Clinical Conundrums
# Clinical Conundrums

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Case Submission – Oral Presentations

**Title:** Toe walking in a toddler – how serious can it be?

**Authors:** Linda-Marie Ustaris, D.O., Stony Brook University Hospital
Ratna Basak, M.D., Stony Brook University Hospital

**Case Presentation:** 4 year old healthy ex-preterm girl with history of toe walking presented with right leg pain, unsteady gait and frequent falls worsening over the last 2 months. Initial neurology evaluation revealed normal strength and reflexes in upper and lower extremities, tight Achilles’ tendons with Babinski sign bilaterally. CPK, Brain and lumbar spine MRI were normal. At two week follow-up, she was unable to stand, with decreased strength in her right greater than left lower limb. She had sustained ankle clonus and +3/4 right patellar reflex. EMG showed normal nerve conduction but reduced motor unit potential activation. MRI cervical spine and thoracic spine including brachial plexus found a large intradural and extradural mass arising from the right C7 nerve root with evidence of cord compression and extension into the right brachial plexus. She underwent C6-C7 laminectomy with excision of tumor. Pathology revealed spindle cell tumor with S-100 staining consistent with schwannoma with intrafascicular growth pattern and ganglion cells consistent with entrapped elements.

**Discussion:** Lower extremity weakness in children can be attributed to an array of causes. Her subacute presentation of worsening leg weakness is suggestive of either an infective, inflammatory, neoplastic or neurodegenerative disease. The lack of constitutional symptoms or systemic signs did not support an infective etiology. A normal CPK ruled out myositis. The absence of neural tube defect stigmata made tethered cord unlikely. While muscular dystrophy could present similarly, the presence of upper motor neuron signs ruled it out. The most alarming features of the patient’s presentation were ataxia, hyperreflexia and the Babinski sign. These are suggestive of long tract myelopathic findings, which are common findings in intradural-extramedullary tumors. When considering upper motor neuron pathology, it is important to consider and image multiple segments from the cortex to corticospinal tracts from cervical to sacral segments.

**Conclusion:** The prevalence of toe walking is between 2-12% in children. Idiopathic toe-walking is a diagnosis of exclusion when it persists after the age of 3 years. It has been associated with neurologic deficits including cerebral palsy, global developmental delay and muscular dystrophy. Common intradural tumors are schwannomas and neurofibromas. Schwannomas are slow growing benign tumors that originate from Schwann cells. Our patient’s age at diagnosis is premature for the typical demographics of patients with a Schwannoma who usually present in their third decade of life. To our knowledge she is one of the youngest presentations of a Schwannoma. Early age presentation of multiple schwannomas or neurofibromas may suggest a possible genetic syndrome such as Schwanomatosis or Neurofibromatosis. Close monitoring for recurrence or new tumor formation is recommended.

**Resources:**

**Presented by:** Linda-Marie Ustaris at: Conundrums Session #1 - Spot A, 7/21/2017 9:45 AM
Title: Asthmatic with Respiratory Distress: Analyzing Beyond Asthma  
Authors: Vanessa McFadden, MD, PhD, Medical College of Wisconsin  
Alyssa Stephany, MD, Medical College of Wisconsin  
Case Presentation: 16 year old male presented with respiratory distress for 4 days. Past medical history included obesity, obstructive sleep apnea, mild intermittent asthma (no history of asthma hospitalizations), and sickle cell trait. Patient developed respiratory distress 4 days prior to presentation and was taking albuterol about 6-8 times a day with some relief. Notably, his mom had recently started smoking indoors and he had not used his prescribed CPAP for 2 years. Patient had no recent doctor visits, other than one for abdominal pain diagnosed as constipation. In his doctor’s office he was hypoxic to 78% on room air. He was transported to the emergency room and was started on continuous duoneb. On exam he was tachycardic, tachypnic, hypertensive, saturating in the low 90s on 12 L/min via facemask, obese (BMI of 53), uncomfortable but able to speak in full sentences. Respiratory exam had equal air movement bilaterally, but diminished in the bases without wheezing or rhonchi. Exam was noted to be limited by body habitus. He was treated with bronchodilators, IV magnesium, and steroids.  
Discussion: He tolerated gradual reduction of respiratory support over the next couple days with weaning of bronchodilators from continuous to 4 times a day and weaning of supplemental oxygen to room air. Then he had a sudden onset of shortness of breath and hypoxia with saturation of 63% on room air. Repeat chest x-ray was similar to prior x-rays with persistent patchy bibasilar and perihilar opacities. At this time, the case was re-examined. In retrospect, his history of no significant asthma flares and absence of wheezing on exam, did not fit with an asthma etiology for his respiratory distress, and the differential was broadened. Due to concern for pulmonary embolism a lower extremity doppler was obtained and revealed a right external iliac deep vein thrombosis. Chest CT revealed a saddle embolism of his pulmonary arteries and also demonstrated a mass near his right kidney along with multiple pulmonary nodules and multiple liver masses. Emergent pulmonary embolectomy was done with nodule biopsy obtained. Ultimately diagnosed with renal medullary carcinoma with metastases.  
Conclusion: Not all respiratory distress in an asthma patient is an asthma exacerbation. Identify the signs and symptoms of pulmonary embolism and remember not to limit the differential. Besides the important lesson of keeping a differential broad, this case has an interesting diagnosis as well. Renal medullary carcinoma is a highly aggressive cancer with an extremely poor prognosis. Neither chemotherapy nor radiation have been shown to be efficacious and the mean survival time is less than 1 year. Metastasis at presentation is extremely common. It almost always affects patients with sickle cell trait and median age is 19 to 22 years old with a male predominance. The most common presenting signs and symptoms are weight loss, hematuria and abdominal or flank pain.  
Resources:  
References:  
Presented by: Vanessa McFadden at: Conundrums Session #1 - Spot B, 7/21/2017 10:00 AM  

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Title: It’s All in Her Head  
Authors: Kelly Ferguson, MD, Resident physician in Internal Medicine and Pediatrics  
Ashley Mehlman, MD, ECU Brody School of Medicine, Amber Lehmann, DO, MPH, ECU Brody School of Medicine  
Case Presentation: A 13 yo with refractory perianal Crohn’s disease s/p diverting colostomy and severe mental illness presented to the ED with subacute progressive ataxia, dysarthria, diplopia, drooling, dysphagia and fatigue. She was seen multiple times in the ED and clinics for these complaints. Her neurologist, PCP and even parents suspected her symptoms to be behavioral, as serial exams and basic labs were consistently normal except for +Romberg. Symptoms progressed, prompting return to the ED. She again had a +Romberg and CT head showed symmetric basal ganglia
abnormalities with minimal local mass effect. She was admitted for further work up, with initial differential of ADEM, vasculitis, or infection. MRI brain showed symmetric edema within the corpus callosum, basal ganglia, mammillary bodies and along the midbrain, concerning for Wernicke’s encephalopathy vs metronidazole-induced encephalopathy. A thiamine level was normal. Metronidazole was a daily med for over 6 months as treatment for her Crohn’s. Once discontinued, the patient’s symptoms and MRI findings resolved 6 weeks later.

Discussion: Metronidazole is usually a safe and effective drug commonly used to treat various infections. Although rare, neurotoxicity, is becoming a more well-known adverse effect. Metronidazole-induced encephalopathy (or MIE) is a unique reaction that has been described to occur at variable doses and treatment durations.1-4 Symptoms reported in the literature most commonly include dysarthria, gait disturbance, and dysmetria, all consistent with cerebellar dysfunction. MRI findings show bilateral symmetric lesions of the cerebellar dentate nuclei, midbrain, dorsal pons, dorsal medulla and corpus callosum, with involvement of the cerebellar dentate nuclei being most characteristic.4 Previous studies have suggested various mechanisms involved in the development of MIE; however, the pathophysiology is not well understood.4,5 Treatment of MIE is immediate drug cessation and the prognosis is generally good, with most cases being reversible. Repeat MRI usually shows improvement or resolution within weeks of discontinuation of the drug, with rare cases of irreversible neurotoxicity reported. 6,3,7

Conclusion: We were fortunate to find a treatable, reversible diagnosis for this teen. A growing number of studies suggest that patients with mental illness experience disparate care for physical disorders due to “diagnostic overshadowing” – the misattribution of physical symptoms to mental illness.8 This bias can delay diagnosis and treatment, which may lead to premature morbidity and mortality. Our patient’s mental illness clouded her caretakers (both professional and parental) judgement and potentially delayed obtaining diagnostic imaging for her complaints. She was also doubly unfortunate to have a rare disease that has yet to even be reported in a pediatric patient. Luckily, there was no permanent damage for our patient, and once metronidazole was discontinued her symptoms completely resolved, as did MRI abnormalities. With all the layers of obscurity, it is clear to see how a rare diagnosis can be missed. It is important for the pediatric hospitalist to avoid diagnostic overshadowing, as well as recognize potential for a rare disease beyond their common knowledge base.

Resources:
References

Presented by Kelly Ferguson at: Conundrums Session #1 - Spot C, 7/21/2017 10:15 AM
A “Bitter” Complaint Leading to an Unusual Diagnosis

Danita Hahn, MD, Medical College of Wisconsin

Case Presentation: A 2-month-old previously healthy term male presented with acute onset of fever and rash. He was ill-appearing and in respiratory distress; rash was characterized by firm, edematous, tender papules with translucent yellow centers and overlying crusting located on the face and extremities. A full septic workup was performed and revealed WBC 20 K/uL, platelet count 820 K/uL, and C reactive protein of 21 mg/dL though was otherwise negative. Despite broad spectrum antibiotics and acyclovir, he developed new skin lesions and remained febrile. Infectious workup, including Herpes simplex virus testing, was negative. Skin biopsy was performed; pathology showed a dermal neutrophilic infiltrate consistent with acute febrile neutrophilic dermatosis (Sweet syndrome). Treatment with systemic corticosteroids was initiated with rapid clinical improvement. In the weeks following discharge, he had complete resolution of symptoms and his steroids were tapered. Etiology in this case was presumed to be a viral illness; no other underlying process was uncovered after extensive workup.

Discussion: Sweet syndrome is characterized by sudden appearance of tender skin lesions accompanied by fevers, leukocytosis, and elevated inflammatory markers. It usually presents in adults but can occur in children, though rarely in infancy. The pathogenesis of Sweet syndrome is unknown. However, viral trigger, usually respiratory or gastrointestinal viruses, is felt to be the most common etiology. It has also been associated with malignancy, autoimmunity, or immunodeficiency; additionally, drug-induced Sweet syndrome has been reported. Biopsy is required for definitive diagnosis, revealing edema and neutrophilic infiltration of the dermis. Systemic corticosteroids are considered the first-line therapy for Sweet syndrome in children and usually leads to swift improvement. Alternative therapies include colchicine, dapsone, potassium iodide, or intravenous immunoglobulin. Recurrences are common as steroids are tapered, occurring in nearly half of pediatric patients, especially when associated with malignancy.

Conclusion: Pediatric hospitalists often encounter children with fever and rash, and the differential diagnosis for this set of symptoms is broad. While rare in the pediatric population, Sweet syndrome should be considered in patients with clinical courses of persistent fevers and atypical rash with an otherwise negative workup. Skin biopsy should be sought for definitive histologic diagnosis, which allows for appropriate treatment and workup of underlying pathology.

References
Title: Losing Sight
Authors: Whitney Rolling, DO, Sanford Children's Hospital
Carl Galloway, MD, Sanford Children's Hospital, Elizabeth Peck, MD, Sanford Children's Hospital, Fernando Bula Rudas, MD, Sanford Children's Hospital

Case Presentation: A 16 month old previously healthy female was admitted to the pediatric hospitalist service with concerns for gross motor regression, poor visual tracking, and peripheral eosinophilia. She originally presented to hematology after referral for eosinophilia and the inability to walk for one month duration. Her symptoms prompted admission for expedited evaluation. Brain MRI had numerous small enhancing lesions, and lumbar puncture revealed 49% eosinophilic meningitis. Funduscopic exam showed right optic nerve granuloma and hyperpigmentation. Her findings and history of living on a farm raised suspicion for parasitic infection. Infectious Disease was consulted, and they discussed testing and treatment with the CDC. Differential diagnosis included parasitic, bacterial, fungal, and viral etiologies. Blood and CSF antibodies were positive for Baylisascaris procyonis, and she was diagnosed with ocular and neural larva migrans. She received a 20 day course of albendazole and corticosteroids, initiated in the hospital for neurologic monitoring, without further complications.

Discussion: Baylisascaris procyonis, or raccoon roundworm, is a rare infection contracted from contact with raccoon feces, potentially causing permanent neurologic impairment or even death. A recent MMWR report from the CDC described 7 human cases since May 2013, including 4 children with ocular or neural larva migrans. B. procyonis is generally an asymptomatic infection in raccoons found throughout North America. Eggs are excreted in feces and can cause infection when ingested. Once ingested, larvae can migrate to eyes, brain, and other organs. With eye involvement, funduscopic exam can visualize larvae. Neural larva migrans can show eosinophilic meningitis (with or without peripheral eosinophilia) and deep white matter changes on MRI. Serologic testing is available through the CDC and detects immunoglobulin G antibodies to B. procyonis. The recommended treatment includes albendazole with systemic corticosteroids to help reduce the subsequent inflammatory reaction, which can include seizures in neural larva migrans. In spite of treatment, permanent neurologic deficits can occur.

Conclusion: Baylisascaris procyonis, or raccoon roundworm, is a rare and potentially severe infection that can cause ocular and neural larva migrans. Due to the parasite’s transmission, young children are at increased risk of exposure. A high degree of suspicion is needed for diagnosis, and should be considered in cases of eosinophilic meningitis or exposure to raccoons or raccoon feces. Prompt treatment with albendazole and corticosteroids is recommended.

Resources: N/A

Presented by: Whitney Rolling at: Conundrums Session #1 - Spot E, 7/21/2017 10:45 AM

Title: A rare cause of chest pain in a healthy teenager: HSV Esophagitis
Authors: Benjamin Mouser, M.D., UT Houston McGovern Medical School
Ann Marshburn, M.D., UT Houston McGovern Medical School, Monisha Shah, MD, UT Health Sciences Center Houston, Adil Solaiman, MD, UT Health Science Center Houston, Baraa Alabd Alrazzak, MD, UT Health Sciences Center

Case Presentation: A 17-year-old male with no past medical history presented to the emergency department with 2 days of fevers and severe, sharp chest pain. Extensive workup including complete blood count (CBC), complete metabolic profile (CMP), D-dimer, cardiac enzymes, chest x-ray, CT angiography, and electrocardiogram was normal except white blood cell (WBC) count was 12.9 x 10^3/µL. Upon further review, the patient’s pain was exacerbated by eating and drinking. Exam was only significant for sinus tachycardia and mild pain on epigastric palpation. Upper GI series was normal. He was started on IV pantoprazole, PO sucralfate, and IV hydromorphone which provided minimal relief. Patient ultimately underwent esophagogastroduodenoscopy (EGD) with biopsies, which showed severe pan-esophagitis (Figure 1). Biopsy results were positive for herpes simplex virus (HSV) on staining. Patient’s girlfriend reported a history of frequent cold sores. HIV testing was negative. Patient received IV acyclovir which provided immediate relief, and he was discharged home to complete 7 days of oral valacyclovir.
Discussion: HSV as a cause of esophagitis has been extensively described in immunocompromised hosts in whom this condition can be fatal (1, 2). While rare in immunocompetent patients, there are now a handful of case reports of severe HSV esophagitis in normal healthy patients (1-10). A review published in 2000 by Ramanathan et al described 38 healthy patients, both adult and pediatric, who were diagnosed with the condition. There was a 3:1 male predominance overall, which increased to 90% in the pediatric population. The typical patient was a young (less than 40 years old in 78% and less than 18 years old in 24% of cases), healthy male presenting with acute odynophagia, dysphagia, or heartburn with or without prodromal symptoms (fever, pharyngitis, respiratory symptoms) or oral lesions (1).

As was the case with our patient, patients with esophagitis frequently present with retrosternal “chest pain” (1, 7, 9). Esophagitis should be considered for all patients presenting with the triad of chest pain, odynophagia, and fever, as early recognition can prevent broad cardiopulmonary workups (10).

