234

Background Metastases to the intradural compartment, extra-medullary or intramedullary, are rare. The majority of spinal metastases are to vertebrae and the epidural space. Patients with intradural spinal metastasis usually present with neurologic symptoms, such as weakness or sensory deficits. Neuroendocrine tumors are uncommon tumors that rarely metastasize to intradural sites in the spine. Also, some spinal

metastases secondary to neuroendocrine tumors do not have an obvious primary site.

Case report 54-years-old woman with history of hypertension and type 2 diabetes mellitus presented with progressive bilateral lower extremities weakness and numbness for 3 weeks. She had history of lower back pain and a 15 pound weight loss during the last 3 months. The family history was positive for breast cancer in her mother. Spinal magnetic resonance images revealed extensive, widespread intradural extramedullary enhancing masses severely compressing the spinal cord at multiple levels. The patient underwent L1 laminectomy and biopsy of an intradural extramedullary lesion. The biopsy result was compatible with a metastatic neuroendocrine tumor. Investigations, including EGD, colonoscopy, CT chest/abdomen/pelvis, failed to reveal a primary tumor site. Tumor marker lab showed CEA 3.2 ng/ml (normal 0.2–3.4), CA 19–9 14.1 units/ml (normal 0–35), AFP 4.0 intl units/ml (normal 0.5–6.9). The patient was treated with chemotherapy (carboplatin and etoposide) and radiation. The radiation therapy improved back the pain and lower extremities weakness.

Conclusion Intradural metastases are relatively uncommon. The prevalence is around 5% of spinal-area metastases. Prevalence of neuroendocrine tumor of unknown primary is around 1% of all tumor of unknown primary site. Treatment of spinal metastasis is individualized but usually involves chemotherapy and radiation. Spinal metastasis treatment aims to relieve pain and preserve neurologic function.

237 WHEN IMATINIB SHUTS DOWN VISION

H Rehman*, A Mansurov, D Jaishankar, K Chakraborty. *East Tennessee State University, Johnson City, TN*

10.1136/jim-2018-000974.235

Case report Discovery of the activating mutation related to c-KIT proto-oncogene resulting in overexpression of c-KIT receptor on tumor cells has dramatically changed the treatment of metastatic gastrointestinal stromal tumors (GIST). Extensive use of Imatinib in GIST and chronic myeloid leukemia requires recognition of a wide spectrum of toxicity. Ocular complications with imatinib therapy such as periorbital edema and epiphora are well known. Acute bilateral vision loss secondary to bilateral optic neuritis is not well established and we report here one such rare but important complication. A 79-year-old man presented with sudden vision loss while receiving palliative imatinib with metastatic GIST. His medical history included locally advanced prostate cancer and poorly controlled hypertension. He initially presented with abdominal pain and work up included abnormalities on imaging studies. He was diagnosed with a 16.5 cm abdominopelvic tumor with peritoneal metastasis on laparotomy. Imatinib 400 mg daily was initiated as therapy with excellent clinical and radiographic response. After six months of treatment patient developed sudden visual loss in both eyes, more so on the right compared to the left. Ophthalmology evaluation was consistent with optic neuritis. MRI Brain and Orbits ruled out space occupying lesions and neurovascular etiology secondary to systemic hypertension. Imatinib was discontinued and tapering steroids administered with significant but

incomplete improvement in visual acuity. Follow up systemic imaging off Imatinib continued to reveal stable disease. Sunitinib as second line therapy is a consideration although there are rare case reports of ocular toxicity with sunitinib as well. Despite being rare, these complications often have significant clinical consequences affecting a patient's quality of life. As a result, we recommend being cognizant of this rare toxicity, advocate expedited ophthalmological evaluation and discontinuation of Imatinib. In an era of personalized medicine with reduced toxicity from targeted therapy or immunotherapy, every clinician, be it a primary physician, hospitalist or subspecialist needs to be alerted about these rare, sometimes bizarre yet devastating side effects.

238 BUT FIRST, DO NO HARM: AN INTERESTING CASE OF CONFOUNDING IATROGENIC COMPLICATIONS

¹D Robbins*, ¹E Dauchy, ²N Nielsen, ¹LS Engel. ¹LSU Health Sciences Center, New Orleans, LA; ²Tulane Health Sciences Center, New Orleans, LA

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Introduction Delayed hemolytic transfusion reaction (DHTR) is defined as a reaction occurring greater than 24 hours, up to 30 days, after blood administration.

Case A 71 year-old woman with hypertension, diabetes, and diverticulosis presented with 5 days of generalized weakness and dizziness associated with intermittent 'dark, dark' stools. She was hypotensive and tachycardic with conjunctival pallor and melena on exam. Laboratory evaluation showed severe anemia with a hemoglobin of 4.4 g/dL, previously 12.4 g/dL. Type and screen was negative for antibodies. She was transfused 3 units of red blood cells with appropriate response. Following endoscopy, she was discharged on propranolol and iron supplementation.

The patient returned 12 days later with hypotension, bradycardia, and respiratory distress. Labs revealed macrocytic anemia with a hemoglobin of 6.6 g/dL. Type and screen was positive for anti-C and anti-Jkb antibodies. LDH was elevated to 774 U/L, and haptoglobin decreased to less than 30 mg/dL. She had a new pre-renal acute kidney injury with a creatinine of 2.51 mg/dL and severe lactic acidosis. Findings were consistent with delayed hemolytic transfusion reaction with gross organ hypoperfusion. She failed medical therapy for beta blocker toxicity and required initiation of transcutaneous pacing and inotropic support. Subsequent review of prior transfusion records revealed a history of anti-C and anti-Jkb alloantibodies. She was transfused antigen-compatible red blood cells with appropriate response. Hemoglobin stabilized without evidence of further hemolysis.

Discussion This case illustrates DHTR confounded by beta blocker toxicity leading to gross organ hypoperfusion. DHTR is characterized by destruction of donor red blood cells due to an anamnestic response to a foreign RBC antigen to which the recipient was previously exposed prior transfusions. In this case, the patient's initial antibody screen was 'negative', however when exposed to the incompatible blood, a robust antibody response was mounted leading to delayed hemolysis. The patient's subsequent hospitalization and complications were direct results of her initial therapy, serving as an important reminder to 'first, do no harm.'

239 ACQUIRED HEMOPHILIA A IN MULTIPLE SCLEROSISA Roy*, A Sasapu. *University of Arkansas for Medical Sciences, Little Rock, AR*

10.1136/jim-2018-000974.237

Case report Acquired hemophilia A (AHA) is a rare bleeding disorder caused by autoantibodies against clotting factor VIII (FVIII) with an incidence of 1.2–1.48 cases per million/year. It occurs most commonly in the elderly population. AHA is seen associated with malignancy, autoimmune diseases, pregnancy, drugs, and blood transfusions. However, up to 50% of reported cases remain idiopathic. AHA is extremely rare in patients with Multiple Sclerosis (MS) and based on the literature only three cases are reported so far. We report a case of AHA in a patient with MS on treatment with glatiramer acetate. A 56-year-old male with MS, diabetes, coronary artery disease presented with 5 week duration of extensive ecchymosis on thighs and arm. There was no personal or family history of bleeding disorders or recent transfusions. Imaging revealed extensive iliopsoas hematoma. He had anemia, normal platelet count, normal liver and renal function. Coagulopathy workup showed normal prothrombin time and prolonged activated partial thromboplastin time (APTT) which was not corrected by mixing study. His FVIII activity was markedly reduced to <1%. Bethesda titer showed elevated inhibitor levels at 30 Units. Fibrinogen, serum protein electrophoresis, von Willebrand factor, cardiolipin antibody were normal. Malignancy workup and hepatitis panel were negative. His acute bleeding improved with Factor Eight Inhibitor Bypassing Activity (FEIBA) and steroids. Rituximab was added for the elimination of inhibitor and steroids were tapered off. FVIII activity and APTT were normalized in 5 weeks and he is currently in remission. AHA should be suspected in any patient with autoimmune disease and new onset of bleeding in the presence of prolonged APTT. Prompt diagnosis and treatment are vitally important as it carries a high mortality. The mainstay of treatment involves acute bleeding control and eradication of the inhibitor. Because of its high relapse rate (15%–33%), continued to follow up of the patients is recommended.

240 PNEUMATOSIS INTESTINALIS FROM CLOSTRIDIUM DIFFICILE INFECTION WITH UNDERLYING TRANSPLANT ASSOCIATED THROMBOTIC MICROANGIOPATHYA Roy*, A Ananthula, M Veeraputhiran. *University of Arkansas for Medical Sciences, Little Rock, AR*

10.1136/jim-2018-000974.238

Case report Thrombotic microangiopathy (TMA) is a lethal complication of allogeneic hematopoietic stem cell transplant (allo HCT). Incidence of TMA post allo HCT is variable between 0.5% and 63.6%. The risk factors include graft-versus-host disease (GVHD), cyclosporin A (CSA), tacrolimus, total body irradiation, high-dose chemotherapy, and infection. We report a case of Clostridium difficile infection in a patient with post-transplant TMA and GI Graft- Versus Host Disease (GVHD) who presented with pneumatosis intestinalis. A 40-year-old female with a history of aplastic anemia, status Day-158 post allo HCT from a matched unrelated donor, presented with abdominal pain, diarrhea and hematemesis. Her post allo HCT was complicated with TMA due to cyclosporine (D+23) which was treated with withdrawal of offending agent CSA and replacement with sirolimus. She developed GVHD of the

gut (D+50). Her GVHD was refractory to steroids but had complete remission to alemtuzumab. She then contracted Clostridium difficile infection (D+147) that was treated with oral vancomycin. Although she responded initially to oral vancomycin she developed abdominal pain and CT scan revealed extensive pneumatosis involving multiple small bowel loops with air within the vessels, extensive portal venous air suggestive for mesenteric ischemia. She had pancytopenia, thrombocytopenia, elevated haptoglobin, abnormal liver enzymes, and creatinine. Her blood smear showed macrocytic anemia with red cell fragments and schistocytes. She underwent surgery with bowel resection not compatible with life. Autopsy revealed transplant-associated TMA leading to intestinal ischemic necrosis and multiorgan failure. Transplant patients with significant intestinal symptoms need to be evaluated for intestinal TMA especially in the presence of infection as they can deteriorate rapidly. Although withdrawal of offending drug is most often the treatment for post allo HCT associated TMA, recent data shows targeted therapy with complement blockade from eculizumab, could be efficacious in selected patient population.

241 SMARCB1 (INI-1) DEFICIENT SINONASAL CARCINOMA PRESENTING AS SCHWANNOMAA Roy*, S Atiq, A Ananthula, O Atiq. *University of Arkansas for Medical Sciences, Little Rock, AR*

10.1136/jim-2018-000974.239

Case report SMARCB1-deficient sinonasal carcinoma is a recently described rare tumor entity of the sinonasal tract characterized by loss of nuclear expression of SMARCB1 (INI1), which acts as a tumor suppressor gene. Loss of function of both alleles gives rise to SMARCB1-deficient tumors. We report a unique case of SMARCB1-deficient sinonasal carcinoma presenting as schwannoma of V2 branch of trigeminal nerve with a morphology entirely composed of high-grade epithelioid malignancy unlike previously described basalioid or plasmacytoid/rhabdoid morphology. A 41-year-old female presented with complaints of right temporal headache, right sided facial numbness, right eye lacrimation, and difficulty opening jaw. Imaging showed nerve sheath tumor of the V2 branch of the trigeminal nerve, suggestive of schwannoma and was excised. Histopathology demonstrated poorly differentiated carcinoma. Postoperative MRI brain showed recurrence of disease. The patient underwent gamma knife stereotactic radiosurgery (GK-SRS) along with stereotactic body radiation therapy (SBRT). Follow-up MRI at 5 months showed extension into pterygopalatine fossa and the right anterior temporal lobe. She underwent temporal craniotomy with decompression. She again underwent GK-SRS and SBRT. Genetic testing, revealing overexpression of EGFR, CDK4, FGFR1, MTOR, MYC, and MYCL, consistent with SMARCB1-deficient sinonasal carcinoma. Palliative chemotherapy with etoposide (VP-16)/cisplatin was initiated. She had received one cycle and then was admitted to the hospital for worsening weakness. Imaging showed extensive dural metastases. She rapidly deteriorated and developed a large hypodense subdural collection with mass effect on the brain causing herniation and death. SMARCB1-deficient sinonasal carcinoma has a very aggressive behavior with most of the patients succumbing to their disease within 2 years of the diagnosis. These tumors can have varied presentations, thus

requiring a high index of clinical suspicion. It is further stated that, while these tumors tend to have an aggressive clinical course, some patients had good outcomes with surgical resection, chemotherapy, and radiation therapy.

242 SQUAMOUS CELL CARCINOMA AND CHRONIC LYMPHOCYTIC LEUKEMIA IN CERVICAL LYMPH NODE BIOPSY – A MIXED PICTURE

P Shelley*, M Fasen, P Reddy. *UF Health – Jacksonville, Jacksonville, FL*

10.1136/jim-2018-000974.240

Background Chronic lymphocytic leukemia (CLL) and Squamous Cell Carcinoma (SCC) are common neoplasias found in the head and neck region. The coexistence of these two malignancies at one location is a rare finding. When a patient presents with CLL, there is a 25% chance that a second primary malignancy exists. There is even less chance that SCC will be the second primary malignancy. Only 12 cases have been reported. In these cases, SCC typically exhibited more aggressive behavior, more frequent recurrence and greater chance of metastasis.

Case description A 58-year-old female with previously diagnosed CLL in a cervical lymph node presented with increased burden of cervical lymphadenopathy. She reported no other past medical history but admitted to extensive smoking history. CT of neck showed mass effect from multiple cervical lymph nodes that was clinically correlated with a recent development of dysphagia. During admission flow cytometry was obtained and showed CD5 +mature b-cell lymphoma with phenotype consistent with CLL. Patient underwent lymph node biopsy to evaluate for Richter transformation. Pathology unexpectedly revealed both mixed CD5 mature B-cell lymphoma and squamous cell carcinoma. Further work-up with laryngoscopy identified a lesion involving the right tonsil that extended into oropharynx and nasopharynx. Biopsies of this lesion showed invasive keratinizing squamous cell carcinoma. She was discharged home with a plan to begin outpatient radiation and chemotherapy.

Discussion This was a fascinating and infrequently encountered case in which a patient was initially diagnosed with CLL and was later found to have SCC. Although the exact pathophysiology responsible for CLL patients' predisposition to harbor additional primary malignancies is unknown, it is believed that CLL leaves the patient immunocompromised, which is a well-known risk factor to the development of malignancies in general. We believe our patient's history of tobacco abuse together with her immunocompromised state put her at risk for multiple malignancies. This emphasizes the importance of recognizing the potential for a second primary malignancy in CLL. Moreover, it is of interest to highlight that the second primary malignancies tend to be more aggressive, which affects prognosis.

243 A DEADLY DUO: ACUTE PROMYELOCYTIC LEUKEMIA COMPLICATED BY CONCURRENT DISSEMINATED INTRAVASCULAR COAGULATION AND TUMOR LYSIS SYNDROME

P Shelley*, B Bajric, J Perez-Downes, M Fasen, P Reddy. *UF Health – Jacksonville, Jacksonville, FL*

10.1136/jim-2018-000974.241

Case report Acute myeloid leukemia (AML) refers to a group of blood cancers that involve myeloid precursor cells. A variant of AML is Acute Promyelocytic Leukemia (APL). It is diagnosed by specific chromosomal translocation, t(15;17), found on bone marrow biopsy. APL is known to have a rapidly fatal course due to the high prevalence of disseminated intravascular coagulopathy (DIC). Tumor lysis syndrome (TLS) is another known complication of many malignancies, including leukemias. It is an oncological emergency that requires aggressive treatment due to its effects on the kidney that leads to metabolic derangements and eventually death. TLS and DIC are oncological complications that need quick diagnosis and rapid treatment. When these diagnoses are present in combination the results can be devastating.

Case A 61-year-old female presented with dizziness. She had a past medical history of stage iia sigmoid adenocarcinoma with previous sigmoidectomy. Initial labs showed anemia and thrombocytopenia, with normal white blood cell count (WBC). Blood smear was found to have numerous blasts concerning for leukemia. Patient was also found to be in TLS with uric acid of 9.1. Bone Marrow biopsy was performed and patient was found to have APL. She quickly decompensated with worsening of her anemia and thrombocytopenia with multiorgan failure. Labs were consistent with DIC. Patient expired 2 days after admission. Her autopsy report showed multiple thrombi throughout myocardial vessels, pulmonary vessels, hepatic blood vessels, with necrotic changes noted in kidney tubules and hemorrhagic infarct of parietal lobe of brain; all findings consistent with DIC.

Discussion AML accounts for approximately 25% of TLS cases. Only 1% of TLS is seen in patients with a WBC count of less than 25 K. In this case, our patient's WBC remained within normal limits. DIC is a distinct coagulopathy disorder noted to be common in APL patients. DIC leads to complicated hemorrhages in these patients. There is a reported 90% of hemorrhages in this group, with hemorrhaging being the most common cause of death. This case stresses the importance of aggressively treating APL along with looking for these secondary manifestations.

244 PROCHLORPERAZINE INDUCED ACUTE CHOLESTATIC HEPATITIS DURING CHEMOTHERAPY

¹K Shou*, ²AS Delmas. ¹Dwight D. Eisenhower Army Medical Center, Augusta, GA; ²Dwight D. Eisenhower Army Medical Center, Fort Gordon, GA

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Introduction Prochlorperazine is a phenothiazine, used commonly for chemotherapy induced emesis. There have been few reported cases of drug-induced cholestasis secondary to prochlorperazine, with a frequency of 0.5%–1% onset within 2–5 weeks. Of those that develop acute cholestatic hepatitis, 7% have a prolonged course. We present a case where clinical picture was initially confounded by chemotherapy initiation with doxorubicin, bleomycin, vinblastine, and dacarbazine (ABVD) for Hodgkin's lymphoma, with prochlorperazine.

Case Our patient is a 58 year old Caucasian male diagnosed with Stage IIA classical, nodular-sclerosing Hodgkin's lymphoma. After his first ABVD treatment he developed scleral

icterus, jaundice, grey colored stools and dark colored urine over the course of one week. Liver enzymes (LFTs) showed cholestatic pattern of injury with peak bilirubin of 20.6 mg/dL, without hemolysis, i.e. grade 4 cholestasis. Work up for autoimmune etiologies was negative and imaging did not support biliary obstruction or gallstones. Liver biopsy was consistent with drug-induced cholestasis. ABVD chemotherapy was initially implicated but there were no reported cases of ABVD induced cholestasis, so prochlorperazine was determined to be most likely culprit. Serial laboratory and clinical assessments showed gradual decline of LFTs and bilirubin. Patient completed chemotherapy with complete remission.

Discussion This is the first reported case of prochlorperazine induced acute cholestatic hepatitis in the setting of ABVD chemotherapy, with symptom onset three days post-exposure. While the mechanism of injury is unknown, prochlorperazine hepatotoxicity is associated with a short latency period, fever, and eosinophilia, suggestive of a hypersensitivity reaction. Dacarbazine does rarely affect the liver, from mild elevation of LFTs to acute liver failure, confounding the initial presentation. It is unknown whether his short symptom onset is due to patient sensitivity versus the combined usage of dacarbazine and prochlorperazine. His alkaline phosphatase and bilirubin improved after restarting ABVD, demonstrating that hepatically metabolized medications can be given safely following drug induced cholestatic hepatitis.

245 METASTATIC BASAL CELL SKIN CANCER MIMICKING BASALOID BREAST CANCER?!

S Singal*, A Mansurov, H Mejbil, K Chakraborty, D Jaishankar. *East Tennessee State University, Johnson City, TN*

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Case report A fifty-year-old lady with Stage I triple negative right breast cancer treated with lumpectomy, chemotherapy and adjuvant radiation in 2015 presented with a lump in the left axilla and neck causing severe left shoulder pain and left arm weakness in 2018. Massive necrotic axillary lymphadenopathy with osseous and pulmonary metastatic disease noted on PET-CT. Biopsy of left neck mass initially reported as metastatic breast carcinoma. Notable outside oncologic history of basal cell carcinoma excision from right forearm in 2011 and multiple excisions left neck/shoulder followed by radiation in 2014. Given timelines, pattern of metastasis and site of relapse in vicinity of previous basal cell carcinoma, review of previous histology requested. Pathology review noted morphology and immunohistochemistry (IHC) concordance between neck biopsy (2018), breast lumpectomy (2015) and previous basal cell carcinoma excisions (2014 and 2011). Tumor characteristics revealed Estrogen receptor (ER), Progesterone receptor (PR) and Her-2 negative status with GATA 3 positive stain common to basal cell skin cancer and basaloid breast cancer. Cancer ID molecular genomic test on breast and neck tissue reported ninety six percent probability of basal cell cancer and less than five percent probability of breast cancer further confirming our suspicion of metastatic basal cell skin cancer. Chemotherapy with carboplatin and

paclitaxel initiated in view of brachial plexopathy and impending loss of limb function with excellent clinical and partial radiologic response. Patient was switched to Vismodegib after completing six cycles of chemotherapy. Metastatic basal cell skin cancer is a rare diagnosis, with metastasis in lymph node, lungs, skin and bone reported in the literature. Basal cell cancer has been very rarely reported to involve nipple areolar complex of breast but to our knowledge metastasis to breast parenchyma is hitherto unreported. Triple negative basaloid breast cancer can have morphology and IHC similar to cutaneous basal cell carcinoma but with widely different tumor biology. This case highlights the importance of assessing pathology in conjunction with the complete clinical picture.

246 FOLLICULAR LYMPHOMA MASQUERADING AS CHOLECYSTITIS AND PANCREATITIS

¹S Singal*, ¹A Mansurov, ²A Patel, ¹K Chakraborty, ¹D Jaishankar. ¹East Tennessee State University, Johnson City, TN; ²ETSU, Johnson City, TN

10.1136/jim-2018-000974.244

Case report Follicular lymphoma (FL), a common type of indolent Non-Hodgkin Lymphoma (NHL), usually presents as peripheral adenopathy with a waxing and waning course. Here we present a case of FL presenting as cholelithiasis and pancreatitis. A 65-year-old male presented with history of nausea, epigastric pain and fever for four days. Labs were notable for transaminitis and hyperbilirubinemia with total bilirubin 4.3. Abdominal imaging revealed cholelithiasis, pancreatitis and mesenteric adenopathy. Patient underwent ERCP with drainage of gallbladder sludge (?) for suspected biliary pancreatitis. Subsequent cholecystectomy and mesenteric lymph node biopsy revealed Grade I follicular lymphoma involving gallbladder and mesenteric lymph node. Persistent transaminitis with no prior history of liver disease precipitated a liver biopsy which revealed hepatic involvement with follicular lymphoma as well. Work up with systemic imaging showed cervical, axillary and mediastinal adenopathy as well as bone marrow involvement with FL. Systemic treatment with Bendamustine and Rituximab was initiated for symptomatic stage IV FL with organ impairment in the form of liver disease. Transaminitis resolved after treatment. Gastrointestinal involvement with FL is infrequent and seen most commonly with involvement of the small intestine particularly the duodenum. Gallbladder involvement is seldom reported and initial presentation of FL as cholecystitis and pancreatitis with transaminitis is rare. Follicular NHL may not need treatment initially, and observation only, is a reasonable approach for asymptomatic patients with low-intermediate prognostic scores. Treatment with chemoimmunotherapy (with CD 20 antibody) is indicated for bulky disease, high stage disease, impending organ dysfunction or symptomatic patients. Treatment is aimed at achieving symptom control and longer disease-free survival given indolent but incurable nature of malignancy. This case highlights the need to keep a low index of suspicion while evaluating patients with seemingly clear-cut diagnoses. The patient was considered to have medication related transaminitis all along. An emphasis on tissue diagnosis is imperative while making a definitive final diagnosis.

247

DEMOGRAPHIC AND PATHOLOGIC CHARACTERISTICS OF TRIPLE NEGATIVE BREAST CANCER DIAGNOSED AT A COMMUNITY HOSPITAL IN SOUTHERN LOUISIANA

¹PM Soto Justiniano*, ²S Donthamsetty. ¹Saba University School of Medicine, The Bottom, Saba, Bonaire, Sint Eustatius and Saba; ²Chabert Medical Center – Ochsner, New Orleans, LA

10.1136/jim-2018-000974.245

Purpose of study Triple Negative Breast Cancer (TNBC) affects African American women disproportionately, it is diagnosed at younger ages with higher tumor grade and it is associated with worse outcomes. This study aims to determine if these demographic and pathologic disparities are present at our institution.

Methods used Hospital electronic medical record system was used to retrospectively study breast cancer cases diagnosed from January 2017 to September 2018. Cases were identified by record keeping of specimens submitted for ancillary testing. The age, race and tumor grade between TNBC and Non-TNBC groups were compared. Contiguous and categorical data were analyzed with T-test and Chi-Square tests, respectively.

Summary of results Amongst 73 diagnoses of breast cancer, 21% were TNBC and 79% Non-TNBC. The average age of diagnosis for TNBC was 61.5 years and 60 years for Non-TNBC. The difference in age between the groups was not significant (p 0.36). The TNBC group was composed of 53.3% Non-Hispanic (NH) Black, 33.3% NH White, and 13.3% Hispanic women. The Non-TNBC had 48.3% NH Black, 48.3% NH White, 3.4% Hispanic women. The difference in ethnicity between TNBC and Non-TNBC was not significant (p 0.25). This study found tumor grade between TNBC and Non-TNBC to be significant ($p < 0.05$). TNBC cases were 60% Grade 3 while 52.6% of Non-TNBC were Grade 2.

Conclusions The majority of TNBC cases were high grade, while Non-TNBC were intermediate grade. The significant difference between the groups is consistent with the known aggressive nature of triple negatives. Age and ethnic composition were not significantly different in either group. Further studies with a larger sample could elucidate this incongruity in our community as compared to the general population and determine whether ethnic and age groups in our population are equally affected by triple negative breast cancer.

248

HEMOLYSIS AND PROFOUND PANCYTOPENIA IN SEVERE VITAMIN B12 DEFICIENCY

J Suria*, K Hoppens, J Dillon, K Zamora, M Bernton, B McConaughy, LS Engel, J Martinez. LSU Health Sciences Center, New Orleans, LA

10.1136/jim-2018-000974.246

Introduction Deficiency of vitamin B12 is a well-known cause of hematologic illness (megaloblastic anemia, pancytopenia, mimicking malign hematologic disorder) and neuropsychiatric illness ('subacute combined degeneration').

Case A 26 year old incarcerated man with history of asthma presented to the hospital with nausea, vomiting, fatigue, decreased appetite, weight loss, and palpitations of 6 weeks duration. He did not experience any bloody or

melanotic stool. He denied any numbness or tingling, easy bruising, skin rashes, recent infections, or hemoptysis. At presentation, he was tachycardic (124 bpm) with an unremarkable blood pressure and respiratory rate. Physical exam revealed conjunctival pallor and delayed capillary refill. He had no lymphadenopathy, bruising, or organomegaly. Serum studies demonstrated a severe pancytopenia with WBC of 2.6 cells/mm³, hemoglobin 2.8 g/dl hematocrit 8.2%, platelets 19/mm³ and MCV of 104.9 fl. Blood smear showed marked anisocytosis, scattered ovalocytes, and increased tear drop cells and schistocytes. Other studies included LDH 4200 mg/dl, haptoglobin <30 mg/dl, B12 <50 ng/ml, Folate 9.4 ng/ml, Ferritin 451 ng/ml, Iron 199 mcg/dl, DAT negative, PTT 28.6 s, INR 1.1, Fibrinogen 383 mg/dl, and absolute reticulocyte count of 0.15 thou/ul. He received 4 units of pack red blood cells with an appropriate response. Intrinsic factor antibody was negative. He was started on daily 1000 mg intramuscular cyanocobalamin injections and daily oral folic acid with improvement of his blood indices.

Discussion We present the case of a previously healthy young man with severe pancytopenia and constitutional symptoms all of which mimic hematologic malignancy but was in fact due to vitamin B12 deficiency. Published data suggest that hematologic and neurologic manifestations are frequently dissociated, with hematologic manifestations almost always preceding neurological manifestations. Both of these manifestations are frequently associated with a very high homocysteine level, very low B12 level and elevated Methylmalonic acid level. Our patient did not manifest neurologic symptomatology despite significant vitamin B12 deficiency.³

249

PULMONARY SARCOMATOID CANCER PRESENTING AS PAINLESS SUBCUTANEOUS NODULES

A Tambe*, H Singh Gambhir, V Tambe. SUNY Upstate Medical University, Syracuse, NY

10.1136/jim-2018-000974.247

Introduction Pulmonary sarcomatoid carcinoma (PSC) is a rare subtype of non-small cell lung cancer comprising of <1% of all lung cancers. Although 1%–12% of all lung cancers can metastasize to skin, cutaneous involvement in PSC has rarely been reported in literature. We present a female who initially presented with subcutaneous nodules and was later diagnosed with PSC.

Case A 67-year-old woman with 50 pack-year smoking history presented with two-week history of cough, shortness of breath and left shoulder pain radiating to left hand. She also endorsed several painless subcutaneous nodules over her back (Left figure 1), spreading to her torso and abdomen over the past two months. On presentation she was tachycardic, hypotensive and tachypneic. Pertinent laboratory findings were leukocytosis and anemia. CT thorax revealed a left lower lobe cavitary lesion with post-obstructive pneumonia secondary to a large left hilar mass. CT abdomen showed a large pancreatic mass with multiple foci of metastatic disease involving the liver, peritoneum, abdominal wall and bones. MRI of the shoulder revealed a mass in the left lung apex abutting the exiting nerve roots at C6 – T2 level (Right figure 1). Biopsy of cutaneous nodule showed sarcomatoid carcinoma with primary in the lung.

Abstract 249 Figure 1

Discussion PSC is highly aggressive tumor more commonly associated with men age 60–70 years and extensive smoking history, the prognosis is poor with median survival rate of 17.4 months. All histological types of lung cancer can metastasize to the skin, and in 20–60 percent of cases the skin lesions present before or synchronously with diagnosis of the primary tumor. Hence patients presenting with subcutaneous nodules in high-risk patients should raise a high suspicion for underlying malignancy such as sarcomatoid carcinoma, metastatic sarcoma, malignant melanoma and pleural mesothelioma.

250

PIPERACILLIN-TAZOBACTAM INDUCED HEMOLYTIC ANEMIA

VV Tambe*, P Desai, A Tambe, M Shah. *SUNY Upstate Medical University Hospital, Syracuse, NY*

10.1136/jim-2018-000974.248

Introduction Drug-induced immune hemolytic anemia (DIIHA) is a rare phenomenon with the use of Piperacillin-Tazobactam. Literature review revealed less than 20 cases that have been described. This can potentially be a life-threatening complication and early recognition of this entity is the key to the patient's survival. We present one such case in an adult male with odontogenic abscess.

Case 56-year-old-man was admitted for evaluation of acute onset anemia from the Infectious disease (ID) clinic where he was being followed for the home infusion of Piperacillin-Tazobactam for management of odontogenic abscess. In ID clinic, his hemoglobin/hematocrit was found to have declined from 15.1/49.6 to 6.5/21 in two weeks and he was advised to go to ER for further evaluation. In the ER, he was hemodynamically stable and apart from lethargy, he had no other symptoms and denied any visible blood loss. His total bilirubin was 2.0 with indirect fraction of 1.1, AST/ALT of 62/67 and ALK of 126. LD was 766. Urine did not show any hemoglobinuria. He also showed significant reticulocytosis of 15.2 and MCV of 112. He was transfused one unit of PRBC and admitted for further workup. He responded well to blood transfusion and lab work showed signs of hemolysis like high LDH, low haptoglobin <20, increased RDW and increased MCV, with bite cells and Heinz bodies seen on the peripheral smear. On abdominal imaging he had hepatomegaly without splenomegaly. Hematology team was consulted for further recommendations who agreed with presence of

hemolysis secondary to Piperacillin-Tazobactam which was immediately discontinued. His hemoglobin recovered slowly over next few days post discontinuation and supportive management.

Discussion Antipseudomonal penicillins are very rarely known to cause hemolytic anemias. The exact mechanism of the DIIHA is not known but possible mechanisms include drug antibodies that are directed at the drug or a combination of drug and erythrocyte membrane protein triggering extravascular hemolysis through interaction with macrophages in the reticuloendothelial system or initiating acute intravascular hemolysis via complement activation. Principle management includes discontinuation of the drug and supportive management.

251

LEPTOMENINGEAL INVOLVEMENT BY PROSTATE CANCER – STRIKE BY AN OMINOUS HYDRA

F Tawadros*, S Manthri, M Zayko, D Jaishankar, K Chakraborty. *ETSU, Johnson City, TN*

10.1136/jim-2018-000974.249

Case report Prostate cancer is the most commonly diagnosed cancer in men. An estimated 1 64 690 new cases of prostate cancer will be diagnosed in 2018 in the United States. Prostate cancer contributes to 9% of male cancer deaths, second only to lung cancer. Skeletal and regional adenopathy are typical metastasis with visceral involvement, infrequent and late. Leptomeningeal involvement by prostate cancer is extremely rare with few cases reported. We present a case of advanced castrate resistant adenocarcinoma of prostate with this highly unusual site of metastatic spread. A 67-year-old male presented with severe nausea and vomiting after failing oral antiemetics. Prior history of stage III prostate cancer for which he underwent radical prostatectomy followed by external beam radiation. Patient was maintained on androgen deprivation therapy (ADT) for five years. Subsequent imaging demonstrated retroperitoneal adenopathy denoting ADT failure. PSA levels continued to rise despite enzalutamide and subsequent abiraterone therapy reflecting disease progression. He maintained excellent functional capacity through 23 cycles of docetaxel chemotherapy. He then developed hip pain with imaging confirming new skeletal metastasis. He received the first cycle of Radium-223 and presented with intractable vomiting. Cranial MRI suspicious for leptomeningeal spread noted. Meningeal biopsy (after negative lumbar puncture-cytology) confirmed pathologic diagnosis. Patient had excellent symptomatic response to high dose dexamethasone and was referred for whole brain irradiation. Currently there is no definitive therapy for leptomeningeal involvement and palliation remains the main aim of treatment with overall poor prognosis. Clinical Oncology is currently witnessing a surge in the incidence of leptomeningeal spread of various cancers due to increased overall survival from effective systemic therapy albeit with poor penetration across the blood brain barrier. Murine studies suggest that Cabazitaxel a new taxane agent may have efficient blood brain barrier penetration. Recognition of this once rare oncologic urgency-emergency by clinicians, is important to effectively diagnose and palliate this condition.

Infectious Diseases/HIV/AIDS

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

252 ECTOPIC ACTH AND NOCARDIA – A DEADLY COMBINATION

A Ananthula*, C Wood, H Goraya, G Krishnan. *University of Arkansas for Medical Sciences, Little Rock, AR*

10.1136/jim-2018-000974.250

Case report Nocardiosis in the context of ectopic Adrenocorticotrophic hormone (ACTH) production is very rare and so far, only 12 cases have been reported. We report a similar case with an aggressive specie; *Nocardia Farcinica*. A 50-year-old Caucasian female with past medical history of perforated peptic ulcer underwent vagotomy, antrectomy, Nissen fundoplication and Billroth I presented with hypertension, tachycardia and altered mental status. She also had recent diagnosis of hyperglycemia, hyperlipidemia, chronic hypokalemia, Vitamin D deficiency, 50 pounds weight gain, and multiple pathological fractures. She had multiple hospital admissions for altered mental status, seizures and pneumonia. Patient had typical cushingoid features. Her cortisol and ACTH level were >123 ug/dL, and 658 pg/dl, respectively. High dose dexamethasone suppression test showed non-suppression of ACTH. CT chest and abdomen showed multiple bilateral non-calcified pulmonary nodules and bilateral adrenal hyperplasia. MRI pituitary protocol ruled out pituitary adenoma. MRI brain showed multiple ring enhancing lesions. She was started on broad spectrum antibiotics for presumed sepsis as well on ketoconazole and eventually also on metyrapone. The source of ectopic ACTH was unknown. Patient further deteriorated and was intubated for hypoxic respiratory failure. Tracheal aspirate showed branching filamentous gram positive bacteria suspicious for *Nocardia*, which later speciated as *Nocardia Farcinica*, which is considered as the most aggressive strain of all *Nocardia* species, and frequently associated with disseminated disease and brain abscesses. Based on sensitivities antibiotics were changed instantly. Bilateral adrenalectomy was deferred due to high perioperative mortality risk given patient further went into multi-organ failure. Family chose the care to be comfort focused and patient died. Hence, early diagnosis and prompt control of hypercortisolism is necessary to prevent life threatening complications and vicious cycle of immunocompromised status, worsening clinical condition due to deadly infection which further leads to high perioperative mortality risk.

253 DISSEMINATED NOCARDIOSIS IN HIV/AIDS

KL Angelle*, AV Varma, LS Engel, S Sanne. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.251

Case A 42 year old homeless man with HIV/AIDS (CD4 count 18, CD4% of 3) who was not taking antiretroviral therapy presented with headaches and shortness of breath. He was previously seen months prior for shortness of breath and treated for community-acquired pneumonia. After discharge,

Culture results from a bronchoscopy grew *Nocardia amikacin-tolerans*. Computed tomographic of his chest on this admission demonstrated consolidation and cavitary focus in the right upper and middle lobe. A brain CT revealed subcortical ring-enhancing lesions in the right fronto-temporal lobe. Intravenous amikacin, and imipenem were added to his a regimen of trimethoprim-sulfamethoxazole. The patient refused further imaging, lumbar puncture and his non-adherence to treatment complicated his hospital course. He developed acute altered mentation with right-side gaze deviation. Repeat CT of the head without contrast showed increased cerebral edema and 8 mm midline shift. The patient was moved to the ICU and switched from imipenem to linezolid due to concern of seizure activity and lowered seizure threshold. His mental status improved and fevers abated. He remained on trimethoprim-sulfamethoxazole, amikacin, and linezolid. The patient developed thrombocytopenia secondary to Linezolid and was switched back to imipenem. He stabilized and was to continue amikacin, trimethoprim-sulfamethoxazole, and imipenem for 6 weeks followed by a prolonged oral antibiotic regimen and repeat imaging. Unfortunately, the patient failed to follow up. **Discussion** *Nocardia* more commonly infects immunocompromised patients, including patients with AIDS. Typically, patients will present with pulmonary findings associated with fever and cough. Infection spreads from the primary site of the lungs to the central nervous system (25%–45%) and skin (10%). The presence of headaches, nausea, vomiting, altered mental status, and seizures may suggest cranial infection. Nocardiosis is usually diagnosed via sputum culture or tissue biopsy. Blood cultures are recommended for immunocompromised patients. Sulfonamides, such as trimethoprim/sulfamethoxazole, have been the drug of choice for *Nocardia*. With disseminated *Nocardia*, multi-therapy with amikacin or imipenem is recommended.

254 SELECTION OF OUTPATIENT ANTIBIOTICS UNCOMPLICATED URINARY TRACT INFECTION IN THE PEDIATRIC EMERGENCY DEPARTMENT

M Hofstetter, S Delozier*, J Bassett, J Dreher, A Murphy, J Stultz, S Arnold. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.252

Purpose of study Urinary tract infection (UTI) is a common reason for antibiotic prescriptions in the emergency department (ED). Antibiotic misuse contributes to antibiotic resistance. We reviewed the microbiology of uncomplicated, UTI in the Pediatric ED determine optimal empiric management.

Methods used This is a retrospective review of children, 1 month to 18 years, with a diagnosis of UTI in 2014–2015, seen at an urban, tertiary-care, Pediatric ED. Patients with underlying genitourinary conditions, immunocompromising conditions or atypical UTI organisms were excluded. UTI was defined as culture growing >10 000 colony forming units (CFU) of a single pathogen. Patient demographics, clinical findings, urinalysis and culture results were collected.

Summary of results There were 827 visits reviewed; 408 (49.3%) met criteria for UTI and were treated for an average of 9 days. The most common etiologic agents were *E. coli* (339 cases, 77.0%), *Proteus* species (16, 3.6%), and *Klebsiella* species (14, 3.2%). Antibiotics prescribed at discharge or after susceptibilities were available included trimethoprim-

sulfamethoxazole (TMP-SMX) (303, 36.7%), cefixime (265, 32.1%), cefdinir (152, 18.4%), ciprofloxacin (48, 5.8%), cephalexin (25, 3.0%). 16.2% of pathogens were resistant to TMP-SMX; 13 discharged with TMP-SMX had a resistant pathogen. Four (1%) patients had pathogens resistant to third generation cephalosporins (3GC) and 12 (2.9%, including the four 3GC resistant) had first generation cephalosporin (1GC) resistant pathogens. Therapy with 3GC covered 99% and 1GC covered 96.8% of organisms for a difference of 2.2%. This means that one would need to treat 52 cases of uncomplicated UTI with a 3GC such as cefdinir or cefixime (instead of a 1GC like cephalexin), to prevent one potential treatment failure due to 1GC resistance.

Conclusions Almost 50% of patients diagnosed with a UTI in our ED did not meet culture criteria for a UTI. Rising resistance to TMP-SMX makes this a suboptimal agent for empiric therapy. 1GC, such as cephalexin, can offer a narrower spectrum alternative to oral 3GC with minimal loss of efficacy due to antibiotic resistance.

255 TOXIC EPIDERMAL NECROLYSIS DUE TO CEFTAROLINE COMPLICATING PERSISTENT MRSA BACTEREMIA

B Bajric*, P Shelley, C Canha, B Attarha, P Reddy. *University of Florida, Jacksonville, FL*

10.1136/jim-2018-000974.253

Case report A 69 year old male with hypertension and end-stage renal disease, on hemodialysis, presented with fevers and general malaise. Work-up revealed blood cultures positive for methicillin-resistant staphylococcus aureus (MRSA). The source was an infected tunneled dialysis catheter. A 2-dimensional trans-thoracic echocardiogram was negative for endocarditis. Treatment with vancomycin was started. Blood cultures remained positive for more than 72 hours. A trans-esophageal echocardiogram was then done, also negative for endocarditis. At this time, daptomycin was added to the vancomycin for the persistent bacteremia. A CT of the Chest, Abdomen, and Pelvis did not show any occult infection. Due to spinal tenderness to palpation, a non-contrast MRI spine was done, showing L3-L4 endplate irregularities and a T2 hyperintense signal within the disc space, suspicious for osteomyelitis. Despite the addition of daptomycin, blood cultures remained positive at 12 days. Vancomycin was discontinued. Ceftriaxone was then added to the daptomycin. Blood cultures were negative on this new regimen 72 hours later. Two weeks later, the patient developed a desquamating, erythematous skin rash, present on his bilateral upper extremities, lower extremities, and much of his trunk and neck. His oral mucosa had multiple crusted and desquamating ulcers. A large punch biopsy of the skin showed a lymphocytic infiltrate with apoptotic keratinocytes at the dermo-epidermal junction. TEN was diagnosed. Given the association between cephalosporins and cutaneous reactions, including TEN, the ceftriaxone was the suspected culprit and was discontinued. The patient was treated supportively, however, ultimately expired from multiorgan failure.

Discussion The therapeutic options for MRSA bacteremia are limited. The first and second line agents, vancomycin and daptomycin, respectively, did not clear the infection. The alternative agents available, including ceftriaxone, are technically not approved for MRSA bacteremias and are often used as salvage therapy in persistent bacteremias. This case illustrates the diagnostic challenge in treating persistent MRSA

bacteremia, further complicated by a life-threatening reaction caused by one of the only available agents for treating the bacteremia.

256 TO CUT OR NOT TO CUT: RIGHT-SIDED INFECTION ENDocarditis WITH PERSISTENT BACTEREMIA

JM Bouchillon*. *University of Alabama Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.254

Case report A previously healthy 6-year-old male presented with fever and suspected bacteremia as a transfer from a community hospital. He was begun on intravenous nafcillin as his initial cultures from the outside facility had speciated as methicillin-sensitive *S. aureus* (MSSA) shortly after transfer. An echocardiogram was obtained which revealed a large (2.5 cm × 1 cm) vegetation adherent to the septal leaflet of the tricuspid valve with trace tricuspid valve regurgitation confirming infective endocarditis (IE). The patient had no history of IE, congenital heart disease, central venous catheters, or rheumatic heart disease. His physical exam was notable for a diastolic heart murmur and palpable splenomegaly but he had no physical exam findings concerning for vascular or immunologic phenomena. Intravenous nafcillin was continued with plans to treat for six weeks. Intravenous gentamicin and oral rifampin were both added on hospital day 13 due to persistent MSSA bacteremia. A multidisciplinary team meeting led to the decision to continue medical management given the risks of operative management despite 16 days of positive blood cultures. His last positive blood culture was obtained on hospital day 16 representing 19 days of positive blood cultures. A peripherally inserted central catheter was placed on hospital day 26 following negative blood cultures for five consecutive days and resolution of fever. He was discharged on hospital day 27 with plans to complete 6 weeks of IV nafcillin in addition to two weeks of intravenous gentamicin and oral rifampin from the time of the last negative blood culture. Shortly after discharge he was re-admitted to for further evaluation of recurrent fever. A repeat echocardiogram demonstrated reduced size of the previously measured cardiac vegetation with disappearance of the pedunculated portion of the vegetation concerning for embolization to the lungs. The Cardiology and Cardiovascular Surgery teams determined that there was low utility for surgical management at that point. He was discharged home following resolution of fever. Outpatient echocardiograms revealed gradual improvement in the size of his cardiac vegetation and a repeat chest CT demonstrated near resolution of prior septic emboli. Intravenous nafcillin was discontinued following roughly 6 weeks of treatment.

257 WHEN THINGS AREN'T AS THEY SEEM: AEROMONAS BACTEREMIA PRESENTING AS AN AKI AND RHABDOMYOLYSIS

BL Boudreaux*, E Dauchy, S Werner, LS Engel, C Sanders. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.255

Introduction *Aeromonas* species are gram negative rods associated with water sources. *Aeromonas* septicemia is rare but is

associated with a high mortality rate of 45%. Immunocompromised patients are at higher risk, especially those who suffered open wounds in aquatic environments and those who have consumed fish or shellfish.

Case A 49 year old African American man presented to the hospital with complaints of chest pain, back and neck pain, and numbness and weakness of his left arm of 2 weeks duration. He had a significant history of alcohol use. On presentation, he was noted to have a leukocytosis, hyperglycemia, elevated lactate, and abnormal liver enzymes, and acute renal injury. During his first night he began to experience leg pain bilaterally which worsened until he could no longer flex or extend his calves without severe pain out of proportion to exam. Out of concern for compartment syndrome, he was taken for left lower extremity compartment fasciotomy. He was started on cefepime as initial blood and urine cultures grew a Gram negative rod which was identified as *Aeromonas hydrophilia* complex. Antibiotic was changed to ceftriaxone. By day 8 he began to develop chest pain and was notably less alert requiring ICU admission and intubation. Antibiotics were broadened to vancomycin, piperacillin/tazobactam, and TMP-SMX. He required a RBC transfusion and hemodialysis (HD). His WBC peaked at 44 cells/mm³ then began to trend down. Antibiotics were de-escalated to ciprofloxacin. His course was further complicated by the development of pericarditis with a large pericardial effusion that demonstrated tamponade physiology. Bloody fluid was drained from his pericardium and cultures of this fluid remained negative as did further blood and urine cultures. He made significant improvements, was able to stop HD, normalized liver enzymes and nearly complete wound healing 2 months after hospital discharge.

Discussion This case was unusual in that the patient had no known water exposures or wounds on his extremities that could have made him susceptible to *Aeromonas hydrophilia*. His undiagnosed type 2 diabetes and alcohol history may have contributed to immunocompromised state.

258 LOWER EXTREMITY LYMPHEDEMA SECONDARY TO KAPOSI SARCOMA

C Canha*, M Morales Hernandez, L Boshnick, P Reddy. *University of Florida, Jacksonville, FL*

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Case presentation A 30-year-old with a history of acquired immunodeficiency syndrome (AIDS), presented with complaints of left lower extremity edema and pain for the past month. On examination, the left lower extremity was markedly edematous and there was minimal edema on the opposite leg. There were multiple violaceous plaques and nodules over the entire cutaneous surface and bilateral inguinal lymphadenopathy involvement. Laboratory studies showed a CD4 T cell count of 52 cells/ μ L. Bacterial, fungal, and mycobacterial blood cultures were unremarkable. Ultrasound Doppler was negative for deep venous thrombosis. Biopsy of a cutaneous lesion and inguinal lymph node demonstrated Kaposi Sarcoma (KS) and the diagnosis was made. Oncology recommended continuous leg elevation and liposomal doxorubicin 20 mg/sq meters every 3 weeks. After one dose, he experienced approximately 20% reduction in tumor burden as well as improvement of his edema.

Discussion KS is an angioproliferative neoplasm that is derived from endothelial cells of blood and lymphatic vessels. Epidemiologic-clinical forms of KS include classic, African, AIDS-associated, and iatrogenic. KS usually manifests as red to violaceous patches, plaques, or nodules and extracutaneous involvement include lymph nodes, gastrointestinal tract, lungs, and liver. Lymph node involvement in AIDS-related KS occurs in 50% of cases and edema is estimated to occur in 20% of patients with KS. The pathophysiology of this lymphedema is not fully understood, proposed mechanisms include obstruction due to the external compression of lymphatics by cutaneous lesions and mixing of blood and lymph secondary to lymphaticovenous anastomoses. Treatment can be local or systemic, and depends on the type and aggressiveness of the KS. Indications for systemic treatment include visceral involvement, extensive lymphedema, extensive and rapidly progressive cutaneous KS and failure to respond to local treatment. Anthracyclines and paclitaxel are typically the agents used to treat KS.

Conclusion KS should be considered as an etiology for lymphedema in patients with AIDS. The ability to recognize this entity can lead to earlier diagnosis, appropriate therapy and prevent the loss of life or limb from this malignancy.

259 SALMONELLA MENINGITIS IN AN INFANT WITH REPTILE EXPOSURE

T Dixon*, M Kaur, F Levent. *Texas Tech Health Sciences Center, Lubbock, TX*

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Introduction *Salmonella* is a motile gram-negative bacteria most commonly associated with food-borne gastroenteritis. In immunocompromised adults, neonates, and people exposed to reptiles, *Salmonella enterica* spp. *arizonae*, one of the non-typhoidal *Salmonella* (NTS) can cause sepsis and bacteremia. Rarely, patients may develop meningitis with high risk for seizures, hydrocephalus, sensorineural hearing loss, and death. Immediate recognition and initiation of treatment is critical.

Case presentation We present an 18-day-old male with a one day history of fever, irritability, and decreased oral intake. He was taken to the local emergency department and was admitted after initial sepsis evaluation. His initial work-up was concerning for meningitis and was empirically started on intravenous (IV) acyclovir, ampicillin, gentamicin, and cefepime. He was transferred to our institution where further history revealed exposure to pet snakes at the aunt's house where the infant and his family were staying. His cerebrospinal fluid (CSF) was later reported to grow gram-negative rods (GNR). Acyclovir was discontinued since the (GNR) in blood and CSF cultures was confirmed to be *Salmonella enterica* spp. *arizonae*. He was treated with ceftazidime at meningitic doses since the isolate was reported to be sensitive and cefotaxime was not available at our institution. Two days after treatment, his symptoms started to improve and repeat blood cultures remained negative. He received total 4 weeks of IV antibiotics. End of therapy CSF parameters, head imaging and hearing screening were within normal limits per age. He had normal physical examination at discharge.

Conclusion Non-typhoidal *Salmonella* meningitis should be on the differential diagnosis in neonates, the immunocompromised, and individuals who were exposed to reptiles who present with signs and symptoms of bacterial meningitis. Although very rare, failure to identify and treat in a timely manner can result in a very poor prognosis.

260

CROUP CAN BE 'COCCI', TOO: A CASE OF A 9-MONTH-OLD BOY WITH SUBGLOTTIC COCCIDIOIDOMYCOSIS MASS PRESENTING AS PERSISTENT CROUP

¹AK Sah, ¹CM Donath*, ²A Heidari, ³L Mandviwala. ¹Ross University School of Medicine, Bakersfield, CA; ²Valley Fever Institute, Bakersfield, CA; ³Kern Medical, Bakersfield, CA

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Introduction Laryngeal coccidioidomycosis is rare and typically presents with upper airway symptoms. Predominant reported cases are seen in adults. Here we present a nine-month-old infant with persistent croup symptoms secondary to laryngeal coccidioidomycosis.

Case report Patient is a nine-month-old Hispanic boy from Bakersfield CA, presented with a two-month history of recurrent laryngotracheobronchitis (croup). The respiratory symptoms started with persistent nonproductive barking cough, inspiratory stridor, hoarseness and a low-grade fever. Initial diagnosis was croup but further workup showed right lower lobe and right middle lobe pneumonia. Patient received two courses of antibiotics, several doses of racemic epinephrine, inhaled and oral steroids over a four months' time with only few days of remission per treatment course. Radiographic imaging showed subsequent subglottic narrowing and retropharyngeal widening. Patient was transferred for higher level of care where he underwent direct laryngoscopy and bronchoscopy.

Laryngoscopy showed an extrapulmonary laryngeal mass encompassing majority of the true vocal cords (TVC) while obstructing the airway and the mobility of the surrounding structures. Biopsies of the laryngeal mass revealed *Coccidioides* spherules with endosporulation. Bronchoscopy was essentially negative. Initial serology was negative but repeat serum complement fixation titer (CF) came back as 1:32 with a serum Immunodiffusion positive for IgG. Patient underwent tracheostomy and started on Amphotericin B. One month later, repeat laryngoscopy showed good TVC movement with no evidence of laryngeal mass; and treatment was switched to fluconazole 120 mg daily. Decannulation of tracheostomy was performed two months after placement. The patient recovered well without swallowing difficulties, speech impairment, hoarseness, stridor or respiratory distress. His most recent serum coccidioidomycosis CF improved to 1:8. He continues to take daily fluconazole 120 mg.

Conclusion Clinicians should be aware of laryngeal coccidioidomycosis in endemic region as one of the differential diagnosis of croup.

261

AN UNSUSPECTED ATTACK OF A RAPIDLY GROWING MYCOBACTERIUM

P Ghimire*, S Khanal, K Paolino. SUNY Upstate Medical University, Syracuse, NY

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Case report We present a 77 year old male with a medical history of Coronary Artery Disease, complete heart block with Cardiovascular Implantable Electronic Device (CIED) and mechanical Aortic Valve on chronic anticoagulation, who was transferred from another hospital for higher level cardiothoracic surgery care for device pocket site pain, swelling, redness, discharge and wound dehiscence, gradually worsening over a week. He also recalled having dry cough and episodes of hemoptysis a few months prior which had since resolved.

The device was promptly removed and replaced with a temporary transvenous pacemaker. He was started on broad spectrum antibiotics vancomycin and cefepime after wound and blood cultures were sent. A transthoracic echocardiogram was obtained to rule out vegetations but was a suboptimal study. After 2 days, he had a permanent pacemaker placed and was transferred back to his other hospital on the same antibiotics. The preliminary stains on the wound culture were positive for acid fast bacilli, which was at the time regarded as skin contaminant.

The patient failed to improve and was sent back to our hospital for worsening collections in the same site along with drainage, pain and erythema. On revisiting the cultures drawn from the first admission, the wound isolate strongly grew *Mycobacterium fortuitum*. New cultures were again drawn and patient was started on empiric treatment with intravenous Amikacin, Cefoxitin and oral Levofloxacin. A CT thorax showed bronchiectasis and subacute appearing reticular opacities diffusely. A transesophageal echo was done which revealed a mobile pedunculated mass on the aortic valve. Thus, a diagnosis of *M. fortuitum* endocarditis and CIED infection was made. His second set of cultures also grew *M. fortuitum*. Progressively through his stay, he started improving and was discharged on a 6 weeks of this followed by further 6 months of doxycycline and linezolid.

Mycobacterium fortuitum is a clinically uncommon rapidly growing mycobacterium which mainly causes pulmonary disease but very rarely can cause surgical site, catheter-related infections or prosthetic valve endocarditis or implant infections. Because our patient's respiratory symptoms prior to presentation and nature of lung CT it was possibly a hematogenous spread, which is even rarer in the immunocompetent.

262

DISSEMINATED TUBERCULOSIS AND DEEP VEIN THROMBOSIS: A CAUSATIVE LINK?

A Goel*, Y Pandey, A Lucas, J Johnsrud, M Burgess. University of Arkansas for Medical Sciences, Little Rock, AR

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Introduction Tuberculosis is a chronic granulomatous infection caused by *Mycobacterium tuberculosis*. Although tuberculosis can affect any organ system, endometrial involvement is uncommon and usually seen with disseminated infection. Deep vein thrombosis is a rare complication of tuberculosis, and very few cases have been reported worldwide. It is proposed to be a consequence of hypercoagulability from systemic inflammation. We present the case of a female patient who was found to have disseminated (pulmonary and endometrial) tuberculosis and common iliac vein thrombosis.

Case presentation A 32 year old, 3 month post-partum, physically active female presented to the hospital with fever, cough, anorexia and weight loss for 2 months. Chest auscultation revealed coarse crackles bilaterally. Chest X-ray showed nodular lesions with central cavitation in both lungs. Beta-HCG was done to rule out gestational trophoblastic disease and was negative. Work-up for HIV and fungal infections was negative. CT of the chest, abdomen and pelvis with contrast revealed miliary nodules in lungs, heterogeneous enhancement of uterus with free fluid and right common iliac vein thrombosis. The patient was started on warfarin and bridged with low-molecular weight heparin. Pelvic ultrasound showed thickened endometrium. Endometrial biopsy revealed chronic granulomatous inflammation and stained positive for acid-fast bacilli. Sputum polymerase chain reaction and culture came back positive for *Mycobacterium tuberculosis*, confirming the diagnosis of tuberculosis. Anti-tubercular therapy with rifampin, isoniazid, pyrazinamide and ethambutol was initiated. Rifampin was switched to rifabutin due to induction of warfarin metabolism by the former. The patient tolerated the therapy well and improved symptomatically.

Discussion This case is unique because it reports the rare complication of deep vein thrombosis in a patient with disseminated (pulmonary and endometrial) tuberculosis. These patients require anticoagulation therapy, in addition to multi-drug anti-tubercular regimen. Rifampin may need to be switched to rifabutin due to induction of warfarin metabolism by the former.

263 FUSOBACTERIUM NECROPHORUM PELVIC ABSCESS IN AN ADOLESCENT MALE

T Harris*, KL Ludwig. *University of Oklahoma Health Sciences Center, Edmond, OK*

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Case report Our patient is a previously healthy 15-year-old male who presented with 12 days of progressive upper left leg pain and fever. Early in his course, he sought outpatient care and was diagnosed with strep pharyngitis based on history alone and was prescribed and completed a 10 day course of amoxicillin. On initial presentation he was found to be febrile with elevated inflammatory markers including: white blood cell count with left shift, erythrocyte sedimentation rate, and a C-reactive protein level. Blood cultures were obtained and X-ray of lower left extremity showed no acute process. Further imaging studies of a left lower extremity magnetic resonance Imaging (MRI) and abdominal/pelvic computed tomography scan (CT) showed a large pelvic abscess at the region of the psoas, with subsequent mass effect and myositis. He was started on antibiotics of ceftriaxone, vancomycin, and metronidazole. Pediatric infectious disease, pediatric surgery, orthopedic surgery, and interventional radiology (IR) were consulted. Recommendations were to have IR place a drain under CT guidance. Drain was placed and initial 300 cc of purulent fluid was drained and sent for culture. Purulent fluid continued to drain for more than 2 weeks. Blood cultures had no growth, but culture of abscess fluid grew *Fusobacterium necrophorum*. Patient continued to be febrile while in hospital and required 2 weeks of IV antibiotics. CT done at 2 weeks of hospitalization showed improvement, but continued to show a large and unresolved

complex collection. Due to difficult anatomical location of abscess, no surgical intervention was recommended. He was transitioned to oral antibiotics without issue and took cefdinir, metronidazole, and sulfamethoxazole/trimethoprim outpatient for 4 weeks. Repeat imaging at follow-up showed improved pathology.

Fusobacterium necrophorum is a common bacterium of gut and oral flora and is a leading cause of pharyngitis and tonsillitis in young adults and adolescents. Cases of *Fusobacterium* abscesses consist of hematogenous spread and mainly occur in the liver. Our patient had a negative blood culture, complicated by use of antibiotics previously prescribed. This case presents a unique location for a *Fusobacterium* abscess in an otherwise healthy, adolescent with an unidentified cause.

264 AEROMONAS SOBRIA: A RARE AND FULMINANT CAUSE OF SEPTIC SHOCK IN HEPATOBIILIARY DISEASE

JM Hernandez*, L Boshnick, KR Green, AE Martinez. *University of Florida College of Medicine, Jacksonville, FL*

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Case report A 59 year-old female presented to the emergency room after being found down at home in large amount of diarrhea. Medical history was significant for cholecystectomy in 2005 and chronic pneumobilia. She was febrile, hypotensive and tachycardic. Reported non-bloody diarrhea and abdominal pain for 3 days. Vancomycin and piperacillin-tazobactam were started. Patient deteriorated requiring intubation and vasopressors. Computer tomography revealed new biliary ductal dilatation with possible distal common bile duct obstruction concerning for ascending cholangitis. Workup showed transaminitis in the 100 s, direct bilirubin 3.0 mg/dL, INR 1.9. Blood cultures grew Gram negative rods. Speciation confirmed *E. coli*. Gastroenterology attempted emergent endoscopic retrograde cholangiopancreatography but patient had cardiac. An emergent percutaneous biliary drain was placed. Her liver function continued to decline, and required continuous renal replacement therapy. *Aeromonas sobria* speciation was later reported along with sensitivities, demonstrating pansensitive *E. coli* and *A. sobria* resistant to piperacillin-tazobactam. Levofloxacin was initiated. Despite antibiotic optimization patient entered acute liver failure and expired due to cardiac arrest.

Aeromonas are motile Gram-negative rods that are ubiquitous in water and can colonize humans. There are 3 defined species: *A. hydrophila*, *A. caviae* and *A. veronii biovar sobria*. These organisms are unusual and emergent pathogens. *Aeromonas* infections occur in healthy patients, but immunosuppression, malignancy, and hepatobiliary disease are the greatest risk factors. *A. sobria* is considered the most pathogenic species, and a serine proteinase secreted at infection sites could be linked to septic shock induction. In our patient, translocation of *Aeromonas* across the hepatobiliary tract is most likely the source of bacteremia. *Aeromonas* bacteremia has a fatality rate 30–70%. These pathogens can produce at least 3 β -lactamases.

Clinicians should verify involvement of *Aeromonas* as infection culprit in patients with gastrointestinal/hepatobiliary disease when treatment with β -lactams is refractory. β -lactams should be used with caution for the treatment of *Aeromonas* infections.

265 BILATERAL CAVERNOUS SINUS THROMBOSIS COMPLICATING PANSINUSITIS IN HIV PATIENT

JM Hernandez*, K Vahdat, JR Rivas, I Serrano, J Ruiz, C Isache. *University of Florida College of Medicine, Jacksonville, FL*

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Case report A 64 year-old male with HIV and polysubstance abuse presented to the emergency room complaining of left (L) eye swelling and tearing for 2 months. Referred decreased vision, pain with eye movement, coryza, fever and vomiting. Physical exam was remarkable for bilateral (BL) severe chemosis. His L eye was down and out with proptosis and hyperemia. He exhibited elevated intraocular pressure (IOP) of 25 mmHg in L eye and 28 mmHg in right (R) eye, and impaired visual acuity of 20/200 in L eye and 20/30 in R eye. He had leukocytosis ($22.57 \times 10^3/\mu\text{L}$) and elevated sedimentation rate (35 mmhr). CT orbits showed complete opacification of the paranasal sinuses. Vancomycin, cefepime and metronidazole were started. CT venogram (CTV) revealed thrombosis of BL superior ophthalmic veins, anterior cavernous sinuses, and L sigmoid sinus and jugular bulb. Functional endoscopic sinus surgery was done. Intraoperative cultures yielded coagulase negative *Staphylococcus aureus*. Patient was instructed to complete 6 weeks of antibiotics and warfarin was started. CTV repeat one month post discharge showed recanalization of the superior ophthalmic veins and continued filling defect within the L transverse sigmoid and L jugular bulb. Blurry vision, photophobia, and worsening visual acuity to L eye persisted 5 months post event.

CST is rare, representing 0.5%–1% of all strokes. *Staphylococcus aureus* is the most common isolated pathogen when infection is the cause. Mortality is around 20%–30%. Most common signs result from direct injury to cranial nerves III–VI and impaired venous drainage from the orbit and eye. This condition can have dramatic local and metastatic complications including carotid thrombosis, subdural empyema, brain abscess, meningitis, and septic embolization. Long-term sequelae occur in up to 50% of patients who recover. Imaging techniques confirm the diagnosis, and antibiotherapy is the mainstay of therapy.

A high index of suspicion is crucial for a prompt diagnosis of CST. Antibiotics are mainstay of treatment when infection is culprit, whereas anticoagulation and steroid use remain controversial. Nonetheless, recent evidence favors their use. Long-term follow up is warranted as relapses can occur after apparent resolution.

266 VARICELLA ZOSTER IN A 2-MONTH-OLD INFANT BORN TO A VARICELLA IMMUNE MOTHER

^{1,2}M Hood*, ^{1,2}C Costello, ^{1,2}B Bagga. ¹University of Tennessee Health Science Center, Memphis, TN; ²Le Bonheur Children's Hospital, Memphis, TN

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Case report It has been reported that maternal acquired antibodies decrease below the 150 mIU/ml threshold at 3–4 months, with a strong inverse correlation between antibody levels and varicella complication. We report a post-natal varicella infection in a 2-month-old born with maternal history of childhood varicella.

A 2-month-old full-term male with no past medical history presented with a 2 day history of generalized vesicular

rash. Both parents immigrated to US from Mexico in adulthood; Maternal history significant for childhood varicella-zoster virus (VZV) infection. Father reported a recent similar exanthematous illness 2.5 weeks prior, work up revealed elevated VZV IgM with normal IgG consistent with acute varicella infection. Physical exam of the infant was notable for generalized numerous erythematous, vesicular lesions in multiple stages of healing. Laboratory workup of CBC, CMP, and CRP were only remarkable for a mildly elevated ALT. Given the young age, and that herpes simplex virus (HSV) was initially considered in the differential, CSF studies were done and were negative. DFA of a vesicle on the patient's thigh was positive for VZV antigen. VZV DNA PCR from skin vesicle was positive. Antibody studies were significant for an elevated IgG level (267.5 mIU/ml) and negative IgM (≤ 0.90). Diagnostics for HSV were negative. He was treated with parenteral acyclovir until plateau in number of lesions was seen on hospital day 4, after which was transitioned to oral therapy to complete a 10 day course.

Despite maternal childhood VZV infection and scars from the illness, elevated seemingly protective serum IgG levels in the infant, a 2-month-old patient presented with clinical VZV infection after post-natal exposure. This report serves to highlight that passively acquired immunity from mother may not always offer total protection in a young infant exposed to varicella. It is thus important for clinicians to counsel varicella immune mothers about avoiding exposure of infant to family members with exanthematous illness. Lastly, the treatment guidelines for VZV infection in this unique age group is not well defined. We successfully used a regimen combining parenteral and oral acyclovir therapy and propose that as an option.

267 THE LUNG THAT CRIED WOLF

Cody Horn*, Unnati Warden, Stuart Shah, Brittany Lyons, K Pramod. Reddy, MD, *University of Florida – Jacksonville, Jacksonville, FL*

10.1136/jim-2018-000974.265

Case report A 61-year-old male with a past medical history of end stage renal disease on hemodialysis, hypertension, diabetes mellitus, recurrent pancreatitis, and left fibrothorax was admitted to the medical intensive care unit for altered mental status and acute respiratory failure. He was intubated and a chest X-ray showed the presence of a left lower lobe pneumonia. A CT abdomen/pelvis displayed nonspecific signs of colitis. The patient was found to have *Salmonella* species bacteremia. Though he was treated with appropriate intravenous antibiotics, he remained persistently febrile with leukocytosis. Urine, sputum and repeat blood cultures had no growth. A CT chest demonstrated a fibrothorax and loculated parapneumonic effusion, which had been present for several years. A bronchoscopy was performed which grew mixed flora. A thoracentesis was performed and 150 cc of brown exudative pleural fluid was removed. Fluid analysis showed 386,000 u/L white blood cells and LDH of 20,000 u/L. Pleural fluid culture grew *Salmonella* species. Cardiothoracic surgery was consulted and the patient underwent a video assisted thoracoscopic surgery with lung decortication. Intraoperative pleural tissue cultures grew *Salmonella* species. The patient was extubated and had no additional postop fevers.

He completed six weeks of antibiotics while inpatient and was discharged in good health.

Salmonella species bacteremia is an uncommon condition with most cases occurring in children. Empyemas and parapneumonic effusions associated with *Salmonella* are even more rare. An online literature search revealed less than fifty published cases with this diagnosis. Our patient had a history of chronic lung disease of a left-sided fibrothorax from recurrent pancreatitis. He had previously undergone several unsuccessful thoracentesis procedures as prior imaging was concerning for empyema. When the patient remained febrile after completing two weeks of appropriate antibiotics but remained febrile with a leukocytosis after subsequent negative cultures, the source remained unknown. Despite years of prior unsuccessful attempts at obtaining pleural fluid, this was ultimately the source of the infection on this admission and the patient was treated appropriately.

268 LEFT VENTRICULAR ASSIST DEVICE INFECTIONS: A NEW ROLE FOR DALBAVANCIN?

J Howard-Anderson*, S Pouch, M Sexton, A Mehta, R Cole, GM Lyon, R Friedman-Moraco. Emory University, Atlanta, GA

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Case report A 59-year-old female with ischemic cardiomyopathy underwent left ventricular assist device (LVAD) placement, which had been complicated by recurrent LVAD-associated infections (LVADIs). She first developed a methicillin-susceptible *Staphylococcus aureus* (MSSA) LVADI 2 years after LVAD placement and was treated with 2 months of cephalexin. Unfortunately, the infection recurred after stopping antibiotics and cephalexin was restarted. Six months later, she was switched to doxycycline out of concern for clinical failure, although cultures persistently grew MSSA. She did well for 1 year before purulent drainage from her driveline exit site resumed. Cultures grew the same MSSA but also revealed a new isolate (resistant to oxacillin, doxycycline, clindamycin; susceptible to vancomycin, rifampin, trimethoprim/sulfamethoxazole). Her blood cultures were negative. Therapy was switched to trimethoprim/sulfamethoxazole; however, she developed a severe adverse reaction necessitating conversion to linezolid. Two weeks later, she was readmitted for an elevated INR without evidence of active infection. Kidney function was at baseline (GFR ~40). Given concerns for toxicities with long-term linezolid, we switched her to dalbavancin 1500 mg IV weekly for chronic suppressive therapy (dose later reduced to 1500 mg every 2 weeks). She has now received greater than 16 weeks of dalbavancin without adverse effects or recurrence.

In this case we illustrate a potential novel role for dalbavancin. As a lipoglycopeptide antibiotic, dalbavancin has activity against many gram-positive organisms including staphylococci. It is FDA-approved for acute bacterial skin and skin structure infections although its use has rarely been reported for LVADIs.¹ Approximately 1/3 of patients with LVADs develop an LVADI within 2 years. Treatment often requires daily administration of IV antibiotics, vascular access device placement with associated infection risk, laboratory monitoring, and home health services. Dalbavancin's unique weekly to biweekly dosing schedule and excellent safety

profile therefore make it an attractive alternative for the treatment of LVADIs.

REFERENCE

1. This could be considered an off-label indication.

269 RARE CASE OF CANDIDA LUNG ABSCESS IN A DIABETIC PATIENT

G Krishnan*, R Parikh, K Bano, M Konda, JC Rico-Crescencio. UAMS, Little Rock, AR

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Case report Candida lung abscess is a rare entity, even rarer is candida lung abscess in a person with no known history of immunocompromise.

A 60-year-old male with diabetes mellitus and coronary artery disease was found down at home and emergently intubated for hypoxic respiratory failure. CT chest showed multiple left lower lobe peripherally enhancing fluid collections, largest being 7.1×5.4×8.3 cm. Vancomycin, cefepime and metronidazole were started for treatment of lung abscesses. Bacterial, fungal and AFB cultures from bronchoscopy and lavage did not grow any organisms. Interventional radiology performed drainage of abscess and cultures grew *Candida albicans*. Patient was initially started on micafungin and later changed to fluconazole based on susceptibilities. He continued to require ventilator support although oxygen requirements came down and was weaned off vasopressors. Repeat CT chest showed decrease in size of abscesses with improvement in consolidative changes. He had open surgical drainage of abscess which grew *Candida albicans* and one colony of *Staphylococcus capitis*. Blood cultures did not grow any organism. He tested negative for HIV and acute hepatitis, urine drug screen was negative, HbA1C was 8.0 and serum cortisol level was within normal limits for sepsis. He does not have a history of solid organ transplant, treatment with long term steroids or chemotherapy. Patient ultimately had tracheostomy and got discharged to complete long course of fluconazole, amoxicillin-clavulanate and doxycycline.

There are handful of published case reports of candida lung abscess in literature. This was a unique case since he developed lung abscesses without any intravenous catheters, critical illnesses or candidemia. His only risk factor was uncontrolled diabetes. Hyperglycemia related impairment in immune system by alteration of neutrophil chemotaxis and opsonization is well established. Diabetics are more prone to buccal and vaginal candidiasis due to glucose inducible proteins in the epithelium. Therapy for candida abscess is guided by clinical response and there is no consensus on the duration of treatment. The preferred initial drug is an echinocandin and this can be changed to an azole based on sensitivities.

270 CAPNOCYTOPHAGA GINIVALIS BACTEREMIA AFTER UPPER GASTROINTESTINAL BLEEDING IN IMMUNOSUPPRESSED PATIENT

^{1,2}FJ Lawal*, ^{1,2}SL Baer. ¹Medical College of Georgia at Augusta University, Augusta, GA;

²Charlie Norwood Veteran Affairs Medical Center, Augusta, GA

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Introduction *Capnocytophaga* is a genus of anaerobic fastidious gram-negative bacilli which is mostly found as part of human,

dog and cat oral flora. *C. gingivalis* is one of 6 identified human colonist species and all are associated with periodontitis, caries and in plaque in adults and in children with malignancies. They are less virulent than the two species are found in animals. *C. gingivalis* is rarely reported as an invasive pathogen in humans, but immunocompromised states predispose to fulminant infections.

Case presentation 70-year-old man who was on methotrexate and prednisone for rheumatoid arthritis and sarcoidosis was being managed for dehydration and COPD exacerbation. He was intubated twice due to hypercapnic respiratory failure. During his second intubation upper gastrointestinal (GI) bleeding was observed with aspiration of blood. He was started on antibiotics and vasopressors after developing a fever of 101.7 Fahrenheit and hypotension. One anaerobic blood culture became positive on 5th day with *Capnocytophaga* species. This was later identified as *Capnocytophaga gingivalis* using 16 s rRNA sequencing. Antibiotics were changed to ampicillin/sulbactam with resolution of hypotension and fever. Upon further questioning, his wife reported she has a dog and he re-chews his snuff after storing it at room temperature.

Discussion We believe this patient developed bacteremia from translocation of oral flora through upper GI bleeding or possible aspiration. Only four cases of *C. gingivalis* have been reported, two of the cases with bacteremia and more severe illness were in immunocompromised patients. Poor oral hygiene in our patient most likely increased the risk in this patient.

Conclusion This case highlights the importance of oral care in immunocompromised patients and the risk of bacterial translocation with GI bleeding.

271

POULTRY RELATED INFECTION: BACTEREMIA WITH CONFOUNDING CAUSALITY

¹FJ Lawal*, ¹A Alexander, ^{1,2}SL Baer, ¹R Colombo. ¹Medical College of Georgia at Augusta University, Augusta, GA; ²Charlie Norwood Veteran Affairs Medical Center, Augusta, GA

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Introduction *Streptococcus gallolyticus* subsp. *gallolyticus* was formerly *Streptococcus bovis* biotype I and is now one of 3 subspecies of *S. gallolyticus*. *S. bovis* biotypes are known to be part of human and animal gut flora but may also be opportunistic pathogens associated with colonic malignancies. Transmission of *S. gallolyticus* subsp *gallolyticus* from colonized or infected animals to humans has been documented and the risk is higher with close contact. Modes of transmission include droplet, contact, or fomites.

Case presentation We present a case of *Streptococcus gallolyticus* bacteremia in a 62-year-old female poultry farmer with a chronic indwelling pacemaker. She suffered a puncture wound from a rooster spike resulting in cellulitis and abscess, treated with cephalexin. Three months later, she developed progressive weakness, thrombocytopenia, fatigue, and high-grade fevers. She was admitted, cultures were obtained, and antibiotics were started. Blood cultures became positive and identified by gram-positive blood culture assay (Verigene nucleic acid assay) as *Streptococcus* spp within 24 hours. She underwent colonoscopy and transthoracic echocardiogram (TEE). She was found on colonoscopy to have a colonic tubulo-villous adenoma with focal high-grade dysplasia. On transthoracic echocardiogram she had a flail mitral leaflet, but no visible vegetations. Bacteria

was then identified by MALDI-TOF as *Streptococcus gallolyticus (bovis)*. She was treated with a 4 week course of IV ceftriaxone for a suspected endovascular infection involving her pacemaker.

Discussion Possible sources of bacteremia with *S. gallolyticus* in this patient include both chronic colonization from long term poultry exposure, or direct inoculation from the rooster spike and resulting abscess and cellulitis. Her tubulo-villous adenoma is also a potential source of infection as *S. gallolyticus* bacteremia has been associated with wide range of bowel disease. The differential diagnosis of infections in poultry farmers should include zoonotic infections such as *S. gallolyticus*. Patients with bacteremia from this organism should be evaluated for colonic abnormalities.

272

MANAGEMENT CHALLENGES OF IMMUNE RECONSTITUTION INFLAMMATORY SYNDROME ASSOCIATED WITH KAPOSI SARCOMA

¹M Lear*, ²H Edriss. ¹Texas Tech University Health Sciences Center, Lubbock, TX; ²Saint Joseph Hospital, Lexington, KY

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Background Kaposi Sarcoma (KS) is a malignant spindle cell tumor derived from endothelial cells caused by human herpesvirus-8 (HHV-8), primarily occurring in patients with low CD4 counts. Initiation of antiretroviral therapy (ART) in HIV patients, especially patients with a low CD4 count, may cause immune reconstitution inflammatory syndrome (IRIS). This is defined as a recovering immune system with an exaggerated inflammatory response and is largely understudied.

Case description A 24-year-old homosexual man with HIV presented with shortness of breath, fever and productive cough for 1 month. Physical exam showed oral thrush with bilateral cervical, axillary and inguinal lymphadenopathy. A 2 cm raised, erythematous lesion in the oral cavity and multiple violaceous skin lesions were noted. Lab testing revealed a CD4 count of 43 cells/mm³ with an HIV viral load of 5 81 000 copies/mL. Computed tomography of the chest showed dense patchy alveolar infiltrates and mediastinal lymphadenopathy. Skin and endobronchial biopsies showed atypical vascular proliferation consistent with nodular KS. Inguinal lymph node dissection revealed KS and Mycobacterium Avium Complex (MAC). Patient received ART, triple therapy for MAC, and radiation therapy for KS. Approximately 8 weeks after starting ART, he developed IRIS with fever of 101.5–102.3 °F, tachycardia, and enlargement of his cutaneous Kaposi lesions. Lab tests showed worsening thrombocytopenia and dysglycemia. During that time, his CD4 count was 78 cells/mm³ and blood cultures were negative for any pathogen. Patient continued on ART. After fever improved a week later, he began vincristine to treat KS and received two doses. On the 36th day of hospitalization, the patient expressed concern to be discharged home to remain closer with family.

Conclusion Kaposi Sarcoma is a common neoplasm in HIV patients with a markedly decreased incidence since the establishment of ART. The occurrence of IRIS in these patients is mostly underrecognized due to the rarity of this disease. It paradoxically worsens KS progression and leads to increased morbidity and mortality. Clinical trials are indicated to identify an effective therapy and improve outcomes.

273 AIDS PRESENTING AS HIGH GRADE DLBCL MASQUERADING AS SCIATICA

W Li*, H Ojeda-Martinez. SUNY Downstate Medical University, Brooklyn, NY

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Case report Our patient was a 46 year-old female with sciatica initially admitted for intractable left hip and lateral thigh pain radiating down to ankle. Described as similar to her usual sciatic pain she had visited an urgent care center who diagnosed her with bursitis and prescribed anti-inflammatory pain medication. After a mechanical fall which exacerbated the pain, she decided to seek evaluation.

Work-up included CT imaging which demonstrated a large infiltrating mass lesion measured $5.8 \times 4.8 \times 4.5$ cm and $3.6 \times 3.7 \times 4.7$ cm involving the piriformis and obturator internus muscles. Subsequently image-guided biopsy with surgical pathology revealed highly malignant DLBCL.

Incidentally a HIV-1 combination antibody test returned positive with absolute CD 4 count of 17/uL and viral load of 1 29 178 copies/mL. The patient described herself as a happily married individual with her husband, having two young children. She denied any promiscuous behavior, extra-marital affairs, previous history of STDs, IVDU, or blood transfusions.

Abstract 273 Figure 1

First described in 1981, HIV/AIDS has grown to become one of the most prolific public health issues of our generation (Mandell). As of 2018, more than 36.7 million individuals worldwide suffer from the virus (AIDS Fact Sheet). While presentation is broad, malignancy is a less often suspected initial presentation. Our patient described complaints not different from her sciatic pain. Despite having a more likely explanation, we must keep high-grade lymphoma in the differential as a presenting finding for HIV/AIDS to limit future morbidity and mortality.

274 OSTEOMYELITIS SECONDARY TO STAPHYLOCOCCUS HAEMOLYTICUS BACTEREMIA

JS Lim*, C Sandlin, RE Begue. Louisiana State University, New Orleans, LA

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Case report *Staphylococcus haemolyticus* is a known cause of coagulase-negative staphylococcal bacteremia, most commonly seen in the elderly and immunocompromised patient populations. Osteomyelitis caused by this pathogen is rare, and case reports are limited. Subsequently, *Staphylococcus haemolyticus* bacteremia causing osteomyelitis in a young, immunocompetent pediatric patient is a rare presentation.