Conclusion: HSV esophagitis should be kept in the differential diagnosis for patients presenting with acute onset of fever, heartburn, retrosternal “chest pain”, odynophagia or dysphagia, though it does not uniformly present with classic oral lesions like gingivostomatitis or coupled to a known exposure to a HSV-positive contact. Prior to initiation of broad and potentially unnecessary cardiac workups in the emergency department, healthy adolescents presenting with “chest pain” should be screened for symptoms of esophagitis. While common in the immunocompromised patient, HSV esophagitis is now increasingly being described in immunocompetent patients as well. When this diagnosis is suspected or confirmed, it is important to evaluate immune function as, in addition to being more common, this condition also has a significantly more severe course of illness in immunocompromised patients. Prompt diagnosis and treatment with antivirals such as acyclovir or valacyclovir typically results in complete resolution in otherwise healthy patients with HSV esophagitis.

Resources:

Figure 1. Esophagastroduodenoscopy images showing severe pan-esophagitis.
Case Presentation: A 17 year-old female with history of anxiety and depression presented with vomiting and weight loss associated with fatigue, constipation, abdominal pain and decreased eating for a month. Last month she was hospitalized for vomiting and diagnosed with cannabinoid hyperemesis syndrome due to her significant marijuana use and relief with hot showers. Both she and her mother denied any restrictive eating behavior or intentional weight loss. She had discoloration of her lips and tongue she attributed to marijuana rolling paper. She lost 12lbs in the previous 8 weeks. She had a normal outpatient CBC, LFTs, BMP, lipase and KUB. Her psychiatrist was concerned she had an eating disorder due to persistent weight loss despite claiming to eat extensively at home. She was orthostatic by HR and BP on admission with a BMI of 17. She was hyponatremic to 130 with a bicarbonate of 17 and a mild respiratory alkalosis on VBG, but otherwise normal electrolytes. She was admitted to the eating disorder team and placed on a restrictive eating protocol with advancement of calories daily.

Discussion: She had daily refeeding labs checked as per protocol. Patient reported unchanged nausea during hospitalization and intermittent dizziness with standing. On hospital day 4 she was persistently tachycardic with retching, agitation and hyponatremia to 129, hyperkalemia to 5.9 and hypercalcemia to 11.1. Patient also had a syncopal event following emesis with significant abdominal pain and feeling of weakness. EKG, echo and repeat KUB were normal. Endocrine was consulted. The ACTH was 3156 pg/ml (9-57), PM cortisol was 1.3 ug/dl (3.0-16) in the setting of vomiting and tachycardia and AM cortisol was 49.3 ug/dl (5.5-20) after 12 hours of stress dose steroids. Prolactin and TSH were normal. Patient revealed to the endocrine team significant salt craving and that prior to admission she had been eating ~5 pickles and a few bags of chips per day. Stress dose steroids were initiated with improvement in her energy and nausea. Patient was diagnosed with primary adrenal insufficiency (Addison’s disease) and discharged with hydrocortisone and fludicortisone maintenance medication.

Conclusion: Her initial normal labs were likely due to her compensation at home with increased salt intake. This compensatory mechanism was restricted when she was placed on the restrictive eating protocol and her diet was carefully regulated. She subsequently had worsening of her symptoms and electrolyte abnormalities. Her hyperpigmentation on presentation was likely the initial clue to her Addison’s disease.
Addison’s disease is caused by impaired adrenal function of destruction of the adrenal cortex. Due to the often vague presentation of fatigue, nausea, abdominal pain and vomiting, less than 30% of women and 50% of men are diagnosed within the first 6 months of symptom onset. Patients are most commonly given either a psychiatric or gastrointestinal diagnosis initially as the biochemical signs of hyponatremia, hyperkalemia, hypoglycemia and ketonemia may not develop until later. This patient’s repeated refusal to admit eating disordered behavior in addition to her hyperpigmentation and persistent vomiting upon admission to the hospital should raise the suspicion for alternative etiologies.

Resources:

Presented by Danielle Halpern at: Super Conundrums - Spot B, 7/21/2017 3:15 PM

Title: Mind the Marrow: A Case of Recurrent Bone Pain
Authors: Martha Elster, MD, Kaiser Permanente Medical Center Oakland
Julia Lam, MD, Kaiser Permanente Medical Center Oakland

Case Presentation: 9 year old male admitted with 4 days of left knee pain and refusal to walk in the absence of trauma or fever. Past history of normocytic anemia, thrombocytopenia, and splenomegaly of unclear etiology despite extensive workup. Multiple clinic visits and two prior hospitalizations for right knee pain with no identifiable source including bone biopsy. On exam, left knee swollen with posterior fossa tenderness, limited ROM, and refusal to walk. Labs with mild anemia (Hgb 11.4), thrombocytopenia (plt 91k), elevated ESR 88 and CRP 6.3. X-ray and US normal. MRI with subperiosteal fluid collection of the femur concerning for hematoma or abscess. I&D drained a complex pus collection with negative gram stain and culture. Started on clindamycin and ceftriaxone for presumed culture negative osteomyelitis. Given bone pain and cytopenias, bone marrow aspirate obtained and revealed clusters of gray-blue macrophages with expanded cytoplasm concerning for lysosomal storage disease. Later testing revealed a low beta-glucosidase level and GBA gene sequence variants consistent with Gaucher disease.

Discussion: Gaucher disease is a rare autosomal recessive lysosomal storage disease caused by a lack of beta-glucosidase, also known as beta-glucocerebrosidase. Inadequate beta-glucocerebrosidase activity interferes in the sphingolipid catabolism pathway, preventing breakdown of glucocerebroside within scavenger cells, predominantly macrophages. Accumulation of this substrate in the cytoplasm causes a change in cell morphology, producing “Gaucher cells,” which is a hallmark of the disease but not pathognomonic (2). Deposition of these cells causes organ damage, manifesting in the liver, spleen, hematologic, and skeletal systems - thus causing the classic presentation of hepatosplenomegaly, anemia, thrombocytopenia, and acute episodes of bone pain or bone “crises.” Bone crises occur in 10-32% of patients with Gaucher disease and can be nearly indistinguishable from acute osteomyelitis as they often include swelling, tenderness, fever, and tachycardia. However, unlike osteomyelitis, acute bone crises of Gaucher disease typically resolve with supportive care alone (3).

Conclusion: Gaucher disease should be included in the differential diagnosis of a child presenting with multiple episodes of acute bone pain, particularly in the setting of cytopenias and/or hepatosplenomegaly. Acute bone crises can be mistaken for acute osteomyelitis and thus early recognition is prudent to potentially prevent unnecessary interventions and prolonged antibiotic treatment. Enzyme replacement therapy has been shown to reverse and prevent systemic manifestations of Gaucher disease (1,4). Therefore, prompt identification and initiation of therapy is important for minimizing physical and emotional consequences of delayed diagnosis.

Resources:
**Title:** The “gold” standard? A 4-year-old boy with somnolence and focal weakness

**Authors:** Eric Zwemer, MD, University of North Carolina School of Medicine
Alexandra Florence, MD, University of North Carolina Hospitals

**Case Presentation:** A 4-year-old previously healthy male presented with headache, abdominal pain, unresponsiveness, left-sided weakness, and right eye deviation. Vital signs were T 37.1°C, BP 126/80, and HR 61. Exam showed somnolence, rightward eye deviation, left-sided hyperreflexia, and a benign abdomen. Brain MRI demonstrated multifocal ischemic infarcts and leptomeningeal enhancement. CSF revealed WBC 66 (52L/46N), protein 41, and glucose <20. Ceftriaxone, vancomycin, and fluconazole were empirically started; blood, urine, and CSF cultures remained negative. MRA, echo, HIV, HSV, hypercoagulability labs, and autoimmune labs were unrevealing. Further history revealed patient had lived in Tanzania for first year of life. PPD, Quantiferon gold testing, and CSF TB PCR were negative. After clinical decompensation and persistent fevers, repeat MRI showed new infarcts and hydrocephalus; doxycycline, steroids, IVIG, and RIPE therapy for TB were empirically initiated. Brain biopsy demonstrated caseating granulomas with rare AFB+ organisms. CSF AFB cultures later grew M. Tuberculosis Complex.

**Discussion:** Children represent about 5% of TB diagnoses in the United States, with the majority being young children age 1 to 4 with foreign-born parents or prior residence outside the U.S. Extrapulmonary TB most commonly presents as CNS involvement or lymphadenopathy. CSF analysis may reveal low glucose, elevated protein, and pleocytosis, often evolving from a neutrophil predominance early to a lymphocytic later. Common complications of TB meningitis in children include ischemic stroke, hyponatremia, and hydrocephalus, all of which our patient developed. In particular, TB meningitis classically presents with deep, basilar infarcts and meningeal enhancement. Although more sensitive when used in combination, PPD and Quantiferon testing rely on a functioning immune system, and overwhelming TB infection can cause false negative results. Many cases of TB in children less than 5 years old are never microbiologically confirmed. Newer molecular tests, such as the PCR assay used for our patient, are subject to sampling error and still have limited sensitivity in children.

**Conclusion:** The constellation of abdominal pain, fever, CNS infarcts, hypoglycorrachia, and hyponatremia should raise suspicion for TB meningitis. Prompt recognition and initiation of anti-tuberculosis treatment is crucial to minimize neurologic morbidity and mortality. This patient’s CSF AFB culture was eventually confirmed as Mycobacterium Tuberculosis by the CDC, and genetic analysis revealed the most likely origin was Africa. This case demonstrates the importance of obtaining a thorough exposure and birth history in children, especially when critically ill and not improving on current therapies. One should never be falsely reassured by negative tests, and it is important to initiate empiric anti-tuberculosis therapy and corticosteroids if clinical suspicion for TB meningitis is high. Our patient was ultimately transferred to inpatient rehabilitation and is currently completing directly observed therapy with no new infarcts and resolution of leptomeningeal enhancement on most recent MRI.

**Resources:**
Brain MRI on Initial Presentation: Axial DWI image demonstrating multiple infarcts in basal ganglia and temporal lobe.

Brain MRA: Sagittal MPR image demonstrating extensive basilar and leptomeningeal enhancement.

Brain MRI before brain biopsy: Axial TRACE weighted image demonstrating progressive and new infarcts with progressive ventriculomegaly.
Is this Effusion just due to Bad Luck with Community-Acquired Pneumonia?

**Title:** Is this Effusion just due to Bad Luck with Community-Acquired Pneumonia?

**Authors:** Lauren Bouton, MD, UTSouthwestern Medical Center  
Kathryn King, MD, UTSouthwestern Medical Center

**Case Presentation:** Our patient (Pt) is an 8-year-old, previously healthy, Hispanic female admitted with 2-3 weeks weight loss, decreased energy, and abdominal pain followed by 5 days of a non-productive cough. No fever, chest pain, shortness of breath, emesis, diarrhea or no sick contacts. Pt immigrated from Mexico 1.5 years ago. Pt weight <3rd%. CT chest showed a large left-sided pleural effusion (Image 1). WBC 11.7, without a left shift, amylase 264, and lipase 1944. Pt had a left chest tube placed, and started on IV ceftriaxone. A work-up for TB, brucella, strongyloides, histoplasmosis, hepatitis B/C, HIV, aspergillosis and echinococcus was negative. A sweat chloride test was indeterminate at 38 mmol/L. CFTR gene study sent and fecal elastase 46. The pt had a persistently draining chest tube and a dilated pancreatic duct/head with a pancreaticopleural fistula on imaging. A MRCP showed signs consistent with chronic pancreatitis. There was fluid tracking up into the chest cavity (Image 2). The CFTR studies were positive with 2 cystic fibrosis (CF) gene mutations: delta-F508, and F508del.

**Discussion:** Pancreaticopleural fistulae are a rare complication of chronic pancreatitis, which in itself is rare in the pediatric population (3). Obstruction and genetic mutations such as CFTR, PRSS1, and SPINK1 are often found as the underlying etiology of pediatric chronic pancreatitis (2). De Boeck et al published a study looking at 3,306 patients with CF and 1.84% had a history of pancreatitis. In their studied population, it was more common in older children with a mean age of 19.9 years (1). Our patient is an example of an atypical presentation of CF in that she is 8 years old with no known history of pancreatitis, recurrent infections, sinusitis, chronic cough, greasy stools or delayed passage of
meconium. She is also pancreatic insufficient. In patients with CF, pancreatitis is more common in those that are pancreatic sufficient compared to those that are pancreatic insufficient (1).

**Conclusion:** Our case represents an unusual presentation of a common condition with the clues of failure to thrive in the setting of pancreatitis leading to the diagnosis. This case reminds us that CF should be on our differential in older patients with low weight and gastrointestinal symptoms even without a history of respiratory issues, and to expand our differential in a pleural effusion without improvement after drainage.

**Resources:**


**Presented by** Lauren Bouton **at:** Super Conundrums - Spot E, 7/21/2017 4:00 PM

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**Title:** It's Not Just All In Your Head

**Authors:** Jeremy Yardley, MD, Eastern Maine Medical Center
George Payne, MD, Eastern Maine Medical Center, Jonathan Wood, MD, Eastern Maine Medical Center, Nathalie Kremer, MD, Eastern Maine Medical Center, Lindsay Williams, MD, Dahl-Chase Diagnostic Services

**Case Presentation:** A 9 year old male with a history of migraines for 3-4 years presented with 3-4 days of worsening headaches and several episodes of emesis. In the ER, he was hypertensive with a blood pressure of 212/145. Brain MRI showed obstructive hydrocephalus and cerebellitis. Labwork was remarkable for a serum sodium of 133 and potassium of 2.4. Exam was remarkable for bilateral blurring of the optic disc margins. Neurosurgery advised VP shunt placement. Urine electrolytes showed elevated potassium losses, raising concern for hyperaldosteronism. This then raised the possibility that the hydrocephalus was caused by hypertension (Posterior Reversible Encephalopathy Syndrome) rather than the hydrocephalus causing hypertension, averting potentially unnecessary Neurosurgical intervention. Further
Endocrine workup included aldosterone, renin, thyroid/parathyroid, and pheochromocytoma testing. Aldosterone returned very elevated. Ultrasound and then MRI of the abdomen were obtained, revealing a left renal tumor. The renin level subsequently returned very elevated, leading to the diagnosis of Reninoma.

**Discussion:** A Reninoma, or Renal Juxtaglomerular Cell Apparatus Tumor, is a very rare tumor, with about 90 reported cases in the literature, and even fewer in the pediatric population (fewer than 20, with only about 1/3 of these being male). Removal of these tumors is “typically” curative, though with such a limited set of reported cases, it was difficult to make the decision for surgical intervention, and required discussions with experienced surgeons across the country, most of whom had never seen a similar case. Even within the small subset of these reported cases, this case is unique in that there was such impressive hydrocephalus (as a result of the significantly elevated blood pressure causing PRES, seen on imaging as cerebellitis, which is a very atypical location for PRES as it is more typically parieto-occipital). The other unique aspect of this case is the hyponatremia. With elevated renin, one would expect HYPERnatremia. In discussion with Neurology and Endocrinology, it was concluded that this was likely secondary to SIADH as a result of increased intracranial pressure.

**Conclusion:** Malignant hypertension as a presentation of Reninoma in the pediatric population is exceedingly rare, so it would be unreasonable to state that a take away point from this case should be to think about this particular tumor with significantly elevated blood pressures. In terms of management of malignant hypertension, however, an important point to draw from this case, is that normalization of the blood pressure immediately is potentially very dangerous, as there is likely a need for at least some degree of elevated systemic blood pressure to maintain cerebral perfusing pressure. Another point from this case that can be extrapolated to other cases is that in the setting of hypertension with associated hydrocephalus, it is important to think about hypertension as both the cause and effect of hydrocephalus. The patient in this case nearly had unnecessary neurosurgery due to lack of recognizing this concept.

**Resources:** N/A

Presented by Jeremy Yardley at: Conundrums Session #2 - Spot A, 7/22/2017 9:45 AM

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**Title:** How Atypical?