We present a case of a 9-year-old male with a history of recurrent dental infections secondary to poor dental hygiene who presented after a minor car accident. Incidentally, the patient was found to be febrile on presentation and on further history had 2 days of tooth pain and 1 day of left mandibular facial swelling. CT head and sinus revealed chronic osteomyelitis of the left mandibular body in association with a periapical abscess of the left mandibular first molar. Blood culture revealed *Staphylococcus haemolyticus* growing at 0.38 days sensitive to Clindamycin and Vancomycin and resistant to Oxacillin and Penicillin. CBC revealed a slightly elevated WBC but no other abnormalities on differential. ESR and CRP were both elevated. The patient was treated with Clindamycin with improvement in his overall clinical picture and repeat blood culture that was negative. Clindamycin was continued outpatient to complete four weeks of antibiotic treatment for osteomyelitis as well as close follow-up with dentistry and plastic surgery.

This case demonstrates osteomyelitis secondary to *Staphylococcus haemolyticus* bacteremia in an immunocompetent pediatric patient. While *Staphylococcus haemolyticus* bacteremia is documented in the literature, it is often in the elderly and immunocompromised patient populations. This case demonstrates not only *Staphylococcus haemolyticus* bacteremia in a healthy pediatric patient, but it presents a case of osteomyelitis secondary to this pathogen. Given the exceedingly rare nature of this pathogen being associated with osteomyelitis especially in a young, healthy pediatric patient, additional case reports are critical for elucidating the full pathogenic nature of this coagulase-negative staph.

275 THE PERICARDIUM: AN UNUSUAL LOCATION FOR METHICILLIN RESISTANT STAPHYLOCOCCUS AUREUS

J Lung*, JR Rivas, P Staiano. University of Florida, Jacksonville, FL

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Case report A 45 year old Caucasian male with past medical history of cirrhosis, esophageal varices, and diabetes presented to the emergency department with 5 days of chest pain, dyspnea, and fever. Physical exam revealed only tachycardia. Electrocardiogram showed no acute ischemic findings; however, troponin resulted positive and was attributed to demand ischemia in setting of sepsis likely due to pericarditis. Empiric antibiotics were then started. Use of nonsteroidal anti-inflammatory drugs and colchicine was deferred due to acute kidney injury and gastrointestinal bleed requiring transfusions. Computed tomography of the chest noted only small pericardial effusion and findings suggestive of cirrhosis. Bacteremia with methicillin resistant *Staphylococcus aureus* (MRSA) was later discovered. He later required intubation due to respiratory distress, muffled heart sounds, jugular venous distension, development of atrial fibrillation with rapid ventricular response, and septic shock. Echocardiogram revealed invagination of the right side of the heart. Emergent pericardiocentesis

released 700 cc of fluid. Pericardial fluid was cultured and revealed MRSA. Atrial fibrillation subsequently resolved, and need for pressors and mechanical ventilation diminished. Improvement in clinical status was attributed to source control and relief of cardiac tamponade. Patient required a total of 6 weeks of intravenous vancomycin based on sensitivities.

Discussion Pericarditis is the inflammation of the pericardium encompassing the heart. Protracted hospital courses and invasive procedures are associated with MRSA infection of the pericardium. Our case differs as the patient was admitted with pericarditis caused by MRSA. Since community-acquired MRSA is not traditionally associated with sepsis or severe disease, increased cognizance of its virulence and spectrum is warranted. Treatment of MRSA pericarditis and associated tamponade requires drainage of pericardial fluid and prompt antibiotic initiation.

Conclusion MRSA has become more prevalent in healthcare and community acquired infections. MRSA is found in up to 60% of patients in the medical intensive care unit; even so, MRSA pericarditis is scarcely reported. Further research must be done to evaluate the incidence of pericardial infections with MRSA.

276 BIOPSY-PROVEN SCHISTOSOMIASIS IN A REFUGEE FROM SUDAN

R Mangat*, A Eranki. *SUNY Upstate, Syracuse, NY*

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Case report Schistosomiasis is a disease caused by blood flukes, and can affect both intestinal and urogenital systems. Fresh-water becomes infested when snails release cercariae and infection occurs when the larvae of the parasite penetrate the skin. There are 5 species that can cause infections in humans. 91% of individuals who require treatment for the disease live in Africa. People more likely to be infected are those that are exposed to infested waters. Symptoms occur secondary to migration of eggs through tissues. Infected individuals can present with urogenital symptoms such as hematuria or granulomatous inflammation, which can result in urinary tract obstruction. Here, we presented a biopsy proven case of schistosomiasis in a refugee presenting with hematuria who recently moved to Upstate New York.

A 30 year old man with no significant medical history presented with intermittent episodes of gross hematuria along with abdominal pain, dysuria and hematospermia. He recalled that hematuria started at age 15. Patient is a refugee from West Sudan and moved to the US in 2017. He spent 5 years in a refugee camp in Kenya prior to coming to the US. He admits to drinking river water as well as swimming in rivers. He worked in agriculture in Sudan and then as a security guard in Kenya. Urology workup revealed distal ureteral narrowing. Cystoscopy showed multiple small yellow lesions in the bladder. Biopsy showed chronic cystitis associated with calcified microorganisms consistent with schistosomiasis (most likely *Schistosoma hematobium*).

Although most infected individuals do not develop any symptoms, schistosomiasis is an important differential to consider in patients who present from endemic countries, or travelers, as treatment is aimed at preventing both chronic complications and neuroschistosomiasis. Acute infection can be seen in travelers, whereas chronic infections are seen in individuals with ongoing exposure in endemic regions. Lab

findings include eosinophilia, anemia, thrombocytopenia and transaminitis. Diagnosis can be made through the detection of parasite eggs in stool or urine specimens or biopsy showing granulomas surrounding eggs embedded in the mucosa. Treatment with praziquantel has been shown to reduce morbidity and mortality by reducing egg production.

277 DELFTIA ACIDOVORANS BACTEREMIA IN A RELATIVE IMMUNOSUPPRESSED STATE

SS Masson*, V Kohli, K Singh, S Karakattu. *East Tennessee State University, Johnson City, TN*

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Case report Infections with *Delftia acidovorans*, a non-pathogenic environmental gram-negative bacillus rarely of clinical significance, have been reported in immunocompromised states (neutropenia, HIV/AIDS, malignancy, immunosuppressive therapy) and rarely even in immunocompetent ones. Our case is of *D. acidovorans* bacteremia in a patient with no confirmed source of entry, but only relative immunosuppression due to diabetes mellitus and chronic kidney disease. A 75 year old female was admitted to the hospital due to increasing somnolence. The patient had a long list of co-morbidities including PVD, chronic lower extremity lymphedema with recurring cellulitis, type II diabetes mellitus and stage 3 CKD. Lab abnormalities on admission included elevated WBC count (25000/mm³), CRP of 8.93 mg/L and ESR of 93. Suspecting an infection, the patient was started on IV vancomycin and piperacillin-tazobactam. Blood cultures collected prior to antibiotic administration showed growth of *D. acidovorans*, while a heel ulcer culture was positive for *Streptococcus* species with light growth of non-lactose and lactose fermenting gram-negative rods. On normalization of her mentation the next morning, she revealed she had taken a handful of tramadol pills for pain relief from her chronic leg pain few hours prior to hospitalization. Given the impaired barrier skin (from chronic lower extremity lymphedema) and immune defenses due peripheral vascular disease, long-standing diabetes mellitus and Stage III CKD, bacteria of even low virulence may have entered the bloodstream. Although the heel cultures did not directly isolate this organism, it did grow host of non-lactose gram negative bacilli, a category to which this organism belongs. Although cases of this bacterium causing serious infection in immunosuppressed patients exist, rarely do they document infection in setting of relative immunosuppressed states such as diabetes mellitus and chronic kidney disease. Our case highlights such conditions as potential risk factors associated with infection with this relatively non-pathogenic microbe. As a result, providers should also consider infections with non-pathogenic organisms in relative immunosuppressed states that are now becoming more prevalent due to the increasing burden of such chronic diseases.

278 TICKBORNE ILLNESS: HEARTLAND VIRUS AND Q FEVER

S McClelland*, M Burns. *University of Arkansas for Medical Sciences, Little Rock, AR*

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Case report 70-year-old Caucasian man with no medical history, who lived on a farm with multiple animals, presented

from an outside hospital (OSH) for further workup of tick-borne illness. Patient developed night sweats, fevers, decreased oral intake, and low back pain one month after hiking a local trail, and removing a tick from his neck. Patient was seen by his PCP two days after symptoms started and labs were consistent with a tick-borne disease. Tick serologies were obtained and patient was started on doxycycline. Patient's symptoms progressed to worsening fatigue, new onset balance difficulties, difficulty writing, blurry vision, and a new tremor in hands and jaw despite doxycycline. He was admitted to OSH and doxycycline was continued. MRI showed minimal periventricular white matter disease, mild ethmoid sinus disease, hypertrophy of right mucosal turbinate, and small left mastoid effusion. Labs at OSH showed WBC:0.15, platelets:46, Na:129, CPK:1526, and transaminitis. LP showed glucose: 62, protein: 69, and WBC: 2. Patient had progression of symptoms despite doxycycline and one dose of Rocephin and had new ascending lower extremity neuropathy. Due to progression of symptoms, heartland virus was suspected and labs were obtained and sent to the CDC and the Arkansas Health Department. Patient was switched to IV doxycycline and transferred to UAMS. While at UAMS, doxycycline was continued and infectious workup was negative, except for prior infection of CMV and EBV. CT chest/abdomen/pelvis significant for a 1.8 cm right inguinal lymph node. Liver enzymes initially elevated on admission but improved. Heartland virus was positive. After discharge from UAMS, patient's CoxiellaBurnettiIgG phase II serology titers was 1:128 (positive). Patient was treated for heartland virus with supportive care and Q fever with two additional weeks of PO doxycycline and close ID clinic follow up.

279 SUDDEN HEARING LOSS! THINK MENINGITIS

A Mohammedzein*, M Malik, K Shepherd. *Texas Tech University Health science center, Amarillo, TX*

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Recognize that acute hearing loss as a fairly common symptoms of meningitis.

Case presentation An 84-year-old woman presents with two days of sudden near complete sensorineural hearing loss. The day of admission she also developed confusion and fever. No reported headache, seizure, weakness, or other neurologic symptoms.

Exam revealed temperature of 39.5°C, heart rate 129 bpm, respiration 37/min, blood pressure 161/79 mmHg. She was only oriented to person. She had notable neck stiffness. We were unable to perform proper neurological exam due to patient's difficulty following commands, but it was grossly unremarkable.

Labs remarkable for WBC 13.8 with 80% neutrophils with remainder of labs within normal limits. Due to high suspicion for meningitis lumbar puncture was performed, with results of WBC 2367 cell/mcl, neutrophils 95%, RBC 106, glucose 4 mg/dl, total protein 627 mg/dl.

She was started on vancomycin, meropenem (penicillin allergy), and dexamethasone. CSF and blood cultures grew *Streptococcus pneumoniae*, accordingly she was switched to ceftriaxone. The source of infection was most likely sinusitis as evident on head CT. She was afebrile within 24 hours and her mentation improved. She was discharged to rehabilitation

facility to continue antibiotics for a total of 10–14 days. Hearing improved significantly by the time of discharge.

Discussion Hearing loss occurs in up to 14% of bacterial meningitis, with a higher rate in pneumococcal meningitis. It's mostly permanent, but improvement does occur in rare cases. Hearing loss can easily be missed as one study demonstrated nearly 40% of patients were found to have hearing impairment after discharge who weren't noted to have it before. Transient hearing loss is usually secondary to a conductive disturbance, whereas permanent hearing loss can result from damage to the eighth cranial nerve, cochlea, or labyrinth induced by direct bacterial invasion and/or the inflammatory response elicited by the infection.

Dexamethasone therapy may reduce the rate of neurologic sequelae, particularly in selected patients with pneumococcal meningitis of intermediate severity. Early recognition, administration of intravenous antibiotic, and glucocorticoids remain the most critical steps in reducing mortality and neurological sequelae.

280 SHEWANELLA ALGAE AS AN UNUSUAL CAUSE OF SEPTIC SHOCK: AN EMERGENT PATHOGEN

M Morales Hernandez*, C Canha, JM Hernandez, W Kogler, P Reddy. *University of Florida College of Medicine, Jacksonville, FL*

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Case report A 94 year-old female presented with acute encephalopathy requiring admission to the intensive care unit. She was found to have hyperglycemic hyperosmolar coma and septic shock. She had history of type 2 diabetes mellitus, stage 3 chronic kidney disease and chronic venous stasis with lymphadenitis. Patient required vasopressor support, continuous insulin infusion and was started on vancomycin and piperacillin-tazobactam. Urine cultures grew mixed flora, but blood cultures yielded Gram negative organism consistent with *Shewanella algae*. She denied recent travel, sick contacts, exposure to water sources, or seafood consumption. Patient's mentation returned to baseline and vancomycin was stopped. She was maintained on piperacillin-tazobactam with excellent clinical response. Repeat blood cultures remained negative, and was discharged to a nursing facility.

Shewanella species are saprophytic Gram negative rods found in marine habitats. Thus far, *S. putrefaciens* and *S. algae* are known to infect humans. These are emergent pathogens and pathogenesis has been correlated with hemolysin production. They are most commonly encountered in the immunocompromised host with soft tissue infections or with exposure to marine environments. Risk factors include age, peripheral vascular diseases, diabetes, renal failure and hepatobiliary disease. *S. algae* is an unusual pathogenic bacterium in humans often confused with *V. vulnificus*. We hypothesized infection was transmitted from breakdowns in the skin in the lower extremities due her chronic lymphadenitis into the circulation. To our knowledge, this is the fifth case of *Shewanella* species reported in the United States. Typically, *Shewanella* is susceptible to aminoglycosides, quinolones, extended spectrum cephalosporins, β -lactamase inhibitor combinations, and carbapenems. Our patient was successfully managed with piperacillin-tazobactam for 14 days.

Shewanella species should be considered an emergent pathogen in the United States. A detailed medical and social history

are warranted for prompt diagnosis. Due known history of emerging drug resistant to quinolones or β -lactams, empiric antibiotherapy should be started early and tailored based on sensitivities.

281 EMERGING LACTOCOCCUS GARVIEAE INFECTION IN LUNG ADENOCARCINOMA: A CASE REPORT

C Ogbonna*, M Khan, M Land. *University of Tennessee Health Sciences Center, Memphis, TN*

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Purpose of study *Lactococcus Garvieae* is a gram-positive, catalase negative, anaerobic cocci that may appear in chains, pairs or single cocci. It has been identified in cow's milk and cheese. We report a case of a man presenting with an epidural abscess associated with *Lactococcus Garvieae* bacteremia.

Summary of results 55 year old male presented with complaints of acute lower back pain with no precipitating events. Physical exam was significant for low-grade fever and tachycardia. Imaging of the lumbar spine revealed L3-L5 epidural abscess with L5-S1 discitis. Thoracic imaging demonstrated a right upper lobe 4.2×3.2 cm mass compared to a 1.7×0.9 cm nodule at the initial time of detection one year prior.

Overnight blood cultures became positive for gram-positive cocci in pairs initially identified as enterococci; but with matrix-assisted desorption/ionization time of flight mass spectrometry [MALDI-TOF MS], *Lactococcus Garvieae* was confirmed as the true pathogen. Other baseline laboratory studies were unremarkable. He was initially begun on Vancomycin at the time of admission, and converted to ceftriaxone plus ampicillin and sulbactam after cultures resulted. Repeat blood cultures 4 days after admission were sterile. A transthoracic echocardiogram was negative for vegetations. CT guided biopsy of the lung mass revealed adenocarcinoma. He was then scheduled for outpatient colonoscopy, and discharged from the hospital with 6 weeks of intravenous antibiotics due to associated spinal epidural abscess.

Conclusion *Lactococcus* may be misidentified as enterococcus as in our case. From literature review, many patients present with endocarditis, spondylitis, hepatobiliary disease, peritonitis and urinary tract infection. An association with the consumption of raw fish or unpasteurized dairy products is well-known. There have been case reports of association with colorectal carcinoma and cholangiocarcinoma.

Our case is unique because our patient had no contact with raw fish, and no intake of unpasteurized milk products. Additionally this is the only case per our literature review of bacteremia with epidural abscess identified in a patient with lung adenocarcinoma. Further studies are warranted to establish an association of *Lactococcus Garvieae* with underlying carcinomas in general.

282 A RARE CASE OF DISSEMINATED MYCOBACTERIUM CHELONAE INFECTION IN AN IMMUNOCOMPETENT ADULT

¹R Parikh*, ¹G Krishnan, ²M Vuong-Dac, ¹L Achanta. ¹UAMS, Little Rock, AR; ²UAMS, Little Rock, AR

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Case report *Mycobacterium chelonae* is a rapidly growing non-tuberculous mycobacterium (NTM) found in water and soil, which usually causes skin and soft tissue infections. It can cause invasive and disseminated disease in immunosuppressed patients. We present a unique case of disseminated disease in an immunocompetent patient, a diagnostic dilemma.

An 83 year old man with ischemic cardiomyopathy and prostate cancer in remission was transferred to our facility for evaluation of multiple, painless, non-pruritic nodules, draining sanguineous fluid, on his extremities and pustules on his cheeks for the past 3 months. He was not exposed to any aquatic animals and denied any preceding trauma to the skin or contamination of skin with soil. He was previously treated with oral antibiotic and empiric steroid courses with minimal improvement and recurrence. On examination, multiple large pustules and erythematous nodules were seen on his cheeks and extremities. Biopsy of the lesions revealed gram-positive, acid fast bacilli in the dermis. *Mycobacterium chelonae* DNA was detected by polymerase chain reaction (PCR) of his skin biopsy sample. Blood cultures also grew *M. chelonae*. He was started on imipenem, clarithromycin and amikacin. His rash had improved upon discharge but he unfortunately passed away suddenly before his follow up clinic appointment in two weeks.

The prevalence of *M. chelonae* and other NTM infections is increasing in southern United States. There are several case reports of *M. Chelonae* causing catheter related bacteremia, traumatic and surgical wound infections (after corneal implants, sclerotherapy or tattooing). There is considerable variation in their geographical distribution and a high degree of suspicion is needed to diagnose them earlier and improve patient outcomes. This case was diagnostically challenging as the patient had no predisposing factors. Failure to respond to empiric treatment should prompt an earlier skin biopsy and culture. Molecular techniques like PCR are needed to identify different NTM species to guide therapy as in our case. Dual therapy with macrolides and aminoglycosides for four to six months is recommended for disseminated disease along with surgical debridement or catheter removal when necessary.

283 SEPTIC SHOCK SECONDARY TO SHIGELLA FLEXNERI BACTEREMIA: A RARE PRESENTATION IN THE CARIBBEAN

P Pastor-Perez*, M Colon. *San Juan City Hospital, Bayamon*

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Case report *Shigella flexneri* bacteremia is a very rare complication with less than 8% of cases documented worldwide of which majority are seen in severely malnourished children and immunocompromised adults. Outbreaks of infection have been rarely documented in travelers who visited developing countries such as India, Haiti and Dominican Republic. Severe malnutrition, dysentery, fever and leukocytosis are among the most common symptoms. Will present a unique case of *S. flexneri* in an immunocompetent non-traveler male native from the Virgin Islands complaining of non-bloody diarrhea.

A 79 year old male who was brought to urgency room after being found on the street disoriented and dehydrated referring episodes of non-bloody diarrhea since one week ago. Patient denied any constitutional symptoms. He is native from the Virgin Islands but recently moved to Puerto Rico. Initial

laboratories were remarkable for leukocytosis, hypernatremia, kidney injury and transaminitis; Human Immunodeficiency virus test was negative. Physical examination remarkable for dry oral mucosa and temporal wasting. Stool workup requested was negative. Following a decrease in leukocytosis, an abrupt elevation was reported. Subsequently patients' clinical status deteriorated, he was hypoactive and anorexic. Blood culture was positive for *Shigella flexneri* (Ampicillin-resistant). Even with antibiotic optimization patient developed septic shock requiring vasopressors and mechanical ventilation. Despite appropriate medical management patient passed away.

Few cases of *Shigella flexneri* bacteremia have been reported, however, due to its susceptible high level of mortality it should be studied further. In this case, even though patient is immunocompetent, is evident the significant clinical deterioration developed regardless of appropriate medical management. In majority of cases of *S. flexneri* bacteremia fecal cultures are reported negative. Extra-intestinal manifestations are rarely seen in *S. flexneri* infection unless patients are children or immunocompromised leading to a more complicated presentation such as septic shock. In this unique case patient presents no such risk factors. Therefore, adequate workup, including fecal and blood cultures should be considered in every invasive diarrhea case.

284 PROGRESSIVE VISION LOSS DUE TO OCULAR SYPHILIS

H Patel*, R Bahloul, E Sottlie. *UF COM Jacksonville, Jacksonville, FL*

10.1136/jim-2018-000974.282

Case report A 53 year old Caucasian male with a medical history significant for human immunodeficiency virus (HIV) presented to the emergency department due to worsening left eye pain and vision loss. His symptoms initially began with blurry vision in his left eye. He had associated erythema, photophobia, and pain with eye movement. A few weeks later began having worsening of his vision to the point of complete loss of vision. He also started developing symptoms in his right eye. Ophthalmological evaluation revealed bilateral anterior uveitis with posterior involvement. As patient was not currently on anti-retroviral therapy for his HIV his differential was broad. CT scan of head was negative for any significant findings. A lumbar puncture revealed low glucose, elevated protein, normal opening pressure, and elevated white blood cells. Extensive work-up for opportunistic infections (including syphilis, toxoplasmosis, tuberculosis, Lyme disease, and herpes-zoster) returned largely negative except for RPR titer of 1:512, reactive CSF VDRL (venereal disease research laboratory) titer of 1:8, and a confirmatory TP-PA (treponema pallidum passive particle agglutination assay). He was treated with intravenous penicillin G daily for 2 weeks.

Neurosyphilis involves infection of the central nervous system by the spirochete *treponema pallidum*. Ocular syphilis is a rarer subset of neurosyphilis. It can involve practically any structure in the eye but most commonly presents as posterior uveitis or pan-uveitis. Despite having an effective therapy with penicillin, the incidence over the last couple decades has been increasing likely due to the increase in HIV infected individuals and men who have sex with men. There are no specific trials or data for the treatment of ocular syphilis therefore it is treated in the same manner as neurosyphilis with 18–24 million units of IV penicillin G daily for 10–14 days.

Therapy doesn't lead to complete resolution of the symptoms but will stop progression of the visual changes which is why early diagnosis and treatment are so vital. This diagnosis should always be kept in mind in patients with uveitis especially in the HIV population.

285 A RARE CASE OF TEMPORALIS MUSCLE ABSCESSES SECONDARY TO MAYFIELD CLAMP PIN SITES IN A POST-OPERATIVE NEUROSURGERY PATIENT

^{1,2}NB Peeples*, ^{1,2}JJ Xie, ²T Bartter. ¹UAMS, Little Rock, AR; ²CAVHS, Little Rock, AR

10.1136/jim-2018-000974.283

Case report The Mayfield skull clamp is a three-prong positioning system commonly utilized in neurosurgery to stabilize the head. Literature discussing the complications from these clamps is exceptionally rare. The few complications that are documented are quite severe, such as skull fracture, epidural hematoma, and air embolus, with the caveat that many lesser injuries likely go unnoticed or unreported. Here we describe a case of bilateral temporalis muscle abscesses occurring at the Mayfield head clamp placement sites.

A 50 year old male status post C5-C7 laminectomy and intradural mass resection presented two weeks after his surgery with dysuria and fever. Initial work-up showed cystitis, and the patient was started on appropriate antibiotic therapy. Despite three days of treatment, he remained febrile. His only other complaint was jaw pain. His differential diagnosis included cervical spine surgical site infection versus perinephric abscess secondary to cystitis; however, these were ruled out based on imaging. The patient remained febrile with bilateral jaw pain, so magnetic resonance imaging (MRI) of the brain was ordered which revealed bilateral temporalis muscle fluid collections with abnormal enhancement and central fluid collections, consistent with abscesses. The locations of his abscesses were consistent with the placement of the head clamps.

Had this patient not been able to guide our diagnostic approach with his history, these abscesses would have likely gone undiagnosed and inadequately treated. To our knowledge, there is no pre-existing literature on abscesses at the surgical sites of Mayfield head clamps, making this the first documented case. Abscess from head-securing surgical clamps is a rare complication, but should remain in the differential of a febrile post-op neurosurgery patient. This complication will require a more aggressive antibiotic course and/or drainage to resolve, and could easily go overlooked in a patient with other potential sources of infection or a patient who is unable to localize symptoms.

286 A CASE OF PNEUMOCYSTIS JIROVECI PNEUMONIA ASSOCIATED WITH IDELALISIB USE

H Rafeek*, S Ali. *TTUHSC, Odessa, TX*

10.1136/jim-2018-000974.284

Case report A 74 y old male presented with 2 week history of fever, productive cough with shortness of breath worse with exertion. Past medical history was significant for chronic lymphocytic leukemia since 10 years for which he had been on treatment with a monoclonal antibody called idelalisib. He did not have significant travel history, no recent hospitalization

and no risk factors for tuberculosis. He had stopped taking Bactrim prophylaxis about 3 months prior. Physical exam showed coarse breath sounds. Laboratory results showed increased arterial alveolar gradient, elevated lactate dehydrogenase and a positive sputum polymerase chain reaction for pneumocystis jiroveci. HIV 4th generation test was neg. Ct chest showed bilateral ground glass opacities. A diagnosis of pneumocystis jiroveci pneumonia was made. Patient was treated with bactrim and prednisone with complete resolution of symptoms.

Conclusion This case highlights the importance of recognizing the specific infections associated with some chemotherapeutic agents.

Idelalisib use is associated with increased risk for pneumocystis jiroveci pneumonia. It is important to place on bactrim prophylaxis while using this monoclonal antibody.

287 STERNOCLEIDOMASTOID PYOMYOSITIS: AN ATYPICAL PRESENTATION OF AN UNCOMMON INFECTION

¹JG Rodriguez Velez*, ²C Frau Nicole. ¹San Juan City Hospital, San Juan; ²UPR Carolina, Carolina

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Introduction Pyomyositis is an infection of skeletal muscle most frequently presenting in the extremities and usually caused by hematogenous spread of *Staphylococcus aureus*. Predisposing factors include immunocompromise, trauma, intravenous drug abuse (IVDA), and malnutrition. Among high risk population, IVDA patients are often hospitalized due to skin and soft tissue infection (SSTI) secondary to direct bacteria inoculation into skin or muscle which may lead to development of pyomyositis. It is a complication of a SSTI which often requires surgical intervention. This report describes a case of sternocleidomastoid (SCM) pyomyositis secondary to direct inoculation in an IVDA patient.

Case description A 40 y/o male with history of IVDA presented to the urgency clinic complaining of right sided painful neck mass which worsened in the last week. Patient referred having constant, sharp pain, 7/10 in intensity, which radiated to the chest and aggravated with movement. He also referred subjective fever, chills and odynophagia. He attributed symptoms to recent drug injection near the affected site.

Physical examination was remarkable for a 3×3 cm non-mobile, non-pulsatile, erythematous, tender, warm area of induration. Neck CT scan reported an abscess at the right SCM muscle with displacement of adjacent structure. ENT services performed incision and drainage. Culture of purulent fluid revealed MRSA. The patient completed 11 days of Vancomycin with clinical improvement. Follow up CT scan was negative for abscess. The patient was discharged to complete 15 days of Doxycycline.

Discussion Pyomyositis rarely occurs in neck muscles, for this reasons it may be overlooked as a differential diagnosis when evaluating neck masses. It is important to take note of risk factors and maintain a low threshold of suspicion when evaluating at risk populations with neck masses. Diagnosis and treatment of pyomyositis includes imaging, antibiotics and surgical intervention, without which response to pharmacologic treatment alone may be limited. Given the sensitivity and proximity of vital structures in the neck, early detection and intervention could prevent progression and complications such as compression of airway, nervous or vascular structures.

288 THE URINE COLOR IS WHAT? A CASE OF PURPLE URINE BAG SYNDROME

J Ruiz*, AE Martinez, M Ganji. UF Health Jacksonville, Jacksonville, FL

10.1136/jim-2018-000974.286

Case report 63 year-old male with no past medical history who presented to the emergency department with 6 month history of progressive fatigue and weakness. Associated with dysuria and decreased urine output with some difficulty voiding. Patient experienced a fall earlier in the day and hit his forehead on the pavement and was found down and rescue was called. Labs on presentation showed a potassium of 6.2, urea of 190, bicarbonate of 7 and creatinine of 13.91. Electrocardiogram showed peak T waves but no ST changes or elevations. Bedside renal ultrasound performed showed severe bilateral hydronephrosis with significant sedimentation present. Foley was inserted and purple urine was found draining in the foley bag. Urinalysis showed WBC>180, positive leukocytes and +4 bacteria. Urine culture grew >1 00 000 colonies/mL of *Escheria coli*. Patient was started on ceftriaxone and later switch ciprofloxacin for urinary tract infection based on sensitivities from urine culture. Nephrology and urology was consulted given obstructive uropathy which patient required replacement of foley and hemodialysis for a couple of days. Patient was discharge in stable condition with follow up with urology for voiding trial.

Discussion Purple Urine Bag Syndrome (PUBS) is a rare seen but real manifestation of a more common pathology, urinary tract infections. Usually seen in women, chronically debilitated patients, use of plastic catheter and bag, and also associated with patients suffering from chronic constipation. Patients who are infected or colonized by high bacterial loads are at an increased susceptibility for this phenomenon. The metabolites of the amino acid tryptophan by the urinary bacteria are what most authors believe to be the mechanism of the purple urine. There are several species associated with PUBS, which are usually primarily associated with urinary tract infections. Most common organism are *Klebsiella pneumoniae*, *Escherichia coli*, *Enterococcus* species, and *Pseudomonas aeruginosa*. As interesting and shocking as purple urine may seem to be, this is a relatively benign process aside from the fact there is colonization by bacteria. Treatment of PUBS is typically the same as that of the treatment of urinary tract infection.

289 EPIGLOTTITIS AND TONSILLAR NECROSIS: AN UNUSUAL PRESENTATION OF DISSEMINATED HISTOPLASMOSIS

N Sawal*. SUNY Upstate, Syracuse, NY

10.1136/jim-2018-000974.287

Case report Histoplasmosis is the most prevalent endemic mycosis in the United States. While most infections are either asymptomatic or produce a self-limiting respiratory illness, dissemination can also be seen. Extra-pulmonary disease can spread to many organs, however tonsillar involvement is rare – with only three documented cases. We present the diagnostic dilemma of an immunocompetent 64 year-old Caucasian male who presented with a unilateral tonsillar ulceration that was histologically proven to be Histoplasmosis.

A 64 year-old Caucasian male with a 40 year history of tobacco abuse presented with a two-month history of non-productive cough progressing to odynophagia and hoarseness of

voice. Vitals were normal and labs were significant for a slight leukocytosis and a marginally elevated CRP. Both mononucleosis and rapid strep test were negative. CT imaging revealed an enlarged epiglottis extending into the vallecular space with effacement of the pre-epiglottic fat, and a non-specific hypodensity within the C3 vertebrae concerning for metastatic disease. Flexible nasopharyngo-laryngoscopy demonstrated an enlarged epiglottis and biopsies were obtained of the ulcerated right tonsillar surface. The patient had minimal improvement with antibiotics and steroids, and GMS stain of surgical pathology showed numerous small, oval fungal yeasts within the ulcer most consistent with Histoplasma. He had complete resolution of his symptoms upon completing a 12 week course of oral Itraconazole.

Due to the unusual location and confounding medical history, our case provided a diagnostic dilemma. Characterized by an area of extra-pulmonary focus, disseminated histoplasmosis occurs only in about one in 2000 patients with acute infection. Although Histoplasmosis of the oral cavity is fairly common, tonsillar lesions are rare and often mistaken for malignancy. In our case, diagnosis was delayed because the oral lesions were attributed to alternative etiologies including bacterial/viral infections and malignancy. This case highlights that tonsils are a potential site of Histoplasmosis and this diagnosis should be considered even in immunocompetent hosts. Although histological examination of the tonsil confirmed the diagnosis, the highly specific and sensitive H. capsulatum PCR may aid in diagnosis and should be considered in order to avoid invasive procedures.

290

A CASE OF CHRONIC INVASIVE FUNGAL RHINOSINUSITIS PRESENTING WITH CHRONIC HEADACHES IN IMMUNOCOMPETENT PATIENT

I Serrano*, P Shelley, H Patel, F Warda. *University of Florida, Jacksonville, FL*

10.1136/jim-2018-000974.288

Case report A 22-year-old African American male with a history of seasonal allergies and nasal polyps presented with bilateral eye pain and eyelid swelling of 3 day duration. Associated symptoms included rhinorrhea and daily headaches for 10 months. Severe bilateral proptosis was noted on physical exam. On ophthalmologic exam, his vision was intact but he experienced severe pain with extraocular movements. He had hyponasal speech, abnormal widening of the nose and his nares were mostly obliterated by mucous, tissue and a small dry blood clot. Laboratory tests were unremarkable and HIV test was negative. An orbital CT-Scan revealed an extensive mixed solid and cystic soft tissue mass occupying the paranasal sinuses with dehiscence of the bilateral superomedial orbital walls. MRI-brain was suggestive of fungal rhinosinusitis with mucocele formation and extension into the left frontal lobe compressing the corpus callosum. He was admitted to the Neurological Intensive Care Unit for a Functional Endoscopic Sinus Surgery (FESS) with submucosal resection of nasal septum and nasal polypectomy. Empiric antibiotic coverage with Vancomycin, Zosyn and IV Liposomal Amphotericin B was started. Final pathology results of biopsy revealed eosinophilia and invasive fungal hyphae. Cultures grew *Curvularia* species (mold), *Pseudomonas aeruginosa* and *Staphylococcus aureus* (MSSA). After 2 weeks of IV antibiotics, he was discharged on oral Trimethoprim-sulfamethoxazole and

Levofloxacin daily to complete a 6 week course and Posaconazole for 6 months. Repeat MRI-brain after 2 months revealed improved opacification of left frontal sinus with minimal mass effect on the left orbital apex.

Discussion Patients with anatomic abnormalities of the paranasal sinuses that impair drainage, such as nasal polyps, are vulnerable to fungal colonization. Although colonization of the upper and lower airways is a common asymptomatic condition, aggressive fungal infections known as invasive fungal rhinosinusitis can develop with the majority of cases involving immunosuppressed patients. Diagnosis is dependent upon histopathologic demonstration of fungal invasion by biopsy and empiric treatment with IV amphotericin B should be started promptly, as overall survival is poor in severe cases.

291

PROLONGED ANTIBIOTIC USE IN PRETERM INFANTS WITH NEGATIVE BLOOD CULTURE

W Sourour*, B Niebuhr, J Burdine, V Sanchez, S Jain. *University of Texas Medical Branch, Galveston, TX*

10.1136/jim-2018-000974.289

Purpose of study Neonatal sepsis in premature infants can present with non-specific clinical presentations. This leads to prolonged (>48 hours) antibiotic use in preterm infants. The purpose of our study was to evaluate the role of prolonged antibiotic use in preterm infants with blood culture negative suspected early onset sepsis in late onset sepsis and hospital stay.

Methods used This is an observational retrospective study from January 2017 to December 2017 (12 months) of very low birth weight infants (<1250 grams) who were treated with antibiotics in first week of life. We included infants <30 weeks gestation and <1250 grams birth weight who received antibiotics for early onset sepsis and survived at least 7 days. All infants with known congenital or chromosomal anomalies were excluded from the study. Detailed prenatal and neonatal data was collected from the electronic medical records at UTMB, Galveston. All subjects were divided into two groups. Prolonged antibiotics group who received antibiotics for first >72 hours for blood culture negative suspected sepsis. Control group included infants who received antibiotics <72 hours for blood culture negative suspected sepsis. We used chi square test and p value <0.05 was considered statistically significant.

Summary of results 81 eligible subjects were included in the study. Of these subjects 50 of them met our inclusion criteria for the study. There were 24 female and 26 male infants with mean gestational age 26.5+1.9 weeks and birth weight 865 +207 grams. 33 subjects received prolonged antibiotics and 17 received antibiotics <72 hours. There was significantly prolonged length of hospital stay in prolonged antibiotic group (106+81 versus 65+30 days p<0.05). There was no significant difference in late onset sepsis, NEC, ROP, IVH or pneumonia.

Conclusions Prolonged antibiotic use in preterm infants with blood culture negative suspected sepsis in preterm infants extends the length of hospital stay. We should be cautious in using prolonged antibiotics in preterm infants with negative blood culture. We are including more infants to see if by increasing sample size we may detect any effect of prolonged antibiotics use on other variables like late onset sepsis, NEC, etc.

292

A SIDE EFFECT OF CEFTAROLINE IN MRSA MENINGITIS TREATMENT

J Verdecia*, JM Hernandez, C Izzo, E Sottile, C Isache. *University of Florida College of Medicine, Jacksonville, FL*

10.1136/jim-2018-000974.290

Case report A 61-year-old Caucasian woman presented to the emergency room with complaints of left-sided chest pain and altered mentation for 3 days. Her medical history included liver cirrhosis and coronary artery disease. Initial chest x-ray demonstrated a left pleural effusion and thoracentesis was performed. The patient required supportive treatment including vasopressor drugs and continuous renal replacement therapy. Cultures from the pleural fluid and blood yielded MRSA. Antimicrobial therapy with vancomycin was started. A lumbar puncture was performed showing turbid cerebrospinal fluid and cultures grew MRSA. Given this finding and the persistence of bacteremia, ceftaroline was added for double MRSA coverage. After initiation of ceftaroline, bacteremia cleared and mental status improved, however, patient experienced an acute drop in hemoglobin from 8.7 to 4.9 g/dL. A direct Coombs test confirmed the acute development of warm autoimmune hemolytic anemia and ceftaroline was stopped. Vancomycin was continued. Hemolysis was controlled with prednisone and blood products. The patient improved significantly, with mental status returning to baseline, but eventually opted for hospice care and discontinuation of hemodialysis. She was switched to oral linezolid in order to complete eight weeks of therapy.

Staphylococcal meningitis is a rare occurrence; found in 1%–10% of bacterial meningitis cases. It can occur in patients with neurosurgical interventions or spontaneous. Most common clinical features include fever, altered mentation, headache, seizures, and septic shock. Ceftaroline is generally tolerated but about 3% of patients experience side effects. A smaller percentage have serious adverse effects that may necessitate discontinuation of the drug.

Ceftaroline seems to be a suitable option for disseminated MRSA infection when the clinical response to vancomycin is inadequate. Caution should be taken when using ceftaroline either at off-label doses or for longer than 2 weeks. Most common adverse effects are hematological and severe dermatological reactions. MRSA meningitis is rare but potentially deadly if not treated promptly. Further studies are warranted in order to establish adequate dosing while avoiding potentially deadly adverse effects of ceftaroline in MRSA meningitis.

293

SCROFULA: AN UNUSUAL MANIFESTATION OF MYCOBACTERIUM TUBERCULOSIS

NI Vergne*. *San Juan City Hospital, Puerto Rico, San Juan*

10.1136/jim-2018-000974.291

Case report Tuberculous Cervical Lymphadenitis also known as Scrofula, is a unique manifestation of tuberculosis infection due to *Mycobacterium Tuberculosis* complex. This unexpected particular presentation of extrapulmonary TB is presented as a chronic, specific granulomatous lymphadenitis involving the cervical region with associated caseous necrosis. It accounts for approximately 6.5% of TB cases in the United States and is prominent in the HIV population. Commonly is manifested

as deep cervical nodes and as an isolated chronic nontender cervical lymphadenopathy, without systemic symptoms other than fever in young adults patients. Diagnostic and therapeutic challenges constitute management frustration as it mimics other pathological processes and yields inconsistent clinical and laboratory findings.

This is a case of 47 y/o Hispanic female with PMHx of hypothyroidism, who presented to our institution complaining of neck discomfort associated with multiple cervical masses of 7 month duration. Patient reported progressive worsening of symptoms, especially in the right cervical area, despite previous antibiotic management. Upon interview, patient denied B symptoms, recent infection, contact with sick persons or any recent travel, but state a voluntary missionary in Cuba in 1993. Physical examination revealed remarkable isolated bilateral cervical and subclavicular lymphadenopathy prominent in the right side area. Laboratory data reported no evidence of ongoing infection nor malignancy. HIV test negative and sputum collection negative, PPD positive, and CXR without pathological findings. Neck/chest CT scan disclose bilateral cervical and mediastinal lymphadenopathy without pulmonary cavitary lesions nor pathological findings. After surgical evaluation, FNA biopsy of the most prominent right cervical lymph node, identified the presence of a granulomatous disease, requiring excisional biopsy for definitive diagnosis. Excisional biopsy confirmed caseating granuloma with positive acid fast bacilli smear. Cervical Tuberculous Lymphadenitis was diagnosed and TB management was initiated. After four weeks of TB drug therapy, cervical lymphadenopathy showed significant improvement with complete resolution of patient's neck discomfort and at the moment patient is undergoing treatment with satisfactory evolution.

294

CLOSTRIDIUM SEPTICUM BACTEREMIA PRESENTING AS A PARA-NEOPLASTIC SEPSIS

¹Y Vorakunthada*, ²W Lilitwat. ¹Texas Tech University Health Sciences Center Lubbock, Lubbock, TX; ²University of Iowa Hospitals and Clinics, Iowa, IA

10.1136/jim-2018-000974.292

Case report A 58-year-old male with metastatic adenocarcinoma of the colon and the pancreas who presented with fever, abdominal pain, diarrhea, and fatigue. He presented with signs of severe sepsis on admission. His labs revealed leukopenia (white cell count was 3,760/uL with an absolute neutrophil count of 2,100/uL), elevated pro-calcitonin (13.11 ng/ml), and elevated lactate (4 mg/dL). The abdominal radiograph was unremarkable. Stool *Clostridium difficile* toxin PCR was positive. However, interestingly, his blood cultures grew *Clostridium septicum*. He was admitted to the MICU and treated empirically with vancomycin and metronidazole. His hemodynamic status improved with fluid resuscitation. Repeat blood cultures were negative, and he was discharged with oral metronidazole. Unfortunately, the patient has moved to California, and we could follow his disease course.

Clostridium septicum is a gram-positive, spore-forming, mobile, obligate anaerobic bacillus. Unlike other *Clostridium* species, *Clostridium septicum* does not appear to be a normal gut flora, but it can be found in soil and animals. It can be differentiated from the more common counterpart, *Clostridium perfringens*, by microscopic examination of subterminal spores.

Factors that promote the growth and proliferation include tissue hypoxia from anaerobic glycolysis and outgrowth of blood supply by the proliferating tumor cells. Additionally, the production of various enzymes, including hyaluronidase, hemolysins, deoxyribonuclease, and fibrinolysin allow evasion of the host's immune response. In our patient, a combination of tumor necrosis and gastrointestinal mucosal damage from chemotherapy agents would account for this infection.

Clostridial septicum bacteremia should always be suspected in cancer patients who present with fever, malaise, and localized pain. On the other hand, previously healthy patients with Clostridium septicum bacteremia should also prompt physicians to perform workups for gastrointestinal, genitourinary, and hematologic malignancy. Rapid diagnostic tests for Clostridial infection is not available, and a high index of clinical suspicion is necessary for early diagnosis. Early institution of antibiotic therapy improves prognosis.

295 LEGIONNAIRE'S DISEASE PRESENTING AS HYPOXIC RESPIRATORY FAILURE IN PATIENT WITH HIV

U Warden*, C Horn, M Oye, H Patel. *UF Health, Jacksonville, FL*

10.1136/jim-2018-000974.293

Case report A 54 year old male with AIDS (CD4 116), G6PD deficiency and noncompliance presented with shortness of breath accompanied with productive cough, pleuritic chest pain and subjective fevers. He denied any weight loss, hemoptysis, night sweats, being homeless, incarcerated or being exposed to TB. He was found to be febrile to 102.9, tachycardic and hypoxic requiring non-rebreather. X-ray and CT chest were both significant for multifocal pneumonia. LDH was 259. CBC was significant for leukocytosis with bands. BMP showed hypokalemia and a sodium of 140. Procalcitonin was 1.3. EKG was significant for sinus tachycardia. Urine Legionella antigen was positive. Streptococcus pneumonia antigen, MRSA, respiratory panel, respiratory culture, AFB and blood cultures were negative. He underwent bronchoscopy which showed purulent secretions in left upper lobe and no obstructive lesions. AFB, PCP and fungal cultures were negative. He was started on vancomycin and cefepime, which was later changed to azithromycin to treat legionella pneumonia. He was also treated with prednisone and atovaquone empirically for PCP, which was discontinued once PCP culture was noted to be negative.

Legionella is a Gram-negative intracellular pathogen. Infection usually occurs after inhalation of aerosols from contaminated water or soil. Incidence of Legionnaire's disease is 1.4–1.8 cases per 1 00 000. Risk factors include age greater than 50 and immunosuppression. Our patient's risk factors included AIDS and age but no known Legionella exposure or other sick contacts. Patients usually present with respiratory symptoms; however associated symptoms may include nausea and vomiting. Hyponatremia is present in the majority of cases; however, our patient had a normal sodium. Diagnosis is confirmed via PCR, sputum culture or urine antigen testing. It must be noted that urine antigen testing only detects serotype 1 and thus, PCR or culture should be ordered if antigen testing is negative and there is a high suspicion for the disease. The urine antigen sensitivity and specificity for Legionella is seventy percent and ninety percent, respectively. Extrapulmonary infections such as cellulitis, septic arthritis, endocarditis,

meningitis are rare but have been reported. Treatment includes use of Levaquin or Azithromycin.

296 ACUTE RETROVIRAL SYNDROME: AN UNUSUAL CAUSE OF SEPSIS IN PEDIATRICS

A Webb*, P Scalici. *UAB, Birmingham, AL*

10.1136/jim-2018-000974.294

Introduction The CDC recommends screening all individuals between the ages of 13 and 64 for HIV, yet an estimated 40% of new HIV infections come from an undiagnosed partner. There are myriad perceived barriers to testing for HIV in the adolescent population. HIV remains an under-recognized cause of severe illness in pediatric patients.

Case A 17-year-old previously healthy male presented to an adult Emergency Department with six days of fever, unintentional weight loss, headache, vomiting, abdominal cramps, and chills. He required acute resuscitation for presumed septic shock due to fever, tachycardia, and hypotension with poor perfusion. He received fluid resuscitation and empiric IV antibiotics were started. Labs revealed hyponatremia, lactic acidosis, acute kidney injury, transaminitis, elevated CK, thrombocytopenia, and leukopenia. He was transferred to our pediatric facility for management of presumed bacterial sepsis. As part of the adult hospital protocol an HIV screen had been obtained on arrival and returned positive for HIV p24 antigen, but was antibody negative. Empiric antibiotic therapy was continued until blood cultures were negative. Hemodynamics improved with fluid resuscitation. HIV viral load resulted greater than 10,000,000 copies/mL. He was worked up for common opportunistic infections, all of which returned negative. He was started on highly active antiretroviral therapy and discharged to follow up with Infectious Disease clinic with a diagnosis of acute HIV infection.

Conclusion There is little information in the pediatric literature regarding acute retroviral syndrome. Often, pediatric providers are not thinking of HIV in the acute setting, therefore acute retroviral syndrome is easily missed or at best diagnosis is delayed. HIV testing is an important part of comprehensive adolescent health care, but it is frequently overlooked in the pediatric world. Keeping this diagnosis on the differential is important to provide complete, accurate, and timely care to pediatric patients.

297 COCCIDIOIDOMYCOSIS

¹ME Ziada*, ²B Morehead, ¹LS Engel, ¹M Modica. ¹LSU Health Sciences Center, New Orleans, LA; ²University of Mississippi Medical Center, Jackson, MS

10.1136/jim-2018-000974.295

Case report A 28 year old male laborer who had a history of tobacco use presented with a 7 day course of intermittent high-grade fevers associated with sweats and generalized body aches which he described as 'bone pain' and mild nausea. He emigrated from Mexico 3 years prior and had no other recent travel. On physical exam, his vital signs were BP 123/51 mmHg, pulse 123 bpm, Temp 100.5° F, Resp 22/min, SaO2 100% on room air. He was diaphoretic, with shallow breathing, but lungs were otherwise clear to auscultation. The remainder of his exam including neuro exam was unremarkable. Shortly after admission, he became hypotensive (88/49 mmHg) and he was not

responsive to a 4 L fluid bolus. He required presser support and was transferred to the ICU. Labs were significant for Leucopenia, thrombocytopenia, BUN 28 mg/dl, Cr 1.88 mg/dl, transaminitis and elevated bilirubin. Computed tomographic imaging of the chest revealed a solitary necrotic nodule with fluid and gas. AFB smear and T.Spot were negative. He was treated for lung abscess and discharged. Further laboratory studies returned including Leptospira IgM which was positive but PCR was negative. Coccidioides antibodies IgM and IgG were positive. Repeat CT chest did not show improvement and lung biopsy was performed. Fungal culture from biopsy material grew *Coccidioides immitis/posadasii*.

Discussion Coccidioidomycosis, an infection caused by the dimorphic fungi of genus *Coccidioides*, usually results from inhalation of spores. Clinical disease ranges from self-limited acute pneumonia to disseminated disease, especially in immunosuppressed patients. In the United States, most cases are concentrated in southwest. Less than one-half of all infections come to medical attention because illness is often subclinical. Primary infection manifests as CAP approximately 7–21 days after exposure. Most common symptoms are chest pain, cough and fever. Routine laboratory findings are frequently unremarkable. Common radiographic abnormalities include unilateral infiltrate and ipsilateral hilar adenopathy. In general, mild disease does not require antifungal therapy. Therapy with either fluconazole or itraconazole is recommended for patients who are at higher risk of developing severe disease.

298 CRYPTOCOCCUS MENINGITIS IN IMMUNOCOMPETENT PATIENT

ME Ziada*, A McCormick, S Werner, H Phillips, LS Engel, J Amoss. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.296

Case report An 83 year old man presented with altered mental status and an unsteady gate. Initial vitals were unremarkable. He was agitated and had poor attention span. Magnetic resonance (MRI) study of the brain demonstrated bi-hemispheric anterior and posterior circulation acute to subacute infarcts. He developed a low-grade fever and was started on empiric antibiotics for meningitis. He had normal pressure on lumbar puncture and his cerebrospinal fluid (CSF) was clear, colorless with a glucose 21 mg/dl, protein 188 mg/dl, WBC 108 cells/mm³ (Neutrophils 4%, lymph 84%, Mono 12%), and RBC 10 cells/mm³. CSF culture and smear were negative. Testing for HSV, West Nile virus, tuberculosis, Lyme disease and Syphilis was negative. Antibiotics were de-escalated. A repeat MRI brain showed new infarcts and several contrast enhancing peripheral lesions which were concerning for vasculitis or metastasis. Brain biopsy was deferred. Malignancy workup was unremarkable. He was started on steroids for possible vasculitis and repeat MRI brain suggested improvement. Three days later his condition worsened and his course was complicated with respiratory failure due to pneumonia. His level of consciousness deteriorated and repeat MRI brain showed enlarged ventricles. A ventricular shunt was placed (normal opening pressure). CSF India ink stain was positive for encapsulated budding yeast and he was started on amphotericin B and flu-cytosine and steroids were tapered. Cryptococcus antigen was positive and later fungus cultures grew *C.neoformans*. Unfortunately, the patient passed away 6 days later.

Discussion Cryptococcus meningitis is most common in immunocompromised patients. Infection usually occurs by inhalation with resultant pneumonia or asymptomatic respiratory infection which may be followed by meningitis. Serum cryptococcal antigen is positive in 80%–95% of patients with meningitis. The sensitivity and specificity of CSF cryptococcal antigen is 99% and ~100% respectively. India ink stain has a sensitivity of 65%. Treatment is initiated with amphotericin B plus flucytosine for 14 days followed by consolidation therapy with fluconazole. Treatment duration in the immunocompetent patient is unclear but an average of 10 weeks has been documented.

Medical Education/Medical Ethics/Advocacy Joint Plenary Poster Session and Reception 4:30 PM

Thursday, February 21, 2019

299 PROCEDURAL COMPETENCE IN PEDIATRIC RESIDENTS

A Doss*, E Bishop, EO Schmit, N Tofil, MH Nichols. *University of Alabama at Birmingham, Vestavia Hills, AL*

10.1136/jim-2018-000974.297

Purpose of study To evaluate the effectiveness of our procedural boot camp, which was designed to increase pediatric residents' hands-on experience and self-assessed competence in performing 6 of the 13 ACGME required procedures for pediatric residents.

Methods used Our intervention group each year includes PGY-1 pediatric residents who complete a procedural boot camp at the Children's of Alabama Sim Center. Residents complete pre- and post-boot camp surveys rating their self-assessed competence using a 5-point Likert scale in performing: bladder catheterization, foreign body removal, IV placement, tracheostomy exchange, UAC/UVC placement, bag-mask ventilation, and neonatal intubation. During the index year, PGY-2s and PGY-3s were surveyed as controls.

Summary of results Pre- and post-boot camp mean responses of the 27 residents from the index year showed an increase in self-assessed competence for every skill. The largest increases were in trach changes (1.53 to 3.88), IV placement (1.94 to 3.70), bladder catheterization (2.47 to 4.11), and pediatric intubation (2.26 to 3.53). All of which had p-values < 0.0001. Skills with the least improvement were UAC/UVC placement (2.52 to 3.53, p-value 0.002), and neonatal intubation (2.47 to 3.48, p-value 0.01). When comparing the intervention group with the controls, the PGY-1s had the highest reported competence for bladder catheterization, immunizations, and IV placement. Data analysis of the subsequent years is still ongoing.

Conclusions Each year PGY-1s demonstrate improvement in self-assessed competence in each skill. Areas of most improvement were trach changes, IV placement and bladder catheterization. We theorize these are skills to which PGY-1s are not often exposed during intern year. When comparing across all classes interns report higher competence, which indicates lack of exposure continues throughout residency. We are in the preliminary stages of completing data analysis

of subsequent years of the boot camp. These early results demonstrate similar findings of our index year. We are also in the process of assessing competence at 6 and 12 month intervals post-boot camp. By continuing this project, residents will be able to appropriately rate their competence level per the ACGME guidelines at the time of graduation and in turn perform such procedures proficiently in their future practice.

300

LITERACY PROMOTION IN PRIMARY CARE CLINICS: IMPROVING UTILIZATION AND ADHERENCE TO THE REACH OUT AND READ MODEL

¹S Jafer*, ²A Caldwell, ²M Dunlap, ²S Gillaspay, ²PM Darden. ¹University of Oklahoma, Oklahoma City, OK; ²University of Oklahoma Health Sciences Center, Oklahoma City, OK

10.1136/jim-2018-000974.298

Purpose of study Pediatric providers have a unique opportunity to intervene and help families during early childhood to improve language development and promote literacy. Reach Out and Read (ROR) is a clinical intervention that promotes early childhood literacy. Despite widespread participation in, and success of ROR among clinics across the U.S., little is known about clinics' utilization of, and adherence to, the ROR model. This study evaluates clinic adherence to the ROR model which includes a medical provider providing an age appropriate book for the patient to keep, literacy based anticipatory guidance, and exposure to a literacy rich waiting room.

Methods used Parents of patients ages 6 months through 5 years of age completed a survey during well child examinations at 10 participating clinics in Oklahoma. Preliminary data was analyzed and descriptive statistics obtained. Data will be analyzed further once all surveys are completed and collected.

Summary of results Of the 313 participants, 97.7% of parents reported receiving a book at their visit and of those receiving a book, 67.2% stated that a physician gave them the book, 7.9% another health care provider, 24.5% a nurse and 0.3% other. When asked about their visit that day, 92.5% of parents reported that their provider discussed reading, 94.3% reported they discussed the benefits of reading aloud, 90.7% reported they discussed how to read to their child, and 97% state the provider discussed the importance of talking to their child. Parents were asked about the waiting room of the clinic that day and 82.7% report that there were books in the waiting room, 24.9% reported that there were volunteers reading and 64.2% stated there were displays about reading.

Conclusions Among the parents surveyed, most reported received a book and literacy based anticipatory guidance at their visit. However, a lower proportion of respondents described a literacy rich waiting room. This component of the ROR intervention could be improved at clinics currently utilizing ROR.

301

BARDET BIEDL SYNDROME WITH EARLY ONSET METASTATIC COLON CANCER

WM Kogler*, J Rivas, M Morales Hernandez, V Seeram. University of Florida-Jacksonville, Jacksonville, FL

10.1136/jim-2018-000974.299

Case report A 39 year old female with a past medical history of Bardet Biedl Syndrome (BBS) presents with progressive functional decline over 2 months with associated fever, constipation, and decreased oral intake. She was found to be hypotensive, tachycardic, and afebrile. She was started on antibiotics that were stopped after infectious workup was negative. Further workup revealed hypercalcemia of 16.8, PTH of 330, and creatinine of 2.04. She was admitted to the Intensive care unit, calcitonin and Continuous Renal Replacement Therapy were initiated. Physical exam was significant for retinitis pigmentosa, mental retardation, thick neck, obesity, and enlarged mons pubis. Computed tomography of the abdomen revealed findings consistent with colon cancer with metastatic mesenteric implants measuring up to 1.5 cm. Patient had no known family history of Colon Cancer. After many discussions the family decided to take their daughter home with hospice care after resolution of hypercalcemia

BBS is a rare autosomal recessive disorder with a prevalence of 1 in 1 60 000. It is associated with consanguinity and is more common in developing countries. It is caused by a mutation in the BBS genes, most commonly BBS1-BBS18 that encode for ciliary proteins. The classic signs of BBS include visual problems, obesity, mental retardation, polydactyly, genital dysfunction, and renal dysfunction. BBS has been linked to renal and endometrial malignancy, but a link to colon cancer has not been described in the literature. Diagnosis is clinical; genetic testing can be used for genetic counseling. Treatment involves managing symptoms and improving quality of life for the patient and caregivers.

BBS syndrome is rare but poses a huge challenge for clinicians and caregivers. It has devastating effects on multiple organ systems making treatment a multidisciplinary effort. Malignancy is often a complication of BBS which increases morbidity and mortality. Colon cancer has not typically been associated with these patients but given the early onset, lack of family history, and the pathophysiology of the disease, further study may be warranted. It is an important thing to consider for this patient population, so that quality of life can be improved for the patient and family.

Neurology and Neurobiology

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

302

NEUROSARCOIDOSIS MISDIAGNOSED AS ENCEPHALOMENINGITIS

A Ananthula*, L Singh, RL Archer. University of Arkansas for Medical Sciences, Little Rock, AR

10.1136/jim-2018-000974.300

Case report Neurosarcoidosis (NS) is a rare clinical entity with an incidence of about 5% in patients diagnosed with systemic sarcoidosis. Due to various clinical presentations and lack of specific diagnostic tests, timely diagnosis becomes difficult. We describe a case of a 28-year-old male who presented with new onset seizures, altered mental

status, asterixis and ataxia, visual hallucination with slurred speech and end gaze nystagmus, who was misdiagnosed as encephal meningitis. On arrival, his vitals were stable. MRI Brain and spinal cord showed extensive involvement of hypothalamus, infundibulum and leptomeningeal enhancement suggestive of an atypical infection versus NS. CSF analysis showed elevated cell count 410 cells/ul and total protein 1250 mg/dl and low glucose – 23 mg/dl suggestive of bacterial meningitis vs Tuberculosis (TB) vs. atypical. CT Chest showed concern for fungal vs. Miliary TB. Hence, he was started on RIPE therapy and broad-spectrum antibiotics. He also diagnosed with pan hypopituitarism and was treated for Diabetes Insipidus and hypothyroidism. RIPE therapy was stopped as TB testing was negative. He was discharged with 8 weeks of antibiotics for polymicrobial encephal meningitis. He was also started on Decadron for cerebral edema and decision was made to taper over next 8 weeks. After stopping Decadron, he was readmitted with altered mental status, hypoxia, hypothermia, and hypotension. Given the concern for NS, he was started on prednisone 60 mg daily and he showed drastic improvement. CT chest showed mediastinal and hilar lymphadenopathy. EBUS biopsy was negative for malignant cytology and no granulomas were identified. Later, he underwent mediastinal biopsy and wedge resection of the lung which showed extensive non-necrotizing granulomatous inflammation proving his diagnosis of systemic sarcoidosis. In conclusion, CSF abnormalities in NS are most pronounced in patients with diffuse leptomeningeal enhancement on MRI which can sometimes be very misleading. Patients with clinically active disease can have a significantly higher CSF cell counts, total protein, lactate and significantly lower glucose levels than patients with stable disease.

303 TUBEROUS SCLEROSIS IN THE ABSENCE OF SEIZURE ACTIVITY

JC Baity*, R El-Abassi. LSU New Orleans, New Orleans, ID

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Introduction Tuberous Sclerosis (TS) is a hereditary disorder caused by a mutation in the TSC 1 or TSC 2 gene. Mutation in TSC 1 results in abnormal hamartin protein and TSC2 in abnormal tuberin protein. With TSC thought to act as a tumor suppressor gene, the loss of function of these proteins results in numerous abnormal cells throughout the body. These can include lesions in the brain (cortical tubers), skin (Ash leaf macules, Shagreen patch, skin tags), retinal phakoma, and angiomyolipomas or the liver and kidneys. To neurologists, patients present with seizures in 79%–90% of the cases. We report a patient with TS misdiagnosed as cervical radiculopathy and tension headache. A comprehensive medical history, thorough physical examination, clinical suspicion and neuroimaging were required to unmask the diagnosis.

Case report A 31 year old otherwise healthy female presented complaining of radicular-type, shock-like generalized body pain and independent supraorbital stabbing headaches for 7 years. MRI brain showed nonspecific enhancement near right caudate nucleus. MRI spine revealed multilevel root impingement at cervical and lumbar spines. She had previously been started on Duloxetine for cervical radiculopathy and tension headaches with improvement in neuropathic symptoms. Years later,

MRI brain showed areas of enhancement near right caudate nucleus and a subependymal lesion. MRI spine revealed T1 and T2 hypointense lesions at C4, T4, and T5 and scattered sclerotic bone lesions throughout the lumbar spine and sacrum. MRI abdomen showed liver hemangiolioma and renal angiomyolipomas. Careful assessment of the skin showed Ash leaf macules on her lower back, periungual fibromas, facial angiofibromas and adenoma sebaceum, connective tissue nevus on her right upper back and hypopigmented lesions on her legs and trunk. She had no history of seizure throughout her life and EEG was normal.

Conclusion Vogt's triad of seizures, mental retardation and adenoma sebaceum occurs in less than half of patients, thus requiring high index of suspicion to diagnose TS. After nearly seven years of complaints, our patient was finally diagnosed with TS, in spite of multisystem peripheral manifestations. Seizures are seen in 70% patient even with normal mental status.

304 OSMOTIC DEMYELINATION SYNDROME WITH SYNCHRONOUS WERNICKE'S ENCEPHALOPATHY

¹I Huml*, ²C Chandravanka. ¹Upstate Medical University, Syracuse, NY; ²Syracuse VA Medical Center, Syracuse, NY

10.1136/jim-2018-000974.302

Case report Osmotic demyelination is a syndrome that occurs from rapid increases in serum osmolality, while Wernicke's encephalopathy is a condition that occurs secondary to acute thiamine deficiency most commonly seen in chronic alcoholics. While the two entities' pathophysiologies are mutually exclusive, they are rarely seen to occur concurrently. Here we describe a case of a middle age man who presented to the hospital with increasing agitation and confusion and was found to have both diseases.

A 55 year old man with past medical history of beta-thalassemia trait presented to the VA hospital in Syracuse, NY with the primary complaint of increasing agitation and forgetfulness. While not initially known, it was discovered that patient presented to an outside hospital 13 days prior to his current presentation after a motor vehicle accident. At that time he was noted to have sodium level 98 mEq/l which was corrected to 135 mEq/l in less than 3 days. During the admission to our hospital the patient also claimed to have unsteadiness on his feet and had increasing number of falls over the past several days. He was a recently retired high ranking military official and had been drinking about 1 liter of rum daily. On exam the patient was noted to have right sided ptosis, horizontal nystagmus, diffusely increased muscular tone, and was ataxic. His labs showed normal sodium, TSH, and B12. MRI was obtained which showed diffusion restriction of central pons consistent with osmotic demyelination syndrome. It also revealed increased T2 signal intensity symmetric within the mammillary bodies reflecting a mild degree of Wernicke's encephalopathy. Patient was admitted to the hospital and started on intensive IV thiamine therapy and eunatremia was maintained.

This case illustrates the importance when diagnosing and treating osmotic demyelination syndrome of keeping a high index of suspicion for concurrent Wernicke's, especially in the alcoholic patient. Wernicke's is a treatable disease and making the diagnosis could prevent unnecessary morbidity and

facilitate faster recovery in patients recovering from osmotic demyelination syndrome. At this time further research is required to be able to identify the exact incidence of the concurrent diseases.

305 A RARE CASE OF TOLOSA HUNT SYNDROME

JJ Maguire*, M El Iskandarani, S Elamparithi. *East Tennessee State University, Johnson City, TN*

10.1136/jim-2018-000974.303

Case report Tolosa-Hunt syndrome is a rare neurological disorder with an incidence of one case per million. It is a granulomatous inflammatory condition that affects the cavernous sinus and is characterized by painful ophthalmoplegia and headaches. We present a 57-year-old male who presents with complaints of double vision and headaches. The patient has a history of cluster headaches controlled with sumatriptan. He reports that several days prior to admission headaches were getting more frequent, sharp, localized to the left side. On physical exam vitals were stable, pupils were equal in size and reactive to light but limited left eye abduction and external rotation were noted. The remainder of physical exam was unremarkable including the remaining cranial nerves. CT head was done which showed no evidence of acute stroke. MRI brain was then performed which showed an asymmetric bulge of the left cavernous sinus which raised suspicion for cavernous sinus inflammation. Systemic high dose steroid trial was given. Patient's symptoms improved within 72 hours and diagnosis of Tolosa-Hunt Syndrome was confirmed. The criteria for diagnosis of Tolosa-Hunt syndrome is given by International Headache Society, which includes: Unilateral headache; MRI or biopsy demonstrating granulomatous inflammation of cavernous sinus, superior orbital fissure or orbit; ipsilateral nerve palsy involving one or more of 3rd, 4th and/or 6th cranial nerves; no alternate diagnosis based on the symptoms; specific history of ipsilateral headache localized to the ipsilateral brow and eye and it should occur 2 week before the oculomotor palsy or along with it. Our patient met all the criteria mentioned above. Ruling out other causes of headache and ophthalmoplegia is important in making the diagnosis. A differential diagnosis includes stroke, vasculitis, myasthenia gravis and multiple sclerosis. Currently, inadequate data is available to determine the best route and duration of treatment with steroids. Our patient received oral steroid 100 mg for 3 days followed by slow steroid taper and had improvement in symptoms. Although a rare disorder, it is important to consider Tolosa-Hunt syndrome in the differential diagnosis of patients who presents with headaches and visual changes, especially after ruling out other common causes.

306 HYDROCEPHALUS AS A CAUSE OF SEIZURE-LIKE EVENTS IN AN INFANT

SL Nelson*, B Wild. *Tulane University School of Medicine, Metairie, LA*

10.1136/jim-2018-000974.304

Case report The differential diagnosis for seizure-like events in infants includes Sandifer syndrome, benign sleep myoclonus, infantile masturbation, infantile spasms, etc. Hydrocephalus is

not routinely considered as causative of abnormal movements in infants without other signs of increased intracranial pressure such as macrocephaly. Chiari 1 malformation, where the cerebellar tonsils extend through the foramen magnum into the spinal canal, may obstruct CSF drainage and increases the risk of hydrocephalus. We report an infant who presented with seizure-like events that were later determined to be due to severe obstructive hydrocephalus due to a congenital Chiari 1 malformation.

She presented to the pediatric neurology clinic for initial evaluation at 5-months-old for a 3 week history multiple myoclonic jerks per day. A recent routine EEG was normal. She also had severe reflux and feeding issues for which was being treated without improvement. Her head size and neurological exam were normal. She was admitted for video EEG monitoring due to concerns for infantile spasms. Multiple events were captured and were not associated with any changes on EEG to suggest seizures. MRI of the brain and total spine demonstrated severe hydrocephalus with enlargement of the lateral and third ventricles, Chiari 1 malformation with cerebellar herniation to 6 mm, and syringomyelia extending from C2 to the filum terminale. She had a VP shunt placed to correct the hydrocephalus, and a posterior fossa decompression for Chiari 1 malformation due to the syrinx. This resulted in resolution of the myoclonic jerks as well as her feeding issues.

This infant presented with symptomatic obstructive hydrocephalus secondary to her Chiari 1 malformation. Although uncommon in infants, Chiari malformations can present acutely, and delay in diagnosis and treatment can dramatically impact outcome. Certainly, the longer hydrocephalus and syringomyelia persist without treatment, the more likely they will have permanent neurological sequelae. Once this infant was treated, her seizure-like episodes and feeding issues resolved, demonstrating that they were due to her previously undiagnosed hydrocephalus. Of note, her head size was normal, suggesting that the obstruction was a new process, and that the seizure-like episodes led to early diagnosis and treatment.

307 LATERAL MEDULLARY SYNDROME IN A MAN WITH FOCAL SEGMENTAL GLOMERULOSCLEROSIS

S Prakash*, MK Islam, T Naguib. *Texas Tech University Health Sciences Center Amarillo, Amarillo, TX*

10.1136/jim-2018-000974.305

Case report Lateral medullary syndrome, also known as Wallenberg syndrome, is a clinical syndrome caused by acute ischemic infarction from blockage of blood vessels supplying the lateral region of the medulla oblongata. We present an interesting case presented with bilateral lower leg swelling that was found to have small subacute infarct in the lateral medullary regions thought to be related to hypercoagulability induced by a biopsy-proven focal segmental glomerulosclerosis (FSGS).

A 46-year-old African-American gentleman with history of uncontrolled hypertension presented with bilateral progressive lower extremity painful swelling for 2 weeks. Problems swallowing liquids and solids began 2 weeks earlier along with altered temperature sensation in the arms but not hands. Long history of tobacco and marijuana use was reported but

otherwise he was healthy. Exam showed BP of 224/132, moderate leg edema with faint pulses and abnormal temperature response in right arm. Besides BUN 11, creatinine 0.8, troponin 0.026, albumin 1.4, total protein 4.3, all lab was normal including BNP. Imaging revealed bilateral superficial femoral artery (SFA) occlusion with suggestion of occlusion of anterior tibial artery with distal reconstitution, but no venous thrombosis. Ejection fraction was 60% with left ventricular hypertrophy. MRI of brain showed left lateral medullary changes and T2 signal hyperintensity bilaterally in thalami and in right upper pons. 24 hour urine protein of 3.2 grams was attributed on kidney biopsy to FSGS of perihilar variant with moderate interstitial fibrosis and mild arteriosclerosis.

This case of bilateral lateral medullary syndrome in the setting of nephrotic proteinuria is likely related to underlying FSGS and is exceedingly rare. Appropriate treatment of FSGS may well mitigate the extension of both central and peripheral ischemic complications of the disease. If proteinuria is noted, aggressive diagnosis and management should be pursued not only to protect kidney function but also to prevent target organs from sustaining significant vascular damage. This case highlights the controversy of prophylactic anticoagulation in the setting of FSGS.

308 ELECTROCLINICAL FINDINGS IMPLICATE WHITE MATTER EDEMA IN THE PATHOGENESIS OF TOXIC LEUKOENCEPHALOPATHY

AB Ramos*, RA Cruz, R El-Abassi, LA Branch, EC Mader. *LSUHSC – New Orleans, New Orleans, LA*

10.1136/jim-2018-000974.306

Purpose of study When confronted with subcortical white matter hyperintensity on FLAIR/T2-weighted MRI images, physicians tend to think of immune, inflammatory, vascular, or metabolic causes of leukoencephalopathy. Toxic leukoencephalopathy due to substance abuse, therapeutic agents, or environmental toxins is often overlooked. Patients with toxic leukoencephalopathy often present with neurobehavioral symptoms due to injury of white matter tracts involved in higher cerebral function. However, the breadth of clinical manifestations—from inattention, forgetfulness, and personality change to delirium, coma, and death—suggests that the mechanism of white matter injury is not the same in all patients.

Methods used A 57-year-old man with substance abuse disorder presented with a 5 day history of strange behavior. He was awake and alert but disoriented to time on admission. He exhibited psychomotor retardation, diffuse hyperreflexia, bilateral dysmetria, and gait ataxia. Brain MRI revealed diffuse confluent subcortical white matter hyperintensities on FLAIR/T2-weighted sequences. EEG showed mild diffuse slowing. Blood tests were all normal except for mild hyperammonemia. Toxicology was positive for cocaine. Nutritional therapy, assistance with activities of daily living, and gait training resulted in normalization of his mental status, behavior, sleep-wake cycle, and ambulation.

Summary of results After 3 weeks, he was discharged to a nursing facility. He returned to clinic 5 months later and was noted to have normal cognitive and neurological function. His

follow-up brain MRI showed near-100% resolution of white matter hyperintensity and his follow-up EEG was normal.

Conclusions Our case demonstrates that MRI evidence of extensive toxic leukoencephalopathy does not preclude full functional recovery in patients with a favorable electroclinical profile. The rapid resolution of MRI white matter hyperintensities in our patient imply a reversible mechanism of injury, such as intramyelinic edema. The fact that EEG is sensitive to subcortical white matter disease, but barely affected by white matter edema, makes it a great tool for differentiating subcortical edema from toxic demyelination and axonal loss. EEG should always be included in the work-up of toxic leukoencephalopathy.

309 A CASE OF RAPIDLY PROGRESSIVE HTLV-ASSOCIATED MYELOPATHY WITH URINARY RETENTION

¹S Smith*, ²A Al-Thani, ²A Al-Abdulmalek, ¹T Huq. ¹Tulane University, New Orleans, LA; ²Weill Cornell Medical College, New York, NY

10.1136/jim-2018-000974.307

Case report Human T-lymphotropic virus type I (HTLV-I) infection, although most commonly asymptomatic, can cause serious, debilitating disease in a small percentage of individuals. HTLV is associated with a disabling inflammatory myelopathy that occurs in less than 2 percent of infected individuals. This condition, termed HTLV-associated myelopathy/tropical spastic paraparesis (HAM/TSP), is characterized by progressive lower extremity weakness, diffuse hyperreflexia and clonus, loss of vibratory sensation, and urge urinary incontinence characterized by overactive bladder. Onset is most commonly insidious, beginning with leg weakness and progressing to wheelchair dependence over a mean of 21 years. A 51 year old patient with positive HTLV antibody titers presented to our clinic wheelchair-bound less than a year after symptom onset, with urinary symptoms that had progressed from overactive bladder with urge incontinence to urinary retention. This patient's disease course was unusually rapid, even considering the association between age over 50 at symptom onset and faster motor deterioration. In addition, this report supplements evidence presented in at least one other case that overactive bladder may progress to urinary retention in HAM/TSP. This case also illustrates how these patients can be wrongly diagnosed with multiple sclerosis.

Abstract 309 Figure 1 Hyper-intense T2 MR signal within midline posterior cervical cord involving C1–C6

310 ASCENDING PARALYSIS IN A YOUNG WOMAN WITH PSYCHIATRIC COMORBIDITY

¹AV Varma*, ²K Garvie, ²S Walvekar, ²M Igi, ²M Garikepati. ¹Louisiana State University Health Sciences Center School of Medicine New Orleans, New Orleans, LA; ²LSUHSC, New Orleans, LA

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Introduction Guillain Barre Syndrome (GBS) is an acute inflammatory demyelinating polyneuropathy affecting both motor and sensory peripheral nerves. Typically presenting after a gastrointestinal or a respiratory tract infection, it manifests as ascending paralysis with concomitant areflexia in patients. Cytoalbuminologic dissociation is a supportive finding on CSF analysis. Due to variability in presentation, misdiagnosis and delay in treatment can occur, and consequently, GBS can become life-threatening.

Case report We report ascending paralysis in a 36-year-old woman with history of bipolar disorder who recently recovered from aspiration pneumonia following a drug overdose event. Given her psychiatric history, her presentation was initially dismissed as conversion disorder. On re-evaluation at a tertiary care facility, initiation of intravenous immunoglobulin (IVIG) therapy was decided before CSF findings were made available due to strong suspicion of GBS, based on history and physical exam findings. CSF analysis and radiological findings subsequently supported our clinical suspicion. Early initiation of IVIG therapy, we believe, prevented her from impending respiratory failure. Concurrent aggressive management of her pain, a symptom frequently unattended to in GBS, along with physical and respiratory therapy and monitoring resulted in recovery, without any complications or ICU admission.

Conclusion A thorough history taking and complete (neurological) exam is our best tool in diagnosing disorders which may have a confounding presentation. Psychiatric comorbidities in a patient should not compromise evaluation of a patient. We discuss the variability of GBS presentation, along with reporting our case. Early initiation of therapy in our patient prevented life threatening complications and resulted in improved outcome.

Pediatric Clinical Case Reports

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

311 INACTIVE MATRIX GLA PROTEIN CONCENTRATIONS ARE ASSOCIATED WITH PREDIABETES AND CARDIOMETABOLIC RISK MARKERS IN OVERWEIGHT CHILDREN

R Ahmad*, M Fain, CF Williams, M Brown, R Bassali, C Davis, NK Pollock. Medical College of Georgia, Augusta University, Augusta, GA

10.1136/jim-2018-000974.309

Purpose of study Matrix Gla protein (MGP) is a vascular calcification inhibitor dependent upon vitamin K for activation. Evidence suggests that elevated plasma inactive MGP

(desphospho-uncarboxylated MGP, dp-ucMGP; indicating poorer vascular vitamin K status) is associated with greater cardiovascular disease (CVD) risk. Given that insulin resistance predisposes children to CVD combined with recent findings in overweight children of increasing rates of prediabetes, it is vital to understand the role of dp-ucMGP on cardiometabolic risk markers in this population. We investigated vascular vitamin K status (*via* plasma dp-ucMGP) between 64 overweight children (aged 8–11 y) with normal glucose levels (n=30) and prediabetes (n=34), and the associations of plasma dp-ucMGP with cardiometabolic risk markers.

Methods used Fasting blood samples were measured for glucose, insulin, lipids, adiponectin, leptin, and C-reactive protein (CRP). Using additional fasting plasma, dp-ucMGP was measured as a marker of vascular vitamin K status (insufficient defined as dp-ucMGP >500 pmol/L).

Summary of results Mean dp-ucMGP levels were 2.6 times higher in the prediabetes vs. normal glucose group (471 ± 320 vs. 180 ± 135 pmol/L, $p < 0.01$). Forty-seven percent of the prediabetes group had elevated dp-ucMGP levels compared to 3% in the normal glucose group ($p < 0.01$). Multiple linear regression, adjusting for age, sex, race, and body mass index, revealed that dp-ucMGP was independently related to fasting glucose ($\beta = 0.43$), fasting insulin ($\beta = 0.27$), homeostasis model assessment of insulin resistance ($\beta = 0.28$), HDL-cholesterol ($\beta = -0.23$), and leptin ($\beta = 0.17$) (all $p < 0.05$). No relations were found between dp-ucMGP and adiponectin or CRP.

Conclusions Our data suggest that the higher plasma dp-ucMGP concentrations found in children with prediabetes may be associated with multiple cardiometabolic risk markers. Experimental trials are needed to determine whether vitamin K reduces progression of CVD development by increasing MGP carboxylation.

312 NEUROPSYCHIATRIC SYMPTOMS IN AN ADOLESCENT: DIG DEEPER

M Ahmed Khan*, A Tanios. St.Louis University School of Medicine, Saint Louis, MO

10.1136/jim-2018-000974.310

Case report A seventeen-year-old female presented with a three-month history of depressed mood, behavioral changes, fifteen-pound weight loss and enuresis. Symptoms included increased fatigue, anorexia, anhedonia and insomnia. Patient identified a recent breakup as a trigger. She also reported intermittent headaches and vomiting. Physical exam was notable for flat affect, increased distractibility, slow thought process, generalized hyperreflexia, bilateral Babinski's sign and postural hand tremors. She did fit the diagnosis of Major Depressive Disorder with severe features. However, associated features like urinary incontinence, headache and vomiting episodes could not be explained by an isolated mood disorder. This warranted neuroimaging to look for intracranial pathology. MRI brain revealed a heterogeneous enhancing lobular mass likely arising from the septum pellucidum resulting in obstructive hydrocephalus. She was emergently transferred to the operating room where she underwent external ventricular drainage followed by resection of the tumor by neurosurgery. On histopathology, Grade I Subependymal Giant Cell Astrocytoma was identified. Post operatively, she recovered well in the intensive care unit, subsequent MRI showed complete resection and external

ventricular drains were removed without signs of hydrocephalus and she was discharged home.

Depression is highly prevalent among adolescents in the United States, predominantly females, 70% of whom have severe impairment. Interpersonal dysfunction is a common trigger. Psychiatric symptoms often have underlying medical causes. However, they are a rare presentation of brain tumors. Here, we describe a case of an adolescent who presented with symptoms of depression, had upper motor neuron signs on exam and neuroimaging revealed an intraventricular tumor.

Our clinical conundrum is a classic case of a neuropsychiatric interface where timely diagnosis was very crucial and required emergent neurosurgical intervention. A cluster of vague symptoms that do not fit into a single psychiatric category should prompt a high index of suspicion for intracranial pathology. A thorough history and complete physical examination including neurological is important and neuroimaging will clinch the diagnosis.

313 ATHLETES: SO FAST, THE IMAGING CAN'T CATCH UP

OC Aimuyo*, C Stanecki, M Bolton, MH Roy. *Our Lady of the Lake Children's Hospital, Baton Rouge, LA*

10.1136/jim-2018-000974.311

Case report Sports injuries are a common chief complaint amongst adolescent athletes. These injuries can precipitate serious conditions that may present non-specifically with negative initial imaging. We present a teenage athlete in whom pyomyositis and osteomyelitis initially mimicked an overuse injury, accompanied by creatine kinase (CPK) elevation from presumed overexertion.