**Authors:** Ratna Basak, M.D., Stony Brook University Hospital
Catlin Keane, DO, Stony Brook University Hospital, Robert Woroniecki, MD, Stony Brook University Hospital

**Case Presentation:** A 17 year old morbidly obese African American (AA) female presented with dyspnea, tachycardia and history of intermittent fever, myalgia, skin rash, vomiting and diarrhea for 1 month. She had hypertension, pitting edema in the legs, decreased breath sounds at right lung base and oliguria. Labs: WBC 15.5k/μL (10% eos), Hb 5gm/dL, platelets (PLT) 14k/μL, retic 7.1%, 3+ schistiocytes, LDH 3330 IU/L, low haptoglobin, elevated CRP and serum creatinine (SCR=3mg/dL), low albumin, elevated transaminases and triglyceride levels. Her ferritin was 5269 ng/mL. ANA, dsDNA, other rheumatologic markers were negative, and C3/C4 were normal. PTT/INR were mildly elevated with normal fibrinogen level. Her urine had large amount of blood and protein, 4 RBC, 17 WBC/hpf. Renal USG showed enlarged kidneys with increased echogenicity. She had enlarged cardiac silhouette with small bilateral pleural effusions on CXR and her ECHO showed a large pericardial effusion. Urine and blood cultures were negative and stool PCR/cultures were negative for E Coli 0157.

**Discussion:** Her differential diagnosis included: Hemolytic Uremic Syndrome (HUS); Hemophagocytic Lymphohistiocytosis (HLH); Thrombotic Thrombocytopenia (TTP); microangiopathic hemolytic anemia. She received PLT and RBC transfusions, plasmapheresis, hemodialysis and pericardiocentesis. With ADAMS13 activity at 56%, TTP was ruled out, as was HLH with absence of other diagnostic criteria and a diagnosis of atypical HUS (aHUS) was presumed. Her renal biopsy showed thrombotic microangiopathy with interstitial nephritis (eosinophils). She received weekly Eculizumab infusions and showed clinical and renal function improvement after the 3rd infusion. Hemodialysis was discontinued, and SCR went down to 1.17 (most recent 0.88) mg/dL. She was sent home on oral steroids (for interstitial nephritis/serosistis) and bi-weekly Eculizumab. Her pericardial and pleural effusions resolved and her steroids were discontinued.

**Conclusion:** We report an atypical aHUS associated with interstitial nephritis, serositis and unusually high serum ferritin. Atypical HUS is now a term preferentially reserved for HUS without a coexisting disease but with a dysregulation of complement cascade due to genetic background. Only 30% of patient with aHUS have low C3 levels. Decreased complement factor H or I are observed in 30-50% of patients with mutated CFH or CF1. Our patient’s genetic testing was
positive for large CRHR1-CFHR3 homozygous deletion (Factor H autoantibody test was negative), two silent variants of unknown significance in exon 2 of CFI and in exon 29 of C3, and heterozygous missense variant in exon 2 of DGKE (more common in AA, aHUS-associated homozygous mutations in DGKE maybe enriched in pediatric patient and may not respond to Eculizumab). Ezulizumab should be instituted early to avoid progression to ESRD, and in subjects with aHUS with genetic mutations this can be both lifesaving and lifelong treatment.

Resources: N/A
Presented by Caitlin Keane at: Conundrums Session #2 - Spot B, 7/22/2017 10:00 AM

Title: Avoiding "Rash" Decisions in the Work up of a Patient with Refusal to Walk
Authors: Allison Ashford, MD, University of Nebraska Medical Center
Cassandra Wehling, Med-Peds Resident, University of Nebraska Medical Center

Case Presentation: An 8-year-old boy presented with 3-months of left knee pain. He refused to bear weight and had developed a rash on his abdomen and legs. There was no trauma, fever, or chills. He was treated for strep infection 1 month prior. There was family history of sarcoidosis. On exam, he had a temperature of 38.7, and a painful left knee worse with hip flexion and knee extension. There was no swelling or erythema of the knee, and his other joints were normal. There were multiple non-tender erythematous nodules on the lower legs. Significant labs included ESR 40, CRP 7.5, WBC 16, Hg 12, and calcium 9.6. MRI of lower extremities was normal except for multiple ill-defined soft tissue lesions. Chest x-ray, done after the differential was expanded, was significant for hilar adenopathy. CT of the chest showed peri-lymphatic lung nodes. Normal labs included complement, ASO titers, ferritin, ACE, ANCA, ANA, histoplasmosis antigen, quantiferon gold, toxoplasmosis antibodies, and bartonella antibodies. His Vitamin D 1,25 was high. Biopsy of a lesion on his leg was consistent with erythema nodosum.

Discussion: The patient was given a diagnosis of presumptive sarcoidosis. This was based on his hilar lymphadenopathy, erythema nodosum, high vitamin D 1,25, and exclusion of more common causes of his symptoms. The patient and was discharged on a course of steroids. On follow up he could walk without pain and the rash was resolved. Sarcoidosis is an uncommon diagnosis in children with an incidence as low as 0.22 in 100,000. Diagnosis is made with clinical signs and symptoms indicative of sarcoid and tissue biopsy demonstrating non-caseating granulomas. Signs and symptoms include hilar adenopathy, peripheral lymphadenopathy, uveitis, erythema nodosum, joint effusions, proteinuria, malaise, fever, and elevated 1-25 dihydroxy vitamin D, calcium, ESR, CRP, and ACE. Treatment of sarcoidosis involves steroids, thus it is especially important to first rule out infectious processes, especially other causes of granulomatous disease such as histoplasmosis, before proceeding with treatment. Definitive diagnosis in this patient would require biopsy of the most accessible affected tissue, often a lymph node.

Conclusion: Joint pain is a common complaint in pediatrics. Causes can range in severity from benign joint strains to potential life or joint threatening conditions such as septic joint. This case had several interesting features including a rash, recent strep infection and family history of sarcoidosis. Nevertheless, evaluation initially should focus on ruling out more common life and limb threatening conditions early so treatment is not delayed. When these conditions have been considered, benign and uncommon causes should be evaluated. Septic joint and rheumatic fever were considered early in this case before making a sarcoidosis diagnosis. Additionally, more common infectious granulomatous diseases such as histoplasmosis and tuberculosis needed to be ruled out before treatment with steroids could begin. This case highlights the importance of keeping a broad differential, but prioritizing evaluation and not jumping to rare conditions too soon.

Resources:
Case Presentation: A 16 year old male with autoimmune polyendocrine syndrome type 1 (APS), failure to thrive, cytopenias of unknown etiology, prolonged QTc presents with intermittent episodes of vertigo, dizziness, and headaches that have increased in frequency over the last 5 months. As a result of his APS, he has chronic autoimmune hepatitis, pancreatitis, and enteritis and has a g-tube to maximize his nutrition. He also reported transient blurry vision and diplopia. Episodes of dizziness last up to two weeks and self-resolve. He is on multiple medications including tacrolimus, ursodiol, and prednisone. He was previously on growth hormone, but this was discontinued.

On exam, he is small and malnourished. Vital signs and orthostatics were normal. His mentation was intact. His neurologic exam was notable for an unsteady, shuffling gait, and dizziness reported with standing. His cranial nerves were intact, there was no nystagmus or papilledema, and he had normal strength and reflexes. CBC, electrolytes, and EKG were stable. Due to his intermittent and chronic symptoms a brain MRI was obtained.

Discussion: Upon admission, due to the patient’s complex medical history and non-specific, intermittent nature of his symptoms the differential diagnosis was broad. It included vestibular causes, an intracranial process, encephalitis, acute disseminated encephalomyelitis, and medication related side effects. There was a lesser concern for venous sinus thrombosis and cardiac etiologies. MRI brain was done which demonstrated nearly symmetric signal abnormality within the mammillary bodies, basal ganglia, and midbrain (Figures 1,2). These findings were consistent with Wernicke’s encephalopathy. A thiamine level was obtained prior to starting treatment which later confirmed thiamine deficiency. He was immediately started on IV thiamine and his symptoms improved dramatically.

Conclusion: Wernicke’s encephalopathy is a rare disorder in pediatric population. The cause of his symptoms was likely due to his long standing history of autoimmune enteritis and malabsorption. Wernicke’s encephalopathy classically presents in alcoholics as a triad of confusion, ophthalmoplegia, and ataxia. The diagnosis calls for a high index of suspicion as the majority of those affected do not present with the classic triad. However, there are non-alcohol causes
typically seen in pediatrics. These cases are usually due to bariatric surgery, malignancy, hemodialysis, or hyperemesis. MRI imaging demonstrates T2 symmetric signal abnormalities in the thalamus and mammillary bodies. The complex medical history and non-specific complaints the patient reported, made this a challenging diagnostic case. Lastly, the intermittent, self-resolving nature of his symptoms also played a role in delaying further work up and eventual diagnosis.

**Resources:**

**Presented by** Teena Hadvani at: Conundrums Session #2 - Spot D, 7/22/2017 10:30 AM

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**Title:** Refractory Diarrhea: When Rotavirus Goes Rogue  
**Authors:** Erin MacLeod, DO, University of Texas at Austin-Dell Children's Medical Center  
Kathryn Neupert, MD, University of Texas at Austin-Dell Children's Medical Center, Sheeba Kunnel, MD, University of Texas at Austin-Dell Children's Medical Center  

**Case Presentation:** A 6 week old male presented to the emergency department for 36 hours of profuse, watery diarrhea, initially requiring stabilization in the intensive care unit for hypovolemic shock. He tested positive for rotavirus. As he failed to improve, the initial diagnosis became a prolonged course of rotavirus and ensuing postenteritis syndrome. He underwent endoscopic evaluation that showed diffuse villous atrophy and inflammation throughout the gastrointestinal tract.

As his course became more protracted he remained fully dependent on TPN despite trials of multiple formulas. Other features emerged, notably moderate eczema and an elevated IgE. Our workup included evaluation for food sensitive enteropathy, congenital malabsorptive disorders and immunodeficiency. This led to evaluation of his T lymphocytes and consideration of immune dysregulation. Ultimately, this patient’s diagnosis was obtained from whole exome sequencing, which showed a de novo mutation within the FOXP3 gene, consistent with a diagnosis of immunodysregulation polyendocrinopathy enteropathy X-linked syndrome (IPEX).

**Discussion:** IPEX is an exceedingly rare illness without well-known incidence. Mutations of the FOXP3 gene result in dysfunction of T regulatory cells, leading to the autoimmunity in IPEX. It has 3 main clinical features: intractable diarrhea, insulin dependent diabetes mellitus and eczema. The rarity of this syndrome and the patient’s presentation with a rotavirus infection, suggesting a postenteritis syndrome, led to consideration of this diagnosis a month into his course. His intractable diarrhea, eczematous cutaneous involvement, and elevated IgE all fit with IPEX. Interestingly, he had a relatively normal T-regulatory cell flow cytometry. This finding has been reported in IPEX as the FOXP3 mutation can cause non-functional proteins though total number of cells may be relatively normal. Of note, as of 4 months of age, he has not yet developed thyroid disease or autoimmune diabetes, the major endocrinopathies involved in this syndrome. This highlights the need for sequencing of the FOXP3 gene for definitive diagnosis of IPEX, as phenotypes may vary.

**Conclusion:** Though extremely rare, IPEX is an important diagnosis to consider in cases of intractable diarrhea of infancy because delay in diagnosis can be fatal. The only known cure for IPEX is immunosuppressive agents and hematopoietic
stem cell transplant (HSCT), ideally before the onset of neonatal diabetes. As with other autoimmune disorders, IPEX can exhibit disease ‘flares’ which may be triggered by infections, among other causes. So although his rotavirus infection can be viewed as a potential detractor, it is possible that the rotavirus infection triggered the initial flare of his IPEX syndrome, ultimately leading to his diagnosis. Our patient’s course has been complicated by multiple bacterial illnesses and he has begun immunosuppressive therapy with the goal of HSCT when medically stable.

Resources:
References

Presented by Erin MacLeod at: Conundrums Session #2 - Spot E, 7/22/2017 10:45 AM

Title: No X to Mark the Spot
Authors: Amanda Hartke, MD PhD, Greenville Health System
Marla Chapman, MD, Greenville Health System
Case Presentation: An 18yo female with XO/Fragile X was admitted for dehydration and oropharyngeal ulcers. Tests for GAS and HSV were negative and she was diagnosed with herpangina. Loose stools were noted and attributed to recent antibiotics. She was admitted a month later for left elbow septic arthritis with CRP 173mg/L, bloody joint fluid, but all cultures negative. She had loose stools again and one hematochezia episode. An anal skin tag was noted but no tears/fissures. Stools improved and no further workup was pursued. She was discharged with 21d of IV Rocephin but was readmitted in 2 weeks for right knee effusion. Joint fluid was bloody without crystals/organisms and all cultures were negative. MRI noted inflammatory arthritis, prompting evaluation of SLE, GAS, Celiac, and HLA-B27 – all negative. Rocephin was stopped and she was discharged on Naproxen/Prevacid. Recurrent loose stools were evaluated with lactoferin(+), calprotectin(>2000mcg/g) and infectious stool studies(negative). EGD/colonoscopy noted chronic gastritis and multiple colitis areas with biopsies consistent with Crohn’s disease.

Discussion: Developmentally delayed patients present diagnostic challenges, more so when they have atypical disease presentations. Consideration of increased disease rates within certain genetic conditions, like IBD in Turner Syndrome (TS), may help guide evaluation of these patients.

Conclusion: This patient presented with oral ulcers and arthritis, which can be extra-intestinal manifestations of IBD, but was not diagnosed until after three admissions. In the 3 months since initial presentation, she had had loose stools, decreased appetite, and an overall 15lb weight loss. She was treated with a prolonged steroid taper and mesalamine enemas with significant improvement in her stools and no recurrent arthritis episodes. While the hallmarks of IBD include chronic abdominal pain, diarrhea, and weight loss, oversight of extra-intestinal manifestations as presenting signs of IBD can delay diagnosis. This patient presented multiple additional challenges, including being non-verbal, having developmental delay, TS and Fragile X. Patients with TS patients have nearly 5 fold increased rates of Crohn’s disease, often with more severe disease course (1). Awareness of this association should be considered when treating TS patients with arthritis or colitis symptoms or female IBD patients with growth restriction despite resolution of IBD symptoms (2,3).

Resources:
Case Presentation: 12-year-old girl presented with altered mental status, hypothermia, and bradycardia following one week of abdominal pain, progressive fatigue and flattened affect. Family denied ingestion, fever, cardiorespiratory symptoms, headaches or unusual exposures. Exam significant for Temperature 33.2°C, Heart Rate 42 bpm, sluggishly reactive pupils, disconjugate gaze, able to intermittently follow commands and localize to pain. Laboratory studies revealed hyponatremia, hypokalemia and negative infectious work-up. CSF analysis had a persistent lymphocytic pleocytosis and oligoclonal bands. MRI Brain showed a non-enhancing, ill-defined lesion involving the hypothalamus and majority of the diencephalon, eventually with scattered grey-matter lesions which showed hypercellularity on biopsy. NMO/AQP4 titer was positive in the CSF, but not the serum. She was diagnosed with probable Neuromyelitis optica (NMO) autoimmune encephalitis with predominantly hypothalamic involvement. IVIG had no effect. Steroid treatment provided minimal improvement. Rituximab resulted in near-resolution of symptoms.

Discussion: Autoimmune encephalitis (AE) is an increasingly recognized etiology of encephalitis. It remains challenging to diagnose due to its varied presentation and limited antibody testing. Common presentations include delirium, seizures, behavioral changes, autonomic instability, respiratory failure, coma, and sepsis (1). NMO typically presents with optic neuritis or transverse myelitis but can present as an AE and has been described in AE syndromes related to diencephalic and brainstem pathology, as in this case. About 65% of children with suspicion for NMO have positive AQP4-IgG in the serum but seronegativity should not exclude the diagnosis as AQP4-IgG may appear as late as 5 years after initial symptoms (2). Early recognition and treatment can lead to reduced morbidity and mortality (3). The absence of antibodies should not delay treatment if AE is suspected (4). Treatment includes immunomodulatory therapies. First line therapies include steroids, plasmapheresis, and IVIG with advancement to second-line therapies such as rituximab or cyclophosphamide if no clinical improvement is seen.

Conclusion: As in this case, AE should be suspected in patients with an acute or subacute onset of neurologic symptoms that correlate with signs of inflammation on CSF studies, histopathological tests, and/or imaging. Other etiologies including toxins, vasculitis, metabolic and infectious should be explored. Though there is often diffuse brain involvement, presentations may have specific symptoms related to the localization of the initial inflammatory process. Hypothalamic involvement is reported to be associated with NMO/AQP-4 antibody-mediated encephalitis, though this patient lacked other typical associated symptoms associated with neuromyelitis optica (NMO) (5). A history of autoimmune conditions as well as a preceding infectious prodrome support the diagnosis. Immunomodulatory therapy should be initiated as soon as infectious etiologies have been excluded to minimize morbidity and mortality. Clinical response should guide therapy, though treatment may take days-to-weeks to take effect and overall improvement in AE is often slow.

Resources:
2. Dalmau et al. Clinical Experience and laboratory investigations in patients with antiNMDAR encephalitis. Lancet Neurol 2011; 10; 63-74
3. Bale Jr, J. Virus and Immune-Mediated Encephalitides; Epidemiology, Diagnosis, Treatment, Prevention. Pediatric Neurology. 2015; 53; 3-12

**Presented by** Alyssa Tilly at: Conundrums Session #3 - Spot B, 7/22/2017 4:30 PM

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**Title:** A Perf-ect Differential  
**Authors:** Carolyn Marcus, MD, Boston Children’s Hospital  
**Case Presentation:** A 6 year old boy, with a history of constipation, presented with two days of abdominal pain, mostly on the left. He was seen in the ED, with a KUB revealing a large stool burden. He was given an enema which led to a bowel movement with pain relief so was discharged home. The pain recurred, so he re-presented to the ED where he had diffuse abdominal tenderness. Abdominal ultrasound did not visualize the appendix and had no signs of intussusception. He was admitted with a diagnosis of likely constipation for serial exams and bowel clean out. He was started on NG Golytely, and his pain improved with stool output; however, he continued to have episodes of severe pain followed by times with no pain, with persistent tenderness to deep palpation in the LLQ. Repeat ultrasound was performed which revealed a small, blind-ending loop of bowel in the midline pelvis, with adjacent fat stranding. A CT was performed, revealing likely Meckel’s diverticulitis. He went to the OR and was found to have a perforated Meckel’s diverticulum.

**Discussion:** Meckel’s diverticulum is the most common congenital anomaly of the gastrointestinal tract. They are usually clinically silent, but when symptomatic, are classically thought to present with painless bleeding secondary to ectopic gastric mucosa. Additionally, they can present with nonhemorrhagic symptoms, including bowel obstruction, inflammation, and perforation. Similar to acute appendicitis, entrapment of a foreign body within the diverticulum can lead to diverticulitis. As compared to appendicitis, the pain is generally located more toward the midline; however, the position of the diverticulum can vary. In this patient with known constipation, a KUB revealing large stool burden, and pain relief with stool output, the diagnosis was initially assumed to be constipation. Anchoring bias was part of the diagnostic challenge in this case. However, this patient’s persistence of pain and tenderness led us to further investigation.

**Conclusion:** Meckel’s diverticulitis should be on the differential for children with acute abdominal pain and can present similarly to acute appendicitis. Clinicians should be aware of their cognitive biases and consider other diagnoses when the clinical picture does not fit.

**Resources:**

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**Presented by** Carolyn Marcus at: Conundrums Session #3 - Spot C, 7/22/2017 4:45 PM
**Title:** A Severe Case of the “Blues”  
**Authors:** Laura Hodo, MD, University of Utah - Primary Childrens Hospital  
            Kirsten Rupp, MD, University of Utah

**Case Presentation:** A 13 year old Hispanic female with past history of depression and suicidal ideation presented to an outside hospital after an overdose. She was found in the bathroom with open pill bottles by her mother, who took her to the ED. Her mother hypothesized the ingested medications were aspirin and phenazopyridine (Pyridium). Upon arrival, her cognition was appropriate and she endorsed abdominal pain, nausea, and tinnitus. Vital signs and exam were normal. Urine drug screen was positive for THC and urine sample was notably orange, but pregnancy test, urinalysis, and salicylate level were within normal limits. Given clinical stability, discharge was planned. Prior to discharge, however, she developed cyanosis and hypoxia. Arterial blood gas (ABG) was performed and significant for pH 7.53, pCO2 20 mmHg, pO2 128 mmHg, SpO2 85%, and methemoglobin (MetHb) 48%. Given her significant methemoglobinemia, 1mg/kg IV methylene blue was administered and she was transferred to the University of Utah for further care. She was admitted to a pediatric hospitalist team for further monitoring and care.

**Discussion:** MetHb is an altered state of hemoglobin in which ferrous iron is oxidized to the ferric state and unable to bind oxygen, shifting the oxygen dissociation curve to the left. Clinical presentation ranges from asymptomatic to respiratory depression, altered consciousness, and death. Consider methemoglobinemia in the setting of cyanosis and hypoxia unresponsive to supplemental oxygen. Minimal MetHb is present in normal serum; proportions above 1% are considered pathological, and may be congenital or acquired. The acquired form results from exogenous agents increasing methemoglobin formation, such as toxic or therapeutic doses of phenazopyridine, especially in infants and the elderly. Phenazopyridine’s mechanism of action is not well-understood, but it is secreted into the urine and acts as a local anesthetic. Treatment of methemoglobinemia includes discontinuing offending agents and administering methylene blue or vitamin C if the patient is symptomatic or for MetHb level >20%. Methylene blue provides an artificial electron transporter, reducing hemoglobin via the NADPH pathway.

**Conclusion:** Despite initial concerns about aspirin toxicity, this patient developed methemoglobinemia secondary to phenazopyridine overdose. After one dose of 1mg/kg IV methylene blue, her MetHb level was 40%. Toxicology recommended repeating the dose, with a subsequent decrease in her MetHb to 3%. Following methylene blue administration, she was monitored for complications of methylene blue, including rhabdomyolysis and hemolytic anemia. Creatinine kinase and complete blood count were normal prior to transfer. Pulse oximetry is unreliable in methemoglobinemia, so the patient was monitored for respiratory distress and received periodic ABG’s. She made a complete recovery and was transferred to a psychiatric facility for further management. Methemoglobinemia is a potentially fatal condition if not accurately and timely diagnosed, and clinicians must have a low threshold for the diagnosis in the proper scenario. Clinicians astutely identified methemoglobinemia in our patient, resulting in appropriate treatment and full recovery.

**Resources:**

**REFERENCES**


**Presented by Kirsten Rupp at:** Conundrums Session #3 - Spot D, 7/22/2017 5:00 PM
Title: Acute Genital Ulcerations
Authors: Weijen Chang, MD, U Mass Medical School / Baystate Children’s Hospital
Rachel Clarke, MD, U Mass Medical School / Baystate Children’s Hospital

Case Presentation: This is a previously healthy 13 year-old girl who experienced malaise, headaches, and fevers 5-6 days prior to admission. She then experienced vulvar pain and erythema that evolved into “black dots” which ulcerated. A targetoid rash then appeared on her left shoulder where a tick bite occurred 2 weeks prior. She was started doxycycline, but worsening vulvar pain, dysuria, and urinary retention led to admission. Exam showed two 0.5x1 cm ulcers with yellow exudate over an indurated left labia majora. A circular blanching erythematous rash 20 cm wide with central clearing was on her left shoulder. She had no leukocytosis, Hgb 10.8 g/dL, an ESR of 37 mm/hr, and unremarkable liver panel. Oral doxycycline was continued and she was treated for vulvar pain. She was discharged after 2 days with improvement in pain and ulcerations. After discharge, Lyme IgG Western blot came back negative but was positive for IgM and remained positive after 2 weeks. EBV viral capsid IgM was initially positive at 57.0, but was negative after 2 weeks, with persistent negative IgG and nuclear antigen antibody.

Discussion: Nonsexual acute genital ulceration (NAGU), also known as Lipschutz ulcers, are a rare complication of Lyme disease, with only one prior case reported in the adult literature.1 This is the first reported case of NAGU resulting from Lyme disease in a pediatric patient. NAGU has been associated with EBV,2 Mycoplasma, HIV, mumps,3 cytomegalovirus,4 and influenza disease. The clinical history and laboratory findings followed by rapid improvement on doxycycline suggested that the etiology was Lyme disease, but positive EBV viral capsid IgM also suggested this as a possibility. Persistently positive Lyme IgM Western blot and negative EBV viral capsid IgM after 2 weeks confirmed our clinical suspicion. The differential diagnosis also included inflammatory disease (e.g., Behcet’s disease) and sexually transmitted infection, but the low ESR and negative STI testing (negative urine GC/chlamydia panel, HIV, RPR) made these unlikely. In retrospect, the initial positive EBV viral capsid IgM was likely due to serologic cross reactivity with Lyme disease, which has been described previously.5

Conclusion: In areas where Lyme disease is endemic, patients with acute non-sexual genital ulceration should be evaluated for Lyme disease if the history and physical are suggestive of this. EBV, mycoplasma, influenza, mumps, CMV and inflammatory conditions should also be considered. Follow up serologies can be helpful in confirming the diagnosis.

Resources:
References:

Presented by: Weijen Chang at: Conundrums Session #3 - Spot E, 7/22/2017 5:15 PM
Case Submissions – Poster Presentations

**Title:** A Rare Cause of Unilateral Cervical Lymphadenitis with Rare Sequelae

**Authors:** Jessica Mayer, M.D., Riley Hospital for Children at Indiana University Health
Frank Solty, M.D., Riley Hospital for Children at Indiana University Health, Kimberly Schneider, M.D., Riley Hospital for Children at Indiana University Health

**Case Presentation:** 11 y.o. female presented from an outside hospital (OSH) after oral antibiotic treatment for OM and unilateral lymphadenitis failed, prompting admission to the OSH. Over the next 9 days, lymph node (LN) increased in size with daily fevers, night sweats and fatigue. Despite treatment with clindamycin and vancomycin for 4 days, fevers continued as did enlargement of the LN. Neck CT showed enlarged left cervical lymph nodes without abscess. Labs notable for leukocytosis of 36.9 with neutrophil predominance and CRP of 33. She was transferred for further work-up. Biopsy was done to rule out lymphoma, and antibiotics were continued. By day 12 she developed intermittent conjunctivitis and peeling of skin on hands and feet. Echocardiogram showed multiple severe coronary artery aneurysms (CAA) diffusely confirming the diagnosis of incomplete Kawasaki disease. With high dose aspirin, IVIG, and warfarin her symptoms resolved. Her course was then complicated by left-sided Bell’s palsy, a rare complication of Kawasaki disease, which was treated with prednisone.

**Discussion:** Atypical or incomplete Kawasaki disease (KD) is a rare presentation of a potentially life threatening condition. It is characterized by fever >5 days, and 2-3 clinical criteria (conjunctivitis, polymorphous rash, oral mucous membrane involvement, desquamation/edema of extremities, cervical lymphadenopathy). Its diagnosis can be aided by elevated inflammatory markers and supplemental laboratory criteria per the AAP and AHA algorithm for the evaluation of suspected incomplete KD. Often, as in this patient, diagnosis is delayed due to incomplete development of clinical symptoms. Due to the delay in diagnosis, patients with incomplete Kawasaki have an increased risk of CAA.1 The typical age range for KD is 6 mo to 5 years, making diagnosis outside of this age range more challenging.2 Patients younger or older than this age range, such as our patient are more likely to present as incomplete KD, thus leading to delayed diagnosis, and increased CAA. 2,3

**Conclusion:** Pediatric providers should continue to be vigilant for the development atypical/incomplete Kawasaki disease in patients with prolonged fever > 5 days. In patients not meeting the full 4+ criteria for KD, continued clinical suspicion, reevaluation, and laboratory work-up is critical as these patients may go on to develop KD. Prompt identification and treatment is crucial to prevent the mortality and morbidity of associated sequelae including both cardiac and non-cardiac manifestations. Patients older than 5 are often misdiagnosed as suppurative lymphadenitis.3 The lymph node in our patient reached 9 cm in size at its largest with little other clinical signs in the first 11 days other than fever, leukocytosis and elevated inflammatory markers. Heightened clinical suspicion when the LN failed to respond to appropriate treatment should prompt consideration of other diagnosis, such as KD or cancer, as in our patient.

**Resources:** References:


**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A101
Case Presentation: An 8-month-old female with feeding intolerance on Neocate formula presented with decreased movement of her right lower extremity. Physical examination revealed pain with range of motion and bowing of her extremities. Radiographs revealed a fractured right fibula and diffuse osteopenia with associated bowing of fibular diaphysis consistent with rickets. Initial laboratory investigations confirmed hypophosphatemic rickets and further evaluation revealed adequate renal absorption of phosphate. The consideration that phosphate bioavailability in the formula may be impaired was suggested and an alternate formula (Elecare) was substituted. A significant increase in phosphate and decrease in calcium levels occurred with the formula change and calcium supplementation was added. Repeat radiographs confirmed improved osseous mineralization after two months of therapy and calcium supplementation was discontinued. Subsequent labwork remained normal on Elecare formula alone. Final diagnosis was hypophosphatemic rickets associated with presumed poor gastrointestinal absorption of phosphate.

Discussion: Phosphate plays a critical role in bone mineralization with 85% of total body phosphate as hydroxyapatite, the mineral form of calcium apatite that makes up human bone. The most widely observed mechanism for hypophosphatemic rickets is associated with renal phosphate wasting, as seen in X-linked hypophosphatemic rickets. In this clinical conundrum, evaluation confirmed appropriate renal phosphate absorption and extrarenal sources for hypophosphatemia were sought. Discussion between Hospital Medicine and Pediatric Endocrinology led to a novel approach to therapy with a change in elemental formula. This formula change was associated with a significant increase in phosphate and subsequent improvement in bone mineralization. There is limited literature evaluating extrarenal causes for hypophosphatemic rickets and few cases reports of gastrointestinal phosphate homeostasis associated with hypophosphatemic rickets. Interestingly, there has been recent reports presented nationwide of similar cases associating Neocate formula with hypophosphatemia and subsequent rickets.

Conclusion: Extrarenal phosphate losses are an uncommon cause of hypophosphatemic rickets. In this clinical conundrum, a novel approach of changing formula was associated with significant elevations in phosphate and underlines the importance of considering non-renal phosphate losses in the differential diagnosis for hypophosphatemic rickets. Currently, there is no clear explanation for the potential association of hypophosphatemia and Neocate formula and further investigation is warranted. Potential hypotheses for this association include differences in phosphate bioavailability between formulas and the role of gastric ph in the maintenance of phosphate homeostasis.

Table 1. Summary of laboratory values in response to changes in management plans

<table>
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<tr>
<th>Variable</th>
<th>Reference Range (“)</th>
<th>Initial Presentation</th>
<th>1 Week Follow Up</th>
<th>2 Week Follow Up</th>
<th>2 Month Follow Up</th>
<th>2 Month Follow Up</th>
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<tbody>
<tr>
<td>Calcium (mg/dL)</td>
<td>10.0</td>
<td>7.4</td>
<td>8.8</td>
<td>10.2</td>
<td>10.6</td>
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<tr>
<td>Phosphate (mg/dL)</td>
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<td>7.2</td>
<td>6.5</td>
<td>5.9</td>
<td>5.7</td>
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<tr>
<td>Parathyroid Hormone (pg/mL)</td>
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<td>373</td>
<td>158</td>
<td>59</td>
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<tr>
<td>Alkaline Phosphatase (ui/I)</td>
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<td>777</td>
<td>416</td>
<td>358</td>
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<td>Vitamin D3 (ng/mL)</td>
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<td>Vitamin D 25-Dihydroxy (ng/mL)</td>
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<td>Urine Ca-Cr</td>
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<tr>
<td>Urine Ph-Cr</td>
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<td>2.10</td>
<td>0.97</td>
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<td>Management Plan</td>
<td>Formula changed to Elecare</td>
<td>Calcium supplement added</td>
<td>No changes made</td>
<td>Calcium supplement discontinued</td>
<td>Continued on Elecare</td>
<td></td>
</tr>
</tbody>
</table>

*Reference ranges are age-adjusted and based on _______
Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A102

Title: New onset seizures in a teen, or something more?