A 17 year old male presented with left hip pain following a weekend of intense physical activity. His hip pain became 'searing' and impeded his gait. Initial labs showed normal renal function, microscopic hematuria, proteinuria, and an elevated CPK (1297 IU/L). Plain films were unremarkable and he was admitted for IV fluids. The next day he continued to endorse pain over his left anterior superior iliac spine (ASIS). His CPK and pain did not improve with IV fluids and he soon became febrile. Further evaluation showed an elevated C-reactive protein (87 mg/L) and a possible nondisplaced avulsion fracture from the left iliac crest to the ASIS on MRI. Orthopedics recommended rest and weight-bearing as tolerated. The patient then developed pleural effusions, hypoxemia, and MRSA bacteremia. He was started on vancomycin and received an echocardiogram which ruled out endocarditis. A repeat MRI was obtained four days after presentation due to persistent fever and bacteremia and revealed microabscesses along the left iliopsoas muscle and anterior abdominal wall consistent with pyomyositis and left iliac crest osteomyelitis. He received a CT-guided percutaneous drainage catheter and then gradually began to improve. He completed an 11 day course of vancomycin, and was discharged with 4 weeks of oral clindamycin.

Pelvic pyomyositis and osteomyelitis are rare entities in children and often present non-specifically. Both of these infections are often caused by hematogenous spread, most commonly of *S. aureus*. Pyomyositis can quickly form abscesses, spread to local bone, cause osteomyelitis, and complicate bacteremia, so IV antibiotics should be started promptly

and IR-guided abscess drainage should be considered. To avoid these complications, pyomyositis and osteomyelitis should be considered in adolescent athletes presenting with altered weight bearing and hip pain, especially when initial imaging is not consistent with an infectious process.

314 PERSISTENT HYPOGLYCEMIA IN A NEWBORN WITH HUMAN HERPESVIRUS 6

S Albracht*, E Los, G Ford, D Macariola. *East Tennessee State University, Johnson City, TN*

10.1136/jim-2018-000974.312

Case report One day after elective circumcision, a 5 day-old term AGA infant is admitted for hypothermia and lethargy. Mother reports poor feeding, taking only 2 ounces of formula in the preceding 24 hours, and decreased responsiveness since circumcision. Birth history is significant only for GBS positive mother who received prophylactic antibiotics.

At outside ER, the patient's temperature is 36.0° C (96.0° F) and point of care glucose is 20 mg/dL. Dextrose bolus and maintenance dextrose-containing fluids are started. Physical exam is notable for scleral icterus, mild jaundice, and healing circumcision. After admission, the infant develops apnea and persistent hypoglycemia despite multiple dextrose boluses. Full septic work-up is performed, and empiric ampicillin, cefotaxime, and acyclovir are started. CBC is noted to have mild neutrophil predominance, but otherwise normal for age. BMP shows glucose 24 mg/dL and potassium 7.1 mEq/L (hemolyzed). Total bilirubin is 10.4 mg/dL, below phototherapy threshold. Meningiti/Encephalitis PCR panel is positive for Human Herpesvirus 6 (HHV-6). Empiric antibiotics and acyclovir are discontinued, and a 10 day course of IV ganciclovir is started. Remainder of infectious work-up is negative. Despite ganciclovir treatment, the patient requires a glucose infusion rate (GIR) of 15–17 mg/kg/min (normal GIR 5–8 mg/kg/min) to maintain normoglycemia. He has normal TSH and free T4. Critical labs drawn during confirmed hypoglycemia show elevated free and total insulin at 12 uIU/mL and 14 uIU/mL respectively.

The patient is diagnosed with hyperinsulinism in the setting of HHV-6 aseptic meningitis. Diazoxide therapy for the patient's hyperinsulinism is trialed but fails to achieve euglycemia. Genetic testing for congenital hyperinsulinism is positive for gene mutation. Focal hyperinsulinism is found on the proximal pancreas with ¹⁸F-fluoro-L-DOPA positron emission tomography performed at outside tertiary center. Ultimately, the patient undergoes 50% pancreatectomy, and his hyperinsulinism resolves.

315 THROMBOCYTOPENIA AS PRESENTING SYMPTOM FOR TYPE 1 GAUCHER DISEASE

¹H Alva*, ²C Falcon. ¹Tulane University, New Orleans, LA; ²Tulane Pediatrics, New Orleans, LA

10.1136/jim-2018-000974.313

Case report A 14-year-old girl with chronic ITP presented to the ER with acute, debilitating right thigh pain. Plain films were normal but CT showed lesions consistent with an acute right femur infarction. MRI confirmed acute and chronic bilateral distal femur infarctions. Lab tests for sickle

Abstract 315 Figure 1 A. Typical Gaucher cell (GC) with fibrillary cytoplasm and an eccentric placed nucleus; B. GC with foamy cytoplasm and central nucleus

cell disease were negative. Her pain improved and she was discharged home. Despite Hematology and Orthopedic consultation, an etiology for the bone infarcts was not discovered. Five months later she again developed extremity pain that brought her to the ER for evaluation. Physical exam revealed a normal neurological exam, splenomegaly, confirmed by ultrasound, as well as a platelet count of 69 K (nl 140 K–450 K). Peripheral smear showed an abundance of abnormally shaped red blood cells and thrombocytopenia, which can be seen with evolving myelodysplasia or other bone marrow disorders. Her bone marrow biopsy revealed hyper-cellular marrow with an unusual predominance of large atypical macrophages consistent with Gaucher cells (figure 1). She was referred to Genetics where testing showed glucocerebrosidase of 1.0 (nl >8.7) and glucosylcholine of 1000 (nl <47), both of which are diagnostic of Gaucher disease. The presentation of splenomegaly, thrombocytopenia, bone infarcts, and absence of neurologic involvement are consistent with classic clinical findings seen in Type 1 Gaucher disease.

316 JUVENILE HUNTINGTON'S DISEASE

R Babriwala*, H Gill, E Gonzalez-Toldeo, R Riel-Romero. *LSU-Shreveport, Shreveport, LA*

10.1136/jim-2018-000974.314

Case report Juvenile Huntington's disease presents before 20 years of age with clinical features including myoclonus, seizures, cognitive and behavioral problems. Chorea is usually absent. Huntington's disease is caused by cytosine-adenine-guanine (CAG) trinucleotide repeats. The Juvenile type is associated with more than 60 CAG repeats. Our patient was a healthy male born term without complications during pregnancy, no significant past medical history, and developmentally appropriate until presentation. He first presented at 6 years old when grandmother noticed he was falling more frequently and had difficulty completing tasks. In the next year he was

Abstract 316 Figure 1

diagnosed with seizures with an abnormal EEG. MRI showed volume loss of putamen and caudate nucleus. Mitochondrial test, 343 gene epilepsy panel, plasma creatine and guanidinoacetate panel, and transferrin isoforms for congenital disorder of glycosylation were normal. He had intractable seizures and developed left sided weakness. He regressed developmentally. Three years after presentation he was diagnosed with Juvenile Huntington's. Blood analysis showed 100 CAG repeating sequences. He passed away within 6 months of his diagnosis. This case illustrates rapid progression of Juvenile Huntington's. Although outcome would not have changed, it is important to recognize these symptoms and consider Huntington's disease as a differential diagnosis when regression is present to prevent a delay in diagnosis.

317 CATATONIA: A UNIQUE MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

^{1,2}C Rikard, ^{1,2}B Bagga*, ^{1,2}E Dobish. ¹University of Tennessee Health Science Center, Memphis, TN; ²Le Bonheur Children's Hospital, Memphis, TN

10.1136/jim-2018-000974.315

Case report A 14-year-old female with treatment-naïve systemic lupus erythematosus (SLE) diagnosed two weeks prior presented with three days of progressive paranoia. Clinically, she had a diffuse macular rash, oral ulcers, and delusions; lab studies revealed stable anemia, leukopenia, and elevated ESR. Initial differential diagnosis was broad and included drug abuse, lupus cerebritis, neoplasm, encephalitis, cardiovascular event, or a primary psychiatric condition. Cerebral spinal fluid had elevated glucose and IgG but was negative for infections, multiple sclerosis, and autoimmune panels. Electroencephalogram showed mild diffuse encephalopathy and a urine drug screen was negative.

On admission she was started on high-dose steroids and anti-psychotics. Brain MRI/MRA revealed no sign of cerebritis. Initially she had dysautonomia and was articulate, but she rapidly progressed to a non-verbal state and developed urinary incontinence; cerebritis was the leading diagnosis since imaging can lag. After the first week, she became catatonic, so olanzapine was stopped and SLE therapy was escalated to monthly cyclophosphamide infusions with five days of pulse-dose steroids followed by low-dose daily steroids. Her catatonia was treated with titrated lorazepam. Repeat MRI was unchanged, eliminating cerebritis. Other diagnoses such as autoimmune encephalitis, heavy metal toxicity, Wilson's disease, macrophage activating syndrome, and infectious encephalopathies were excluded. After this extensive workup, she was diagnosed with neuropsychiatric SLE catatonia, a rare presentation of pediatric lupus without well-defined treatment guidelines. Since she remained nonverbal and altered, a trial of IV immunoglobulins (IVIG) was administered, which led to marked clinical improvement.

As she improved, quetiapine, mycophenolate, and hydroxy-chloroquine were started. She was discharged with that regimen, a lorazepam wean, and outpatient rehabilitation. She returned to school this fall. This case report represents the importance of understanding the broad neuropsychiatric manifestations of SLE and highlights a rare case of SLE-related catatonia which was uniquely and successfully treated with IVIG.

318 DELAYED DIAGNOSIS OF ANNULAR PANCREAS IN 11 YEAR-OLD GIRL WITH DIGEORGE SYNDROME – CASE REPORT

¹MM Barr*, ²J Gilbert, ²Z Murrell. ¹UAB School of Medicine, Birmingham, AL; ²Huntsville Hospital, Huntsville, AL

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Case report DiGeorge Syndrome is a collection of symptoms stemming from a heterozygous microdeletion on chromosome 22. Most patients have palate abnormalities and/or cardiac defects, but additional features vary in presentation. DiGeorge syndrome is loosely associated with various GI malformations, but annular pancreas has not been reported elsewhere in medical literature.

An eleven-year-old girl with DiGeorge syndrome presented to Pediatric Surgery with a history of episodes of vomiting since birth and a recent hospitalization for aspiration pneumonia. Upper GI series demonstrated a large, distended stomach and duodenum with severely delayed passage of contrast into distal bowel.

At birth, she was noted to have a cleft palate, but no cardiac abnormalities and was later confirmed to have DiGeorge Syndrome. She had recurrent non-bilious emesis, sometimes projectile, throughout infancy and childhood. Pyloric ultrasound was negative for pyloric stenosis, and her presumed gastroesophageal reflux disease was treated medically. Over time, she limited her diet to sauces and other soft foods; avoiding meat entirely. Her weight and linear growth were normal for age. While being evaluated for recent aspiration pneumonia episode, an upper GI series revealed a dilated stomach and duodenum from a suspected obstructing duodenal web. At surgery, an annular pancreas was noted as the cause of obstruction. The obstruction was treated by a

diamond duodenoduodenostomy and her postoperative course was unremarkable. She was discharged tolerating a mechanical soft diet.

Most annular pancreas patients present with complete obstruction in infancy, yet this patient with DiGeorge syndrome presented with moderate symptoms in early adolescence. Although DiGeorge is known to involve gastroesophageal reflux, a thorough evaluation should be performed to determine anatomic causes of delayed gastric emptying. This is the first case of DiGeorge syndrome reported with annular pancreas; her clinical course represents a delayed diagnosis as our patient was able to modify her diet to allow for foods that could fit through the stenotic duodenal lumen and grow appropriately.

319 JAK INHIBITOR FOR TREATMENT OF STEROID REFRACTORY STAGE IV ACUTE GVHD IN A PEDIATRIC HEMATOPOIETIC STEM CELL TRANSPLANT PATIENT

A Bauchat*, R Craver, L Yu. LSUHSC, New Orleans, LA

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Case report Acute graft versus host disease (aGVHD) is a common complication and the leading cause of mortality in allogeneic hematopoietic stem cell transplantation. First-line treatment is high dose steroids, however, approximately 40% have steroid-refractory (SR) disease and carry a dismal outcome. Unfortunately, no second-line standard of care has been established. Ruxolitinib is a Janus-associated Kinase (JAK) inhibitor, which causes reduced inflammation and inhibition of alloreactive T-cells. Recent studies utilizing Ruxolitinib demonstrate encouraging response rates, however, clinical experience remains very limited especially for the Pediatric patients.

Our patient is a 1 year old male with X-linked lymphoproliferative syndrome who underwent an unrelated 5/6 molecularly matched cord blood transplant after receiving RIC conditioning with campath, fludarabine, thiotepa and melphalan. He received Tacrolimus and Mycophenolate for GVHD prophylaxis as well as a gut decontamination regimen. Post-transplant, he developed idiopathic pneumonia syndrome requiring treatment with Methylprednisolone 2 mg/kg/day and Enbrel with resolution of his symptoms. While on this therapy, he developed persistent diarrhea. Methylprednisolone was increased to 5 mg/kg/day due to concern for aGVHD which was confirmed on day +20 by rectal biopsy. Clinically, he met the criteria for stage IV with abdominal pain, hematochezia, and stool output >2000 mL/day. Infliximab was initiated. Supportive care measures included Protonix, Novo7, and Octreotide. Despite these interventions, he required multiple transfusions and daily adjustments for electrolyte imbalances. On day +26, Ruxolitinib 2.5 mg twice daily was initiated. Subsequently, the patient's hematochezia resolved, the transfusion requirements declined rapidly, his supportive care measures were gradually discontinued, and a steroid taper was started without any flare up of the GVHD. He has received Ruxolitinib for over 2 months without recurrence. This case report demonstrates the successful utilization of Ruxolitinib as salvage therapy for SR, grade 4 gastrointestinal aGVHD in a pediatric patient. Further clinical trials are necessary to thoroughly research the role of Ruxolitinib in the management of aGVHD.

320 ALTERED MENTAL STATUS PRESENTING TO EMERGENCY DEPARTMENT

H Baumann*, P Ghosh, M Johnson. *University of Alabama at Birmingham, Birmingham, AL*

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Case presentation Our patient is a 9 yo previously healthy African American female who presented to our ED with altered mental status. She had blurry vision and been 'off balance' with slurred speech for a month. She had intermittent vomiting with weight loss, polyuria and polydipsia. She was worked up by her PMD for behavioral issues with normal labs but concern for thyroid issues so an endocrine referral was made. One day prior to presentation she reported bugs crawling on her skin, worsening vomiting with increased sleepiness so mom presented to the ED. Only medications in the home were Amoxicillin and cetirizine. No headache, neck pain, fever, weakness, numbness, incontinence, or trauma.

ED Course Physical exam: Her vital signs were T 97, P 114, BP 105/68, Sats 100% on RA. She was somnolent but arousable, able to follow commands. Pupils- BERRL. Cranial nerves, power, tone were intact. Speech was slurred. No abnormal movements or nystagmus. Remaining physical exam normal. Differential diagnosis included ingestion, meningitis, electrolyte imbalance or intracranial mass.

Labs were significant for sodium 183, chloride 138, BUN 117, Cr 2.8, H/H 14.5/43. Negative UDS, acetaminophen and aspirin levels. CT head was notable for suprasellar mass. She received a normal saline bolus and nephrology, neurosurgery and endocrine were consulted. Patient had hypernatremic dehydration with secondary acute kidney injury. She had polyuria and polydipsia for 2 months likely due to diabetes insipidus from her suprasellar mass. She was started on D2.5 NS and transitioned to D5 NS while replacing urine output to slowly bring down the sodium. DDAVP was held initially due to low UOP. Further imaging and staging in the OR was held until she was medically stable. She was admitted to the pediatric intensive care unit for further management.

Discussion The differential for altered mental status is broad. Quick physical exam and assessment with blood gas, electrolytes, kidney and liver function as well as a CT scan is crucial in ruling in or out life threatening causes. In this case, the patient had compounding factors leading to her altered mental status. Severe hypernatremia in children is rare and can lead to significant brain injury. It should lead you to examine for further neurologic causes in a cause with altered mental status.

321 UNUSUAL CAUSE OF CHEST PAIN IN A PEDIATRIC PATIENT

LE Bradley*. *LSUHSC-Shreveport, Shreveport, LA*

10.1136/jim-2018-000974.319

Introduction Chest pain is a common complaint seen in pediatric practice and can cause a great deal of concern for parents and physicians. In most cases, chest pain is likely due to MSK pain or GI distress that will likely resolve on its own. However, though rare, more serious pathologies should be considered if presentation is not consistent with usual causes. Here we report an uncommon cause of chest pain in a young patient.

Case report A 13 year-old female presented to her local ER with gradual onset of chest pain. She denied wheezing, cough, or shortness of breath. The patient had no significant PMH, no recent trauma, surgery, or respiratory infection. The patient was not tachypneic and appeared to be in no distress. Upon auscultation, she had no decreased breath sounds or wheezes. Cardiac examination was benign except tachycardia. EKG was unremarkable. A chest X-ray was ordered that suggested moderate pneumomediastinum (PMS). A chest CT was performed that confirmed the diagnosis. Contrast esophagogram was used to rule out the possibility of an esophageal tear. It should be noted that several hours before the onset of the patient's chest pain, she was playing the French horn for many hours. At this time, we believe it may be possible that the PMS was caused due to the excessive force required to play this instrument. She was managed conservatively with analgesics and rest.

At her follow up visit after 2 weeks, we found that her chest pain had resolved and that her respiratory exam was benign. Repeat imaging showed near total resolution of PMS. The patient was advised stay well rested for next few weeks.

Discussion PMS occurs when air is able to leak into the mediastinal cavity. PMS can be classified as spontaneous when a precipitating factor is not otherwise apparent, or secondary when an obvious cause can be identified such as trauma, acute severe asthma, respiratory infection, or following an invasive procedure. Assuming the patient is stable, PMS is most often managed conservatively as the trapped air will likely be reabsorbed by the surrounding tissues.

Conclusion Though spontaneous PMS is rare, it should be considered in a patient who presents with otherwise unexplained chest pain, even without an obvious cause.

322 MYOSITIS WITH FASCIITIS FOLLOWING INTRAMUSCULAR INJECTION OF PENICILLIN IN A CHILD-A UNIQUE COMPLICATION

K Bybee*, A Bogie. *University of Oklahoma Health Science Center, Oklahoma City, OK*

10.1136/jim-2018-000974.320

Case report 7 y/o presents to the ED w/left leg pain and swelling. Pt was seen by PCP 3 days prior and diagnosed with strep pharyngitis and given IM Bicillin in the left thigh. Pt presented to the ED that same day several hours later secondary to thigh pain, swelling and discoloration. Pt had CT of thigh that showed no obvious hematoma or other significant abnormalities. Pt discharged home w/warm compresses and NSAIDs.

Pt returns on day of presentation with worsening swelling and pain. Unable to ambulate. The swelling has now spread beyond the thigh and includes the knee. No erythema. No fever, sorethroat, headache. No numbness or tingling in the left leg but does have 'shooting' pains in the left knee and thigh with movement. No hx of clotting disorders.

HR 101, o/w NML appears to be in pain HR 101, pulses 2+ in all extremities including left popliteal and left dorsalis pedis Left thigh with edema, firm and warm to the touch. No erythema, +mottling. Small circular hematoma on lateral thigh. Left knee with effusion and TTP. Lower leg normal skin color/temp and no swelling. Decreased ROM of left knee secondary to pain. Sensation, pulses and reflexes normal LLE.

Labs:

WBC 19.3, o/w nml
 CMP nml
 CRP 23.5 (Nml 0–10), ESR 28 (Nml 0–20)
 Significant pain and unable to ambulate despite IV meds.
 Ortho consulted secondary to concern for developing compartment syndrome.
 Admitted, IVF, IV Toradol, MRI w/o contrast.

Repeat labs:

WBC 19.3 →15.8 →12.2
 ESR 28 →45 →9
 CRP 23.5 →17.9 →7.4
 CK 75 k →59 k
 AST 2141 →1556 →1305
 ALT 888 →745 →629

MRI LE w/o: Diffuse edema and abnormal enhancement in muscles of anterior compartment of left thigh with fluid extending along fascial plains. Muscles also appear enlarged. Findings consistent with diffuse myositis and fasciitis. Imaging findings also concerning for compartment syndrome but that is not a radiographic diagnosis.

Diagnosis Chemical myositis and fasciitis secondary to IM Bicillin

Daily measurement of thigh circumference with initial increase followed by decrease prior to dc home. MIVF 2/2 increased CK. Afebrile during stay. No abx given. D/C home 2 days later. 1 week follow up with near resolution of swelling and pain.

323

GORLIN SYNDROME: A MILD CASE DUE TO UNREPORTED *PTCH2* VARIANT AFFECTING AT LEAST TWO GENERATIONS

¹KR Casano*, ²H Meddaugh, ^{2,3}R Zambrano, ^{2,3}M Marble, ^{2,3}J Torres, ^{2,3}Y Lacassie. ¹LSU New Orleans School of Medicine, New Orleans, LA; ²Children's Hospital, New Orleans, LA; ³LSU Health Sciences Center, New Orleans, LA

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Case report Gorlin syndrome, also known as Nevoid Basal-Cell Carcinoma Syndrome, is an autosomal dominant tumor syndrome that presents early in life with congenital malformations and tumors. This syndrome is caused by mutations of the *PTCH1* tumor suppressor gene. Major diagnostic criteria include two or more palmar pits, odontogenic keratocysts, calcification of the falx cerebri, a first-degree relative with Gorlin syndrome, and one basal cell carcinoma (BCC) prior to the age of thirty or more than five BCC in a lifetime. Minor criteria include macrocephaly, cleft lip or palate, hypertelorism, ovarian or cardiac fibroma, medulloblastoma, rib and vertebral anomalies, polydactyly, and ocular anomalies. Various frameshift, missense, and nonsense mutations of the *PTCH1* gene have been reported as a cause of this rare syndrome. Recently, two separate case reports have implicated mutations in the *PTCH2* gene as a cause of Gorlin syndrome. In both cases, mutations of *PTCH2* lead to mild clinical presentations of Gorlin syndrome.

We report a patient with an unpublished *PTCH2* mutation inherited from his father. The proband displays several typical yet milder phenotypic features of Gorlin syndrome, thus confirming the pathogenic role of this gene. Major features in the proband include macrocephaly, a wide face and large forehead, hypertelorism/telecanthus, large eyes, cleft lip and palate, thin vertical creases on his palms, penoscrotal inversion, and a pigmented spot on his penis. His father displays evident macrocephaly, several nevi on his back, and a single palmar pit in

his left hand. Whole exome sequence (trio) showed the proband is heterozygous for Pro1116Leu in exon 21 of the *PTCH2* gene inherited from his father. This semi-conservative amino acid substitution has not been reported as either a benign or pathogenic variant. This report supports and confirms the role of *PTCH2* in mild cases of Gorlin syndrome.

324

METABOLIC STROKE AND THE UTILIZATION OF ARGININE IN MITOCHONDRIAL DNA DELETION SYNDROMES

JS Chawla*, L Puttock, S Sarkar. LSU Health Sciences Center, Metairie, LA

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Case report Mitochondrial DNA deletion syndromes can present as three well-described, overlapping phenotypes: Kearns-Sayre syndrome (KSS), the triad of pigmented retinopathy, progressive external ophthalmoplegia (PEA), and onset before age 20; Pearson syndrome (PS), a combination of sideroblastic anemia and exocrine pancreas dysfunction often fatal in infancy; and isolated PEA. Stroke-like events and their effective treatment with arginine have been studied in patients with other mitochondrial disorders. Similarly, KSS and PS can exhibit cellular respiratory chain dysfunction, but there is limited documentation of stroke-like episodes associated specifically with these MiDNA deletion syndromes, and data on these patients' responsiveness to arginine therapy is limited.

We present the case of a 13 year-old male with PS, KSS, and dyslipidemia who presented with altered mentation and emesis following an unwitnessed fall. Patient denied head trauma but displayed ataxia, hemiparesis, aphasia, and drowsiness. Head CT was unremarkable, and patient was treated for a concussion. Neurology was consulted for persistent altered mental status and neurologic deficits. MRI/MRA brain showed acute ischemia on a background of chronic global atrophy consistent with an acute stroke. Patient was transferred to PICU and started on IV arginine chloride. Following initiation of therapy, he exhibited a swift return to baseline cognition. He was discharged on oral arginine, referred to physical therapy, and exhibited eventual return to baseline of all gross motor function and coordination at subsequent neurology clinics.

It is critical to recognize stroke-like episodes in patients with mitochondrial disease presenting with acute neurologic changes, as rapid initiation of treatment has been shown to improve outcomes. There is significant research available regarding the efficacy of arginine to treat metabolic strokes in MELAS patients; however only recently has there been research that arginine can similarly benefit patients with mitochondrial diseases such as KSS and PS in the context of acute metabolic strokes. Our case study highlights this atypical manifestation of MiDNA deletion syndromes and supports the efficacy of this treatment option.

325

ACUTE PANCREATITIS AS PRESENTATION OF DIFFUSE LARGE B-CELL LYMPHOMA

N Zeky, M Collins*, G Aljomah, E Klepper, K Sims. Our Lady of the Lake Regional Medical Center, Baton Rouge, LA

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Case report Diffuse large B Cell lymphoma is a type of non-Hodgkin lymphoma (NHL). Each year there are about 800

new cases of NHL diagnosed in pediatric patients in the United States. Large B-Cell Lymphoma is the most common type. Traditionally, it presents with symptoms of compression from lymphadenopathy and can include wheezing, facial swelling, difficulty breathing, or abdominal pain. Despite NHL being a common type of pediatric cancer, only about 2% of them have pancreatic involvement.

A seventeen year old female presented to the Emergency Room for intermittent, cramping abdominal pain that was worse with eating. Initial work up demonstrated an elevated lipase of 1564 U/L, normal liver function tests, and a normal complete blood count. Abdominal computed tomography confirmed the diagnosis of pancreatitis. She was admitted for routine pancreatitis care, including intravenous hydration and pain control. She continued to struggle with abdominal pain with advancement of her diet. Repeat labs were obtained on hospital day four showing a new elevation in her liver function tests (ALT 373 U/L, AST 230 U/L), an elevated gamma-glutamyltransferase (489 U/L) and elevated direct bilirubin (1.3 mg/dL). Abdominal ultrasound showed gallbladder sludge with a normal common bile duct diameter. To further evaluate for possible obstruction, a magnetic resonance cholangiopancreatography was done, which showed nodularity of the pancreas with scattered nodular areas in the kidneys suspicious for lymphoma. Persistently elevated liver function tests, lipase, and direct bilirubin led her to have an endoscopic ultrasound for biliary stent placement and pancreatic nodule biopsy. Pathology report was consistent with high grade CD10 positive B-Cell Lymphoma. Bone marrow biopsy confirmed evidence of disease as well. Positron emission tomography scan showed widespread disease with activity in the neck, chest, abdomen and pelvis with skeletal involvement concerning for stage 4 disease.

Acute pancreatitis in children is most often secondary to gallstone obstruction, medications or an autoimmune cause. Similarly to lymphoma, pancreatitis can present with abdominal pain, anorexia, emesis and weight loss.

326 BOY VS. NAIL GUN

M Collins*, N Zeky, K Iheagwara. *Our Lady of the Lake, Baton Rouge, LA*

10.1136/jim-2018-000974.324

Case report A 9-year-old male presented to the Emergency Department (ED) with a puncture wound to his back after sustaining an injury from a nail gun. He was ambulatory on arrival to the ED and his Glasgow Coma Scale was 15. He complained of significant back pain but had no evidence of weakness, decreased sensation, or altered mental status. Peripheral pulses were adequate in all extremities, and his vital signs were stable. A puncture wound was noted on the patient's back just left of the midline at the level of T7. A chest x-ray was obtained immediately, which confirmed the presence of a metallic object over the T7–8 left paracentral area. Computed tomography (CT) of the thoracic spine without contrast and CT angiogram of the chest showed a metallic nail traversing the left T8 pedicle and reaching the anterior cortex. The images also showed the distal tip of the nail abutting and causing slight contour abnormality of the descending aorta without evidence of perforation. Trauma surgery, pediatric surgery, pediatric neurosurgery, and cardiothoracic surgery were consulted. Laboratory studies obtained

Abstract 326 Figure 1

showed normal hemoglobin and hematocrit as well as normal coagulation studies. The patient was given intravenous morphine for pain, and after review of imaging with all subspecialists, he was taken to the operating room with pediatric surgery for video assisted thoracoscopy and foreign body removal. During the procedure, it was noted that lungs, diaphragm and chest wall surfaces appeared normal. The nail penetrated through the left trapezius and erector spinae muscles. It was removed with moderate bleeding that resolved with pressure. Repeat CT scan performed postoperatively showed no adverse findings. He was discharged with seven days of prophylactic antibiotics and close follow up with surgery and his pediatrician. He remained well and asymptomatic at his follow up visits.

327 HOSPITALIZATION FOR ELEVATED LEAD LEVEL – BUT IS IT TRULY ELEVATED?

S Cohen, H Foote, SU Das*. *Duke University School of Medicine, Durham, NC*

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Case report A 12-month-old previously healthy male was admitted to the hospital following screening capillary blood draw at his PCP's office demonstrating a blood lead level (BLL) of 79.4 mcg/dL. He was developmentally appropriate for age. He stayed at home with his mother in a house built in 2002 in Wake County, NC. He had age-appropriate vital signs, a normal physical examination, and normal growth curves. A repeat venous BLL was drawn, and after discussion with medical toxicology, he was started on a course of succimer as recommended per CDC guidelines. The venous BLL returned as 1.8 mcg/dL and succimer was discontinued. His family subsequently realized that just prior to the fingerstick he had been playing with a keychain that had been purchased in Asia. Molecular analysis of the keychain at his father's office revealed its surface to be largely composed of lead. Routine screening for elevated BLLs is typically performed by capillary blood draw, with subsequent venous blood draw to confirm BLLs greater than 5 mcg/dL. Possible reasons for a higher BLL on capillary as compared to venous blood draw include lab error, true variation in BLLs, and sample contamination. In a study using state lead poisoning surveillance system data, 73 percent of elevated capillary screening tests (2.2 percent of all capillary screening tests) were false positives. False positive results were less likely for higher capillary BLLs; these were hypothesized to be secondary to sample contamination as in our case. 11 out of 15 150 children screened had

false positive capillary BLLs high enough to meet criteria for chelation therapy per CDC guidelines. Considering our patient with a BLL of 79.4 mcg/dL, the CDC recommends urgent repeat venous draw, hospitalization for chelation therapy in conjunction with toxicology consultation, in addition to evaluation with a history and physical exam including detailed neurological and developmental assessment, hemoglobin and iron studies, and an abdominal x-ray, all of which were done for our patient. In conclusion, it is rare to have false positives for such high BLLs like our patient. However, it is possible and the most likely cause is sample contamination. Through this case, we want to highlight the importance of proper sample collection methods for capillary BLL screening.

328 OTITIC HYDROCEPHALUS IN A 5 YEAR OLD WITH ACUTE OTITIS MEDIA

AL de la Torre*, A Edwards. *University of Florida College of Medicine- Jacksonville, Jacksonville, FL*

10.1136/jim-2018-000974.326

Case report 5 year old female presenting to the Emergency Department with 3 day history of right acute otitis media for worsening headache. A CT scan revealed suspicion for a lateral sinus thrombosis. She was admitted for intravenous antibiotics, anticoagulation, and MRI/MRV, which confirmed a sinus thrombosis spanning from her right transverse sinus to the proximal internal jugular vein. She began displaying signs of mastoiditis and a mastoidectomy with bilaterally myringotomy were performed when her pain worsened. She tolerated the procedure well, however on POD#1 she was noted to have mild ipsilateral abducens palsy thought to be secondary to inflammation. There was minimal improvement to her pain and on POD#3 she developed significant bilateral abducens palsy. Neurosurgery and ophthalmology were consulted. A repeat MRI/MRV revealed improvement in the thrombosis but ophthalmology exam confirmed bilateral papilledema and she was started on acetazolamide. On POD#5 she underwent therapeutic LP with opening pressure >35 mmHg, 30 cc of clear CSF were removed. Post procedure she reported significant improvement in pain. She was stable and discharged on a 4 week course of Ceftriaxone and Clindamycin as well as enoxaparin, with planned followup for repeat therapeutic LPs as needed.

329 RICKETS AND HEART FAILURE: A BREAST-FED STORY

KA Degatur*, A Olanrewaju. *LSUHSC-Shreveport, Shreveport, LA*

10.1136/jim-2018-000974.327

Case report A 5 month old black male presented with a 2 month history of noisy breathing and poor feeding. Prior to admission, he had two clinic and one ER visit for his noisy breathing and was diagnosed with a viral URI. Of note, his weight had progressively dropped at each visit with a 400gram loss over a month. He was exclusively breast-fed with no vitamin D supplementation. He was also taking longer to feed and was sweating with feeds.

Vitals signs revealed a heart rate of 148 beats per minute, respiratory rate of 54 breaths per minute, and oxygen saturation of 100% on room air. He was afebrile, weighed 6.13 kg

(4th percentile) and measured 0.605 m (0.49th percentile). Physical exam showed an alert, and smiling infant. Cardiac exam was normal. He had noisy breathing, with subcostal retractions. No cyanosis, clubbing or hepatosplenomegaly noted.

Due to increased work of breathing and evidence of obstructive breathing pattern, ENT was consulted for nasal airway placement. Nasal trumpet improved infant's ability to breast-feed, and he was more comfortable. CXR was notable for evidence of left atrial enlargement as well as evidence of rickets. An echo was obtained which showed EF 30%–35% with dilated cardiomyopathy. BNP was 3275. He was transferred to ICU for treatment of heart failure with initiation of captopril and Lasix. He also had hypocalcemia, elevated alkaline phosphatase, low vitamin D and elevated PTH. He was started on vitamin D and Ca for treatment of nutritional rickets. Mother's breast milk was also fortified to 24 kcal.

Patient tolerated the initiation of cardiac medications, he was breathing well on room air and tolerating all feeds without difficulty and showed considerable weight gain prior to discharge.

After 2 months of calcium and vitamin D supplementation, his ejection fraction improved to 55% and to 65% after 4 months of treatment. His calcium level trended up while alkaline phosphatase and PTH trended down.

This case demonstrates the potential for rickets in industrialized countries. Previous case reports document rickets occurring among immigrant families. In 2008, the AAP increased the recommended intake for vitamin D to 400 IU/day. Vitamin D is crucial for the absorption of calcium. Deficiency leads to hypocalcemia, rickets, osteomalacia, and cardiomyopathy. Breast milk contains an average of 22 IU/L of vitamin D.

330 LUNG TRANSPLANTATION IN CYSTIC FIBROSIS: A CASE REPORT

KA Degatur*, S Kilaikode, R Toner, J Treadwell, K Scott. *LSUHSC-Shreveport, Shreveport, LA*

10.1136/jim-2018-000974.328

Case report Cystic fibrosis (CF) is a genetic disease causing complications due to thick secretions in various organs. Patients with advanced lung disease in CF may need to be evaluated for lung transplantation (LT). CF is the most common underlying etiology for pediatric LT. We report a pediatric CF patient who underwent LT with improvement in symptoms and lung function.

13 year old boy was diagnosed with CF at the age of 1 month. He initially presented with meconium ileus and bowel obstruction at birth, positive sweat chloride testing levels (92.8 and 92.7 mmol/L) and genetic mutation F508del and N1303K on newborn screening. He was regularly followed in our CF center. His respiratory cultures grew pseudomonas aeruginosa and MRSA during his first year. He had recurrent respiratory symptoms and infections requiring multiple hospitalizations for aggressive airway clearance and IV antibiotics. X ray showed extensive bronchiectasis with severe hyperinflation. His respiratory status, weight, and lung function progressively declined. He eventually required home oxygen therapy. At this point we sent him to a transplant center for evaluation. He was listed for LT and eventually underwent

bilateral LT. He was seen about 3 months after LT in our CF Center. He remains asymptomatic with normal lung functions. He does not require any respiratory treatments. He is on post-transplant immunosuppression. Respiratory culture are negative. He will continue to follow up in our CF center and the transplant center.

Bilateral lung transplantation has been shown to be an important therapeutic option for end-stage CF pulmonary disease. Transplant function and patient survival after transplantation are better in CF than in most other indications for this procedure among children. Our patient had advanced lung disease with pre-transplant FEV1 of 20%. He is currently doing well 3 months post-transplant. Lung functions showed significant improvement and he is well without requiring any respiratory treatments. Timely evaluation and referral for lung transplant is crucial in CF patients with advanced lung disease. Lung functions and quality of life significantly improve after LT as evident from our patient.

331 A CASE REPORT OF PEDIATRIC AUTOIMMUNE NEUROPSYCHIATRIC DISORDER

¹JT Dille*, ²WC Wilson, ²S DeLeon. ¹University of Oklahoma, Edmond, OK; ²University of Oklahoma Health Science Center, Oklahoma City, OK

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Case report We present the case of a 15 year old male with a history of autism, obsessive-compulsive disorder (OCD), and one-time seizure 15 months prior, admitted with generalized tonic-clonic witnessed seizures and 3 months of cognitive decline, increased OCD symptoms, new onset motor and vocal tics, decreased oral intake, bladder/bowel incontinence, decreased interests, poverty of speech, and social withdrawal. Vital signs, initial labs, EEG and MRI were unremarkable. The patient intermittently responded with single word answers and was unable to cooperate with the neurological exam, though motor function and cranial nerve observation were grossly unremarkable. With neurology's assistance, a broad differential was evaluated with the following labs: TSH, B12, ammonia, folate, serum/urine amino acids, rapid strep, thyroid antimicrosomal Ab, anti-thyroglobulin Ab, Anti-NMDA Ab, ASO-Ab titer, Anti-DNase B, lactate, pyruvate, very long chain fatty acids, copper and ceruloplasmin. Psychiatry evaluated the patient and found overmedication contributed to his neuropsychiatric status as he was on risperidone, alprazolam and lorazepam. As the patient's ASO titer and anti-DNase B were significantly elevated in the setting of recent worsening of OCD symptoms, new onset of vocal and motor tics and parental description of worsening OCD symptoms during times of sickness in the past, he was diagnosed with pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS). He was started on a 5 day course of high dose azithromycin to be followed by a one year prophylactic dose, and levetiracetam was started for seizure prophylaxis.

PANDAS is thought to be an autoimmune-related encephalitis which occurs in a subset of children who have either new or exacerbation of existing OCD or tic disorder symptoms with temporal relationship to group A streptococcus infection. The diagnosis is controversial as there are not standard diagnostic or treatment protocols. Further investigation into cases

such as this to clearly define evidence-based diagnostic criteria and treatment guidelines.

332 METASTATIC RHABDOMYOSARCOMA PRESENTING WITH FEATURES OF GRADENIGO'S SYNDROME

^{1,2}K Dillon*, ^{1,2}C Stallings, ^{1,2}E Dobish. ¹University of Tennessee Health Science Center, Memphis, TN; ²Le Bonheur Children's Hospital, Memphis, TN

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Case report A seven-year-old previously healthy girl presented with right-sided facial droop with the inability to close the right eye. She complained of right frontal headaches for the preceding three weeks, and parents reported a history of fever several days prior to presentation. On exam she had palsy of cranial nerves VI and VII as well as right serosanguinous effusion behind her right tympanic membrane. Non-contrasted CT scan of the head was negative for any intracranial abnormalities. Cerebrospinal fluid studies were not consistent with meningitis. Contrasted MRI noted marrow edema with enhancement of the right petrous apex suggesting osteomyelitis and Gradenigo's syndrome. She was started on vancomycin, ceftriaxone, metronidazole and steroids. Per infectious disease and ENT consults, repeat CT of the sella, posterior fossa, and ear was ordered due to patient being afebrile and having a normal CBC and CRP. This showed a nine-millimeter soft tissue lesion of the right mesotympanum. Biopsy of the lesion indicated a high-grade, small round blue cell neoplasm confirmed to be alveolar rhabdomyosarcoma (ARMS). Complete staging noted T1N0M1 stage 4, group IV ARMS with the right foot as presumed primary site and diffuse bony metastases. She was treated with chemotherapy and craniospinal irradiation. Gradenigo's syndrome, as classically described, consists of suppurative otitis media, abducens nerve palsy, and trigeminal neuralgia due to infection of the petrous apex of the temporal bone. Once a common complication of acute otitis media prior to the availability of antimicrobial therapy, the now rare triad of Gradenigo's syndrome has been associated with noninfectious causes including trauma and malignancies. Regardless of underlying etiology, the symptoms require prompt evaluation and management. This case emphasizes the importance of maintaining a broad differential diagnosis in the setting of classic symptoms as well as the possibility of malignancy presenting with acute or subacute, rather than chronic, symptoms in the pediatric patient.

333 SEVERE VARIANT OF EPISPADIAS-EXSTROPHY COMPLEX: OEIS SYNDROME

S Drennan*, A Ruiz-Elizalde, A Rensing, L White. OUHSC, Oklahoma City, OK

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Introduction Epispadias-Exstrophy Complex is a spectrum of midline anomalies caused by embryological defects in multiple organ systems early in gestation. Clinical manifestations of the most severe form include omphalocele, exstrophy, imperforate anus, and spinal defects (OEIS). This syndrome is as rare as it is complex with incidence of 1 in 3 00 000 live births.

Case A 2340 g male was born to a 17 year old, Caucasian primigravida at 35 3/7 weeks for preterm labor. Fetal

ultrasound (US) showed omphalocele, bladder exstrophy, spinal defect, bifid penis, VSD. Infant required intubation at delivery and was transported to the NICU. Admit exam was consistent with prenatal US and also revealed widened cranial sutures, prominent occiput, low set ears, cloacal exstrophy with exposed hemibladders and cecal plate, anorectal malformation, ambiguous genitalia with non-palpable testes, skin covered lumbosacral defect, bilateral clubbed feet and hip laxity. Family history was positive for paternal aunt with abdominal wall defect. Genetic evaluation revealed 46 XY, SNP array pending. Cranial/Spinal US: left choroid plexus cyst, myelomeningocele. Echocardiogram: structurally normal heart, no VSD. Abdominal US: liver within omphalocele, spleen posterior to left kidney. Consults included pediatric surgery, urology, neurosurgery, orthopedic surgery, wound/ostomy care, and physical therapy. On day 2, infant underwent exploratory laparotomy, separation of GI from GU exstrophy, excision of hindgut duplication, creation of loop ileostomy, and approximation of bladder halves. After convalescence in NICU and extensive education for family, he was discharged home on day 29 with Amoxicillin for UTI prophylaxis, home health visits, and appropriate subspecialty follow-up.

Discussion OEIS Syndrome is a rare, but severe variant of Epispadias-Exstrophy Complex. Patients with OEIS are at risk for multiple physical and psychological morbidities (i. e. incontinence, UTI, impaired mobility, sexual dysfunction). Infants like the one in this vignette require multi-disciplinary care starting at birth, intricate staged operations, and life-long medical treatment. Planning should start early with prenatal consultations and delivery at a tertiary care center.

334 HYPERPHOSPHATEMIC FAMILIAL TUMORAL CALCINOSIS: CASE REPORT

VM Thomas*, M Blazek, V Parker, SS El-Dahr. *Tulane, Metairie, LA*

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Background Hyperphosphatemic familial tumoral calcinosis (HFTC) is a rare disorder characterized by hyperphosphatemia and abnormal deposits of phosphate and calcium in body tissues. Mutations in the fibroblast growth factor 23 (FGF23) gene, GALNT3 gene, or Klotho gene involved in phosphate homeostasis cause HFTC. These mutations disrupt the FGF23 signaling pathway through defective protein production or end-organ resistance causing decreased renal phosphate excretion. We report a rare case of HFTC associated with supra-physiological levels of FGF23.

Case A 7-year-old boy presented to clinic with a recurrent mass on his left elbow and history of surgical excision of a similar mass twice in the past two years, pathology diagnosed as calcinosis. The current recurrence was noted 2 months prior to clinic visit. The patient denies any pain to the elbow unless there is a forceful impact, and his family denies any visible swelling of the arm, fevers, recent trauma or fractures. No pertinent known family history. On examination, the left elbow showed a non-tender raised ulcerated lesion of size 2.5 cm with a smaller adjacent 1 cm hypopigmented lesion and mild soft tissue swelling. Healed surgical scar seen. Physical exam and vitals were otherwise unremarkable. Investigations showed elevated phosphorous of

8.6 mg/dL, vitamin D insufficiency (13 ng/mL), decreased hemoglobin of 10.7 g/dL, hematocrit of 34.3%, and MCH 24.2. Serum calcium, PTH and CRP normal. Urine phosphate 94 mg/dL. FGF23 was found to be 2050 (ref ≤ 230 RU/mL). Normal renal ultrasound without nephrocalcinosis. X-ray showed soft tissue calcifications lateral and posterior to distal humerus, with a negative skeletal metastatic survey. Surgical pathology of excised mass showed dense fibro-connective tissue with extensive dystrophic calcification and histiocytic reaction. Genetic testing was deferred due to patient's financial constraints. The patient was placed on a low phosphorous diet with phosphate-binder sevelamer.

Conclusion FGF23 acts on kidneys to cause phosphate loss and decrease in vitamin D synthesis. The patient had supra-physiologic levels of FGF23 with hyperphosphatemia, suggesting a mutation possibly in either GALNT3 or Klotho gene. Interestingly the patient had vitamin D insufficiency unlike most cases of HFTC with increased vitamin D levels.

335 FURTHER EVIDENCE SUPPORTING VPS8 AS A CANDIDATE GENE IN DEVELOPMENT OF CONGENITAL ARTHROGRYPOSIS

¹G Espiritu*, ²H Meddaugh, ^{1,2}R Zambrano. ¹Louisiana State University School of Medicine in New Orleans, New Orleans, LA; ²Children's Hospital New Orleans, New Orleans, LA

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Case report Arthrogryposis is a clinical term used to describe multiple joint contractures. It can be observed in association with other anomalies including micrognathia, shortened limbs, intrauterine growth restriction, lung hypoplasia, and short or immature gut. It is considered a feature of many disorders, with over 200 genes with different modes of inheritance implicated. However, the etiology of many cases remains unknown.

We report a patient with congenital arthrogryposis (CA), micrognathia, acute respiratory failure, microcephaly, congenital heart anomaly, and a complicated neonatal course with development of necrotizing enterocolitis. Initial Genetic testing included a SNP array that was normal. Whole exome sequencing identified variants of uncertain clinical significance (VUS) in several genes: ASXL1, KMT2D, LGI4 and VPS8.

Features on clinical evaluation are not consistent with a diagnosis of Bohring-Opitz syndrome (ASXL1) or Kabuki syndrome (KMT2D). Pathogenic variants in LGI4 are associated with a type of CA with myelin defect. However, this is a recessive disorder, so a single variant would not explain the CA in the proband. Three VUS in VPS8 were identified. VPS8 has been proposed as a candidate gene responsible for CA, and other VPS genes have also been implicated in disorders featuring CA.

The VPS, or vacuolar protein sorting genes, play a role in intracellular trafficking; they have been found to be involved in vesicular transport from early to recycling endosomes in mammals. Experiments involving depletion and knockout of VPS proteins show subsequent defects in integrin-dependent cell adhesion and spreading, cell migration, and regulation of cellular polarity. Disruptions in these molecular processes have been linked to development of disorders that involve CA.

The clinical and molecular findings in this case further support the proposed candidate gene status of VPS8 in relation to a CA phenotype.

336 INDOLENT MEDIASTINAL ABSCESS MASQUERADING AS LARGE RIGHT VENTRICULAR PSEUDOANEURYSM

¹W Fogarty*, ^{1,2}J Ettegui. ¹University of Florida College of Medicine Jacksonville, Jacksonville, FL; ²Wolfson Children's Hospital, Jacksonville, FL

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Case report Right ventricular pseudoaneurysm (RVPA) is a known, but rare complication of surgical repair of right ventricular outflow tract (RVOT) obstruction. This case aims to broaden the differential of the incidental finding of what appeared to be a large RVPA on transthoracic echocardiography (TTE) and subsequently on chest computed tomography (CT). On surgical intervention a large mediastinal abscess was found. The abscess was secondary to staphylococcus lugdunensis which is a rare but known cause of endocarditis, but not yet reported in this setting.

Our case describes a two-year-old female with Noonan's Syndrome who was diagnosed with multilevel RVOT obstruction at birth. She previously underwent successful RVOT reconstruction with partial pulmonary valvectomy and arterioplasty with transannular patch. During a routine evaluation, she was found to have a large RVPA on TTE. She was asymptomatic but was admitted to the CVICU for further evaluation and intervention. Chest CTA revealed a large RVPA with compression of the main pulmonary artery. She underwent surgical evacuation of a large mediastinal abscess cavity and was placed on broad spectrum IV antibiotics. Antibiotic coverage was narrowed per sensitivities to IV clindamycin when culture grew oxacillin resistant staphylococcus lugdunensis. Follow up chest CT on day of discharge demonstrated near resolution of the abscess. She was discharged home with a PICC to complete a 4 week course of IV clindamycin and an additional 4 week course of oral clindamycin.

Abstract 336 Figure 1

337 COMMUNICATING HYDROCEPHALUS IN AN INFANT WITH TUBERCULOSIS MENINGITIS

^{1,2}E Foreman*, ^{1,2}S Surapa Raju. ¹UAB, Birmingham, AL; ²Children's of Alabama, Birmingham, AL

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Case report An 8-month-old Hispanic female presented to the hospital with a 1 day history of vomiting and fever. She had a chest x-ray that showed a pneumonia and a normal head CT. She was admitted to the inpatient service for 6 days of IV antibiotics. Three days after discharge, she became sleepier and began having seizure activity. She returned to the hospital for a repeat head CT that showed communicating hydrocephalus. She was loaded with Keppra for status epilepticus and intubated. An emergent EVD was placed by neurosurgery. CSF indices from the EVD with 14 WBC, 128 RBC, glucose of 34, and protein of 50. These indices were concerning for meningitis so she was started on broad-spectrum antibiotics. A lumbar puncture was subsequently performed which showed 463 WBC, 23 RBC, glucose less than 20, and protein greater than 6000. Due to severely elevated protein, a workup for tuberculosis was initiated. Her PPD returned positive, and she was started on five-drug tuberculosis therapy. Brain MRI with findings of basilar meningeal enhancement in addition to multifocal areas of diffusion restriction. The patient's father was found to have a cavitary lesion on chest x-ray in addition to his sputum being smear positive for AFB therefore resulting in a diagnosis of tuberculosis meningitis. After control of seizures was obtained, she was able to be extubated and EVD removed after ICP normalized. She continued to have frequent episodes of agitation with an overall poor neurologic exam. She was discharged home to continue a prolonged course of tuberculosis medications. One month after discharge, she presented again with increased seizure frequency and was found to have worsening hydrocephalus on head CT requiring VP shunt placement.

Tuberculosis meningitis is a difficult diagnosis due to the non-specific nature of the symptoms and sometimes lack of definitive lab data. It is extremely less common compared to other bacterial or viral causes of meningitis especially in developed nations. It is important to keep tuberculosis meningitis on your differential especially since the morbidity and mortality associated with it are extremely high. The importance of taking a careful travel and family history are also paramount when tuberculosis is suspected in order to procure timely evaluation and treatment of family members or contacts.

338 SIROLIMUS: ITS ROLE IN THE TREATMENT OF GENERALIZED LYMPHANGIOMATOSIS IN AN ADOLESCENT MALE

CE Garcia-Canet*, U Narsinghani. Mercer University School of Medicine, Macon, GA

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Case report We report the case of a 17 year old African American male presenting with respiratory distress and hypoxia. A CT chest revealed a large pleural fluid collection and splenic and lytic bone lesions in the spine. He was admitted to the PICU for management including chest tube placement.

Infectious, autoimmune, and work up for malignancy were normal. A bone biopsy was inconclusive. The patient was noted to have a chylous effusion. Follow-up MR imaging showed a rounded mass-like density along the right hemidiaphragm that was suspicious for a lymphangioma in addition to the multiple bone and splenic lesions. Chylothorax in addition to these findings pointed to a rare, congenital anomaly called Generalized Lymphangiomatosis (GLA). He was started on Sirolimus with close monitoring of drug levels. Initially on total parental nutrition and lipids, his low fat diet was advanced as the drainage from the chest tube decreased. At time of discharge, the pleural effusion had resolved and Sirolimus levels were therapeutic. The patient was continued on Sirolimus and was advised to eat a low fat diet. Six months into therapy, he continues to steadily improve. Lymphangiomas are rare and, if left untreated, can impinge upon critical organs and lead to poor outcomes. As many as 65% arise during infancy and childhood. CT and MRI are the preferred imaging modalities and demonstrate large, multicystic fluid-filled masses. Presence of these masses in multiple organs substantiates the diagnosis of GLA. GLA is closely related to Gorham-Stout Disease, a condition that involves abnormal growth of lymphatic vessels in bone. In early childhood, GLA diagnoses are incidental findings of lytic lesions during a trauma evaluation. In puberty and adolescence, lytic lesions are associated with disease progression. The treatment for GLA varies. Traditional options include surgery, radiation, and medical therapies such as interferon, bisphosphonates, and cyclophosphamide. Some successes with targeting growth factors such as VEGF and PDGFR-beta have been reported. The use of Sirolimus, an inhibitor of T and B cell activation via reduction of sensitivity to IL-2 through the mTOR pathway, has shown promise as it has allowed for improved management for patients with poor response to the aforementioned options.

339 SEVERE ACUTE INTERSTITIAL NEPHRITIS SECONDARY TO MINOCYCLINE USE IN AN ADOLESCENT GIRL

N Geagan*, KP Sharma. *University of South Alabama Childrens and Womens, Mobile, AL*

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Introduction Acute interstitial nephritis (AIN) is defined as a decline in renal function due to inflammation and edema within the renal interstitium. We are reporting an adolescent girl with severe acute renal failure secondary to minocycline-induced acute interstitial nephritis.

Case report A 14-year-old African American female who was being treated with oral minocycline presented to the emergency department with a generalized erythematous rash and anasarca for 10 days. The patient was started on oral minocycline for acne 2 weeks prior. The patient was taking diphenhydramine for rash and pruritus for last 1 week with partial relief. At admission to the hospital, the patient had significant renal failure with azotemia, elevated blood urea nitrogen (75 mg/dL; reference 7–18 mg/dL) and high serum creatinine (12.9 mg/dL; reference 0.40–0.70 mg/dL). Renal ultrasound showed the presence of bilaterally enlarged, echogenic kidneys and percutaneous renal biopsy revealed the features of acute allergic interstitial nephritis confirming the diagnosis. Fluid resuscitation and steroid therapy led to the uneventful clinical recovery and renal function returned to normal within 1 week.

Conclusion Acute interstitial nephritis should be considered in patients with renal failure if minocycline has been used recently. These patients can present late as they may be asymptomatic in the early stage. Early steroid therapy is beneficial for prompt clinical recovery.

340 EOSINOPHILA, ECZEMA, EDEMA-OH MY!

J Ghassan*, N Zeky, MH Roy, J Albery, T McCormick. *Our Lady of the Lake, Baton Rouge, LA*

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Case report Eosinophilic colitis (EC), the rarest form of eosinophilic gastrointestinal disorders, is characterized by increased eosinophils in the colonic wall found on biopsy. It has a peak incidence in the third to fifth decade of life. EC is an IgE and non IgE mediated response that increases eosinophils in the colonic mucosa which results in an increase in cytokine release. The cytokines can lead to severe diarrhea, malabsorption and ultimately protein losing enteropathy (PLE). The most common food triggers are cow's milk, soy, egg, wheat and fish.

Our patient presented at 6 months of age with a history of worsening eczema and loose, non-bloody stools since three weeks of age. Physical exam was notable for anasarca, hair loss, and a diffuse, desquamating rash. Her growth chart revealed no weight gain for 2 months and downward change across two major growth percentiles. Laboratory evaluation on admission revealed hypoalbuminemia (1.2 g/dL) and an absolute eosinophil count of 5900/ μ L. Initial diagnostic considerations included an allergic disorder, nutritional deficiency, immunologic dysregulation and parasitic infection. Further work up revealed low IgG of 236 mg/dL and an elevated IgE of 4127 kU/L. The severity of her eczema was concerning for acrodermatitis enteropathica and her zinc level was low at 0.32 mcg/mL. Stool cultures and stool ova and parasite were negative. EGD was normal and colonoscopy was remarkable for mild edema. Pathology reported dramatically increased eosinophils in the colonic mucosa confirming a diagnosis of EC. She was treated with systemic steroids, zinc supplementation and exclusive elemental formula with significant improvement.

The mucosal injury noted in EC can lead to an increase in permeability of the colon, allowing for PLE. Our patient's alpha 1 anti-trypsin was found to be within normal limits (<12 mg/dL) which was attributed to a dilution effect from the profuse watery diarrhea. Serum zinc levels are typically low in patients with hypoalbuminemia; however in practice it is recommended that zinc should be supplemented. Although eczema and diarrhea are two common chief complaints, the presence of chronic diarrhea, edema and malnutrition should prompt pediatricians to initiate a work up for etiologies such as eosinophilic colitis which lead to both protein-losing enteropathy and nutrient deficiencies.

341 A CASE OF CORTICAL BLINDNESS IN AN INFANT WITH VIRAL MENINGOENCEPHALITIS

A Glinky*, N Godiwala. *LSUHSC, New Orleans, LA*

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Case report Acute visual loss is a rare complication of childhood intracranial illnesses and injuries. Causes of visual loss

include malignancy, trauma, inflammation, bacterial ocular infections, toxins and rarely, non-herpetic viral illness. Infants are unable to express themselves verbally, leaving visual changes largely unrecognized until an extreme change has occurred. The lack of expressive skills in this age group further impedes physicians' ability to track successful response to treatment.

We present a 5 month old male with no significant past medical history who presented with one day of increased sleepiness, fussiness, rhinorrhea, rightward gaze preference and erythematous papular rash over the trunk. Upon admission, the infant had two episodes of right upper extremity jerking, consistent with seizure-like activity, resolved with benzodiazepine administration. Laboratory work-up was significant for mildly elevated liver function tests and viral panel positive for adenovirus and rhino/enterovirus. Initial head CT was negative but MRI obtained the next day was concerning for viral meningoencephalitis. The child's hospital course was complicated by continued seizures, refractory to single drug therapy but ultimately controlled with levetiracetam and phenobarbital.

On hospital day three, the child had a perceived deterioration in vision. Ophthalmology was consulted and found the child to have reduced central vision, consistent with inflammation at the apices of the visual cortex. The patient completed three days of high dose methylprednisone therapy with some subjective improvement in his vision. The infant was transferred to Texas Children's Medical Center for further evaluation with pediatric neuro-ophthalmology where he was treated with intravenous immunoglobulin and continued to have subjective improvement in his vision.

Transient cortical blindness has been associated with several viral meningoencephalitides including herpes and mumps. However, our team was unable to find any other cases of rhino/enterovirus or adenovirus causing cortical blindness. Given the relative rarity of cases with this complication, treatment options are limited to steroids and IV immunoglobulin, as were used in this case. Close follow up with ophthalmology and neurology will reveal the longterm success of this treatment combination in this unique case.

342 GUILLAIN-BARRÉ SYNDROME COMPLICATING MENINGITIS

V Godsey*, C Rutledge. University of Alabama-Birmingham, Birmingham, AL

10.1136/jim-2018-000974.340

Case report *Neisseria meningitidis* (*N. meningitidis*) is the leading cause of bacterial meningitis in the United States. Despite vaccination attempts, mortality secondary to *N. meningitidis* is still approximately 15%. Presenting symptoms of *N. meningitidis* include fever, chills, myalgias, headache and meningismus. *N. meningitidis* rarely presents with areflexia. It is important to perform a complete physical examination, as well as, diagnostic studies including a lumbar puncture on patients presenting with symptoms of meningitis because there are a few case reports of Guillain-Barré syndrome complicating the clinical course of meningitis. The following is a case of a patient with a unique presentation of Guillain-Barré syndrome in the setting of *Neisseria meningitidis*.

A 13-year-old male presented with a two day history of sore throat, fever, vomiting, headache and progressively worsening lower extremity weakness. He required assistance to

stand and developed loss of sensation in his lower extremities. On examination he was alert and cooperative. He was unable to tightly close his eyes. He had a weak cough and a soft voice. He was able to shrug his shoulders but unable to lift his head off the bed. He was unable to move his lower extremities to painful stimuli. He had poor respiratory effort secondary to his neurologic symptoms prompting intubation for airway protection. An MRI brain and spinal cord was obtained and did not show any gross lesions. Given the patient's symptoms and viral prodrome, the initial concern was for Guillain-Barré syndrome (GBS); however, his cerebrospinal fluid was analyzed and showed 12 000 white blood cells, glucose <20 mg/dL and protein >700 mg/dL. The gram stain showed gram-negative cocci and gram-positive cocci. The culture grew *Neisseria meningitidis*. The patient was treated with ceftriaxone for a total of ten days. The patient continued to worsen neurologically and became areflexic in his extremities with continued respiratory muscle weakness. He was given intravenous immune globulin (IVIG) for treatment of GBS. After the first dose of IVIG he had significant neurologic and respiratory improvement and was successfully extubated.

343 EARLY MANDIBULAR DISTRACTION OSTEOGENESIS FOR MANAGEMENT OF SYMPTOMATIC MICROGNATHIA

B Google*, AB Google, I Hoppe. University of Mississippi Medical Center, Jackson, MS

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Case report Micrognathia is a congenital abnormality that can lead to both aesthetic and functional deficits. Frequently seen as a component of Pierre-Robin Sequence, which involves micrognathia, glossoptosis, and cleft palate, patients may struggle with respiratory obstruction and feeding difficulties. Appropriate referral for mandibular distraction osteogenesis may preclude the need for tracheostomy and/or gastrostomy and improve outcomes. We present a case of severe micrognathia in a premature infant at our institution managed with bilateral mandibular distraction osteogenesis (MDO).

We were consulted for a 2 month-old-male born at 28 weeks with severe micrognathia. He was intubated shortly after birth but eventually self-extubated. He was maintained on non-invasive airway support and was unable to be weaned to room air. Additionally, he was unable to tolerate oral feeds

Abstract 343 Figure 1 Severe micrognathia with class II malocclusion. Parent/Guardian consent obtained.

without associated bradycardia and desaturations. Polysomnography (PSG) revealed obstructive sleep apnea with an apnea-hypopnea index of 25/hr. It was decided the patient would benefit from bilateral MDO. Surgery was performed without complication, and he was distracted to a slight class III occlusion over 2 weeks. He was successfully weaned to room air and able to tolerate oral feeds without difficulty 2 weeks into consolidation. Repeat PSG revealed a normal breathing pattern.

Early mandibular distraction placement can significantly improve outcomes. Consultation to a pediatric craniofacial team should be an early consideration.

344 A MASS OF A DIFFERENT KIND: MASSIVE HEMOPERITONEUM FOLLOWING OVARIAN CYST RUPTURE

KB Grizzle*, C Wu. *Children's of Alabama, University of Alabama at Birmingham, Birmingham, AL*

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Case report 1. Consider the diagnosis of ruptured hemorrhagic ovarian cyst in a patient who presents with abdominal pain and radiographic evidence of intraperitoneal fluid. 2. Hemoperitoneum is a rare, but serious complication of ovarian cyst rupture.

Case presentation A 16 year-old female presented after a syncopal episode leading to a fall down the stairs. She had acute onset of abdominal pain, emesis, and fever just prior to the episode. Imaging was obtained to evaluate for injuries from her fall. Abdominal ultrasound disclosed abnormal soft tissue adjacent to the uterus and bilateral ovaries. CT abdomen and pelvis revealed ascites and a homogeneous mass in the adnexal region, concerning for malignancy. Oncology and Surgery were consulted to biopsy the mass and bone marrow. On laparoscopic visualization of the abdomen, her 'mass' was found to be a massive hemoperitoneum, which was evacuated. Inspection of the abdomen following evacuation was notable for a previously ruptured right ovarian cyst. She was transfused two units of blood due to intra-abdominal bleeding. Following evacuation of the hemoperitoneum and blood transfusion, she had symptomatic improvement with stabilization of her hematocrit and vitals.

Discussion The reproductive tract, specifically ovarian cyst rupture and ectopic pregnancy, is the most common source of spontaneous hemoperitoneum in post-pubertal females. While ovarian cyst hemorrhage is relatively common, subsequent hemoperitoneum is rare. Diagnosis is made with a combination of clinical symptoms and radiographic findings. These findings mimic that of many other disorders, therefore they have been called 'the great imitator'.

Cyst rupture is characterized by acute onset of lower abdominal pain and emesis, though may be asymptomatic. Fluid is usually seen in the area surrounding the uterus and adnexa. Imaging findings include a fluid filled mass, internal septations and echogenicity, air fluid levels, or a homogeneous mass. The cyst may or may not be visualized.

Patients who are hemodynamically stable may be managed conservatively with fluids and observation, including serial labs and vital sign trending. Laparoscopic evaluation and evacuation is required to control ongoing hemorrhage in unstable patients, or for further work up of etiology.

345 INFLUENZA AS A TRIGGER FOR VENTRICULAR ARRHYTHMIAS

Y Nathani, L Gunderson*, RM Berent, A Shah. *OUHSC, Oklahoma City, OK*

10.1136/jim-2018-000974.343

Case report Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an inherited channelopathy, characterized by ventricular tachyarrhythmias – bidirectional PVCs, ventricular tachycardia (VT), and ventricular fibrillation (VF), without cardiac structural abnormalities. Majority of cases are related to mutations in the ryanodine receptor 2 gene (RYR2). Common triggers include exercise and physiologic stressors. VT/VF storm is an uncommon presentation of CPVT. We report a case of CPVT due to a previously undescribed RYR2 mutation presenting with VT/VF storm triggered by Influenza B.

A 12 y.o healthy male presented with seizure-like episodes in the setting of cold symptoms. Holter monitor was placed after PVCs were noted on assessment and revealed frequent PVCs and slow monomorphic VT. Concurrent with Holter results, nasal swabs were positive for influenza B, leading to suspicion of myocarditis or cardiomyopathy. However, clinical exam and labs were reassuring and echocardiogram was normal. Due to Holter results in the presence of influenza, he was admitted for monitoring. Subsequently, he developed rapidly progressive ventricular arrhythmias resulting in pulseless VF arrest, requiring resuscitation with CPR and defibrillation. Despite anti-arrhythmics, he had another two VF events requiring CPR at progressively shorter time intervals indicating VT/VF storm. Repeat echo showed worsening biventricular dysfunction. Given rapid deterioration, patient was placed on ECMO, resulting in improved cardiac function. After 72 hours of ECMO, he remained in sinus rhythm with good control and was decannulated; 14 days later, he underwent ICD placement for secondary prevention prior to discharge.

Genomic DNA analysis found a previously unidentified point mutation of the RYR2 gene specifically c.12244G>C, resulting in a missense mutation from glutamic acid to glutamine, leading to a conformational change suspected to sensitize RYR2 to Ca²⁺ activation. Influenza is known to change intracellular calcium concentration during viral entry through plasma membranes of cardiomyocytes. We speculate that this triggered the latent CPVT, resulting in VT/VF storm. This report describes a new viral trigger of CPVT, as well as the first description of this specific mutation.

346 MAYBE IT'S IN HER HEAD, OR HER PELVIS – A 15YO FEMALE WITH ANTI-NMDA RECEPTOR ENCEPHALITIS & OVARIAN TERATOMA

K Gutermuth*, A Sorrentino. *University of Alabama at Birmingham, Birmingham, AL*

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Introduction Anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis occurs when the immune system generates antibodies against the central NMDA-type glutamate receptors. As a result, a wide variety of psychiatric and systemic symptoms can manifest, often making diagnosis difficult, and if left untreated can be lethal. We present the case of a 15yoF presenting with neurologic and psychiatric symptoms who was found to have NMDAR with concurrent ovarian teratoma.

Case A 15yo AAF with history of recent meningitis, was re-hospitalized due to altered mental status (AMS). At an OSH, she was noted to be tachycardic, hypertensive, febrile, agitated and hallucinating. She was diagnosed with a UTI and admitted on empiric antibiotics. After negative cultures, she remained altered and agitated despite multiple doses of antipsychotics. She was transferred to an inpatient psychiatric facility and was refused admittance due to her AMS. She was referred to the pediatric ED, where she was tachycardic, hypertensive, and febrile. Upon examination, she was encephalopathic, had rhythmic lip smacking, depressed mental status, and rigidity. She was empirically received broad spectrum antibiotics, and fluid resuscitation, and was admitted to the PICU for further management. CT Head showed leptomeningeal enhancement, and her CSF had 22 lymphocytes but normal glucose and protein. LTEEG showed no seizure activity and infectious workup was negative. She had a pelvic U/S and MRI which showed right ovarian teratoma, and was resected shortly after. Her pathology revealed a mature ovarian teratoma without evidence of neoplastic process. Her CSF studies concluded the diagnosis of NMDAR encephalitis.

Discussion Anti-NMDAR encephalitis is becoming an increasingly common diagnosis, that often presents with similar signs and symptoms as this patient. There is a female predominance with a median age of 19 years. In female patients older than 18 years, the frequency of an underlying ovarian teratoma is nearly 60%. In addition, African American women are more likely to have an underlying ovarian teratoma than patients of other ethnic groups. Consider NMDAR encephalitis in patients with a viral-like prodrome, followed by a period of psychiatric symptoms, especially in young female patients.

347 TAKE MY BREATH AWAY: PULMONARY MANIFESTATIONS OF RF+ POLYARTICULAR JIA

¹EN Bufkin, ²K Gutermuth*. ¹University of Alabama at Birmingham, Birmingham, Afghanistan; ²University of Alabama at Birmingham, Birmingham, AL

10.1136/jim-2018-000974.345

Case report A 12 yo AAM presented with 5 day history of fever, malaise, cough, and shortness of breath. Past history notable for intermittent cough-variant asthma and polyarticular juvenile idiopathic arthritis which was diagnosed 5 months prior to presentation. He was maintained on MTX before being transitioned to prednisone due to elevated liver enzymes. Upon presentation, he was ill-appearing and hypoxic to 87% on room air. Exam showed respiratory distress, diffuse rales throughout, and digital clubbing. Initial lab work notable for arterial blood gas of PaO₂ of 76 on 35% FiO₂. CXR in the ED revealed diffuse mixed interstitial alveolar pattern. CT Chest without contrast showed diffuse ground glass and reticular interstitial pattern throughout both lung fields. He underwent bronchoscopy with bronchial alveolar lavage (BAL) early in his hospitalization which revealed alveolar disease, and studies were notable for neutrophilic predominance and *Mycoplasma pneumoniae*. BAL pathology revealed mucoid debris with rare extracellular bacteria, negative fungi/mycobacteria/pneumocystis. Additional work-up revealed a markedly elevated cyclic citrullinated peptide antibody and rheumatoid factor. Patient was diagnosed with pulmonary manifestations of RF +JIA and was treated with pulse-dose steroids, rituximab, and cyclophosphamide.

Discussion RF+polyarticular JIA is the equivalent of adult-type rheumatoid arthritis and progresses aggressively. It is diagnosed with positive tests for RF and anti-CPP antibodies in the setting of polyarticular joint involvement. Interstitial lung disease is the most common pulmonary manifestation of rheumatoid disease.

Interstitial lung disease (ILD) is rare in children and tends to cluster in infancy (~50% of cases). Due to its rarity, ILD diagnosis can be delayed for months to years. As with our patient, most clinicians misdiagnose the patient's presenting symptoms as asthma. Common presenting symptoms include tachypnea, nonproductive cough, and failure to thrive/weight loss due to chronic inflammation. This case of ILD in RF +polyarticular JIA may increase awareness of this disease, expand our understanding of disease presentation, and therefore, assist in earlier clinical suspicion, diagnosis, and potential treatment.

348 GLOBICATELLA SPP. AS THE CAUSE OF NEONATAL MENINGITIS

J Guzman*, K Martin. University of Oklahoma School of Community Medicine, Tulsa, OK

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Case report While the most common cause of neonatal bacterial meningitis in the US is group B Streptococcus (GBS), other potential pathogens include the viridans streptococci. *Globicatella* spp are phenotypically similar to the viridans streptococci. One of its types, *G. sulfidifaciens* has only previously been reported in humans once before. Given phenotypic similarities to more common organisms and the possibility of their misidentification, these organisms should be considered possible causes of neonatal meningitis when unusual viridans streptococci are isolated.

We present a 5 day old female born by uncomplicated vaginal delivery presenting with fever. Physical exam demonstrated an irritable infant with a bulging fontanelle. Routine sepsis evaluation including blood culture, lumbar puncture, and urine culture revealed leukocytosis to 19,400 WBC/mL, CSF WBC count of 9,888/mm³, CSF protein of 275.6 mg/dL, and CSF glucose of <1 mg/dL. Both CSF and blood cultures identified *G. sulfidifaciens* by Vitek Gram-positive identification card, prompting confirmation by matrix-assisted laser desorption/ionization time-of-flight (MALDI-TOF). MALDI-TOF identified *G. sulfidifaciens*. Relatively increased mean inhibitory concentrations to 3rd generation cephalosporins led to treatment with intravenous ampicillin, and the infant's illness resolved without apparent sequelae.

This case of meningitis due to *Globicatella* spp. is remarkable as one of only two cases due to this genus among neonates reported in the literature. While our isolate was identified as *G. sulfidifaciens*, the novelty of these organisms and variability in isolates since their discovery means that newer routine identification methods used in the clinical laboratory may be insufficient to discriminate between species. This case highlights the need to consider unusual organisms as potential causes of meningitis, as this may have important therapeutic implications.

Neonatal meningitis due to *Globicatella* spp. is a rare entity. Its detection in a healthy infant demonstrates its capability of producing disease in immunocompetent children. While uncommon, it should be considered when viridans-like streptococci are isolated in clinical specimens.

349 STREPTOCOCCAL TOXIC SHOCK SYNDROME AFTER STREP THROAT

^{1,2}WC Hancock*, ^{1,2}S Gangu, ^{1,2}B Bagga. ¹University of Tennessee Health Science Center Memphis, Memphis, TN; ²Le Bonheur Children's Hospital, Memphis, TN

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Case report A 5-year-old nonverbal male with multiple comorbidities presented to an outlying ED with a brief history of fever, halitosis, and drooling. Chest x-ray, rapid flu, and Strep screens were negative. A throat culture was sent prior to discharge with a diagnosis of viral syndrome. The patient returned in 2 days with worsening fevers and lethargy. Notable lab findings included elevated troponin I, lactic acidosis, leukocytosis, elevated inflammatory markers, and renal insufficiency all concerning for severe sepsis. This prompted initiation of broad spectrum antimicrobials and prompt transfer to our hospital, where he was found to be in cardiorespiratory failure requiring urgent intubation, fluid resuscitation, and pressors. He rapidly deteriorated developing fulminant multi-organ failure necessitating veno-arterial extracorporeal membrane oxygenation support and continuous renal replacement therapy. He was empirically started on vancomycin, cef-tazidime, and clindamycin. Culture from his throat swab returned positive for 2+ *Streptococcus pyogenes* (GAS) soon after admission to the ICU, raising suspicion that the clinical picture was likely due to Streptococcal toxic shock syndrome (STSS). GAS was isolated from the subsequent blood culture with a time to positivity of 5.7 hours. Antimicrobials were narrowed to a two-week course of penicillin and clindamycin to treat STSS. Neck imaging was negative for deep neck abscesses. Our patient had full recovery prior to discharge. Our case makes many illustrative points for clinicians. It reiterates that the rapid strep test can be falsely negative and highlights the importance of sending reflex throat cultures. This case also highlights the importance of empiric anti-streptococcal therapy when the initial symptoms are suggestive of strep throat even if the rapid test is negative. Thirdly, the classic symptom of throat pain may not be obvious in a non-verbal child such as our patient, thus it is important to maintain a high degree of suspicion if there are other signs such as halitosis, drooling, or refusal to eat or drink. Lastly, although STSS typically has a soft tissue source, our report presents a unique case of pharyngeal Strep as the source for toxic shock.

350 LATE ONSET NEONATAL ALLOIMMUNE THROMBOCYTOPENIA IN AN INFANT PRESENTING WITH VIRAL MENINGITIS

MV Hongo*, O Sanchez. University of South Alabama Health System, Mobile, AL

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Introduction Neonatal alloimmune thrombocytopenia (NAIT) is the most common cause of thrombocytopenia typically presenting in the first 24–72 hours of life. Mild cases only include thrombocytopenia. Severe cases are denoted by the presence of intracranial hemorrhage. Screening for NAIT is not routine and often only discovered in symptomatic patients.

Case description A 7 day old infant was admitted to the pediatric intensive care unit after presenting to the ED with rectal temperature of 100.7F. He was also noted to have 2 small bruises at this time and septic workup revealed

thrombocytopenia without any decrease in other cell lines. Patient was given empiric antibiotics and platelet transfusion. Patient was transferred to the PICU where he was given a second platelet transfusion and lumbar puncture was obtained. Antiplatelet antibodies returned positive and patient received 2 more platelet transfusions and IVIG. Patient was diagnosed with enterovirus meningitis and antibiotics were discontinued 2 days later when blood and cerebrospinal fluid cultures returned negative. After treatment with IVIG platelets rose. At this time a diagnosis of immune thrombocytopenic purpura (ITP) was made. Fever resolved and patient was discharged home to follow with pediatric hematology. Hematology suspected NAIT, as ITP is extremely rare in neonates, and obtained antiplatelet antibodies from mother. Platelets continued to remain in normal range at subsequent visits and maternal antiplatelet antibodies returned positive indicating NAIT. Of note patient had normal platelet count at time of birth. Mother and siblings have no history of thrombocytopenia. All of mother's children have the same father. Mother and older sister had been ill with symptoms consistent with enterovirus infection at time of patient's delivery.

Discussion NAIT typically presents prenatally or within the first 72 hours of life with marked thrombocytopenia. In this case the patient presented at 7 days old in the face of a viral meningitis, indicating either very late sensitization of mother to platelet antigens or viral induction of antiplatelet antibodies. Viral induction of ITP is a known phenomenon, but has not been thoroughly investigated in NAIT.

351 CASE OF A VIGOROUS BLUE INFANT

^{1,2}A Iacob*, ¹A Antonio. ¹Miller Children's and Women's Hospital, Long Beach, CA; ²University of California Irvine, Orange, CA

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Case report A 22 day old term female presented with severe dehydration to an outside hospital, and was found to appear 'blue' and with severe metabolic acidosis. History revealed that she was born at 40 weeks gestation by Cesarean section due to nuchal cord and had appropriate weight, length and head circumference. The delivery was uneventful and infant went home at 3 days of life. At 20 days of life, the infant developed projectile vomiting with diarrhea and was taken to an emergency room. Triage vital signs were normal. Abdominal ultrasound ruled out pyloric stenosis. Mother and infant then left without formal discharge. The infant continued to have daily non-bilious, non-bloody emesis with watery, non-bloody, non-malodorous diarrhea. At 22 days of life, mother took the infant to the hospital. Physical exam revealed an infant with normal vital signs, ashy gray/blue in color, and good tone with appropriate reflexes. Initial workup included normal hemoglobin/hematocrit, no evidence of bandemia nor leukocytosis. A capillary gas in room air revealed a pH of 6.87, PCO₂ of 38 mmHg, bicarbonate of 7 mEq/L (7 mmol/L), base deficit of -26, and 7.1% methemoglobin. The infant was transferred to the Neonatal Intensive Care Unit and upon arrival her methemoglobinemia had increased to 19%. She immediately received two doses of methylene blue 0.1 mL/kg with improvement in color and significant drop to 4% methemoglobin. Further investigation included a sepsis rule out by blood culture and cerebrospinal fluid culture, chest X-ray imaging, glucose-6-phosphate dehydrogenase testing, plasma

amino acids and urine organic acids. Her methemoglobin percentage was trended with the Masimo rainbow SET[®], a non-invasive monitoring device that features co-oximetry. This allowed for quick interventions without repeated venipunctures. The infant's exam and blood gas values quickly improved with appropriate hydration. Her symptoms of diarrhea and emesis also resolved. She was started on breastmilk which she tolerated well and was discharged the next day. This case illustrates the presentation of methemoglobinemia in the neonate, the necessary differential evaluation, a review of the management of methemoglobinemia, and describes the use of new technology to evaluate the progression and/or resolution of methemoglobinemia.

352 ATYPICAL KAWASAKI DISEASE PRESENTING AS ABDOMINAL DISTENSION AND SEVERE DEHYDRATION

C James*, C Gregg, A Prudhomme. *LSUHSC, New Orleans, LA*

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Case report Kawasaki Disease is a multi-system vasculitis of childhood and the leading cause of acquired heart disease in developed countries. Infants, particularly those less than 3 months, often present with an incomplete form carrying increased risk for cardiac manifestations that is often refractory to treatment. A high index of suspicion is necessary in patients with several days of high fever and no clear etiology.

We present the case of a 3 month old male with severe dehydration, high fever, and cracked lips for 3 days. Initially admitted to PICU, he underwent full infectious work up with lumbar puncture, positive only for nasopharyngeal rhinovirus/enterovirus. On fever day 5, he developed abdominal distension with rigidity and hypoactive bowel sounds, concerning for obstructive vs infectious process. Significant time was dedicated to the evaluation of the abdomen, including imaging, cultures, and the initiation of antibiotics. Abdominal x-ray showed paucity of gas, and inflammatory markers were elevated. Work up was otherwise unrevealing while fevers and distention continued. Patient underwent echocardiogram on day 7 of fevers, which showed coronary arteries at the upper limit of normal. Patient was started on IVIG and then subsequently developed polymorphous rash, pedal edema, and conjunctivitis. With the diagnosis of Kawasaki Disease, aspirin was initiated. Recurrent fevers led to a second dose of IVIG and ultimately steroids, after which he defervesced. Repeat echocardiograms revealed progressive, severe ectasia of coronary arteries, and dual antiplatelet therapy was begun.

Prolonged course of high fevers with unknown etiology in all children should lead to a high suspicion for Kawasaki disease. Infants younger than 3 months are at particularly high risk for atypical presentation with serious sequelae, including more severe coronary artery dilatation and disease refractory to initial treatment. As with our patient, abdominal involvement can lead to a delayed diagnosis; however, a growing body of evidence supports diverse abdominal findings at initial presentation, including pseudo-obstruction, surgical abdomen, and pancreatitis. Because classic clinical features may not be present simultaneously, a careful history and close physical exam monitoring are necessary in patients who lack a clear source of a fever.

353 A DIGEORGE OF A DIFFERENT COLOR

K Jeffries*, EO Schmit. *University of Alabama, Birmingham, AL*

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Introduction DiGeorge syndrome (DGS) is typically caused by deletion of region on chromosome 22q11.2. However, this case demonstrates the variable DGS phenotype due to an alternative genetic deletion.