Authors: Lajiness Jacquelyn, MD/PhD, Riley Hospital for Children at Indiana University Health
Fatima Jafri, MD, Riley Hospital for Children at Indiana University Health

Case Presentation: A healthy 15 yo girl had new onset generalized tonic clonic seizures. Initial ED negative workup included head CT, EEG, CBC, UA and UDS. She was discharged with neuro followup. With her 3rd seizure she developed altered mental status (AMS) leading to admission and loading with levetiracetam. On Hospital day (HD) 1 fevers began with minimal improvement in her AMS. Brain MRI and LP were done. CSF had a white blood cell count of 87 and all other studies negative including a meningitis encephalitis panel PCR for the common bacterial and viral causes of CNS infection. On HD4 her AMS worsened and status epilepticus developed prompting transfer to the ICU and intubation. Repeat MRI showed infarct in the left temporal and parietal regions. Repeat CSF studies were positive for West Nile virus and GABA-B-R antibody indicating a primary West Nile viral infection compounded by an autoimmune response. She was given IVIG for 5 days as treatment of both the West Nile Virus and the autoimmune process. Her mental status improved some, but has not returned to her baseline.

Discussion: West Nile virus is one of the most widely disseminated arboviruses in the world and can produce severe neurological sequelae including seizure, coma, and death. As of November 2016, there have been 1,617 cases of West Nile reported to the CDC. Of these 855 were considered neuro-invasive (encephalitis or meningitis).(1) Treatments for
West Nile virus are limited, but there is some data to show that alpha-interferon, ribavirin, or IVIG can help resolve clinical symptoms.\(^{(1,2,3)}\) In patients who are severely affected, such as ours, the benefits of a trial of treatment far outweighed the risks. In addition, the presence of a rare autoimmune antibody further complicates the picture. It is unclear whether the West Nile is a false positive, or more likely, that the West Nile infection triggered the autoimmune process. Regardless of cause, IVIG was found to be a possible treatment for both conditions, and was thus given to our patient with resolution of fevers, and some improvement of mental status.\(^{(1,4)}\)

**Conclusion:** Unprovoked new onset seizures warrant careful management, especially in a completely healthy high achieving teen. While West Nile Virus is not widespread in Indiana, it has been seen infrequently. It is important in cases that are not consistent with typical expected course, such as this one, that providers continue to look for answers that are less common in order to provide the best chance for treatment and recovery. Based on continued pursuit of the etiology, our patient has been able to receive treatment and show some recovery. When a patient is not improving as expected for the diagnosis they have received, continued workup and pursuit of other etiologies or complication factors should ensue. In this patient, the West Nile virus could have been the sole cause, but further investigation was done due to her profound AMS and severe status epilepticus, leading to the diagnosis of autoimmune encephalitis as well.

**Resources:** References:

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**Title:** Fatal strokes caused by fibromuscular dysplasia in a child with cystinosis  
**Authors:** Jonathan Fliegel, MD FAAP, University of Wisconsin School of Medicine and Public Health  
Jeff Clark, MD, University of Wisconsin School of Medicine and Public Health

**Case Presentation:** An 8 y.o. girl with cystinosis collapsed on a playground, was unresponsive and had “stiffening and clenching”. She was confused for 2 hours then recovered. Vital signs and neurologic exam were normal. A non-contrast head CT showed no fracture or hemorrhage. She had a headache but no fever nor recent illness and she had no history of syncope or seizures. During a 4 day hospitalization vital signs and neurologic exams were normal, except she did not recall the event. Labs, ECG, telemetry and video EEG were normal, even during a brief episode of confusion. Two nights after discharge, she returned to a local ED for right-sided stiffening and facial droop that resolved. Neurology was consulted, evaluated her in clinic the following day and admitted her. Overnight video EEG captured multiple spells but no seizures. An MRI/MRA showed a left PCA stroke, multiple infarcts and basilar artery occlusion. Over the next day, her condition worsened and she underwent lysis of a basilar artery thrombosis. After a repeat MRI/MRA showed extensive new infarcts, her family withdrew life support.

**Discussion:** Her initial stiffening and confusion were consistent with a seizure. Her quick recovery after both events, her CT with no findings of stroke, her normal EEG and neurologic exams all aligned with a seizure diagnosis. Since her diagnosis of cystinosis at 10 weeks of age, our patient had been compliant with cysteamine therapy. Some children with cystinosis exhibit neurologic findings but there are no reports of seizures or stroke. We rely on clinical neurologic exams to guide our evaluation. Remarkably she had a normal neurologic exam even after her initial MRA that revealed extensive vasculopathy. In retrospect her persistent headaches and dizziness may have been symptoms of stroke, but may also occur with seizures. Her autopsy confirmed acute infarcts of her pons, midbrain, cerebellum and left temporal and occipital lobes. Intimal and medial changes in her basilar, carotid and middle cerebral arteries were diagnostic of fibromuscular dysplasia (FMD). These non-inflammatory arteriopathies may be asymptomatic or may result in occlusion, stenosis, or dissection as we saw in her case.

**Conclusion:** - Clinical diagnosis of pediatric stroke is challenging.
In this case, her normal neurologic exams were misleading in the face of her extensive abnormalities in cerebral vasculature caused by FMD.

- Strokes involving the posterior circulation may present with subtle, non-specific and transient symptoms.
- Head CT scans have limitations in diagnosing strokes.

Non-contrast head CTs are useful in acute settings to assess for fractures or hemorrhages. Without contrast, they cannot outline vascular abnormalities. In addition, the thick mastoid bone leads to loss of sensitivity and detail in the posterior fossa including areas of posterior cerebral circulation.

- An anchoring bias (that her case represented an atypical seizure) may have led to a delay in performing her MRI/MRA and subsequent stroke treatment. Nonetheless it is unlikely that her outcome would have been different given her extensive FMD.
- Though common things do occur commonly, uncommon things can occur. There are no previous reports of cystinosis and FMD occurring together.

**Resources:** N/A

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A104

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**Title:** More Than Meets the Eye: A Rare Cause of Proptosis

**Authors:** Victoria Rodriguez, MD, Ann and Robert H. Lurie Children's Hospital of Chicago
Allison Mariani, MD, Ann and Robert H. Lurie Children's Hospital of Chicago, Kaitlyn Olson, MD, Ann and Robert H. Lurie Children's Hospital of Chicago, Ronit Lever, MD, Ann and Robert H. Lurie Children's Hospital of Chicago

**Case Presentation:** A 14 year old previously healthy boy presented to the ER with 5 days of diffuse throbbing headache and worsening neck pain. He denied fevers, emesis, visual changes, numbness, weakness, and head trauma. Vital signs were normal. Exam was notable for neck stiffness with severely restricted range of motion, normal eye and throat exams, and no neurologic deficits. WBC count was normal yet inflammatory markers were elevated. Head and neck CT scans were normal except for paranasal sinus opacification. On hospital day 2, he developed meningismus. LP revealed CSF pleocytosis so empiric ceftriaxone and vancomycin were started. The next day he developed upper eyelid edema with erythema, proptosis, and painful extraocular movements. CT orbits showed ophthalmic vein thickening and internal carotid artery narrowing concerning for cavernous sinus thrombosis. MRI confirmed these findings, showing filling defects of the cavernous sinus, paranasal sinus opacification, bony enhancement indicative of osteomyelitis, and dural enhancement consistent with meningitis. He was transferred to the PICU.

**Discussion:** The differential diagnosis of unilateral proptosis and headache includes orbital cellulitis, Graves’ disease, malignancy, orbital pseudotumor and cavernous sinus thrombosis (CST). Orbital cellulitis is an infection of the ocular muscles and/or periorbital fat. It typically presents with eyelid swelling, erythema, pain, and vision changes. As a known complication of acute sinusitis, orbital cellulitis is usually caused by S. aureus or streptococcal species. In contrast, proptosis caused by Graves’ disease has a more gradual onset and is uncommon in children. Malignancy, most commonly leukemia and rhabdomyosarcoma, must always be considered in a child with proptosis. However, malignancy was less likely in this case due to rapid symptom onset. Orbital pseudotumor is an idiopathic inflammation of the orbit that is treated with steroids. Patients typically present with proptosis, ocular pain, ophthalmoplegia, and erythema. CST occurs when a blood clot develops within the cavernous sinus. MRI confirmed these findings, showing filling defects of the cavernous sinus, paranasal sinus opacification, bony enhancement indicative of osteomyelitis, and dural enhancement consistent with meningitis. He was transferred to the PICU.

**Conclusion:** The patient was diagnosed with a cavernous sinus thrombosis as a complication of bacterial sinusitis. CST is uncommon in children. The cavernous sinus is located posterior to the sphenoid sinus and contains the internal carotid artery (ICA), portions of CN II-VI, and venous drainage from the eye and cerebrum. Early symptoms include headache, fever and emesis. Proptosis occurs as a result of impaired venous drainage, followed by cranial nerve palsies and stroke from ICA narrowing. Infectious complications include meningitis and subdural empyema. MRI/MRA is diagnostic. Treatment involves antibiotics, surgery, and anticoagulation. Despite treatment, the mortality rate approaches 10% and 20-40% of children have permanent vision loss. Our patient underwent endoscopic sinus surgery, was treated with 6 weeks of IV ceftriaxone and PO metronidazole, and 3 months of enoxaparin. He made a complete recovery. Although an uncommon cause of proptosis in children, CST is important to consider in light of its high morbidity and mortality, as prompt treatment can improve survival and save vision.

**Resources:**
Proptosis with inflammation of the orbit

Bilateral ethmoid and sphenoid sinusitis

Cavernous sinus expansion and thrombophlebitis

Osteomyelitis

Right ICA narrowing

Meningeal enhancement

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A105

**Title:** Be Wary of the Blueberry: A Newborn with Rash and Hepatosplenomegaly

**Authors:** Sara Duffus, MD, University of North Carolina
Eric Zwemer, MD, University of North Carolina School of Medicine, Philip Roehrs, MD, University of North Carolina, Department of Hematology-Oncology

**Case Presentation:** A male infant born at 34 weeks was noted on newborn exam to have significant abdominal distension, hepatosplenomegaly, and a diffuse maculopapular rash with petechiae (figure 1). Birth history was notable for normal routine prenatal ultrasounds, maternal exposure to ParvoB19 in the 3rd trimester, and induction of labor due to decreased fetal movements. Initial CBC showed anemia (Hgb 6.3) and thrombocytopenia (PLT 4), followed by leukopenia (WBC 6.8 on DOL2). Abdominal ultrasound revealed marked hepatosplenomegaly. Differential diagnosis included TORCH infections, Parvovirus, neuroblastoma, and Langerhans cell histiocytosis. He subsequently developed ascites and worsening liver function with direct bilirubin of 18.7. All viral testing (including HSV, EBV, CMV, HHV 6, ParvoB19, enterovirus, adenovirus) and toxoplasma were negative. Skin biopsy of an arm lesion revealed dense, histiocytic infiltrates most consistent with hemophagocytic lymphohistiocytosis (HLH). Additional labs consistent with HLH included elevated ferritin (3670) and elevated soluble interleukin-2 receptors.

**Discussion:** Hemophagocytic lymphohistiocytosis is a disorder of impaired T-cell and natural killer cell activity characterized by cytopenias, hepatosplenomegaly, and persistent fevers. When presenting in the first days of life, symptoms can significantly overlap with more common neonatal disease processes, particularly congenital infections. This patient had hepatosplenomegaly and a rash that was initially thought to be the classic maculopapular “blueberry muffin” rash associated with TORCH infections. However, as opposed to congenital infections that typically present with isolated thrombocytopenia, patients with HLH have at least 2, if not all 3, cell lineages affected. The clinical picture can further be confused as neonatal HLH can also be triggered by viral infections, particularly HSV, CMV, and enterovirus. HLH should be considered in the differential if TORCH infection is suspected and treatment is initiated, but the infant continues to demonstrate clinical decline. Ferritin can serve as an initial screening test, and levels above 500 should prompt further evaluation.
Conclusion: The clinical presentation of neonatal onset HLH considerably overlaps with that of TORCH infections, including cytopenias, maculopapular rash, and hepatosplenomegaly. Although rare, clinicians should consider HLH in the differential diagnosis in the following circumstances: when 2 or more cell lineages are affected or there is continued clinical decline despite appropriate treatment of the suspected infectious etiology. Ferritin can be used as a screening test with levels above 500 prompting further testing and involvement of an HLH expert.

Resources:

Be Wary of the Blueberry: A Newborn with Rash and Hepatosplenomegaly

Figure 1:
Maculopapular rash (arrows) shown over the scalp (A), trunk and extremities (B, C)

Cutaneous Manifestations of Hemophagocytic Lymphohistiocytosis

Dean S. Morell, MD; Marie A. Pepping, MD; J. Paul Scott, MD; Nancy B. Esterly, MD; Beth A. Drolet, MD

Table 3. Pathological, Treatment, and Cutaneous Features of Patients With Hemophagocytic Lymphohistiocytosis at Children’s Hospital of Wisconsin, 1990-2000*

<table>
<thead>
<tr>
<th>Patient No</th>
<th>Bone Marrow Results</th>
<th>Liver Biopsy Results</th>
<th>CSF Results</th>
<th>Chemotherapy</th>
<th>BMT</th>
<th>Skin Condition</th>
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*CSF indicates cerebrospinal fluid; IT MTX, intrathecal methotrexate; Mtg, intravenous immunoglobulin; and BMT, bone marrow transplantation.

References


Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A106
Title: Do You See What Eye See? A Case of Orbital Lymphangioma
Authors: Cyrus Heydarian, MD, Eastern Virginia Medical School

Case Presentation: A healthy one year old female presented with acute onset of right eye proptosis and fever. Six days ago, she was diagnosed with bacterial conjunctivitis, and was prescribed a topical antibiotic ointment. The conjunctivitis has improved, but her proptosis worsened. She returned to the ED, and was febrile to 102 F, with prominent right eye proptosis and lateral conjunctival injection noted. She was not ill appearing, and her ocular movements and pupillary reflexes were normal. A CT of the orbits was concerning for orbital cellulitis, and she was admitted on intravenous antibiotics. Her fever resolved, but the proptosis worsened. An MRI was performed on day 2, showing a cystic mass with fluid-fluid levels in the right intraconal space, consistent with an orbital lymphangioma with intralesional hemorrhage. Mild mass effect of the adjacent optic nerve was noted. Antibiotics were discontinued and a dose of oral steroids was given. Her proptosis quickly resolved, and her ophthalmic exam remained normal. She did not require additional medical or surgical interventions upon discharge.

Discussion: Orbital lymphangiomas are rare, benign vascular lesions that often present in childhood. Deep malformations often present following an intralesional hemorrhage incited by infection or trauma. Common presenting symptoms include proptosis, ophthalmoplegia, pain, and vision changes. Presentation is often confused initially with orbital cellulitis or malignancy. Diagnosis is made through CT or MRI. Lesions are managed with close observation. Medical options include sclerotics, corticosteroids, and sildenafil. Surgical excision is reserved for cases of visual compromise due to the high rate of recurrence, intraoperative hemorrhage, and proximity to the optic nerve during surgery. Rare complications include optic nerve compression with permanent vision loss.

Conclusion: Orbital lymphangiomas, though rare, may be mistaken in children for orbital cellulitis due to similar presentations. Radiography can help differentiate the diagnoses, limiting the unnecessary usage of antibiotics. Most cases self resolve without medical or surgical interventions needed, though permanent visual loss is possible in severe cases.

Resources:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A107

Title: A Shocking End to a Bad Hair Day
Authors: Courtney James, MD FAAP, MD, Our Lady of the Lake Children's Hospital
Jennifer Bentley, MD, MPH, Our Lady of the Lake Children's Hospital, Erin Hauck, MD, Our Lady of the Lake Children's Hospital

Case Presentation: A 10 year old female presented with rash for 10 days. She initially developed bilateral ocular erythema with crusting and a fine, erythematous, sandpaper rash to the face. Tobramycin eye drops were used for 2 days, but her eyes became markedly erythematous and edematous periorbitally. One week later she developed erythroderma to her face and extremities. She was prescribed olopatadine and cetirizine due to concern for allergic reaction. Over the next 3 days she developed bumps to the face with desquamation. On the day of presentation she had subjective fever, headache, decreased oral intake, and decreased urine output which prompted evaluation. Upon further questioning, her mother had applied an at home chemical relaxer to the patient's hair 3 weeks prior. She had chemical burns to areas of the scalp post relaxer treatment. These lesions worsened now with crusting and scaling in her scalp. On admission, she had hypotension with persistent tachycardia after multiple normal saline boluses. Vancomycin and clindamycin were started for presumptive diagnosis of toxic shock syndrome.
**Discussion:** Toxic shock syndrome (TSS) is a potentially life threatening condition. Clinical manifestations typically include hypotension, fever, and skin findings. Progression of symptoms to hemodynamic instability can be rapid. Chemical relaxers are widely used in the African American population to straighten coarse, curly hair via active alkaline agents. These products are sold over the counter with widespread in-home use versus application by a professional cosmetologist. Due to variability in application and processing time, there is significant risk of chemical burns as a result of over processing. It is well documented that TSS is a possible complication from burns to the skin, however, scalp chemical burns as a preceding injury leading to TSS are not widely documented. Similar to this case, parents often do not recognize scalp chemical burns as being equally dangerous as other skin burns and therefore often do not report these lesions to clinicians.

**Conclusion:** The patient in this case presented with classic signs and symptoms of TSS, however an uncommon nidus of infection led to delayed diagnosis and proper treatment. Thorough history and careful physical exam are imperative for early identification of these uncommon sources of infection. Timely recognition is essential to preventing significant morbidity and mortality.

**Resources:** N/A

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A108

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**Title:** Atypical CSF Findings in the Setting of Presumed Guillain-Barré Syndrome

**Authors:** Samuel Dillman, MD, Medical College of Wisconsin
Anika Nelson, MD, Medical College of Wisconsin

**Case Presentation:** A 7-year-old previously healthy boy presented to his local emergency department with a two-day history of progressive lower extremity weakness. Parents reported two days of “clumsiness,” abnormal gait, and frequent falls. Two weeks prior, he had a 7-day course of non-bloody diarrhea associated with fevers. He described paresthesias that preceded the onset of weakness. He did not have bowel or bladder dysfunction, headache, back pain or fever on presentation and no history of trauma. The ED providers noted a “targetoid” rash on his arm that resolved within 12 hours and his parents attributed this to the tape used to secure his IV. There was no known history of tick bites, but he had been on fishing trips in an endemic area of Wisconsin and admitted to having “bug bites.” On neurological exam, sensation was intact but the patient was unable to move his feet or lift his legs. Patellar reflexes were absent with trace Achilles reflexes present bilaterally. The patient was admitted to the hospital and started on IVIG for presumed Guillain-Barré Syndrome (GBS).

**Discussion:** He was treated empirically with Vancomycin and Ceftriaxone for 48 hours. An MRI of the brain and spine was consistent with acute inflammatory demyelinating polyradiculoneuropathy. A lumbar puncture was performed and CSF demonstrated 68,000/µL white blood cells with 71% lymphocytes, 22% monocytes, 6% neutrophils, glucose of 53 mg/dL and protein of 46 mg/dL with a negative gram stain and culture. His CSF studies did not show the expected cytoalbuminologic dissociation seen with GBS and his ankle reflexes were present while patellar reflexes were absent, which is atypical for GBS. Tick paralysis was considered, but no tick was found on thorough examination. A stool culture was positive for Campylobacter jejuni. The patient’s weakness slowly improved. Several days into his hospitalization, Lyme Elisa and Western titers resulted with a positive IgM, negative IgG. Given this, the history of possible tick exposure, targetoid rash on presentation, and CSF findings that are consistent with Lyme meningitis, he was treated with Ceftriaxone for a 28-day course.

**Conclusion:** Acute nervous system infection with Lyme disease commonly manifests as meningitis with a lymphocytic predominance and cranial nerve palsies. An association between Lyme disease and GBS-like illness has been described but is rare. There has previously been reported a case of GBS-like illness in a patient with positive Lyme antibodies. The patient was unresponsive to IVIG but improved with antibiotic treatment. Our patient initially received empiric antibiotics making it difficult to discern if the antibiotics had partially treated Lyme infection or if the IVIG was effective therapy. Due to the atypical CSF findings, an anti-ganglioside antibody panel was also acquired for our patient and was positive. This supports an autoimmune mediated neuropathy, perhaps triggered by acute Lyme infection or his preceding diarrheal illness with Campylobacter. Findings of leukocytosis with a lymphocytic predominance and normal protein in the CSF of a patient with GBS-like illness should prompt a clinician to explore atypical triggers of GBS and consider Lyme disease in the right clinical context.

**Resources:**
Title: Not a Scorpion Sting: An Unusual Presentation of Pancreatitis

Authors: Rebecca Epstein, MD, Resident, University Hospitals Rainbow Babies and Children's Hospital
Paul Shaniuk, MD, Clinical Instructor of Medicine and Pediatrics, Rainbow Babies and Children's Hospital, Ramy Sabe, MD, Assistant Professor, Pediatric Gastroenterology, University Hospitals Rainbow Babies and Children's Hospital

Case Presentation: A 7-year-old Ashkenazi boy presented with one week of fever, jaundice and epigastric/right upper quadrant abdominal pain, in addition to non-bilious emesis and acholic stool. He denied any recent trauma or travel. Physical exam showed normal vital signs, but revealed diffuse jaundice with scleral icterus, and also guarding with tenderness to palpation of the right upper quadrant and epigastric region. The liver was palpable 2 cm below the costal margin. Bowel sounds were normal, Murphy's sign was negative and there was no splenomegaly or CVA tenderness.

Initial labs showed leukocytosis (15.5 x 10^9/L) with an elevated lipase (2,270 U/L), amylase (622 U/L), bilirubin (Total: 4.6 mg/dL, Direct: 2.1 mg/dL) and CRP (6.50 mg/dL), but normal urinalysis, alanine aminotransferase, and aspartate aminotransferase. Abdominal ultrasound and x-ray showed hepatomegaly, and IgG4 levels were normal. A magnetic resonance cholangiopancreatography (MRCP) was performed which led to the diagnosis of autoimmune pancreatitis, and the patient was started on steroids leading to clinical improvement.

Discussion: Autoimmune pancreatitis (AIP) is very rare in pediatrics, and its prevalence in the children is still unknown. There are two distinct variants of AIP. Type 1, which is typically seen in older males, is thought to be the pancreatic manifestation of an IgG-4-associated systemic disease, and often presents with other IgG4 related diseases such as IgG4-associated sclerosing cholangitis and sclerosing adenitis. In contrast, Type 2 appears to be a more pancreas-specific disease, except for possible association with inflammatory bowel disease, and has no known serological markers. The vast majority of reported pediatric AIP cases are Type 2.

The diagnosis of AIP is based on a combination of imaging, histology and response to steroids. Suggestive findings on ERCP or MRCP include a diffusely enlarged or sausage-like pancreas, delayed pancreatic enhancement, a beaded appearance of the pancreatic duct, or strictures of the pancreatic or biliary ducts. AIP responds well to steroids, with Type 2 in particular having almost no relapse post-treatment, and does not impact long term survival.

Conclusion: Our patient’s fever, jaundice and abdominal pain were due to his AIP with associated obstructive cholangitis. He responded well to steroids, and after two weeks his pancreatic enzymes had normalized and his abdominal pain had almost completely resolved. While AIP is quite rare in children, this case emphasizes the importance of including AIP in the differential diagnosis for children with pancreatitis, as most patients do well after receiving steroids with complete resolution of symptoms.

Resources:

References:
Sah RP, Chari ST, Pannala R, et. al. Differences in clinical profile and relapse rate of type 1 versus type 2 autoimmune pancreatitis. Gastroenterology 2010; 139-140

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A109
Title: Acute Pancreatitis with Prolonged Feeding Intolerance

Authors: Anya Kleinman, Pediatric Hospitalist, Rainbow Babies and Children’s Hospital

Case Presentation: 12 year old Flu A (+) female admitted with 1 week of vomiting and abdominal pain. She was admitted for IV fluids due to inability to tolerate all oral intake. Admission labs included: bicarbonate-32, chloride-90, albumin 3.3, CRP-25.1 with no leukocytosis on CBC, and lipase-860. On day #3, she was transferred for further care to the Pediatric GI team given persistent acute pancreatitis (AP) and evolving hypoalbuminemia. CT abdomen was significant for several loops of non-dilated small bowel with mild mesenteric stranding adjacent to the terminal ileum. She was maintained on TPN with lipids after PICC placement given longstanding enteral feeding intolerance. Endoscopy and colonoscopy were performed following an additional week of persistent vomiting despite improving pancreatic enzymes. The studies were consistent with inflammatory bowel disease; patient showed significant clinical improvement after initiation of IV solumedrol. She was discharged on oral steroids, SQ Methotrexate, oral contraceptives, and Vitamin D repletion.

Discussion: Several studies have found an association between AP and new diagnosis of IBD. Retrospective examination of IBD patients has demonstrated that approximately 2% of pediatric IBD patients initially presented with AP; this is over 30 times more common than in adult patients. Development of IBD followed AP by a median of 24 weeks. It is important to consider IBD on the differential given the significant overlap in laboratory results between AP and IBD- including elevated inflammatory markers and hypoalbuminemia- that can complicate the final diagnosis.

Conclusion: Inflammatory bowel disease should be considered on the differential for acute pancreatitis in an otherwise healthy child with no clear triggers for pancreatic inflammation, especially in children with prolonged abdominal pain and/or feeding intolerance.


Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A111

Title: Seeing Double: A Readmission Leads to a Rare Diagnosis

Authors: Sumeet Banker, MD MPH, NYP Morgan Stanley Children’s Hospital/Columbia University
Patricia Hametz, MD MPH, Children’s Hospital at Montefiore

Case Presentation: The patient is a 3 year-old healthy boy who presented for readmission with one day of crossing of the eyes. His course began 7 weeks prior with left acute otitis media complicated by mastoiditis and otorrhea, treated with tympanostomy tube placement and IV then oral amoxicillin-clavulanate and ciprofloxacin/dexamethasone otic drops. He subsequently developed palsy of the 7th cranial nerve as well as headache and facial pain. He was started on a steroid taper and linezolid was added. He improved and was discharged to complete the course. However, he was readmitted with a new complaint of double vision. Examination revealed new esotropia with lateral gaze palsy of the left eye, consistent with cranial nerve VI palsy. MRI demonstrated enhancement of the left petrous apex, subadjacent prevertebral soft tissues, and clivus, suggestive of petrous apicitis/osteomyelitis. Findings were confirmed on bone scan and gallium scintigraphy. The diagnosis of Gradenigo’s syndrome was made. The patient was discharged on an extended course of meropenem and symptoms completely resolved.

Discussion: In 1907, Giuseppe Gradenigo described a constellation of three findings: 1) suppurative otitis media, 2) abducens nerve (cranial nerve VI) palsy, and 3) pain in the distribution of the trigeminal nerve (cranial nerve V). Now known as Gradenigo’s syndrome, it classically originates from infection in the inner ear which spreads to the mastoid bone. Subsequent inflammation at the petrous apex of the temporal bone, known as petrous apicitis, results in cranial nerves deficits. Potentially fatal complications include meningitis, intracranial and deep neck abscess, and cavernous sinus thrombosis. Once a surgical diagnosis, prompt medical management with antibiotics and steroids is typically effective.

Though streptococcal and staphylococcal species are known causative organisms, petrous apicitis is most often caused by Pseudomonas aeruginosa. This may explain why our patient did not initially fully respond to typical antibiotics. Once petrous apicitis was identified, antibiotics were switched to cover this pathogen and symptoms improved.

Conclusion: Gradenigo’s syndrome is a rare and potentially life-threatening complication of otitis media and is an unusual finding in the era of antibiotics. Though our patient manifested each of these findings over the course of his illness, it is reported that only about 42% of patients present with the classic triad of otitis media, diplopia, and...
headache. Rapid identification and medical treatment are critical to avoid complications and obviate the need for surgical intervention.

Resources:

References

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A112

Title: Utility of Newborn Screening in Diagnosis of Omenn Syndrome in a Newborn

Authors: Nikita Goel, MD, Rainbow Babies and Children’s Hospital, Cleveland Medical Center, Cleveland, Ohio
Hemangini Bhakta, MD, Rainbow Babies and Children’s Hospital, Cleveland Medical Center, Cleveland, Ohio, Lukasz Weiner, MD, Rainbow Babies and Children's Hospital, Cleveland Medical Center, Cleveland, Ohio

Case Presentation: A 36 5/7 week old neonate was admitted to the NICU with collodion appearance, respiratory failure, hypoglycemia, and hypothermia. She was started on empiric antibiotic and antifungal treatment. On physical examination, she had diffusely erythematous, shiny skin with peeling and weeping and alopecia totalis. Labs were significant for a WBC count of 12.4 x10E9/L with eosinophilia (3.5 x10E9/L), thrombocytopenia, and elevated IgG level. Differential diagnosis included neonatal sepsis, inherited ichthyosis, hydrops, and staphylococcus scalded skin syndrome. On day of life 5, newborn screen showed decreased amount of T cell receptor excision circle (TREC), suggesting severe combined immunodeficiency (SCID). Flow cytometry revealed low CD3+ (T cell marker), absent CD19 (B cell marker) and normal natural killer (NK) cells. The abnormal T cell maturation study indicated that the patient’s T cells were entirely maternal memory T cells with no naïve T cells present. Based on constellation of clinical and laboratory findings, our patient was diagnosed with Omenn syndrome.

Discussion: Omenn syndrome is a rare form of SCID characterized by erythroderma, alopecia, failure to thrive, chronic diarrhea, pneumonitis, hepatosplenomegaly, lymphadenopathy, eosinophilia, and elevated serum IgE levels (1). It is most commonly autosomal recessive and caused by mutations in recombination activating gene (RAG) 1 or 2, which affects VDJ recombination in lymphocytes. While our patient had dermatologic and diagnostic laboratory findings, she did not exhibit other signs of Omenn syndrome given early presentation. There was no family history of SCID or immunologic conditions. Omenn syndrome is fatal without definitive treatment with hematopoietic stem cell transplant (HSCT) (2). Due to increased risk of infection, patients without HSCT most commonly die of pneumonia and sepsis.
between 2 and 6 months of life (3). HSCT for SCID before 3.5 months of age offers a 5-year survival rate of 94% (4). Our patient developed CMV viremia without evidence of disease, and is scheduled for HSCT at 4 months of age.

**Conclusion:** Given high risk of infection and mortality, timely diagnosis, management, and treatment are necessary for optimal outcomes. In the critically ill neonate, Omenn syndrome can resemble a number of other conditions and thus has potential to result in misdiagnosis or late diagnosis in early phase. The newborn screen tests for SCID via TREC analysis (5). As of October 2016, 42 states in the US have implemented newborn screening of SCID (6). TREC analysis is inexpensive compared to the monetary equivalent of deaths and complications of disease (7). Diagnosis of SCID occurs sooner with newborn screening, allowing for HSCT at an earlier age, which has been associated with improved survival and outcomes (8). Newborn screening in our case allowed for timely diagnosis, preventative measures against other opportunistic infections and complications, and early preparation for treatment with HSCT.

**Resources:**

**References**


**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A113

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**Title:** Common Gastrointestinal Illness with Uncommon Neurological Sequelae

**Authors:** Hayley Redmond, DO, Advocate Children's Hospital - Park Ridge
Lauren McClure, DO, Advocate Children's Hospital - Park Ridge
Heidi Greening, DO, Advocate Children's Hospital - Park Ridge

**Case Presentation:** A healthy 3 year old F was admitted with 4 day history of emesis and diarrhea, generalized tonic clonic seizure, and acute altered mental status. Initial labs unremarkable. After admission, she continued to have seizure activity and worsening mental status. Lumbar puncture revealed CSF pleocytosis and analysis consistent with viral process. Antibiotics and antivirals were empirically started. Extensive infectious workup (from both blood and CSF) which included all typical pathogens associated with encephalitis was negative. The patient continued to be encephalopathic with severe aphasia, apraxia, and hypotonia without improvement. Differential was broadened to include autoimmune and metabolic etiologies, for which workup was negative and trial of steroids was ineffective. Stool studies were later sent due to continued diarrhea, revealing positive Norovirus group II. CSF was unable to be tested for Norovirus. The patient completed antibiotic course without improvement. She required intensive therapy and rehabilitation but eventually returned to baseline.
Discussion: Norovirus is a common pathogen causing gastroenteritis. Gastroenteritis caused by norovirus is usually self-limited. In this case, the patient suffered from encephalopathy associated with Norovirus gastroenteritis. Encephalopathy is a rare complication of viral gastroenteritis and has been well described in rotavirus infections but has not commonly been associated with norovirus. There are several case reports of similar presentations in patients of similar age (1). Unlike in our case, in 2 other cases CSF was able to be tested for Norovirus PCR and returned positive (1,3). In the literature there have only been 2 case reports with proven positive viral genome from CSF (1,3). There were also cases where CSF was either not tested or negative for viral genome, but CSF did show pleocytosis (2). It is possible that encephalopathy resulted from inflammatory response in the CSF rather than actual viral penetration. Several cases also reported a poor neurologic outcome similar to our patient, who suffered from severe functional impairment as a result of her illness (2).