Case The case patient is a 5 month old female full-term infant, previously hospitalized after birth due to meconium aspiration requiring intubation for 2 weeks, who presents to the ED with decreased oral intake and fever. Labs were concerning for acute kidney injury with creatinine of 0.8 and leukocytosis of 42 K with left shift. Urine culture and blood cultures quickly grew gram negative rods. Additionally, there was concern for low iCa of 0.8. She was noted have dysmorphic features with frontal bossing, low set rotated ears, small mouth, wide spaced nipples, syndactyly of fingers, decreased general tone and systolic ejection murmur. Her height and weight plotted only at the 50th percentile for a one month old. Mother noted that patient's aunt had 'chromosome 10 abnormality.' Outside records demonstrated a karyotype with deletion of 10 p12.2 region. FISH demonstrated unbalanced translocation p and q arms of chromosome 10. She required gastrostomy tube due to feeding dysfunction. She has subsequently had two additional febrile UTIs (including ESBL positive *Klebsiella*), now on prophylactic antibiotics due to grade 3 reflux. Endocrine monitors her calcium levels on calcium carbonate and calcitriol. T-cells were moderately decreased for age and echocardiogram showed small PDA. She was also noted to have stenotic ear canals with bilateral sensorineural hearing loss. At 13 months old, she remains developmentally delayed receiving multiple therapies.

Discussion DGS affects an estimated 1 in 4000 people and is frequently associated with deletion of 22q11.2 region. However, there is rarely an association with a 10 p monosomic terminal deletion, which contains the HDR locus and DGCR2. The HDR phenotype includes Hypoparathyroidism, Deafness and Renal anomalies while DGCR2 is known as DiGeorge critical region 2, which can present as variable clinical symptoms of DGS. Common clinical features of DGS include cardiac anomalies, immunodeficiency and hypocalcemia. Many patients also have developmental delays.

Conclusion Patients with 10 p terminal deletions can present with variable symptoms of DGS, which can prolong the diagnosis and treatment of the condition.

354 WHEN CELIAC MET CAMPYLOBACTER

¹BL Johnson*, ^{1,2}R Kassel. ¹University of Alabama School of Medicine, Madison, AL; ²Children's of Alabama, Birmingham, AL

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Case report A 17-month-old male was transferred from an outside hospital (OSH) for hypokalemia and non-anion gap metabolic acidosis secondary to acute on chronic diarrhea. He tested positive for *Campylobacter* at the OSH. In addition, he had recent positive celiac serology, but had not yet undergone confirmatory endoscopy. On admission, the child appeared sleepy and chronically ill with minimal subcutaneous fat,

distended abdomen, and 3+oedema of hands and feet. Labs: potassium 2.7, bicarb 15, albumin 1.7. Weight was 34th percentile and length, 18th percentile. After becoming NPO, he received 1 mL/kg albumin, as well as azithromycin for *Campylobacter* enteritis. Later the day of admission, the patient developed hyponatremia and hyperchloremia.

EGD on hospital day #2 revealed globally denuded duodenum, at which point a gluten-free diet was started. By this time, diarrhea had resolved and electrolytes were normalizing. Edema also improved. The patient began eating voraciously and, by hospital day #5, had begun to gain weight. Biopsy results confirmed celiac disease (CD).

However, on hospital day #6, the patient again developed diarrhea, emesis, and electrolyte abnormalities. Since his GI tract could not tolerate sufficient enteral nutrition, the patient began total parenteral nutrition (TPN). Thereafter, electrolyte abnormalities normalized. After discharge home on TPN, the patient was gradually weaned to oral nutrition. By 2 months after discharge, he tolerated an oral gluten-free diet without diarrhea and had gained 4.5 kg.

Teaching Points Although CD usually presents chronically, the rare patient develops celiac crisis, an acute, life-threatening condition characterized by severe diarrhea, hypoalbuminemia, and resulting metabolic abnormalities. Celiac crisis remains poorly studied. Case reports have described gastrointestinal infection precipitating celiac crisis.

As with more typical presentations of CD, a gluten-free diet remains the mainstay of management. Many patients also require IV fluid support and electrolyte replacement. However, a minority of celiac crisis patients do not respond to these interventions. Clinical trials are lacking, but multiple case reports describe dramatic improvement with corticosteroids. Occasionally, patients may not tolerate oral feeds and require parenteral nutrition.

355 SEVERE CASE OF HEMOLYTIC UREMIC SYNDROME IN A PEDIATRIC PATIENT

S Karim*, DH McVadon, D Gilliam. University of Mississippi Medical Center, Jackson, MS

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Background Hemolytic Uremic Syndrome (HUS) is a microangiopathic hemolytic anemia that can cause non-immune thrombocytopenia, hemolytic anemia, and organ dysfunction, particularly acute kidney injury (AKI) in pediatric patients. The most common cause in this population is *Escherichia coli* O157:H7 strain or other Shiga toxin-producing organisms that can initially present as abdominal pain with emesis and bloody diarrhea. Although AKI is the most common complication of HUS, other complications include bowel perforation, pancreatitis, thrombosis, and cardiac dysfunction, leading to severe cases of this disease. We present a unique case of HUS in which the patient subsequently developed severe complications including AKI requiring dialysis, bowel perforation, pericardial thrombosis and pancreatitis.

Case description A previously healthy 3-year-old white male initially presented to the hospital with bloody diarrhea and non-bilious emesis found to Enterohemorrhagic *Escherichia coli*-gastroenteritis and HUS complicated by hypertension and AKI requiring hemodialysis (HD). Three days after discharge and three weeks after initial presentation, he returned to emergency room with increasing abdominal pain and distention and well as fever. Abdominal CT revealed bowel

perforation and pelvic abscess. This prompted exploratory laparotomy and segmental sigmoid colectomy followed by a colostomy was performed. Abscess cultures were positive for *E. coli* and *Enterococcus Faecalis*; blood cultures positive for *Fusobacterium nucleatum*. He completed a course of ampicillin/sulbactam. Echocardiogram was obtained and revealed thrombosis initially believed to be in the main pulmonary artery and patient was started on heparin; on repeat imaging (CTA heart) thrombosis found to be in the pericardial sac. Heparin was stopped. His admission was then further complicated by hematemesis, tachycardia, and elevated lipase levels that prompted a CT abdomen, demonstrating pancreatitis and a hematoma in the left lower quadrant as well as a peripancreatic hematoma. Despite these complications, the patient's abdominal symptoms improved with an abdominal ascites drain as well as hemodialysis.

Conclusion This case report demonstrates the severity of symptoms and complications that can occur in a case of typical HUS.

356 A CASE OF NEONATAL HERPES IN THE SETTING OF MATERNAL HERPES MASTITIS

TC Kingdon*, J Aita-Levy. Childrens Hospital New Orleans, New Orleans, LA

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Case report Neonatal infection with Herpes Simplex Virus (HSV) occurs in approximately 1 out of every 3,200–10 000 live births. HSV mastitis is an extremely rare condition and has only been documented in a few case reports. Here we present a case of neonatal infection with HSV transmitted from a mother with HSV mastitis.

A 2-week-old female presented to Children's Hospital, New Orleans ED with low-grade fevers, blisters throughout the oral cavity, and poor feeding. Our initial differential diagnosis included: hand, foot, and mouth disease, Staphylococcus scalded skin syndrome, or a TORCH infection. The patient was stable at presentation and underwent a work-up that included HSV blood PCR, HSV lesion PCR, and wound viral culture. Patient was admitted and empirically started on acyclovir. Her basic labs were unremarkable but her CSF revealed a low glucose, an elevated protein and a lymphocytosis. The viral culture, HSV PCR of the lesions, and HSV PCR of the CSF all were positive for HSV-1. Further history obtained from the mother revealed she never had genital lesions, no household members had a history of herpetic lesions, and there were no complications at birth. However, the patient's

Abstract 356 Figure 1 Lesions present on patient's tongue and buccal mucosa, highlighted by arrows

mother did reveal she had lesions on both of her nipples and was diagnosed and treated for mastitis one week prior to patient's presentation. A culture of her nipple lesions yielded a positive HSV PCR. The rarity of herpes mastitis makes this case unique and mothers should be questioned regarding a history of mastitis or nipple lesions whenever neonatal or infantile herpes is considered as a possible diagnosis in a patient.

357 KAWASAKI'S DISEASE VS. ROCKY MOUNTAIN SPOTTED FEVER IN A 15 MONTH OLD

R Klinger*, C Poole. *University of Alabama in Birmingham, Vestavia, AL*

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Introduction Rocky Mountain Spotted Fever (RMSF) is a tick-borne illness that causes a systemic vasculitis. It is classically characterized by fever, rash, headache. In the adult, the differential becomes narrow but in the pediatric population this constellation of symptoms can be seen in many conditions ranging from meningitis to Kawasaki's disease to viral exanthem. The struggle becomes even more challenging when the child is too young to vocalize his/her symptoms and pains and the parental history, physical exam, and labs become more important than ever.

Case report We report a case of 15 month old male from rural Alabama who presented with fever, full body maculopapular rash, conjunctivitis, white buccal lesions and fussiness for 5 days. He was diagnosed with Kawasaki's Disease based on clinical criteria and was subsequently treated with 2 courses of IVIG and Rituximab with no response. Rheumatology was consulted and concerned he had developed macrophage activation syndrome (MAS) and as a consequence he was treated with steroids and Anakinra (an interleukin-1 receptor antagonist). However, he failed to respond and on review of history with his family a tick exposure was mentioned. Serology for RMSF was obtained, and he was started on Doxycycline. Serologies later returned positive and the diagnosis was confirmed. In an illness where untreated fatality rates range from 20%–80% and median time to death is 8 days, our patient had antibiotic therapy initiated on day 9 of illness. He fortunately survived but not without sustaining neurological deficits for the foreseeable future.

Conclusion Febrile illnesses associated with rashes are common presentations in pediatrics with the vast majority due to viral infections. The diagnostic challenge is to recognize the features of the much rarer conditions that require prompt management to avoid negative sequelae. Kawasaki's Disease is a clinical diagnosis and should be considered a diagnosis of exclusion as there are many other conditions that can meet the diagnostic criteria for Kawasaki's Disease as was the case of our patient. RMSF should be considered on the differential of patients presenting with fever in endemic areas regardless of a history of known tick exposure.

358 RECURRENT HYPOGLYCEMIA IN A THREE YEAR OLD GIRL

¹R Koothirezhi*, ¹B Beard, ¹E Luther, ²R McVie, ¹C Murugesan. ¹LSU Health Science Center-Shreveport/Monroe, Monroe, LA; ²LSU Health Science Center-Shreveport, Shreveport, LA

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Introduction Timely identification and treatment is crucial in children presenting with recurrent hypoglycemia to prevent adverse neurodevelopmental consequences. We report an uncommon cause of recurrent hypoglycemia in a child.

Case report A 3 year old girl presented to ER with jitteriness, loss of balance and seizure like activity. Mother denied fever, diarrhea, decreased activity, appetite changes or rash. Physical examination was unremarkable with stable vital signs and reassuring neurological findings. Her blood glucose was low at 55 mg/dl (she received sugary liquids on the way to the ER). Her symptoms responded to IV glucose. She had prior similar episodes treated with glucose. Initial labs revealed unremarkable CBC, mild metabolic acidosis and elevated serum ammonia (73 umol/L). Insulin level was 6.2 uU/ml (elevated in the setting of hypoglycemia). Differential diagnosis of congenital hyperinsulinism, glycogen storage disorder or fatty acid oxidation defects were considered. Detailed metabolic work up was inconclusive. Further on, genetic study revealed a novel variant of GLUD1 mutation (p. Glu349Gly) which is an uncommon cause of congenital hyperinsulinism. It should be noted that child ate chicken thighs few hours before the presentation that had raised question about protein induced hypoglycemia. A protein tolerance test was performed which was positive. After confirming the diagnosis, she was started on Diazoxide and a diet with carb to protein ratio of 4:1 at each meal/snack, with no further reported episodes of hypoglycemia.

Discussion GLUD1 gene mutation is an uncommon cause of congenital hyperinsulinism associated with asymptomatic hyperammonemia and recurrent episodes of profound hypoglycemia precipitated by fasting or protein rich meals. High index of suspicion is required to diagnose this condition when patients present with otherwise unexplained hypoglycemia. Early diagnosis and management are essential to avoid adverse neurodevelopmental consequences associated with recurrent hypoglycemia. Our patient had a heterozygous variant mutation of GLUD1 gene which, to our knowledge, was never reported before.

359 ANOMALOUS LEFT CORONARY ARTERY ARISING FROM THE NON-CORONARY CUSP

¹CP Larmue*, ^{1,2}K Parikh, ^{1,2}L Haddad. ¹Our Lady of the Lake Children's Hospital, Baton Rouge, LA; ²Pediatric Cardiology Associates of Baton Rouge, Baton Rouge, LA

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Case report Congenital anomalies of the coronary arteries are defined as variations in coronary anatomy that occur in <1% of the population. Many of these variations are of no clinical significance. However, some forms have been associated with serious clinical consequences secondary to ischemia. In fact, congenital coronary artery anomalies are the second most common cause of SCD among young athletes. There are several risk factors associated with the development of ischemia, including the origin of a coronary artery and its course.

Common anomalous sites of origin of the left coronary artery (LCA) associated with ischemia include the right coronary cusp and the pulmonary artery. Origin of the LCA from the noncoronary sinus of Valsalva, however, is an extremely rare and was previously suggested to be benign. However, subsequent case reports have described life-threatening arrhythmias and SCD associated with this condition.

This is a case report of a 15 year old female who presented with chest pain and syncope while running in a track meet. A treadmill stress test elicited nonspecific ST changes with J-point depression in early recovery. Anomalous LCA from the non-coronary cusp was subsequently demonstrated on cardiac CT. She was then sent to Boston Children's Hospital where she underwent marsupialization of the anomalous LCA. Stress test at 3 months post-op was normal.

Discussion The patient above experienced symptoms and ECG changes suggestive of cardiac ischemia resulting from an anomalous LCA originating from the noncoronary cusp. If left untreated, this ischemia ultimately could have led to significant morbidity and possibly mortality in this patient. However, due to prompt identification and surgical treatment, the patient's symptoms and ECG abnormalities appeared to have resolved.

Conclusion Although previously thought to be benign, several cases reports have associated significant morbidity and mortality with origin of the LCA from the noncoronary cusp. Although rare, morbidity and mortality may be preventable with surgical treatment in many of these cases. Continued reporting of such cases is essential to further understanding and to develop evidence-based guidelines concerning management of these lesions.

360 RASBURICASE INDUCED METHEMOGLOBINEMIA IN LESCH-NYHAN DISEASE AND PREVIOUSLY UNRECOGNIZED GLUCOSE-6-PHOSPHATE DEHYDROGENASE DEFICIENCY

M Leibman*, R Natarajan, S Arsenegas. UNLV, Las Vegas, NV

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Case report 18-month-old non-verbal, non-ambulatory developmentally delayed African-American male with history of bilateral uric acid nephrolithiasis, irritability and self-mutilating behavior presenting with a one-month history of lower lip swelling and bleeding and one-day history of tactile fevers, cough and congestion. Patient was admitted to the pediatric floor for antibiotic treatment of lip cellulitis.

Patient was previously suspected to have Lesch-Nyhan Disease (LND) as his older brother has confirmed LND. He was prescribed citrate and allopurinol by nephrology, but mother denied medication compliance. On admission, patient was started on maintenance Allopurinol and received a dose of Rasburicase due to a uric acid of 8.9 mg/dl. Uric acid improved to 0.1 mg/dl the next day, but patient became hypoxemic to 85% despite 100% oxygen. He continued to have low saturations while sleeping calmly with no clinical signs of increased work of breathing. He was found to be RSV positive and CXR showed peribronchial cuffing. VBG showed adequate ventilation but methemoglobin level was 6.8%. Patient was also noted to have hemolytic anemia manifested by a hemoglobin of 6.7 g/dl, bilirubin of 3.3 mg/dl and reticulocytosis. Subsequently G6PD enzyme levels, drawn before transfusion, were low leading to a new diagnosis of G6PD deficiency. Additionally, a formal diagnosis of LND was eventually confirmed by positive Hypoxanthine-Guanine Phosphoribosyl Transferase (HPRT) sequence analysis sent during the admission.

Discussion This case highlights that methemoglobinemia induced hemolytic anemia is an important complication of rasburicase with potentially severe consequences. An asymptomatic drop in oxygen saturation by oximetry within 24 hours of rasburicase administration, with or without cyanosis, should

alert the clinician to the probability of methemoglobinemia and in turn test for G6PD deficiency. Interestingly, methylene blue should be avoided in G6PD deficiency as it may exacerbate oxidative hemolysis. Only a few other case reports have highlighted G6PD deficiency in the setting of LND. Further studies are needed to see if all LND patients, or those with specific risk factors, would benefit from G6PD enzyme screening prior to rasburicase administration.

361 INFANTILE SPASM IN A 7-MONTH-OLD RIGHT-HANDED INFANT

M Li*. UF Health Jacksonville, Jacksonville, FL

10.1136/jim-2018-000974.359

Case report A 7 months old previously healthy male presents with new-onset spastic movements. The character of these movements is sudden head and body flexion with synchronous bilateral upper extremities extension. Spasms usually last for one second and occur in a cluster-like manner over the ensuing several minutes. The onset was two weeks ago, gradually worsening in intensity and frequency. Parents have no concern about his development but endorse infant seems to be right-handed. Infant was born via vaginal delivery at term, overall uneventful pregnancy and delivery. Physical exam revealed well-nourished male infant with normal vital signs. No skin abnormality. Infant was fully responsive to environment. Neurological exam findings were all normal except for decreased voluntary movement in left arm compared to right side. 24 hour video EEG was obtained and recorded several typical events with hypsarrhythmia EEG finding, consistent with flexor type infantile spasm. MRI brain was obtained and showed old right middle cerebral artery (MCA) territory infarct (figure 1). Patient was started on high dose prednisolone and discharged home.

Conclusion Common causes of symptomatic IS include diffuse Central Nervous System (CNS) malformation, tuberous sclerosis complex, inborn error of metabolism, congenital infections, perinatal and postnatal insults. IS caused by focal CNS malformation like MCA stroke is exceedingly rare. In this case, using dominant hand before 2-3 years of age, also suggests abnormal development and requires further evaluation. It is important to identify the underlying cause of systematic IS since the treatment may differ depending on etiology, with the mainstream being glucocorticoids or ACTH, and vigabatrin being the first-line treatment for tuberous sclerosis related IS.

362 KNEE PAIN IN A 14-YEAR-OLD FOOTBALL PLAYER

¹ME Liberty*, ²J Read. ¹University of Florida College of Medicine – Jacksonville, Jacksonville, FL; ²Nemours Children's Specialty Care, Jacksonville, FL

10.1136/jim-2018-000974.360

Case report Knee pain is a common condition affecting athletes involved in contact sports and sports that require repetitive jumping, sprinting or quick directional changes. The purpose of this case report is to present an unexpected cause of knee pain in an otherwise healthy athlete.

Our patient is a 14-year-old male football player who presented to sports medicine clinic with the chief complaint of left knee pain and swelling. Onset of pain was 2 months prior

Abstract 361 Figure 1

and he began to complain of intermittent knee swelling over the last several weeks prior to this clinic visit. There was no reported history of trauma or injury. Pain worsened with activity and was localized to his anterior knee. NSAIDs and knee brace did not alleviate symptoms. He denied any associated knee instability, popping or locking. Neurological signs and symptoms were absent. Review of systems was otherwise negative. Examination revealed a moderate sized left knee effusion with tenderness to palpation around the patella and medial joint line. There was no ecchymosis or erythema. Strength exam was normal. Range of motion was decreased in flexion and extension due to swelling. He had a negative Lachman and McMurray test as well as negative anterior and posterior drawer tests. No ligament laxity was appreciated with stress testing. Examination of the contralateral knee was normal. He was otherwise well appearing with normal gait. X-rays were normal. MRI of the knee revealed distal femoral osteomyelitis with associated intraosseous and subperiosteal abscess with deep posterior knee soft tissue abscess. Labs significant for CRP 9.45 mg/dL, ESR 57 mm/hr. He was taken to the OR for I and D and cultures returned positive for oxacillin sensitive staphylococcus aureus. A PICC line was placed and he completed a 6 week course of IV clindamycin. Follow up x-rays 1 month post-op were normal. A full return to sports is expected.

In children, osteomyelitis is primarily hematogenous in origin and affects the most vascularized regions of the growing skeleton. Complications of delayed diagnosis include growth arrest, septic arthritis and chronic infection. It is important to have a broad differential and high index of suspicion for conditions such as osteomyelitis as to prevent delays in diagnosis and treatment.

363

SPONTANEOUS RUPTURE OF THE URINARY BLADDER LEADING TO PROTEUS PERITONITIS

A Magliolo*, KB Grizzle. *Children's of Alabama, University of Alabama at Birmingham, Birmingham, AL*

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Case report

1. Avoid premature closure following diagnosis in hospitalized patients.
2. Discuss diagnosis and treatment of spontaneous rupture of the urinary bladder.

Case presentation 10-year-old male with complex history including an unspecified genetic disorder, static encephalopathy, and g-tube dependence who presented with fever and hematuria. Urinalysis was consistent with UTI. He had a mild AKI, presumably from dehydration. Admitted on Ceftriaxone and IV fluids. Urine culture grew pan-sensitive *Proteus mirabilis*.

Abdominal distention, worsening fevers, and up trending CRP noted on hospital day 4. CT abdomen showed significant ascites. Exploratory laparotomy was performed due to concern for bowel perforation, however instead revealed a bladder perforation. Urology was called and his bladder was repaired. Peritoneal cultures from the procedure also grew *Proteus*. A foley was left in place to allow for bladder decompression during healing. Antibiotics changed to Cefepime and Metronidazole per infectious disease recommendations due to risk of inducible resistance with *Proteus* infection. His bladder rupture was deemed to be secondary to a combination of acute infection and longstanding neurogenic bladder. Therefore, intermittent in and out catheterizations were started after his foley was removed.

Discussion Spontaneous rupture of the urinary bladder is rare, though has the potential for high morbidity and mortality. It is usually the result of an underlying pathology that weakens the bladder wall, including malignancy, outflow obstruction, or neurogenic bladder. The dome of the urinary bladder is a common site of rupture, as it is the weakest point within the bladder wall.

The most frequent CT finding is accumulation of ascites, though free air may also be seen. Labs frequently show 'pseudo-renal failure', which is an elevation of BUN and creatinine due to intra-abdominal resorption of urine. Intraperitoneal contamination leading to peritonitis is common. It is difficult to differentiate from intestinal perforation, as the clinical symptoms and radiographic findings are similar. It is often diagnosed intraoperatively during a laparotomy for acute peritonitis. Treatment includes drainage of the bladder, closure of the perforation, and antibiotic therapy.

364

CASE OF THE CROOKED SMILE

DT Mani*. *Saint Louis University, St Louis, MO*

10.1136/jim-2018-000974.362

Case Presentation A 3 month male presents for wheezing and aspiration. A barium swallow study showed reflux and aspiration to thick and thin liquids. Laryngoscopy did not show any anatomical abnormalities. Continued intolerance of feeds prompted surgery for a gastrostomy tube and fundoplication.

Thereafter, he was diagnosed with hemifacial microsomia (HM), a rare genetic disorder that falls under the umbrella of oculo-auriculo vertebral spectrum.

HM affects the right side of the face, as supported by his right sided hearing loss, muscle weakness on right side of the face caused the left sided droop. Absent hair whorls were noted, indicating a poor neurological and cognitive outcome.

Discussion Hemifacial microsomia is a sporadically acquired defect about 1 per 5000 live births. It causes abnormalities of

Abstract 364 Figure 1

structures arising from the first and second branchial arch. The defects are unilateral majority of the time, mainly on the right. The underlying pathogenesis is uncertain, could be a cause of a vascular insult in-utero or maternal diabetes.

Absence of hair whorls implies gross defects of early brain development, the hair directional patterning may be secondarily altered. The abnormal scalp hair patterning reflects the altered growth of the fetal brain. Gross aberrations of hair patterning imply an insult that took place at 10 to 16 weeks. Hair follicles grow at 18 weeks of fetal life. The crown is the focal point where the posterior scalp skin is under growth tension exerted by the dome-like outgrowth of the brain.

Conclusion Prognosis and quality of life is dependent on IQ. Lower IQ is also noted with microphthalmia.

365 A CASE OF CYSTIC FIBROSIS MISSED BY NEWBORN SCREENING

C Glendye, A Manners*, O Rodriguez. *University of Mississippi Medical Center, Jackson, MS*

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Case report Cystic Fibrosis (CF) occurs in 1/3000 Caucasians vs 1/15000 African Americans. Pulmonary disease is the most common cause of morbidity and mortality, but infants commonly present with meconium ileus or failure to thrive (FTT). The disease is caused by mutations of the CF transmembrane conductance regulator (CFTR) gene, but the CF newborn screening (NBS) only measures serum immunoreactive trypsinogen (IRT) reflecting pancreatic function. Presently, most of NBS in the U.S. only detect up to 60% of cases. CF can be undiagnosed with a normal IRT or incomplete gene analysis. Therefore, sweat tests are still the diagnostic gold standard. The patient presented with several CF manifestations, but a normal NBS. A 5 m.o. male with history of FTT presented with wheezing, hypoxemia without fever, and a clear chest X-ray. He was born term at 3.12 kg by C-section, complicated by respiratory failure requiring a 10 day NICU stay. He was discharged off oxygen, but had few hospital visits for respiratory illness or FTT. Improper formula mixing was suspected given a high-risk social situation. He was admitted with Bronchiolitis on oxygen support. Weight was <0.01% and weight for length was 0.38%. Initial work-up showed negative metabolic or endocrine etiology. Sweat testing was unsuccessful. He

had poor weight gain despite nasogastric tube feedings. Due to loose foul smelling stools, fecal elastase was sent. Hospital course was complicated by respiratory decompensation due to pseudomonas pneumonia requiring intubation in the PICU. Both CF and immunodeficiency were considered, but fecal elastase was abnormal at <15 mcg/g confirming pancreatic insufficiency. While CF genetic testing was pending, standard acute and chronic CF care were initiated. He immediately gained weight with improved respiratory status. Outpatient sweat tests confirmed CF. Finally, CFTR gene sequencing also confirmed the diagnosis with heterozygous pathogenic mutations (c.2988+1G>A, c.825C>G). Newborn screening has been crucial in increasing the early diagnosis and management of CF. It is important to know that CF is a clinical diagnosis that requires confirmation by testing, but a negative NBS does not rule out the disease. CF should be considered in patients with FTT and symptoms of malabsorption regardless of newborn screening results or ethnicity.

366 DOUBLE ANEUPLOIDY 48,XXY,+21 ASSOCIATED WITH HYPERTROPHIC CARDIOMYOPATHY IN AN INFANT

K Mascia*, C Gooch, NA Poteh, A Hurst. *University of Alabama at Birmingham, Irondale, AL*

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Case report A neonate with a double aneuploidy 48,XXY,+21 suspected on prenatal cell free DNA (cfDNA) testing was admitted to the Regional Neonatal Intensive Care Unit (RNICU) due to respiratory distress and multiple congenital anomalies and features consistent with Trisomy 21 including broad nasal bridge, upslanting palpebral fissures, and macroglossia. Diagnosis of 48,XXY,+21 confirmed with karyotyping. At approximately 3 months of life, echocardiogram showed evidence of biventricular hypertrophy particularly involving the septum. Cardiomyopathy gene panel was sent and resulted with no abnormalities found in the genes tested. Only 8 cases of double aneuploidy associated with congenital heart defects have been previously reported in the literature.

367 A HIDDEN OSTEOMYELITIS LEADING TO DEEP VENOUS THROMBOSIS AND PULMONARY EMBOLISM CONFOUNDED BY RECENT TRAUMA

¹KR McManus*, ¹JJ Zorrilla, ²S Bradfield. *¹UF College of Medicine- Jacksonville, Ponte Vedra, FL; ²Nemours Children's Specialty Care- Jacksonville, Jacksonville, FL*

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Case report Deep Venous Thrombosis (DVT) and Pulmonary Embolism (PE) are very uncommon in the pediatric population. For this reason, an underlying reason should be sought. Several risk factors have been implicated in the development of DVT/PE such as central venous access devices, trauma, and infection. Namely, the timely diagnosis of osteomyelitis is of importance to preserve bone integrity. Here, we present the case of a 16 year old male who developed DVT/PE in the setting of a recent knee injury and was later found to have an extensive osteomyelitis. This case highlights the importance of considering osteomyelitis with a patient presenting with a DVT/PE.

368 EPILEPSIA PARTIALIS CONTINUA AS A PRESENTING SYMPTOM OF LATE SEROPOSITIVE NMDA RECEPTOR ENCEPHALITIS

AW McRae*, H Kaur, R Al-Ramadhani, J Ingram. *University of Mississippi Medical Center, Jackson, MS*

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Study purpose To demonstrate a case of late seropositive NMDA receptor encephalitis with Epilepsi partialis continua (EPC) as a clinical presenting symptom.

Methods Multiple avenues of labs, imaging and encephalogram studies reveal the eventual diagnosis of NMDA encephalitis, despite an unusual presentation.

Summary An 11 year old African American male with no past medical history presents with seizures consisting of right hand, leg, and face paresthesia progressing to right tonic hemibody events with myoclonic jerks. The patient never lost mentation.

Initial lab workup and MRI were unremarkable. Initial electroencephalogram (EEG) revealed left hemispheric continuous slowing and seizures arising from the left centroparietal area. He was treated with success by Lorazepam and Fosphenytoin. After negative work up, the patient was discharged on Oxcarbazepine. The following day he returned with seizures, new dysarthria, near continuous focal motor seizures, and a new right hemibody weakness. Clinical presentation at this point was most consistent with EPC, concerning for Rasmussen's encephalitis.

Lumbar puncture was performed and CSF studies were unremarkable, including NMDA receptor antibodies. Brain MRI with and without contrast was unremarkable. Plasma autoimmune investigation and a metabolic work up were within normal range.

He required transfer to the PICU. The patient was started on IVIG, steroids, aggressive anti-epileptic drugs, including a pentobarbital coma, and ketogenic diet. He continued to have seizures, and plasma exchange was completed. Repeat LP was performed due to poor clinical improvement and rapid progression of the disease. Repeat MRI brain, MRA of the head and neck and MR spectroscopy were unremarkable.

Results After plasma exchange, seizure cessation was noted. At that time repeat CSF NMDA receptor antibodies returned positive. The decision to proceed with Rituximab treatment given the clinical course and presentation fit the diagnosis of anti-NMDA receptor encephalitis.

Conclusion Early presentation of EPC, worrisome for Rasmussen's encephalitis, should be followed up with serial NMDA receptor antibody labs. Positive NMDA receptor antibodies can occur late in clinical course and NMDA receptor encephalitis can present with EPC.

369 SURFACTANT PROTEIN C DEFICIENCY: A CASE REPORT

M Abraham*, N Mehdi, R Katz. *OUHSC, Edmond, OK*

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Introduction Childhood interstitial lung disease (chILD) comprises various physiologic abnormalities of pulmonary tissue leading to a reduction in efficiency of gas exchange. These patients often present with persistent tachypnea, hypoxemia, and chronic cough. We present this case of surfactant protein C (SP-C) deficiency as a unique diagnosis often overlooked when caring for an infant with respiratory distress.

Case report Fifteen week old female with past medical history of persistent tachypnea, chronic cough, poor weight gain, gastroesophageal reflux, and failure to thrive was admitted to PICU for acute respiratory distress and hypoxemia. The infant required up to fourteen liters of high flow nasal cannula. Complete blood count and comprehensive metabolic panel were within normal limits. Chest radiographs showed no focal areas of consolidation, with indication of reactive airway process of viral or environmental origin. Of note, the infant was admitted one month prior for hypoxia and found to be positive for Rhino/enterovirus.

The infant was treated with antibiotics for presumed aspiration pneumonia secondary to gastroesophageal reflux. Because of persistent tachypnea, hypoxemia, and chronic cough, pulmonology was consulted and the diagnosis of interstitial lung disease was considered. Further imaging with HRCT was obtained, which showed diffuse groundglass opacities throughout the lungs bilaterally with a few small scattered subpleural cysts. Subsequently, genetic testing was obtained revealing SP-C deficiency. The infant was treated with methylprednisolone, azithromycin, and hydroxychloroquine. Due to the history of failure to thrive, continued increased work of breathing and persistent reflux, placement of gastrostomy tube with fundoplication was completed.

Discussion The most common clinical features of SP-C deficiency are persistent cough, tachypnea, and hypoxemia, which makes the illness difficult to distinguish from an infectious process. As the most common cause of respiratory distress is likely an infectious etiology, chILD is often overlooked. This case demonstrates the significance of further imaging with HRCT in an infant with persistent respiratory symptoms. Early diagnosis and intervention is essential for improving the prognosis of an infant with chILD.

370 THE TREATMENT FOR PSYCHOGENIC EPILEPSY RESULTING FROM DEPRESSION, ANXIETY, AND MENTAL TRAUMA

K Mihaylo*, A Bax. *The University of Oklahoma Health Sciences Center, Oklahoma City, OK*

10.1136/jim-2018-000974.368

Case report Our patient is a 12-year-old female with history of anxiety and depression who presented to our emergency department (ED) with recurrent shaking episodes. One and a half years prior, she was diagnosed with anxiety with persistent fears of failing school and of abduction by stranger. She was also traumatized by witnessing her sister holding a knife to her own throat. During this time, the patient underwent individual/family counseling and received fluoxetine at increasing dosages. She then had increasing mood changes followed by an episode of syncope accompanied by quivering hands and feet, lacking postictal symptoms. Later, she experienced spells of 'whole body rocking' lasting 10–15 s with quick recovery to baseline. After various episodes without returning to baseline, she was brought to an ED where she was started her on levetiracetam load and maintenance doses; however all labs were within normal limits, including Head CT and EEG. The patient continued having episodes, was brought to our ED, and withheld from antiepileptics due to the non-epileptic nature of the suspected psychogenic spells. One month after discharge, due to side effect profile of fluoxetine, she was switched to sertraline which lead to mood

improvement. Meanwhile, the spells continued and the patient underwent 48 hour video EEG that was negative. It was then postulated that the psychogenic seizures were manifestations of emotional stressors which were then addressed with an increasing doses of sertraline along with the addition of cognitive behavioral therapy. For 6 months, her emotional symptoms gradually improved as she developed consistent coping techniques, and her psychogenic seizure-like episodes largely resolved.

Discussion Our patient presented with multiple episodes concerning for neurologic seizures. After full neurological workup indicated these episodes were not consistent with epilepsy, episode resolution has been achieved through management of her emotional health. When her depression, anxiety, and mental trauma were addressed, her seizure-like activity resolved. Pharmacotherapy alone was not enough to control these psychogenic seizures, but when proper CBT was introduced and maintained, that these episodes resolved.

371 TRANSIENT HYPOALDOSTERONISM IN A PEDIATRIC PATIENT

S Mohan*. UT Houston, Houston, TX

10.1136/jim-2018-000974.369

Case report Transient defects in aldosterone secretion that are not associated with low renin have been reported in critically ill adults, however there are few reported cases in pediatric patients. A 7 month old female with a history of eczema presented from clinic with hyponatremia (Na 118), hyperkalemia (K 7.9), non-gap acidosis (HCO₃ 14, anion gap 13), anemia (Hb 7.9), hypoalbuminemia (albumin 2.1) and failure to thrive. Patient's diet at the time of presentation consisted mainly of breastfeeding supplemented with soft foods and small amounts of free water. On admission, patient was fluid resuscitated and started on hydrocortisone due to concern for possible adrenal insufficiency, as patient was intermittently hypotensive on presentation. Within 48 hours of admission, patient's sodium and potassium had normalized, although patient was still mildly acidotic. Within 72 hours patient's acidosis resolved. Hydrocortisone was tapered off over the next week. Patient's sodium remained within normal limits, although patient was intermittently hyperkalemic with questionable hemolysis. Feedings were switched from breast milk to an elemental formula due to concerns for a milk protein allergy. Upon switching to the elemental formula, weight increased and rash improved. Aldosterone on admission was <1 ng/dL (normal 5–90 ng/dL) with normal renin activity of 20.3 ng/ml/hr. Cortisol was 19.5 ug/dL and DHEA-Sulfate level was less than 15 ug/dL, which was normal for the patient's age. Urine Na was 13 mEq/L, which was not consistent with hyponatremia due to renal losses. On day 10 of hospitalization, repeat aldosterone was elevated at 477 ng/dL with an elevated renin activity of 118 ng/ml/hr. Sodium was normal at this point. Endoscopy and sigmoidoscopy demonstrated eosinophilic/allergic enterocolitis, for which patient was started on prednisone with improvement of symptoms. This patient appears to have had a transient defect in aldosterone secretion due to her critical illness, malnourishment and dehydration on presentation. This defect was not associated with hypocortisolism or low levels of renin. Her low aldosterone levels explain her significant

electrolyte abnormalities on admission. Her electrolytes improved and her aldosterone level increased with hydration and improved nutrition.

372 A CASE OF CONGENITAL CMV

NR Natha*, BA Shah. OU Children's Hospital, Oklahoma City, OK

10.1136/jim-2018-000974.370

Case report Our patient is a newborn male delivered at 36 4/7 weeks GA via vaginal delivery to an 18 year old G1P0 Hispanic female. Maternal labs negative for GBS, HIV, RPR, Hepatitis B, and GC/CT. Rubella status unknown at time of delivery due to early prenatal care in Mexico. Glucose screen negative for gestational diabetes. Pregnancy complicated by oligohydramnios leading to induction of labor for non-reactive NST. Patient delivered 4 hours after AROM. Maternal Tmax 37.3. Patient born without complications with no resuscitation steps required, strong cry and HR >100. APGARS 7, 8 at 1 and 5 min. Pt SGA with weight 2150 g, length 46.5 cm, and head circumference 30 cm.

Infant initially admitted to mother-baby unit but later transferred to NICU for persistent hypoglycemia. Treated with IV D10-W for 3 days with resolution of sx's and retention of euglycemic state with regular feeds. Patient with failed hearing screen x3 and incidentally found to have thrombocytopenia in the 30 s–40 s. IgM level and Urine CMV sent to assess for underlying cause of thrombocytopenia, hypoglycemia, and failed hearing. Urine CMV returned positive and patient started on Valganciclovir therapy. Thrombocytopenia improved but patient became neutropenic secondary to abx side effect. Valganciclovir continued with close monitoring of ANC to ensure it remained >500. MRI brain at 13 DOL with punctate acute infarct in the left frontal periventricular white matter and numerous foci. Patient's abx txt extended to 6 mos. Thrombocytopenia resolved by time of discharge and patient passed hearing screen bilaterally.

Congenital cytomegalovirus occurs in 5–7/1000 live births each year in the US. Only 10% of affected babies are symptomatic at birth with majority of infected infants showing no clinical manifestations during newborn period. The most common complication of CMV is sensorineural hearing loss (SNHL), followed by cognitive impairment, retinitis, and cerebral palsy. There are no current universal screening protocols for early detection of CMV, though some institutions have implemented targeted screening for infants with SNHL at birth. Given relatively high incidence of congenital CMV and associated comorbidities, there should be increased awareness and education for pregnant women, emphasis on early detection given potential treatability, and consideration for universal newborn screening for CMV.

373 PULMONARY INTERSTITIAL GLYCOGENOSIS: A RARE DIAGNOSIS

SM Nimri*, S DeLeon. OUHSC, Oklahoma City, OK

10.1136/jim-2018-000974.371

Case report Our patient is an 8 day old full term male admitted with persistent tachypnea. He was born via vaginal delivery with meconium-stained amniotic fluid and required PPV

after delivery. At 24 hours of life he was noted to be tachypneic; blood cultures were negative, and he was discharged following 72 hours of antibiotic therapy. He presented again at day eight of life with persistent tachypnea and decreased oral intake. CBC, CMP, Chest X-ray, and blood gases were unremarkable. Oxygen sats were in the 90 s on room air. Though tachypneic with respiratory rates up to 100 breaths per minute at times, he appeared comfortable. High flow nasal cannula was initiated without improvement in respiratory rate. Antibiotics were restarted to cover for neonatal pneumonia; meconium pneumonitis was suspected as well.

Echo showed a small PFO without findings suggestive of pulmonary hypertension. Subsequent X-rays showed minimal haziness and metabolic workup was negative. HRCT scan showed prominent ground glass opacities in the lungs bilaterally with bibasilar predominance, which was suspicious for genetic surfactant deficiency, infection, aspiration, or interstitial lung diseases. Extensive genetic testing was negative. Dysphagogram and upper GI series were without evidence of aspiration. Infectious workup was negative including viral and bacterial cultures. After approximately 2 weeks in the hospital with persistent tachypnea but on room air, he was discharged home on NG feeds. At outpatient pulmonology follow ups, his clinical status was unchanged. A subsequent thoracotomy with right lung biopsy showed pulmonary interstitial glycogenosis; a gastric tube was placed at the same time. He was then started on oral prednisolone with significant improvement in respiratory rate and is now able to take the majority of his feeds orally.

PIG is an interstitial lung disease in which glycogen is abnormally stored in pulmonary tissue. Though the etiology remains unclear and this disease was only first described in 2002, it has an overall good prognosis; most patients respond well to steroid therapy with eventual resolution of symptoms. Our patient did not present with hypoxia or increased work of breathing, as is most common, and so was a diagnostic challenge. Physicians caring for neonates should be aware of this condition and the spectrum of presenting symptoms.

374 PEDIATRIC TICK PARALYSIS

K Oelsner*, M Rowin. University of Tennessee College of Medicine, Chattanooga, TN

10.1136/jim-2018-000974.372

Introduction Tick paralysis is a rare life-threatening reversible paralysis. This report is the first case of tick paralysis documented in Tennessee in 2018.

Clinical course A 2 year-old female presented with a complaint of lower extremity weakness and inability to walk. She was in her normal state of health until the day of admission when she was unable to get out of bed or bear weight. Exam showed strength to be 3/5 in her lower extremities. Lower extremities also demonstrated areflexia. Sensation and position sense were intact. Family lives in a heavily wooded rural area, but denied any history of tick bites. Complete exam identified a tick attached to her scalp (figure 1). The tick was removed and the patient was admitted for observation. By that evening, she was walking without assistance. By the next day, she had returned to her baseline neurologic exam.

Discussion Tick paralysis should be considered in cases of rapidly ascending paralysis. Clinical exam shows ascending lower

Abstract 374 Figure 1

extremity weakness, loss of deep tendon reflexes but preservation of sensation. Tick paralysis is caused by a neurotoxin which decreases presynaptic acetylcholine release at the neuromuscular junction. In the southern US, the gravid female *Dermacentor variabilis* is the most common vector. Symptoms typically begin 3–7 days after attachment and resolve 24 hours after removal. It is crucial to distinguish tick paralysis from other causes of progressive weakness as patients may undergo unnecessary and expensive therapies.

375 PERICARDITIS TAKES A TURN FOR THE WORSE

NA Poteh*. UAB, Birmingham, AL

10.1136/jim-2018-000974.373

Case report A 9-year-old male presented with 1 week of fever, chest pain, intermittent abdominal pain and fatigue. He had vomiting but no diarrhea or weight change. He

Abstract 375 Figure 1

Abstract 375 Figure 2

initially went to an urgent care center where rapid strep and monospot tests were negative. Symptoms persisted so his pediatrician obtained an abdominal ultrasound revealing pericardial effusion and splenomegaly. Given these findings, the patient was referred to our institution for further evaluation. He was admitted for further work up and remained stable until the following day when he became hypotensive, tachycardic and hypoxic. He was ill-appearing, complained of abdominal and chest pain, and had a bout of emesis. He had muffled heart sounds on exam and abdominal tenderness. Repeat echocardiogram revealed severe cardiac tamponade. Cardiology was consulted and took him to the cath lab for pericardiocentesis. About 450 ml of serosanguinous fluid was aspirated from the pericardium and surgical pathology revealed acute on chronic inflammation but no bacteria or fungi.

376 LET'S KICK IT OLD SCHOOL: THE ART OF THE PHYSICAL EXAM

C Marbrey, N Reyes*, B Dillard, MN Frascogna. *University of Mississippi Medical Center, Jackson, MS*

10.1136/jim-2018-000974.374

Case report The Pediatric Emergency Department (PED) often sees children who are followed by multiple medical providers for various medical problems. Despite their proximity to care, the total extent of illness may not be readily apparent, or even missed. We present a case involving the acute management of a 4-week-old term infant with a history of a moderate atrial septal defect (ASD) and congenital club foot who presented to the PED in severe shock.

A 4 week old male presented to the PED with decreased oral intake, vomiting and lethargy for one day. Upon arrival, he was ill appearing and his vital signs were consistent with decompensated shock and signs of impending respiratory failure. Hepatomegaly was present and, with snug bilateral leg casts extending to the inguinal crease, his femoral pulses were difficult access, let alone palpate. Critically ill, he underwent multiple unsuccessful vascular access attempts, ultimately requiring emergent removal of his casts by orthopedics to obtain access. Chest radiography showed cardiomegaly. A point of care cardiac ultrasound was notable for systolic function of 15%, moderate ASD and enlarged right atrium. He

was intubated for airway protection and a treatment course for presumed sepsis and myocarditis was initiated.

In the intensive care unit, he required ongoing inotropic support without evidence of infectious etiology or preceding upper respiratory infection symptoms. A complete echocardiogram was performed shortly after admission revealing the unexpected diagnosis of severe coarctation of the aorta (COA). Treatment was streamlined after definitive diagnosis was made. When his shock improved, he underwent repair of his COA.

We highlight several confounding factors related to the recognition and management of COA. An echocardiogram performed at eight days of age showed an ASD, but no mention of a COA. Furthermore, the presence of bilateral leg casts likely delayed the diagnosis of COA and distracted providers from recognizing signs earlier. Finally, the diagnosis of COA in the PED was much less obvious due to the patient's hemodynamic instability and shock. This case emphasizes that simple yet critical physical exam findings can have devastating consequences if missed.

377 ATYPICAL PRESENTATION OF CAT SCRATCH DISEASE WITH NEUROLOGIC SYMPTOMS

C Saylor*, T Turner, D Miller, S Palumbo, C Sandlin. *Louisiana State University Health Sciences Center New Orleans, New Orleans, LA*

10.1136/jim-2018-000974.375

Case report Cat Scratch Disease (CSD) is typically a disease of self-limited, tender, regional lymphadenopathy caused by *Bartonella henselae* through exposure to an infected cat or cat fleas. Occasionally, there can be dissemination of disease with neurologic, ocular, and visceral organ involvement leading to life-threatening complications. We present two patients diagnosed with Cat Scratch Disease after presenting with neurologic symptoms.

The first case is a 10-year-old male who presented after a first-time seizure. He was noted to have fever, transient altered mental status and tender, enlarged lymph nodes in his left axilla and in the medial aspect of his left elbow. Neurology was consulted for the seizure. The family reported living in a mobile home community with poor hygiene and exposure to feral cats. An ultrasound revealed an abscess near the axillary lymph node, so he was treated with a 10 day course of clindamycin. He was empirically treated for CSD with a 5 day course of azithromycin. *Bartonella henselae* titers later resulted with an IgG level of 1:512, suggesting active or recent infection.

The second case is a 13-year-old female who presented with subjective fevers and occipital headaches associated with photophobia, blurred vision, and neck pain. She also had swelling and tenderness in her right antero-medial thigh near the inguinal region. An ultrasound revealed enlarged inguinal lymph nodes. The patient reported having a kitten at home that had scratched her several times. She was empirically treated with a 5 day course of azithromycin for presumed CSD. *Bartonella henselae* titers resulted with an IgM level of 1:64, suggesting acute disease.

Both patients were evaluated by a physician for their lymphadenopathy prior to presentation and admission at our hospital. An earlier diagnosis of CSD may have prevented both patients from progressing to neurologic sequelae, ultimately

preventing their hospitalizations. These two cases highlight the importance of obtaining a thorough social history including questions about recent animal exposures. These histories allow us to consider a robust list of differential diagnoses for effective patient management.

378

SEVERE RENAL ARTERY STENOSIS IN A NEUROFIBROMATOSIS TYPE 1 PATIENT, REVEALED VIA ROUTINE BLOOD PRESSURE SCREENING IN AN OUTPATIENT PEDIATRIC CLINIC

C Saylor*, M Jones, M Benak, D Aviles. *Louisiana State University Health Sciences Center New Orleans, New Orleans, LA*

10.1136/jim-2018-000974.376

Purpose Neurofibromatosis Type 1 (NF1) is an autosomal dominant disorder seen in 1 of every 3000 people. A common finding in this population is hypertension, often secondary to renal artery stenosis caused by vascular lesions. This is a case of such stenosis in a patient with known NF1 that was initially discovered through blood pressure screening at a general pediatrician's office.

Methods Chart Review

Summary of results An 8-year-old male was sent to the Emergency Department from his pediatrician's office, where he had presented with new-onset headaches, sore throat, and cough, and was subsequently found to have a blood pressure of 172/104. Hospital imaging showed severe right renal artery stenosis and accessory right renal artery, requiring admission to the nephrology service with a consult to vascular surgery. Upon further interview, it was disclosed that the patient had not seen a pediatrician regularly, nor had he ever established care with a neurologist despite having been diagnosed with NF1 at 6 months of age.

During the admission, patient required initiation of both Enalapril and Labetalol to control his hypertension. Lab workup revealed an elevated renin level with a normal aldosterone level. He was discharged home to continue these medications; he later had a stent placed by vascular surgery to alleviate stenosis. The patient has since established care with neurology, endocrinology, and hematology-oncology for other newly discovered complications of his disease process.

Conclusion Through review of this case, the importance of patients with NF1 establishing regular primary and subspecialty care with close follow-up is emphasized. Under the close surveillance of an outpatient pediatrician, along with the regular blood pressure screenings recommended for these patients by the Genetics Committee of the American Academy of Pediatrics, this patient's stenosis would have likely been discovered sooner. Earlier identification of this stenosis would have in turn allowed for earlier medical management, which may have ultimately kept this patient from undergoing the risky vascular surgery procedure that was eventually required.

379

IMMUNODEFICIENCY PRESENTING AS DIARRHEA AND FAILURE TO THRIVE

C Seaton*, K Ordemann, A Byrd, E Hauck. *Our Lady of the Lake Children's Hospital, Baton Rouge, LA*

10.1136/jim-2018-000974.377

Severe combined immunodeficiency (SCID) is a rare, fatal genetic syndrome that involves the absence of both T- and B-lymphocytic cells. The condition leads to severe susceptibility to infections; therefore, it is considered one of the most serious primary immune deficiencies. We describe a case of an infant who presented with failure to thrive (FTT), diarrhea, and elevated liver enzymes who was found to have SCID.

A 3 month old female was admitted with diarrhea and FTT. She was asymptomatic until 1 month of age when she developed increased spit-ups and cough with feeds, watery stools, and poor weight gain despite multiple changes in formula. She was initially thought to have milk protein allergy, reflux and malnutrition due to transaminitis and eosinophilia found on admission. However, despite change to an elemental formula, symptoms persisted. Stool studies and work-up for infectious causes of hepatitis were negative. Review of labs noted worsening and severe lymphopenia. Further immunologic workup revealed T-cell lymphopenia raising concern for combined immunodeficiency, likely SCID. Suspicion was confirmed by T-cell Receptor Excision Circles. She was then transferred to a bone marrow transplant center for definitive management.

SCID is caused by mutations in genes that affect the development and function of T- and B-cells, and sometimes natural killer cells. Classic symptoms of SCID include recurrent severe infections, chronic diarrhea, and FTT. Although diarrhea is commonly found in the presentation of SCID, this symptom without association with recurrent infections can be misleading. In fact, prior to newborn screening programs, diagnosis of SCID was often delayed due to protection from maternal antibodies which temporarily prevent infections in young infants. Timely diagnosis of SCID is important as the best outcomes have been demonstrated in patients receiving hematopoietic cell transplantation prior to 3.5 months of age and prior to development of any infectious complications of the immunodeficiency. Pediatricians should maintain a high index of suspicion for SCID in patients presenting with chronic diarrhea, unexplained lymphopenia, and FTT, especially in states such as Louisiana in which newborn screening programs do not include screening for this disorder.

380

LATE STAGE HSV II SKIN INFECTION REQUIRING LONG TERM IMMUNOSUPPRESSION

L Settle*, D Hahn. *OUHSC, Oklahoma City, OK*

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Case report Our patient is a female infant, who was born at 35 weeks via spontaneous vaginal delivery with bullous lesions on her scalp, face, upper extremities, and trunk. Mom denies any sexually transmitted infections or vaginal lesions during pregnancy. At 4 days old, she was seen by her primary care provider for these lesions and was started on clindamycin and topical mupirocin with no improvement. She was admitted to our hospital at 8 days old due to pain and worsening of the bullae. On exam, there were bullous lesions at various stages on the scalp, neck, upper extremities, and trunk. Nikolsky's sign was negative. Punch biopsies were sent for H and E stain and electron microscopy. She was started on intravenous acyclovir and clindamycin, and emollients with gauze wrapping to decrease fluid loss and infection risk. Cultures from multiple sites were positive for skin flora. On day of life 16, DNA

PCR for HSV I was negative but HSV II was positive. CSF was obtained and was negative for HSV I and II. Brain MRI showed scattered foci of hemorrhage and white matter insult with no meningoencephalitis. Punch biopsy results suggested late stage herpes infection. Our patient was treated for SEM (skin, eyes, and mouth) HSV disease with acyclovir for 21 days. The lesions improved with treatment and at discharge all lesions showed healthy, hypopigmented skin.

Our patient was readmitted 10 days after discharge due to return of bullous lesions on face, back and fingers that recurred 4 days after discharge. She completed an additional 14 day course of intravenous acyclovir. She was discharged on a 6 month course of suppressive therapy with oral acyclovir.

Herpes Simplex Virus infection in the newborn period can present as SEM, CNS, or disseminated disease. Having a high level of clinical suspicion of HSV infection is important in order to start acyclovir treatment early and prevent progression of disease. This case is unique because she had recurrence of lesions after 21 days of IV acyclovir treatment. Current literature lacks incidence of recurrence of HSV skin infections or the benefits of long term immune suppression with oral acyclovir. The benefits of long term oral acyclovir likely outweigh the risks of possible neurologic damage from HSV recurrence.

381 PEDIATRIC PULMONARY HYPERTENSION: A RARE CASE PRESENTING AS ABDOMINAL PAIN

N Sharma*, H Morgans, SB Sood, K Bielefeld, L Stuemky. *University of Oklahoma School of Community Medicine, Tulsa, OK*

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Introduction Primary pulmonary hypertension (PPH) is a life threatening disease with an annual incidence of 1 to 2 per million people. It is characterized by the progressive increase of pulmonary artery pressures and vascular resistance leading to right ventricular failure. The mean survival of patients with severe PPH without treatment is less than three years.

Case report We report a rare case of a previously healthy 12-year-old female who presented with epigastric and right upper quadrant abdominal pain as her predominant symptoms of PPH. She was evaluated multiple times in the month prior with outpatient ultrasound of the gallbladder showed a thickened gallbladder wall, so she was admitted for a surgical consultation. Inpatient cardiac workup was obtained due to lower extremity edema and heart murmur on physical exam. Lower extremity edema had been present during outpatient workup by parental report. She was subsequently diagnosed with severe idiopathic pulmonary hypertension and right heart failure based on echocardiographic findings. The thickened gallbladder was secondary to right heart failure. Immediately after therapeutic balloon atrial septostomy, the patient went into pulmonary hypertensive crisis leading to cardiorespiratory arrest requiring prolonged resuscitative efforts. She was placed on veno-arterial extracorporeal membrane oxygenation but ultimately developed pulmonary hemorrhage and died.

Discussion PPH in newborns is relatively well studied, but presentation during adolescence is less common and often missed. Her age is unique because she had been previously healthy prior to one month ago, and it is unusual for PPH to be asymptomatic for longer than 18 to 24 months before the diagnosis is made.

Conclusion Abdominal pain in an adolescent patient is a rare presentation of primary pulmonary hypertension. Further studies and case reports are needed to better understand treatment and prognosis among adolescents. This case emphasizes the importance of complete history and physical examination and maintaining an appropriate differential diagnosis.

382 WEIGHT LOSS IN AN ADOLESCENT FEMALE

KJ Shell*, VA Harrison. *University of Mississippi Medical Center, Jackson, MS*

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Central nervous system (CNS) germ cell tumors are rare and occur mainly in the pediatric population¹. The majority are discovered in older pediatric patients and occur primarily in the midline CNS region, with 75% originating from the pineal gland and 10%–20% from the suprasellar region^{1,2}. Of the CNS germ cell tumors, germinomas are the most common and can be characterized by their secretion of tumor markers, including alpha-fetoprotein (AFP) and the beta subunit of human chorionic gonadotropin (β -hCG), and immunohistochemical staining positive for c-kit, placental alkaline phosphatase (PLAP), and Oct-4¹. Our patient is a 15 year-old female who presented with severe weight loss, recent seizure-like activity, and a history of anorexia nervosa, recently diagnosed by a physician. Nine months prior to presentation, she transferred to a new school and began refusing to eat and hiding or throwing away food. She blamed this behavior on abdominal pain associated with intermittent episodes of diarrhea and constipation. Her BMI on presentation was 11.03 kg/m². Upon admission, she was found to have significant vision loss, bilateral optic disk pallor, and progressively declining mental status. Imaging revealed a 4.1×4.1×4.1 suprasellar mass. After subtotal resection, germinoma was confirmed with biopsy and immunohistochemical stains positive for PLAP and c-kit.

Our patient had some symptoms consistent with a neoplastic process in the CNS, such as weight loss, seizures, and vision changes. Her age and gender are also consistent with the more common presentation of suprasellar germinomas in older pediatric females. However, her weight loss appeared to be intentional and both the patient and her family endorsed a diagnosis of anorexia nervosa at presentation. Anorexia nervosa is more common in adolescent-age females and her remarkable weight loss and history of food aversion further supported the diagnosis. This unusual presentation of a suprasellar germinoma in a pediatric patient should encourage pediatricians to consider broadening their differential diagnosis in an adolescent with weight loss.

383 RHIZOMELIC CHONDRODYSPLASIA PUNCTATA TYPE 1

V Sivils*, TL Summerlin, J Philips. *University of Alabama, Irondale, AL*

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Newborns with a prenatal diagnosis of skeletal dysplasia have a broad differential that have different clinical features and genetic mutations. Though rare, rhizomelic chondrodysplasia punctata type 1 has unique clinical findings to suspect diagnosis prior to genetic mutation results.

Case A 39 wga male infant was born to a 25yo mother, diagnosed prenatally with an unknown skeletal dysplasia by amniocentesis. Maternal cell free DNA was negative and FISH was normal. Exam was significant for symmetrically SGA and lower leg contractures. Skeletal survey revealed bony abnormalities consistent with diagnosis of chondrodysplasia punctata. An abdominal ultrasound revealed mild bilateral caliectasis. Head ultrasound was normal without hemorrhage or ventriculomegaly. ECHO only showed a small PFO and small PDA. On day 2 of life the skeletal dysplasia panel resulted positive for 2 different pathogenic variants in PEX7 gene, which is consistent with the diagnosis of rhizomelic chondrodysplasia punctata type 1. An ophthalmology evaluation revealed bilateral cataracts. MRI spine revealed foramen magnum compression and cervical stenosis extending to T4/T5. Based on extent of involvement, neurosurgery did not feel patient was a candidate for surgical correction. At 5 weeks of age, patient was transferred to a skeletal dysplasia center for second opinion on neurosurgical intervention, g-tube placement, and cataract surgery.

Discussion Rhizomelic chondrodysplasia punctata type 1 is a peroxisome biogenesis disorder (PBD) characterized in neonatal period by shortening of long bones, punctate calcifications in epiphyseal cartilage, and vertebral body clefts. The diagnosis is made based on clinical findings and is confirmed by genetic testing with gene mutations. The incidence is less than one per 1 00 000 births. Mortality risk is approximately 40%. Only 39% of affected individuals are expected to survive beyond 2 years of age. All individuals have severe intellectual impairment. Treatment is focused on supportive care and limiting secondary complications. Care for these infants require a multidisciplinary approach in order to optimize long-term management plans for the patient and their family.

Conclusion Rhizomelic chondrodysplasia punctata type 1 is a rare genetic disorder with poor prognosis and characteristic clinical and genetic findings.

384 BACK PAIN – A WARNING SIGN IN CHILDREN?

^{1,2}AM Smith, ^{1,2}E Dobish. ¹UTHSC, Memphis, TN; ²Le Bonheur Children's Hospital, Memphis, TN

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A 2 year old previously healthy male presented with a right supraclavicular mass, anorexia, fatigue, night sweats, and a remote history of back pain. Back pain preceded all other symptoms by 3 months and was deemed secondary to constipation after a KUB confirmed large stool burden and a CBC and CMP were normal. The supraclavicular mass was initially noted one month prior to presentation with fever, cough, and congestion. At that time, influenza B screen was positive and while the mass was endorsed by mother, it was non palpable on exam. The patient was discharged home with supportive care. The fevers resolved but the mass persisted.

Upon return to the ED there was a 2x2 cm hard, immobile, tender supraclavicular mass. CMP, LDH, uric acid, and CRP were within normal limits. CBC was notable for thrombocytopenia. Chest x ray demonstrated a lytic lesion at the medial right clavicular head. An ultrasound of the mass showed an expansile lesion with an echogenic rim. CT showed a lytic lesion at the medial right clavicle; vertebra plana of T8; a lytic lesion of T7; a healing fracture of rib 6; erosive

changes of rib 5; a lytic lesion of the right iliac bone; and erosive changes of the right anterior superior iliac spine. Findings were consistent with Langerhans cell histiocytosis (LCH). In retrospect, the radiologist also reviewed the prior KUB showing large stool burden. On second review of this KUB, early vertebral changes were noted in the context of our suspicion for LCH. Oncology was consulted. The patient was started on a regimen of vinblastine and prednisolone.

This case emphasizes the difficulty of diagnosing LCH, along with the importance of a thorough history and review of records. KUB initially confirmed constipation, playing into the availability bias of a common diagnosis. In contrast, LCH is a rare disease, often left off of differential diagnoses as it consists of non specific signs and symptoms often seen in various minor illnesses of childhood. Diagnosis requires a combination of a certain clinical presentation, specific radiologic findings, as well as histologic characteristics. The initial presentation of back pain in a 2 year old should prompt medical providers to consider a broad differential diagnosis.

385 MULTIDISCIPLINARY MANAGEMENT OF EMANUEL SYNDROME IN A NEONATE

P Snyder*, J Phillips. UAB, Birmingham, AL

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This term male infant was transferred for management of complex medical problems due to prenatally diagnosed Emanuel Syndrome ('ES'). Mother had a history of multiple miscarriages and a balanced translocation of chromosomes 11 and 22. The baby was found to have unbalanced translocation consistent with ES. He was initially intubated for respiratory distress. Following sleep endoscopy, consulting oral maxillofacial surgery and ENT decided on tracheostomy given occlusion of the epiglottis by the tongue base. In further work-up, echocardiogram showed normal structure and peripheral pulmonary stenosis. Renal U/S showed mid pelviectasis. Other abnormalities included 13 sets of ribs (cervical ribs), fractured left clavicle, hypotonia, narrow chest and trunk, redundant skin at nape of neck, cleft soft palate, paralyzed vocal cords, microretrognathia, and low set nipples. The patient required gastrostomy and Nissen fundoplication due to poor nutritional intake. He was discharged home in stable condition, tolerating tracheostomy collar and g-tube feeds.

Discussion Emanuel syndrome, or Supernumerary der(22) is a rare chromosomal disorder with unknown prevalence. The prevalence of balanced t(11;22) carriers is unknown and these individuals are phenotypically normal. Suggestive findings of ES include severe intellectual disability, microcephaly, central hypotonia, failure to thrive, preauricular tag/pit, ear anomalies, cleft/high-arched palate, micrognathia, kidney abnormalities, congenital heart defects, and genital abnormalities in males. The long-term prognosis is directly related to the severity of the congenital malformations. Diagnosis is made by genomic testing methods including karyotype, CMA, and FISH.

Conclusion Emanuel Syndrome is a rare condition associated with high mortality rates in the first few months of life due to the severity of the congenital malformations. Management is multidisciplinary with long-term survival possible. Genetic counseling is important given the increased risk of future pregnancies for ES, balanced t(11;22), or other errors in meiotic segregation.

386 A RARE CASE OF THORACIC DESMOPLASTIC SMALL ROUND CELL TUMOR IN A THREE-YEAR-OLD MALE

S Suhag*, K Jaiswal. *University of Oklahoma School of Community Medicine, Tulsa, OK*

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Desmoplastic small round cell tumor (DSRCT) is a relatively new and very aggressive, often fatal, tumor typically presenting as an abdominopelvic mass in the second or third decade of life. DSRCT finds its origins as a small round blue cell tumor with similar characteristics as lymphoma, Ewing sarcoma and Wilms' tumor among others. For this reason, DSRCT can be easily misdiagnosed. Here, we present a rare case of thoracic DSRCT in a 3 year old male. This is the first report of DSRCT presenting as a mediastinal mass in this age group.

A 3 year old male was admitted to the pediatric intensive care unit (PICU) with a working diagnosis of pneumonia with complete opacification of the left lung on chest X-ray. Further evaluation in the PICU revealed considerable hepatosplenomegaly. CT chest showed an anterior mediastinal mass with large pericardial effusion, nodules suggestive of metastatic disease, prominent spleen, and left basilar atelectasis.

An echocardiogram indicated a very large pericardial effusion and a mass measuring 48 mm × 44 mm. Cytology results of the pericardial fluid were non-diagnostic but did show malignant cells, therefore a thoracotomy with lung biopsy was performed.

The biopsy indicated widespread pleural, parenchymal, and intralymphatic involvement by a poorly differentiated malignancy. The overall appearance of this tumor including nested growth in a densely fibrotic background, evidence of both epithelial differentiation with strong keratin positivity, and expression of mesenchymal intermediate filaments desmin and vimentin with strong perinuclear staining were most concerning for a desmoplastic small round cell tumor (DSCRT). A tumor block was sent for evaluation of EWS-WT1 translocation typical of DSRCT and was positive.

DSCRT is a rare, aggressive tumor. Diagnosis is difficult and often delayed, as patients usually present with vague symptoms. Metastatic disease is frequently present at diagnosis due to the aggressive nature of the tumor. This case indicates that DSCRT may have a more varied presentation than previously described. DSCRT can present in patients younger than formerly outlined and should remain in the differential diagnosis for undifferentiated small round cell tumors of the thorax.

387 A CASE OF FEVER IN A 5 WEEK OLD TWIN PRESENTING TO THE PEDIATRIC EMERGENCY DEPARTMENT

N Teekappanavar*, P Ghosh. *University of Alabama-Birmingham Medical Center, Birmingham, AL*

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Sepsis is defined as a systemic response to infection. Culture-negative sepsis can pose a diagnostic challenge in infants.

Case Our patient is an ex 38 wga, previously healthy 5 wk old twin who presented with tactile fever, emesis and abdominal distension for 1 day. Mom reported mild fussiness and

decreased urine output. No sick contacts. Review of systems otherwise negative.

PE: Vitals: HR 210, RR 52, Temp 104.2. Gen: crying, pale and tachypneic. HEENT: PERRL, dry lips. Pulm: grunting with increased work of breathing, suprasternal, subcostal and intercostal retractions, coarse breath sounds, no wheezing or crackles. CV: tachycardic without murmur or gallop, cap refill 3–4 secs, no edema or petechiae. Abd: distended. Rest of her exam was normal.

Differential diagnosis included sepsis vs metabolic vs necrotizing enterocolitis vs intussusception vs volvulus vs urosepsis. Initial blood gas revealed acidosis with pH 6.99, Na 165, K 6, and lactate of 5.9. WBC 30 k with a left shift. Urine was cloudy but had no leukocytes or nitrites. She received 50 ml/kg of NS bolus and was intubated. She received vancomycin, zosyn and acyclovir and was later switched to vancomycin, ceftriaxone, and flagyl. LP significant for xanthochromia. Head CT demonstrated concern for hypoxic ischemic encephalopathy and intraventricular hemorrhage. EEG was negative. MRI/MRV showed a right transverse sinus thrombus.

Her blood, urine, and post-treatment LP yielded no growth, and no causal organism could be identified. She was diagnosed with culture negative sepsis with concomitant dehydration, resulting in sinus thrombus formation. We treated her with antibiotics at meningitic dosing. Her fraternal twin remained healthy.

Conclusion In summary, an infant with fever, tachycardia, tachypnea, and abdominal distension with emesis was found to have culture negative sepsis and a venous thrombus secondary to dehydration. While some febrile infants may present with bacteremia close to the neonatal period, most only have clinical sepsis. International studies have found culture-negative sepsis is more common than culture-positive sepsis, and factors such as CBC may not correlate with the clinical picture. As a result, antibiotic choices may depend solely on maternal risk factors and symptoms at time of presentation.

388 AN INFANT WITH SEVERE HYPOKALEMIC METABOLIC ALKALOSIS

P Thakore*, K Matthew, M Anderson, O Akingbola, I Yosypiv. *Tulane University School of Medicine, New Orleans, LA*

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A five-month-old full-term female was transferred from outside ER to our Pediatric Intensive Care Unit (PICU) with recent irritability, dehydration due to poor oral intake and weight loss. Review of system was negative except for constipation since birth. Initial investigations showed hyponatremia, hypokalemia and hypochloremic metabolic alkalosis (Na-130, K-2.2, Cl-65, HCO₃-48) and VBG showed metabolic alkalosis with partial respiratory compensation (PH: 7.66, pCO₂: 54, HCO₃: 61, BE:+32). Septic work up, liver and thyroid function tests and urine toxicology were normal. Chest X-ray, ECHO and abdominal ultrasonography were normal. KUB showed a large amount of stool in the colon. After receiving normal saline bolus, and KCl, IV fluid was started. Upon arrival to PICU, BP was normal at 93/54 mmHg weight and body length was at <2nd percentile. Urine output was increased >7 ml/kg/hr. Serum magnesium was reduced at 1.4 mg/dl, and normal total (10 mg/dl) and ionized (1.19 mg/

dl) calcium. Spot urine electrolytes showed increased Cl (119 mmol/L), Na (124 mmol/L), normal K (16.6 mmol/L) and Ca to creatinine ratio <0.2. Plasma renin was high-normal (34 ng/dl) and aldosterone was low-normal (4.4 ng/dl). Bartter/Gitelman syndrome spectrum disorder was suspected. Oral NaCl, KCl, Spironolactone and Ibuprofen were started to correct electrolyte and acid-base abnormalities and reduce urine output. Genetic studies for Bartter/Gitelman syndrome were sent. The patient was eventually discharged home in stable condition. This case illustrates a case of likely Bartter syndrome due to mutations of the genes that direct synthesis or membrane insertion of the Na, Cl or K transporters at the thick ascending loop of Henle. The age of onset, severity and specific manifestations can vary from one patient to another. Molecular genetic testing should aid in confirming the specific diagnosis. The goal of treatment is to provide adequate hydration, correct electrolyte abnormalities and NSAID therapy to blunt prostaglandin-induced polyuria.

389 INFANT WITH 16P11.2 MICRODELETION SYNDROME WITH MULTIPLE CONGENITAL MALFORMATIONS

¹P Thakore*, ¹R Kiani, ¹M Steinhardt, ^{1,2}H Andersson, ¹A Dunbar. ¹Tulane University School of Medicine, New Orleans, LA; ²Hayward Genetic Center, New Orleans, LA

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16 p11.2 microdeletion syndrome is a rare disorder with an autosomal dominant inheritance pattern, although the majority of cases are *de novo*. This deletion involves 26 protein-coding genes and has mainly been associated with neurological sequelae such as autism, developmental delay and intellectual disability. We report a 37 week gestational age female infant born via elective C-section due to absent end diastolic flow to a 27 y/o G2 P1 mother. Fetal imaging demonstrated absent radii and fibula bilaterally and missing digits, suspected Tetralogy of Fallot and IUGR. Amniocentesis microarray demonstrated a microdeletion of 16 p11.2. Baby was born SGA with APGAR scores of 7 and 8 at 1 and 5 min respectively. She received blow by oxygen for desaturations prior to admission to the NICU. Physical exam was significant for large anterior fontanelle, severe micrognathia, cleft palate, single bone in forearms and legs, three digits on each hand and toe with syndactyly bilaterally. ECHO confirmed Tetralogy of Fallot. Complete skeletal survey showed incompletely ossified cranial bones, absence of radii and fibula bilaterally, absence of 4th and 5th digits bilaterally, and dextroconvex scoliosis of the thoracolumbar spine. Abdomen sonogram and MRI brain normal. Postnatal microarray confirmed a 534 KB deletion at chromosome 16 p11.2. Despite initial nominal respiratory requirements, the baby developed slowly progressive respiratory failure, characterized by worsening hypoxia and respiratory acidosis not amenable to noninvasive support. This case is noteworthy because observed deletion is one of the common microdeletion associated with autism and there are only few case reports with different congenital anomalies, however to our knowledge this is the first case that includes both Tetralogy of Fallot and agenesis of both long and small bones. As the phenotypic spectrum of this syndrome can range from mental retardation and/or multiple congenital anomalies to autism, learning and speech problems, to a normal phenotype, predicting outcome and necessary therapeutic interventions in each case from microarray results is difficult.

390 A CONGENITAL CAUSE OF RECURRENT BACTERIAL MENINGITIS IN A PEDIATRIC PATIENT

A Tufton*, C James, M Arriaga, A Prudhomme. LSUHSC, New Orleans, LA

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Bacterial meningitis is a medical emergency requiring prompt diagnosis and early initiation of appropriate antibiotic treatment. Complications can range from hearing loss and cognitive delays to neurological deficits and death. *Streptococcal pneumoniae* meningitis, in particular, carries a mortality rate of roughly 8%. Recurrence of meningitis is rare and warrants immediate immunologic and possibly anatomic work up to ascertain potential areas for intervention and prevent further episodes.

We present the case of a 4 year old vaccinated male with a history of speech delay and previously-treated *Streptococcus pneumoniae* meningitis who presented with new-onset meningitic symptoms. He presented with decreased oral intake, emesis, fever, and nuchal rigidity. Laboratory findings were notable for elevated white cell count with bandemia and CRP. Lumbar puncture CSF results showed white cell count of 2110, protein of 121, and glucose of 24. He was started on vancomycin, Rocephin, and dexamethasone. CSF culture speciated *Streptococcus pneumoniae*. With the previous episode, immunologic workup was completed and negative for immunodeficiency. Due to a history of speech delay and questionably passed hearing tests, anatomic anomaly was investigated with this episode via CT temporal bone. The scan showed right cystic cochleovestibular anomaly with wide communication between the internal auditory canal and the inner ear along with mastoid and middle ear effusions. Immediately upon myringotomy, cerebrospinal fluid was encountered, and the ear canal was packed. The patient completed a full 14 day course of Rocephin before undergoing a right tympanomastoidectomy, CSF leak repair, perilymphatic fistula repair, and encephalocele repair.

Recurrent meningitis poses a unique challenge in management of the pediatric patient. Though recurrence is rare, the complications from multiple episodes directly affect child development and can have lasting neurologic effects. As such, a careful history is necessary to determine the presence of an underlying cause, such as immunologic or anatomic. Children with a history of speech delay and failed hearing screens, especially in the setting of meningitis, may need to be screened for anatomic anomalies.

391 DELAYED DIAGNOSIS OF PRIMARY ADRENAL INSUFFICIENCY MIMICKING ACUTE GASTROENTERITIS

T Turner*, C Saylor, C Mironovici, C Sandlin. Louisiana State University Health Sciences Center New Orleans, New Orleans, LA

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Primary adrenal insufficiency is an impaired synthesis or release of hormones produced in the adrenal cortex: glucocorticoids, mineralocorticoids, and adrenal androgens. Pediatric patients with this diagnosis present with fatigue, nausea, vomiting, hyperpigmentation, and growth impairment. However, signs and symptoms can vary based on which hormones are deficient. Electrolyte imbalances, particularly hyponatremia and hyperkalemia, are common in the setting of acute illness and

present potentially life-threatening complications due to adrenal crisis. Our patient did not have hyperpigmentation or failure to thrive, and he appeared healthy between hospital visits. Such atypical presentations may delay diagnosis leading to increased morbidity and mortality.

A 2-year-old male was admitted to the pediatric intensive care unit (PICU) from an outside hospital. He had hyponatremia, metabolic acidosis, and hypoglycemia despite aggressive fluid replacement secondary to an acute gastrointestinal illness with fever and vomiting. The patient had two prior PICU admissions the previous year for similar presentations following illness. Endocrinology and genetics were consulted for the repetitive nature of events. A work-up that included thyroid studies, free and total insulin, and c-peptide was within normal limits. Testing of his pituitary-adrenal axis revealed cortisol and aldosterone levels were normal. ACTH levels were greater than 10-times the normal limit, and plasma renin activity was elevated. The patient was diagnosed with primary adrenal insufficiency. Anti-21 alpha hydroxylase antibodies were negative, ruling out an autoimmune etiology. The patient was discharged home on daily fludrocortisone and hydrocortisone.

It is important to consider adrenal insufficiency in pediatric patients who present with hypoglycemia and hyponatremia in the setting of acute illness. Our patient's diagnosis may have been delayed due to his disease process mimicking acute gastroenteritis. His case emphasizes the need to remain vigilant regarding all signs and symptoms, as further delays in diagnosis could have resulted in adrenal crisis and been life-threatening.

392

PPHN IN A PATIENT WITH WOLF-HIRSCHHORN SYNDROME-NEW FEATURE OF A RARE GENETIC DISEASE?

¹I Varier*, ²M Steinhardt, ²A Dunbar. ¹Tulane University School of Medicine, Metairie, LA; ²Tulane University School of Medicine, New Orleans, LA

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Background Wolf-Hirschhorn Syndrome (WHS) is a rare genetic disorder with an estimated frequency of 1/50,000 to 1/20,000 births.¹ Median survival was documented to be 34 +years with a significant infant mortality rate (17%).² There is a paucity of data on the natural course of the disease and etiology of early mortality. We discuss a patient with WHS with mortality related to prematurity and severe PPHN refractory to medical therapy.

Case details Infant is a 30 6/7 WGA male, twin B, born by C-section for twin-twin discordance and reversal of end diastolic flow in twin B. Apgar score: 6 and 8 at 1 and 5 min respectively. Prenatal ultrasound demonstrated for cleft lip, cleft palate and IUGR which was thought to be non-syndromic. On physical exam, the infant had dysmorphic facial features including cleft lip, cleft palate, exophthalmos, hypopspadias. He had an initial stable course in the NICU with the exception of refractory hypoglycemia, which resolved with higher GIR infusions. At 48 hours of life, he developed significant acute metabolic acidosis and hypotension. ECHO obtained was consistent with severe PPHN, requiring nitric oxide and inotropes. Other minor cardiac defects (aneurysmal atrial septum and PFO) were also identified. Head ultrasound showed grade 2 IVH. He also developed severe perfusion abnormality of left lower extremity with suspected thrombosis of the left distal femoral artery. On day of life 4, he suffered

cardiopulmonary arrest, despite initial resuscitative efforts unable to maintain heart rate >100. Parental decision was made to stop resuscitative efforts and provide comfort care. Chromosomal microarray confirmed large deletion of 4 p16.3p15.33 consistent with WHS.

Conclusion PPHN has not been described as an association of WHS. It is important to keep comprehensive information on rare genetic conditions as there is limited information regarding causality of death and life expectancy. This will help facilitate discussions with parents regarding early resuscitation efforts and expectant management of affected infants.

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393

YOU ARE WHAT YOU EAT: AN UNUSUAL CAUSE OF SEIZURES

P Redmond, B Vu*, B Dillard, C Faulk, MN Frascogna. University of Mississippi Medical Center, Jackson, MS

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Seizures are one of the most common neurologic complaints presenting to emergency departments, especially in the childhood years. Unprovoked focal seizures or evolving neurologic deficits following a generalized seizure should be considered warning signs of serious systemic or neurologic disease and require immediate investigation.

A 17 year old Guatemalan male presented to the emergency department with arm stiffness and seizure-like activity. The day prior to presentation, he had one episode of full body shaking lasting about 20 min with unilateral facial droop. After the episode, he returned to his baseline with no confusion or drowsiness and was monitored at home without seeking medical care. On the day of presentation, the family noted that his right arm had remained stiff and painful since the episode the day before and had seemed paralyzed for about five minutes that day. His mental status was otherwise normal during this episode. Exam by emergency department (ED) staff noted pain and decreased sensation of the right arm. While in the ED, labs showed a mildly elevated AST of 68 with an otherwise unremarkable chemistry profile and complete blood count. Given his symptoms a brain MRI was obtained and showed a one centimeter cyst-like mass in the left frontal lobe. He was admitted and started on Trileptal by neurology for seizure prophylaxis. Due to the cyst-like nature of the brain mass, an extensive work up for an infectious etiology was initiated with the help of infectious disease specialists. A presumptive diagnosis of cysticercosis was made and a sample of cerebrospinal fluid was sent for cysticercosis enzyme-linked immunoelectrotransfer blot (EITB) for confirmation. He completed a full treatment course of albendazole and steroids. Repeat CT scan two months after discharge showed a decrease in the ring enhancing lesions from the prior study. He has not suffered any recurrent neurologic symptoms since initiating treatment and currently remains medically stable.

The differential diagnosis for seizures and focal neurologic deficits can be quite broad. A detailed history, including travel and dietary information, may yield significant clues to help focus your evaluation.

394 PSEUDOMONAS EPIDIDYMO-ORCHITIS IN A PREPUBERTAL MALE

M Wael*, R Welliver. *University of Oklahoma, Edmond, OK*

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Our patient is an 11 year old male who presented with sudden onset of left hemi-scrotal pain and swelling which started few days after visiting an amusement water park. Patient denied sexual contact, fever, vomiting, abdominal pain, change in bowel habit, dysuria, frequency and urgency.

Initially the patient was afebrile and had left sided hemi-scrotal swelling, erythema, tenderness and had no penile discharge. An ultrasound exam showed evidence of left epididymo-orchitis. A urinalysis showed RBC 20–25 per hpf and WBC 5–10 per hpf. A urine culture showed no bacterial growth. Testing by PCR for gonorrhea and chlamydia was negative.

The patient was managed initially as outpatient with oral cefdinir and ibuprofen but two days later was hospitalized due to worsening scrotal pain, swelling, and new fever. Labs showed an elevated white blood cell count. A repeat ultrasound showed progressive epididymo-orchitis. He was started on ceftriaxone, vancomycin and azithromycin.

By day four, the patient had only minor improvement. His fevers seemed to be decreasing, but he still reported pain in the left scrotal region. Exam was largely unchanged.