Conclusion: In this case, our patient had a rare complication from a common pediatric illness. Serious neurologic sequelae with Norovirus is not well documented and testing of CSF for norovirus is not a common practice. Patients presenting with altered mental status and new neurologic symptoms undergo an extensive and costly workup to determine infectious or autoimmune etiologies. In many cases, workup is negative. When there is a clear history of preceding viral gastroenteritis, Norovirus should be included in the differential. To avoid additional and costly testing, it may be beneficial to include Norovirus in the initial infectious workup. Given the extensive workup, severe functional impairment and prolonged course for this patient, further investigation into the association of norovirus and encephalopathy may benefit similar patients.

Resources:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A114

Title: 12-year old presenting with acute vision loss after remote mild head trauma
Authors: Shiyu Sherry Bai-Tong, MD, Rainbow Babies and Children's Hospital
Nicholas Kucher, MD, Rainbow Babies and Children's Hospital

Case Presentation: AH is a 12-year old previously healthy boy presenting with acute vision loss. He had visited the ED several times after falling at a water park two weeks earlier and hitting his head. He endorsed headaches, neck pain, intermittent blurry vision, anxiety, anhedonia, and behavioral changes and was diagnosed with a concussion. His workup included a CT-spine and head and MRI c-spine, all were normal. He had been evaluated and cleared by neurosurgery and psychiatry consult had suggested somatization. Over the several days preceding this visit, the patient developed worsening to complete vision loss. He presented to an optometrist who noted right ptosis, sluggish pupils, and optic nerve scarring. Ophthalmologic evaluation in the ED showed bilateral papilledema. MRI brain with MRV/MRA showed diffuse dural venous sinus thromboses. LP had elevated opening pressure and he underwent urgent lumbar drain placement and thrombectomy. Hematologic workup found heterozygous factor V Leiden and he was started on anticoagulation therapy. Unfortunately, patient is not expected to regain vision.

Discussion: What initially makes this case less typical is the history of relatively mild trauma. Dural venous sinus thrombosis (DVST) after head injury is more frequently related to skull fracture or intracranial hematoma and it is a rare presentation after mild close head injury such as this case. However, of more interest than the lack of severe head trauma is the prolonged course until diagnosis. While the headache and neck pain can be seen in DVST, the personality changes were more consistent with post-concussive syndrome. His repeated normal neurological exams and unremarkable imaging further lowered the concern of severe pathology. While case reports show CT findings as early as 36 hours after, our patient had a normal head CT one week after initial insult. Not until severe symptoms occurred was the DVST found on imaging. While work up and management was appropriate and the clinical picture was confounded by a diagnosis of concussion, the case demonstrates uncertainty in the progression of the pathology of DVST in mild head trauma.
Conclusion: We presented an unusual case of DVST after a mild closed head injury that presented with progressive personality change and acute vision loss. DVST, although uncommon after mild close head injury, is a diagnosis that cannot be missed. Although more often found in the immediate days after injury, it can follow a subacute clinical course occurring weeks after the initial insult. Symptoms can be mimicked or covered by comorbidities such as concussion or anxiety. Therefore, patients with persistent symptoms need close follow up and warrant further work up if clinical picture deviates from expected. Medical providers must maintain a broad differential when dealing with nebulous diagnosis such as concussion or psychiatric illness in this patient population. Further research on the progression of DVST in pediatric patients is warranted.

Resources:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A115

Title: A 5-Month-Old Female with Unilateral Lower Extremity Swelling
Authors: Christine Chang, MD, Children's Hospital Los Angeles

Case Presentation: A 5-month-old healthy female presented with two weeks of leg swelling below the left knee. Parents denied history of trauma, pain, fevers, weight loss, and easy bruising or bleeding. Family history was negative for cancer. Physical exam showed significant swelling below the left knee without tenderness, erythema, or warmth. The patient had normal vital signs, intact pulses and sensation, and no hepatosplenomegaly or lymphadenopathy. She moved all extremities equally with normal range of motion and strength. Labs revealed leukocytosis, thrombocytosis, and elevated inflammatory markers. X-rays preliminarily showed periosteal reaction involving the left tibia, fibula, and multiple ribs. Differential diagnosis included bone tumors and cysts, fracture, non-accidental trauma, and osteomyelitis. Upon further review of the x-rays, multiple specialists agreed it showed marked cortical hyperostosis of the mandible, ribs, left tibia and fibula, diagnostic of infantile cortical hyperostosis (Caffey disease). Treatment included observation without need for medical or surgical intervention.

Discussion: Caffey disease is a rare, self-limiting disorder that presents prior to 6 months of age. It is characterized by irritability, soft tissue swelling, and cortical bone thickening. Its diagnosis may be difficult because of associated nonspecific symptoms such as fever. Swelling usually occurs suddenly and laboratory tests may reveal leukocytosis, thrombocytosis, and elevated inflammatory markers. Diagnosis is made by physical exam and radiographic findings. Though imaging may be variable during the clinical course, the classic finding is cortical diaphyseal periosteal new bone formation. Involved bones typically include the mandible, clavicles, ribs, and long bones. The involvement of the mandible – the most frequently affected bone – is usually pathognomonic.

Caffey disease usually resolves within 6-12 months. Treatment typically consists of analgesics as needed. In rare cases, it can cause enough long-term bony deformity that surgical correction is pursued. Other complications include exophthalmos, pleuritis, and recurrence of disease.

Conclusion: Caffey disease is a rare, self-limiting condition that should be included in the differential diagnosis of soft tissue swelling in infants. Delayed diagnosis may occur because the disorder can present with nonspecific symptoms, and therefore can mimic neoplastic, traumatic, and infectious processes. While the differential can be broad, many other conditions can be ruled out by age of presentation and distribution of bone involvement. With good clinical suspicion and multidisciplinary input, early identification of Caffey disease based on physical exam and radiographic imaging can prevent unnecessary work-up and invasive procedures.

Resources:
Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A116

Title: A Shocking Presentation of Kawasaki Disease
Authors: Rebecca Dang, MD, Pediatrics, Kaiser Permanente Oakland Medical Center, Oakland, California
           Amelia Castro

Case Presentation: A 10 year old boy presented with three days of fevers, peri- and intra-oral blisters, macular rash/papules on trunk and extremities, and tender cervical lymphadenopathy. Upon admission, he developed capillary leak with subsequent pleural effusions, ascites, and hypotension, prompting transfer to the PICU for respiratory support, fluid management, and pressors. Echo was normal. Broad infectious workup including pan-cultures, RVP, enterovirus, HSV, and strep PCR was negative. Significant labs were normocytic with 45% bands, Hb 7.6, Na 135, albumin 2, coags with elevated d-dimer and PTT, 21.1 CRP, and ferritin>3000. HLH ruled-out on bone marrow biopsy. Skin biopsy showed eosinophils, which can be consistent with Kawasaki Disease (KD). Patient received IVIG with noticeable improvement after. He also received clindamycin for possible Toxic Shock Syndrome given rash and hypotension. Patient was discharged on low-dose aspirin. At his one month follow up with cardiology, repeat echo was again normal, deeming KD prognosis excellent. Aspirin was discontinued with follow-up as needed only.

Discussion: Kawasaki Disease (KD), a medium-sized vasculitis, is a leading cause of acquired pediatric heart disease, primarily coronary artery aneurysms (CAA). As with this patient, 5-7% of KD presents more acutely, with systolic hypotension and/or poor perfusion, now recognized as Kawasaki Disease Shock Syndrome (KDSS). In contrast to KD, KDSS by definition is associated with shock. Patients always require aggressive fluid resuscitation, and often also receive pressors. Higher CRP and bandemia, worse anemia, hyponatremia, and hypoalbuminemia, and a consumptive coagulopathy are seen with KDSS. Cardiac abnormalities are more frequent and extensive, expanding beyond CAA to ventricular and valvular dysfunction. This is due to severe and prolonged inflammation. Given normal bone marrow, this patient’s ferritin supports such severe inflammation. Further delaying improvement, IVIG resistance is common in KDSS, often requiring repeat doses and other immunosuppressive medications. Fortunately, if treated earlier, derangements of vitals, labs, and echos tend to improve promptly without sequelae.

Conclusion: KDSS should be considered early in the presentation of a patient with fever, rash, and shock as early intervention is critical in decreasing coronary artery abnormalities and mortality. The differential diagnosis is wide, with Toxic Shock Syndrome (TSS) as a common misdiagnosis. In addition to obtaining lab work to reflect significant inflammation, an echocardiogram to search for ventricular dysfunction, mitral valve regurgitation, and especially CAA should be obtained for further diagnostic support of KDSS. As in this patient, it is often challenging to rule-out KDSS or TSS in the setting of a negative infectious workup and normal echocardiogram. Lack of initial response to IVIG commonly seen in KDSS further challenges the diagnosis of KDSS. This case highlights the importance of recognizing and treating KDSS early, even if management with both anti-inflammatory medications and antibiotics for KDSS and TSS, respectively, need to be instituted. A delay in diagnosis and intervention the setting of KDSS increases the risk for cardiac defects and death.

Resources:
References

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A117

**Title:** A Toddler with Tremor

**Authors:** Rajesh Sood, MD, Pediatric Resident, Inova Children's Hospital
Kamilah Haltmon, MD, Pediatric Hospitalist, Inova Children's Hospital

**Case Presentation:** A 2-year old previously healthy female presented with a 1-month history of progressive ataxia and tremors, now with inability to ambulate.

Initially, the tremors occurred only upon awakening. Over the course of the month prior to admission, the tremors became constant. The tremors were more pronounced with movement. She was initially unsteady with walking but on admission was no longer able to ambulate. Her mother reported that she was drooling; however, she did not have any difficulty breathing. She was speaking quieter than usual and was more irritable over the past two weeks. Her neurologic symptoms were preceded by a viral upper respiratory infection.

She was an otherwise healthy child with no significant past medical history. She was born full-term and met all developmental milestones on time. Apart from a maternal history of migraines, but there was no family history of any neurological conditions or other problems.

She underwent evaluation by a neurologist as an outpatient which included a physical exam remarkable for tremor and ataxia and a normal MRI of her brain.

**Discussion:** She was diagnosed with an essential tremor and referred to physical therapy. Her physical therapist noticed worsening tremor and ataxia over a 2-week period and advised she go to the ER where she was admitted to the hospital for further work-up.

Her admission physical exam was notable for myoclonic jerks in all four of her extremities. These jerks worsened with movement and appeared more pronounced in her lower extremities. She had no facial asymmetry. Cranial nerves two
through twelve were intact, but she was noted to have rapid upward jerking of both eyes. She was unable to ambulate due to ataxia. She had down going plantar reflex bilaterally, and patellar and Achilles deep tendon reflexes were normal. Laboratory testing was performed including complete blood count, comprehensive metabolic panel, and inflammatory markers. A lumbar puncture was performed to evaluate cell count, protein, glucose as well as cerebrospinal fluid neurotransmitters and oligoclonal bands. She underwent MRI of brain and spine. All of these results were normal, but her neurologic dysfunction persisted. **Conclusion:** After thorough review of her case with neurology, she was ultimately diagnosed with opsoclonus-myoclonus syndrome. She was evaluated by the pediatric oncology team due to the strong correlation between opsoclonus-myoclonus syndrome and neuroblastoma. She underwent an abdominal ultrasound, chest xray, CT scan of abdomen and pelvis, and MIBG scan to evaluate for malignancy. Urinary catecholamines were also assessed. All results were negative, and neuroblastoma was removed from the differential diagnosis. She then began treatment for opsoclonus-myoclonus syndrome. Initial treatment consisted of intravenous immunoglobulin and intravenous steroids followed by oral steroids for one month. The initial treatment was unsuccessful; therefore, she was started on a monoclonal antibody, infliximab, in addition to intravenous immunoglobulin and intravenous steroids. Her symptoms improved with combination therapy and she was close to her neurologic baseline after three to four months. **Resources:** N/A

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A118

**Title:** Blood really is thicker than water: a surprising case of neonatal oliguria

**Authors:** Melissa Villars, MD, UNC Children’s Hospital
Erin Sukhu, MD, UNC Children’s Hospital, Patrick Taus, medical student, UNC School of Medicine

**Case Presentation:** A term 36-hour-old SGA female presented with oliguria. APGARs were 8 and 9. Mother had hypertension and diabetes but normal ultrasounds; she received magnesium for preeclampsia and antibiotics for GBS. Infant had delayed cord clamping. In first 36 hours, she had five stools, emesis, and oliguria so was transferred to floor. Vitals and exam were normal on transfer. Weight was down 11% from birth so IV fluids were given. Creatinine was 1.14 (maternal 0.57), with normal renal US/Dopplers and UA. Ammonia, urine organic acids, abdominal and chest X-rays were normal. Repeat labs DOL3 revealed bicarbonate of 11 but pCO2 of 28; due to floor protocol no further labs could be obtained for 24 hours to address discrepancy. Head US showed no bleeding and UGI no malrotation as causes of emesis. Echocardiogram showed normal anatomy and systolic function.

On DOL4, a CBC was finally obtained, indicating polycythemia with hematocrit 67.5. Fluids were continued and urine output, feeds, and labs normalized. Bilirubin remained normal. Infant was back to birth weight DOL6 and discharged next day.

**Discussion:** Neonatal oliguria’s differential includes obstructive uropathy, intrinsic renal failure, or most commonly, prerenal failure, characterized by inadequate renal perfusion/prerenal azotemia. This can be precipitated by hypovolemia, shock, myocardial failure, or as in this case, by polycythemia/hyperviscosity.

Neonatal polycythemia is a venous hematocrit >65%; incidence is 0.4-5.0%. The consequence is hyperviscosity, which presents as nonspecific symptoms and leads to decreased organ perfusion. Oliguria is a rare finding (incidence 0.4-0.9%). Though some renal perfusion is maintained via vasodilation, hyperviscosity decreases GFR due to decreased renal plasma flow, and can lead to oliguria.

Risk factors of polycythemia include being SGA and term, maternal hypertension, diabetes, and preeclampsia. Delayed cord clamping has negligible increased risk of polycythemia, but little data exist for the SGA population. There is research on plasma exchange transfusion for treatment but no demonstrated benefit in mild cases. Studies assessing its role in severe cases are lacking.

**Conclusion:** With the infant’s fluid resuscitation prior to diagnosis and physiologic neonatal hematocrit trends, the hematocrit on DOL4 was likely deflated and the initial polycythemia/hyperviscosity severe. There was emesis in addition to oliguria, and multiple risk factors for polycythemia, but diagnosis was delayed by low clinical suspicion. Oliguria dictated initial workup; renal perfusion was assessed via renal Dopplers and an echocardiogram with normal function, but there was no consideration of effects of potential hyperviscosity on renal
filtration or to oliguria as a possible sign of polycythemia. After primary renal disease was ruled out, workup unfortunately shifted toward GI/metabolism due to emesis and acidosis.

Providers should recognize polycythemia as a cause of neonatal oliguria, particularly in infants with risk factors such as SGA status, delayed cord clamping, and maternal diabetes and hypertension. Though protocol delayed a CBC in this case, higher suspicion of polycythemia would have prompted a screening heel stick, expedited diagnosis and limited extraneous workup.