Thirteen days after admission, he was found to have a testicular abscess on ultrasound. Surgical exploration revealed a moderate-sized abscess with gross purulence. The testicle was deemed salvageable, and a surgical drain was placed. The patient was started on vancomycin and piperacillin-tazobactam, which was narrowed to ceftazidime when cultures grew *Pseudomonas aeruginosa*. He ultimately required thirty days of therapy.

While epididymitis in adults is usually bacterial, in prepubertal males epididymitis would not usually require antibiotic therapy, even cases with pyuria could be caused by viral infections. These cases generally resolve within a few days from onset. We'd suggest saving antibiotics for those epididymitis/orchitis patients who don't respond equally quickly. Initial treatment possibly should include antibiotics effective against *pseudomonas*, along with other Gram-negative bacteria. Our literature review did not reveal previous reports of *pseudomonas* epididymitis in prepubertal males, suggesting this is the first reported case. It is unclear what role his recent visit to a water park may have had in his infection, though *pseudomonas* is known to thrive in wet environments.

395 KAWASAKI DISEASE: THE GREAT MIMICKER

L Wagner*, Wu CL. *University of Alabama, Birmingham, Birmingham, AL*

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Case report Learning Points: 1) Understand the diagnosis and treatment of atypical Kawasaki disease, 2) A high suspicion required to prevent premature closure of diagnosis

Case presentation A 4 year-old male presented to the ER for neck swelling and fever of two days. CT showed lymphadenopathy without abscess. He was discharged on clindamycin for lymphadenitis to follow up with pediatrician. He returned 6

days later with continued fever and no clinical improvement. Review of systems showed red eyes the day prior to presentation. Labs showed elevated CRP (47.58 mg/dL) and WBC (22.86), anemia, and thrombocytosis. Electrolytes were unremarkable. A repeat CT showed a phlegmonous neck collection and adenopathy. He was admitted for failed outpatient treatment of lymphadenitis with IV ampicillin-sulbactam. Lack of clinical improvement prompted an ultrasound, which showed bilateral cervical adenopathy without abscess. A Kawasaki workup was pursued with repeat labs and echocardiogram. He was diagnosed with atypical Kawasaki with elevated CRP, platelets, and WBC, anemia, low albumin, and abnormal echocardiogram (a right coronary and left anterior descending coronary z-score of 3.05 and 2.34). He was treated with IVIG and low-dose aspirin due to flu season. His neck swelling improved after IVIG, and he was afebrile prior to its completion. He was sent home on low-dose aspirin with Cardiology follow up. Repeat echocardiogram at 8 weeks was normal.

Discussion Kawasaki disease is diagnosed with 5 days of fever and 4 of 5 clinical criteria. Atypical Kawasaki is diagnosed when a patient fails to meet clinical criteria but meets 3 of 6 lab criteria or has an abnormal echocardiogram. Treatment for both is IVIG and aspirin for anti-inflammatory and anti-platelet effect until cleared by Cardiology. Current evidence is controversial between high-, moderate-, or low-dose aspirin. Repeat echocardiograms are recommended based on initial findings (at least in 1–2 weeks and 4–6 weeks post treatment).

Often when a diagnosis is made by another physician, physicians fall into premature closure of diagnosis. This leads to missed diagnoses and inappropriate treatment. Therefore, it is necessary to always consider alternative diagnoses, especially if patients fail to respond to the initial treatment.

396 WHEN CELLULITIS DIAGNOSES VON HIPPEL-LINDAU: A CASE OF SIGNIFICANT INCIDENTAL FINDINGS

L Wagner*, M Orr, Wu CL. *University of Alabama, Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.394

Case report Learning Points: 1. Recognize von Hippel Lindau has many presentations. 2. Consider head imaging in workup of long standing headache with vomiting

Case presentation A 15 year-old male was admitted for preseptal cellulitis with incidental bilateral papilledema. A CT of the orbits revealed preseptal edema consistent with cellulitis as well as mild dilation of the visualized portions of the lateral and third ventricles. On further questioning, he had one year of headaches and vomiting associated with 20 lbs of weight loss. He was followed by Gastroenterology, ENT, Endocrinology, and Ophthalmology without diagnosed etiology. He was started on IV clindamycin but was switched to IV vancomycin when wound culture showed MRSA resistance. He required bedside incision and drainage of multiple eyelid abscesses. MRI of brain and spine showed two large posterior fossa masses and a thoracic spinal nodule. One mass was at the level of the fourth ventricle causing obstructive hydrocephalus. The nodule was associated with a T3-T12 syrinx. An external ventricular drain was placed. The 4th ventricle mass was resected and was found to be a hemangioblastoma, which prompted a genetic work up and subsequent diagnosis of von

Hippel-Lindau syndrome (VHL). He underwent resection of the thoracic nodule two months later. There are no plans to remove the posterior fossa mass due to location and lack of symptoms. After the first resection his symptoms resolved, and he gained 11 kilograms.

Discussion Increased intracranial pressure (ICP) is an important cause of headache. Signs and symptoms include visual changes, ataxia, poor coordination, and papilledema. Causes of increased ICP can include tumors, trauma, central nervous system infections, stroke, and hypertension.

VHL is an autosomal dominant syndrome that leads to development of multiple types of benign and malignant tumors. While most cases are inherited from parents, approximately 20% are *de novo* mutations. Symptoms are dependent on the type and location of tumors. Hemangioblastomas of the brain and spinal cord often present with headache, vomiting, and ataxia. VHL is most commonly diagnosed in the third decade of life; however, it can develop any time in childhood through adulthood.

397 THROMBOEMBOLIC STROKE AS AN INITIAL PRESENTATION OF A CARDIAC MYXOMA

EC Walroth*, J Loberger, P Prabhakaran, C Rutledge. *University of Alabama, Fultondale, AL*

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Cardiac myxomas are rare tumors typically located within the left atrium. Typically patients with a cardiac myxoma either suffer cardiac symptomatology at presentation or are found post mortum. Our case illustrates a unique presentation of an embolic event originating from a left atrial myxoma with involvement of the middle cerebral artery and abdominal aorta with extension to bilateral femoral arteries.

Case A 12 year old female with a 4 month history of headaches presented to the emergency department with acute onset of slurred speech, altered mental status and gait abnormality. Initial concern was for an occult ingestion, however due to a lack of response to naloxone administration and progressive worsening of her speech and evolving right sided facial weakness, magnetic resonance imaging was obtained revealing a left middle cerebral artery infarct. She was emergently taken to interventional radiology for a thrombectomy where she was noted to have cool, pale lower extremities with diminished pulses. During the procedure, she was found to have near complete occlusion of the abdominal aorta with extension to bilateral femoral arteries. All arterial thromboemboli were successfully removed with return of normal pulses and perfusion to her lower extremities and were described as being yellow and gelatinous in appearance. She was electively intubated prior to the procedure and remained intubated due to worsening mental status. Differential on presentation was broad, however since her initial inflammatory markers were grossly normal, a vasculitic process was less likely. An echocardiogram obtained for further evaluation revealed a left atrial mass of uncertain etiology. The pathology report of the emboli subsequently revealed a final diagnosis of a cardiac myxoma. Over the next 72 hours, her neurologic status improved and was subsequently extubated. She was taken to the operating room on hospital day 6 for resection of the atrial myxoma. Incidentally, on post operative echocardiogram she was found to have a right atrial and small right ventricular mass, presumed to be myxomas as well.

Conclusion Although rare, it would be prudent to consider a cardiac myxoma in a child presenting with arterial embolic events, especially when there is no venous involvement.

398 VASOPLEGIA IN VERAPAMIL OVERDOSE

A Watson*, J Loberger, V Godsey, M Kong. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.396

Purpose Discuss the presentation and available therapies for treatment of calcium channel blocker (CCB) overdose and severe vasoplegia syndrome

Discussion A 13-year-old African-American female with type 1 diabetes mellitus presented after an intentional ingestion of 80 tablets of 240 mg extended-release Verapamil. She was drowsy but oriented on arrival with hypotension to 50/30 mmHg. Initial treatment included activated charcoal and intubation for airway protection. She then required multiple high-dose vasopressor infusions including epinephrine, norepinephrine, vasopressin, and dopamine. Intravenous (IV) intralipids and IV insulin titrated as high as 9 units/kg/hr were also given within 24 hours. Methylene blue was later added as a bolus followed by a continuous infusion. Despite transient stabilization of her hemodynamics with each aforementioned intervention, she ultimately required cannulation for veno-arterial Extra Corporeal Membrane Oxygenation (ECMO). She also experienced several dysrhythmias including cardiac stun and complete heart block that resolved with correction of electrolyte derangements. After four days on ECMO and eight days of vasopressor support, her blood pressure normalized, and she was weaned from all therapies with no obvious neurologic or cardiac sequelae.

Conclusion CCB overdoses are responsible for a significant percentage of fatal substance exposures due to profound vasoplegia, impaired cardiac contractility, and cessation of pancreatic insulin release. Initially, there is some role for activated charcoal and whole bowel irrigation to minimize gastrointestinal absorption. Lipid emulsion therapy intravascularly binds the drug. Targeted therapies to treat vasoplegia include vasopressors and methylene blue, which inhibits endothelial nitric oxide synthase to improve vascular tone. Hyperinsulinemia-euglycemia therapy propels carbohydrate metabolism within cardiac myocytes, and ECMO rests the myocardium, allowing time for the body to excrete the offending drug. CCB ingestions can pose a challenge to providers as initial signs and symptoms may not reflect the severity of the ingestion, making frequent reassessment and often aggressive management necessary to prevent morbid outcomes.

399 EVALUATION OF INTRAPERITONEAL CALCIFICATIONS SECONDARY TO MECONIUM PERITONITIS IN A NEONATE

¹L Weaver*, ²J Ladowski, ¹C Campbell. ¹University of Alabama, Birmingham, AL; ²University of Alabama, School of Medicine, Birmingham, AL

10.1136/jim-2018-000974.397

Introduction Meconium peritonitis is a chemical peritonitis secondary to intrauterine intestinal perforation. The reported incidence is 1:35 000 live births. Presentations can range from

abdominal distention, peritoneal meconium, persistent ascites, recurrent bilious vomiting, or incidental finding on ultrasound. Although the pathophysiology is generally unknown, it is suspected to result from intra- and extra-luminal, genetic, or infectious processes.

Case presentation A three-day old male infant born at 38.3 weeks via repeat cesarean was evaluated with concern for an area of calcification adjacent to liver that was noted on prenatal ultrasound. He had an otherwise unremarkable gestation, besides a mildly elevated AFP on prenatal laboratory evaluation. At birth the child had an unremarkable exam, but received further abdominal imaging which showed multiple highly echogenic foci on the visceral surface of the liver and diaphragm; however, no evidence of dilated bowel/fetal anomalies was detected. Given this information, a TORCH evaluation was completed with concern for other possible etiology of abdominal calcifications. The workup was within normal limits apart from an ophthalmological finding of bilateral peripapillary atrophy, which has been associated with congenital toxoplasmosis. He had a routine newborn course, with no notable abnormalities passing meconium and no gastrointestinal symptoms. The remaining labs, including CMV and toxoplasmosis, were negative, which helped support this as an isolated meconium peritonitis.

Discussion This case illustrates a mild meconium peritonitis. More severe findings include persistent ascites or intestinal obstruction or injury. Recent studies have indicated higher survival rates, suggesting improved antenatal diagnosis and resultant outcomes. Increasing awareness of the disease process could prompt early postnatal surgical interventions and reduce morbidity and mortality. A thorough evaluation of all differential diagnoses is important, particularly in this case, due to the acute gastrointestinal symptoms possible with Meconium Peritonitis and the long term sequelae that could result from a congenital infection.

400 PERSISTENT HYPOGLYCEMIA PRESENTING IN AN 8 MONTH OLD

M Wester*, R Sheets, V Godsey. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.398

Summary of results GH is an 8-month-old African-American male with a history of reflux and malnutrition who presented to the Emergency Department (ED) with lethargy and hypoglycemia. Mom reported a two-week history of URI symptoms and one week of decreased solid food intake. Two days prior to presentation the patient had an episode of vomiting and diarrhea. The patient presented from the pediatrician's office to the ED due to lethargy and persistent hypoglycemia with a blood glucose of 30 despite breastfeeding. Upon arrival to the ED, the patient was afebrile, heart rate of 117, blood pressure 102/55, 100% on room air. He was in no distress but noted to be small for age. He had dry mucous membranes, flat nasal bridge, and small eyes. He was initially listless, but became more alert with glucose administration. A septic workup was initiated. The lumbar puncture results were unremarkable. Other labs were normal with the exception of thrombocytosis of 9 00 000 and glucose of 24. Despite numerous dextrose boluses, the blood glucose remained below 60. Ultimately his glucose improved

once he received IV fluids with a glucose infusion rate of 11.5. Within thirty minutes of stopping dextrose-containing IV fluids, the patient's glucose dropped from 101 to 34. While hypoglycemic, his insulin level was inappropriately elevated at 5, suggesting hyperinsulinism. C-peptide was decreased and cortisol was normal. The glucagon challenge was normal so he was started on diazoxide for therapy and had no further episodes of hypoglycemia.

Conclusion The differential for hypoglycemia is broad and varies depending on the age of the patient. It is most commonly attributed to disorders of metabolism, hyperinsulinism, toxic ingestions, and hormonal imbalances, though in our patient's age group, endocrine causes must be strongly considered. Electrolytes, liver function tests, and specific metabolic labs are required to decipher the etiology of hypoglycemia. Careful and experienced interpretation of the results is necessary to prevent further episodes of hypoglycemia and permanent neurologic damage.

401 FULL FRONTAL ACUITY: FROM STUFFY TO PUFFY

S Whitwell*, K Schneider. *University of Mississippi Medical Center, Jackson, MS*

10.1136/jim-2018-000974.399

Case report Pott's Puffy Tumor (PPT) is a rare complication of sinusitis characterized by frontal bone osteomyelitis and subperiosteal abscess that can involve the epidural space via venous communication. We present a 13 year old female with a history of recurrent sinusitis who presented with one week of headache, vomiting, and rhinorrhea. She developed significant facial swelling localized to the forehead and peri-orbital regions, as well as radiating forehead pain and pain with eye movement. Upon presentation, she was afebrile. Complete blood count and metabolic panel were normal, but CRP was elevated (6.8 mg/dL). CT of the face confirmed extensive sinusitis, significant soft tissue swelling over her frontal sinus, and frontal bone osteomyelitis suggestive of Potts puffy tumor. She began treatment with IV vancomycin and ceftriaxone, but she had no improvement in her symptoms and her CRP was increasing (18.7 mg/dL). MRI face and brain confirmed the presence of a subperiosteal abscess with epidural spread. Intracranial extension called for increasing ceftriaxone to meningitic dosing, and metronidazole was added to her therapy regimen. She was taken to the OR on hospital day 3 for frontal abscess drainage (11 cc), subperiosteal abscess drainage, sinusotomy, and ethmoidectomy, and again on hospital day 8 for further frontal abscess drainage and sinus debridement. Blood and wound cultures showed no growth. Antibiotic therapy was narrowed to ceftriaxone and metronidazole. Her CRP trended down (1.3 mg/dL) and her symptoms improved. She was discharged on hospital day 12 with a PICC line to continue IV ceftriaxone and PO metronidazole for 8 weeks. Repeat imaging showed improvement but not resolution, so antibiotics were continued for a total of 10 weeks. PPT can be a threatening entity with the possibility of intracranial extension which can include epidural abscess, subdural empyema, dural sinus thrombosis, and brain abscesses. All patients are treated with high dose antibiotics, and many undergo craniotomy. Although PPT can have serious sequelae, prognoses tend to be favorable with few long-term consequences if diagnosed and treated early.

402 PRESENTATION AND MANAGEMENT OF A PATIENT WITH COHEN SYNDROME AND 16P11.2-P12.2 MICRODELETION

TM Yusin*, C Falcon. Tulane University School of Medicine, New Orleans, LA

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Purpose To describe a case of a newborn with two rare genetic abnormalities, Cohen syndrome and 16 p11.2–p12.2 microdeletion syndrome, highlighting the clinical features of each and the early interventions that should be initiated.

Case The patient was born with left hand polydactyly and microcephaly. He also has congenital neutropenia and has developed bilateral esotropia. In addition to the physical abnormalities, he has had delayed developmental milestones impacting motor, social, and language skills.

Microarray Comparative Genomic Hybridization revealed a deletion of exon 4–33 and a c.1015dupA of VPS13B, diagnostic of Cohen Syndrome. The c.1015dupA mutation has been reported in the literature, however the exon 4–33 deletion has not been reported in individuals with Cohen syndrome. The patient also had a 16 p11.2–16 p12.2 microdeletion.

Discussion Cohen Syndrome is an inherited, autosomal recessive disorder that leads to a pathogenic variant of the VPS13B gene, a gene involved in glycosylation in the Golgi apparatus. Since glycosylation is vital, dysfunction of the VPS13B gene can result in a variety of impacts. This includes developmental delay, microcephaly, intellectual disability, hypotonia, neutropenia, and progressive retinchoroidal dystrophy. There have been 200 confirmed cases of Cohen Syndrome.

16 p11.2–p12.2 microdeletion syndrome is also a rare genetic disorder. It is an autosomal dominant, but usually non-inherited, genetic disorder that can lead to developmental delay and facial dysmorphisms. This mutation has been clinically and molecularly characterized in 5 patients.

The combination of mutations seen in this patient highlights the distinct characteristics of each mutation as well as some overlapping and contrasting qualities. This is the first known case reported in the medical literature. More important than the characteristics of these genetic disorders are the management of their manifestations. Early medical interventions are vital to improve the quality of life in these patients. This includes a comprehensive medical team consisting of a geneticist, developmental pediatrician, ophthalmologist (bilateral esotropia), hematologist (neutropenia), and a wide range of ancillary specialists such as physical, occupational, and speech therapists, among others.

403 PRIMARY ADRENAL INSUFFICIENCY OF UNKNOWN ETIOLOGY

N Zeky*, C Smith, E Klepper, J Gardner. Our Lady of the Lake, Baton Rouge, LA

10.1136/jim-2018-000974.401

Case report A ten day old male presented to the Emergency Room after two days of emesis, increased fussiness and increasing lethargy. He was a previously healthy full term,

formula fed infant with a normal newborn screen. Pertinent positive findings on physical exam were an ill appearing baby with jaundice, delayed capillary refill, and increased skin turgor. His initial lab evaluation showed a sodium of 124 mmol/L, potassium of 9.3 mmol/L, chloride of 98 mmol/L, glucose of 57 mg/dL, bicarbonate of 9 mmol/L, and lactic acid of 4.2 mmol/L. During his evaluation, he developed supraventricular tachycardia followed by ventricular tachycardia. Airway stabilization occurred quickly with intubation. Initially, he received amiodarone, calcium gluconate, insulin with dextrose, albuterol, and kayexylate to stabilize his heart and correct his hyperkalemia. Due to the hyponatremia and hyperkalemia, endocrinology was consulted to evaluate for adrenal insufficiency (AI) with crisis. Further labs showed an elevated plasma renin activity value with a low serum aldosterone, supporting a diagnosis of AI. Stress dose hydrocortisone was initiated and empiric, broad spectrum antibiotics were also started to cover for sepsis.

Adrenal insufficiency in the neonatal period often manifests as an adrenal crisis within 2 weeks of life. Typical presentations include a sepsis-like picture with electrolyte abnormalities and hypotension. Common causes of primary adrenal insufficiency in young infants include congenital adrenal hyperplasia, in particular 21-hydroxylase deficiency, and very rarely 3-beta hydroxysteroid dehydrogenase (3-beta-HSD) deficiency. Our patient had a normal 17-hydroxyprogesterone, ruling out 21-hydroxylase deficiency, as well as normal male genitalia, making 3-beta-HSD very unlikely. Other rare forms of AI have been considered, including adrenoleukodystrophy and adrenal hypoplasia congenita, but his serum very-long chain fatty acid profile was normal, and sequencing of the NR0B1 gene was normal.

He was discharged on hydrocortisone and fludrocortisone treatment. Work up for his AI etiology has been extensive but continues to be inconclusive. Adrenal crisis is life threatening and can have a wide variety of presenting symptoms so it is important for all pediatricians to have it of their differential for most acutely ill pediatric patients.

404 AN INTERESTING CASE OF A BOY WITH IGA DEFICIENCY PROGRESSING TO CVID IN THE SETTING OF NON-REFRACTORY THROMBOCYTOPENIA

¹JJ Zorrilla*, ¹N Tariq, ¹D Nordli, ^{1,2}PA Pitel. ¹University of Florida College of Medicine-Jacksonville, Jacksonville, FL; ²Nemours Children's Specialty Care, Jacksonville, Jacksonville, FL

10.1136/jim-2018-000974.402

Case report Combined Variable Immunodeficiency Disorder (CVID) is characterized by a variety of clinical features including recurrent infections, autoimmune manifestations, and allergic diseases. It is often difficult to diagnose due to the heterogeneity of presentation. Here, we present a 16 year old male with a history of IgA deficiency and recurrent infections presenting with Immune Thrombocytopenic Purpura and autoimmune hemolytic anemia (Evan's Syndrome). Rarely, IgA deficiency may be the presenting manifestation of CVID, but this case highlights the importance of considering CVID in symptomatic patients with IgA deficiency.

Perinatal Medicine

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

405

EFFECTS OF CHANGE IN THE DELIVERY ROOM MANAGEMENT WITH THE NRP-RECOMMENDED ECG MONITORINGST Ahmed*, M Anderson, EG Szyld, A Wlodaver, BA Shah. *OU Medical Center, Oklahoma City, OK*

10.1136/jim-2018-000974.403

Purpose of study In seventh edition, NRP® recommended ECG monitoring for babies receiving positive pressure ventilation (PPV) or higher support. The purpose is to study the impact of change on practice interventions and outcomes.

Methods used A cohort study comparing 100 newborns requiring positive pressure ventilation (PPV) retrospectively before implementation of new NRP guidelines to 100 newborns requiring PPV prospectively after implementation. Comparisons of categorical variables between groups were made using the Chi-square or Fisher's exact test as appropriate. Continuous variables were assessed for normality and comparisons between groups were made using Student's t-test, or the Wilcoxon-Mann-Whitney test, as appropriate.

Summary of results Newborns delivered prior to implementation of new guidelines who required PPV were endotracheally intubated more, and less patients were given continuous positive airway pressure (CPAP) or face-mask ventilation. Compared to preimplementation, more newborns received CPAP (52 vs 79, p-value of <0.05) and face-mask ventilation (76 vs 96, p-value of <0.05). Less newborns were intubated (100 vs 88, p-value of <0.05) compared to preimplementation group. Chest compressions were increased in post-implementation (5 vs 14, p-value of <0.05). Increased chest compressions was not associated with a statistically significant increase in epinephrine use. There was no difference in mortality or incidence of hypoxic ischemic encephalopathy between the two cohorts.

Conclusions Our facility has successfully implemented NRP-recommended cardiac monitoring in the delivery room. There was an increased use of CPAP and face-mask ventilation using the new guidelines. ECG monitoring has allowed us to detect the need for more effective ventilation without needing to endotracheally intubate neonates. Strategies to provide effective ventilation is ongoing. Further studies with increased power are needed to validate these findings.

406

PRETERM INFANTS WITH BRONCHOPULMONARY DYSPLASIA DEVELOP A LESS DIVERSE INTESTINAL MICROBIOME

¹MH Althouse*, ^{2,3}A Venkatachalam, ^{1,4}S Ford, ¹K Lingappan, ^{2,3}R Luna, ¹AB Hair. ¹Baylor College of Medicine, Houston, TX; ²Texas Children's Hospital, Houston, TX; ³Baylor College of Medicine, Houston, TX; ⁴University of South Florida, Tampa, FL

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Purpose of study The study of the developing microbiome in the context of neonatal lung health and disease is a growing area of interest. Bronchopulmonary dysplasia (BPD) is a debilitating lung disease affecting preterm infants and leads to significant morbidity. In neonates, there is a complex relationship between the development of the gastrointestinal microbiota and the regulation of immune function. The purpose of this study is to elucidate the differences in the intestinal microbiota between preterm infants who develop BPD and those who do not, as well as the effect of antibiotic exposure on the neonatal microbiota in these cohorts.

Methods used Stool specimens were collected from very low birth weight (VLBW) infants born <1500 g BW during the first six weeks of life. 16S rRNA gene sequencing was performed. Overall bacterial diversity and relative abundance were determined in each sample. BPD was defined as need for supplemental oxygen >21% for >28 days. The primary outcome was diversity of gut microbiota at the genus level among infants with/without BPD along with antibiotic exposure taken into consideration for each comparison.

Summary of results Among all preterm infants exposed to early antibiotics, microbial diversity was significantly decreased in those who developed BPD compared to those who did not. In infants who developed BPD and were exposed to antibiotics, there is a significant increase in the relative abundance of *Escherichia/Shigella* and *Streptococcus* in comparison to infants that developed BPD without antibiotic exposure. Among the antibiotic exposed cohort, the relative abundance of *Veillonella* is significantly decreased in infants with BPD compared to the infants who did not develop BPD.

Conclusions Our study suggests that VLBW infants who develop BPD have decreased microbial diversity and display significant changes in the composition of their intestinal microbiota compared to those without BPD. Early antibiotic exposure also leads to distinct microbiota changes in this cohort. These results highlight the potential for restoration of a healthy gut microbiome in the prevention/treatment of BPD.

407

ACCURACY OF CONTINUOUS CAPNOGRAPHY IN VENTILATED NEONATES ADMITTED TO THE CARDIOVASCULAR INTENSIVE CARE UNIT

¹A Arias-Shah*, ²C Rusin, ²S Acosta, ¹J Hagan, ¹CJ Fernandes, ¹M Pammi. ¹Baylor College of Medicine/Texas Children's Hospital, Houston, TX; ²Baylor College of Medicine, Houston, TX

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Purpose of study End-tidal carbon dioxide (ETCO₂) monitoring is widely used in the adult and pediatric intensive care units, but its use in neonates has not received the same degree of acceptance. The purpose of this study is to assess the accuracy of ETCO₂ monitoring in neonates in the cardiovascular intensive care unit (CVICU).

Methods used This was a retrospective, observational, single center study conducted between January 1, 2013 to August 8, 2018 in neonates receiving mechanical ventilation and had an indwelling arterial catheter in the CVICU. ETCO₂ measurements were simultaneously compared with PaCO₂ drawn from an indwelling arterial line. A total of 15 314 dyad measurements were obtained from 578 patients.

Abstract 407 Figure 1 Number of point = 15314

Summary of results In general, PaCO₂ is higher than ETCO₂ with an average difference of 10.4 mmHg (SD=7.3) and limits of agreement -4.2 to 25.0 mmHg in this population. The correlation coefficient between the two measurements is 0.614.

Conclusions There is moderate agreement between ETCO₂ and PaCO₂ in mechanically ventilated neonates with a cardiac condition. Our study suggests that continuous ETCO₂ monitoring can provide useful trending information even in a population that is expected to have both lower tidal volumes and poor perfusion. Future studies should include stratification by gestational age or weight to further understand its impact on accuracy in neonates.

408 CONGENITAL ACUTE MYELOID LEUKEMIA IN A PRETERM INFANT

ME Barbian*, J Miller, MA Pakvasa. Emory University, Decatur, GA

10.1136/jim-2018-000974.406

Case report Congenital leukemia is a rare disease, with an incidence of less than 5 per 1 million births that develops in utero and presents in the first month of life. Among cases of congenital leukemia, acute myeloid leukemia (AML) is more common than acute lymphoblastic leukemia. Hepatosplenomegaly and leukemia cutis, are described in 80% and 60% of patients, respectively. In contrast, hyperleukocytosis is seen in half of patients with congenital AML. We report the case of a preterm infant presenting with congenital AML at birth.

Case presentation A 30 2/7 weeks' gestation infant was born via cesarean section for non-reassuring fetal status in the setting of acute on chronic placental abruption. At delivery, she required positive pressure ventilation but was transitioned to continuous positive airway pressure prior to admission to the neonatal intensive care unit. Her physical exam was significant for a preterm infant with coarse facial features, midface hypoplasia, posteriorly rotated, low set and edematous ears, bulbous nose, tachypnea with intercostal retractions, 2/6 systolic murmur in the left lower sternal border and hepatomegaly

extended midline and into the pelvis. Following admission, she was intubated for respiratory failure. An echocardiogram demonstrated an enlarged right atrium, mild pericardial effusion and tricuspid regurgitation. On laboratory evaluation, she had a white blood count of 57.1 10E2/MCL with 0% neutrophils, 17% myelocytes, platelet count of 71 10E3/MCL, hemoglobin of 6.1 GM/DL and hematocrit of 18.5%. She was coagulopathic and had hyperbilirubinemia. Flow cytometry demonstrated abnormal myeloid cells with monocytic differentiation and aberrant CD56 expression, consistent with AML.

Discussion Our patient's presentation was complex, confounded by her prematurity and maternal history of abruptio. Her coarse facial features and edematous ears, which resolved with chemotherapy, were due to leukemic infiltrates. Her respiratory distress and findings on echocardiogram were supportive of hyperviscosity syndrome versus leukemic infiltration. Our case highlights some of the various ways congenital AML can present, particularly in a preterm infant, in which the presenting symptoms might be easily explained by other conditions typically seen with prematurity.

409 OPTIMISING PERFORMANCE OF VENTILATORY CORRECTIVE STEPS IN NEWBORN RESUSCITATION BY PEDIATRIC RESIDENTS

¹D Sambalingam, ²L Bautista*. ¹Texas Tech University Health Sciences Center, El Paso, TX; ²University of Texas Southwestern Medical Center, Dallas, Dallas, TX

10.1136/jim-2018-000974.407

Purpose of study NRP recommends integrating corrective ventilatory steps as MR-SO-P-A.

The aims of this project are

1. To assess ease and effectiveness in performing corrective steps in 3 different ways
2. To determine the residents' adherence and practice and to explore differences amongst the residents based on their year of training.

Methods used Simulation-based prospective testing of the NRP was performed with NRP certified residents in all 3 years of training. Residents were randomly assigned in groups of 3 and asked to perform the corrective steps 3 times as MR-SO-P-A, MR-SO-P-A, AND M-R-S-O-P-A. The resuscitations were videotaped for assessment. The residents were asked to perform the initial assessment, effective ventilation and corrective steps

Abstract 409 Figure 1 Variation in performing corrective steps

as per NRP. Time taken to reach various time points was measured. A self-assessment questionnaire was given to the residents after the scenario.

Summary of results 45 residents participated in the study in 15 groups of 3 each. There was no significant difference in time taken to reach corrective steps in any of the groups. The consistency for the time taken to perform corrective steps and hence to initiate chest compressions and cohesive teamwork were best in the MRSO-P-A group. Trainee awareness of current recommendations of corrective steps was 48%.

Conclusions Integrating corrective steps into resuscitation most effectively to maintain consistency and without delaying chest compressions could be achieved better with MRSO-P-A technique. Continuous education could alleviate the confusion in NRP certified providers.

410 GIANT OMPHALOCELES: A CLINICAL CHALLENGE IN NEONATAL CARE

CC Beaulieu*, C Sawyer, AM Khan. University of Texas Health Science Center at Houston, Houston, TX

10.1136/jim-2018-000974.408

Purpose of study Omphaloceles, abdominal wall defects that occur in neonates, are often associated with additional anomalies typically coinciding with chromosomal abnormalities, which have a significant effect on neonatal outcomes. Omphaloceles have been classified into 3 groups: small, giant and ruptured. Giant omphaloceles are defined in our institution as a defect larger than 4 cm in diameter in term infants, often involving partial or complete liver herniation. They are associated with increased morbidity and mortality usually due to common co-morbidities. In this case series, we focus on a group of neonates diagnosed with giant omphaloceles and describe the complexities of their clinical courses.

Methods used We reviewed 33 cases of infants born with omphaloceles at our institution between January 2015 and July 2018. Among those, 20 were classified as giant defects. We conducted a chart review to assess baseline characteristics, associated anomalies and different outcomes among the selected population.

Summary of results Of the 20 giant omphaloceles identified in our review, 15 infants (75%) survived until discharge. Among the 5 neonatal deaths, 2 cases were complicated by rupture while another 2 were associated with chromosomal abnormalities. Major predictors of poor outcomes defined as death or prolonged hospital stay included omphalocele rupture (30%), chromosomal anomalies (25%), or multiple

associated anomalies (45%). Only 4 of the infants with giant defects did not have additional associated anomalies. In comparison, 7 (35%) were suspected to have pulmonary hypoplasia with 3 progressing to tracheostomy and long-term mechanical ventilator dependence. Additionally, 7 (35%) had an associated cardiac lesion. Multiple infants developed feeding difficulties with 6 (30%) requiring TPN long-term (≥ 30 days).

Conclusions Infants with giant omphaloceles often have complex medical courses. Predictors of morbidity and mortality including omphalocele rupture, chromosomal abnormalities, and multiple associated anomalies, as indicated by our case review, can guide for long-term prognostics.

411 VALIDATION OF SEPSIS RISK CALCULATOR IN A HIGH EARLY ONSET SEPSIS INCIDENCE POPULATION

E Benaim*, K Upadhyay, AJ Talati. UTHSC, Memphis, TN

10.1136/jim-2018-000974.409

Purpose of study Early onset sepsis (EOS) incidence has declined over the last 2 decades. However, the incidence has been variable based on population. A web-based EOS calculator (CAL) has recently been used to evaluate the risk in newborns ≥ 34 weeks. Our purpose was to validate the CAL in a setting with an EOS incidence of 2/1000 live births.

Methods used Retrospective review of all newborns born ≥ 34 weeks admitted to NICU from Jan 1, 2016 to Dec 31, 2017. The CAL was applied to all neonates using an incidence of 0.6/1000 and 2/1000. Data were divided into four cohorts of

Abstract 411 Figure 1

Abstract 411 Table 1 Demographic characteristics

	Total (n=1367)	Q1 (n=343)	Q2 (n=349)	Q3 (n=351)	Q4 (n=324)
African-American, n (%)	1043 (76.3)	263 (76.7)	274 (78.5)	270 (76.9)	236 (72.8)
Male, n (%)	716 (52.4)	187 (54.5)	187 (53.6)	183 (52.1)	159 (49.1)
EGA (weeks), median (IQ-UQ)	38.0 (36.3–39.3)	38.2 (36.3–39.4)	37.6 (36.2–39.2)	38.2 (36.3–39.3)	37.6 (36.4–39.3)
Intrapartum temperature ($^{\circ}$ C), mean $\pm$ SD	37.2 $\pm$ 0.66	37.2 $\pm$ 0.66	37.2 $\pm$ 0.68	37.2 $\pm$ 0.68	37.1 $\pm$ 0.63
Rupture of membranes (hours), median (IQ-UQ)	3.35 (0.03–11.51)	3.57 (0.07–11.93)	3.31 (0.03–10.81)	3.82 (0.02–12.70)	2.88 (0.03–9.52)
Positive/unknown maternal GBS status, n (%)	736 (53.8)	197 (57.4)*	186 (53.3)	175 (49.9)	178 (54.9)
Intrapartum antibiotic use, n (%)	475 (34.7)	141 (41.1)†	112 (32.1)	115 (32.8)	107 (33.0)

* vs. Q3 ($p < 0.05$)

6 month periods for comparison. The rate of abx use was compared between local protocol and the CAL.

Summary of results Of the 1367 newborns, 679 received abx (table 1). Over 2 years, abx use has declined significantly (figure 1). The CAL would have recommended abx for 468 patients (31% decline) for an incidence of 0.6/1000, but when local rate of EOS was applied (when available in July 2018) the CAL recommended abx for 673 patients (1% decline) overall. Figure 1 shows declining use of abx with local protocol and comparison to CAL.

Conclusions The EOS CAL could be helpful in reducing abx, but local incidence of EOS should be known and applied when using the CAL. Our local protocol seemed to be comparable to the CAL, especially for the last 6 months of the study period.

412 DIAGNOSTIC ACCURACY OF ADJUSTED GROWTH CURVES TO PREDICT COGNITIVE SCORES AT 24 MONTHS IN EXTREMELY PRETERM INFANTS

A Bhatia*, AA Salas. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.410

Purpose of study Traditional or unadjusted growth curves do not account for early postnatal physiologic weight loss or changes in growth velocity. Growth curves adjusted for such variations could redefine growth restriction, determine ideal catch-up growth, and predict risk of neurodevelopmental impairment. We compared the diagnostic accuracy of adjusted and unadjusted growth curves to define postnatal growth and predict neurodevelopment at age 2 years in extremely preterm infants.

Methods used We performed a retrospective cohort study assessing infant growth at 36 weeks post-menstrual age (PMA) in 350 extremely preterm infants born ≤ 26 6/7 weeks gestational age (GA). Postnatal growth was defined as below, within, or above target using adjusted and unadjusted growth curves. Linear regression models were used to compare adjusted and standard growth trajectories at 36 weeks PMA. The primary outcome was Cognitive Composite Score (CCS) of the Bayley Scales of Infant Development-III (Bayley-III) at 24 months.

Summary of results Mean birthweight (BW) was 750 ± 138 g and median GA was 25 weeks (interquartile range: 24 to 26). A multivariate analysis of postnatal growth defined with adjusted curves and eight covariates (GA, BW, weight-Z-score at birth, sex, race, antenatal steroid use, singleton birth, and corrected age at follow-up assessment) predicted higher CCS-Bayley-III scores at 24 months in infants with postnatal growth within target (adjusted mean $\pm$ standard error: 89 ± 3) and lower scores in infants with postnatal growth below and above target (85 ± 3 vs. 83 ± 6 , respectively) at 36 weeks PMA ($p=0.04$). A multivariate analysis of postnatal growth defined with unadjusted curves and the same covariates did not predict significant differences in scores of infants with postnatal growth below (86 ± 3), within (86 ± 3), or above target (86 ± 6) at 36 weeks PMA ($p=0.99$).

Conclusions Adjusted growth curves identified an inverted 'U-shaped' association between postnatal growth and CCS-Bayley-III scores at 24 months. Additional well-powered studies are needed to validate the diagnostic ability of adjusted growth curves in routine clinical practice.

413 DONOR BREAST MILK IMPROVES FEEDING TOLERANCE IN INFANTS WITH CRITICAL CONGENITAL HEART DISEASE

A Bhatia*, S Abernathy, A Moellinger, R Toms. *University of Alabama at Birmingham, Birmingham, AL*

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Purpose of study Neonates with critical congenital heart disease (CCHD) are prone to feeding intolerance resulting in growth failure. A standardized pre-operative feeding protocol emphasizing feeding mother's milk and donor breast milk was implemented in 2015. All neonates admitted to Children's of Alabama Cardiovascular Intensive Care Unit were fed according to this protocol. Our objective was to compare the impact of breastmilk feeding versus formula feeding on weight gain, tolerance to advancement to full feeds, and the incidence of NEC.

Methods used A case control study was performed of infants with CCHD requiring corrective or palliative surgery at a single tertiary center between 2013 and 2017. Outcomes of infants admitted following feeding protocol initiation in March 2015 who received mother's milk or donor milk (MM/DM) were compared to infants admitted prior to protocol initiation who were routinely formula fed (FF). We confirmed type of feed received and assessed weight gain from birth to hospital discharge, tolerance to advancement to full feeds (defined as 120 ml/kg/day), and incidence of NEC. Infants in both groups received standard bovine milk fortification. Analyses were conducted utilizing chi-squared and Student's T-tests.

Summary of results Of 131 infants, 63 infants were in the MM/DM group and 68 infants in the FF group. CCHD diagnoses, sex, gestational age, and birth weight were similar between groups. No significant difference was detected in discharge weights of infants receiving MM/DM vs. formula (3.95 ± 1.53 vs. 3.97 ± 1.23 , $p=0.9$). Tolerance to feeding advancement to 120 ml/kg/day post-operatively was greater in the MM/DM group ($p=0.05$). No significant difference was identified in NEC rates between MM/DM and FF groups ($p=0.5$).

Conclusions Feeding MM/DM improves tolerance to advancement to full feeds compared to FF group. MM/DM also effectively promotes weight gain comparable to FF infants with CCHD.

414 TRICUSPID VALVE MASS IN A NEONATE WITH BECKWITH-WIEDEMANN SYNDROME

A Bhatia*, A Freeman, J Philips. *University of Alabama at Birmingham, Birmingham, AL*

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Case report Beckwith-Wiedemann syndrome is an overgrowth syndrome characterized by hemihyperplasia and risk of intra-abdominal tumors, though cardiac tumors are rarely associated with this disorder. We report a case of a neonate diagnosed with Beckwith-Wiedemann Syndrome and intracardiac tumor.

A female neonate was delivered at 36.3 weeks to a 26-year-old woman whose pregnancy was complicated by fetal cardiac anomaly and hydrops fetalis. Mother had adequate prenatal care with previously unremarkable findings. Ultrasound performed six days prior to delivery showed new-onset polyhydramnios, small pericardial effusion, and a 2.0×1.7 cm tricuspid valve mass causing downward valvular displacement.

Repeat ultrasound on day of delivery demonstrated new-onset scalp edema and persistent pericardial effusion with associated tricuspid valve mass.

At delivery, she required bag-mask ventilation for apnea. Physical exam revealed a macrosomic infant with macroglossia, erythematous, blanching vascular malformation overlying the glabella, short, upturned nose, and right posterior ear pits. A grade II/VI systolic ejection murmur was heard loudest at the left upper sternal border. Abdominal exam showed a firm mass in the left lower quadrant. Mild hemihypertrophy was noted in right upper and lower extremities. The remainder of her exam was unremarkable.

She required intubation and surfactant administration following delivery. Chest radiograph revealed cardiomegaly. Follow-up echocardiogram showed a severely dysplastic tricuspid valve with significant redundant, cyst-like tissue on the septal leaflet with prolapse and normal biventricular function. She was started on prostaglandin infusion, inhaled nitric oxide, and dopamine for pulmonary hypertension.

She weaned off prostaglandins on hospital day (HD) 3 as the lesion was not ductal-dependent. Multiple repeat echocardiograms exhibited persistence of the valve mass. Subsequent evaluations revealed nephromegaly, midline facial hemangioma, and hepatomegaly. Methylation panel confirmed the diagnosis of Beckwith-Wiedemann Syndrome. Karyotype confirmed 46 XX, and chromosomal analysis showed uniparental disomy of chromosome 11. She was extubated and weaned to room air on HD 17. She was discharged home on HD 30 and did not require cardiac surgery.

415 AN INSTITUTIONAL REVIEW OF INTESTINAL MALROTATION AND MIDGUT VOLVULUS OVER LAST 7 YEARS

C Boyle*, JM Davidson, AJ Talati. *University of Tennessee Health Science Center, Memphis, TN*

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Purpose of study Midgut volvulus is a surgical emergency for a newborn. However, symptoms could be subtle and progression rapid, leading to high morbidity if diagnosis is delayed. We aimed to review cases of malrotation with or without volvulus at our institution and identify differentiating characteristics of infant with midgut volvulus.

Methods used A retrospective review was done of all the cases of malrotation with or without midgut volvulus at our children's hospital over a 7 year period. Patients were identified from the NICU database and charts reviewed to record clinical characteristics and outcomes. Comparisons were made between group with volvulus and only malrotation.

Summary of results There were 51 infants diagnosed with malrotation in this time. Of these 51 babies, 23 (45%) had midgut volvulus at the time of surgery. Higher proportion of males had volvulus (82% vs 46%, $p=0.01$). The primary presenting symptom of the volvulus cases was bilious emesis with 16/23 (69%) compared to only 2/28 (7%) in the malrotation group ($p=0.0001$). There were no cases of volvulus in any infant less than 31 weeks gestation at birth and only 3 (10%) cases of malrotation in this age group. The mean time from symptom onset to surgery for the volvulus group was 0.81 days with a range 0–2 days. The majority of the malrotation group had no gastrointestinal symptoms at presentation and

were only found incidentally on screenings or at time of surgery for other reasons, 23/28 (82%, $p=0.0001$). Also, 22/28 (79%) patients in the malrotation group had other clinical associations or anomalies compared to only 4/23 (17%) volvulus patients ($p=0.0001$).

Conclusions As expected, bilious emesis was the most common presenting symptom of volvulus in neonates. In the volvulus group, the time from symptoms onset to surgery varied and was not related to bowel loss or death. Isolated malrotations are usually associated with other anomalies and are incidental findings. In our cohort, volvulus was more likely in males, closer to term gestation and as an isolated anomaly.

416 NEONATAL OUTCOMES OF VERY LOW BIRTH WEIGHT INFANTS BORN TO TEENAGE MOTHERS

SK Chilakala*, R Pollack, R Dhanireddy. *UTHSC, Germantown, TN*

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Purpose of study Adolescent pregnancy (10–19 years) is a matter of public concern as it affects maternal health, infant outcomes and will have negative socioeconomic consequences. Birth rates in 15–19 years old for United States in 2016 is 20.3/1000 females. For the State of Tennessee though it has declined by 51% between 2008 and 2017, It still stands 10th highest at 28/1000. Shelby county, the rate is 32/1000. Previous studies reported adverse maternal and neonatal outcomes associated with teen pregnancy, but none specifically explored the neonatal outcomes of the VLBW infants born to teenage mothers. We sought to determine the prevalence of VLBW infant births in adolescent population in a Level 3B NICU in Shelby County, TN and evaluate whether neonatal outcomes of VLBW infants born to mothers ≤ 19 years old are any different from outcomes of the infants born to mothers > 19 years of age.

Methods used VLBW infants delivered at our facility between January 2012 until December 2017 and admitted to the NICU were identified from the data base. Mothers age obtained from social work data. Retrospective review of all VLBW infants was conducted. Demographic information, neonatal outcomes at discharge were recorded and compared between the two groups. Chi-square, Fisher exact test and t tests were used to compare the two groups

Summary of results A total of 958 VLBW infants were admitted to the NICU between January 2012 and December 2017. = 124 of these infants (12.9%) were born to mothers who are ≤ 19 years of age. Demographic and obstetric characteristics of both groups were analyzed. Mortality and morbidity outcomes at discharge of both groups were analyzed. There were no statistically significant differences in demographic and obstetric characteristics between the two groups. Although there was a trend of more male births (50.3% vs 41%) in mothers > 19 years of age, it was not statistically significant. Infants born to mothers ≤ 19 years old were more likely to have increased oxygen requirement at 28 days of life compared to the other group (36.8% vs 20.5% respectively. $p < 0.05$) though the oxygen requirement at 36 weeks did not show significant difference. All other outcomes were similar

Conclusions VLBW infants of younger age (≤ 19 years) mothers have similar morbidity and mortality outcomes to discharge during the initial NICU hospitalization

417

RED BLOOD CELL DISTRIBUTION WIDTH (RDW) AS MARKER FOR NEONATAL SEPSIS

M Choe*, M Zayek, F Eyal. *University of South Alabama, Mobile, AL*

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Purpose of study Neonatal sepsis is a major cause of mortality in extremely low birth weight (ELBW) infants. Early detection is imperative for improved outcome, but is challenging due to non-specific clinical signs and symptoms. Further, inflammatory markers for neonatal sepsis for ELBW infants are limited and often misleading. Elevated red blood cell distribution width (RDW) has recently been found to be a marker for higher risk of sepsis or death in adults, children, and term infants. Yet there is no information available about association of RDW and neonatal sepsis in ELBW infants.

Objective To determine whether RDW is elevated during early stages of sepsis in infants with gestational age (GA) <25 weeks and birth weight (BW) <1000 g.

Methods used We identified neonates who had a culture positive sepsis in the first 10 days of life (DOL) and were born between January 2002 and September 2017, with GA <25 weeks and BW <1000 g (n=28). We matched these infants (1:1) with non-infected infants for BW and history of a recent blood transfusion. Because RDW values increase during the first week of life, we compared RDW values obtained on the day of sepsis with RDW from the matched controls obtained on the same DOL, using Whitney rank sum test. If RDW values were missing on the study day, in the matched control group, we inserted predicted values from a linear regression equation from all serial RDW values in the control infants. Other inflammatory biomarkers were also analyzed.

Summary of results RDW was significantly elevated in the sepsis group (21.5; 95% CI, 19.6–23.3) compared to the matched control (18.2; 95% CI, 17.2–19.2), $p<0.01$. While C-reactive protein was also higher in the sepsis group compared to control, immature to total neutrophil ratio, and neutrophil to lymphocyte ratio were not different between the groups.

Conclusions Higher values of RDW can be a potential marker of sepsis in the ELBW infant. Further studies are warranted to determine its predictive role in neonatal sepsis.

418

USE OF CONSULTATIVE SERVICES FOR DELIVERIES AT PERIVIALE GESTATIONAL AGES

T Coleman*, J Desai, A Hayslett, B Ferguson, M Tollison, M Famuyide. *University of Mississippi Medical Center, Jackson, MS*

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Purpose of study No data to date reports on the use of consultative services such as social work (SW), chaplaincy (CH), or palliative care surrounding the delivery of infants at periviable gestational ages (GA). As part of a larger study exploring decisions surrounding periviable births, we explored how these services are currently used.

Methods used Inborn periviable births in 2011–2015 at 6 centers in the United States were included in the study. Mothers with living fetuses delivered between 22 0/7 and 24 6/7 weeks GA were included unless there were major congenital

	Total			Before Birth			After Birth		
	22 Weeks N=128	23 Weeks N=181	24 Weeks N=261	22 Weeks	23 Weeks	24 Weeks	22 Weeks	23 Weeks	24 Weeks
Any Consultative Service	105 (82%)	153 (84.5%)	222 (85%)						
Social Work Consultation									
Involvement, N (%)	89 (69.5%)	133 (73.5%)	200 (76.6%)	17 (13.3%)	35 (19.3%)	74 (28.4%)	72 (56.3%)	100 (55.2%)	126 (48.3%)
First SW visit and birth, Median (IQR), hours*	-14 (-25, -2)	-14 (-29, 2)	-16 (-36, 4)	23 (1.52)	26 (16.8)	73 (9.14)	-16 (-37, 3)	-19 (-34, 10)	-30 (-46, 18)
First SW visit and death, Median (IQR), hours*	-9 (-20, 1)	-3 (-16, 9)	55 (12.619)						
Chaplain Consultation									
Involvement, N (%)	49 (37.5%)	66 (36.5%)	62 (23.8%)	8 (6.3%)	18 (9.9%)	26 (10%)	49 (38.3%)	48 (26.5%)	36 (13.8%)
First CH visit and birth, Median (IQR), hours*	-36 (10, 2)	-4 (30, 1)	-9 (45, 24)	11 (2.7)	29 (15.7)	42 (15.5)	-4 (-17, 1)	-18 (-34, 3)	-31 (-58, 3)
First CH visit and death, Median (IQR), hours*	-9 (-16, 0.8)	-1 (-17, 5)	1 (8.66)						

Abstract 418 Figure 1

anomalies, life-limiting genetic conditions, fetal demise soon after admission, or missing data.

Summary of results 570 periviable deliveries were analyzed from 498 mothers (66 twin and 6 triplet gestations). GA at delivery was 22 weeks (22%), 23 weeks (32%), and 24 weeks (46%). Consultative services were involved in 85% (480/570) of all deliveries; SW and CH were involved in 75% and 31% of births, respectively. Caucasian mothers were less likely to get a SW (63% vs 79%, $p=0.002$) consult and more likely to have a CH visit (37% vs 26%, $p=0.03$) as compared to African American mothers. Single mothers without family support and employed mothers were least likely to receive a SW consult ($p=0.000$).

Involvement was significantly lower before delivery (SW: 21% vs 52%, $p=0.000$; CH: 9% vs 22%, $p=0.000$). Consults were more likely after death at 22–23 weeks GA but before death at 24 weeks GA. When parents made a choice for ‘resuscitation’ as compared to physicians, they were more likely to get a SW consult (79% vs 70%, $p=0.000$) and a CH visit (37% vs 24%, $p=0.000$).

Conclusions Variability exists in the use of consultative services in our cohort. Paucity of psychosocial, spiritual, and palliative care support prior to delivery suggests inadequate perinatal anticipatory guidance and support. Racial and economic disparities in consultation are concerning and need further exploration.

419

UNUSUAL CASE OF NECROTIZING ENTEROCOLITIS

FM Dawoud*, JG Lawson, DS Shah. *Quillen COM East Tennessee State University, Smyrna, TN*

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Case report Spontaneous intestinal perforation (SIP) usually happens within the first week of life. Major risk factors include extreme prematurity and prophylactic indomethacin along with delayed onset of feeding and chorioamnionitis. Necrotizing enterocolitis (NEC) is characterized by ischemic necrosis of intestinal mucosa and its presentation is inversely related to gestational age with peak incidence between 28–32 weeks post menstrual age. Other risk factors beside prematurity include hypoxia, sepsis, and blood transfusion.

To our best of knowledge there is no literature discussing necrotizing enterocolitis that occurs after SIP in extreme pre-term infants.

A 24.1 week infant was born via stat C/S to a 20 yo G2P0 who presented with ROM for >24 hours with purulent fluids and maternal fever. This pregnancy was complicated by active herpes and chorioamnionitis. The neonate received resuscitation measures upon birth including compressions, intubation, and surfactant. Neonate was then diagnosed with isolated perinatal bowel perforation on the 8th day of life following changes in abdominal discoloration and distention. Triple

antibiotic therapy of Vancomycin, Cefepime and Flagyl was administered and a JP drain was then placed in RLQ. Due to critical condition, small bowel follow through was not conducted until the 15th DOL. Clinical picture was complicated by decompensation, worsening acidosis, hypotension, IVH, and elevated WBC count. This revealed development of significant free intraperitoneal air. Exploratory Laparotomy was conducted with abdominal irrigation and JP drain removal on the 18th DOL revealing healed perforation and normal bowel wall. The neonate was stabilized and resumed trophic feeding which was gradually advanced until the 35th DOL when condition began deteriorating with evidence of abdominal discoloration and distention with emesis. NEC was diagnosed per abdominal x-ray on 36th DOL. Infant's condition progressed worsened and died in 36 hours despite aggressive medical management with multiple organ failure.

This presents an unusual case where isolated bowel perforation and subsequent management with laparotomy occurred previous to development of necrotizing enterocolitis. SIP is not considered risk factor for NEC but whether this case illustrates it could be risk factor for NEC.

420 RELATIONSHIP BETWEEN MATERNAL CHORIOAMNIONITIS AND RETINOPATHY OF PREMATURITY

S Debnath*, H Hong, P Giannone, J Bauer. *University of Kentucky, Lexington, KY*

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Purpose of study Retinopathy of prematurity (ROP) is a major morbidity in preterm infants with debilitating consequences including blindness. Prenatal infection has been recognized as a contributor to preterm birth and chorioamnionitis can be identified via clinical symptoms, or via placental histopathology after birth. Several postnatal factors enhance ROP risks but less is known about prenatal factors. Defining relationships between prenatal inflammation and postnatal morbidities may help direct therapeutics for preterm infants.

Methods used Design/Methods: Medical records of 105 infants born <30 wks from 2015 to 2017 were studied including demographics, birth anthropometric measurements, and postnatal morbidities. Maternal clinical data and placental histology were also collected. ROP assessment was based on ophthalmologist examination using standard guidelines, and evaluated at three levels: any stage of ROP, ROP stage 2 or greater, ROP requiring intravitreal Avastin and/or Laser photocoagulation. Statistical comparisons were done using the Fishers exact tests.

Summary of results Of 105 preterm infants, there were 11 cases of clinical chorioamnionitis and 41 cases of histopathologic chorioamnionitis; 100% of the clinical diagnoses were confirmed by placental pathology. Chorioamnionitis defined by placental pathology showed a ~3 fold increase in occurrence of any stage of ROP in the postnatal period in premature infants ($p < 0.05$); ROP stage 2 or greater or ROP requiring treatment were not different. Prolonged rupture of membrane was strongly associated with histopathologic ($p < 0.001$), but not clinically identified chorioamnionitis. In contrast, clinical chorioamnionitis was not statistically associated with any aspect of ROP

Conclusions The timely and accurate identification of chorioamnionitis is important but challenging and we found that the clinical definition of chorioamnionitis missed many pathologically detected cases. The insensitivity of clinical identification may lead to an underestimation of inflammation as a contributor to ROP risks. Further investigation of prenatal inflammation-driven angiogenesis pathways and its impact on postnatal ROP risks, are clearly warranted. Furthermore, taking placental pathology findings into consideration for early management of premature infant care may have value for this high-risk patient group.

421 PULMONARY SEQUESTRATION: A RARE CAUSE OF NON IMMUNE HYDROPS AND RECURRENT PLEURAL EFFUSION

K Dolma*, J Philips. *University of Alabama at Birmingham, Birmingham, AL*

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Introduction Non immune hydrops is associated with many diseases and an effort to determine etiology should be made. We report a case of antenatally diagnosed non-immune hydrops with recurrent pleural effusion in the neonatal period due to pulmonary sequestration.

Case presentation Baby girl was born at 31.3 wks to a 25 y/o black mom with CHTN. Pregnancy was complicated by prenatal diagnosis of hydrops fetalis, polyhydramnios and a fetal left pleural effusion. Mother was B+ and Ab screen was weakly positive for anti E. FISH, CMV, toxoplasma, syphilis, HSV, and Kleihauer Betke tests were negative. She had amniocentesis, fetal thoracentesis, and thoraco-amniotic shunt placement at 30.3 wga. After 5 days, recurrent polyhydramnios with severe left pleural effusion was seen with shunt dislodgement. Concern for fetal lung damage from persistent effusion led to induction. At birth, intubation and immediate left thoracentesis were done. Apgar scores were 1, 5 and 7. Workup including EBV, Parvo B19, CMV, and echo was negative. Infant was extubated on DOL 7. She had reaccumulation of the effusion on DOL 28. Thoracentesis and chest tube placement were done. Fluid analysis was consistent with transudate. CT showed a left extralobar pulmonary sequestration supplied by arterial feeder from descending thoracic aorta, consistent with the cause of recurrent pleural effusion. Infant improved and chest tube was removed after 7 days. Pulmonology and pediatric surgery were consulted and the decision was made to follow as an outpatient with surgical intervention in future. Infant was stable on room air and was discharged on DOL 62.

Discussion Pulmonary sequestrations are nonfunctional masses of lung tissue that lack normal communication with the tracheobronchial tree and receive blood supply from systemic circulation. Routine prenatal ultrasound screening may allow for the early detection of pulmonary sequestration. In the majority of cases, the lesion regresses during the course of gestation. Occasionally, antenatal complications such as massive pleural effusion or even hydrops can develop requiring fetal intervention.

Conclusion Pulmonary sequestration causing non immune hydrops and recurrent pleural effusion is rare. Appropriate diagnosis will help in optimal management.

422 NOVEL BIOMARKERS FOR LOW CARDIAC OUTPUT SYNDROME IN INFANTS FOLLOWING BYPASS

S Drennan*, EG Szyld, K Burge, J Eckert, A Mir, AK Gormley, R Schwartz, S Daves, J Thompson, H Burkhardt, H Chaaban. *OUHSC, OKC, OK*

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Purpose of study Low cardiac output syndrome (LCOS) is a common complication of cardiopulmonary bypass (CPB). This study aims to assess the perioperative changes in markers of inflammation, specifically extracellular histones, in infants undergoing CPB, and to determine if higher levels correlate with the development of LCOS. Histones are biomarkers and therapeutic targets in pathological conditions where inflammation and thrombosis occur. Whether histones are released during CPB and if they play a role in LCOS is unknown.

Methods used Observational pilot study designed as a convenience sample. Infants <6 mos of age with congenital heart disease were divided into groups based on anatomical defect: group 1 is a non-surgical control (n=5); group 2 are surgical patients without bypass (n=4), group 3 are surgical patients with bypass (n=21). Baseline markers of inflammation were measured in group 1. In surgical patients, histones and cytokines were drawn serially: preoperatively, intraoperatively, on PICU admission, 6, 12, and 24 hours later. Histones were measured by Cell Death Detection ELISA.

Summary of results Group demographics were comparable. LCOS incidence was 47%. Histone levels were significantly elevated post operatively over serial time points in groups 2 and 3 ($p < 0.0001$). Histone levels were significantly elevated at 6 and 24 hours post-operatively in group 3 vs 2

Abstract 422 Figure 1 Cell death ELISA by group

Abstract 422 Figure 2 Cell death ELISA-LCOS

($p=0.03$ and $p=0.01$, figure 1). Neonates with LCOS had higher histone levels at all time points, peaking at 6 hours ($p=0.0065$; figure 2).

Conclusions Infants undergoing CPB have elevated histone levels at 6 and 24 hours following PICU admission compared to infants not requiring bypass. Patients with LCOS have consistently higher histone levels across all time points, most significantly at the six-hour time point which coincides with the onset of LCOS. Future studies are needed to confirm this association and determine if histones can be used as biomarkers and therapeutic targets in LCOS.

423 A COMPARISON OF METHODS OF DISCONTINUING NASAL CPAP IN PREMATURE INFANTS <30 WEEKS GESTATION- A FEASIBILITY STUDY

BL Duyka*, CC Beaulieu, AM Khan. *University of Texas- Houston, Houston, TX*

10.1136/jim-2018-000974.421

Purpose of study Nasal continuous positive airway pressure (CPAP) mode of respiratory support is widely used to support premature infants. While there is significant evidence justifying the use of CPAP, very little is known about how and when to wean off CPAP. In clinical practice, CPAP is either stopped regardless of the pressure or the pressure is gradually reduced before stopping CPAP, based on individual preferences. We hypothesize, based on the review of the literature and clinical experience, that there is no difference in the number of days on CPAP between the two groups.

Methods used This is a randomized control comparative effectiveness trial of discontinuation of CPAP with or without weaning CPAP pressure in patients with gestational age <30 weeks. Patients were randomized once the provider made the decision to discontinue CPAP and met pre-specified criteria. In the wean arm of the study, the CPAP pressure was decreased by 1 from 6 to 4 daily, while in discontinuation arm, CPAP was stopped from 6. If the patients met failure criteria, they were placed back on CPAP for one week before trying again. The study stopped 28 days from randomization or if the subject failed discontinuation twice.

Summary of results 139 eligible patients have been screened, and 42 infants completed the study. Both groups were similar except for higher birth weight and randomization weight in the wean group as compared to the no wean group (988 gm vs 860 gm) and (1450 gm versus 1190 gm). Duration of mechanical ventilation was longer in the wean than in the non-wean group (16d vs 12d, $p=0.054$). Patients in the wean arm were more likely to succeed the first time than the no wean arm (10 vs 5, $p=0.087$). There were no significant differences in number of attempts to come off CPAP, study failure, post menstrual age at discharge, length of stay, time to full feeds, or weight gain.

Conclusions Our study shows that CPAP can be discontinued in premature infants without weaning the pressure. Weaning pressure may expose the infants to prolonged duration of mechanical ventilation. These results need verification with a larger definitive trial.

424

NEWBORN STATE SCREENING FOR CONGENITAL HYPOTHYROIDISM IN THE NICU: ARE WE USING THE RIGHT SCREEN FOR PREMATURE INFANTS?

TO Findley*, A Shah, AM Khan. *University of Texas Health Science Center at Houston, Houston, TX*

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Purpose of study Newborn screening (NBS) allows for early identification of congenital hypothyroidism (CH). In Texas, two NBS are mandated per newborn with primary thyroxine (T4) used for CH screening. In neonates, the limitations of primary TSH in NBS have been well-described. The purpose of our study is to evaluate the role of NBS with T4 in the neonatal intensive care setting.

Methods used We reviewed all NBS results collected from a single, large, Level IV neonatal intensive care unit (NICU) during a nine-year period (2007–2016), omitting those who had less than two state screens collected to exclude healthy newborns discharged soon after birth (n=6100). We also reviewed the charts of infants medically treated for CH during the same time period. Patients were excluded from analysis if treatment was started at an outside hospital or if NBS results were incomplete. Duration of medical therapy was tracked in discharged patients who followed up in our pediatric endocrinology clinic.

Summary of results The overall incidence of treated hypothyroidism in our population was 1:103, with higher incidence associated with lower birth weight. Among infants treated for CH (n=59), 42 (0.71) and 51 (0.86) had abnormal first and second CH NBS results, respectively, and 35 (0.59) had abnormal results for both NBS. Primary T4 NBS at birth showed sensitivity and specificity of 0.74 and 0.85, respectively; second NBS showed improved sensitivity and specificity of 0.86 and 0.87, respectively. The positive predictive value on initial NBS was 0.046 and 0.058 on the second NBS.

Conclusions Abnormal T4 NBS is common for preterm neonates due to transient hypothyroxinemia of prematurity. Frequent false positives make interpreting NBS results challenging for clinicians as published guidelines and recommendations target healthy, term newborns and are directed by TSH levels. These findings suggest that the NBS is not an ideal screening tool for CH in NICU patients, and premature and ill neonates require a separate screening modality.

Population Health and Precision Medicine Joint Plenary Poster Session and Reception 4:30 PM

Thursday, February 21, 2019

425

RESPIRATORY FAILURE IN A NEONATE WITH A NOVEL GENE VARIANT OF LETHAL OSTEOGENESIS IMPERFECTA TYPE II

J Gallois*, B Barkemeyer, R Zambrano. *LSUHSC School of Medicine, New Orleans, LA*

10.1136/jim-2018-000974.423

Case report Osteogenesis Imperfecta (OI) is a heterogeneous connective tissue disorder caused by mutations in the *COL1A1*

and *COL1A2* genes. These genes code for type I procollagen, which is a primary matrix protein for bone, dentin, sclerae, and ligaments. There is a classification scheme for OI in which those with type II are the most severely affected, often with in utero or perinatal death. Fetuses with OI type II are characterized by short limbs, clinical and radiographic evidence of severe osseous fragility and defective ossification. Respiratory insufficiency can result from underdeveloped lungs and an abnormally small thorax. Biochemically regulated processes, as well as mechanical factors, may impede pulmonary development in cases of OI type II.

We present a case of a full term female infant with perinatal suspicion of OI.

At birth, the patient required high flow nasal cannula (HFNC) for mild respiratory distress. Postnatal exam and bony radiography were consistent with OI type II. A skeletal survey showed extensive changes of OI with innumerable bilateral rib cage and long bone fractures. She was transferred on day of life (DOL) 23 to a tertiary care unit on HFNC respiratory support for poor feeding. Respiratory distress acutely progressed on DOL 30 requiring non-invasive positive pressure support. Given the clinical diagnosis of OI type II, the family chose not to pursue more aggressive interventions. The baby died on day of life 35 of respiratory failure.

Subsequent results from DNA sequencing revealed a likely pathogenic variant in the *COL1A1* gene (c.1382c; p.Gly461aIs variant). This variant has not been described previously in the medical literature or in gene-specific databases, and is not present in general population databases. It disrupts a sequence motif of the collagen triple helix predicting impaired collagen function. Other variants at the codon and surrounding glycine substitutions in this exon have been classified as pathogenic. Given the clinical findings we feel this variant is clinically significant and responsible for the phenotype, and should be classified as pathogenic.

426

INCIDENCE OF ACUTE KIDNEY INJURY AND OUTCOMES IN NEONATES WITH GASTROSCHISIS – SINGLE CENTER FIVE-YEAR EXPERIENCE

¹PM Garg*, ¹J Patel, ¹B Ferguson, ²DJ Askenazi, ¹M Famuyide. ¹UMMC, Jackson, MS; ²UAB, Birmingham, AL

10.1136/jim-2018-000974.424

Purpose of study The purpose of the study is to determine the incidence of AKI in infants with gastroschisis and describe outcomes based on AKI status

Methods used A single center retrospective cohort review of 62 patients was done in neonates undergoing gastroschisis repair between 2010 and 2015. The incidence of AKI was determined using the Kidney Disease: Improving Global Outcomes (KDIGO) Clinical Practice Guideline.

Data on urine output and total fluid intake on postnatal days 1, 2, 3, 4, 7 and 14, timing of silo placement and surgical closure were collected. Outcome measures such as mortality, length of stay, TPN days, direct hyperbilirubinemia, time to full feeds and culture-positive sepsis were collected. A p-value below 0.05 was considered significant. Data was analyzed by using JMPv13.1®.

Summary of results AKI was seen in 11% of gastroschisis cases in the first week of life. The median GA (IQR) was 36.2 (27.1–34.8) weeks and the mean BW was 2.3 Kg (SE

Abstract 426 Table 1 AKI vs Non AKI group

	No AKI n=55	AKI n=7	P value
Gestational Age Mean	35.5 (SE 0.27)	36.04 (SE 0.77)	0.55
Birth Weight (kg)	2.33 (SE 0.076)	2.32 (SE 0.21)	0.96
SCr Day 1 (Post silo mean)	0.75 (SE 0.02)	1 (SE 0.05)	0.0001
SCr Day 2	0.63 (SE 0.02)	1.1 (SE 0.06)	<0.0001
SCr Day 3	0.55 (SE 0.02)	0.92 (SE 0.05)	<0.0001
SCr Day 4	0.48 (SE 0.02)	0.79 (SE 0.05)	<0.0001
SCr Day 7	0.43 (SE 0.01)	0.54 (SE 0.04)	0.0228
SCr Day 14	0.37 (SE 0.01)	0.46 (SE 0.04)	0.06
Mortality	3/55 (5.45%)	1/7 (14.29%)	0.38
Length of Stay (Days)*	67.7 (SE 55.9)	120.6 (SE 37.6)	0.04
Positive Blood culture	10 (18.18%)	1 (14.29%)	1
Time to Full feeds (Days)*	41.1 (SE 7.28)	19.3 (SE 6.17)	0.47
Mean Direct bilirubin (mg/dl)*	2.2 (SE 0.53)	6 (SE 1.57)	0.02
No of TPN days*	58.5 (SE 8.2)	103.6 (SE 24.3)	0.08

0.07 kg). Fifty seven percent of patients were delivered by C-section and were mostly inborn (87%). All patients had initial reduction via Silo method and median age of surgical closure was 4 (IQR 3–7) days.

The AKI group had a significant increase in serum creatinine during the first week of life as compared to the non-AKI group (p 0.001) [table 1]. The length of stay (p 0.04) and direct bilirubin (p 0.02) levels were significantly higher in the AKI group. In the AKI group, infants with early closure (<3 days) had a statistically significant longer length of stay compared to late (>3 days) closure (p 0.012). However, there was no significant difference in the time to full feeds and culture positive sepsis between both groups.

Conclusions In this cohort of neonates with gastroschisis, stage 1 AKI was seen in 11% of patients. In patients with AKI, early closure (<3 days) was associated with a longer length of stay. Large prospective multi-centric studies are needed to define patient specific risk factors for AKI in gastroschisis patients.

427 MONITORING READINESS FOR WEANING FROM NASAL CPAP IN PRETERM INFANTS USING OXYGEN SATURATION HISTOGRAMS: A CASE CONTROL STUDY

SJ Gentle*, N Ambalavanan, C Travers, WA Carlo. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.425

Purpose of study Continuous positive airway pressure (CPAP) is widely used to support preterm infants with respiratory distress. Identification of readiness for weaning from CPAP can balance benefits and risks such as air leak syndromes, impaired venous return, and nasal septal injury. This case control study's objective was to determine if oxygen saturation histograms in preterm infants are associated with successful weaning from CPAP in preterm infants.

Methods used This was a single center matched case control study in infants <1250 g at birth receiving $\leq 30\%$ FiO₂ on CPAP during the first postnatal week using a case: control ratio of 1:2. Control infants were infants who remained off

Abstract 427 Figure 1 Oxygen saturations in 24 h prior to CPAP discontinuation

Abstract 427 Table 1

	Control (n=24)	Cases (n=12)
Gestational age, weeks $\pm$ SD	28 $\pm$ 1	28 $\pm$ 1
Weight, grams $\pm$ SD	899 $\pm$ 217	1019 $\pm$ 199
Antenatal corticosteroids, n (%)	19 (79)	9 (75)
Mechanically ventilated, n (%)	9 (38)	4 (33)
Surfactant, n (%)	9 (38)	4 (33)
Days following birth when trialed off CPAP, days $\pm$ SD	4 $\pm$ 1	4 $\pm$ 1
FiO ₂ prior to trial off CPAP $\pm$ SD	23 $\pm$ 3	23 $\pm$ 2

n = number; SD = standard deviation; CPAP = continuous positive airway pressure; FiO₂ = fractional inspired oxygen concentration

CPAP for seven consecutive days following CPAP discontinuation, and cases were infants placed back on CPAP within seven days of being taken off CPAP. Individuals were matched based on gestational age at birth (± 10 days) and frequency matching within each group included antenatal corticosteroid, surfactant, and mechanical ventilation exposure. Prospectively collected 24 hour histograms of oxygen saturations prior to CPAP discontinuation were compared between cases and controls.

Summary of results Over a 12 month monitoring period, 36 infants met inclusion criteria. The rates of exposure to antenatal corticosteroids, surfactant, and mechanical ventilation did not differ between cases and controls (table 1). Oxygen saturation histograms between cases and controls differed with controls demonstrating more time at saturations 95–97 and 97%–100% ($p < 0.05$) compared to cases (figure 1).

Conclusions Histogram monitoring of achieved saturations in preterm infants with respiratory distress receiving CPAP may enable care providers to predict readiness to wean from support.

428 PARENTERAL NUTRITION IN EXTREMELY LOW BIRTH WEIGHT INFANTS: INCREASED PHOSPHORUS AND EARLY POTASSIUM DELIVERY

¹SR Glenn*, ²C Finch, ³DM DellaValle, ⁴S Taylor. ¹Medical University of South Carolina, Charleston, SC; ²Virginia Commonwealth University Medical Center, Richmond, VA; ³Marywood University, Scranton, PA; ⁴Yale University, New Haven, CT

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Purpose of study To determine if increased parenteral phosphorus delivery and early parenteral potassium delivery resulted in fewer IV replacements required for hypophosphatemia and hypokalemia.

Methods used This study retrospectively reviewed extremely low birth weight (ELBW) infants who received increased

parenteral nutrition (PN) phosphorus (1.2 mmol/100 mL; calcium-to-phosphorus molar ratio 1:1) and potassium (1.4 mEq/100 mL) by 24 hours of life (increased-electrolyte (IE) group; born 2016). PN electrolytes, serum electrolyte lab values, and number of IV phosphorus and potassium replacements were compared with a standard-electrolyte (StE) group (born 2015) given 0.97 mmol/100 mL PN phosphorus (calcium-to-phosphorus ratio 1.2:1) and no potassium in the first 24 hours of life. Data analysis used descriptive statistics and repeated measures ANOVA. P value<0.05 was considered statistically significant for main effects and P value<0.2 for interaction effects.

Summary of results Thirty-three ELBW infants were included in this study (IE n=13, StE n=20). There were no significant differences between groups in birth weight, gestational age, or gender. There were no differences between groups in the number of IV phosphorus and potassium replacements. The IE group did receive more PN potassium without subsequent hyperkalemia. A significant effect of time was observed for PN phosphorus and calcium intakes in the IE group (p<0.001) compared to the StE group. A significant group by time interaction was observed for PN phosphorus (p=0.018). Significant time effects were observed for both serum calcium and potassium (p<0.001 and p=0.03, respectively).

Conclusions Early delivery of PN phosphorus and potassium was safe for ELBW infants receiving early PN protein and frequent monitoring for deficiency was needed. In our unit, PN phosphorus was increased to 1.2 mmol/100 mL, and PN potassium safely provided by 24 hours of life.

429 EVALUATING THE NEED FOR A DELIVERY ROOM CHECKLIST IN EXTREMELY LOW BIRTH WEIGHT DELIVERIES VIA SIMULATION

¹AB Gonzalez*, ¹RC Farner, ^{1,2}MM Vasquez. ¹University Hospital, San Antonio, TX; ²Long School of Medicine at UT Health San Antonio, San Antonio, TX

10.1136/jim-2018-000974.429

Purpose of study The Neonatal Resuscitation Program (NRP) has outlined equipment and team organization needed for the birth of an extremely low birth weight (ELBW) newborn. This includes not only a standardized equipment checklist, but also a pre-event time-out or briefing. The Vermont Oxford Network has advocated for the use of a delivery room check-list (DRCL) since 2010. In view of the above, two quality improvement questions were raised. In the context of fellow-led delivery room teams (DRTs): What degree of measurable team readiness is present at the birth of high risk ELBW infants? Would using DRCLs improve team readiness?

Methods used Six scenarios were developed to each simulate a high risk Cesarean section delivery of an ELBW newborn. The first three scenarios primary objective is to obtain baseline data about team preparation activities before the introduction of a DRCL in the subsequent three scenarios.

There are six drills per scenario (one fellow per drill). The delivery room team (DRT) consists of: one fellow (team leader), two NICU nurses, one NICU respiratory therapist, and one neonatologist. All DRT members are blinded to drill objectives except for the neonatologist. Four NRP recommended pre-delivery activities are used to help measure team readiness: team leadership, assignment of key roles and duties,

gathering equipment and medications, and anticipating ELBW thermoregulation needs.

Summary of results

1. We report the data of the first three scenarios (seventeen non-DRCL drills).
2. There are inconsistencies in performing various required tasks in all four measured groups.
3. Team 'order call-backs' are performed at a rate of 67% to 75%, but brake down to 43% when epinephrine is ordered.
4. There appears to be a 'dis-connect' between team leader orders given and team performance of required actions.

Conclusions In view of measured baseline data, the proactive use of a DRCL may be helpful in fellow-led DRT attendance of ELBW births.

430 INTESTINAL ALKALINE PHOSPHATASE AS A DIAGNOSTIC BIOMARKER FOR NECROTIZING ENTEROCOLITIS

¹M Heath*, ¹R Buckley, ²Z Gerber, ¹P Davis, ¹B Barkemeyer, ³M Good, ¹D Penn, ¹S Kim. ¹LSU Health Science Center, New Orleans, LA; ²Pediatrics, Las Vegas, NV; ³Washington University in St. Louis, St. Louis, MO

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Purpose of study Necrotizing enterocolitis (NEC) in premature infants is an inflammatory gastrointestinal disease with significant mortality and morbidity. Diagnosis and management is difficult due to nonspecific symptoms that overlap with other diseases and limited diagnostic methods for advanced NEC. Our hypothesis is aberrant intestinal alkaline phosphatase (iAP) biochemistry in stool is a biomarker for late and early stages of NEC in preterm infants, but is not correlated with sepsis.

Methods used Premature infants in 3 hospitals [median birth weight 1050 g and gestational age 28.4 wks] were enrolled in a prospective, observational study: 25 with advanced NEC (Bell stage II/III), 19 with early NEC (Bell stage I), and 92 non-NEC infants. Of these 136 patients, 26 were diagnosed with sepsis and 15 had non-GI infection. Using stool samples, relative amounts of iAP were determined by immunoassay; enzyme activity was fluorometrically detected. iAP measurements were evaluated using cross-sectional analyses.

Summary of results Compared to non-NEC patients, a 20-fold increase in iAP content was measured in patients with NEC at time of disease. iAP content, shed in the gut and detected by stool, was 1.02, 1.46, and 0.06-fold for Bell II/III, Bell I, and non-NEC samples, respectively. In contrast, iAP enzyme activity was lower in NEC with a decrease by 50% in Bell I and by 75% in Bell II/III. There was no significant difference for iAP measures of samples at the time of sepsis or matched control samples. For other infections, only iAP activity statistically differed with control samples. Area under the curve (AUC) values were 0.94, 0.96, 0.56, and 0.51 for iAP abundance at time of Bell I, Bell II/III, sepsis, and other non-GI infections; values for enzyme activity were 0.62, 0.77, 0.55 and 0.62, respectively.

Conclusions High fecal levels of iAP and low enzyme activity are reliable biomarkers specific for NEC. Preliminary sensitivity and specificity studies suggest iAP can serve as noninvasive diagnostic and monitoring biomarkers for NEC.

431 PRIMARY CUTANEOUS ASPERGILLUS IN A PREMATURE IMMUNOCOMPROMISED NEONATE

J Hendricks*. Emory University, Atlanta, GA

10.1136/jim-2018-000974.429

Introduction Primary Cutaneous Aspergillus (PCA) is seen in immunocompromised patients. In neonates, aspergillosis usually presents in its disseminated form, often with lethal outcomes. As such PCA is rarely a differential for a scab in a preterm neonate. We present an immunocompromised preterm neonate with PCA who presented with an eschar.

Case A female 30 w 5 d neonate being treated for congenital acute myeloid leukemia developed a dark, superficial scab on the right subscapular region without surrounding induration presumed to be a decubitus ulcer at age 10 weeks. She previously had a Steven-Johnson reaction with skin desquamation after her first cycle of chemotherapy and sepsis with respiratory failure treated with several courses of antibiotics and fluconazole prophylaxis.

Despite wound care, it enlarged. Detachment of the eschar revealed a dark base. The excised specimen showed budding hyphae identified as *Aspergillus flavus*. Imaging of the chest, brain and abdomen was negative for aspergillomas. Spinal fluid, blood and urine cultures were sterile.

She was treated empirically with liposomal amphotericin, then voriconazole and micafungin after identification of *A. flavus*. With resumption of chemotherapy, serum voriconazole levels fell, so amphotericin was resumed. The lesion re-epithelialized in 5 weeks. 12 weeks of antifungals were completed.

Discussion *A. Flavus* is a ubiquitous saprophytic mold. Reports of neonatal aspergillosis often present in the disseminated form and culminate in death. Premature infants are susceptible due to their immature immune system, multiple invasive procedures, and antibiotic use. The most common non-neonatal risk factor for PCA is immunosuppression. Our case is unique as she was both neutropenic from antineoplastic chemotherapy and premature.

Her lesion was thought to be a decubitus ulcer, but the rate of growth and her immunocompromised state raised the index of suspicion of PCA.

Voriconazole's use for aspergillosis treatment pediatric population is limited by lack of consensus on age-appropriate plasma levels. The species of *A. flavus* in our case was sensitive to amphotericin, which is well tolerated in children.

Conclusion We present this case as a reminder that a growing eschar in a premature neonate, may represent PCA especially in an immunosuppressed state. This should prompt systemic antifungals and biopsy.

Pulmonary and Critical Care Medicine
Joint Plenary Poster Session and Reception
4:30 PM
Thursday, February 21, 2019

432 STOP IN THE NAME OF STEWARDSHIP: SAFETY OF AN ANTIMICROBIAL 36-HOUR 'HARD STOP' IN THE NEONATAL INTENSIVE CARE UNIT

B Her*, MO Montgomery, J Cantey. University of Texas Health Science Center San Antonio, San Antonio, TX

10.1136/jim-2018-000974.430

Purpose of study Automatic discontinuation of empiric antibiotic therapy via the electronic medical record ('Hard stop') is an effective stewardship strategy to prevent unnecessary antibiotic use. Safety concerns of hard stops include inadvertent discontinuation, late culture positivity, and need for subsequent re-initiation of therapy. We investigate the safety of a 36 hour hard stop program in a level 4 neonatal intensive care unit (NICU).

Methods used Prospective cohort study of all infants admitted to the neonatal intensive care unit from 9/15 to present. Demographic, clinical, laboratory, and outcome data were collected for all infants. 36 hour hard stops were implemented beginning 9/1/17. Safety signals included 1) inadvertent discontinuation, 2) positive culture result after 36 hours incubation, and 3) restarting antibiotics within 72 hours of discontinuation.

Summary of results 1531 infants were admitted to the NICU during the study period; 1004 infants received ≥ 1 antibiotic course (66%). There were 1452 distinct antibiotic courses; 1145 before hard-stop implementation and 307 afterwards.

There were no inadvertent discontinuations during the study period.

Positive culture >36 hours: 200 (13.8%) of the 1452 antibiotic courses were associated with a positive culture from any site. 25 cultures were positive after 36 hours, 24 before hard-stop implementation and 1 after. The antibiotics had been continued empirically for 17 of these infants; cultures from 8 infants whose antibiotics had been discontinued yielded coagulase-negative *Staphylococcus* in the blood (n=5), *Capnocytophaga* sp. in the blood (n=1), *Enterococcus* in the urine (n=1), and *Enterobacter* in the urine (n=1). These infants had antibiotics restarted and all did well.

Restarting antibiotics within 72 hours: Before hard-stop implementation, antibiotics were restarted within 72 hours after 34 of 1145 courses (3%); 10 infants had sepsis (n=2) or necrotizing enterocolitis (n=8). After hard-stop implementation, antibiotics were restarted after 5 of 307 courses (1.6%), 1 infant had necrotizing enterocolitis.

Conclusions Implementation of a 36 hour hard stop was not associated with increased safety signals in a single-center NICU.

433 BREASTFEEDING DURATION MODERATES THE EFFECT OF PRENATAL STRESS ON INFANT ATTACHMENT CLASSIFICATION

¹SM Foroushani, ²S Drury, ²C Herman*. ¹Tulane University, New Orleans, LA; ²Tulane University School of Medicine, New Orleans, LA

10.1136/jim-2018-000974.431

Purpose of study High maternal prenatal stress (PNMS) is associated with decreased maternal sensitivity and may influence later maternal-infant attachment. One factor associated with higher maternal sensitivity is breastfeeding. To better understand how PNMS and later maternal behavior influence attachment this study tested the relation between PNMS and attachment at 12 months of age and whether breastfeeding moderated this relation.

Methods used Mothers were recruited during pregnancy and measures of prenatal stress were collected. PNMS was defined

using factor analyses and included the Prenatal Life Events, Rini Pregnancy Related Anxiety, Perceived Stress scale, and Edinburgh Depression and Chronic Strain Questionnaire. Breastfeeding duration was obtained by maternal report at 4 months of age. Attachment classification was determined with The Strange Situation Procedure at 12 months of age. Data analyses were performed in SAS 9.4. Linear regression examined the relation between maternal prenatal stress and attachment. Secondary analyses tested whether breastfeeding duration moderated the association between attachment and prenatal maternal stress.

Summary of results Of 153 dyads, 98 (64.1%) were classified as secure, 17 (11.1%) as avoidant, 25 (16.3%) as resistant, and 13 (8.5%) as disorganized. Controlling for race and socioeconomic status, higher PNMS was significantly associated with attachment classification ($p=0.02$). Duration of breastfeeding at 4 months also significantly predicted attachment security ($p=0.04$). Breastfeeding duration was a partial moderator of the association between PNMS and attachment.

Conclusions Mothers reporting high prenatal stress were less likely to have a secure attachment relationship with their infants. Breastfeeding duration through 4 months of age partially moderated this effect, suggesting that breastfeeding promotion, particularly in high risk mothers, may have benefits for maternal and child health that include strengthening the attachment relationship.

434 RACIAL DISPARITIES IN THE DEVELOPMENT OF ADIPOSITY AMONG PRETERM INFANTS

ML Jerome*, P Chandler-Laney, AA Salas. *University of Alabama at Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.432

Purpose of study At term-equivalent age, all preterm infants have higher percent body fat (%BF) than infants born at term. This difference in %BF is often the result of early exposure to enriched postnatal diets prescribed to promote rapid weight gain among preterm infants. Because rapid weight gain and obesity are more prevalent among blacks than among whites during early infancy, racial disparities in the development of adiposity expressed as %BF need to be explored. The purpose of this study was to compare %BF in two major racial groups using normative data as reference.

Methods used We analyzed data from a prospective study that assessed body composition in preterm infants using air displacement plethysmography (PeaPod®). After stratifying data according to race, we analyzed differences between mean %BF values of preterm infants at the time of hospital discharge and compared these differences to existing references of %BF in preterm infants. A linear regression analysis was performed to account for differences in baseline characteristics.

Summary of results We assessed body composition in 84 preterm infants, of which 47 were black and 37 were white. Mean birthweight was 1471 g and median gestational age was 30 weeks. In preterm infants assessed at the time of hospital discharge, mean %BF was 14.6 ± 3.6 (14.5 ± 2.9 in white infants and 14.6 ± 4.1 in black infants; $p=0.90$). The measured %BF at the time of hospital discharge was higher

than the expected %BF at equivalent postmenstrual age (mean difference: 4.7 ± 3.5 ; $p<0.05$). After adjustment for BW, GA, sex, and length of hospital stay, this difference between measured and expected %BF was not significantly higher among black infants compared to white infants (5.1 vs. 4.2; $p=0.28$).

Conclusions Black race is not associated with higher %BF at the time of hospital discharge in preterm infants. If racial disparities in body composition exist among former preterm infants, those differences may occur after hospital discharge. Both black and white preterm infants exposed to enriched postnatal diets develop higher than expected %BF by the time of hospital discharge.

435 LATE-ONSET OCHOBACTERUM ANTHROPI SEPSIS IN A NEONATE

N Kabani*, J Phillips. *University of Alabama, Birmingham, AL*

10.1136/jim-2018-000974.433

Case report This 2 mo old former 27 wk male was born via stat c section for NRFHT tones and pre-eclampsia. Mom received betamethasone prior to delivery. Baby required EEP for poor respiratory effort but was intubated for increasing O2 requirement and given surfactant. Baby weaned to oxygen-environment but was reintubated at 2 weeks. KUB revealed stage 2 NEC and antibiotics were initiated. He clinically worsened at 40 DOL, and blood culture grew *Candida albicans*; he was started on amphotericin B. At 50 DOL, baby's blood cultures from PICC grew GNR; Zosyn and Tobramycin were initiated. Blood cultures speciated as *Ochrobacterum anthrophi* in two sets of cultures on two separate dates; it was sensitive only to meropenem. He received a 14 day course of meropenem and cleared cultures. Echo showed no signs of endocarditis. He remains critically ill on HFOV, dopamine and stress-dose hydrocortisone.

Ochrobacterum anthrophi is a rare bacterium for infection in humans and the typical course is not well described. There are very few case reports of *Ochrobacterum anthrophi* in children, fewer in neonates. It belongs to the *Bucellaceae* family and is an opportunistic and nosocomial pathogen. It is an aerobic, oxidase-positive, urease-positive, Gram-negative, motile, non-lactose-fermenting bacillus, formerly known as 'Achromobacter group Vd'. Most human cases are due to line infections and tend to be in elderly and immunocompromised populations. It can cause many complications such as osteomyelitis, endocarditis, pancreatic abscess, UTI, meningitis, and pelvic abscess in addition to line infections. It is rare but tends to present in patients with severe underlying disease; it is usually of very low virulence. It is identified via blood culture. If suspected, one must cover empirically for gram negative rods until speciation. It is widely resistant to chloramphenicol and all beta lactams except imipenem and meropenem. It can be susceptible to Bactrim, gentamicin and fluoroquinolones. Once identified, the physician must look for other sources of infection as the cause of bacteremia such as endocarditis, osteomyelitis, and abscesses. Once these have been ruled out, a course of treatment can be decided. Given how ill this infant was, he received a 14 day course for bacteremia.

436 INFANTS OF DIABETIC MOTHERS: GROWTH AND FEEDING OUTCOMES

¹HD Kelley*, ¹JC Newman, ²S Taylor. ¹Medical University of South Carolina, Charleston, SC; ²Yale School of Medicine, New Haven, CT

10.1136/jim-2018-000974.434

Purpose of study Infants of diabetic mothers (IDM) have prolonged in utero exposure to high insulin levels and are at increased risk of being born large for gestational age (LGA) which can affect feeding outcomes and growth trajectories. We aim to determine if it takes IDMs longer to achieve full PO feeding volumes than their non-IDM counterparts. As a secondary outcome, we believe that infants of diabetic mothers have different initial growth trajectories when compared to the general population.

Methods used We are conducting a retrospective cohort study of late preterm infants (34 0/7–36 6/7 weeks gestation) admitted to the neonatal intensive care unit (level II or III) at a single academic center. Data was obtained from institutional, neonatal databases with comparisons between IDM and non-IDM cohorts. Outcomes to be measured include time until full PO feeds and initial weight gain trajectories. Weight gain trajectories will be described and compared to traditional growth charts.

Summary of results 502 subjects (129 IDM and 373 non-IDM) were included. Based on the preliminary information collected, the two groups were similar when comparing gestational age, gender, race, NICU level of admission, length of hospitalization and respiratory support. The IDM group had a significant higher mean birth weight [2748 g vs. 2278 g; $p < 0.0001$], a higher incidence of being LGA [27.9% vs. 2.7%; $p < 0.0001$] and higher incidence of hypoglycemia [51.2% vs. 27.4%; $p < 0.0001$]. Currently the databases are being queried for time to full PO feeds (days) and daily growth parameters [weight (g), length (cm) and head circumference (cm)]. A survival curve will be created to analyze time to full PO feeds in the groups. Daily weights of the IDM group will be plotted along traditional growth curves to determine if there is a difference in initial growth when compared to traditional growth curves. We will also analyze the subgroups of IDMs that are LGA against non-IDMs who are not LGA.

Conclusions At baseline, IDM infants in our cohort have higher birthweights, higher incidence of being LGA and higher incidence of hypoglycemia. We hypothesize that IDM infants have longer time to full PO feeds and different growth trajectories than their non-IDM counterparts, including the potential for catch-down growth. Further analysis is ongoing.

437 BURDEN OF RESPIRATORY READMISSIONS IN BRONCHOPULMONARY DYSPLASIA INFANTS IN A MIDSOUTH CHILDREN'S HOSPITAL

L Key*, AJ Talati, B Bagga. University of Tennessee Health Science Center, Memphis, TN

10.1136/jim-2018-000974.435

Purpose of study Bronchopulmonary Dysplasia (BPD) is a chronic illness and many preterm infants are discharged with active but stable illness. Readmissions are frequent and management during readmissions varies. The purpose of this study was to examine the characteristics and management strategies

Abstract 437 Table 1

	Mild	Moderate	Severe	Total
Total Patients	22	28	22	72
Total Re-admissions	18	30	29	78
Median length of stay in days (range)	6 (1–23)	4 (1–29)	3 (1–111)	4 (1–111)
Median days requiring increased FiO2	2 (0–23)	1 (0–28)	1 (0–22)	1 (0–28)

of re-admissions for respiratory illnesses of patients discharged from the NICU with a diagnosis of BPD.

Methods used We reviewed medical records of infants born January 2012 to December 2016 and discharged from a level IV NICU with a diagnosis, coded in the discharge summary, of any degree of BPD. Readmissions to our institution were also reviewed. We collected demographics, medical history, clinical course and outcomes during these admissions. Medical records were reviewed for viral or bacterial triggers, antibiotic and steroids use.

Summary of results 72 BPD infant charts were reviewed. 8 patients were excluded because they died of severe BPD prior to discharge from the hospital. 36 of the 64 patients included were re-admitted with respiratory complaints during the first two years after discharge. 39 (50%) of admissions were given a discharge diagnosis of a viral illness and of these, 16 admissions had a positively identified viral trigger (9 RSV, 3 Human Metapneumovirus, 4 Influenza). RSV positive patients spent a median of 6 days hospitalized, ranging from 2–32 days. 64% of admitted patients received steroids during admission. 70% of patients required increased FiO2 during admission and those who required increased FiO2 were on it for an average of 5.3 days.

Conclusions The burden of readmissions with respiratory illness in our BPD patients was high with more than half the patients readmitted within the first two years of life, with a hospital stay averaging over a week. Patients diagnosed with RSV had a longer length of stay. A majority received steroid therapy. Our study demonstrated that readmission with a respiratory illness was common in children with BPD and the clinical practices during these readmissions are not standardized and requires further clinical trials.

438 THE ASSOCIATION BETWEEN MATERNAL ADVERSE CHILDHOOD EXPERIENCE AND MATERNAL DEPRESSION: A LONGITUDINAL COHORT STUDY

J Kim*, T Clarke, C Jones, S Drury. Tulane School of Medicine, New Orleans, CA

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Purpose of study Though adverse childhood experiences (ACE) has been linked to negative mental and physical health outcomes in adulthood, no studies to date have examined maternal depression prenatally and postnatally across the first years of life in relation to maternal ACE score. This study investigates the association of maternal ACE with both the prevalence and persistence of depression across the prenatal and postnatal time period.

Methods used Data from a longitudinal parent study ($n=355$) was used to investigate the impact of ACE on rates and

persistence of depression at six different time points: prenatal, 4, 12, 18, 36, 48 months. ACE was captured through ACE Questionnaire, and depression was measured by Edinburgh Postnatal Depression Scale (depression if ≥ 10) and Beck Depression Inventory (depression if ≥ 14). The difference in rates of depression between high (≥ 4 , 28.5%) vs. low (< 4 , 71.5%) ACE groups at each time point was analyzed using the Chi-square statistics, and the association between ACE and depression scores was evaluated with linear regression. Secondary analysis tested the effect of ACE on the persistence of depression through each time point, defined as the proportion of times a mother screened positive for depression.

Summary of results Mothers characterized by high ACE, compared to low ACE group, showed higher rates of depression at prenatal (51.5% vs. 31.5%; $\chi^2(1)=12.362$, $p<0.001$, $n=355$), 18 mo (34.7% vs. 17.4%; $\chi^2(1)=5.726$, $p=0.017$, $n=158$), and 36 mo (54.2% vs. 17.9%; $\chi^2(1)=10.813$, $p=0.001$, $n=80$); the higher rates at 4 mo ($p=0.284$, $n=206$), 12 mo ($p=0.146$, $n=163$), and 48 mo ($p=0.373$, $n=27$) were statistically not significant. Linear regression revealed that ACE score was positively associated with depression scores at prenatal ($\beta=0.239$, $p<0.001$), 18 mo ($\beta=0.195$, $p=0.02$), and 36 mo ($\beta=0.299$, $p=0.009$) after accounting for SES, race, and maternal age. Higher ACE score predicted increased depression persistence, most notably at 48 months ($\beta=0.216$, $p=0.001$).

Conclusions Maternal ACE is associated with elevated risk of perinatal maternal depression. Despite the blunt and retrospective nature of the ACE score it robustly predicts depression risk. The lack of significance only 4 and 12 months, suggests the first year of a child's life may be a unique window relative to the chronic cross-domain health effects of maternal ACEs.

439

PREDICTORS OF LENGTH OF STAY IN GASTROSCHISIS INFANTS

¹SR Komic*, ¹F Aderibigbe, ²A Rao, ¹B Stansfield. ¹Medical College of Georgia, Augusta University Health, Augusta, GA; ²Augusta University, Augusta, GA

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Purpose of study Simple gastroschisis is a developmental defect of the periumbilical wall resulting in a persistence of herniated abdominal organs outside of the abdomen. While surgical correction of the abdominal defect is theoretically straightforward, neonates with gastroschisis have varied lengths of hospitalization (LOH) and healthcare costs. The purpose of this study was to 1) identify factors associated with and 2) build a prediction model for LOH for neonates with simple gastroschisis.

Methods used We identified and reviewed 106 cases of gastroschisis at the Children's Hospital of Georgia from 2000 to 2017. Subjects with complex gastroschisis, bowel atresia and/or necrosis, and multiple congenital anomalies were excluded. 73 subjects with simple gastroschisis were identified and included in the analysis. Multivariable linear regression was used to identify relationships between demographic data and LOH. These relationships were used to develop a predictive model of LOH for simple gastroschisis.

Summary of results Female sex ($-2.4d$, $p=0.04$), shorter time to silo placement ($-0.9d$, $p=0.004$), and supplementation of

breastmilk with formula ($-3.4d$, $p=0.04$) associated with reduced LOH. Dependence on nasogastric feeding ($+11.4d$, $p<0.001$) significantly increased LOH, but allowed some neonates to be discharged sooner than expected. Birth weight, gestational age, maternal age, mode of delivery, and time to repair did not significantly modify LOH. Based on these variables, we built a mathematical model to predict LOH for neonates with simple gastroschisis and compute estimated LOH for sub-groups that were highly associated with LOH.

Conclusions We identified several variables that directly associate with LOH for simple gastroschisis and propose a mathematical model to predict LOH for neonates with simple gastroschisis.

440

ANOTHER YELLOW BABY

M Martinez*. LSU Shreveport, Bossier City, LA

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Cholelithiasis is very uncommon in the neonatal period, previous cases association with prolong exposure to total parenteral nutrition and lack of enteral feeds. We present a case of premature infant with hyperbilirubinemia found to have gallstones.

Case Infant is a 96 day old born at 28 w 5 d corrected gestation 42 w 1 d seen in clinic for follow up after neonatal intensive care discharge. Physical exam was consistent with jaundice which prompted laboratory evaluation. Labs significant for total bilirubin of 9.5 mg/dl, direct bilirubin 8.1 mg/dl, AST 140 and ALT 194 u/dl. Infant was admitted for further evaluation. Medical history significant for 94 day neonatal intensive care unit stay. Birth APGARs of 3,6 and 8 at 1, 5, 10 min, he was intubation and received surfactant at birth, antibiotics initiated as part of sepsis protocol and discontinued after negative blood cultures. NICU course complicated by 2 episodes of necrotizing enterocolitis modified bells 1B treated with NPO and antibiotics, 4 blood transfusions, severe feeding intolerance with 76 days of total parenteral nutrition, jaundice treated with phototherapy for 9 days, and 1 episode of hematochezia prior to discharge that resolved with hydrolyzed formula. He was discharge home on room air and tolerating full oral feeds. During hospital floor admission an abdominal ultrasound was obtained along with HIDA scan showed cholelithiasis and abnormal biliary imaging with no tracer activity. An intra-operative cholangiography was performed and noted a stone in the distal common bile duct. A cholecystectomy with wedge biopsy of the liver was performed and showed portal hepatitis grade 2, portal fibrosis grade 1 and hepatic parenchyma showing bile stasis. Gallbladder pathology showed mild chronic cholecystitis with cholelithiasis. Infant was treated with Actigall for 7 days and discharge on poly-vi-sol and pregestimil 24 kcal formula. At time of discharge his total bilirubin was 5.7 mg/dl and direct bilirubin 5.3 mg/dl both trending down.

Discussion TPN use in premature infants has been associated with hepatobiliary complications including steatosis, cholestasis and steatohepatitis. Although a rare finding, neonatal cholelithiasis has also been implicated. Our infant had multiple risk factors including prematurity, prolonged TPN with lack of enteral nutrition leading to cholestasis and possibly contributing to cholelithiasis.

441 GLYCOGEN STORAGE DISEASE TYPE IV: CLINICAL PRESENTATION AND DIAGNOSIS IN A NEONATE

CN Massey*, D Dannaway. *University of Oklahoma, Oklahoma City, OK*

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Our patient is an infant female born at 32 weeks gestation to a 25 year old G1P0 female. Prenatal history included polyhydramnios and intrauterine marijuana exposure. She required intubation shortly after birth due to minimal spontaneous respirations. Apgar scores were 3/6/7 at 1/5/10 min of life. She was transferred to our NICU for further care on day of life 6. Admission exam showed a 2637 gram, large for gestational age female who was jaundiced and mildly tachycardic. Her pupils tracked, were mildly dilated and non-reactive. There was no spontaneous respiratory effort, and she was very hypotonic with slight recoil to stimulation. Multiple imaging studies and labs were unremarkable except for mildly elevated creatine kinase and ammonia levels. Over the subsequent weeks, the patient had no improvement of her respiratory status or hypotonia. She developed left ventricular hypertrophy on EKG plus hepatomegaly. An extensive metabolic workup and multiple genetic tests including single nucleotide polymorphism array failed to provide a diagnosis. At 11 weeks of life, a muscle biopsy was obtained. Concurrently, blood samples were sent for whole exome sequencing (WES), which indicated that the patient had two pathogenic variants in the *GBE1* gene known to cause glycogen storage disease type IV (GSD IV). Muscle biopsy results showed the accumulation of diastase-resistant round PAS-positive globules, which confirmed the diagnosis. The patient died the following month. Cause of death was cardiopulmonary arrest secondary to GSD IV.

GSD IV is an autosomal recessive disease that causes a deficiency of glycogen branching enzyme, which leads to accumulation of abnormal glycogen molecules in affected tissues. The age of onset, tissue involvement, and severity are variable. Our patient's presentation correlates with the congenital neuromuscular subtype of GSD IV. This subtype presents in the newborn period with profound hypotonia, respiratory distress, and dilated cardiomyopathy. Prenatal findings include polyhydramnios and decreased fetal movement.

Diagnosis can be difficult and delayed due to the broad differential of neonates with hypotonia. GSDs should be considered early during the workup and WES and muscle biopsy should be utilized.

442 A CASE OF PULMONARY SEQUESTRATION IN A NEWBORN

R McPeters*, EN Chiccarelli, LS Engel, JS Krulisky. *LSU Health Sciences Center, New Orleans, LA*

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Introduction Bronchopulmonary sequestration (BPS) is a rare congenital lung abnormality of the lower airway typically composed of a dysplastic and nonfunctioning lung tissue. The lung tissue lacks normal communication with the tracheobronchial tree which receives its arterial blood supply from the systemic circulation and has various forms of venous drainage.

Case A 5 day old infant, with in utero exposure to opiates and THC, born to a G1P1 mother via spontaneous vaginal

delivery was admitted for hyperbilirubinemia secondary to dehydration and neonatal abstinence syndrome. Mother noted decreased oral intake since nursery discharge along with increased fussiness and decreased urine output. Upon presentation, the patient had stable vital signs with no fever but was noted to have a total bilirubin of 14.6 mg/dL. A grade 2/6 systolic ejection murmur was noted on physical examination prompting an echocardiogram. 2D-echocardiography showed normal pulmonary venous return with a patent foramen ovale with left to right shunting, mild left ventricular enlargement and a moderate AP collateral/artery arising from the descending aorta suggestive of a possible pulmonary sequestration. EKG showed normal sinus rhythm with no signs of left ventricular enlargement. CT angiography of the chest showed an extralobar pulmonary sequestration within the right lower lobe with systemic arterial supply arising from the anterior aspect of the aorta just proximal to the takeoff of the superior mesenteric artery. The feeding artery and the draining veins returning to the left atrium were prominent and dilated, suggesting an associated arteriovenous malformation vs fistula.

Discussion BPS varies depending on the size, type, and location of the lung lesion. The affected newborn/individual is usually asymptomatic but in certain cases may present with recurrent pneumonias. Computed tomography (CT) angiography is the gold standard for diagnosis of BPS and allows for visualization of the anatomy, identifies the anomalous systemic arterial supply, and shows the venous drainage. Management of asymptomatic BPS is controversial but infants with BPS causing respiratory symptoms are treated with excision as surgery is curative and associated with minimal mortality.

443 AMPLE SAMPLE? SAMPLING STRATEGIES FOR NEONATAL ANTIBIOTIC STEWARDSHIP

MO Montgomery*, B Her, J Canteley. *University of Texas Health Science Center San Antonio, San Antonio, TX*

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Purpose of study Prospective audit and feedback is a critical aspect of antibiotic stewardship programs. Studies on antimicrobial stewardship in the neonatal intensive care unit (NICU) have collected information from the entire infant cohort, but this may not be economical for long-term surveillance if stewardship program resources are limited. Our objective was to determine the accuracy of limited sampling strategies.

Methods used Prospective cohort study of all infants admitted to the neonatal intensive care unit from 9/15 to present. Demographic, clinical, laboratory, and outcome data were collected for all infants. Samples of approximately 50%, 25%, and 10% of NICU admissions were taken by including infants born on calendar days 1–14 (50% group), 15–21 (25% group), and 22–24 (10% group) of each month. Demographic information and antibiotic use, as measured by days of therapy (DOT) per 1000 patient-days, were compared between sampling strategies.

Summary of results 1531 infants were admitted to the NICU during the study period, accounting for 41 190 hospital days and 10,642 days of antibiotic therapy (258.4 DOT/1000 patient-days). Accuracy of different sampling strategies is shown in table 1. Sampling 50% or 25% of the cohort was associated with only 0.4%–1% sampling error in DOT/1000

Abstract 443 Table 1

	Entire cohort	50% sampling	25% sampling	10% sampling
N	1531	730	339	154
Inborn, %	82	80	84	82
Female, %	45	56	42	49
Median gestational age, weeks (IQR)	35 (33–38)	35 (33–38)	35 (33–38)	35 (33–38)
Median birth weight, g (IQR)	2360 (1860–3090)	2379 (1863–3102)	2450 (1860–3140)	2316 (1896–2987)
Vaginal delivery, %	43	43	42	45
Indication for admission, %				
≤34 weeks	45	45	44	45
≥35 weeks, respiratory distress	19	19	21	24
Congenital anomaly				
Other	9	10	8	6
DOT/1000 patient-days	258.4	259.4	264.4	227.5
Variance	n/a	+0.4%	+1%	–12%

patient days compared to the gold standard. Sampling error increased (12%) in the 10% sampling group.

Conclusions Smaller sampling strategies may be an option for resource-limited antibiotic stewardship programs in the NICU. Reduced time and effort must be weighed against increased sampling error with smaller samples.

444 NEPHROTOXIC ANTIBIOTIC EXPOSURE AND ACUTE KIDNEY INJURY IN A GENERAL NEONATAL INTENSIVE CARE UNIT POPULATION

K Allen, T Hardy, TC Dixon, K Twombly, A Wahlquist, HJ Murphy*. *Medical University of South Carolina, Charleston, SC*

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Purpose of study We aim to quantify nephrotoxic antimicrobial exposure in a general NICU population and determine what characteristics differ between those who do and do not develop AKI.

Methods used We are conducting a retrospective study, collecting data from the records of infants admitted within 48 hour of life to a single center NICU from 7/1–12/31/2017 and underwent ≥1 sepsis evaluation with antimicrobial receipt. Comparisons were made between those who did (n=5) and did not develop AKI (n=43; 1 infant excluded) to date with AKI defined by modified KDIGO criteria ≥stage 1.

Summary of results During the study period 364 infants were admitted to the NICU. To date, 49/100 infants screened met inclusion criteria, 43% females and 57% males (data collection ongoing). Median GA was 34 weeks (IQR 31–36) and median BW was 2220 grams (1620–2670). Median Apgar scores were 7 (4–8) and 8 (7–9), at 1 and 5 mins. 9 infants had >1 sepsis evaluation (range 2–6). 48/49 (98%) survived and 1/49 (2%) died. Those with AKI have lower GA, BW, APGAR scores, more sepsis evaluations, and a greater number of antibiotic doses (table 1).

Conclusions Preliminary data suggests that infants who require sepsis evaluations and antibiotics in our NICU are premature and low birth weight. Those who develop AKI are more

Abstract 444 Table 1 Characteristics of those with and with acute kidney injury

Characteristics	No Acute Kidney Injury (n=43)	Acute Kidney Injury (n=5)	p-value
Gestational age (weeks)	34 (32–36)	25 (24–25)	0.005
Birthweight (grams)	2370 (1695–2910)	800 (680–808)	0.006
APGAR (1 min)	8 (5–8)	2 (1–3)	0.02
APGAR (5 min)	8 (7–9)	4 (4–5)	0.003
Number of sepsis evaluations	1 (1–1)	3 (2–4)	0.0002
Total antibiotic doses	8 (6–23)	55 (33–93)	0.005
Ampicillin doses	6 (4–8)	8 (7–21)	0.1
Gentamicin doses	2 (1–3)	4 (4–4)	0.01
Vancomycin doses	6 (5–10)	4 (3–25)	0.7
Piperacillin+Tazobactam doses	14 (6–23)	23 (7.5–38.5)	0.5
Ceftazidime doses	4 (3–5)	2 (2–2)	0.3
Nafcillin doses	2 (1–7)	12.5 (6–19)	0.4
Other antibiotic doses	8.5 (1.5–16)	2 (2–2)	>0.9

Data expressed: median (IQR).

premature, have lower birthweights and APGAR scores, experience more sepsis evaluations, and are exposed to a larger number of antibiotic doses.

445 NEONATE WITH INTERSTITIAL DELETION WITHIN THE LONG ARM OF CHROMOSOME 10

¹S Hagan, ²T Sandhu, ²RR Nandyal*. ¹O. U. Medicine, Oklahoma City, OK; ²Oklahoma University Health Sciences Center, Oklahoma City, OK

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Partial deletion of distal chromosome 10q was first reported by Lewandowski (1978). Interstitial deletions within bands 10q25e10q26.3 are rare. Only seven such cases were reported, so far.

We report a rare case of a male newborn with an interstitial deletion within the long arm of chromosome 10 between bands 10q25.1 and 10q26.1, with dysmorphic features, Pierre-Robin sequence, right anorchia and bilateral dorsal horn ventriculomegaly.

A term male newborn delivered at a perinatal center because of antenatal diagnosis of unbalanced translocation of chromosomes 10 and 12 and fetal cleft lip and cleft palate. Blood sent for chromosome analysis, done using a GTG banding method.

Baby was AGA. Dysmorphic features include frontal sloping with normal anterior fontanel, hypertelorism, flattened nasal bridge, cleft lip on the left with cleft of soft and hard palate, cleft of the soft and hard palate on the right without cleft lip, intact nasal septum, normal ears and micrognathus. Other findings include normal shaped chest, appropriately spaced nipples, no heart murmur, 3 vessel cord, no organomegaly, suspected right cryptorchidism, patent anus and base visible sacral dimple.

Echo showed trivial tricuspid regurgitation; brain MRI revealed dilated dorsal horns of lateral ventricles with decompressed 3rd and 4th ventricles and prominent perivascular spaces; passed hearing screening; abdominal ultrasound

showed absence of right testis in inguinal canal and abdomen (anorchia); TSH and T4 were normal; blood glucose screens were all normal. Chromosomal analysis was abnormal.

Hospital course was remarkable for feeding problems requiring feeding team, plastic surgery consult (surgery at 2–3 months of age), and taping of the cleft lip. Discharged home on day 7.

This was a case of a dysmorphic newborn with an interstitial deletion within the long arm of chromosome 10 between bands 10q25.1 and 10q26.1, with a few new associations. Despite their rarity, routine referral to a maternal fetal medicine specialist for common anatomical abnormalities of the fetuses such as cleft lip, cleft palate or Pierre- Robin sequence may help earlier detection of both common and rare chromosomal- genetic abnormalities to prepare the family and health care team to plan optimal care.

446

EXTREMELY LOW BIRTH WEIGHT INFANTS WITH ANTENATAL EXPOSURE TO MAGNESIUM SULFATE ARE LESS LIKELY TO HAVE CEREBRAL PALSY

R Pandey*, C Arnold, A Duncan. *The University of Texas Health Science Center at Houston, Houston, TX*

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Purpose of study To compare neurodevelopmental outcomes of extremely low birth weight (ELBW) infants with antenatal exposure to magnesium sulfate for neuroprotection (MgSO₄) and antenatal steroids (ANS) to those exposed to ANS only. We looked at Bayley Scales of Infant Development, 3rd ed. (Bayley-III) Cognitive and Language Composite Scale scores and Cerebral Palsy, at 18–24 months corrected age.

Methods used This was a cohort study of ELBW (<1000 grams at birth and <28 weeks) infants born between April 2011 and May 2015 with antenatal exposure of ANS. Infants were grouped into two groups based on exposed to MgSO₄ for neuroprotection. Infants were excluded from analysis if administration of MgSO₄ was for maternal pre-eclampsia, Bayley-III scores or CP data were missing. STATA was used for analysis and *t* test and logistic regression were done to compare groups.

Summary of results One hundred fifty-four infants were included. One hundred thirty-three were exposed both to MgSO₄ and ANS and 21 were exposed to ANS only. When we analyzed all infants born in the study period, 13.9% of infant exposed to ANS only died prior to 36 weeks compared to 9.5% infants exposed to ANS and MgSO₄ (*p*=0.37). Using logistic regression to assess for confounding, adjusting for

Abstract 446 Table 1 Characteristics and outcomes of infants

	ANS (21)	ANS and npMgSO ₄ (133)	p value
Birth weight	785 (153)	785 (153)	NS
Gestational Age (wks)	25.5 (1.2)	25.3 (0.93)	NS
Male	62%	52%	
Cerebral Palsy	19%	4.1%	0.031
Bayley cognitive score	81 (14)	84 (13)	NS
Bayley language score	76 (15)	78 (15)	NS

Expressed in mean (SD)

Abstract 446 Table 2 Cerebral palsy in two groups:

Cerebral Palsy	Number	CP incidence	Risk Difference (95% CI)	Risk Ratio (95% CI)	NNT	p value
ANS only	21	4 (19.1%)				0.031
ANS and MgSO ₄	133	6 (4.5%)	−0.14 (−0.31 to 0.2)	0.23 (0.07 to 0.76)	6.8 (3.1 to 38.4)	

CI=Confidence Interval, NNT=Number needed to treat

gestation age, birth weight, gender, or year of birth did diminish the magnitude of the protective effects of magnesium. Number needed to treat with MgSO₄ to prevent one CP was 6.8 (95% CI 3.1 to 38).

Conclusions Magnesium sulfate for neuroprotection for ELBW infants is associated with lowered risk of CP.

447

EVIDENCE FROM NATIONAL DATABASE FOR PRENATAL COUNSELING STATEMENT SUCH AS 'MOST PREMATURE BABIES ARE DISCHARGED HOME AROUND EXPECTED DATE OF DELIVERY'

¹JM Patel*, ²M Patel, ¹J Desai. ¹UMMC, Madison, MS; ²UMMC, Jackson, MS

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Purpose of study To compare actual length of stay against the calculated expected day of delivery (EDD) for the corresponding Gestational age group.

Methods used Data from national multicenter Kids' Inpatient Database of the Healthcare Cost and Utilization Project from the years 2000, 2003, 2006, 2009 and 2012 were analyzed using complex survey design using SAS. We included infants with ICD9 codes (76521–76528) indicating Gestation age groups. Expected day of delivery (EDD) was calculated as a difference between corrected 40 week gestation and birth GA. Actual Length of stay was compared against calculated EDD for each group for discharged and deaths after excluding transfers.

Summary of results A total of 2 10 274 cases with weighted frequency of 2 72 480 were included in analysis. For GA ≤28 weeks, 70.9% neonates were discharged by Calculated EDD while >28 weeks, 98.11% infants were discharged by

Abstract 447 Figure 1

calculated expected day of delivery (EDD) derived from birth GA till corrected 40 week gestation. (χ^2 $p < 0.001$). Percent discharged by median EDD were significantly different between different GA category groups (P value < 0.001). Higher variability was seen in discharges for lower GA categories. Lower the GA, percent discharged by EDD were lower. For neonates who died, 75th percentile LOS were less than 16 days across all GA categories.

Conclusions Majority of infants > 28 weeks were discharged by EDD while, for neonate ≤ 28 weeks, about a quarter of infants stayed beyond their calculated EDD. Lower the gestational age group, variability in discharge prediction by EDD was higher.

448 EXTRACORPOREAL LIFE SUPPORT IN PEDIATRIC PATIENTS WITH BRONCHOPULMONARY DYSPLASIA AND ASSOCIATED PULMONARY HYPERTENSION

¹A Pena Hernandez*, ²N Carr, ¹V Armijo-Garcia, ¹DC McCurnin. ¹UT Health San Antonio, San Antonio, TX; ²Brooke Army Medical Center, San Antonio, TX

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Purpose of study To examine if a prior diagnosis of bronchopulmonary dysplasia with secondary pulmonary hypertension (BPD +PH) is associated with decreased survival of patients receiving ECLS when compared to patients with BPD alone.

Methods used Retrospective review of the ELSO registry (1982–2016) for pediatric patients with a prior diagnosis of BPD that received ECLS for noncardiac indications. Patients with congenital heart disease, congenital diaphragmatic hernia, congenital anomalies of airway or lungs (excluding tracheoesophageal fistula) or required additional ECMO run were excluded. Data was analyzed with Fisher's exact if nominal, student-t test if continuous. Multiple logistic regression analysis was used to identify a correlation of survival with PH and other categorical variables.

Summary of results 260 patients with BPD were identified with a 5% subset with associated PH. The overall survival rate was 70% in all BPD patients and 77% in BPD +PH patients ($p = 0.759$). A review of a contemporary subgroup (2012–2016) demonstrated 17% of 63 total patients with BPD had BPD +PH. Mean oxygenation index (OI) prior to ECLS was similar (BPD 48 ± 32 mean $\pm$ SD vs. BPD +PH 55 ± 14 mean $\pm$ SD). There were no differences in survival or duration of ECLS support. Multiple logistic regression failed to show an association between survival and PH, weight, gender, duration of ECLS, or mode of ECLS support. The most common ECLS reported complications were hypertension requiring vasodilators (BPD 38%, BPD +PH 27%), inotrope use on ECLS (BPD 35%, BPD +PH 45%), and hemofiltration requirement (BPD 21%, BPD +PH 36%). Neurologic and pulmonary complications were uncommon in both groups presenting in $< 10\%$ of the cases.

Conclusions Limited retrospective data suggests a reasonable survival of patients with BPD +PH, similar to patients with BPD alone. Severe complications and prolonged duration of ECLS were not noticed in these patients.

Disclosure The view(s) expressed herein are those of the author(s) and do not reflect the official policy or position of Brooke Army Medical Center, the U.S. Army Medical Department, the U.S. Army Office of the Surgeon General, the Department of the Air Force, Army, Navy, Department of Defense or the U.S. Government.

449 FEEDING PROBLEMS IN IN LATE PRETERM AND TERM INFANTS BORN TO MOTHERS WITH DIABETES MELLITUS

S Peravali*, SK Chilakala, AJ Talati. University of Tennessee, Collierville, TN

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Purpose of study Maternal diabetes mellitus (DM) is a growing problem, leading to increased infant morbidity. Infants of diabetic mothers (IDM) seem to have delayed maturation and more difficulty with feeding. We reviewed hospital stay of late preterm and term infants with maternal DM to identify the risk of feeding difficulties and associated clinical risk factors.

Methods used All infants ≥ 34 wk gestation admitted to our level 3 B NICU between Jan 2014 and Dec 2015 were included. A retrospective chart review was done; infants with major congenital anomalies HIE, NAS and NEC were excluded. Groups were divided based on maternal DM history. Age, race, parity of the mother, obstetric and neonatal characteristics during hospital stay were collected and compared between the two groups.

Summary of results 656 babies were included in the study, 159 (24%) of those were IDM (after exclusion of 34 infants). There were no significant differences in race, maternal age, mode of delivery or Apgar scores. Time to reach full oral feeds was higher in the IDM group compared to the control group (3 d v. 1 d, $p < 0.0001$). IDM babies also had a significantly longer median length of stay compared to the control group (7 d v. 6 d, $p < 0.0001$). On subgroup analysis of the study group, IDM infants born at 37 and 38 week completed gestation age had much higher need of tube feeding (17/53 [32%]) compared to non-IDM (7/131 [5.3%]), $p = 0.0001$. IDMs who had blood glucose levels ≤ 45 mg/dl in the first 24 hour were more likely to require tube feeding (40% vs 20%, $p = 0.0118$).

Conclusions IDM status is associated with increased time to reach full oral feeds, which may lead to prolongation of hospital stay. This is more pronounced if the IDM infants are hypoglycemic in the first 24 hours.

450 ORAL GENTAMICIN AND NEONATAL ONSET BACTEREMIA OF INFANTS HOSPITALIZED IN A NEONATAL INTENSIVE CARE UNIT

¹N Pettit*, ¹K Bryant, ^{1,2}J Sullivan, ³J Duong, ¹C Espinosa. ¹University of Louisville, Louisville, KY; ²Norton Childrens Hospital, Louisville, KY; ³Washington University, Saint Louis, MO

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Purpose of study The use of oral gentamicin to prevent necrotizing enterocolitis (NEC) in very low birth weight (VLBW) infants (< 1500 g), is controversial. Some studies suggest a protective effect against NEC, but impact on the neonatal microbiome is unknown. We assessed the antibiotic resistance profile of organisms isolated in the blood in infants exposed to oral gentamicin.

Methods used A retrospective study with descriptive analysis was performed. NICU patients that received oral gentamicin from 2/18/13 to 01/6/17 were identified using pharmacy records from a health care organization system with 150 NICU beds. Gestational age, birth weight, CRIB II score, duration of oral gentamicin and the presence of hospital-onset bacteremia, were extracted from health records. The resistance patterns of bacterial bloodstream isolates were described.

Abstract 450 Table 1 Gram negative minimum inhibitory concentrations

Organism	Age (days) culture obtained	PO GEN Start Day	AMK	AMC	AMP	SAM	CFZ	FEP	CTX	CRO	CIP	GEN	LVX	MEM	TZP	TOB	SXT
E. coli	6	0	≤16	16/8	*>16	*>16/8	*>4			≤1	≤1	*>8				*>8	≤2/38
E. coli (ESBL)	20	1		≤8/4	*>16	*>16/8	*≤8	*≤8	*≤1	*≤8	*>2	≤4	*>4	≤4	≤16		*>2/38
K. pneumoniae	7	5	≤8	16/8	*>16	*>16/8	*>16	*>16	*16	*>32	*>2	*>8	≤1		≤8	*>8	*>2/38
S. marcescens	17	0		16/8	*>16	*>16/8	*>16	≤8	*>16	32	≤1	≤4			*>64		≤2/38
C. freundii	241	0		*>16/8	*>16	*>16/8	*>16	*≤2	*>16	*32	≤0.5	≤0.5			≤0.5		≤0.5/9.5
complex																	

*Resistant

Summary of results A total of 289 infants were included in the analysis. Most infants were white (n=196) and the mean gestational age was 27.6 weeks. Eight (3%) had NEC, 8% (n=23) had hospital onset bacteremia and 5% (n=14) died. Median duration of oral gentamicin therapy was 30.3 days [IQR 20–41]. There were 5 gram-negative and 18 gram-positive isolates. Two gram-negative isolates were gentamicin resistant (table 1). Gentamicin synergy was absent in two *Enterococcus faecalis* isolates.

Conclusions Prophylactic oral gentamicin may negatively influence the antibiotic resistance patterns of bacteria in subsequent bloodstream infections. Further studies are needed to confirm that oral gentamicin contributes to antibiotic resistance in VLBW infants.

451

COMPARISON OF OUTCOMES IN VERY LOW BIRTHWEIGHT NEONATES WITH HUMAN MILK FORTIFICATION BY ACIDIFIED LIQUID PROTEIN HUMAN MILK FORTIFIER OR HYDROLYZED PROTEIN HUMAN MILK FORTIFIER

R Pollack*, N Alshibi, AC Skeen, R Krishnan, AJ Talati. UTHSC, Memphis, TN

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Purpose of study To compare growth measures, metabolic outcomes, specifically acidosis, BUN, hematocrit, as well as adverse effects of abdominal distension, loose stools, and emesis in VLBW neonates on enteral feeds of human milk with either acidified liquid protein fortifier (aHMF) versus liquid hydrolyzed formulas (hHMF).

Methods used A retrospective review of VLBW neonates fed human milk with fortification was done. Babies were designated into two groups based on the liquid fortifier additive. Group 1 included acidified liquid human milk fortifier (aHMF) and group 2 included hydrolyzed human milk fortifier (hHMF). Data was recorded for 4 weeks after initiation of fortifier with weekly growth parameters, laboratory values for acidosis, BUN, hematocrit, as well as adverse outcomes including abdominal distension, loose stools, and emesis. Metabolic acidosis was defined as a base deficit of >5, bicarbonate of less than 18, and/or a pH <7.25.

Summary of results A total of 58 VLBW neonates were analyzed: 24 patients in the aHMF group and 34 patients in the hHMF group. Comparative clinical characteristics are shown in the table. No differences in weight gain over 4 week period was noticed (p=0.399). Adverse events were similar in two groups.

Abstract 451 Table 1 Comparative data

n=58	aHMF n=24	hHMF n=34	p-value; t-test/Chi-square
Gender (Male)	17	21	p=0.579
GA (weeks)	27±2.5	27±5.6	p=0.60
Mean BW (gm)	809±269	1025±314	p=0.008
Mean weight gain over 4 week period after initiation of fortifier (gm)	1231±313	1122±560	p=0.399
Mean change in OFC over 4 weeks (cm)	6.47±2.59	6.5±3.55	p=0.707
Mean change in length over 4 weeks (cm)	6.952±3.6	6.89±4.32	p=0.81
Acidosis (%)	14 (32%)	21 (27.4%)	p=0.78
Mean BUN (mg/dL) at wk 4	10.4±4.53	9.5±5.4	p=0.82
Mean Hematocrit at wk 4	33.3±4.87	31.6±3.9	p=0.96
Number of transfusions	5.5±3.4	1.8±2.5	p=0.002
Abdominal Distension	13 (54%)	16 (47%)	p=0.21
Loose stools	12 (50%)	13 (38%)	p=0.427
Emesis	7 (29%)	5 (15%)	p=0.21

Conclusions Though mean birthweight was lower in aHMF group, there was no significant difference between aHMF and hHMF groups in growth, metabolic acidosis, BUN, Hct, or adverse effects. Both bovine fortifiers seem to be equally tolerated in our cohort.

452

LONG-TERM OUTCOMES FOLLOWING INTESTINAL REHABILITATION OF INFANTILE ONSET ULTRA-SHORT BOWEL SYNDROME

¹MH Premkumar*, ¹L Gollins, ²S Shah, ¹AB Hair. ¹Baylor College of Medicine, Bellaire, TX; ²Texas Children's Hospital, Houston, TX

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Purpose of study Intestinal Failure (IF) is a condition of malabsorption resulting from small bowel resection, anatomical defect, or functional dysmotility requiring long-term parenteral nutrition (PN). Ultra-short bowel syndrome (USBS) is defined as residual small bowel length of <30 cm. Ensuring proper growth and nutrition while minimizing significant mortality and morbidities in infants with USBS is challenging. Management by specialized multidisciplinary intestinal rehabilitation teams has improved outcomes for these high-risk infants. Our

Abstract 452 Table 1 Diagnosis resulting in USBS

Diagnosis	Number
NEC	4/9 (45%)
Gastroschisis with volvulus	3/9 (33%)
Gastroschisis	1/9 (11%)
Midgut volvulus	1/9 (11%)

objective was to evaluate the treatment and outcomes of infantile-onset USBS in a single quaternary NICU.

Methods used A retrospective review of patients treated for infantile-onset USBS from 2015–2017 was performed. Demographic information, patient characteristics, treatment details, and short-term outcomes were collected. Descriptive Statistics were used to quantify the data.

Summary of results During a three-year follow-up 9 infants were diagnosed with USBS. The diagnoses resulting in USBS is shown in table 1. Mean gestational age was 32 5/7±11 1/7 weeks and birthweight was 1849±926 grams. Residual small bowel length was 14 cm (IQR, 10 to 25) and 11% (1/9) had preserved ileocecal valve and colon. One infant (11%) was lost to follow-up and one infant (11%) died during NICU stay. Enteral autonomy was achieved in 2 (22%) patients by 1.4 years. 7/9 (78%) infants had IF associated liver disease which resolved with the use of fish oil-based lipid emulsion. No patients received transplants or autologous intestinal reconstruction (AIR) procedures.

Conclusions During a three-year follow-up, 2/9 (22%) infants with USBS achieved enteral autonomy while only 1/9 (11%) died. None required AIR or transplant, and despite long-term PN, none developed liver failure. These results describe the highly encouraging outcomes in this high-risk population of infants with USBS.

453 LONGITUDINAL COST UTILIZATION FOR COMMON NEONATAL SURGICAL CONDITIONS

¹J Miller, ¹I Adams, ²M Raval*, ¹A Piazza. ¹Emory, Atlanta, GA; ²Northwestern University, Chicago, IL

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Purpose of study Decreasing cost and improving healthcare utilization remains a focus for patients and insurance companies. Infants born with major congenital malformations incur a heavy economic burden. Creating a cost of care timeline and identifying the time point when cost of care approximates the cost of care for an unaffected child would provide important information for families and health economists.

Our objective was to evaluate longitudinal cost of care during the first five years of life for the following defects: abdominal wall defect (AWD), congenital diaphragmatic hernia (CDH), tracheoesophageal fistula and esophageal atresia (TEF/EA), and intestinal atresia (IA).

Methods used Data were analyzed from the 2008–2014 Truven Health MarketScan Commercial Research Database. Patients were identified by ICD-9 codes for the surgical defect and associated repair procedure. Inpatient and outpatient claims were extracted over a five-year period for infants who were continuously enrolled for a minimum of 6 months. A

Abstract 453 Figure 1

control population of term infants will be matched based on the birth year during the study period.

Summary of results TEF/EA, followed by CDH, had the highest median cost for the initial year after birth. Cost for all defects decreased exponentially during the first year of life (figure 1). Ongoing cost varied over time with a notable increase in inpatient costs in years 3 to 4 of life. There is ongoing analysis to determine the cost of care for an unaffected child born during the same time period to use a control for our cohort population.

Conclusions The cost of care was highest during the first year of life for all four defects. The cost of care decreases over time, however, these patients may incur additional costs early in life. Further analysis will determine if and when these costs are similar to those in a healthy, term control population.

454 IMPROVING PRETERM INFANT CLINICAL OUTCOMES THROUGH IMPLEMENTATION OF A FEEDING PROTOCOL

J Robinson*, R Gupta, J Surcouf, M Knecht. LSUHSC School of Medicine, New Orleans, LA

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Purpose of study The purpose of the study is to standardize enteral feeding practices in preterm infants via a feeding protocol leading to improved time to initiation of enteral feeds, improved time to full enteral feeds/overall feeding tolerance, reduced total parenteral nutrition (TPN) days ultimately leading to less central venous line (CVL) days, and reduced incidence of necrotizing enterocolitis (NEC) and late-onset sepsis (LOS).

Methods used The study was conducted via a retrospective chart review evaluating preterm infants ≤2000 g or ≤34 weeks gestation at Touro Infirmary one year before and one year after implementation of a standardized feeding protocol in January 2017. Exclusion criteria included infants with chromosomal or congenital malformations. Eligible infants were identified through neonatal intensive care unit (NICU) admission records. Electronic medical records were analyzed for nutritional parameters including day of life (DOL) of initiation of enteral feeds, DOL full enteral feeds (150 ml/kg/d) achieved, and DOL of enteral feed fortification (24 kcal/oz) as well as clinical outcomes including days of TPN/CVL and incidence of Feeding Intolerance (FI), NEC, and LOS.

Summary of results After implementation of the standardized feeding protocol, there were significant improvements in earlier initiation of enteral feeds (DOL 2.3 to 1.9, p 0.01) and earlier fortification of enteral feeds (DOL 15.4 to 11, p

0.025). There was some improvement in earlier achievement of full enteral feeds (DOL 11.6 to 10.4, p 0.09). Number of days of TPN did not improve (6.8 to 6.4, p 0.74), but number of days of maintained CVL did dramatically decrease by 47% (8.5 to 4.5, p 0.013). There were also some mild improvements in incidence of FI (20.5% to 15.9%, p 0.25) and NEC (4.5% to 3.7%, p 0.68). The most clinically significant improvement was in reduced incidence of LOS by 55% (8.3% to 3.7%, p 0.085).

Conclusions Through the initiation of a preterm infant standardized feeding protocol, we achieved earlier initiation of enteral feeds, progression to full enteral feeds, and fortification of enteral feeds, as well as improved our incidence of feeding intolerance. We reduced our CVL days as well as our incidence of NEC and LOS, which now are below national standards per Vermont Oxford Network (NEC 4.9%, LOS 8.1%).

455 GENERALIZED LYMPHATIC ANOMALY IN TWO PRETERM INFANTS, DIAGNOSIS AND MANAGEMENT

A Rose*, ME Barbian, A Gill, M Hawkins, M Briones, MA Pakvasa. *Emory University, Decatur, GA*

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Case report Generalized lymphatic anomaly (GLA), a rare disease affecting children and young adults, is a multifocal lymphatic malformation that involves skin, soft tissue, bone, pleural and abdominal viscera. Pathologic effusions may occur. A phase II trial showed sirolimus, a mammalian target of rapamycin (mTOR) inhibitor, to be effective for GLA; however, little is known about sirolimus use in preterms. We report 2 cases of preterm infants with GLA diagnosed by MR lymphangiography and treated with sirolimus.

Case 1: A 1.6 kg female infant born at 34 weeks required intubation for respiratory failure. Chest x-ray revealed bilateral pleural effusions (PE), and fluid studies were consistent with chyle. She was managed with chest tube (CT) drainage, nil per os and an octreotide drip. At 6 weeks of age MR lymphangiography revealed pulmonary lymphangiectasia, consistent with GLA. Sirolimus was initiated and octreotide weaned off. In 4 weeks, her CTs were removed and she was extubated. She went home at 5 months on sirolimus.

Case 2: A 580 g male infant born at 27 weeks required intubation, surfactant and ventilation for respiratory distress syndrome. By 1 month, he developed a right PE which evolved to bilateral PEs requiring CT drainage. Fluid studies suggested chyle. Octreotide was ineffective. MR lymphangiogram revealed diffusely abnormal lymphatic channels, consistent with GLA. Treatment with sirolimus was initiated. In 1 month, both CTs were removed and he was extubated.

Discussion GLA typically presents in childhood, although perinatal manifestations occur. Histology demonstrates lymphatic channels of variable sizes, lined with flattened endothelium. Lymphangiography can identify lymphatic malformations and is an alternative diagnostic tool when tissue biopsy is unavailable. The pathogenesis of GLA involves mTOR signaling within the phosphatidylinositol 3-kinase/AKT pathway, which regulates normal vascular development and angiogenesis. By inhibiting mTOR, sirolimus decreases angiogenesis and lymphangiogenesis. Sirolimus is orally administered and should be initiated once the infant tolerates feeds. Both infants were

treated with sirolimus, resulting in clinical improvement with resolution of pleural effusions and successful extubation, without adverse effects.

456 THE EFFECT OF IMPLEMENTING A PRACTICE GUIDELINE FOR METABOLIC BONE DISEASE

¹E Sabroske*, ^{1,2}C Motta, ²K Kohlleppe, ³D Payne, ²C Kather, ²C Stine, ^{1,2}M Pierce, ^{1,2,4}KA Ahmad. ¹Baylor College of Medicine, San Antonio, TX; ²Pediatric Medical Group, San Antonio, TX; ³University of Texas Health Science Center, San Antonio, TX; ⁴MEDNAX, Sunrise, FL

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Purpose of study Metabolic bone disease (MBD) has been estimated to occur in 16% to 40% of very preterm infants. In 2015 we implemented a policy regarding the screening, diagnosis, and management of MBD at The Children's Hospital of San Antonio. We hypothesized this would lead to more frequent screening and detection of MBD in VLBW infants.

Methods used Retrospective review of VLBW infants admitted to the NICU at The Children's Hospital of San Antonio for the two calendar years prior to (2013 and 2014) and after implementation (2016 and 2017). We included all VLBW patients surviving at least 4 weeks after birth (excluding infants with major birth defects). We reviewed demographics, timing and results of MBD screening laboratory tests, therapies, and outcomes. Statistics utilized included students t-test and chi squared.

Summary of results A total of 65 VLBW infants were studied prior to policy implementation and 78 after. These cohorts had similar gestational age at birth (28.2 ± 2.9 vs 27.8 ± 3 weeks) and birth weights (1051 ± 272 vs 1043 ± 255 grams). After policy implementation we found significantly increased rates of patients having alkaline phosphatase (AP, 55.4% vs 92.1%), phosphorous (49.2% vs 92.1%), total calcium (53.8% vs 88.2%), and vitamin D levels evaluated (7.7% vs 34.2%, all $p < 0.0001$) after 21 days of age. Enteral supplemental of Vitamin D by 14 days of age increased from 21.5% to 39.5% ($p = 0.029$) with the median age for initiation of Vitamin D decreasing from 26 days to 15 days ($p < 0.001$). The median number of AP levels obtained increased from 1 to 4 ($p < 0.0001$). There was a non-significant decrease in the percent of patients with an AP > 800 (16.7% vs 7.1%, $p = 0.18$) and no rickets was identified by X-ray.

Conclusions Implementation of a standard prevention and management protocol for MBD was associated with a significant increase in enteral Vitamin D supplementation by 14 days of age and a significant increase in the frequency of screening lab utilization. The percentage of infants with laboratory evidence of osteopenia of prematurity decreased, but not significantly.

457 DAY-TO-DAY VARIABILITY IN HUMAN MILK USE DURING THE FIRST 2 WEEKS AFTER BIRTH AND GROWTH OUTCOMES OF EXTREMELY PRETERM INFANTS AT 36 WEEKS OF POSTMENSTRUAL AGE

AA Salas*, ML Jerome, M McLaughlin, AB Attawala, WA Carlo. *University of Alabama at Birmingham, Birmingham, AL*

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Purpose of study Human milk use as primary source of enteral nutrition for extremely preterm infants has been

associated with poor in-hospital growth when compared with preterm formula. However, by reducing feeding intolerance, early human milk use either as mother's own milk (MOM) or donor human milk (DHM) could offer important advantages to promote higher daily enteral intake and improve growth outcomes of extremely preterm infants. The objective of this study was to determine the association between day-to-day variability in human milk use during the first 14 days after birth and growth outcomes of extremely preterm infants at 36 weeks of postmenstrual age (PMA).

Methods used In this secondary analysis, data on daily intake of maternal and donor milk during the first 14 days after birth were collected from medical records of extremely preterm infants that had growth outcome data at 36 weeks PMA. Human milk intake in infants that developed postnatal growth restriction at 36 weeks PMA (weight below the 10th percentile) was compared to human milk intake in infants that did not develop growth restriction at 36 weeks PMA. A linear regression analysis was performed to account for differences in baseline characteristics between groups.

Summary of results Of 50 extremely preterm infants included in this analysis, 22 had normal postnatal growth (referent group) and 28 had restricted postnatal growth at 36 weeks PMA (PGR group). The mean birthweight was 887 g (SD: 240 g) and the median gestational age was 27 weeks (IQR: 25–28). Birthweight, gestational age, and weight-for-age Z-score at birth differed significantly between groups. Day-to-day variability in MOM/DHM ratios did not differ between groups. In an analysis adjusted for weight Z-score at birth, sex, and daily human milk intake, higher cumulative human milk intake between postnatal day 7 and postnatal day 14 was associated with higher weight Z-scores at 36 weeks PMA ($p=0.04$).

Conclusions Higher intakes of human milk between postnatal day 7 and postnatal day 14 are associated with a lower risk of postnatal growth restriction at 36 weeks PMA in extremely preterm infants. This association appears to be irrespective of the proportion of enteral intake as mom's own milk or donor milk.

458 LUTEIN IMPROVES NEURO-BEHAVIORAL OUTCOME IN NEWBORN RATS WITH HYPOXIC ISCHEMIC (HI) BRAIN INJURY

D Sambalingam*, X Gong. *Texas Tech University Health Sciences Center, El Paso, TX*

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Purpose of study The aim of the study is to assess effect of lutein supplementation in neuro-behavioral outcomes in newborn rats with HI brain injury

Methods used Newborn rat Vannuchi model of perinatal HIE was adopted where the interventional rats are subjected to unilateral carotid ligation on PND 7% and 8% hypoxia for 1.5 hours. SD rat pups were randomised into 3 groups – Control rats, HI and HI +Lutein. The pregnant dams were treated with lutein special diet (200 μ mol/day) and newborn rats are treated with 0.5 ml intra-peritoneal lutein injections (30 mg/kg) from PND 8–10. Neuro-developmental tests were performed in weeks 3 and 6 of life. The tests were focused on particular regions of maximal injury. Measurement of Forelimb Use Asymmetry test (spontaneous motor activity) forelimb

Abstract 458 Figure 1 Cylinder test (Forelimb use asymmetry test)

Placing test and corner turn test (sensorimotor functional behavior).

Summary of results 13 newborn rat pups born to single Dam in 2 different pregnancy cycles were used. In this study, neonatal HI brain injury mice showed motor deficits that corresponded to left hemisphere damage. Lutein treated group performed significantly better than the HI group in neuro-behavioral testing.

Conclusions Prenatal and post-natal supplementation of lutein improves neuro-behavioral outcomes in perinatal HI injury.

459 CASE OF NEONATAL FATALITY FROM NEUROMUSCULAR VARIANT OF GLYCOGEN STORAGE DISEASE TYPE IV

T Sandhu*, M Polan, Z Yu, A Makkar. *The University of Oklahoma, Oklahoma City, OK*

10.1136/jim-2018-000974.457

Case report Glycogen storage disease type IV (GSD-IV), or Andersen disease, is a rare autosomal recessive disorder that results from the deficiency of glycogen branching enzyme (GBE). This in turn results in accumulation of abnormal glycogen molecules that have longer outer chains and fewer branch points. GSD-IV manifests in a wide spectrum, with variable phenotypes depending on the degree and type of tissues in which this abnormal glycogen accumulates. Typically, GSD-IV presents with rapidly progressive liver cirrhosis and death in early childhood. However, there is a severe congenital neuromuscular variant of GSD-IV that has been reported in the literature, with fewer than 20 patient cases thus far. We report an unusual case of GSD-IV neuromuscular variant in a late preterm female infant who was born to non-consanguineous healthy parents with previously healthy children. Prenatally, our patient was found to have decreased fetal movement and polyhydramnios warranting an early delivery. Postnatally, she had severe hypotonia and respiratory failure, with no hepatic or cardiac involvement. Extensive metabolic and neurological workup revealed no abnormalities. However, molecular analysis by whole exome sequencing revealed two pathogenic variants in the *GBE1* gene. Our patient was thus a compound heterozygote of the two pathogenic variants: one of these was inherited from the mother [p.L490WfsX5 (c.1468delC)], and the other pathogenic variant was a *de novo* change [p.E449X (c.1245G>T)]. As expected in GSD-IV, diffuse intracytoplasmic periodic acid schiff positive, diastase-resistant inclusions were found in the cardiac myocytes, hepatocytes, and skeletal muscle fibers of our patient.

460

ROUTINE USE OF SMOF LIPID REDUCES TREATMENT OF PARENTERAL NUTRITION ASSOCIATED CHOLESTASISA Schwalm*, M Sorrell, C Klesel, M Piepkorn. *Texas Tech Health Science Center, Lubbock, TX*

10.1136/jim-2018-000974.458

Purpose of study Parenteral nutrition associated liver disease (PNALD) accounts for significant morbidity in neonates receiving prolonged parenteral nutrition (PN). SMOF lipid (SMOF) has been shown to improve liver function in neonates with PNALD compared to soy based lipid emulsions and was found to be efficacious and safe when used as the sole lipid source in pediatric patients receiving PN. Therapies for PNALD, including ursodiol and phenobarbital, are often ineffective, so prevention of PNALD remains the most effective strategy. The purpose of this retrospective cohort study was to evaluate the safety and efficacy of routine SMOF administration to neonates who received PN compared to a historical cohort of neonates who received only Intralipid (IL).

Methods used All neonates who received PN starting in October of 2017 received SMOF at a dosage ranging from 0.5 g/kg/day to 2 g/kg/day based upon the clinical discretion of the neonatologist. The dosage of SMOF was then adjusted daily to a max ranging from 3 g/kg/day to 3.5 g/kg/day. Clinical data including liver function testing, growth trends, common neonatal morbidities, and the use of ursodiol and phenobarbital was collected. This data was then compared to a historical cohort of neonates who received IL as their lipid source. Infants who received 7 days or more of PN were analyzed. A *p*-value of <0.05 was used as threshold for statistical significance.

Summary of results Demographic data including gestational age, birthweight, and gender was similar between groups. Total days of PN was also similar between groups. The use of ursodiol and/or phenobarbital was significantly reduced in the SMOF group compared to IL (9.8% vs. 21.4%, *p*<0.05). The mean number of days ursodiol and phenobarbital was prescribed was reduced in the SMOF group (13 days vs. 29 days, 18 days vs. 34 days respectively, *p*<0.05). Liver function testing, growth trends, and common morbidities including chronic lung disease, central line infections, PDA, and ROP was similar between groups.

Conclusions The reduction in the use of ursodiol and/or phenobarbital supports the continued routine use of SMOF instead of Intralipid in neonates. Similar rates of common neonatal morbidities and growth trends suggests SMOF is safe to use in critically ill neonates and supports necessary postnatal somatic growth.

461

INFANTILE HEMANGIOMAS MASQUERADING AS CONGENITAL HYPOTHYROIDISMBL Scott*, JB Cahill. *Medical University of South Carolina, Charleston, SC*

10.1136/jim-2018-000974.459

Case report An eight-week-old premature female presented with feeding intolerance and abdominal distension relieved by intermittent bowel movements. She had been unable to tolerate human milk fortifier and various formula trials, including elemental formula. She was otherwise well with two normal newborn screens, obtained at two and thirty days of life. Her physical exam was notable for a distended abdomen with

normal bowel sounds and no organomegaly. Abdominal radiographs revealed gaseous distension throughout her gastrointestinal tract. A contrast enema was notable for redundant sigmoid colon without colonic strictures. A rectal biopsy ruled out Hirschsprung's disease. New onset mild hypothermia coupled with constipation led to a clinical concern for hypothyroidism. Screening thyroid stimulating hormone was high while free thyroxine was normal. The patient was immediately started on levothyroxine for suspected congenital hypothyroidism missed by newborn screen. A later plain film suggested hepatomegaly, and a subsequent ultrasound identified innumerable hypoechoic lesions throughout the liver concerning for hemangiomas. Her skin exam revealed numerous very small hemangiomas. Echocardiography and bronchoscopy were unremarkable. The dermatologic and ultrasound findings prompted initiation of propranolol for infantile hemangiomas (IH). Her feeding intolerance is slowly improving, and she is steadily increasing the dose of propranolol with plans for repeat hepatic imaging and thyroid studies.

Infantile hemangiomas occur in 5% of the population with premature infants at higher risk than term infants. Complications include ulceration, sensory impairments, congestive heart failure due to arteriovenous shunting, airway obstruction and hypothyroidism. Hypothyroidism is specifically found with diffuse hepatic IH, due to production of type 3 iodothyronine deiodinase by the hemangiomas, which catalyzes the conversion of thyroxine to reverse triiodothyronine, and triiodothyronine to 3'3'-diiodothyronine; both are biologically inactive. Despite normal newborn screens the patient was profoundly hypothyroid most likely from widespread hepatic IH. Providers should be aware of the associations between IH and prematurity, and the relationship between diffuse hepatic hemangiomas and clinical hypothyroidism.

462

IDENTIFYING DIAGNOSTIC ERRORS IN THE NEONATAL INTENSIVE CARE UNIT USING THE SAFER DX NICU INSTRUMENT: A PILOT STUDY^{1,2}G Shafer*, ^{1,3}H Singh, ^{1,2}G Suresh. ¹Baylor College of Medicine, Houston, TX; ²Texas Children's Hospital, Houston, TX; ³Michael E. DeBakey VA Medical Center Department of Veteran Affairs, Houston, TX

10.1136/jim-2018-000974.460

Purpose of study To test the feasibility of a new method to determine the frequency of diagnostic errors in the Neonatal Intensive Care Unit (NICU).

Methods used Retrospective chart review of 60 randomly-selected NICU admissions from 2015–2018 using a data collection tool – adapted version of the Safer Dx Instrument – with questions to evaluate diagnostic processes and identify presence/absence of diagnostic errors. A trained physician used the instrument to review the electronic medical record for the first 7 days of NICU admission, and categorized cases into 'yes,' 'unclear,' or 'no' for diagnostic error. After all charts were reviewed by the primary reviewer, we used a two-step process for reliability. (1) An individual blinded-review by trained secondary reviewers (4 NICU physician leaders) of all charts in the 'yes' category, plus a random sample of charts in the other categories; and (2) Non-blinded, group review by all 4 secondary reviewers of charts with discordance between primary and secondary reviewers.

Summary of results Primary review classified 5 charts as 'yes,' 8 as 'unclear,' and 47 as 'no' for diagnostic error. Blinded

Abstract 462 Table 1 Descriptions of identified diagnostic errors

Identified Diagnostic Error	Summary of the Diagnostic Error
1	Missed pneumonia on chest radiograph
2	Perforation of the small bowel after misinterpreted dilation of the bowel on abdominal radiograph
3	Missed sacral dimple with hair on physical exam
4	Missed tension pneumothorax on chest radiograph
5	Delayed diagnosis of a humeral fracture after shoulder dystocia during delivery
6	Concern for Dandy-Walker Syndrome on prenatal ultrasound not followed-up postnatally

secondary review evaluated 13 charts with discordance in 5 cases. After group secondary review, a total of 6 diagnostic errors were confirmed in 5 patients with 8% of study patients having at least 1 diagnostic error, and a rate of 1.4 errors per 100 patient days.

Conclusions Chart review using an objective instrument followed by confirmatory reviews is a feasible method to study the frequency of diagnostic errors in the NICU. We will use this method to perform a comprehensive study on a large sample (n=600) of patients.

463 DEVELOPMENT AND INTEGRATION OF A PERINATAL PALLIATIVE CARE PROGRAM AT A TERTIARY CARE CENTER

BA Shah*, R May, L White, A Wlodaver. *University of Oklahoma Health Sciences Center, Oklahoma City, OK*

10.1136/jim-2018-000974.462

Purpose of study Advances in fetal medicine have increased referrals to tertiary perinatal care centers. NRP® guidelines recommend that only neonates with anencephaly and less than 400 grams are not to be resuscitated. In 2015, Oklahoma passed a law stating 'it is the duty of the physician to inform the woman who carries a fetus with anomalies that perinatal hospice services are available and that this is an alternative to abortion'. We aim to describe the process of developing a perinatal palliative care (PPC) program for newborns with prenatal diagnoses of life-limiting conditions.

Methods used Focus groups were conducted with responsible parties. Existing resources were identified. Implementation approaches were discussed.

Summary of results The need for a PPC team was identified. Nurses, neonatologists, chaplains, social workers and child life specialists were integrated into this service with the support of an existing pediatric palliative care team. A new PPC program was introduced. Prenatal consults initiated by maternal-fetal medicine are an integral part of the prenatal care. Options of intensive care versus comfort care are offered. A neonatologist attends all these deliveries. The newborn stays with the mother. Special nurses were assigned for comfort care in the mother's room. Guidelines and specific PPC orders are now available in the electronic medical record. Examples are listed in the table 1. A website was created to inform referring physicians about this new program.

Abstract 463 Table 1

Case	Diagnosis	Outcome
1	Trisomy 18	G-tube at 4 months; alive at a 1 year
2	Alobar holoprosencephaly	VP shunt at 3 months; died one year later
3	Hypoplastic left heart with intact septum	Died at home at one week of age with hospice
4	Renal agenesis	Died at 2 days of age
5	22 weeks gestation	Died on 1 st day

Conclusions PPC was successfully established at a regional tertiary center. Parents of newborns diagnosed with lethal conditions have the choice of often non-beneficial intensive care or care focused on comfort after birth. This specialized care allows parents to celebrate the limited life of their precious newborns. Providers across the state have now access to a referral center offering perinatal neonatal palliative care.

464 MATERNAL PRENATAL FACTORS AND CHILD RETINAL VASCULATURE

S Shovers*, C Jones, J Kim, S Drury. *Tulane University, New Orleans, LA*

10.1136/jim-2018-000974.462

Purpose of study Maternal prenatal obesity and smoking increase her child's health risk possibly via changes to the infants' developing vasculature. Maternal obesity contributes to umbilical cord endothelial dysfunction, and maternal smoking has been linked to higher fetal middle cerebral artery diameter; we posit that changes in children's microvasculature (MV) may precede the development of poor health linked to maternal obesity and smoking.

Measuring retinal microvasculature (RM) is a novel and noninvasive method to measuring cerebral MV in children but has limited data.

To address the gap, we examined if maternal BMI or prenatal smoking predicted offspring venule and arteriolar diameter.

Methods used Prenatal BMI and smoking habits were self-reported and compared with medical record data and a postnatal interview at 4 months. Retinal images of children (n=30) were taken at 4 years of age and analyzed with IFLEXIS software; Central Retinal Venule Equivalent (CRVE) and Central Retinal Arteriolar Equivalent (CRAE) were calculated from an average of 6 vessels. Linear Regression compared continuous BMI to CRVE/CRAE; T-tests compared prenatal smoking to CRVE/CRAE.

Summary of results Mean maternal age was 29 (SD 5.4), BMI 27.7 (SD 7.7). Mean gestational age was 39.3 weeks (SD 1.3). 20% of mothers were positive for prenatal smoking. No relation was found for BMI and venule or arteriole diameters. Prenatal smoking was not associated with venule diameter (p=0.25) but significantly predicted child arteriolar diameter (p=0.05).

Conclusions Similar to prior studies, CRAE was significantly lower (155μ vs 172μ) for children whose mothers smoked prenatally compared to those whose did not. Smaller arterial diameter has been linked to hypertension in adults, suggesting early changes in MV due to prenatal smoking may add to elevated health risks for children.

Maternal BMI did not predict CRVE/CRAE. Small sample size may contribute to the negative finding; alternatively, MV changes may not be clear at age 4, or the relation between maternal obesity and future child health may involve other pathways.

Larger studies with repeated measures of RM coupling child BMI and blood pressure are needed to grasp the predictive value of RM and the role of MV changes in transgenerational effects. The noninvasive, inexpensive nature of RM imaging offers significant potential for possible detection of CVD or risk.

465

OPTIMUM TIME FOR RESUMPTION & ADVANCEMENT OF ENTERAL FEEDS FOLLOWING STAGE 2 NECROTIZING ENTEROCOLITIS

A Smazal*, A Massieu, J Hagan, AB Hair, MH Premkumar. *Baylor College of Medicine, Bellaire, TX*

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Purpose of study Resumption and advancement of enteral feeds following stage 2 necrotizing enterocolitis (NEC) is often not based on evidence. We hypothesized that earlier initiation and rapid advancement of enteral feedings after stage 2 NEC in neonates was associated with improved short-term outcomes.

Methods used Neonates with stage 2 NEC at our institute between 2014–2017 were included. Information on patient characteristics, enteral feeding practices, and short-term outcomes were collected. Pearson's correlation was used to assess the relationship between variables.

Summary of results The demographic and outcome data are summarized in table 1. All infants survived to discharge. One infant developed stricture. Diagnosis of NEC at a younger chronological age and not GA or PMA was associated with longer duration of fasting [NPO] ($p=0.04$). Longer duration of NPO was associated with longer duration of PN ($p=0.01$) and central venous catheter (CVC) days ($p=0.03$), but not time to reach full feeds, or length of hospitalization (LOS). Slower advancement of feeds was associated with longer LOS ($p<0.01$), duration of PN requirement ($p<0.01$) and CVC days ($p<0.01$).

Conclusions In infants with stage 2 NEC younger chronological age at the time of diagnosis of NEC, rather than GA or

Abstract 465 Table 1 Patient demographics, treatment and outcomes

Variable	
GA (wks)*	31.9±4.6
Birth weight (g)*	2021±1137
Male: female	9:6
PMA at time of diagnosis(wks)*	34.2±3.5
Age at time of diagnosis (days)*	15.7±15.4
Days to resumption of feeds (NPO)*	9.3±1.9 (Range 7 to 12)
Days to achieve full feeds*	10.8±12.5 (Range 5 to 55)
PMA at full feeds* (wk)	36.6±4.1
Duration of CVC (d)*	19.3±14
Duration of PN (d)*	23±14.4
Length of hospitalization (days)*	63.9±45.8

PMA, was associated with delayed resumption of enteral feeds. Earlier resumption and faster advancement of enteral feedings after diagnosis of stage 2 NEC could shorten LOS, duration of PN and CVC days. We are extending the study time frame to include additional years to increase the study population.

466

EVALUATION OF ANTIBIOTIC USE FOR LATE-ONSET SEPSIS IN A LEVEL 3 NICU

WH Stafford*, K Upadhyay, AJ Talati. *University of Tennessee Health Science Center, Memphis, TN*

10.1136/jim-2018-000974.464

Purpose of study Nonspecific clinical symptoms lead to frequent evaluation for late-onset sepsis (LOS) and subsequent antibiotic use in the NICU. Overuse of abx can lead to several problems. The purpose of this study was to evaluate abx use in our NICU for LOS and identify signs and symptoms that lead to antibiotic use.

Methods used Charts were reviewed for LOS after identification of babies by use of vancomycin initiation. Data were collected regarding symptoms, abx duration, and laboratory values from January 1, 2015 to December 31, 2017. Data were divided into three cohorts based on year.

Summary of results 235 babies with 352 events were identified with characteristics shown in table 1. Table 2 shows abx use patterns. Abx initiation decreased overall, but 39% of babies with a negative culture were still receiving abx for greater

Abstract 466 Table 1 Cohort characteristics

Year	2015	2016	2017
Total Admissions (n=3028)	1095	979	954
Infants Receiving ABX (n=235)	94 (8.6%)	81 (8.3%)	60 (6.3%)
Sepsis Evaluations (n=352)	152 (13.9%)	111 (11.3%)	89 (9.3%)*
Median EGA (Completed Weeks)	28 (23, 31)	27 (25, 30)	27 (25, 30)
Mean Birth Weight (gms)	1195.3±734.0	1075.3±559.1	1011.0±655.5
Male(%)	44 (46.8%)	37 (45.7%)	31 (51.7%)

* – $p=0.0015$ (2015 to 2017)

Abstract 466 Table 2 Evaluation data

Year	2015	2016	2017
Negative Culture, No NEC/PNA/Death (#)	91 (59.9%)	63 (56.8%)	41 (46.1%)
Negative Culture receiving >48 hours ABX (#)	59 (64.8%)	32 (50.8%)	16 (39.0%)*
Average Peak CRP in Negative Culture (mg/L)	4.9±7.2	4.3±6.2	3.8±7.2
Time on ABX with Negative Culture, No NEC/Death/PNA (Hours)	70 (38, 138)	48.4 (36.6, 91.9)	39.2 (34.4, 92.6)**
Positive Culture±NEC (#)	50 (32.9%)	41 (36.9%)	33 (37.1%)
Duration of Therapy, All evaluations (Hours)	120 (50, 230)	72.4 (39.7, 178.4)	101.1 (37.2, 168.4)
Day of Life at Evaluation (Days)	21 (13, 36)	25 (13, 41)	25 (15, 47)
Deaths while on ABX (#)	10 (6.6%)	11 (9.9%)	4 (4.5%)

* – $p=0.0077$ (2015 to 2017); ** – $p=0.00804$ (2015 to 2017)

than 48 hours. Symptoms like anemia, asymptomatic elevation in CRP, and hypoglycemia do not seem to be associated with true infection.

Conclusions There was an overall decrease in the number of evaluations for LOS requiring abx. The number of babies with negative culture receiving abx greater than 48 hours decreased, but a significant number of babies are still receiving abx for longer than 48 hours in a culture negative setting. Non-specific signs and symptoms may not be associated with a true infection.

467 CHROMOSOME 13Q31 DELETION SYNDROME

S Staples*, J Phillips. *University of Alabama, Birmingham, AL*

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Introduction Improved prenatal genetic diagnosis allows families time for counseling and delivery planning. However, identification of a rarely documented condition leaves families and providers with prognostic uncertainty and limited anticipation of possible postnatal complications.

Case presentation A term female was delivered via cesarean for IUGR and occipital encephalocele without brainstem involvement. Additional anomalies on prenatal imaging included abnormal brain tissue, microcephaly, and VSD. Amniocentesis revealed a 13q31 à QTER deletion. Infant required intubation at birth. Physical exam was notable for IUGR <1%, microcephaly with head circumference 50% for 25 WGA infant, small closed midline occipital encephalocele, hypotelorism, low-set ears, absent thumbs, absent toe on left foot with bilateral clubbed feet and decreased spontaneous movement. Brain MRI was suggestive of but not typical of lobar holoprosencephaly. Echo revealed muscular VSD, bilateral SVC and right aortic arch. Skeletal survey showed proximal fusion of radius and ulna bilaterally and the absent digits. Ophthalmologic exam and renal ultrasound were normal. Infant was unable to wean from the ventilator and given poor prognosis family chose compassionate extubation allowing natural death at 10 days of age.

Discussion Chromosome 13 deletion syndrome was first described in 1969 with variable phenotype including developmental delays, facial anomalies, limb malformations, CNS defects including posterior encephalocele, holoprosencephaly and NTDs, cardiac, eye, lung, kidney, GU and GI tract anomalies. Clinical characteristics and severity depend on the location and size of the deleted region, but specific genotype-phenotype relationships for anomalies have not been elucidated due to rarity of the disorder.

Conclusion Chromosome 13 deletion syndrome demonstrates significant phenotypic variance and like other rare disorders, insufficient experience makes prognosis challenging particularly when diagnosed prenatally. Genetic testing is important for early intervention and management and may have implications for long-term family planning, however, counseling should be considered prior to diagnostic amniocentesis given the risks of the procedure in addition to the uncertainty of identifying a causative gene or being able to predict outcomes.

468 ENDOPLASMIC RETICULUM STRESS RESPONSE IN VASCULAR ENDOTHELIAL CELLS OF INFANTS WITH PLACENTAL INFLAMMATION

TL Summerlin*, C Ren, P Crowder, H Wongprasert, B Halloran, N Ambalavanan, T Jilling. *University of Alabama at Birmingham, Gardendale, AL*

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Purpose of study Endoplasmic reticulum (ER) stress is a physiological cellular response to metabolic and environmental insults such as inflammation. ER stress dysregulation contributes to endothelial cell dysfunction. Chorioamnionitis, an inflammatory disorder of the chorion, amnion and placenta, is associated with adverse neonatal outcomes. Objective: Establish primary cultures of human umbilical vein endothelial cells (HUVECs) obtained from pregnancies with and without chorioamnionitis to determine if chorioamnionitis increases endothelial ER stress.

Methods used 25 umbilical cords were collected from term pregnancies in which placental pathology was evaluated. HUVECs were isolated, exposed at early passages *in vitro* to hyperglycemia (30 mM glucose) or tunicamycin for 24 hour and then analyzed for ER stress. ER stress was estimated by Western Blotting for GRP78, P-PERK, ATF4 and CHOP, proteins associated with the unfolded protein response (UPR) during ER stress.

Summary of results We report preliminary findings on 4 HUVEC isolates for which placentas had confirmed inflammation, and 4 for which placentas did not have inflammation on histology. GRP78, P-PERK and ATF4 were detected on Western Blots, but no CHOP signal was present. ATF4 band intensity was increased by both hyperglycemia and tunicamycin. Tunicamycin induced the most ER stress, based on band intensity, and hyperglycemia induced a milder ER stress. GRP78 and P-PERK appeared unaffected by the stimuli. No significant differences were seen between the no inflammation and inflammation groups.

Conclusions Although no significant differences were observed between the two groups to date, we established a model to determine whether placental inflammation correlates with altered ER stress signaling in HUVECs. Changes in ATF4 quantity were the most robust in response to stressors at the 24 hour time point post-stimulus. ER stress in response to hyperglycemia in HUVECs has implications for future studies on the effects of hyperglycemia in endothelial dysfunction, possibly in infants of diabetic mothers or hyperglycemia secondary to sepsis.

469 CONGENITAL MYASTHENIC SYNDROME 6

TL Summerlin*, J Phillips. *University of Alabama at Birmingham, Gardendale, AL*

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Introduction Newborns with hypotonia can have abnormal neuromuscular transmission, leading to muscle fatigability and weakness at birth. Though rare, congenital myasthenic syndromes (CMS) can cause neuromuscular junction failure in newborns presenting with hypotonia and weakness.

Case presentation A 33 wga male was born to a 30yo G2P1 via C/S due to decreased heart tones. Pregnancy was

complicated by maternal obesity, chronic hypertension, A2DM and mild polyhydramnios, otherwise normal US. Mom received 2 doses of betamethasone. Infant was born with APGARs 1, 2, requiring intubation and surfactant. Cord gases were >7.24. Exam showed diffuse edema, arthrogryposis, hypotonia with trace spontaneous movements, absent reflexes, single palmar creases and club feet. Neurology and genetics evaluated infant for a neuromuscular disorder. An extensive work up was negative for TORCH, chromosomal abnormalities, SMA, congenital myotonic dystrophy, MuSK and ACh-R Abs. He had a normal eye exam, CPK level, EEG and brain/spine MRI. Whole genome sequencing showed 2 mutations in the choline acetyltransferase (CHAT) genes and the diagnosis of CMS 6. Infant was started on an acetylcholinesterase inhibitor, which led to mild improvement in his apnea and spontaneous movements. Infant remains ventilator-dependent, with ultimate plans for g-tube and tracheostomy placement.

Discussion CMS are rare but important disorders characterized by a dysfunction in neuromuscular transmission leading to hypotonia in infancy. Diagnosis is usually made with targeted genetic testing to determine the CMS subtype, which guides treatment, prognosis and genetic counseling. Genes commonly associated with CMS are CHAT, CHRNE, COLQ, DOK7, GFPT, and RAPSN. The CHAT subtype is autosomal recessive and can present with arthrogryposis, hypotonia and respiratory failure. Stress-induced apnea is common with CHAT and RAPSN, therefore prophylactic anticholinesterase therapy is recommended. Severity and prognosis are variable between CMS subtypes. Early diagnosis of the CHAT subtype can lead to potentially life-saving treatments, and some symptom improvement are expected with age.

Conclusion CMS should be considered in hypotonic infants with an otherwise negative extensive work up. Differentiating between the CMS subtypes can guide treatment strategies and determine prognosis.

470 CHARACTERISTICS OF EXTREMELY LOW BIRTH WEIGHT INFANTS REQUIRING VASOPRESSORS IN 1ST WEEK OF LIFE

P Zaveri*, AM Walker, K Upadhyay, AJ Talati. *UTHSC, Memphis, TN*

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Purpose of study A significant variability exists for diagnosis and treatment of hypotension in ELBW infants. Benefits of use of vasopressors (VP) remain unclear. We wanted to identify the risk factors associated with use of VP in the 1st week of life and their impact on outcomes of ELBW infants.

Methods used Retrospective review of all newborns ≤28 weeks EGA and admitted in NICU from Oct 1, 2012 to Oct 31, 2015 was done. Data regarding antenatal and neonatal characteristics and outcomes were recorded. Study infants were divided into 2 cohorts and compared based on VP use. Chi-square, t-test and multiple logistic regression were performed as appropriate and significance set at $p < 0.05$.

Summary of results Of 213 ELBW infants, 90 (42.3%) received VP in 1st week of life. Antenatal and neonatal characteristics are listed in table 1. The mean blood pressure at admission in these infants was significantly lower than that of non-VP group. (27 ± 8 vs 30 ± 6 mm Hg, $p < 0.05$). VPs were initiated within 24 hour in 91% of babies. After controlling for other variables, use of VP was significantly higher in lower

Abstract 470 Table 1

	VP (n=90)	No VP (n=123)
EGA (median wk)*	25	26
BW gm (mean±SD)*	668±173	796±209
Male (n,%)	51 (56%)	58 (47%)
African American (n,%)	81 (90%)	106 (86%)
C-sec delivery (n,%)*	71 (78%)	79 (64%)
multiple births (n,%)*	23 (25.5%)	15 (12%)
Antenatal steroids (n,%)	86 (95.5%)	114 (92.7%)
SGA (n,%)*	29 (32%)	18 (14.6%)
5 min Apgar (median)	5	6
Admission hypothermia (n,%)*	22 (24%)	16 (13%)
Mean arterial pressure on admission (mean, SD)*	27 mmHg (SD-8)	30 mmHg (SD-6)

* p

birthweight (OR 3.2, CI [1.6 to 8.3]), with 5 min Apgar score ≤5 (OR 1.8, [CI 1.2–3.12]) and admission hypothermia (OR 2.7, CI [1.3 to 4.9]). The use of VP was significantly associated with severe IVH, even after controlling for other significant variables (OR 5.9, CI [1.6 to 9.3]).

Conclusions Lower birth weight, low 5 min Apgar score and admission hypothermia are risk factors for early use of VPs in ELBW infants. Infants treated with VPs are at a higher risk of developing severe IVH.

471 A RISK-BASED ALGORITHM FOR INFANTS EXPOSED TO MATERNAL CHORIOAMNIONITIS IMPROVES ANTIBIOTIC STEWARDSHIP

S Vander-Plas*, M Piepkorn, M Sorrell. *Texas Tech Health Science Center, Lubbock, TX*

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Purpose of study Maternal chorioamnionitis (MC) is a known risk factor for early onset neonatal sepsis. The AAP currently recommends empiric antibiotic therapy for all neonates exposed to MC. Universal empiric antibiotics may expose low risk neonates to unnecessary antibiotics, alter the development of the microbiome, increase costs, and reduce mother-infant bonding. However, the potential mortality of neonatal sepsis can not be overlooked. The purpose of this study was to develop and implement a simple, risk based algorithm for the management of infants exposed to MC in an attempt to improve antibiotic stewardship.

Methods used An algorithm was developed to risk stratify infants exposed to MC. Risk factors included gestational age (GA), duration of rupture of maternal membranes, infant's complete blood count at 6 and 24 hours of life, and clinical status based on 48 hours of observation in the NICU. High risk infants received 36–48 hours of empiric ampicillin and gentamicin. Low risk infants remained in the NICU for observation but did not receive empiric antibiotics. Maternal and neonatal demographic, clinical, and laboratory data was collected and compared to a historical, pre-intervention (PRE) cohort of neonates exposed to MC, all of whom received empiric antibiotic therapy.

Summary of results A total of 45 neonates exposed to MC were identified in the PRE group and 26 neonates in the post-intervention period. Infants exposed to MC following algorithm implementation received antibiotics at a lower rate compared to the PRE group (73% vs. 100%, $p < 0.01$). No

significant differences were observed between the two groups in GA, birth weight, white blood cell count, immature-to-total neutrophil ratio, absolute neutrophil count, or length of stay. Blood culture results remained negative for all patients in both groups.

Conclusions Empiric antibiotic use was reduced in infants exposed to MC following implementation of our risk-based algorithm. All blood cultures remained negative in both groups suggesting empiric therapy based on MC may be unnecessary. Future approaches to the management of infants exposed to MC may include initial observation only followed by antibiotic therapy based on postnatal clinical status potentially improving both antibiotic stewardship and preventing NICU admission for MC alone.

472 NONINVASIVE GLUCOSE MEASUREMENT IN NEWBORN INFANTS

¹D Varma*, ¹J Carns, ¹L Zhang, ¹K Gerhardt, ¹G Bao, ¹J Tabor, ²AM Khan, ²EW Reynolds, ¹R Richards-Kortum. ¹Rice University, Houston, TX; ²University of Texas Health Science Center Houston, Houston, TX

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Purpose of study Serum glucose measurement in neonates requires blood sampling via heelstick. Previous studies have reported transcutaneous glucose (TG) measurements in adults and preschool-age children. Higher transepidermal water loss (TEWL) (~30 g/m²/hr) from preterm neonate skin suggests that this method may be useful in neonates and decrease the need for heelsticks. This study investigates the feasibility of measuring TG in neonates and develops a standard collection technique.

Methods used 6 neonates (36–38 weeks gestation) undergoing serum glucose monitoring as part of their medical care were included. 250 µL of phosphate buffered saline (PBS) was held against the infant's palm or forearm skin via 2 different collection methods: (1) PBS was held in an inverted Eppendorf tube against the skin for 2 and 5 min; (2) PBS was held in a well, created by an adhesive ring for two 5 min periods. The samples were frozen for later processing. Measurements were made with a glucose binding protein (GBP) tagged with a fluorescent dye, allowing glucose measurement in the µmolar range. TEWL of the sites was measured with a Vapometer.

Summary of results Mean TEWL of 11.65±3.38 g/m²/hr suggested that the infant skin was mature. Mean TG was 0.73

Abstract 472 Figure 1 TG vs serum glucose

±0.58 µM but did not statistically correlate with serum glucose levels (figure 1). Early efforts were hampered by sample loss. Sample recovery improved by changing the site and sampling method.

Conclusions This is the first report of noninvasive TG measurements in infants using GBP. Low TEWL suggests limited diffusion of glucose across the skin and warrants further study in more preterm infants. Improvements made to the sampling technique developed in this pilot study can serve as the foundation for future studies of this method.

473 ATTITUDES, COMFORT LEVEL, AND KNOWLEDGE OF NICU HEALTH CARE WORKERS TOWARDS CARING FOR NEONATES WITH DISORDERS OF SEXUAL DEVELOPMENT

¹M Walsh*, ^{1,2}A Diaz-Thomas, ^{1,3}K Upadhyay, ¹A Eddington. ¹University of Tennessee Health Science Center, Memphis, TN; ²LeBonheur Children's Hospital, Memphis, TN; ³Regional One Health, Memphis, TN

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Purpose of study Disorders of sexual development (DSD) occur at a frequency of 1 in 4500 to 5500 live births. Newborn nursery healthcare workers are a chief point of medical contact for these neonates and their families. For families of DSD patients, the time spent in the neonatal nursery can be filled with uncertainty. For newborn healthcare workers at our hospital, an educational intervention dedicated to the care of DSD patients and families does not exist. Our purpose in this study is to assess current attitudes, comfort levels, and knowledge of newborn healthcare workers towards caring for DSD patients. Our aim is to create a standardized educational program to prepare workers to care for these patients. Specifically, we aim to emphasize the psychosocial support and an interdisciplinary care team model that is essential to offer these patients in addition to traditional medical services.

Methods used Study population includes nurses, nurse practitioners, and other healthcare workers employed in our perinatal center (level III) and children's hospital (level IV) neonatal ICU. Participants will receive a pre-post educational questionnaire to assess knowledge gained from a recorded educational presentation on DSD. The recorded presentation focuses on the pathophysiology behind DSDs, commonly encountered DSDs and complications, and the role of psychosocial support. The pre-post educational questionnaire consists of twenty-six questions divided into three domains: previous experience, current knowledge of normal and atypical genital development, and level of comfort in caring for these patients. Participants will complete the surveys and lecture from personal or work devices.

Summary of results This survey will be sent to an estimated 200 nurses, physicians, and other healthcare providers. Results from the pre-post educational surveys will be recorded and assessed for changes in knowledge following the educational program.

Conclusions We expect to see substantial improvement in knowledge and comfort levels via the post-educational survey. Future goals include implementing a standardized educational program to improve NICU knowledge of DSD to enhance patient care.

474

THE INTESTINAL COMMENSAL BACTERIA PROMOTE RESISTANCE TO LUNG INJURY IN EXPERIMENTAL BRONCHOPULMONARY DYSPLASIA

¹KA Willis*, ²D Siefker, ³MM Aziz, ³CK Gomes, ³JF Pierre, ²SA Cormier, ¹AJ Talati. ¹The Univ. of Tennessee Health Science Center, Memphis, TN; ²Louisiana State University, Baton Rouge, LA; ³The University of Tennessee Health Science Center, Memphis, TN

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Purpose of study We investigated if alteration of the intestinal microbiota by maternal antibiotics exposure (MAE) influenced the development of hyperoxic lung injury in neonatal mice

Methods used We randomized pregnant C57BL/6J mice on the tenth day of gestation to MAE (penicillin 500 mg/L) or control. After birth, offspring were randomized to hyperoxia (FiO₂ 0.60 or 0.85) or control (air) for 14d. Alveolarization was analyzed by mean linear intercept (MLI), alveolar heterogeneity (D₂) and septal thickness. Pulmonary fibrosis was characterized by immunohistochemical staining. Inflammasome activation was quantified. The colonic microbiome was analyzed by 16S rRNA amplicon sequencing and predictive metagenomics (PICRUSt).

Summary of results MAE induced mild pulmonary fibrosis independent of oxygen exposure. In combination with FiO₂ 0.60, MAE increased the MLI, D₂, septal thickness and fibrosis. When combined with FiO₂ 0.85, interalveolar septal thickening from fibrosis was so severe that MLI and D₂ were only minimally elevated. Mortality also increased with the combined exposure. Oxygen, but not MAE, increased inflammasome activity. As quantified by qRT-PCR, alpha diversity, and principle component analysis, MAE reduced abundance and diversity of the gut microbiome in pups. PICRUSt predicts extensive metagenomic changes resulting from MAE.

Abstract 474 Figure 1 Maternal antibiotic exposure alters lung architecture

Conclusions The commensal intestinal bacteria influence BPD development in our mouse model of neonatal hyperoxic lung injury.

475

ACCURACY OF POINT OF CARE BLOOD GLUCOSE MEASUREMENTS IN HEALTHY NEWBORN INFANTS

AJ Zayek*, R Bhat, M Zayek. College of Medicine, Mobile, AL

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Purpose of study Point-of care (POC) glucose estimation is widely used across newborn nurseries although these bedside

glucometers are not reliable at low blood glucose (BG) levels usually seen in healthy newborn infants. At present, various hospitals have different critical BG values below which a blood sample is routinely sent for confirmation. These cut-off BG values may range from 40 to 50 mg/dl. Our nursery cut-off is a POC glucose value of 50 mg/dl.

The purpose of this study is to assess the accuracy of POC blood glucose values ranging from 40 to 49 mg/dl. Thus we paired them with confirmatory blood glucose values measured by a Rapidlab 1265 analyzer.

Methods used We retrospectively paired 220 POC blood glucose levels (Accu-Check;Inform-II, Roche) with their respective 220 laboratory confirmed BG levels which were obtained from healthy newborn infants with risk factors for hypoglycemia and measured by a Rapidlab 1265 analyzer (Siemens). Collection and analysis time were retrieved from all infants' charts. Samples were excluded if lapse of collection time between the 2 paired specimen was >10 min or if the sample for confirmation was analyzed >20 min from collection.

Summary of results A total of 53 blood sample values were excluded. To examine the agreement between these 2 methods, the 167 paired glucose values were examined on a Bland-Altman plot. The mean values of confirmatory BG values were 3.6 mg/dl lower than POC glucose values, showing a systematic difference. Among newborn infants with POC glucose values of 40 mg/dl and 49 mg/dl, 50% and 15% had a BG levels <40 mg/dl on a confirmatory blood sample, respectively. In addition, 23% and 4% of these infants had a confirmatory BG levels <35 mg/dl, respectively.

Conclusions When using a POC glucometer, the critical cut-off point for low levels of BG should not be set at a BG value below 50 mg/dl. Even a cut-off point >50 mg/dl could be considered because undetected hypoglycemia may lead to neurological impairment.

Population Health and Precision Medicine
Joint Plenary Poster Session and Reception
4:30 PM
Thursday, February 21, 2019

476

LONG NON-CODING RNA IN FABRY DISEASE DIAGNOSIS AND THERAPY

MA Al-Obaide*, T Vasylyeva. Texas Tech University Health Science Center, Amarillo, TX

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Purpose of study So far, mutations in α -galactosidase-A enzyme encoded by the *GLA* gene are the main cause of the Fabry disease. MicroRNAs are evolving as effective functional regulators in Fabry disease (Oncotarget 2018, 9:27333–27345). Although long non-coding RNAs (lncRNAs), 200 nucleotides to multiple kilobases in length, showed critical functions in the tissue-specific regulation of gene expression (Nature Rev. Genetics 2011, 12:861–874), there are no reports on their influence on *GLA* expression. The objective of this study was to investigate the unexplored regulatory function of lncRNAs in the expression of the *GLA* gene.

Methods used Identification of un-reported miRNA mature sequences (miR) of three lncRNA transcripts performed by using miRBase BLASTN search tool sequence option (NAR 2014, 42:D68-D73). The miR recognition element (MRE) in the *GLA* mRNA analyzed and identified by RNA22 v2 micro-RNA target detection tool (Cell 2006, 126:1203–1217). The lower E-value or p-value represent, the greater chance that the loci contain a valid MRE.

Summary of results The *GLA* gene showed differential expression in the kidney, brain, heart, liver and other tissues, which is an indication of a tissue-specific pattern associated with promoters, enhancers, and lncRNA. The lncRNA can produce miR sequences that can target the predicted MRE of *GLA* mRNA at 3-UTR region or exons. Up-to-date there is no report on the regulatory functions of lncRNAs in the *GLA* gene. Our analysis identified several miR sequences originated from lncRNA transcripts that can target *GLA* mRNA. For example, the identified miR sequences of lncRNAs LOC105373233, LINC02358, and WD repeat containing antisense to TP53 (WRAP53/TP53-AS) can target the identified MRE of the *GLA* NM_000169.2 mRNA and consequently downregulate the α -galactosidase-A enzyme production.

Conclusions This study provided an insight into the functions of lncRNA to downregulate *GLA* gene expression at transcriptional and posttranscriptional levels in various tissues. The study showed the potential of lncRNA use in the diagnosis and therapy of Fabry disease. A better understanding of the mechanisms underlying *GLA* expression can initiate more effective methods for diagnosis and therapy of disease.

477 RACIAL DIFFERENCES IN EPIGENETIC MODIFICATIONS OF CIRCADIAN GENES AND THEIR EFFECTS ON NIGHTTIME BLOOD PRESSURE

A Daoudi*, C Sabogal, S Su. *Medical College of Georgia, Augusta, GA*

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Purpose of study To assess the racial disparities in methylation levels of circadian genes; to examine the association between methylation levels of circadian genes and nighttime blood pressure; and to investigate whether the racial differences in nighttime blood pressure can be partially explained by the racial differences in methylation levels of circadian genes.

Methods used We performed a secondary data analysis using genomic data collected from the Georgia Stress and Heart study (n=480) which consisted of a roughly equal number of European Americans (EA, n=252) and African Americans (AA, n=228). Only 299 individuals were included in the final analysis and 181 participants were omitted due missing data in at least one of the following categories: age, sex, race, BMI, supine systolic blood pressure, nighttime systolic blood pressure. We identified 159 CpG methylation sites that are associated with 8 known circadian genes. We then constructed linear models using these methylation sites to test correlations between methylation score and race, methylation score and nighttime blood pressure, and interactions between race and nighttime blood pressure. The models were controlled for age, sex, body mass index (BMI), and supine systolic blood pressure.

Summary of results When controlling for age, sex, BMI and supine systolic blood pressure, there was a significant variation

between the EA and AA populations in methylation status in 8 out of 159 CpG sites localized to 3 of the 8 circadian genes: *CRY2*, *ARNTL* and *PHLPP1* (p<0.05). The direction of change in each population is not indicated in this type of analysis. No significant correlations were detected between methylation scores and nighttime blood pressure, or race and nighttime blood pressure.

Conclusions The results of this study suggest that race accounts for some of the changes in DNA methylation scores of circadian genes, specifically *ARNTL*, *CRY2* and *PHLPP1*. Further investigating these findings and understanding the factors affecting these methylation scores may contribute to the development of innovative and personalized intervention strategies that could help reduce health problems associated with circadian disruption.

478 LINE-1 EXPRESSION AND REGULATION IN INDUCED PLURIPOTENT STEM CELLS

T Kaul*, P Deininger, M Morales. *Tulane Cancer Center, New Orleans, LA*

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Purpose of study Long interspersed element-1s (L1s) are autonomous, mobile elements that are able to copy and insert itself throughout the genome. These elements make up 17% of the human genome with over 5 00 000 copies, though the vast majority of these elements are defective and only a few dozen are potentially responsible for L1 activity. It is reported that there is increased retrotransposon activity in induced pluripotent stem cells (hiPSCs) and human embryonic stem cells (heESCs). hiPSCs hold the promise of broad application in the biomedical field including regenerative treatment. However, there is an increased risk of tumorigenesis when these reprogrammed cells are implanted. As L1 has the potential to contribute to tumor progression through insertional mutagenesis and increased genomic instability, we investigated its expression in hESCs and hiPSC.

Methods used To better understand the potential of L1-mediated mutagenesis in stem cells, it is imperative to first identify the few culprit L1s at specific loci that are actively transcribing to RNA. Using Illumina RNA-Sequencing with PacBio sequencing of full length L1 RNAs, we mapped the sequence results to the reference genome using our in-house bioinformatics pipeline. We then examined the genomic environment around the expressed L1s by determining distances from expressed genes.

Summary of results Here we characterized full-length expressed L1s in 2 human cord blood derived endothelial cell lines (hCBEC), 1 fibroblast cell line (hFF), 3 hESC lines, and 8 iPSC lines using RNA and Pacbio Sequencing. We characterized L1 expression patterns at the specific loci level in the four types of cells lines and saw an increase in L1 expression in hESC and iPSC cell lines. We also found that expressed L1s are significantly more likely to be in or near an expressed gene (p<0.05), which indirectly indicates that expressed L1s are in regions of open chromatin.

Conclusions We begin to understand at the loci-specific level where and why certain L1s are expressed in pluripotent stem cells. As there is an increase in L1 expression in induced pluripotent stem cells, there is greater potential for L1-mediated mutagenesis. This is important when considering induced pluripotent stem cells as potential therapy.

479

LOW SELF-REPORT ANTIHYPERTENSIVE MEDICATION ADHERENCE (USING KROUSEL-WOOD MEDICATION ADHERENCE SCALE) IS ASSOCIATED WITH UNCONTROLLED BLOOD PRESSURE (BP), USING ESTABLISHED ($\geq 140/90$ MMHG) AND 2017 DEFINITIONS ($\geq 130/80$ MMHG)

¹JJ Marsh*, ¹J Silver, ¹T Johnson, ²B Mohundro, ¹E Peacock, ^{1,3}M Krousel-Wood. ¹Tulane University, New Orleans, LA; ²Blue Cross and Blue Shield of Louisiana, New Orleans, LA; ³Ochsner Health System Research Division, New Orleans, LA

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Purpose of study Validated, open-access tools which measure antihypertensive medication adherence and are associated with BP among hypertensive patients are needed. The self-report 4-item Krousel-Wood Medication Adherence Scale (K-Wood-MAS-4) is open-access and associated with uncontrolled BP in adults ≥ 65 years old in managed care. We assessed the association with K-Wood-MAS-4 with uncontrolled BP using established and 2017 definitions in a broader sample of adults with respect to age and insurance status.

Methods used A cross-sectional survey of 200 community-dwelling adults ≥ 55 years old collected demographic, clinical, and adherence data. Low adherence was defined as a score ≥ 1 on the K-Wood-MAS-4. Uncontrolled BP was defined using the established definition (systolic BP (SBP) ≥ 140 mmHg or diastolic BP (DBP) ≥ 90 mmHg) and the 2017 definition by the American College of Cardiology (ACC)/American Heart Association (AHA) Hypertension Guidelines (SBP ≥ 130 mmHg or DBP ≥ 80 mmHg). Multivariate logistic regression models assessed the association between low adherence and uncontrolled BP for each definition.

Summary of results The sample was 50.0% female and 42.5% nonwhite with a mean age of 64.2 years. Uncontrolled BP prevalence was 30.4% (established definition) and 58.8% (2017 definition). Low K-Wood-MAS-4 adherence prevalence was 43.3%. Compared to adults without low adherence, the multivariable-adjusted odds ratio (OR) for uncontrolled BP associated with low adherence was 3.09 (95% CI 1.52 to 6.30) for the established definition, and 4.94 (95% CI 2.33 to 10.44) for the 2017 definition.

Conclusions Antihypertensive medication adherence measured by K-Wood-MAS-4 is associated with uncontrolled BP among community-dwelling hypertensive adults ≥ 55 years old for both established and 2017 definitions of uncontrolled BP. This validated scale provides an open-access tool for measuring medication adherence.

480

THE ASSOCIATION BETWEEN ADMISSION GLUCOSE LEVELS AND OUTCOMES IN ADULTS ADMITTED TO A TERTIARY CARE HOSPITAL

P McGrade*, S Yang, K Nugent. Texas Tech University Health Science Center, Frisco, TX

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Purpose of study Hyperglycemia at the time of hospital admission has been associated with poor outcomes in several patient groups, including patients with trauma, patients in emergency departments, and patients admitted to coronary care units. There is little information about the association of glucose levels at admission and outcomes in patients with diverse diagnoses admitted to the hospital.

Methods used We identified all adult patients (>18 years, <89 years) who have had glucose levels measured during the first 24 hours of admission to the hospital between 10/1/2015 and 9/30/2016. Clinical information included age, gender, glucose levels within 24 hours of admission, diagnoses based on ICD 10 discharge coding by the medical record department, length of stay, disposition, and mortality. Patients were classified into quartiles based on glucose levels and into clinically relevant glucose range categories (<55 mg/dL, 55–140 mg/dL, 140–200 mg/dL, and >200 mg/dL).

Summary of results This study included 18 478 adult patients admitted to University Medical Center in Lubbock, Texas. The median age was 53 years (25th percentile-33 years; 75th percentile –67 years). The median length of stay (LOS) was 4 days (25th percentile-2 days; 75th percentile-6 days). The overall mortality was 3.8%. The mortality increased in each age quartile. The median admission glucose level was 117 mg/dL (25th percentile- 97 mg/dL; 75th percentile-155 mg/dL). The mortality increased in each glucose quartile; it varied in the clinically relevant categories and was highest in patient admitted with a glucose <55 mg/dL and patients admitted with a glucose >200 mg/dL. The LOS was significantly shorter in patients in glucose quartiles 1 and 2 compared to quartiles 3 and 4. Glucose levels were not associated with mortality in patients with sepsis or COPD.

Conclusions Admission glucose levels were associated with mortality and length of stay in this large cohort of hospitalized patients. Attention to the glucose levels, especially high levels, can help clinicians focus on the underlying level of stress related to the admitting diagnosis, important comorbidities which may or may not be known prior to admission, and the potential need for glucose management strategies during hospitalization.

481

TRUST IN HEALTHCARE PROVIDERS IS ASSOCIATED WITH ANTIHYPERTENSIVE MEDICATION ADHERENCE AMONG OLDER BLACK HYPERTENSIVE ADULTS

¹J Silver*, ¹JJ Marsh, ¹T Johnson, ²B Mohundro, ¹E Peacock, ¹M Krousel-Wood. ¹Tulane University, New Orleans, LA; ²Blue Cross Blue Shield of Louisiana, Baton Rouge, AZ

10.1136/jim-2018-000974.479

Purpose of study Low adherence to antihypertensive medications remains a significant public health and clinical challenge, particularly among blacks. Provider trust has been associated with patients' medication adherence. However, data are limited on the impact of provider trust on racial disparities in medication adherence. This analysis assessed the association between patient trust in healthcare providers and low antihypertensive medication adherence among community-dwelling adults, stratified by self-reported race.

Methods used We conducted a cross-sectional analysis of survey data from 199 men and women with established hypertension, age 55 and older, recruited through a commercial health insurance partner and via community outreach in the Greater New Orleans area. An interviewer-administered survey assessed demographic, clinical, and medication adherence and attitudes data. Trust in the provider managing hypertensive care was measured by the Trust in Physician Scale, with low trust defined as scores in the lowest tertile. Low medication adherence was measured using the 4-item Krousel-Wood Medication Adherence Scale (K-Wood-MAS-4), with low adherence

defined as a score ≥ 1 . Multivariate logistic regression models assessed the association between provider trust and medication adherence; results were stratified by black and white race.

Summary of results The sample was 42.9% black and 50.5% female, with a mean age of 64 years and a prevalence of low adherence of 44.4%. Compared to adults without low trust in providers, the multivariable adjusted odds ratio and 95% confidence interval for low K-Wood-MAS-4 adherence associated with low trust in providers was 4.97 (1.61, 15.33) for blacks, and 1.10 (0.42, 2.87) for whites.

Conclusions Black, older adults with low trust in their health-care provider are 4.97 times more likely to report low antihypertensive medication adherence compared to their counterparts without low provider trust. In contrast, there was no association between low provider trust and low adherence among whites.

482 HEALTH LITERACY AMONG MINORITY AND UNDERSERVED TEENS AND PARENTS

S Sojourner*, S Looney, M Tingen. *Medical College of Georgia, Augusta University, Augusta, GA*

10.1136/jim-2018-000974.480

Purpose of study Health literacy is essential for promoting personal health and chronic disease self-management. There are disparities in health literacy across minority and underserved populations. We explored whether differences existed among teens and parents from different schools and a faith-based organization, all from minority and underserved backgrounds.

Methods used Health literacy was assessed on children and parents/guardians (Ps/Gs) of seven cohorts: 4 cohorts from School A, 1 cohort from School B, and 2 cohorts from a church. All cohorts were participating in the Strengthening Families Program (SFP), which focuses on substance abuse prevention, increasing family cohesion, resiliency, and communication. Three instruments for health literacy included: Rapid Estimate of Adolescent/Adult Literacy in Medicine (REALM), Single Item Literacy Screener (SILS) and Newest Vital Sign (NVS). To compare the test results for the cohorts, non-parametric methods were utilized including the k-sample median test with Bonferroni adjustment, Kruskal-Wallis test, Fisher's exact test, and Dwass, Steel, and Critchlow-Fligner (DSCF) pairwise comparisons.

Summary of results 109 teens and 100 parents completed health literacy measures. The seven teen cohorts differed significantly in terms of median score on the REALM ($p=0.028$) and in REALM categories ($p=0.049$), also in the median score of the NVS ($p=0.003$) and NVS categories ($p\leq 0.001$), yet did not differ significantly on median SILS response ($p=0.287$) or SILS categories ($p=0.257$). There was no significant difference among percentage of teens needing assistance with the SILS. There was no difference among the seven P/G cohorts on median score on the REALM ($p=0.068$) or in REALM categories ($p=0.212$). Ps/Gs differed significantly on median score on the NVS ($p=0.001$) and NVS categories ($p=0.002$). The P/G cohorts did not differ significantly on either median SILS response ($p=0.146$) or SILS categories ($p=0.186$). There was also no significant difference in the percentage of Ps/Gs needing assistance with the SILS.

Conclusions Differences existed among the teen and P/G cohorts despite similar backgrounds. Poor scores indicate that further efforts and research are needed to improve health literacy in this disparate population.

Pulmonary and Critical Care Medicine

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

483 EVALUATION OF THE SAFETY OF ENTERAL FEEDING IN PEDIATRIC PATIENTS WHO ARE ON VASOACTIVE INFUSIONS

¹I Justement, ²H Winters, ¹C Garbe, ¹HC Allen*. ¹Oklahoma University Health Sciences Center, Oklahoma City, OK; ²University of Oklahoma, Oklahoma City, OK

10.1136/jim-2018-000974.481

Purpose of study To determine the safety of enteral feedings in pediatric patients on vasoactive infusions.

Methods used We performed a retrospective chart review of infants and children who were on vasoactive infusions in the ICU over a 1 year period. Records were reviewed to determine what vasoactives were being given, their doses at two set times per day, and when enteral feeds were started. We collected data regarding the route of enteral feeding, formula, volume per day, percent of goal calories per kg, and any symptoms of feeding intolerance. Participants were excluded if they were on vasoactive infusions for less than 48 hours, or if they did not receive enteral feeds within 7 days of ICU admission. Primary outcomes were VIS scores and daily percent of goal calories. Secondary outcomes were frequency and reasons feeds were held, symptoms and frequency of feeding intolerance and gastrointestinal complications.

Summary of results In total, 54 patients were included. Of those, 43 (79%) were on a ventilator. Diagnoses varied with 43 (79%) cardiac, 3 pulmonary, 3 infectious, 2 neurological, 1 post-operative, 1 trauma, and 1 hematologic-oncologic. The median hospital LOS for all patients was 19 (range 4, 333) days with the median number of PICU days was 11.5 (range 1, 27) and the median number of ventilator days was 8 (range 1, 252). There was no difference in the median VIS score for both time points on day and day 3. Participants achieved a higher percentage of goal caloric intake on day 3 (65.85%) of enteral feeds compared to day 1 (14.5%). The majority of patients (>60%) had no feeding intolerances on any day. For patients with feeding intolerance, constipation, diarrhea, and emesis were the most common. A single patient was noted to have NEC, this patient had a shunt dependant cardiac lesion, with a max VIS score of 5.

Conclusions The majority of our patients tolerated their enteral nutrition well and did not experience symptoms of feeding intolerance. This small retrospective study suggests that enteral feeds while on low dose vasoactive infusions are safe and should be considered by physicians.

484 NEVER LET CT SCAN FINDING FOOL YOU!

MA Alsharif*, M Abdou, T Vo. *Texas Tech University Health Science Center, Amarillo, TX*

10.1136/jim-2018-000974.482

Background Necrotizing soft tissue infection is a life-threatening disease that is often difficult to diagnose. The likelihood of necrotizing fasciitis cannot be predicted reliably using laboratory parameters or any imaging. Here we present a mortality case of worsening cellulitis which did not receive timely surgical exploration despite a worsening clinical picture.

Description A 61-year-old female with history of bilateral osteoarthritis and morbid obesity presented with fever and worsening left leg cellulitis for two weeks which was not improved with oral antibiotics. She was hypotensive, tachypneic and tachycardic. On exam, she had left lower extremity circumferential erythema and edema up to mid-leg. She also had tenderness with warmth, normal peripheral pulses, and no cyanosis. Chest x-ray was unremarkable. Lower extremity venous duplex was negative for DVT. Urinalysis was positive for red blood cells, blood, and granulocyte casts. Her labs were significant for acute kidney injury with creatinine of 1.9, BUN of 27, CPK of 6880 and procalcitonin of 17. She was admitted to intensive care for septic shock secondary to cellulitis. Despite appropriate IV antibiotics, her condition worsened over the next two days and the cellulitis continued to extend proximally with purulence. Kidney function continued to decline. Necrotizing fasciitis was suspected. Surgery was consulted and computed tomography without contrast of the leg was ordered, which revealed edematous changes within the subcutaneous tissues without any soft tissue abscess or abnormal air collection identified. Surgery recommended to continue medical management with IV antibiotics. Blood cultures were negative and wound culture grew group G beta-hemolytic *Streptococcus*. Two days later, she was intubated due to worsening septic shock and respiratory distress. Eventually the patient's husband decided on palliative measures due to her worsening condition and poor outcome.

Discussion Our case demonstrates a vital teaching point – that early diagnosis of necrotizing fasciitis may be life saving. Patients require aggressive treatment to combat the associated high morbidity and mortality. Standard teaching rejects CT scanning as a reliable method of diagnosing necrotizing soft tissue infections. Surgical exploration is recommended as the diagnostic standard of reference.

485 A SLEEPING SUBDURAL HEMORRHAGE: HICKAM'S DICTUM VERSUS OCCAM'S RAZOR IN THE INTENSIVE CARE UNIT

CM Baldeo*, K Seegobin, L Jones. *UF College of Medicine, Jacksonville, FL*

10.1136/jim-2018-000974.483

Case report A 57-year-old male with DM, HTN presented for encephalopathy. Unresponsive to naran. Glucose was critically high. CT head was negative for any intracranial abnormalities. ECG was concerning for STEMI. Troponin was 0.19. Cardiology called off STEMI alert. Enoxaparin started for NSTEMI. Intubated for GCS of 3. Insulin drip for DKA started. Became bradycardic into 30 s overnight. ECG showed sinus bradycardia. Transcutaneous pacing initiated. Agitated with observed patient-ventilator dyssynchrony and hypertensive with SBPs

170–190 s. Multiple doses of midazolam given with improvement in agitation. Hypotensive the next day, MAPs in 50–60 s. Blood pressure improved with norepinephrine drip. Throughout day, remained unresponsive off all sedation, attributed to sedation given overnight. Repeat neurological exam showed GCS 3, loss of pupillary/gag reflexes, negative of Doll's eyes. Repeat CT head with diffuse cerebral edema, new large right subdural hemorrhage (SDH), right-to-left shift, significant mass effect with descending transtentorial and cerebellar tonsillar herniation. Family opted for comfort. Patient compassionately weaned off ventilator, death pronounced soon after extubation. Clinical diagnosis of SDH after initiating anticoagulation therapy was missed in this case. Features suggesting SDH with increased intracranial pressure and brain herniation include respiratory depression, hypertension and bradycardia (Cushing's triad). Agitation was likely in response to this. Unfortunately neurological sequelae was masked by sedation received for agitation the night prior. As with all anticoagulants bleeding is major adverse effect of enoxaparin. Hemorrhage may occur at any site with risk dependent on multiple variables. There is up to 0.8% risk of intracranial hemorrhage. Unclear if he suffered head injury prior to hospitalization that may have precipitated his bleed. Normal CTH on admission argues against it. Case highlights need for frequent neurological exam of ICU patients particularly with changes in mentation or clinical status after starting anticoagulation. Sustained intracranial hypertension and acute brain herniation signify catastrophic neurological events requiring immediate recognition and treatment to prevent irreversible injury and death.

486 USE OF DIURETIC FOR BPD INCREASES THE RISK OF METABOLIC BONE DISEASE IN PRETERM INFANTS

S Bhatia*, M Abraham, B Niebuhr, S Jain. *University of Texas Medical Branch, Galveston, TX*

10.1136/jim-2018-000974.484

Purpose of study Increased survival of preterm infants has led to increased incidence of bronchopulmonary dysplasia (BPD). Diuretics are routinely used in the management of BPD. Use of diuretics in the management of BPD is controversial. Preterm infants are at increased risk of developing metabolic bone disease (MBD). MBD is multifactorial; there is insufficient data to assess the effect of diuretics on MBD in preterm infants. The aim of the study was to assess association of diuretics with MBD.

Methods used After IRB approval, in a retrospective study, we included all infants born at our institute between January 2017 to August 2018 (20 months) who were ≤ 30 weeks gestation and birth weight ≤ 1500 grams. All infants were divided into Diuretics and control group. Diuretic group infants received diuretics for BPD. Control group did not receive diuretics. BPD was defined as infants requiring respiratory support at 36 weeks post menstrual age and diuretics use was hydrochlorothiazide and spironolactone for at least 7 days. All infants were assessed for development of MBD. MBD was defined as parathormone (PTH) > 100 pg/mL. We also assessed if diuretics helped to improve the respiratory status. Data was analyzed using SPSS, Chi Square Test and T test and $p < 0.05$ were considered significant.

Summary of results 154 infants were included in the study. Mean gestational age was 27.6 ± 2.7 weeks; mean birth weight

999±312 grams. There were 53 (35%) in BPD +Diuretic group and 101 infants (65%) in control group. 37 V/S 21% ($p<0.05$) infants in BPD +Diuretics developed MBD. 87% V/S 13% ($p<0.001$) infants with BPD were treated with diuretics. 55% V/S 45% ($p<0.05$) infants with BPD develop MBD. When compared the respiratory response to diuretics, diuretics did not improve the respiratory status. At the same time, we found infants given diuretics remained on ventilation for longer ($p=0.002$).

Conclusions Infants treated with diuretics for BPD are more likely to develop MBD without any significant improvement in respiratory status. Diuretics should be used with caution and monitor closely for MBD. Further studies may be useful to set standardized guidelines for diuretic use.

487 HEMODIALYSIS INITIATED DURING ACLS FOR TREATMENT OF CRITICAL HYPERKALEMIA

D Campbell*. UAB, Birmingham, AL

10.1136/jim-2018-000974.485

Case report Hyperkalemia is a common abnormality found in the ICU for a variety of reasons. Hemodialysis is the most definitive therapy for hyperkalemia, but often not performed in a hemodynamically unstable patient. We would like to present the case of a patient who went into cardiac arrest due to hyperkalemia and was successfully treated with iHD started during chest compressions.

Mr. C was a 25 y/o male admitted to the MICU for ARDS after massive aspiration during status epilepticus. Post-seizure rhabdomyolysis failed to improve with IV fluids and he progressed to renal failure. He was started on continuous renal replacement therapy (CRRT), but potassium rapidly escalated from 4.5 mEq/L to 6.9 mEq/L despite aggressive medical management. He became progressively bradycardic with widened QRS and went into cardiac arrest with ACLS initiated. ROSC was achieved multiple times, but always with deterioration and pulselessness within minutes. Potassium remained >7 mEq/L on POC testing during the codes. Nephrology was readily available and hemodialysis was started within 20 min. He was placed on the circuit during a period of relative stability, but again lost pulse during the startup cycle for the dialysis machine. Dialysis was actually initiated during a round of chest compressions. He had ROSC and one further episode of pulselessness with ROSC while on iHD. After about 30 min on dialysis, his potassium improved and he had no further episodes of pulselessness. Despite his improved hemodynamics, his acidosis and hypoxemia worsened and he was emergently placed on VV ECMO. Unfortunately, despite some improvement, he had episodes of VT requiring defibrillation while on ECMO and the family decided to withdraw care.

Even though this patient did not survive his critical illness, we were able to successfully treat his hyperkalemia and keep him stable long enough to be cannulated for ECMO. While medical management and CRRT will remain the mainstays of therapy for hyperkalemia in the critically ill, hemodynamically unstable patient, it is important to remember that hemodialysis is much faster in clearing potassium. Hemodialysis should be considered in the critically hyperkalemic patient, even if the patient is hemodynamically unstable, especially if increased support with additional vasopressors is possible.

488 PREVALENCE OF GLAUCOMA AMONG PATIENTS WITH OBSTRUCTIVE SLEEP APNEA: A CROSS-SECTIONAL STUDY

¹HR Cintrón-Colón*, ¹WD Marrero, ¹V Fonseca-Ferrer, ¹C Rivera-Franceschini, ²AC Doval-López. ¹San Juan City Hospital, San Juan; ²University of Puerto Rico, San Juan

10.1136/jim-2018-000974.486

Purpose of study To examine the association between OSA and Glaucoma in a population representative of the United States.

Methods used This cross-sectional study included 6760 participants in the 2005–2008 National Health and Nutrition Examination Survey (NHANES), age ≥ 40 years, who reported a presence or absence of glaucoma. Participants were interviewed regarding their sleep patterns and whether they have been previously diagnosed with sleep apnea. Information regarding the presence or absence of glaucoma, as well as demographics, comorbidities, and health-related behaviors were obtained via interview. Multivariate logistic regression models were created and refined by adding confounding variables. These models were used to examine the possible independent association of OSA with self-reported glaucoma.

Summary of results Of the 6760 participants, 620 reported to have a previous diagnosis of a sleep disorder and 387 reported a diagnosis of sleep apnea. The diagnosis of glaucoma was self-reported in 46 patients with sleep disorders and 30 with OSA. Self-reported glaucoma was compared between participants with and without a previous diagnosis of OSA while adjusting for demographics, health-related behaviors, and comorbidities (OR 1.38, 95% CI: 0.51 to 3.70).

Conclusions Contrary to popular belief, our study reiterates the lack of association between OSA and Glaucoma.

489 HIV-1 TRANSGENE EXPRESSION INDUCES PROSTANOID SIGNALING DYSFUNCTION IN RAT ALVEOLAR MACROPHAGES

¹C Cook*, ^{1,2}SK Cribbs, ¹M Ahmed, ¹X Fan, ^{1,2}D Guidot, ¹BS Staitieh. ¹Emory University, Atlanta, GA; ²Atlanta VA, Decatur, GA

10.1136/jim-2018-000974.487

Purpose of study Despite the advent of anti-retroviral therapy, people living with HIV (PLWH) remain at a higher risk of bacterial pneumonia. We have previously determined that the increased risk of pneumonia is due, at least in part, to the effects of HIV-related viral proteins on alveolar macrophage (AM) innate immune function. Using an HIV-1 transgenic rat model to simulate the conditions in the lungs of PLWH, we previously characterized alterations in prostanoid receptor expression and impaired phagocytic function in HIV-1 rat AMs. The latter defect was reversed by the addition of an EP4 receptor agonist. To extend our prior work, we designed a series of experiments to determine the effects of HIV-related viral proteins on the cellular response to prostaglandin E2 (PGE2), the endogenous EP receptor ligand.

Methods used AMs obtained by whole lung lavage from HIV transgenic rats and their wild-type littermates were treated with PGE2 for twenty-four hours prior to assessment of inducible nitric oxide synthase (iNOS) and CD14 levels by qRT-PCR. In parallel, a rat alveolar macrophage cell line (NR8383) was also treated with either PGE2 or EP4 receptor agonist for 24 hours prior to assessment of EP2 and EP4 receptors by qRT-PCR.

Summary of results Treatment of WT rat AMs with PGE2 increased iNOS and CD14. The response of HIV transgenic AMs to PGE2 was markedly more robust in both cases, though. Treatment of NR8383 cells with PGE2 decreased both EP2 and EP4 receptor gene expression in a dose-dependent fashion, and treatment with an EP4 agonist significantly increased EP4 receptor gene expression.

Conclusions HIV dramatically amplifies the inflammatory response induced by prostanoid signaling. As key innate immune effectors, the aberrant response of iNOS and CD14 to PGE2 in the setting of chronic viral protein exposure suggests a markedly dysregulated immune response in HIV. Interestingly, PGE2 seems to induce a negative feedback loop on EP receptors, causing their expression to decrease as its levels increase. Treatment with a specific EP4 agonist, however, appears to promote EP4 receptor expression. Modulating EP receptor expression may be a fruitful avenue for improving innate immunity in HIV.

490 SMOKING-RELATED INTERSTITIAL LUNG DISEASE IN A PATIENT WITH CONNECTIVE TISSUE DISEASE

J Crane*, D Sotello, K Nugent. *Texas Tech University Health Science Center, Lubbock, TX*

10.1136/jim-2018-000974.488

Case report A 44-year-old female with a 6 month history of worsening dyspnea on exertion, nonproductive cough, and newly developed hypoxemia was referred for further evaluation. She had a medical history of fibromyalgia, obesity, and OSA, and was an active cigarette smoker with a 30 pack-year history. She reported morning stiffness in both hands, fatigue and malaise. Room air oxygen saturation of 90%, mild clubbing of both hands, and telangiectasias of the face and chest without joint deformity. Labs were significant for a chronically elevated neutrophilic leukocytosis and thrombocytosis, ESR of 77, CRP of 34, and an ANA titer of 1:1280. Chest x-ray suggested faint interstitial opacities in the lower lung fields. HRCT chest revealed diffuse bilateral ground glass opacities throughout. A 6MWT showed desaturation to 83% after 360 meters. PFTs revealed FVC 2.4 L (63% pred), FEV1 1.8 L (57% pred), FEV1/FVC 0.75, normal TLC, RV 3.6 L (181% pred), DLco (36% pred), consistent with a mixed obstructive-restrictive defect. She was placed on continuous oxygen supplementation of 2 L/min. She had signs of a connective tissue disease. She underwent a wedge resection of the left lower lobe. Histology revealed regions of alveoli filled with macrophages consistent with DIP, and intraluminal macrophages in the respiratory bronchioles and peribronchiolar fibrosis consistent with RB-ILD. She was diagnosed with smoking related ILD. The primary

Abstract 490 Figure 1

Abstract 490 Figure 2

intervention was smoking cessation rather than immunosuppression. Obtaining a tissue diagnosis is important in cases of ILD of unclear etiology.

491 A RARE CASE OF LANE-HAMILTON SYNDROME

R Dean*, R Subedi. *Upstate Medical University, Liverpool, NY*

10.1136/jim-2018-000974.489

Case report Idiopathic pulmonary hemosiderosis (IPH) associated with celiac disease, known as Lane-Hamilton Syndrome (LHS) is a rare phenomenon. This disease process requires prompt recognition as treatment generally includes a gluten free diet and possibly immunosuppression. Here, we present a case of LHS in a 19 year old male.

A 19 year old male, with no past medical history, presented with dyspnea on exertion and fatigue that had worsened over the previous month in addition to a chronic cough with intermittent hemoptysis of 1 year. His review of systems was otherwise negative. He was found to be anemic, with a hemoglobin of 4.1 g/dL. He was transfused 4 units of packed red blood cells with an appropriate response. A CT with contrast of the thorax showed diffuse bilateral ground-glass opacities consistent with pulmonary hemorrhage. A bronchoscopy with biopsy and lavage showed normal anatomy and bronchial mucosa and the presence of old blood and with progressively bloody return. Infectious workup as well as cytology was negative. An autoimmune workup was negative. Serology for celiac disease was positive, with an elevated tissue transglutaminase of IgA 100 U/mL and deamidated gliadin antibodies elevated at 84 U/mL. He remained clinically stable and did not require further transfusion during his admission. He was discharged on a gluten free diet and with endoscopic follow up to confirm celiac disease.

IPH and CD are believed to be related through an immunologic mediated pathway, though the exact mechanism is not clear. Hypotheses include that gluten stimulates an increased T cell mediated response, forming circulating immune complexes which are stored in the alveolar basement membrane or complexes form in the lungs when food allergens are inhaled.

Many cases present with respiratory complaints or anemia rather than gastrointestinal symptoms related to CD. Despite this, experts recommend performing serologic testing in all those diagnosed with IPH. Some are even proponents of performing duodenal biopsy, the gold standard in diagnosis of CD, in all those diagnosed with IPH.

Treatment of LHS is a gluten free diet, which in many cases leads to a partial or complete regression of the disease process. Gluten avoidance significantly reduces the need for immunosuppressive therapy, which is frequently used in IPH.

492

CONGENITAL DIAPHRAGMATIC HERNIA (CDH): MORTALITY TRENDS AND FACTORS ASSOCIATED WITH SURGICAL REPAIR

J Desai*, AJ Bhatt, M Famuyide, R Savich. *University of Mississippi Medical Center, Jackson, MS*

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Purpose of study Aims (i) to describe mortality trends associated with surgical repair and extracorporeal membrane oxygenation (ECMO) in patients with CDH (ii) to examine the demographic factors associated with surgical repair.

Methods used National multicenter Kids' Inpatient Database (KID) was used; admissions at age ≤ 30 days, term gestational age at birth with diagnosis of CDH were selected with ICD-9 codes (756.6, 551.3, 552.3 and 553.3) during years of 2003, 2006, 2009 and 2012. Analysis was performed using SAS Enterprise Guide 7.1.

Summary of results Among 3879 term infants mortalities showed significant decrease from 2009 to 2012 despite of same rates of incidence and surgical repair but increased rates of ECMO. Mortality was higher if repair was performed after 7 days of life (35% vs 9%, $p < 0.0001$); this relationship remained significant for infants who did not required ECMO at all (12% vs 1.7%; OR: 8.3 (3.7, 18.8), $p < 0.0001$). Mortality among infants who went for ECMO before surgery was not different from who went for surgery before ECMO (54% vs 60%, $p = 0.44$). Time of surgical repair was significantly early in survivors (Median, IQR: 3, 1–4 vs 8, 3–13, $p < 0.0001$) while timing of ECMO was not different. Presence of other anomalies including critical congenital heart defects (CHD), hypoxic-ischemic encephalopathy (HIE), African American race, public insurance, Midwest or south geographic location of hospital (vs west coast) and small hospital size were significantly associated with lower odds of having surgical repair.

Abstract 492 Figure 1

Conclusions To our knowledge this is the largest cohort based on national multi center database describing mortality trends and demographic factors associated with surgical repair of CDH. Significant decrease in mortalities from year 2000 to 2012 despite of increasing ECMO rates indicating improved management strategies.

493

THE ROUTINE USE OF POST-OPERATIVE NSAIDS IN THE PEDIATRIC PATIENT IS EFFECTIVE AND SAFE

E Gaid*, J Waller, R Mehta. *Medical College of Georgia at Augusta University, Augusta, GA*

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Purpose of study Effective and appropriate post-operative pain management in the pediatric population is important for a safe and comfortable recovery process. Opioids, non-steroidal anti-inflammatory drugs (NSAIDs), and acetaminophen remain the current pharmacologic therapy for post-operative pain management, with an increasing preference for the use of scheduled NSAIDs to avoid opioid-related side effects. The objective of this study is to examine the renal and bleeding complications of scheduled post-operative NSAID use in the pediatric population (ages 0–18). We hypothesize that pediatric patients started on a scheduled regimen of NSAIDs immediately post-operatively are at an equal risk of bleeding and renal complications for the first 5 days post-op compared to those not receiving the medications.

Methods used A retrospective chart review of patients admitted to the pediatric intensive care unit from July 2015-May 2018 status post congenital heart defect or other surgery requiring pain control was conducted. Renal effects were evaluated by serum creatinine level, while significant bleeding events were assessed by clinically documented intracranial hemorrhage, GI consult or significant upper GI bleed, or bleeding requiring additional surgical exploration or transfusion.

Summary of results Analyses were conducted using descriptive statistics to evaluate the significance of abnormal creatinine values by age. Due to a low frequency of abnormal creatinine levels, the percent of days with abnormal creatinine was used to assess for differences between groups. There was no overall statistically significant difference in creatinine levels among cardiac surgery and NSAID groups over time. There was no significant association between patients with scheduled NSAIDs and any bleeding event in cardiac or non-cardiac surgeries.

Conclusions There is no association with increased bleeding or adverse renal effects following the use of NSAIDs in the pediatric post-operative setting in both cardiac and non-cardiac surgeries. A randomized prospective study is needed to confirm the safety and efficacy of NSAIDs in the post-operative pediatric population.

494

SPONTANEOUS HEMOTHORAX: LATE MANIFESTATION OF CAVITARY LESION BY ASPERGILLUS

JM Garcia Puebla*, K Hernandez Moya. *San Juan City Hospital, Guaynabo*

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Case report Spontaneous hemothorax is an accumulation of more than 400 mL of blood within the pleural space in absence of trauma with a median presentation of 22–34 y/o. Symptoms of hemothorax include chest pain and dyspnea. If there is 1,000 mL or more of blood in the pleural space it can present with restlessness, pallor, pleuritic pain, hemoptysis, and shortness of breath. If there is hemodynamic instability and shock, prognosis worsens and it can be life threatening. Currently, there is not widely available literature of spontaneous hemothorax when unrelated to trauma.

The case is a 33 y/o male patient with history of pulmonary aspergillosis infection 5 years ago, treated for 1 month with unknown medications. He presented to the emergency room with complaint of dry cough, chest pain, shortness of breath, which progressed to weakness, dizziness and hemoptysis of approximately 10 mL of blood. Physical exam was remarkable for decreased breath sounds on left hemi thorax.

Chest X-ray showed left sided apical pneumothorax with associated pleural effusion and right mediastinal shifting, along with a cavitary lesion. Chest CT scan confirmed the cavitation measuring 10.8 cm × 6.9 cm × 6.1 cm, along with a left sided hydropneumothorax, and presence of a complex hemorrhagic left-sided hemopneumothorax collapsing the left lung. Labwork with WBCs in 25 K with left shifting, stable hemoglobin and platelet levels, hyperglycemia, creatinine level of 1.49, lactic acid of 43.7 mg/dL, and normal coagulation profile. Upon evaluation, the patient became hemodynamically unstable, with decrease in Hgb to 9 mg/dL, for which left sided chest tube was placed that drained 1 400 mL of bright red blood. Intravenous antifungals and antibiotics were started and a left lower lobe lobectomy was performed with complete resolution of symptoms.

Spontaneous hemothorax is uncommon in absence of trauma, and this type of clinical event is critical and life threatening. Most importantly, spontaneous hemopneumothorax cases are not common and have not been previously reported in Puerto Rico nor the United States, much less likely related to previous infections like Aspergillosis as in this case. Awareness should be encouraged among physicians regarding clinical findings, life threatening complications, and available management for this manifestation.

495

EGGSHELL CALCIFICATION ON X-RAY

WC Green*, G Berdine. *Texas Tech University Health Sciences Center, Lubbock, TX*

10.1136/jim-2018-000974.493

Case report Agent Orange contained a human toxin known to cause many health problems and pro-inflammatory effects. Silica is also known to cause respiratory disease and lymph node calcification. We present a case of a 74-year-old man who presented with dyspnea and a cough and subsequently found eggshell calcification on chest x-ray clearer than many textbooks. His history was significant for Agent Orange exposure during the Vietnam War along with a later occupational exposure to silica. Such clear eggshell calcification could be

Abstract 495 Figure 1

Abstract 495 Figure 2

due to exposure to both Agent Orange coupled with silica. The case highlights the importance of prior films for comparison and must consider Agent Orange as a possible etiology of the eggshell calcification.

496

SEVERE CALCIUM CHANNEL BLOCKER INTOXICATION IN A PEDIATRIC PATIENT

V Habet*, N Godiwala. *Louisiana State University Health Sciences Center, New Orleans, LA*

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Case report Calcium channel blocker intoxication is uncommon in children but is associated with high mortality. Extended release forms increase morbidity and mortality of overdoses as they are thought to decrease gastric motility, thereby increasing drug absorption and causing latency of symptoms. We discuss a case of severe verapamil overdose resulting in a prolonged hospital course.

A 14 year old female with major depressive disorder and prior suicidality presented after intentional ingestion of 15–20 extended release 240 mg verapamil tablets. She developed multiple complications, including prolonged hypotension requiring several vasopressor infusions, bradycardic arrest, atrioventricular block, acute respiratory distress syndrome, pulmonary edema, hyperglycemia, and lactic acidosis. Initial ECG revealed widened QRS complex and prolonged QT interval. Interventions included IV calcium gluconate, lipid emulsion therapy, and both high-dose insulin and glucagon infusions. Due to persistent bradycardic dysrhythmias, she was transitioned to an isoproterenol infusion. Repeat ECG showed a first degree atrioventricular block. Echocardiogram revealed a right atrial thrombus or vegetation. After cultures confirmed a central catheter infection, she was treated for possible endocarditis, and heparin was initiated for possible thrombus. Isoproterenol was weaned after 2 days, and her atrioventricular block resolved on successive ECGs. She was extubated after 9 days of mechanical ventilation and completed 4 weeks of ceftriaxone. Repeat echocardiogram showed resolution of the atrial mass.

Intoxication with extended release calcium channel blockers can cause delayed onset of dysrhythmias, shock, and sudden cardiovascular collapse. Treatment is often complicated and multimodal. Standard therapies in adults include IV calcium, high-dose insulin, lipid emulsion therapy, glucagon, and vasopressor support. Veno-arterial extracorporeal membrane oxygenation or cardiac pacing are options in refractory cases. There is a paucity of literature available on the treatment of calcium channel blocker toxicity in children. Further studies are needed to assess if it is reasonable to apply adult management guidelines to the pediatric population.

497 FEVER ASSOCIATED WITH ABNORMAL CHEST RADIOGRAPHY IN PEDIATRIC PATIENTS PRESENTING TO THE EMERGENCY DEPARTMENT FOR ASTHMA EXACERBATION

S Shipman, B Hyatt*. *Integris Southwest Medical Center, Edmond, OK*

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Purpose of study Acute asthma exacerbations account for many Emergency Department (ED) visits in the pediatric population. Chest radiography (CXR) is occasionally performed in these patients to identify practice changing findings such as pneumonia. There is limited data regarding which clinical and historical factors in these patients may warrant the cost and radiation exposure of CXR. The purpose of this study is to determine if there is a correlation between historical factors and clinical findings and an abnormal CXR in pediatric patients presenting to the ED for asthma exacerbation.

Methods used This was a prospective observational non-interventional cohort study. Sequential pediatric patients (age 0 to 18 years) with a history of asthma or previous wheezing presenting to the ED with acute wheezing/dyspnea and whom the treating physician ordered a CXR were enrolled. Multiple logistic regression models examined univariate associations between patient characteristics and historical factors and abnormal CXR.

Summary of results 54 cases were enrolled (age range 3 months to 18 years). The clinical findings tested included accessory muscle use or crackles on exam, room air hypoxia, age adjusted tachypnea or tachycardia, concurrent upper respiratory symptoms, and fever (temperature $\geq 38^{\circ}\text{C}$). Additionally, the patient's current need for admission and historical factors to include: steroid use in the last 60 days, prior admission for asthma, prior ICU admission for asthma, and if patient was taking asthma control medication were tested. Only a fever was noted to reach statistical significance under linear regression with an abnormal chest x-ray ($p=0.020$). Calculation of test characteristics for fever and abnormal chest X-ray demonstrate Sensitivity 0.31, Specificity 0.81, PPV 0.63 and NPV 0.68.

Conclusions In pediatric patients presenting to the ED for asthma exacerbation, most clinical findings and historical factors are not associated with an abnormal CXR. This study demonstrates a level of correlation between an abnormal chest x-ray and fever, but a larger study is needed to examine the potential significance of fever in this patient population.

498 MIDODRINE USE IN INTENSIVE CARE UNIT PATIENTS RECOVERING FROM SEPTIC SHOCK

¹K Iwuji*, ²B Dziuk. ¹Texas Tech University Health Science Center School of Medicine, Lubbock, TX; ²Texas Tech University Health Sciences Center, Lubbock, TX

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Purpose of study With rising cost of ICU care and vasopressor associated complications, clinicians need oral medications that can hasten recovery from septic shock in patients requiring low dose IV vasopressor agents.

Methods used We retrieved all the available studies published on PubMed and Embase (until September 2018) that reported the use of midodrine in septic shock.

Summary of results Levine, et al. reported a decrease in intravenous vasopressor rate after administration of midodrine from $-0.62+1.40$ mcg/min per hour before midodrine to $-2.20+2.45$ mcg/min per hour during the first four doses of midodrine ($p=0.012$). Whitson, et al. reported a two-day decrease in mean ICU length of stay when midodrine is used with IV vasopressors ($p=0.017$).

Conclusions Midodrine may have the potential to hasten the vasopressor weaning process in patients recovering from septic shock, but there are a limited published data available to support its use in this setting.

499 LARGE SECONDARY SPONTANEOUS PNEUMOTHORAX

S Jadeja*, J Verdecia, CM Baldeo. *UF Health Jacksonville, Jacksonville, FL*

10.1136/jim-2018-000974.497

Case report A 62 year old male presented with sudden onset chest pain and shortness of breath. Chest X-ray showed a large right-sided pneumothorax. A computed tomography scan of the chest showed underlying emphysematous bullae. Cardiothoracic surgery was consulted and the patient underwent

Abstract 499 Figure 1 Large right-sided pneumothorax

Video Assisted Thoracoscopic surgery with pleurodesis and resection of bleb. Repeat chest x-ray showed resolution of the pneumothorax and the chest tube was removed after 72 hours.

Spontaneous pneumothorax is classified into primary and secondary. Primary spontaneous pneumothorax usually affects healthy, young people without lung disease. Secondary pneumothorax is most commonly seen in older individuals that have underlying lung disease. Secondary spontaneous pneumothorax can be life-threatening due to already compromised lungs, and therefore, requires immediate intervention in the hospital. The British Thoracic Society and the American College of Chest Physicians outline the management of pneumothorax. British Thoracic Society looks at the distance between the chest wall at the hilar level and the lung surface and American College of Chest Physicians uses the distance from the chest wall to the lung apices. If the distance is greater than 2 cm or 3 cm respectively, a chest tube should be placed. All individuals with secondary spontaneous pneumothorax should have pleurodesis on their first occurrence. Our patient was treated as per guidelines and returned to baseline a few days after admission.

500 CLOSTRIDIUM DIFFICILE COLONIZATION IN A PEDIATRIC INTENSIVE CARE UNIT

¹RM Keilman*, ¹M Rowin, ²J Holcombe, ²H Spratt. ¹University of Tennessee College of Medicine, Chattanooga, TN; ²University of Tennessee Chattanooga, Chattanooga, TN

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Purpose of study The hospital environment is an increasingly recognized source of hospital-acquired infections, especially in locations of high antibiotic use such as the pediatric ICU (PICU). Our previous study demonstrated significant bacterial colonization in the PICU by *Staphylococcus* and enterobacter, primarily on in-room air filters. Our current study investigates the prevalence of *Clostridia difficile* in the PICU environment.

Methods used Six locations in a PICU room were studied including the sink, multiple floor sites, the patient bed and the air duct. Samples were collected weekly in triplicate with sterile swabs and inoculated in Hardy Diagnostics *C. difficile* Banana Broth within 4 hours of collection. A total of 66 samples were analyzed over a 6 month period. *C. difficile* detection was determined at 48 and 120 hours of incubation by colorimetric analysis with degree of growth determined through densitometry.

Summary of results In the PICU, the patient's sink was most heavily contaminated with 40.9% of our samples detecting *C. difficile*. Significant contamination was also noted on the floor 2 meters (36.4%) and 4 meters (31.8%) from the sink. Minimal colonization was noted on the patient's bedrail (1.5%). In our earlier study, we noted the air ducts heavily contaminated with *Staph aureus* (98%). However, in this study we found only 23.1% contamination with *C. difficile*. Additionally, our study noted routine cleaning with detergents did not affect the degree of *C. difficile* contamination in any of our sampled locations.

Conclusions *C. difficile* is a known human pathogen. Historically, the reservoir of *Clostridia* was presumed to be infected individuals. Most hospital decontamination efforts therefore focus on hospital staff-directed measures. Little is known on the location and effects of *C. difficile* environmental

colonization. This study is the first to demonstrate that the PICU sink, and the floor around the sink up to 4 meters, act as potential reservoirs for *Clostridium difficile*. Our study suggests focused decontamination procedures in these areas may decrease the potential spread of hospital-acquired *C. difficile*.

501 MASSIVE HEMOPTYSIS OF AN INCARCERATED PATIENT

DT Lake*, S Werner, K Ellard, LS Engel, E Dauchy. LSU Health Sciences Center, New Orleans, LA

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Introduction Hemoptysis is caused by bleeding anywhere in the respiratory tract from trachea to alveoli and is considered massive if volume is >300 mL. The underlying process leading to hemoptysis may be traumatic (foreign body aspiration), infectious (bronchitis, pneumonia, tuberculosis), cardiovascular (pulmonary embolism, volume overload, arteriovenous malformation), hematological (supratherapeutic anticoagulation), oncological (lung cancer), or rheumatological (lupus, sarcoidosis).

Case A 54-year-old incarcerated man with hypertension, chronic kidney disease, lupus nephritis, and history of pulmonary embolism on warfarin presented to the emergency department with dyspnea on exertion for one month. His symptoms had a gradual onset and were accompanied by generalized malaise and fatigue. Two weeks prior to arrival, his urine output decreased, prompting an empiric course of prednisone for concerns of lupus flare. One week prior to arrival, he noticed bilateral lower extremity swelling, intermittent melena, and hemoptysis. At presentation, vitals were stable and physical exam revealed active hemoptysis, rales auscultated at the lower lung fields, and pitting edema above the knees. Notable laboratory studies included hemoglobin 5.8 g/dL, serum potassium 4.1 mEq/L, BUN 101 mg/dL, serum creatinine 15.9 mg/dL, BNP 484 ng/L, and INR 8.8. Bedside ultrasound revealed grossly normal ejection fraction, and chest X-ray showed increased opacification diffusely. Testing for tuberculosis was unremarkable. His symptoms resolved gradually over several days after initiating treatment with oral vitamin K and hemodialysis.

Discussion A complex history in this patient led to a broad differential diagnosis of the etiology of hemoptysis. Pulmonary embolism was unlikely given the supratherapeutic INR. Tuberculosis was more seriously considered given a long history of incarceration; negative PPD and serial AFB's made tuberculosis less likely. Diffuse alveolar hemorrhage is a possible sequela of lupus; normal serum complement levels made lupus flare less likely. Supratherapeutic INR combined with new onset end-stage renal disease leading to volume overload most likely explained hemoptysis in this patient.

502 CORTICOSTEROID FOR TREATMENT OF HANTAVIRUS PULMONARY SYNDROME

P Laoveeravat*, W Vutthikraivit, K Nugent. Texas Tech University Health Sciences Center, Lubbock, TX

10.1136/jim-2018-000974.500

Case report Hantavirus pulmonary syndrome is a rare respiratory disease and has a high mortality rate. Early diagnosis and

supportive treatment in intensive care unit during severe respiratory distress show to provide better prognosis in patients with HPS. Role of corticosteroids is still controversial.

Case description A 62-year-old male with no significant past medical history presented to the emergency room because of acute worsening shortness of breath. He reported having runny nose, nonproductive cough, and progressive shortness of breath for 4 days. He was desaturated and rapidly progress to acute respiratory failure. Intubation was done, and he was admitted to intensive care unit. Chest x-ray showed diffuse bilateral coalescent opacities in both lungs. The diagnosis was made as acute respiratory distress syndrome with septic shock. He was initially treated with IV fluid, vasopressor, and empirical antibiotics. Labs finding showed leukocytosis, thrombocytopenia and erythrocytosis. Electrolyte and kidney function labs showed acute kidney injury with hypovolemic hyponatremia. After supportive treatment with low-tidal volume ventilator along with vasopressor for hemodynamic support, his clinical status on the night of admission was stable. High-dose methylprednisolone was started on day 3 of admission. Methylprednisolone had been given for 3 days, then tapered down. His chest x-ray revealed a significant decrease in diffuse alveolar damage on day 5 of the admission. Regarding of his improvement, he was extubated and transferred to the floor for continuity care on day 5. He was discharged after 13 days of admission without any complications. Serologic tests for bacterial infection, including Mycoplasma and Legionella came back negative. Histoplasma galactomannan antibody and Beta-D glucan were negative. Hypersensitivity pneumonitis panel was negative. Serologic test for Hantavirus IgM was done on day 4 which came back positive after the patient was discharged.

Discussion In conclusion, the early aggressive approach to full supportive care using low tidal volume ventilation and vasopressor for hemodynamic support resulted in rapid recovery from cardiorespiratory failure. In addition, our case demonstrated the clinical benefit of early using high-dose methylprednisolone in HPS.

503 PULMONARY HEMOSIDEROSIS AND RECURRENT PULMONARY EMBOLISM: A MANAGEMENT DILEMMA IN A PATIENT WITH PRIMARY ANTIPHOSPHOLIPID SYNDROME

¹H Edriss, ²GD Bedanie, ²M Lear*, ²K Nugent. ¹Saint Joseph Hospital, Lexington, KY; ²Texas Tech University Health Sciences Center, Lubbock, TX

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Introduction Antiphospholipid syndrome (APS) is an uncommon autoimmune disease. Small vessel (venous and arterial) thrombosis is the most frequent pulmonary manifestation. Recurrent alveolar hemorrhage causing hemosiderosis is rare (<2% of cases).

Case summary A 35-year-old woman with a history of APS, protein S deficiency and recurrent bilateral pulmonary embolism (PE) who was on full dose low molecular weight heparin presented with worsening shortness of breath and increased oxygen requirement. Physical examination revealed HR of 130 BPM, RR of 22 BPM, normal blood pressure, normal temperature, and hypoxemia with SpO₂ of 83% on FiO₂40%. Laboratory testing revealed an elevated cardiolipin IgG and IgM. Protein S was functionality low. Anti-double stranded DNA

and ANA were negative. Computed tomography (CT) angiogram of the chest revealed chronic bilateral sub-segmental PE and bilateral parenchymal nodular and ground glass infiltrates. Since she had missed few doses of LMWH, the initial thought was that she had another episode of acute PE. However, the CT chest finding did not support this. She underwent bronchoscopy and bronchoalveolar lavage (BAL). Lung biopsy was deferred because of pulmonary hypertension. The BAL was blood tinged with a WBC count of 1820 cells/mm³ and RBC count of 697 cells/mm³. BAL cytology revealed abundant hemosiderin-laden macrophages. She was treated with 40 mg of oral prednisone and was discharged to follow up in pulmonary clinic.

Conclusion DAH is associated with high mortality and morbidity. Although APS induced capillitis is described as the likely pathogenesis, the exact pathogenesis of DAH in primary APS is not certain. We hypothesize that another potential pathogenesis would involve elevated pulmonary venous pressures due to venous microthrombosis causing chronic subtle pulmonary hemorrhages and secondary hemosiderosis. This diagnosis may be delayed or missed due to lack of suspicion among clinicians. Based on the currently available evidence, the primary management of DAH includes high dose glucocorticoids with either cyclophosphamide or rituximab.

504 MSSA BACTEREMIA COMPLICATED BY SUSPECTED DAPTOMYCIN-INDUCED EOSINOPHILIC PNEUMONIA

MW Lelli*, LW Aldred, A Wilhelm. University of Mississippi Medical Center, Jackson, MS

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Introduction Eosinophilic pneumonia is a rare, but dangerous, complication of many antibiotics including daptomycin. This condition is defined by cough, new infiltrates on chest X-ray (CXR), and more than 25% eosinophils on bronchoalveolar lavage (BAL). The differential diagnosis for eosinophilia on BAL includes eosinophilic granulomatosis with polyangiitis, fungal and parasitic infections, chronic eosinophilic pneumonia, and acute eosinophilic pneumonia. Daptomycin has been shown to cause acute eosinophilic pneumonia 2 to 4 weeks after initiation. Here, we present a case of daptomycin induced acute eosinophilic pneumonia.

Case report A 47-year-old male with uncontrolled diabetes was found to have methicillin-sensitive *Staphylococcus aureus* (MSSA) bacteremia secondary to tricuspid endocarditis and complicated by vertebral osteomyelitis. He was started on cefazolin but had persistently positive blood cultures. Daptomycin was added on day 10 of admission. His blood cultures cleared and he remained afebrile for 18 days. On day 29 of admission, he became febrile to 101.6° F with a leukocytosis of 13×10⁹/L. Daptomycin and cefazolin were stopped. He was started on vancomycin and cefepime. He remained febrile with an increasing oxygen requirement; a CXR showed diffuse pulmonary infiltrates. He subsequently required intubation. A BAL was performed showing 944 WBC/mm³ with a 37% eosinophilia. He also demonstrated a 16% peripheral eosinophilia. Daily prednisone was started. Over the next two days, he quickly improved. His peripheral eosinophilia resolved and CXR showed improvement. He was successfully extubated and quickly transitioned down to 3 liters nasal cannula. Blood cultures, sputum cultures, and BAL studies never grew a causative organism.

Discussion Daptomycin is a rare cause of eosinophilic pneumonia. This condition typically occurs 2 to 4 weeks after initiation of daptomycin and resolves with discontinuation of the medication and corticosteroids. Of note, this typically recurs if the patient is re-exposed. Although this complication is rare, it should be considered in patients with respiratory decompensation after daptomycin exposure.

505 FATAL CASE OF GROUP A STREP AND INFLUENZA LEADING TO TOXIC SHOCK SYNDROME

J Lucier*, S Lappin. *SUNY Upstate, Syracuse, NY*

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Introduction Influenza leads to thousands of hospitalizations and deaths yearly. Though less common, superinfection with influenza virus and group A strep has been reported and can precipitate invasive processes including acute respiratory distress syndrome, renal failure, toxic shock syndrome, and sepsis.

Case A 51 year old male with a PMH of AML s/p allogeneic bone marrow transplant two years prior, currently in remission, presented to an outside hospital with flu like symptoms that started five days prior. He had a chest xray that showed bilateral infiltrates described as pneumonia vs ARDS. He was intubated for respiratory distress, started empirically on Vancomycin and Zosyn, and transferred to our facility for a higher level of care. He was found to be flu positive and started on Tamiflu. A BMP showed worsening renal function, for which he was started on CVVH. He also developed hypotension and required multiple pressors to maintain his blood pressure. It was decided to start vvECMO for his ARDS. On day three of his hospitalization he began developing a petechial rash on his abdomen that spread to all of his extremities. Blood cultures returned as strep pyogenes. Infectious disease was consulted and recommended Zosyn, Clindamycin, and Vancomycin for toxic shock syndrome secondary to group A strep bacteremia. During his hospitalization the patient was minimally responsive and not improving on his current medical therapies. On day twelve of his hospitalization the patient unfortunately was made comfort care and passed.

Discussion Toxic shock syndrome is a rare complication associated with influenza, though it has been seen in patients with coinfection of group A strep. It is characterized by shock and multiorgan failure secondary to the release of inflammatory cytokines by streptococcal toxins. Treatment includes maintenance of blood pressure with fluids or vasopressors, antimicrobial therapy, and possible surgical debridement. Our patient also developed ARDS, which is associated with a high rate of mortality. ARDS can be treated with ventilator support, prone-positioning, and lastly ECMO, which was attempted in our patient.

Conclusion It is important to keep bacterial coinfections in mind when treating severe cases of the flu, as timely diagnosis and aggressive management may prevent negative outcomes in future patients.

506 TERBINAFINE INDUCED FATAL FULMINANT HEPATIC FAILURE

A Mahajan*, P Sankhyan, T Bhandari, G Hoskere. *East Tennessee State University, Johnson City, TN*

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Case report A 72 year old male with no significant past medical history presented to the hospital with worsening cough, dyspnea, decreased appetite, weight loss and malaise for 2 months. He was previously treated with a 10 day course of Levofloxacin and then a 10 day course of prednisone with minimal improvement. A few days prior to admission he developed dark urine with changes in mental status. On physical examination the patient was noted to have significant scleral icterus, decreased mentation and right upper quadrant tenderness. On admission his labs were significant for alkaline phosphatase 541, aspartate transaminase 557, alanine transaminase 94, total bilirubin 8.6, lactate 11.7; with computed tomography scan showing hepatosplenomegaly, mild ascites and trace bilateral pleural effusions. His acute viral hepatitis serology, cryptococcal antigen (Ag), histoplasma Ag, Respiratory virus panel, Ebstein barr virus were negative. Anti nuclear Antibodies (Ab), anti mitochondrial Ab, Anti Actin Ab were also negative. Amylase, lipase, tylenol, alcohol levels were negative on admission. The patient was initially started on broad spectrum antibiotics, empiric N acetyl cysteine and aggressive IV hydration. However, his condition worsened with profound shock requiring mechanical ventilation, bicarbonate, stress dose steroids, and pressor support.

On further investigation, the patient was noted to be taking terbinafine daily for toe onychomycosis (Day 112). Ferritin levels were elevated at 1596 with 93% iron saturation. Ceruloplasmin levels were within normal limits. The patient was not a transplant candidate due to multi-organ failure. He was subsequently palliatively extubated as per the family's wishes.

Terbinafine is an antifungal agent commonly prescribed for onychomycosis. Minor abnormalities in liver functions tests have been reported in upto 4 percent of patients taking it, however fulminant hepatic failure is a rare but fatal complication. Although anti fungals are a necessary and sometimes life saving category of drugs, their side effects can be dangerous. We believe that the presence of previously undiagnosed hemochromatosis in our patient predisposed him to this rare side effect.

507 HEMOTHORAX FOLLOWING CPR

A McCormick*, S Sanders, A Chimakurthy, LS Engel, D Janz. *LSU Health Sciences Center, New Orleans, LA*

10.1136/jim-2018-000974.505

Case report A 69 year old man with a history of non-small cell lung cancer presented to the hospital for left upper lobe lung resection. Following surgery, the patient suffered cardiac arrest. Return of spontaneous circulation was achieved after one round of chest compressions and one round of epinephrine and he was transferred to the ICU for management of shock and respiratory failure. He regained consciousness but subsequently became hypoxic, hypotensive and required vasopressors. Chest x-ray revealed a new left lung opacity and he underwent bronchoscopy, revealing extrinsic compression of left lower lobe. Computed Tomography (CT) imaging of the chest demonstrated findings consistent with an active bleed and coagulated blood in the left chest. A chest tube was placed and drained approximately one liter of blood. Chest compressions following cardiac arrest were thought to likely be the cause of hemothorax, and the patient underwent surgical evacuation of hematoma the following morning. An

additional 1.5 L of coagulated blood was removed, and patient began recovery in the intensive care unit.

Discussion The most common thoracic injuries following CPR include rib fracture, sternal fracture, lung contusion and pneumothorax. Hemothorax may also result from traumatic chest compressions. Diagnosis can prove to be difficult as the patient may only have vague chest pain complaints, with physical exam as the only indicator of internal chest trauma. Tachycardia, hypotension, decreased/absent breath sounds on the corresponding side, and other nonspecific signs of shock are common. Initial preferred diagnostic imaging is a chest x-ray. In a critical care or trauma setting, an ultrasound may be favored due to its rapidity. CT scan requires more time and a supine patient, making it a less than ideal study for initial diagnosis. Management involves controlling the source of the bleed and draining blood that has accumulated in the thorax. Complications can occur even in the setting of prompt treatment; chest tube and surgery may be required to stop continued bleeding or evacuate blood that does not drain through a chest tube.

508

IMMUNOSUPPRESSANTS CAUSING A DEVASTATING MALIGNANCY

M Milan Esteve*, R Fernandez, RA Hernandez, V Fonseca-Ferrer. *Hospital Municipal de San Juan, San Juan, Puerto Rico*

10.1136/jim-2018-000974.506

Case report Solid Organ Transplantation (SOT) provides a life-saving therapy for patients with end-stage organ disease nevertheless, recipients have an elevated risk of cancer due to immunosuppression, mostly after lung and kidney transplant, few cases have been reported after liver transplant. This is a 67-year-old female with past medical history of hypothyroidism, chronic back pain related to thoracic compression fracture who underwent an Orthotopic Liver Transplant (OLT) in 2017 secondary to Primary Biliary Cirrhosis. Her post-operative course had no complications and she was started on immunosuppressive therapy with Tacrolimus, Prednisone, Mycophenolate Mofetil and Valgancyclovir. She presented to the Emergency Room complaining of left side back pain and shortness of breath 2 days duration. Denied chest pain, fever, chills, cough, palpitations, but recalls having slipped from bed a few weeks back and pulled a back muscle. Chest CT revealed no filling defect and large left pleural effusion with complete collapse of left lung. Thoracostomy tube was placed and pleural fluid analysis was remarkable for an exudative effusion, negative gram stain, RBC 152,000 and hematocrit >50%; last values consistent with a Non-Traumatic Hemothorax. Thoracostomy tube was removed 5 days later after full expansion of the lung without residual fluid and eventually she was discharged home. After 2 months she came back with shortness of breath. At that time images revealed small loculated pleural effusion and diffuse pleural thickening throughout the left lung. Due to history of OLT and the use of immunosuppressive medications and a restrictive pattern on pulmonary function tests, Pleuropulmonary Fibroelastosis was suspected but since it has not been associated with liver transplant she underwent a pleural biopsy which was positive for malignant mesothelioma. Days after procedure she developed worsening shortness of breath requiring mechanical ventilation support for which she never recovered. This case is unusual due to the rare relation between SOT and development of aggressive pleural mesothelioma after OLT. To the best of

our knowledge, only 1 case has been documented with mesothelioma after liver transplant without the usual and known risks factors.

509

ARDS AS A RESULT OF SEROTONIN SYNDROME

T Moffitt*, FC Moore, A Ramage. *Eisenhower Army Medical Center, Grovetown, GA*

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Introduction We present a case of a 34 year-old woman who was admitted for serotonin syndrome from a Venlafaxine overdose which resulted in ARDS (acute respiratory distress syndrome). There have been prior cases of ARDS resulting from serotonin syndrome. Our case represents one of the first reported cases of ARDS as a complication of serotonin syndrome resulting from Venlafaxine overdose.

Our patient is a 34 year-old woman with a history of multiple suicide attempts and diagnosed MDD who was admitted for observation following an intentional overdose of unknown amount of venlafaxine and alcohol. She initially presented to the emergency department approximately 6 hours post-ingestion, endorsing visual hallucinations, tremor, and inability to ambulate. Review of systems was otherwise negative, with the exception of suicidal ideation.

On initial exam she was noted to be tachycardic and tachypneic with normal oxygen saturations and blood pressure. Patient was not hyperpyrexia. She was easily distracted and oriented to person only. Neurologic exam was significant for upper extremity tremors and increased muscle tone.

She initially improved but later worsened with increased tremors and agitation. She had diffuse, severe muscle rigidity with sustained ankle clonus on repeat exam.

She developed worsening hypoxemia with SPO2 measurements in the low 80 s despite methods of non-invasive oxygenation. She was transferred to ICU and required endotracheal intubation. On chest x-ray there were bilateral alveolar infiltrations typical of non-cardiogenic pulmonary edema and consistent with ARDS. PaO2/FiO2 ratios were noted to be less than 200. She had no cardiac history nor physical exam findings to suspect heart failure.

Cyproheptadine was given via oral-gastric tube. She remained intubated for several days but eventually recovered and was discharged without any lasting consequences.

Discussion/conclusions The Serotonin Syndrome can present with variable spectrum of signs and symptoms. Clinicians need to have a low index of suspicion for adverse complications of serotonin syndrome, even in patients without other significant medical comorbidities. ARDS, though relatively rare, is a known complication of serotonin syndrome.

510

A RAPID DIAGNOSTIC MAY IMPROVE HEALTH CARE DELIVERY TO PATIENTS WITH SEPSIS IN THE EMERGENCY DEPARTMENT

¹KA Morgan*, ²K Vance. ¹LSU New Orleans Medical School, Baton Rouge, LA; ²LSU Health Science Center, New Orleans, LA

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Purpose of study Sepsis is a common and deadly syndrome. Effective treatment requires rapid identification and intervention by the treating clinicians. Delays in care result in increased

morbidity and mortality. Unfortunately, indiscriminate treatment results in the untoward consequences of increased resource utilization and antibiotic prescription. A rapid and accurate early diagnostic can both improve outcomes of sepsis while minimizing excess resource utilization and achieve the goals of antibiotic stewardship. We conducted an analysis of a prospective cohort of patients to determine if a rapid diagnostic could improve our institution's compliance with recently published sepsis guidelines.¹

Methods used Patients were prospectively identified and enrolled in two similar cohorts consisting of potentially septic patients presenting to the Emergency Department (ED) of a large academic medical center between February and July of 2018. Data abstracted from the patients' charts included time of triage in the ED, time of initial laboratory orders, blood draw, and initial laboratory results [Complete Blood Count (CBC)], as well as orders for fluids, parenteral medications, and intravenous antibiotics. We also collected times of administration for these parenteral interventions. We performed descriptive statistical analysis representing triage as Time 0.

Summary of results A total of 68 patients were included in our analysis. All times reported as means. Clinicians placed the initial laboratory order at 22 min with the collection at 40 min and results at 70 min. Clinicians placed the initial antibiotic order at 123 min with administration at 175 min. They ordered fluids at 36 min with administration at 64 min. Clinicians also ordered parenteral drugs at 68 min with administration at 100 min.

Conclusions Current clinician behavior is not adherent to the most recent guidelines for the treatment of sepsis. A rapid diagnostic which results with a CBC could potentially reduce the mean time to the ordering and administration of antibiotics by up to 53 and 105 min respectively.

REFERENCE

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511 LARYNGEAL PENETRATION WITHOUT ASPIRATION AND PERSISTENT RIGHT UPPER LOBE ATELECTASIS: THINK LARYNGEAL CLEFT

K Morgan*, N Mehdi. *University of Oklahoma Health Sciences Center, Oklahoma City, OK*

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Case report 6 month old girl with a history of laryngomalacia and chronic respiratory illnesses presented with respiratory distress and increased oxygen requirement for sixth admission. She had five prior hospitalizations for similar episodes of respiratory distress requiring increased oxygen, including one pediatric intensive care unit admission requiring non-invasive ventilation. During prior admissions, she tested positive for parainfluenza and human metapneumovirus and chest xray (CXR) findings of persistent right upper lobe opacities. Three prior dysphagiagrams showed laryngeal penetration without aspiration. Upper GI unremarkable. Nuclear medicine esophageal motility study without evidence of aspiration. Immunology work up negative. Chest CT showed right upper lung consolidation with air bronchograms and bilateral mosaic pattern of perfusion likely small airway disease and bilateral groundglass and linear opacities representative of chronic lung changes. During sixth admission, she had coarse breath sounds, intermittent wheeze, tachypnea, and subcostal retractions that resolved

on high flow oxygen. Otorhinolaryngology (ORL) and Pulmonology performed airway evaluation with direct laryngoscopy and bronchoscopy and identified a type II laryngeal cleft. She had failed conservative therapy with thickened feeds and reflux precautions. ORL performed injection of laryngeal cleft with water based gel implant. Bronchoalveolar lavage showed high WBC with neutrophilia and grew multiple bacteria. She did well after laryngeal cleft injection and was discharged home to complete a 10 day course of augmentin. At follow up, she had clinically improved and was breathing comfortably with clear breath sounds. Although laryngeal cleft is a rare diagnosis, it is important to consider in patients with chronic respiratory illnesses with other negative work up. Microlaryngoscopy is the gold standard for diagnosis. Type II laryngeal clefts often delayed diagnosis due to vague history of chronic respiratory and feeding issues that gradually worsen over time. Therefore, maintaining a high index of suspicion for laryngeal cleft in patients with chronic respiratory infections and CXR suggestive of aspiration is important in order to have an early diagnosis and prevent pulmonary complications.

512 MICRORNA-144 MEDIATES ALVEOLAR MACROPHAGE DYSFUNCTION IN HIV-1 TRANSGENIC RATS

¹SC Murray*, ¹X Fan, ^{1,2}D Guidot. ¹Emory University, Atlanta, GA; ²Atlanta VA, Decatur, GA

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Purpose of study Despite anti-retroviral therapy, individuals with HIV are at high risk of pneumonia. HIV-1 transgenic (HIV-1 Tg) rodent models implicate HIV-related proteins, such as gp120 and Tat, as mediators of multiple complications of chronic HIV infection. Previously we determined that these proteins inhibit Nrf2, which activates anti-oxidant and immune defenses, and impair alveolar macrophage (AM) phagocytic function. Several microRNAs are known to inhibit Nrf2; based on a recent screen we focused on microRNA-144 (miR-144) and examined its role in HIV-mediated AM dysfunction.

Methods used Primary AMs from wild type (WT) and HIV-1 Tg rats were isolated and analyzed *ex vivo* for expression of miR-144, Nrf2, and Nrf2-dependent antioxidants (by RT-PCR and/or western immunoblotting) as well as bacterial phagocytic capacity (using fluorescent bacteria). In selected experiments, AMs from WT rats were treated *ex vivo* with a miR-144 mimic; in other experiments WT and HIV-1 Tg rats were treated with a miR-144 antagomir intratracheally (3 doses over 6 days) and AMs assessed for Nrf2 expression and phagocytic function. Finally, we quantified the expression (by RT-PCR) of two transcription factors (AP-1 and CP2) known to induce miR-144 in AMs from HIV-1 Tg rats.

Summary of results miR-144 expression was increased ~4 fold in AMs from HIV-1 Tg rats, and treating WT AMs with the miR-144 mimic inhibited their phagocytic function. In contrast, treating HIV-1 Tg rats *in vivo* with the miR-144 antagomir increased AM expression of Nrf2 as well as the Nrf2-dependent antioxidants NQO1 and GCLC and, in parallel, restored phagocytic capacity. Finally, the expression of both AP-1 and CP2 were increased in AMs from HIV-1 Tg rats but only the c-Fos subunit of AP-1 was suppressed by the miR-144 antagomir.

Conclusions Chronic expression of HIV-related proteins in the airways induces miR144 expression in AMs and this in turn

inhibits Nrf2 and Nrf2-dependent antioxidants as well as bacterial phagocytic capacity. These chronic pathophysiological effects can be reversed in this experimental model by antagonizing miR-144 *in vivo*. Taken together, these results suggest that targeting miR-144 and/or its downstream effects such as Nrf2 inhibition and AM immune dysfunction could enhance lung health in individuals living with HIV.

513 TNFALPHA MODULATES TSP-1 IN A549 CELLS

M Pacurari*. Jackson State University, Jackson, MS

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Purpose of study TNF α plays a key role in inflammation particularly in acute lung injury (ALI) and initiates a cascade of signaling pathways. Thrombospondin -1 (TSP-1) a matricellular glycoprotein has been shown to play a role in acute inflammation. To further identify the mechanism by which TNF α modulates inflammation, we used bioinformatics tools and identified TSP-1 is a target of microRNA-1. Moreover, microRNA-1 also has binding sites for Notch 2 that plays a key role in cell fate determination during lung development and lung remodeling. In the present study, we investigated whether TNF α regulates TSP-1 in human alveolar epithelial cells. In the present study, we investigated whether TNF α regulates TSP-1 in human alveolar epithelial cells.

Methods used Using A549, the regulation of TSP-1 by TNF α were analyzed using real-time qPCR, Western blot, and IHC. A549 cells were exposed to TNF α (1 or 10 ng/ml) for 6 or 24 hour. Total RNA was extracted using TRIzol. and cDNA were generated and analyzed by real-time qPCR with specific primers. Western blot and IHC were performed using specific antibodies and fluorescent microscopy.

Summary of results Our preliminary results show that exposure of A549 cells to TNF α , suppressed the expression of TSP-1. TNF α at 1 ng after 6 and 12 hour of exposure suppressed TSP-1 mRNA by 32% and 45% vs control, whereas TNF α at 10 ng after 6 and 24 hour further suppressed TSP-1 mRNA by 47% and 55%. Target scan analysis revealed that TSP-1 is a target of miR-1. To further investigate the possible mechanism responsible for suppression of TSP-1 by TNF α , we investigated the effect of TNF α on miR-1. miR-1 was differentially suppressed by TNF α ; low dose (1 ng) suppressed miR-1 whereas a higher dose, 10 ng, had no effect after 6 hour of treatment. Notch 2 mRNA was upregulated regardless of TNF α dose and time.

Conclusions These results suggest TSP-1 downregulation is independent of miR-1. However, up-regulation of Notch 2, a well-known regulator of cell fate determination, suggests that alveolar epithelial cells under inflammatory signaling undergo remodeling more-likely based on cell-cell communication rather than matrix-mediated signaling.

514 IMPACT OF MUCOLYTIC PAAG ON KLEBSIELLA PNEUMONIAE BIOFILM

JT Powner*, WE Swords. University of Alabama at Birmingham, Birmingham, AL

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Purpose of study Polyacetyl Arginyl Glucosamine(PAAG) is a soluble nontoxic polycationic polysaccharide that has purported

activity on bacteria biofilm. The mechanism of action involves interaction of the cation with bacterial membranes causing disaggregation and pore formation. These actions reduce bacteria, disrupt biofilm, and prevent biofilm formation. PAAG has possibly been shown to reduce and treat biofilm forming bacteria with multi-drug resistance strains.

Klebsiella pneumoniae(Kp) is a gram negative rod that is a common hospital-acquired infection in humans. It can be multi-drug resistant and produce biofilm. Due to these traits, Kp is an ideal bacteria to test the ability of PAAG on biofilms and test for synergy with an antibiotic.

Methods used Microtiter dish biofilm formation assay was used to quantify the biofilm formation and effectiveness of the treatments. Kp was grown in fresh BHI media overnight and then diluted to 1.5 ODU. Then 200 μ l of the dilution was put in 4 columns of a microtiter dish and incubated for 24 hours at 37°C. Each well was then washed with sterile water. One column was left untreated and had 100 ml of BHI added to each well, one was treated with cefuroxime 0.1 mg/ml and BHI solution, one was treated with 200 μ l/ml of PAAG and BHI solution, and one received both treatments. The dish was then incubated for 6 hours. Each well was washed and then dyed for 30 min with 100 ml of 0.1% crystal violet. After washing again, the wells received 100 ml of 100% ethanol. After 10 min the spectrophotometer was used to quantify the biofilm in each well. Graphpad Prism 7.0 was then used to run two-way anova tests of the data.

Summary of results Kp grew measurable biofilm by microtiter biofilm assay. Treatment with PAAG caused a reduction in biofilm mass. Cefuroxime produced a reduction in biofilm that was more than PAAG alone. PAAG and Cefuroxime together produced the highest reduction in biofilm. All treatments arms were significant when compared against each other ($p < 0.05$).

Conclusions PAAG and Cefuroxime was able to reduce the biofilm produced by Kp. When used together PAAG and Cefuroxime further reduced the biofilm. PAAG appears to have antimicrobial effects that are further enhanced when used together with an antibiotic.

515 INFLAMMATORY RESPONSE INDUCED BY NONTUBERCULOUS MYCOBACTERIA IS ASSOCIATED WITH ALTERED CELL ENERGY METABOLISM IN MACROPHAGES

¹Z Prasla*, ^{1,2}C Bedi, ^{1,2}RT Sadikot. ¹Emory University, Atlanta, GA; ²VA Hospital, Atlanta, GA

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Purpose of study The incidence of nontuberculous mycobacterial (NTM) lung disease is rising worldwide and accounts for most cases of NTM infection. Mycobacterium avium complex (MAI) is a well-known cause of infection in immunocompromised patients. However, pulmonary NTM infections are not limited to immunocompromised hosts, and Mycobacterium abscessus (MAB) is particularly difficult to treat in patients with COPD, non-CF bronchiectasis, and CF. Despite abundant knowledge of the clinical significance of NTM, relatively little is known about host immune response to NTM infection. Our objective was to define the immune-metabolic response induced by NTM in macrophages.

Methods used We infected RAW cells (murine immortalized macrophages) and MH-S cells (murine alveolar macrophages)

with clinical strains of MAI or MAB (MOI of 10, 25, or 50) for 6, 24, and 48 hours. We determined the expression of key inflammatory genes (COX-2, IL-1 β , and TNF α) that contribute to immune response to infections by using RT-qPCR to calculate their fold change relative to GAPDH. We used this infection model to perform a cell energy phenotype assay in MH-S cells infected by MAI.

Summary of results We found that in RAW cells, COX2 and TNF α were increased at 48 hours, with an increase in TNF α noted as early as 24 hours. There was no increase in IL-1 β production in RAW cells. In comparison, in MH-S cells, there was a robust increase in IL-1 β that was noted as early as 24 hours with a modest increase in TNF α seen at 24 hours. These results were confirmed with an IL-1 β ELISA assay, which showed peak concentration at 24 hours but was noted as early as 6 hours. Cell energy phenotype analysis showed that at baseline, ECAR was increased in infected MH-S cells relative to control cells.

Conclusions These data suggest that there is a difference in the immunologic phenotype of macrophage host response to NTM, which may be dependent on the type of macrophage. In MH-S cells, MAI appears to shift cells to a more glycolytic phenotype. The functional impact on bacterial killing and phagocytosis of changes in these inflammatory genes and their associated alterations in cell energy metabolism need to be further investigated to elucidate new mechanisms of host response that aid treatment of NTM infections.

516

WHERE IS THE AIR? PNEUMOTHORAX VERSUS PULMONARY BULLA

K Ramakrishna*, P Desai, A Pathmanathan, H Singh Gambhir. *SUNY Upstate Medical University, Syracuse, NY*

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Case report A pulmonary bulla is defined as an airspace in the lung that is greater than 1 cm in diameter. A giant bulla is

one that occupies more than 30% of a hemithorax. It is a rare but important mimicker of pneumothorax

A 43 year old male with a history significant for COPD and known unexplained, bilateral pulmonary bullous disease presented to the ER with acute dyspnea and right sided pleuritic chest pain. The patient was hemodynamically stable but hypoxic to 88% on pulse oximetry, requiring 4 L of supplemental oxygen to maintain saturations above 95%. There was decreased air entry on the right side on auscultation and hyperresonance noted on percussion. Bilateral rhonchi were also heard. Clinical exam thus appeared to suggest a right pneumothorax which was supported by the chest X ray which demonstrated a new, large pneumothorax in the right hemithorax. A CT Thorax was done (given history of bullous disease) revealing the presence of a giant bulla almost obliterating the right lung with only collapsed lung inferiorly. Chest tube insertion was therefore not pursued. The patient's respiratory distress was treated as COPD exacerbation with steroids and bronchodilators, with improvement in symptoms. Differentiating between pneumothorax and a pulmonary bulla which is not immediately apparent on clinical exam or CXR requires further imaging such as chest ultrasound or CT scan. This can definitively identify a bulla and rule out pneumothorax, thus avoiding the unnecessary and potentially hazardous insertion of chest tubes.

517

MATRIX METALLOPROTEINASE-9 IS ENHANCED BY INTERFERON GAMMA AND HIV-1 TRANSGENE EXPRESSION IN RAT ALVEOLAR MACROPHAGES

¹AT Roth*, ¹M Ahmed, ¹SC Auld, ¹X Fan, ^{1,2}D Guidot, ¹BS Staitieh. ¹Emory University, Atlanta, GA and; ²Atlanta VA, Decatur, GA

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Purpose of study HIV is known to increase the risk of emphysema, but the underlying mechanism remains a mystery. Given the known association of matrix metalloproteinase 9 (MMP-9)

Abstract 516 Figure 1 CXR showing an apparent right pneumothorax. CT thorax showing giant bulla in right hemithorax with definite bleb wall (arrow)

and emphysema, one possibility is the elevation of MMP-9 levels in the lungs of persons living with HIV (PLWH). Using an HIV transgenic rat model to simulate the lungs of PLWH, we previously determined that chronic exposure to HIV-related viral proteins increased MMP-9 protein levels. We therefore designed a series of experiments to assess MMP-9 activity (using its known cleavage target RAGE, the receptor for advanced glycation end products) and to begin examining the mechanism of MMP-9 elevation.

Methods used To determine the effects of macrophage activation state on MMP-9 expression, a rat alveolar macrophage cell line (NR8383) was treated with interferon-gamma (IFN- γ) or interleukin-4 (IL-4) prior to assessment by qRT-PCR. In parallel, AMs isolated from whole-lung lavage of both HIV-1 transgenic rats and their wild-type littermates were stained with tissue inhibitor of MMP 1 (TIMP-1) for fluorescence microscopy. AMs from HIV-1 transgenic rats were also treated with or without an MMP-9 inhibitor for 24 hours prior to staining for RAGE for immunofluorescence.

Summary of results IFN- γ markedly increased MMP-9 expression relative to IL-4. MMP-9 AMs from HIV-1 transgenic rats demonstrated significant impairment in TIMP-1 expression by immunofluorescence. RAGE levels were significantly decreased at baseline in HIV transgenic macrophages, but were increased markedly by treatment with an MMP-9 inhibitor.

Conclusions Shifting macrophages to an inflammatory state with IFN- γ markedly increased MMP-9 expression, suggesting one potential mechanism for MMP-9 modulation. HIV transgene expression also increases TIMP-1, which suggests another potential mechanism by which MMP-9 activity could be increased, as TIMPs suppress baseline metalloproteinase activity. The decrease in membrane-bound RAGE levels, and the significant increase in levels with direct MMP-9 inhibition strongly suggests that MMP-9 activity is increased in HIV-1 transgenic rat macrophages. These data support the underlying hypothesis that MMP-9 activity may be responsible, at least in part, for the increased risk of emphysema in PLWH.

518

SURVIVING A CORE BODY TEMPERATURE OF 70.5 FAHRENHEIT

¹P Sankhyan*, ²G Singh, ¹N Amilineni, ³A El-Abbassi. ¹East Tennessee State University, Johnson City, TN; ²Dayanand Medical College, Ludhiana, India; ³East Tennessee State University, Johnson City, TN

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Case report We present the case of a 29 year old male, with a past medical history of alcoholism and intravenous (IV) drug abuse, who was presented to the hospital with a core body temperature of 70.5 Fahrenheit (F). The patient was passed out all night in an open garage in the winter, with a temperature of 20–24 F. He was found gasping and unresponsive in the morning. He also had frostbite of both his toes going up to the metatarsals bilaterally. He was placed on mechanical ventilation for airway protection. Initially an IV access could not be obtained, so he was given normal saline through an intra-osseous line and then a central venous catheter was placed, he received aggressive IV hydration with warm saline and an external warming blanket. He was also severely hypokalemic, acidotic with total creatine kinase (CK) level 24845 U/L; with benzodiazepines and amphetamines in his urine drug screen and a blood alcohol level of 462 mg/dL. We cautiously replaced his potassium for concern of

hyperkalemia from muscle breakdown. His electrolytes were closely monitored while we continued aggressive IV hydration with warm saline and bicarbonate to protect his kidneys from myoglobin induced damage. The patient was brought to a core body temperature of 99 F within 10 hours. However, his feet continued to be cyanotic and was not found to be a candidate for hyperbaric oxygen therapy. He was given gapapentin for hyperaesthetic pain. Despite his high CK levels, because of timely and adequate hydration and renal protective measures, he never developed evidence of acute kidney injury, with his highest Creatinine level being 1.17 mg/dL.

Hypothermia can become a life threatening condition unless acted upon quickly and aggressively. There are many rewarming techniques described in the available literature including re-warming with internal alius catheters, peritoneal catheters, intra thoracic re-warming and so on. We only used warm IV saline and external heating blanket and our patient responded very well to the treatment. However, when using an external cooling blanket, care has to be taken to include all the extremities of the patient to avoid the phenomenon of core temperature afterdrop.

519

AMIODARONE INDUCED MYXEDEMA COMA IN A PATIENT WITH CONGESTIVE HEART FAILURE

¹N Mahajan, ²P Sankhyan*, ²A Mahajan, ²A El-Abbassi. ¹BJ Medical College, Pune, India; ²East Tennessee State University, Johnson City, TN

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Case report We present the case of an 83 year old gentleman with a history of atrial fibrillation with rapid ventricular response for which he was on amiodarone and apixaban. The patient was recently seen at another hospital for shortness of breath and was found to have an ejection fraction of 10%, with severely decreased systolic function of the left and right ventricles. He did not have evidence of acute coronary syndrome at the time with negative troponins and electrocardiogram showing no acute changes suggestive of ischemia. He was referred to our hospital with worsening renal failure and shock liver. The family wanted a second opinion after the previous hospital had suggested a hospice evaluation. His son mentioned that the patient had been lethargic, somnolent and had increasing memory problems for a few months but this was attributed to his underlying dementia. On exam he was minimally responsive and had to be placed on non invasive positive pressure ventilation for respiratory distress. On a routine check, his TSH level was 169.60. He did not have a prior history of thyroid disease. He was then started on levothyroxine 200 micrograms, hydrocortisone 100 milligrams thrice a day and a furosemide drip for anuric acute kidney injury in the setting of severe congestive heart failure. The patient next morning was more awake as compared to admission but not oriented and continued to have worsening liver failure. He was subsequently made hospice per the family's request.

Amiodarone is commonly used for rate control in atrial fibrillation, especially when blood pressure control is difficult. However, it is not a harmless drug. It has been implicated in severe adverse effects like liver failure, pulmonary fibrosis and thyroid dysfunction. It is important to educate patients about signs of pulmonary toxicity and to seek timely help in case they develop symptoms of hypersensitivity pneumonitis or

alveolar pneumonitis. However, the treatment of myxedema coma, due to amiodarone along with acute exacerbation of congestive heart failure, remains unclear, with the risk of inducing arrhythmias and worsening heart failure with high dose levothyroxine therapy.

520 A PEDIATRIC CASE OF UNILATERAL RENAL ARTERY STENOSIS PRESENTING WITH POLYURIA

F Sifers*, V Godsey, C Rutledge. *University of Alabama-Birmingham, Birmingham, AL*

10.1136/jim-2018-000974.518

Case report Secondary hypertension is the most frequent form of hypertension in children. Renal artery stenosis (RAS) is a rare but important cause of childhood hypertension. Early diagnosis and treatment is essential to prevent target organ damage. Polyuria can occur in the setting of RAS due to activation of the renin-angiotensin system causing secondary hyperfiltration and pressure diuresis leading to sodium loss from the contralateral non-stenotic kidney. This hyperfiltration can lead to the loss of solutes causing hypokalemia, alkalosis, and proteinuria.

We present a case of a patient with polyuria and hypertensive urgency secondary to RAS.

A 4 year old female with no significant medical history presented with a two day history of headache and vomiting. She was found to be profoundly hypertensive with a blood pressure of 200/150. Her initial laboratory workup was notable for hypokalemic metabolic alkalosis and mild hyponatremia. Her urinalysis had moderate proteinuria and trace ketones. A nicardipine infusion was started to slowly lower her blood pressure. She was noted to be polyuric with urine output greater than 7 ml/kg/hr. Hyperhydration was initiated to prevent further kidney injury. Diagnostic workup was performed to evaluate the cause of her hypertension and included renal ultrasonography, urine toxicology screen, plasma catecholamines, renin and aldosterone activity, and echocardiogram. Her echocardiogram showed left ventricular hypertrophy. Renal ultrasound revealed hazy echogenic material in the junction between the pyramids and papilla of the left kidney concerning for nephrocalcinosis versus proteinuria. A computed tomography angiography of the abdomen was performed and revealed a normal single right renal artery and left renal artery stenosis and abnormal cortical thinning within the upper pole of the left kidney. She was diagnosed with unilateral RAS as the cause of her hypertension. She was continued on a nicardipine infusion being transitioned to oral medications. An angiotensin-converting enzyme (ACE) inhibitor was started initially. She continued to be hypertensive despite maximizing the ACE inhibitor prompting the addition of a beta blocker. Her hypokalemic metabolic alkalosis and polyuria resolved with normalization of her blood pressure prior to discharge home.

521 DOUBLE TROUBLE IN AN IMMUNOCOMPROMISED HOST

D Sotello*, J Crane, K Nugent. *Texas Tech University Health Sciences Center, Lubbock, TX*

10.1136/jim-2018-000974.519

Case report A 76-year-old female with past medical history of rheumatoid arthritis (RA), COPD, and history of tobacco use (25 pack/years).

Her RA was treated with weekly abatacept and methotrexate, with good control. She was admitted for evaluation of progressive dyspnea, chronic productive cough associated with generalized fatigue. For this reason, a chest x ray was requested and showed bilateral lung nodules. A chest computed tomography (CT) showed a 2.1×1.8 cm right upper lobe (RUL) nodule and a 2.4×2.4 cm left lower lobe (LLL) nodule, both with spiculated borders and central cavitation. A respiratory culture was requested and Gram stain showed 1 + gram positive rods with beaded and branching filaments, the culture eventually confirmed *Nocardia* spp. A CT of the head ruled out central nervous system involvement. The patient was initially treated with linezolid and trimethoprim/sulfamethoxazole (TMP/SMX), and after susceptibilities were reviewed the antibiotic regimen was changed to doxycycline and TMP/SMX, the patient experienced some clinical improvement. A repeat CT was done 3 months later, it showed: the RUL lesion grew to 2.7×1.8 cm and the LLL lesion grew to 3.6×4.1 cm. Subsequently CT-guided biopsy of the pulmonary lesions was performed and confirmed invasive squamous cell carcinoma in the LLL mass and chronically inflamed scarred lung tissue in the RLL nodule. The tumor had high expression of PDL-1, for this reason pembrolizumab was added started. The patient expired 5 months later due to sepsis.

Abstract 521 Figure 1

Immunocompromised patients are at risk of opportunistic infections, such as Nocardiosis, in our case this could have explained the etiology of the original pulmonary lesions, it is important to keep an open mind about alternative diagnosis when there is clinical or radiological worsening despite adequate management.

522 PERSISTENT TACHYPNEA SECONDARY TO PULMONARY INTERSTITIAL GLYCOGENOSIS

N Soulages Arrese*, N Mehdi, R Katz. *University of Oklahoma Health Sciences Center, Oklahoma City, OK*

10.1136/jim-2018-000974.520

Case report This report highlights a case of a term neonate with persistent tachypnea who was found to have Pulmonary interstitial glycogenosis (PIG), a rare pediatric interstitial lung disease. Our patient is male neonate who was born at 38 weeks gestation, by spontaneous vaginal delivery, received short period of PPV after delivery, and required NICU admission at 24 hours of life for tachypnea with respiratory rate (RR) to maximum of 120 without hypoxemia. He presented to the general pediatric service at 8 days of age, after being discharged to home, with persistent tachypnea, reported RR

of 100 while sleeping and choking with feeds. On exam he had persistent tachypnea with RR greater than 60, without hypoxemia, and intercostal retractions. Extensive work-up was completed: infectious work-up was negative, echocardiogram with small patent foramen ovale, high resolution computed tomography with scattered groundglass opacities in the lungs bilaterally most prominent in the bilateral lower lobes, genetic testing included all negative. During hospitalization, the patient received supplemental oxygen without clinical improvement in respiratory rate. Given that patient was well appearing without hypoxemia, he was discharged to home at one month of age, with persistent tachypnea and a nasogastric tube in place for feeding. Lung biopsy was completed at two months of age. On microscopy, sections showed partially inflated lung tissue with alveolar growth abnormality characterized by enlarged and simplified alveoli. On electron microscopy, spindle-shaped cells containing large amounts of monoparticulate glycogen within the interstitial compartment were seen, indicative of pulmonary interstitial glycogenosis. Prior to diagnosis with biopsy, the patient was already improving clinically with respiratory rate in the 40's while asleep. Once diagnosed with PIG, the patient was started on oral steroids. This case highlights a likely under-recognized disease that supports the need to maintain a broad differential for tachypnea and respiratory distress and consider interstitial lung disease, specifically PIG under two years of age for which lung biopsy is required.

523 TRIPLE THERAPY FOR DIFFUSE ALVEOLAR HEMORRHAGE INDUCED BY LEPTOSPIROSIS

G Veloz Irizarry*, A Rodriguez, D Boodoosingh. *San Lucas, Ponce, PR, Puerto Rico*

10.1136/jim-2018-000974.521

Case report Leptospirosis is a bacterial disease that affects humans and animals. It can cause a wide range of symptoms, with a severe form known as Weil's disease, complicated by jaundice, acute kidney injury, pulmonary hemorrhage, acute respiratory distress syndrome, hepatic failure, thrombocytopenia and shock.

39-year-old Puerto Rican male patient without past medical history who presented with dyspnea, fever, and chills that started 3 days before admission. Vital signs in the emergency room were notable tachycardia, fever and oxygen saturation 73% – room air. Physical exam was remarkable for bilateral crackles and respiratory distress. Chest x-ray show bilateral patchy alveolar opacities suggestive of acute respiratory distress syndrome. He was intubated and transferred to the Intensive Care Unit (ICU). The next day his condition complicated with severe pulmonary hemorrhage confirmed by bronchial lavage and bilateral pneumothorax. He continued with refractory hypoxemia despite conventional treatment for which ICU team decided to start high dose intravenous steroids, inhaled nitric oxide and desmopressin. Patient was monitored with arterial oxygen tension (PO₂), fractional inspired oxygen (FiO₂) and Pao₂/FiO₂ ratios daily. He improved after triple treatment and was successfully extubated without major complications.

This case demonstrated Weil's disease with severe pulmonary involvement and hypoxemic respiratory failure. He was treated with high dose intravenous steroids, inhaled nitric oxide and desmopressin with resolution of symptoms, such therapy helped reduce further complications by targeting

Abstract 523 Figure 1 PaO₂/FiO₂ after triple therapy

inflammatory process, hemostasis and pulmonary vasoconstriction.

Renal, Electrolyte and Hypertension

Joint Plenary Poster Session and Reception

4:30 PM

Thursday, February 21, 2019

524 MINIMAL CHANGE DISEASE IN AN ELDERLY PERSON WITH SPONTANEOUS RESOLUTION: THE TIP OF THE ICEBERG?

M Abdou*, MA Alsharif, M Naguib, RS Urban. *Texas Tech Univ HSC Amarillo, Amarillo, TX*

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Case report Minimal change disease (MCD) is the predominant form of primary nephrotic syndrome (PNS) in children. However, adult and elderly onset MCD is much less common and counts for 10%–15% only of PNS cases. Although the exact underlying cause of MCD is not fully understood, some evidences suggest T cell dysfunction.

We are reporting a case of new onset MCD, biopsy proven in a 78 year-old woman with history of hypertension, hypothyroidism, and osteoarthritis who presented with 5 days of progressively worsening generalized swelling, fatigue, puffy eyes, and nausea. She also noticed worsening blood pressure for 2 weeks that required adding a second antihypertensive agent.

The patient denied any urinary symptoms including hematuria. Her home medications were losartan, hydrochlorothiazide, levothyroxine and occasional over the counter ibuprofen.

Initial evaluation in the emergency room showed blood pressure of 149/80. Lab revealed serum sodium 128 mmol/L, potassium 3.2, and chloride 92. Serum creatinine was 1.1 mg/dL and eGFR 48 mL/min. Albumin was 1.3 g/dL and total protein 4.5. Urine was yellow and cloudy with 3+proteinuria, 0–2 WBCs/HPF, 3–5 RBCs/HPF but negative nitrate and leuko-esterase. 24 hours urine collection had 12.1 g of protein. Workup was negative for protein electrophoresis, light chain, autoimmune and hepatitis panels.

Kidney biopsy showed minimal change glomerulopathy with moderate foot process effacement under electron microscopy. Over the next few days her edema improved with furosemide but also serum creatinine declined to 0.8 and eGFR was 70 mL/min on discharge. She did not receive any steroids

during the hospital stay and was instructed to follow up in the nephrology clinic.

MCD is relatively rare in elderly and is associated with less relapse. However, the condition is important due to the higher hypertension and renal impairment and the slower response to steroids therapy. Some studies suggested yearly incidence rate to be 3 per million. However, the spontaneous resolution is of particular interest as it may suggest under recognition since some of the persons who will not present promptly to care spontaneously recover.

525 SEVERE HYPOKALEMIA: HOW EMPTY CAN YOU GET!

M Abdou*, G Sindi, M Syed, DJ Feilds, M Naguib. *Texas Tech Univ HSC Amarillo, Amarillo, TX*

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Case report Although hypokalemia can be transiently induced by intracellular potassium shift, most cases result from gastrointestinal or urinary losses. An estimate of 10 mEq of potassium chloride is reported to increase serum levels by 0.1 mEq/L.

We report a case of severe hypokalemia due to GI losses required massive amounts of potassium replacement to replenish serum potassium levels with no concurrent potassium losses.

58 year-old male with hypertension and chronic kidney disease stage2 presented with fever, non-bloody diarrhea of 6 episodes per day, and weakness for a week. Physical exam was unremarkable besides temperature of 38.8 C and BP 137/65. Leukocyte count was 10.7 K/uL, serum potassium was 2.2 mg/dL, creatinine 2.2 mg/dL. Home medications were Carvedilol, Diltiazem, Clonidine and statin.

Stool was positive for Campylobacter antigen and azithromycin was followed by resolution of diarrhea in 3 days.

Despite aggressive replacement, potassium level remained low for 6 days and finally corrected to 3.6 mg/dL after a total of 640 mmol of potassium chloride (220 mmol intravenously and 420 mmol orally) along with daily regular diet with estimated potassium of 80 mmol/day (total of 420 mmol/diet) in absence of diarrhea. An adequate collection of 24 hour urine potassium content was only 45 mmol.

The patient displayed lower motor neuron weakness in his lower extremities that worsened during his hospital stay. LP and MRI of the brain and spine were unremarkable and his weakness thought to be secondary to his hypokalemia. He started to show some improvement after normalization of potassium level but still required rehab on discharge.

Potassium is primarily an intracellular cation. Studies showed that a decrease in serum potassium concentration of 0.3 mmol/L may reflect a 100 mmol reduction in total body potassium stores. However, in our patient it took about 640 mmol of potassium to raise his level by 1.4 mmol/L, much more than expected, still not taking into account the full daily diet potassium intake. This case suggests the underestimation of the true potassium deficit and how profound the intracellular depletion can be in the setting of severe GI loss. Also, our patient's severe potassium repletion was associated with hypokalemia periodic paralysis that did not resolve until 2 weeks after potassium was replenished.

526 ASSESSMENT OF UTILITY OF URINE SEDIMENT MICROSCOPY IN HEPATORENAL ACUTE KIDNEY INJURY

A Alghamdi*, J Velez. *Ochsner Health Foundation, New Orleans, LA*

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Purpose of study Microscopic examination of the urinary sediment (MicrExUrSed) is a useful diagnostic tool in acute kidney injury (AKI). However, its performance has not been examined in AKI in patients with end-stage liver disease (ESLD). Hepatorenal syndrome type 1 (HRS-1), a type of AKI in ESLD, is difficult to diagnose despite the existence of the International Club of Ascites (ICA) criteria. We hypothesized that MicrExUrSed improves accuracy of diagnosis of acute tubular injury (ATI) in ESLD patients with AKI.

Methods used MicrExUrSed was performed in patients with AKI stage ≥ 2 with or without ESLD over a 6 month period. HRS-1 was defined by the ICA criteria. Urine Na < 20 mEq/L, urine volume < 500 ml, mean arterial pressure < 75 mmHg and serum Na < 135 mEq/L were supportive criteria. Definite HRS-1 was defined as: ICA + supportive criteria were met. Possible HRS was defined as: only ICA criteria were met. No HRS was defined as: ≥ 1 ICA criteria was not met. Urinary cast scores (based on Chawla *et al* and Perazella *et al*) were assigned to each specimen. Chawla scores (CS) 3–4 and Perazella scores (PS) 2–3 were deemed consistent with ATI and not HRS-1.

Summary of results Distribution of casts ($> 25\%$ lpf with ≥ 1 cast) differed between the ESLD (n=35) and non-ESLD (n=44) groups. Hyaline casts (HC), renal tubular epithelial cell casts (RTECC) and waxy casts (WC) were identified more often in ESLD compared to non-ESLD [57 vs 29% ($p < 0.05$); 26 vs 5% ($p < 0.01$) and 20 vs 5% ($p < 0.05$) for HC, RTECC and WC], but not granular casts [43 vs 61%]. In the ESLD group, total bilirubin was significantly higher for those with RTECC [35.9 vs. 6.4 mg/dL ($p < 0.0001$)] suggesting pathogenesis of bile cast tubulopathy. A diagnosis of Definite HRS-1 was assigned to 6 (17%), Possible HRS-1 to 14 (40%) and No HRS-1 to 15 (43%) patients. Addition of MicrExUrSed changed the diagnosis in 11 (31%) patients: Definite HRS-1 was changed to No HRS-1 in 2 and Possible HRS-1 was changed to No HRS-1 in 9, changing the final No HRS-1 count to 26 (74%) and HRS-1 to 9 (26%). Nonetheless, vasoconstrictor therapy to treat HRS-1 was given to 23 (66%) patients.

Conclusions MicrExUrSed can aid in the diagnosis of AKI in ESLD by identifying those with evidence of ATI, i.e., not consistent with HRS-1, with potential implications on the use of vasoconstrictor therapy and/or dialysis decisions.

527 A RARE CASE OF CONTAMINATED COCAINE-INDUCED GLOMERULONEPHRITIS

M Arevalo*, A Ismail, W Vutthikraivut, RM Medrano, C Pena-Hernandez. *Texas Tech University Health Sciences Center, Lubbock, TX*

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Introduction Levamisole has long been used as a cutting agent in cocaine for unknown reasons but presumably to potentiate the stimulant effects of cocaine. It was developed as an anti-helminthic agent and later used in cancer and autoimmune disease therapy due to its strong immunomodulatory effects. It

was withdrawn in 2000 due to serious side effects including agranulocytosis.

Anti-neutrophil cytoplasmic antibody (ANCA) vasculitis has been recently linked to levamisole-contaminated cocaine.

Case summary A 50-year-old man presented to the emergency center complaining of shortness of breath and decreased urine output for 2 days. He snorted cocaine regularly but denied intravenous drug use. Upon admission, creatinine was 10.5 mg/dl. Urinalysis with microscopy showed proteinuria, hematuria, and red blood cell casts. Urine drug screen was positive for cocaine. Both Anti-Myeloperoxidase and anti-Proteinase 3 antibody index was high at 4.3 and 4.6 respectively. Anti-glomerular basement membrane antibody and antinuclear antibody were negative. Antiphospholipid antibodies were positive. Complement C3 was low, and C4 was normal.

Renal ultrasound showed increased echogenicity with normal kidney size, no hydronephrosis, and no stones.

Renal biopsy showed focal (20%) necrotizing crescentic, focal endocapillary proliferative, sclerosing glomerulonephritis and severe interstitial fibrosis and tubular atrophy (60%). Despite therapy with pulse intravenous steroids patient progressed to end-stage renal disease.

Discussion This case highlights the importance of having a high index of suspicion; even in the absence of classic vasculitis signs or other commonly described levamisole side effects, as detection of levamisole in standard urine samples is difficult.

Clinical awareness of severe side effects of levamisole-contaminated cocaine has improved over the past decade, but the full spectrum of kidney disease remains unknown. Pauci-immune glomerulonephritis, as well as membranous nephropathy, have been described.

Current treatment includes cessation of cocaine, supportive care, and intravenous steroid therapy. However, the efficacy of immunosuppression and the optimal regimen remain unclear.

528

VASCULAR ENDOTHELIAL GROWTH FACTOR MODULATION AND RENAL EFFECTS

M Arevalo*, C Pena-Hernandez. *Texas Tech University Health Sciences Center, Lubbock, TX*

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Introduction Angiogenesis is known to be vital in cancer growth. The vascular endothelial growth factor (VEGF) family promotes neovascularization, nutrition, and metastasis; thus VEGF blockers have a key role in cancer therapy. Some experts refer to the manifestations associated with the disruption of renal nutrition as Anti-VEGF Nephropathy.

Case summary A 71-year-old man presented with shortness of breath, nausea, and decreased urine output for two days. History included hypertension, chronic kidney disease stage III, and stage IV renal cell carcinoma treated with right radical nephrectomy and chemotherapy with gemcitabine. He was on sunitinib therapy for at least 3 years before presentation and had been recently started back on gemcitabine.

Initial laboratory showed pancytopenia, elevated creatinine (2.9 mg/dl from a baseline of 1.2 mg/dl), high anion gap metabolic acidosis, hyperbilirubinemia, and transaminitis. A peripheral blood smear showed macrocytosis, burr cells, target cells, and teardrop cells with large platelets. d-dimer was 44,946 ng/mL, lactate dehydrogenase was 961 U/L. ADAMTS13 activity was mildly decreased (30%) with high

inhibitor 0.5 (reference range <0.4). Urine microscopy showed hematuria and muddy brown casts. The fractional excretion of sodium was 2%, and urine eosinophils were negative. He was diagnosed with Thrombotic Microangiopathy (TMA) in addition to combined pre-renal and chemotherapy-induced acute kidney injury. Chemotherapy was held; however his clinical condition quickly deteriorated and family opted for hospice and patient died shortly afterward.

Discussion The podocyte produces and secretes VEGF, which is essential for survival, nutrition, and development of glomerular capillaries, endothelial cells, and mesangium. With endothelium disruption due to VEGF inhibition, there is fibrin deposition, enhanced clotting and ultimately TMA. TMA is considered the most common complication of VEGF blockers. In contrast to nephrotoxicity from medications, VEGF blockers may take up to 3 years to cause renal injury. Discontinuation of VEGF blockers is standard management however prognosis is variable and dependent on timing of diagnosis so clinicians should closely monitor patients while on therapy.

529

LIMITATIONS OF BEDSIDE ULTRASOUND FOR VOLUME DETERMINATION IN CONTINUOUS RENAL REPLACEMENT THERAPY PATIENTS

¹C Boyle*, ¹M Saleem, ¹S Padala, ¹M Hinnant, ^{1,2}JJ White, ^{1,2}S Nahman, ^{1,2}AA Mohammed. ¹Augusta University, Augusta, GA; ²Charlie Norwood VA Medical Center, Augusta, GA

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Purpose of study We have previously shown that bedside ultrasonography (US) may help determine volume status and ultrafiltration goals in ESRD patients. Increased IVC diameter (IVCD) and increased number of lung B-lines (comets) were shown to be useful in gauging fluid removal with dialysis (Mohammed, JASN; Saleem JASN). Septic patients requiring CRRT may have confusing volume assessments on exam. Thus, we theorized that bedside US may help determine UF goals in this setting.

Methods used Ten septic patients requiring CRRT were studied. Following informed consent, a single operator (CB) used a Philips Lumify portable ultrasound device to assess the number of lung comets and IVC diameter (cm) at the initiation of dialysis (T-0) and after 24 hours (T-24) of therapy. Data are expressed as mean $\pm$ SD.

Summary of results Nine patients were on pressors, 5 were ventilated. For all patients, lung comets were only detectable in 2 (T-0; 13 and 9 comets), but the number did decrease at T-24 (6 and 0 comets, respectively). For IVCD measurements, two groups of patients were studied: intubated and ventilated (VENT, n=5) and masked oxygen only (MASK, n=5) during inspiration (I) and expiration (E) at each time point. At T-0, IVCD decreased with E in MASK (1.48 ± 0.4 vs 1.33 ± 0.8 for I vs E, respectively). A similar trend was observed at T-24 (1.21 ± 0.1 vs 1.07 ± 0.7 for I vs E, respectively). In contrast, VENT showed paradoxical increases in IVCD during E at both time points (T-0; 1.64 ± 0.4 vs 1.87 ± 0.5 and T-24; 1.69 ± 0.7 vs 1.92 ± 0.7 for I vs E, respectively).

Conclusions The clinical utility of bedside US is limited in CRRT patients. Not surprisingly, lung comets were uncommon in these septic patients. In contrast, determination of IVCD in MASK patients may offer information on volume status. Increases in IVCD with expiration render the procedure unreliable in ventilated patients on CRRT.

530

A CASE OF PYODERMA GANGRENOSUM MASKING UNDERLYING HENOC-SCHONLEIN PURPURA

JA Carlson*, H Sadiek, JL Chique, R Makary, James IR. University of Florida – Jacksonville, Jacksonville, FL

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Case report Henoch-Schonlein purpura (HSP) is a systemic vasculitis characterized by tissue deposition of IgA containing immune complexes in different organs, resulting in a tetrad of non-thrombocytopenic palpable purpura, arthritis, gastrointestinal (GI) symptoms, and renal involvement; the latter commonly associated with poor prognosis. On histopathology, leukocytoclastic vasculitis (LKV), characterized by neutrophil infiltration, with IgA deposition on immunofluorescence is commonly seen.

A 21 year old woman with CHF and hypothyroidism was admitted for CHF exacerbation. In addition, she developed nausea, a pyoderma gangrenosum (PG) rash, and pink to tea-colored urine. Urinalysis on admission was significant for protein (100 mg/dl), negative leukocyte esterase, and large blood. Her eGFR was >60 ml/min and electrolyte panel was normal. The rash was biopsied and diagnosed as a LKV. Due to active urine sediment and proteinuria, kidney biopsy was performed which proved IgA nephropathy.

Given findings on skin biopsy (LKV), renal biopsy (IgA nephropathy), and GI symptoms, a diagnosis of HSP was made.

Abstract 530 Figure 1 A: Skin biopsy with leukocytoclastic vasculitis; B: HE stain showing increased mesangial matrix, cellularity in glomerulus; C: PAS stain suggestive of early crescent; D: Gomori Trichrome stain showing global and segmental sclerosis; E&F: Electron microscopy consistent with proliferative immune mediated glomerulonephritis

PG is not currently associated with the diagnosis of HSP, and HSP usually occurs in pediatric populations. Likewise, vasculitis or findings consistent with a neutrophilic vascular reaction or LKV are not consistent with the usual diagnostic findings of PG. In this patient, PG was the first manifestation of underlying HSP; in specific cases, PG may be suggestive of underlying HSP.

531

NOCARDIOSIS IN RENAL TRANSPLANT PATIENTS: INCIDENCE AND RISK FACTORS

¹M Gibson*, ^{1,2}S Baer, ¹J Waller, ^{1,2}S Nahman, ¹M Kheda, ¹AA Mohammed, ¹L Young, ¹W Bollag. ¹Augusta University, Augusta, GA; ²Charlie Norwood VA, Augusta, GA

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Purpose of study Nocardiosis is an opportunistic infection commonly occurring within 2 months to 2 years after renal transplant. *Nocardia* is an important cause of mortality in these patients with a subtle clinical presentation manifesting primarily as an unexplained fever. Identifying additional clinical risk factors for *Nocardia* may allow for earlier diagnosis and therapy. To address this question, we queried the United States Renal Data System (USRDS) for the incidence and clinical risk factors for *Nocardia* in post-transplant patients.

Methods used Demographic and clinical risk factors for *Nocardia* in incident kidney transplant patients were identified using ICD-9 codes. Generalized linear models incorporating the number of person years at risk were used to examine risk factors for *Nocardia* and the adjusted relative risks (aRR) and 95% confidence intervals (CI) were determined.

Summary of results Among 1 91 332 renal transplant patients from the USRDS, 630 (0.3%) were diagnosed with *Nocardia*. The following were associated with an increased aRR of infection: age >65 (aRR=2.04, CI 1.65 to 2.52); tacrolimus (aRR=2.14, CI 1.73 to 2.64); corticosteroids (aRR=1.94, CI 1.16 to 3.24); and thymoglobulin (aRR=1.56, CI 1.32 to 1.86). There was a decreased risk of infection with azathioprine (aRR=0.60, CI 0.50 to 0.74), sirolimus (aRR=0.61, CI 0.48 to 0.76), mycophenolate (aRR=0.76, CI 0.61 to 0.94) hepatitis C (aRR=0.56, CI 0.37 to 0.85), diabetes (aRR=0.85, CI 0.73 to 0.99), and tobacco use (aRR=0.75, CI 0.62 to 0.92).

Conclusions Nocardiosis is uncommon following kidney transplant occurring in 0.3% of patients. Older patients and those on tacrolimus, corticosteroids, or thymoglobulin were at higher risk for infection. Diabetes, tobacco use and hepatitis C infection were associated with a decreased risk. These data may allow clinicians to consider Nocardial infection earlier in the assessment of unexplained fever in kidney transplant patients, and thus help to minimize morbidity and improve mortality.

532

UNUSUAL PRESENTATION OF GRANULOMATOSIS WITH POLYANGIITIS

¹K Greer*, ¹N Algu, ^{1,2}I Ashoor. ¹LSU Health Science Center, Metairie, LA; ²LSU HSC and CHNOLA, New Orleans, LA

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Case report Granulomatosis with polyangiitis is a rare pediatric disorder with high morbidity and mortality if diagnosis is delayed. A presentation of granulomatosis with polyangiitis consisting of primarily gastrointestinal and dermatologic symptoms is exceedingly rare, and if not recognized, may lead to delay in diagnosis and treatment.

We employed a retrospective chart review and literature review to explore this particularly uncommon presentation.

Our case is a 14 y.o. male who initially presented with abdominal pain, intractable vomiting with significant weight loss, cola colored urine, and erythematous conjunctiva. Over the next several days, he developed diffuse purpuric rash, bloody stools, hypertension, and arthralgias. Henoch Schonlein Purpura was initially suspected based on the patient's primary presentation with GI symptoms and new onset

purpuric rash. Renal biopsy was obtained due to nephrotic range proteinuria and active urine sediment concerning for nephritis. The biopsy confirmed necrotizing pauci immune glomerulonephritis. Serologic testing revealed a positive C-ANCA titer. He was diagnosed with glomerulonephritis with polyangiitis based on these findings. High dose steroid treatment followed by 4 weekly rituximab infusion successfully induced remission.

This case demonstrates the potential variability in a rare but serious pediatric disease. Granulomatosis with polyangiitis very uncommonly (<40%) presents with GI symptoms (i.e. nausea, vomiting, abdominal pain) and even fewer (<5%) present specifically with bloody stools. A high index of suspicion based on evaluation of asymptomatic renal abnormalities is critical to establish a definitive histological diagnosis and initiate timely therapy to induce remission and limit long term morbidity.

533 INTRACRANIAL ANEURYSM IN A PEDIATRIC PATIENT WITH ARPKD

^{1,2}M Hajirawala, ^{1,2}M Shapiro, ^{1,2}B Casey, ^{1,2}D Aviles. ¹LSUHSCNO, New Orleans, LA; ²Children's Hospital of New Orleans, New Orleans, LA

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Case report Five percent of the U.S. population is estimated to have at least one ICA in which 0.2% rupture each year. On the other hand, a systematic review found an 11.5% prevalence rate in patients with autosomal dominant polycystic kidney disease (ADPKD) and 1.9% of these ICAs rupture every year. This link has not been established in autosomal recessive polycystic kidney disease (ARPKD). We present the fifth known case of intracranial aneurysm in a patient with ARPKD and only the third pediatric case.

A 7-year-old female with autosomal recessive polycystic kidney disease (ARPKD) and hypertension was admitted to pediatric ICU for hypertensive emergency with blood pressure of 200/120 mmHg and headache. CT brain demonstrated severe diffuse global cerebral edema, obstructive hydrocephalus and impending downward central herniation. MRI/MRA confirmed suspicion of subarachnoid hemorrhage and an underlying intracranial aneurysm. An external ventricular drain was placed, the patient was electively intubated, and cerebral angiogram was performed with coil embolization of right A1 saccular aneurysm in the anterior cerebral artery. The rest of her hospital course was significant for frequent fluctuations in blood pressures that were controlled by day 14 of admission with EVD removal by day 15. She did not have any neurological sequelae by the day of discharge.

One of the previously reported cases presented with recurrent headaches and another with sudden onset neurologic symptoms (lethargy, dizziness, and vomiting). The remaining two presented similarly to ours with sudden onset severe headache and were found to have subarachnoid hemorrhages, as in our patient. Following initial presentation, all ICAs were diagnosed with neuroimaging and were subsequently treated with surgical intervention except in the one case where it was contraindicated. Although rare, aneurysms can present in children with ARPKD. Early diagnosis with appropriate neuroimaging should be considered to decrease morbidity and mortality associated with a ruptured aneurism.

534 AN UNUSUAL CASE OF INTERSTITIAL NEPHRITIS IN AIDS

G Kaur*. Emory University, Decatur, GA

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Case report Acute kidney injury (AKI) is common in HIV. We describe a case of HIV-infected female (F) with many possible differential diagnoses of AKI; renal biopsy played an important role in determining the cause. She had granulomatous interstitial nephritis (GIN) due to disseminated mycobacterium avium complex infection (DMACI).

35 year old F with h/o HIV/AIDS (CD4 count of 14, viral load 2.93 log 10 copies/mL) and DMACI presented for vision loss due to CMV retinitis and AKI and was admitted for treatment with IV Foscarnet. Baseline creatinine (CR) was 1.3–1.4 mg/dL which increased to 3.0 mg/dL. Her CR did not improve beyond 2.7 mg/dL after 2 days of IV fluids. Further investigation revealed urinalysis negative for protein with no cellular casts or crystals. Renal ultrasound showed b/l increased echogenicity. Spot urine revealed protein: creatinine ratio of 1.3 mg/g with microalbumin 92 mg. ANA, ANCA, were negative, HBA1c was 3.4%, SPEP was negative, hepatitis B surface antigen was positive, RPR was negative. Renal biopsy was performed on day 4 of admission: interstitial, non-necrotizing granulomatous inflammation; moderate interstitial fibrosis/tubular atrophy, 1/20 glomeruli were sclerotic, non-sclerotic glomeruli were normal on light microscopy, focal areas of glomerular podocyte foot process effacement on ultrastructural analysis. Given the patient had a severe DMACI 1 month prior with MAC bacteremia and MAC associated necrotizing mesenteric lymphadenopathy with chylous ascites, these granulomas were likely progression of the patient's MAC infection, which was the cause of the AKI. She completed 21 days of renally dosed of IV foscarnet and her creatinine on discharge was 2.1 mg/dL. We did not treat her with steroids, since benefit risk ratio was not in favor of treatment and she was getting pathogenetic treatment for MAC. She was asked to follow up in clinic.

GIN is an uncommon histological finding. Most common etiology of this in developing nations is tuberculosis and fungal infections. This renal disease is insidious and may go undetected for several years. Recognition and treatment of underlying cause can lead to improvement in some cases and renal biopsy is key to determine prognosis. Our case represents a complex case with various possible etiologies of AKI where a renal biopsy helped to establish diagnosis and guide therapy.

535 TO DIURESE OR NOT TO DIURESE, THAT IS THE QUESTION

G LaBier*, D Summers, J Doan, LS Engel. LSU Health Sciences Center, New Orleans, LA

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Introduction Minimal Change Disease (MCD) is a common cause of nephrotic syndrome in both adults and children. While its etiology remains largely unknown, MCD generally presents with a number of symptoms including proteinuria, hypoalbuminemia, edema, weight gain, and in some cases acute renal failure.

Case A 33 year old man with a past medical history significant for minimal change disease presented to the Emergency Department with a chief complaint of swelling in his arms and legs for five days with increasing shortness of breath. Upon examination, the patient was found to have significant lower extremity edema extending from his feet to his knees. He had bibasilar crackles on lung auscultation. Laboratory studies were notable for a K of 2.2 mmol/L, magnesium of 1.7 mg/dL, total CK of 29,316 U/L, CRP of 1.50 mg/dL, sedimentation rate of >120 mm/hr, and albumin of 1.8 gm/dL. Blood urea nitrogen and creatinine were found to be 14 mg/dL and 1.54 mg/dL respectively. Baseline creatinine was 1.06 mg/dL. Urinalysis revealed a random total protein of 1060 mg/dL and was negative for significant myoglobinuria. The patient was subsequently admitted to the internal medicine service and underwent diuresis with intravenous furosemide. Additionally, the patient was given both intravenous and oral potassium replacement. His home azathioprine (200 mg daily) and prednisone (20 mg daily) were continued upon admission and during his hospitalization. With diuresis, the patient's total CK level decreased to 2553 U/L over a course of six days. On day 3 of hospitalization, the patient was switched from intravenous furosemide to oral spironolactone. The patient was discharged on spironolactone and potassium chloride tablets.

Discussion Minimal Change Disease is often characterized by sudden onset of edema, hypoalbuminemia, and acute renal failure. Initial treatment consists of volume removal and immunosuppressive therapy with glucocorticoids. In this case, the decision to diurese was complicated by the patient's concurrent diagnosis of rhabdomyolysis and hypokalemia. This case illustrates the complexity in clinical decision making when faced with diagnoses that require opposite treatments.

536

GABAPENTIN TOXICITY IN THE SETTING OF ACUTE RENAL FAILURE SECONDARY TO NON-STEROIDAL ANTI-INFLAMMATORY DRUG USE

JN Leibovitch*, S Knohl. *Upstate Medical University, Syracuse, NY*

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Introduction The exact mechanism of action of gabapentin is unknown. It is believed to act on GABA receptors, and is primarily approved for treatment of partial seizures and neuropathic pain. Adverse effects of gabapentin tend to be neurologic in nature, and include dizziness, confusion, and sedation. These tend to be more prevalent in patients with renal disease, especially when doses are not adjusted to the glomerular filtration rate. Concrete studies, however, have not been performed. Our patient is an example of this infrequently documented phenomenon.

Case report A 46-year-old man with a past medical history of chronic shoulder pain and alcohol abuse presented to the emergency department with altered mentation. His wife noted that the patient had been 'unwell' and began hallucinating hours before presentation. Exam demonstrated a delirious, hypotensive patient with myoclonic jerking movements. Admission labs demonstrated a creatinine of 10.87 mg/dL, sodium of 115 mmol/L, ALT of 739 U/L, AST

of 983 U/L, total bilirubin of 3.6 mg/dL, and a negative blood alcohol level. Nephrology and Toxicology were urgently consulted while the patient was admitted to the intensive care unit where he received isotonic volume expansion and N-acetylcysteine for potential acetaminophen poisoning. The patient's condition improved over the next 48 hours, and he endorsed that 2 weeks prior to admission his shoulder pain worsened such that he binged on alcohol, started taking 600 mg ibuprofen four times daily, and was told to increase his gabapentin to 300 mg twice daily (from 200 mg twice daily). We surmised that patient's recent anorexia combined with heavy NSAID use resulted in pre-renal azotemia resulting in an accumulation of gabapentin leading to acute liver injury and neurocognitive deficits. He was ultimately discharged home with plans for frequent monitoring of electrolytes.

Conclusion Few case reports have documented the neurologic side effects of gabapentin toxicity. Given that it is completely renally excreted, any situation compromising kidney function can result in serious drug-related complications. A case can be made for close monitoring of serum levels of gabapentin in patients susceptible to kidney disease in order to prevent development of gabapentin toxicity.

537

CORRECTION OF DIALYSIS-INDUCED METABOLIC ALKALOSIS WITH SALINE INFUSION

A Mohammedzein*, PP Kyaw, M Naguib. *Texas Tech University Health Science Center, Amarillo, TX*

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Purpose of study Metabolic alkalosis is rare in end-stage renal disease (ESRD) patients on hemodialysis (HD) and carries significant mortality and morbidity. In this study we present an innovative approach to manage metabolic alkalosis in this population.

Methods used An established patient on HD with primary metabolic alkalosis, was infused with 3 L of normal saline solution (NSS) prefilter in addition to utilizing low bicarbonate dialysate (30 mEq/L). Excess fluid was removed through ultrafiltration (UF) with a net negative balance of 1.5 L. Venous blood gases and basic metabolic panels were obtained around the first HD session.

Summary of results Improvement in pH from 7.50 to 7.43 and in total CO₂ from 36 to 30 mEq/L were noted after the treatment, while serum chloride increased from 91 to 101 mmol/L (table 1). Subsequently, we used the same membrane (Optiflux 160 NR[®]) for 2 consecutive daily treatments to remove excess fluid and prevent worsening alkalosis using

Abstract 537 Table 1 Venous blood gases and chloride around first dialysis session

	Before HD	During HD	After HD
pH	7.50	7.47	7.43
pCO ₂	42	48	47
Total CO ₂	36	30	30
Chloride	91	100	101

the same minimal bicarbonate bath, but no further NSS was given. Volume was controlled as needed with UF. The bicarbonate did not drop after the second HD session, suggesting low organic acid production in the interdialytic period. By shortening the duration of HD to 3 hours and improving nutritional intake, we achieved dry weight and the patient was sent home with a total CO₂ of 25 mEq/L.

Conclusions Metabolic alkalosis is rare in patients with ESRD on HD and is often challenging to correct. Prevention should be aimed at improving nutritional status, limiting bicarbonate content in the dialysate, and establishing lab alerts in chronic dialysis clinics for high concentrations of pre-treatment serum bicarbonate. An individualized approach utilizing prefilter isotonic saline infusion with concomitant ultrafiltration during HD is a simple and effective in correcting alkalosis and can be used safely in both inpatient and outpatient settings.

538 COLLAPSING GLOMERULOPATHY: ASSOCIATION WITH SUBSTANCE ABUSE

L Nahapetyan*, N Elzubeir, M Naguib, R Bharadwaj. *Texas Tech Univ HSC Amarillo, Amarillo, TX*

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Background Collapsing glomerulopathy (CG) is a form of focal segmental glomerulosclerosis (FSGS) that has been commonly associated with HIV. Other risk factors are parvovirus B19, hepatitis C, *APOL1* genotype, African American race, SLE, and ANCA vasculitis. This case report highlights the role of substance abuse in the development of CG.

Case A 29-year-old Hispanic male with a 10 year history of methamphetamine and marijuana abuse was admitted for bilateral lower extremity edema, pain, and erythema. He was hypertensive with elevated creatinine (3.3) and BUN (68). Urinalysis showed proteinuria, hematuria and 24 hours urine protein excretion of 17 grams. Urine toxicology was positive for marijuana and amphetamines. Extensive work up was negative for HIV, Hepatitis B and C, ANA, anti-DNAse, ANCA. He had unremarkable complement levels, ASO and urine protein electrophoresis. Kidney biopsy showed CG. Patient was treated with prednisone with some improvement in creatinine to 2.4. He was counseled on refraining from further substance abuse.

Discussion Since CG was described in 1970s as a 'malignant' subvariant of FSGS, the etiology is still not entirely clear with most cases defined as either idiopathic or HIV associated. Although a relationship between heroin and CG has been established, limited studies identified associations with other substances such as amphetamines and synthetic cannabinoids. Recent research indicated high potency of synthetic cannabinoids to bind Type 1 and 2 cannabinoid receptors in kidney as a potential mechanism of kidney injury. Given the high prevalence of substance use in HIV patients, it is probable that the association of HIV with CG is confounded by substance abuse. Additionally, substance abuse patterns have changed since 1970s with methamphetamine and synthetic marijuana becoming among the predominantly used substances in 21st century. Therefore, it is important for primary care physicians to obtain detailed substance abuse history in patients with nephrotic syndrome and to also consider

screening patients who chronically abuse amphetamines and marijuana for proteinuria. Given the ominous prognosis of CG and changing prevalence of substance abuse, further research is needed to identify the association of CG with other abused substances besides heroin.

539 IMMUNE-COMPLEX MEDIATED GLOMERULONEPHRITIS WITH ANCA POSITIVITY

P Nguyen*, B Birkelo, S Nahman, P Fall, JJ White. *Medical College of Georgia at Augusta University, Augusta, GA*

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Case report A 76 year old female was transferred for evaluation of non-oliguric AKI. Her medical history was significant for CKD stage 3 (baseline Cr 1.5), diabetes, coronary artery disease and peripheral arterial disease, with no history of autoimmune disease. Her medications included hydralazine 100 mg TID. Initial work up at our institution was notable for Cr 5.22 mg/dL, urinalysis with >300 protein and TNTC normal-appearing RBCs, proteinuria of 1.2 g/day, and no casts. Labs revealed low C3 (77 mg/dL), positive ANA 1:640, anti-dsDNA, anti-histone, RF, and ANCA (MPO Ab 7.6 (nl <0.4), PR3 Ab 3.5 (nl <0.4)), and SPEP with polyclonal increase in gamma globulins. Hepatitis panel, HIV and cryoglobulins were all negative. Hydralazine was held and she was started on methylprednisolone for likely drug-induced lupus nephritis versus drug-induced ANCA vasculitis. A renal biopsy showed mesangial matrix expansion and mesangial and endocapillary hypercellularity with one crescent on light microscopy, IgM (4+) and C3 (4+) on immunofluorescence, and by EM, mesangial and subendothelial immune complex deposits. The final pathologic diagnosis was of IgM-dominant immune complex mediated focal proliferative glomerulonephritis. Dialysis was initiated during hospitalization, and she remained dialysis-dependent at time of discharge.

Drug-induced lupus (DIL) and ANCA-associated vasculitis (AAV) are documented adverse effects of hydralazine. AAV can cause a necrotizing glomerulonephritis with little or no immune complex deposition. Our patient had serum markers suggestive of hydralazine-associated vasculitis, including MPO-ANCA and anti-dsDNA; however the renal biopsy did not support this diagnosis due to the lack of necrosis and the presence of immune deposits. Renal involvement in DIL is rare; reported cases have shown an immune complex-mediated glomerulonephritis on biopsy. A 1984 case report by Ilhe et al described six cases of hydralazine-induced lupus with renal involvement; of those, all had ANA positivity, half had hypocomplementemia, and 66% had anti-dsDNA. Given our patient's serum and histopathologic findings, she would best fit a diagnosis of DIL.

540 PRIMARY HYPERPARATHYROIDISM WITH HEPATIC ENCEPHALOPATHY: A CLINICAL ENIGMA

S Prakash*, MK Islam, T Naguib. *Texas Tech University Health Sciences Center-Amarillo, Amarillo, TX*

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Case report The diagnosis of hepatic encephalopathy should exclude other metabolic causes. We present a rare case who

presented with recurrent hepatic encephalopathy, persistent hypokalemia, and hypercalcemia due to adenomatous primary hyperparathyroidism.

A 62-year-old female with history of cirrhosis and hepatic encephalopathy due to hepatitis C was admitted with confusion for the last 2 days. She had multiple hepatic encephalopathy episodes that resolved with lactulose and rifaximin therapy and had transjugular intrahepatic portosystemic shunt (TIPS) done for esophageal varices in the past year. She had previous episodes of hypokalemia and hypercalcemia, with previous parathyroid hormone (PTH) level of 82 pg/mL. She had no diarrhea or diuretic use. She had flapping tremors and right upper abdominal tenderness. INR was 1.5, potassium 2.6 mmol/L, calcium 12.6 mg/dL, phosphorus 0.9 mg/dL, CO₂ 24 mmol/L, and PTH 44 ng/dL. Computed tomography (CT) of head without contrast and chest x-ray were negative. CT abdomen/pelvis showed retained stools, cirrhosis and gallstones. TIPS was patent on ultrasound. Urinary 24 hour calcium collection was not done due to disorientation. Hypokalemia improved after medication Zepatier was stopped and spironolactone was added. Ultrasound of parathyroid gland showed hypoechoic 1 cm round mass in the lower inferior thyroid pole that showed increased uptake on nuclear sestamibi scan. Cinacalcet was initiated and she was discharged to outpatient better serum calcium and mentation.

Evaluating persons with cirrhosis and hepatic encephalopathy can be challenging due to other etiologies contributing to mental confusion. This case highlights the importance of identifying other etiologies for confusion in hepatic encephalopathy to arrive at correct management. Primary HPTH with hypercalcemia can complicate hepatic encephalopathy presentation. Management with bisphosphonates and long-term cinacalcet without surgery is appropriate in advanced liver disease.

541

HYPERPROLACTINEMIA AS INITIAL PRESENTATION IN AN ADOLESCENT GIRL WITH CHRONIC KIDNEY DISEASE (CKD)

D Ravindra Reddy*. *Children Hospital of Georgia, Augusta, GA*

10.1136/jim-2018-000974.539

Case report Hyperprolactinemia is uncommon in children, where most cases are due to pituitary adenomas, secondary to medications or hypothyroidism. We report a case of hyperprolactinemia with an uncommon etiology that posed a diagnostic challenge that could have been overcome with systematic investigation.

A 16-year-old female presenting with history of amenorrhea and galactorrhea for 2 years, was initially evaluated by Gynecologist and was noted to have elevated serum prolactin level of 194 ng/mL (Normal <20). Other relevant investigations include normal thyroid functions, MRI Brain with contrast showed no evident pituitary lesions. She was given a trial of oral bromocriptine. She was non-compliant due to poor tolerance.

Her care was transferred to endocrinology, where she underwent further diagnostic evaluation. Her review of symptoms included daily headaches and nausea. No history of decreased urine output, hematuria, facial or extremity edema.

Blood pressure was at 90–95th percentile. Initial investigations were significant for normal serum electrolytes with acidosis, BUN 83 mg/dL and serum creatinine 14 mg/dL suggestive of renal failure with GFR 5.9 mL/min/1.73 m². Subsequent work up shows normocytic normochromic anemia with hemoglobin of 6.7 g/dL, normal iron studies and elevated parathyroid hormone 2,323 pg/mL favoring CKD. No hematuria or proteinuria. Immunological work up was normal. Renal biopsy suggestive of primary tubulointerstitial disease with severe interstitial fibrosis and tubular atrophy. Hemodialysis was initiated for uremia. Prolactin level was down trending with recent prolactin level 39 ng/mL with return of menstrual cycles.

Hyperprolactinemia secondary to CKD is a rare and the prevalence is unknown in children. Altered dopaminergic suppression of pituitary prolactin by uremia, decreased tubular clearance and decreased breakdown of prolactin by kidneys are some of the proposed hypotheses for this occurrence. Prolactin levels normalizes after renal transplant and the response is uncertain with dialysis. Dopaminergic agonists can be beneficial in some cases. This case highlights that a detailed medication history, baseline lab investigations such as a pregnancy test, comprehensive metabolic panel, thyroid panel, and brain imaging are essential in the evaluation of a case with hyperprolactinemia.

542

RENAL BIOPSY IN THE ASSESSMENT OF TMA

¹J Ro*, ¹S Padala, ^{1,2}S Nahman, ¹D Kleven, ^{1,2}AA Mohammed, ¹G Bowers. ¹Augusta University, Augusta, GA; ²Charlie Norwood VA, Augusta, GA

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Case report Diseases manifesting as thrombotic microangiopathy (TMA) may result in serious morbidity and mortality if therapy is delayed. Thus, TMAs presenting as thrombotic thrombocytopenia purpura, atypical hemolytic uremic syndrome (aHUS), or malignant hypertension may share similar presentations, raising the question of empiric treatment with plasma exchange (PEX) and/or eculizumab, with or without the addition of a renal biopsy. This case assessed the addition of a renal biopsy in this situation.

A 34-year-old African American woman with a past history of hypertension, chronic kidney disease and cocaine abuse presented with chest pain and shortness of breath. On admission, the BP was 207/146 and pulse rate was 108. The serum creatinine was 11.9 mg/dL, hemoglobin 7.1 g/L, platelets 62,000/L, haptoglobin 8 mg/dL and LDH 872 U/L. On peripheral smear there were moderate schistocytes. One month earlier, the creatinine was 1.8 and a renal ultrasound showed 8.9 cm and 9.7 cm kidneys. She was admitted to the ICU for advanced AKI and TMA. Empiric therapy with PEX was instituted after sending ADAMST13 levels. After three PEX, there was no improvement in the TMA and ADAMST13 returned at 59%. The diagnosis shifted to include aHUS vs malignant hypertension. The risk/cost to benefit ratios were discussed for renal biopsy vs empiric eculizumab, and a biopsy was pursued.

The renal biopsy showed endothelial swelling with RBC fragments in the arterioles and glomeruli, but no glomerular clots. There was interstitial fibrosis with tubular atrophy and advanced renal scarring. A diagnosis of advanced hypertensive

nephrosclerosis was made, and the TMA was attributed to malignant hypertension. With aggressive therapy, the BP was controlled and the TMA resolved. The patient remained on dialysis.

This case illustrates the difficulty in distinguishing between different etiologies of TMA and the potential diagnostic role of renal biopsy. Empiric eculizumab may be beneficial but is extremely expensive. A renal biopsy may not distinguish between every cause of TMA but may prove useful in narrowing the differential. Further, as seen in this case, it is helpful in determining the extent of chronic and irreversible disease.

543 THIN BASEMENT MEMBRANE DISEASE (TBMD) IN A PEDIATRIC PATIENT WITH CROHN'S DISEASE

¹S Singh*, ²I Yosypiv, ²L Kidd. ¹Tulane University, Metairie, LA; ²Tulane University, New Orleans, LA

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Case report 13 year old male with past medical history of Crohn's disease controlled with mesalamine, presented with 2 episodes of gross hematuria and left flank pain for 2 days. Physical examination revealed absence of edema or skin rashes and normal blood pressure. Laboratory investigations showed plasma creatinine of 0.7 mg/dL, normal electrolytes, albumin, C3/C4 complement levels, CBC and platelet count, urine calcium/creatinine ratio, negative urine protein and 3 + blood with 11–30 RBCs in the urine. Renal ultrasonography demonstrated presence of two kidneys with normal size, structure and echogenicity. Given that patient continued to have hematuria, a renal biopsy was performed and showed segmentally thin glomerular basement membrane with no areas of fragmentation, consistent with TBMD. Genetic testing for COL4A3, COL4A4 and COL4A5 was recommended. Family was informed of benign course of the TBMD. Awareness of the association of Crohn's disease with TBMD will allow early diagnosis and optimal clinical care of patients with Crohn's disease.

544 A SIMPLE, CLEAR-CUT, STRAIGHT FORWARDLY CONFUSING HYPONATREMIA CASE

M Touchy*, DS Rees, J Maldonado, MV Nalajayan, EA Aguilar, S Barry, F Yazdi, S Morse, RB Varelzdis, E Reisin. LSUHSC-NO, New Orleans, LA

10.1136/jim-2018-000974.542

Case description A 51-year-old Hispanic female with past medical history significant for meningioma status post resection/radiation 8 years ago who was admitted for hyponatremia. On presentation, the patient was hypotensive and had a sodium level of 116 mEq/L. It was discovered through further work-up that the patient was also hypothyroid (TSH 7.0 mIU/L and free T4 0.48 ng/dL) and adrenal insufficient (cortisol 5.6). The patient was experiencing increased altered mental status on our evaluation, and we began the patient with a one time 100 mL 3% saline bolus over 15 min followed by hypertonic saline continuous infusion; solu-cortef and levothyroxine were also initiated. 48 hours after the patient's nadir sodium level of 110, her sodium had

corrected to 124 mEq/L. The patient was maintained on a 1.5 liter per day fluid restriction and continued on 1300 mg TID sodium bicarbonate. The patient remained in the hospital another 48 hours, and on discharge, her serum sodium was 135 mEq/L.

Discussion Both the United States (2013) and European (2014) guidelines recommend hypertonic saline bolus for acute or severely symptomatic hyponatremia, regardless of etiology. Because our patient had an acutely altered mental status at a serum Na level of 110 mEq/L, we initiated a hypertonic saline bolus of 100 ml. Hypertonic saline is an effective and potentially life-saving treatment for cerebral edema due to hyponatremia, as the high extracellular sodium concentration immediately removes water from the intracellular space.

545 PHYSICAL ACTIVITY, DISEASE BURDEN AND MORTALITY IN DIALYSIS

JA Williams*, J Waller, S Nahman, S Baer, AA Mohammed, M Kheda, L Young. Augusta University, Augusta, Georgia

10.1136/jim-2018-000974.543

Purpose of study End-stage renal disease (ESRD) is the final stage of chronic kidney disease leading to permanent loss of kidney function and requires dialysis or kidney transplant. An estimated 40% of all ESRD patients die within 5 years of initial dialysis treatment. The impact of physical activity and perceived disease burden on mortality has not been reported in dialysis patients. Therefore, the purpose of this study was to examine the associations between physical activity, disease burden, and 5 year mortality in ESRD patients receiving dialysis.

Methods used This retrospective, secondary study used data from the Comprehensive Dialysis Survey, a subset of the United States Renal Data System. The association of physical activity, perceived disease burden, and other demographic and clinical variables with 5 year mortality was examined using Cox Proportional Hazard models.

Summary of results Among the total studied participants (n=1257), 5 year mortality was approximately 52% (n=648). Significant associations with mortality were found for age, Charlson Comorbidity Index (CCI), race, sex, employment status, and physical activity. Thus, the risk of 5 year mortality decreased with increasing physical activity scores (aHR=0.98, 95% CI: 0.97 to 0.98). Other variables which showed a decreased risk of mortality included increasing BMI (aHR=0.99, 95% CI: 0.97 to 0.99), black race compared to white race (aHR=0.75, 95% CI: 0.62 to 0.92), female sex (aHR=0.82, 95% CI: 0.69 to 0.96), and being employed (aHR=0.62, 95% CI: 0.44 to 0.88). The risk for mortality increased for every one-year increase in age (aHR=1.02, 95% CI: 1.02 to 1.03) and for every 1-unit increase in the CCI (aHR=1.09, 95% CI: 1.06 to 1.12). Perceived disease burden was not significantly associated with 5 year mortality in this population.

Conclusions ESRD patients should be closely examined for physical functioning and specific risk factors before and during hemodialysis. Considering the clinically significant impact of ESRD on physical activity, disability and mortality, additional studies examining the significance of disease burden on mortality are needed.

Correction: *Fitz-Hugh-Curtis Syndrome, an unexpected diagnosis in an early adolescent patient*

N Hidalgo Corral. Fitz-Hugh-Curtis Syndrome, an unexpected diagnosis in an early adolescent patient. *J Investig Med* 2019;67:376. doi: 10.1136/jim-2018-000974.58

The co-author for this abstract is: Dr. Maria Rene Roca Garcia, University of South Alabama College of Medicine, Mobile, AL

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Since the publication of the Abstracts the authors have noticed that Sottile E was misspelled as Sottlie E in 4 publications. They should have been listed as the following:

Morales Hernandez M, Omar M, Hernandez JM, Rivas JR, Sottile E. Brugada Pattern in Severe Hypothyroidism. *Journal of Investigative Medicine*, February 2019, 67 (2). 10.1136/jim-2018-000974.128

Patel H, Bahloul R, Sottile E. Progressive visual Loss due to Ocular Syphilis. *Journal of Investigative Medicine*, February 2019, 67(2). 10.1136/jim-2018-000974.282

Verdecia J, Hernandez JH, Izzo C, Sottile E, Isache C. A side effect of Ceftaroline in MRSA Meningitis Treatment, *Journal of Investigative Medicine*, February 2019, 67 (2). 10.1136/jim-2018-000974.290

Bahloul R, Patel H, Serrano I, Sottile E. An interesting case of Necrotizing Autoimmune Myopathy, *Journal of Investigative Medicine*, February 2019, 67(2). 10.1136/jim-2018-000974.573

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