**Resources:**

References:

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A119

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**Title:** Femur Mass in a Patient with HIV and Poor Medication Compliance

**Authors:** Laura Finley, MD, Our Lady of the Lake Children's Hospital. Emily Klepper, MD, Our Lady of the Lake Regional Medical Center, Rebekah Dickman, Pediatric Resident, Our Lady of the Lake Children's Hospital; Jeffery Deyo, MD, PhD, St. Jude, Our Lady of the Lake Children's Hospital

**Case Presentation:** The patient was an 11 year old male who presented with fever, abdominal pain and right thigh pain affecting his gait. Work-up showed mild elevation of ESR and CRP. Plain films and a CT scan demonstrated a small fibrous cortical defect. An MRI was suggestive of a Brodie’s abscess (subacute osteomyelitis). Incision and drainage of the lesion returned a small amount of seropurulent material, which was negative for infection or malignancy. The patient was discharged from the hospital with four weeks of Clindamycin therapy. Two months later, the patient returned with worsening leg swelling and pain. Repeat MRI demonstrated pronounced periostitis, a large abscess and cortical erosion. Pathology of bone and tissue showed malignancy and extensive necrosis. The patient was subsequently diagnosed with Burkitt’s non-Hodgkin’s lymphoma. Following referral to Pediatric Oncology, he began chemotherapy using a standard regimen for his disease. Concurrently he was maintained on HIV medical therapy with the Pediatric Infectious Disease service. He has since completed treatment for his cancer.

**Discussion:** Burkitt’s lymphoma was first reported in Eastern Africa as childhood facial tumor and subsequently classified as a non-Hodgkin’s type lymphoma. Since the emergence of the Human Immunodeficiency Virus (HIV) infection, an association has been noted between Burkitt’s lymphoma and HIV. Three subtypes have been identified: endemic, sporadic or AIDS related. In United States, sporadic and immunosuppression-related cases have been reported. Patients usually present with an abdominal mass and generalized “B symptoms”. We present a patient with a history of congenital HIV infection and poor medication compliance, who developed Burkitt’s lymphoma with an atypical presentation. As our case illustrates, it is important to consider the immunosuppressed status of a patient with HIV as well as medication compliance when evaluating vague complaints. Although Burkitt’s lymphoma can present with lytic lesions, this is an uncommon initial finding. Presentation of this form may be subtle, and there are previous reports in the literature of Burkitt’s lymphoma masquerading as an abscess – as was seen with our patient. The discerning clinician
should be wary when considering common diagnoses such as infection in the immunocompromised patient. Malignancy must be carefully considered and abnormal presentations should not be forgotten in these individuals.

Resources: N/A

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A120

Title: What Swollen Gums and Genital Sores Have In Common

Authors: Victoria Chu, MD, Pediatrics, Kaiser Permanente Oakland Medical Center, Oakland, California
Rebecca Dang, MD, Pediatrics, Kaiser Permanente Oakland Medical Center, Oakland, California, Eva Padilla, MD, Pediatrics, Kaiser Permanente Oakland Medical Center, Oakland, California

Case Presentation: An 11 year old female was admitted for worsening chronic genital ulcers despite antifungal and antibiotic treatment. She endorsed mild weight loss, gingival hyperplasia of 5 years, and intermittent non-bloody loose stools. Denied sexual activity or abuse. On physical exam, there were multiple vulvar lesions bilaterally and one perianal lesion.

The differential diagnosis included Lipshutz ulcer, lymphogranuloma venereum, pyogenic infections, Crohn’s disease (CD), sarcoidosis, syphilis, and hidradenitis suppurativa. Lab results were inconsistent with a primary infectious etiology for the vulvar lesions. She had a negative GC/CT, syphilis, and HSV. ESR was mildly elevated with a normal CRP. Rheumatology labs (ACE, HLAB27, and ANCA) were negative except for ANA. Colonoscopy and upper endoscopy was grossly negative. Ultimately, the intestinal and vulvar biopsies showed noncaseating granulomatous lesions consistent with CD.

The patient was treated with topical tacrolimus and oral prednisone. At her one month follow up, the genital lesions had resolved and gingival hyperplasia were improv

Discussion: CD is a granulomatous inflammatory bowel disease that can affect any part of the gastrointestinal tract (GI) from the mouth to the anus. 20-40% of patients will present with extraintestinal symptoms as well. Interestingly, this patient presented with minimal GI symptoms and only metastatic lesions of the gingiva and vulva. Metastatic Crohn’s disease (MCD) can present with isolated swelling or ulcers of the oral cavity and vulva. Duration between appearance of MCD and development of intestinal CD still needs to be elucidated as 25% of patients with MCD do not have accompanying GI symptoms at presentation. Maintaining a high index of suspicion for CD in patients with oral or genital lesions is important, and there should be a low threshold for endoscopy in those with even mild GI symptoms. In cases of MCD, the oral and vulvar lesions themselves serve as a separate biopsy site. The histopathology is similar to that of intestinal CD (transmural inflammation, neutrophilic infiltration, patchy architectural distortion, and non-caseating granulomas) and can confirm the diagnosis.

Conclusion: Although Crohn’s disease typically presents with gastrointestinal symptoms, it should still be considered when evaluating children presenting with persistent isolated mucocutaneous findings such as oral ulcers, gingival hyperplasia, or vulvar lesions. In these cases, a thorough history regarding any unreported gastrointestinal or extraintestinal symptoms should be taken as well as an examination of the rectal and genital mucosa. If there are symptoms or signs for MCD, additional workup should be pursued. However, as this case demonstrates, CD can present with minimal gastrointestinal symptoms, negative stool WBCs, negative calprotectin, and grossly normal intestinal mucosa on endoscopy. The diagnosis was eventually made via intestinal and vulvar histopathology. Of note, had the oral lesions been biopsied earlier, she may have been diagnosed with CD years prior. This case highlights the challenges of diagnosing CD in children presenting with isolated mucocutaneous lesions, leading to delayed diagnosis and inappropriate management.

Resources:
REFERENCES

SUPPLEMENTAL FIGURES:

Vulvar ulcers seen at presentation to the hospital
Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A121

**Title:** “Gang”ing up on the skin: An 11-month-old with ulcerative skin lesions

**Authors:** Laura Cannon, M.D., University of North Carolina Children's Hospital
Eric Zwemer, MD, University of North Carolina School of Medicine, Eveline Wu, M.D., University of North Carolina Children's Hospital

**Case Presentation:** A previously healthy 11-month-old female was admitted to the hospital with ulcerative skin lesions. Her pediatrician had trialed topical and oral antibiotics for 4 weeks without improvement. Exam revealed diffuse well-circumscribed erythematous papules and pustules of varying sizes on her face and extremities intermixed with erosions with central crusting and violaceous borders (Figures 1-3). Differential diagnosis included atypical mycobacteria infection, nodular vasculitis, T-cell cutaneous lymphoma, and infantile pyoderma gangrenosum (IPG). Labs revealed a leukocytosis with neutrophilia and skin biopsy was consistent with IPG. Lesions improved with oral steroids. She was discharged but returned 2 weeks later with worsening pain and new lesions. She was admitted again for IV steroids and later started on Infliximab therapy. Evaluation for underlying disorders included normal immunoglobulin levels, blood smear, LDH, uric acid, diphtheria and tetanus antibody titers, leukocyte adhesion deficiency panel, and neutrophil function test. To date, no other conditions have been diagnosed.

**Discussion:** Infantile pyoderma gangrenosum (IPG) is very rare with less than 20 cases reported in the literature. It is a neutrophilic dermatosis that presents as ulcerations of the skin with inflammation. Infants diagnosed with IPG warrant further work-up for associated systemic diseases including inflammatory bowel disease, Takayasu’s arteritis, leukocyte adhesion deficiency, and chronic granulomatous disease. Most cases, however, are idiopathic—though it can be difficult to rule out an associated condition, given skin findings may precede other systemic symptoms by several years. At this time, standardizing treatment is challenging given the rarity of IPG, but includes immunomodulation with steroids and/or biologic agents.

**Conclusion:** Though rare in infants, pyoderma gangrenosum should be considered in the differential diagnosis for an infant with diffuse ulcerative skin lesions.

**Resources:**
Case Presentation: 13 y.o. male presents for evaluation of 2 weeks of left hand paresthesia, “funny sensation” bilaterally in lower extremities, and 1 week of shoulder & neck pain. Physical examination was notable for decreased left hand grip strength. A head CT angiogram was negative but MRI of the cervical spine showed T2 hyperintensity & expansion of the cord from C1-C7.

A lumbar puncture was performed and CSF was significant for elevated protein, IgG, and albumin. The remainder of infectious/inflammatory disease workup was negative. He was presumptively diagnosed with Transverse Myelitis, treated with IVIG, and given 5-day course of steroids with notable improvement in symptoms. The patient was readmitted within days of completing the steroid course with worsening symptoms. Repeat MRI of the cervical spine revealed diffuse expansion of the cervical spinal cord. Lumbar puncture was repeated and CSF cytology was negative. He underwent cervical spinal biopsy of the lesion and pathology showed a high-grade diffuse infiltrating glioma consistent with diffuse midline glioma of the spinal cord.

Discussion: Intramedullary spinal tumors are primary spinal cord lesions that arise from glial cells in the spinal cord. Spinal cord tumors are extremely rare in children, representing only 1% of all CNS tumors. Approximately one-third of these spinal tumors are intramedullary. Of those that are intramedullary, high grade gliomas only account for 1-3%. Despite advancements in therapy high grade diffuse gliomas are a challenging diagnosis with ultimately poor prognosis. This case proved to be challenging as the acute onset of symptoms and clinical presentation initially seemed consistent with an inflammatory process. However, with progression of symptoms, further diagnostic workup was necessary. Given the significant risks of a cervical spine biopsy, the least invasive diagnostic studies were completed initially. This included collaboration and consultation with national oncology centers with expertise in primary CNS tumors. Ultimately, a cervical spinal biopsy was necessary as pathology confirmed the diagnosis.

Conclusion: Although primary spinal tumors are extremely rare in pediatric populations, patients who present with multiple neurological complaints including motor and sensory deficits should raise a high index of suspicion for a CNS tumor. Clinicians should be aware that the differential also includes inflammatory or autoimmune processes including multiple sclerosis, neuromyelitis optica, and transverse myelitis, in addition to vascular lesions. When symptoms don’t improve, continued re-evaluation is always necessary and should include a multidisciplinary team of pediatric hospitalists, neurologists, hematologist/oncologists, and neurosurgical specialists. A family-centered approach is also crucial to ensuring the patient and family understand the differential diagnosis and the plan for diagnostic evaluation. Pediatric palliative and supportive care consultation was integrated early into the care management plan, and proved invaluable as the poor prognosis for the confirmed diagnosis became evident.
Resources:
Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A123

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**Title:** A Superficial Presentation of a Deeper Diagnosis

**Authors:** Angela Dietrich-Kusch, MD, Riley Hospital for Children at Indiana University Health
Sarah Puffer, MD, Indiana University School of Medicine, Laurie Wilkie, MD, Indiana University School of Medicine

**Case Presentation:** 14 year old female with history of IBS presented with bilateral shin “abscesses.” Weeks prior, she noted a small painful papular lesion on her right shin that grew in size; she then developed similar lesions on her left shin. Lesions worsened despite treatment with Keflex, an I&D, and Bactrim. On admission, she denied fevers, fatigue, joint pain, trauma, or change in stools. Exam revealed a draining, violaceous, bullous lesion on right leg and two intact bullae on left leg (Figs 1&2). Labs revealed mild anemia and thrombocytosis with elevated ESR and CRP. Culture of fluid was negative. Dermatology was consulted, and skin biopsy showed neutrophilic infiltration of the subcutis suggestive of atypical infection or other neutrophilic panniculitis. Lesions appeared to improve on clindamycin and ciprofloxacin. After discharge the fecal calprotectin returned markedly elevated. The patient was referred to GI, and endoscopy/colonoscopy revealed erosions and aphthous ulcers in the recto-sigmoid colon. Pathology confirmed chronic active colitis, thought to most likely represent Crohn’s disease.

**Discussion:** The differential diagnosis was initially quite broad, including abscess, erythema nodosum (EN) and pyoderma gangrenosum (PG). Despite improvement on antibiotics, the calprotectin level and colon pathology ultimately led to a dermatologic manifestation of IBD as the diagnosis. While GI symptoms are the hallmarks of IBD, extraintestinal manifestations can be the only presenting symptoms. After the GI tract, the skin is one of the most frequently affected systems, with the two most common lesions being EN and PG. These conditions are especially rare in pediatric patients, with EN affecting 4% and PG 0.75% of children with IBD. Our patient's skin pathology was not classic for either diagnosis (lacking the septal changes of EN and the dermal/epidermal findings of PG), but rather revealed neutrophilic panniculitis (NP). NP is a skin disorder caused by neutrophilic infiltration of the subcutaneous fat causing painful, indurated, subcutaneous nodules. NP is more often associated with alpha-1-antitrypsin deficiency and myelodysplasia but has rarely been linked with IBD in the literature.

**Conclusion:** Skin lesions in the pediatric population are frequently secondary to infection, and timely initiation of antimicrobial therapy is appropriate when the history and exam suggest an infectious process. However as this case highlights, it is important to maintain a broad differential diagnosis, particularly when the clinical course is unexpected. On presentation, our patient’s symptoms had been refractory to multiple appropriate antibiotic regimens. This unusual response to therapy inspired a broader work-up, without which the ultimate diagnosis of IBD would certainly have been further delayed. This case serves as an important reminder of the importance of investigating more deeply when what may appear to be a typical diagnosis responds to treatment in an atypical way. Sometimes it is not the treatment that needs broadening, but rather the differential diagnosis.

**Resources:**
References:
Allen CP, Burge SM. Extraintestinal Manifestations of Inflammatory Bowel Disease: Cutaneous and Oral Manifestations of Inflammatory Bowel Disease. Crohn’s Disease and Ulcerative Colitis. December 2011:611-630.
**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A124
Case Presentation: 12 mo old male admitted for developmental regression. Normal development until 9 months when he had otitis media and developed progressive hypotonia and motor regression to 3 mo milestones. Found to have elevated AST/ALT/GGT. GI, neurology, genetics, and metabolic evaluations inpatient revealed no clear etiology (Fig 1). NG tube placed after dysphagia study showed silent aspiration. Admitted twice in next 3 months for bronchiolitis. Aspiration pneumonia treated outpatient. Hospitalized again at 16 months for recurrent respiratory symptoms, persistent transaminitis and regression. Liver biopsy c/w nonspecific hepatocellular disease. Repeat dysphagia study confirmed aspiration. Gtube/fundo done, complicated by gastric perforation. 2 days later intubated for respiratory failure from presumed chronic aspiration and post-op ARDS. Treated with meropenem. Abdominal US showed ascites with possible abscess and developed pancreatitis. 2 weeks post-op, vesicular rash noted and was Varicella +. Had persistent pancreatitis and developed colitis. HIV testing sent and returned positive (Fig 2).

Discussion: Gross motor regression alone has a broad differential. We considered metabolic disease, leukodystrophy, neuromuscular disease, HIE, and TORCH infections. Mother had documented negative HIV testing during pregnancy, lowering our suspicion. This patient had several iatrogenic factors with perforation and aspiration, also confounding our workup. Pediatric HIV often presents with lymphadenopathy, oral candidiasis, and failure to thrive which our patient did not have. AIDS-defining conditions in children include Pneumocystis jirovecii pneumonia, recurrent bacterial infections, wasting syndrome, candida esophagitis, HIV encephalopathy, and cytomegalovirus (CMV). Our patient initially demonstrated unrecognized HIV encephalopathy alone, with persistent transaminitis. He eventually developed varicella, leading to the diagnosis, and was then found to have CMV. With HIV treatment, his development has normalized. The case highlights the need to recognize the broad range of HIV presentations and consider perinatal HIV when evaluating developmental regression despite negative prenatal testing.

Conclusion: This case represents a rare presentation of a treatable diagnosis, confounded by several complications and a large multidisciplinary workup. HIV encephalopathy as a treatable cause of developmental regression is especially important. Negative prenatal testing emphasizes the potential pitfall of lowering suspicion for HIV based on prenatal testing alone. The key discussion points include 1) the approach to evaluation of developmental regression and key differential, 2) described presentations of early childhood HIV, and 3) maintaining suspicion for HIV despite negative prenatal testing. Our case highlights how the “classic” findings can be masked or easily attributed to other complications, such as mild cytopenias presumed from secondary infections rather than primary HIV. This case would be presented chronologically, highlighting the evolution of each new symptom, piecing together separate clues leading to the diagnosis, and describing key unifying factors recognized in hindsight.

Resources:
Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A125

Title: Elevated Ketoacids and Branch Chain Amino Acids in a Hypoglycemic Patient
Authors: Samantha Heisler, DO, Jersey Shore University Medical Center
Jamie Pinto, MD, FAAP, Jersey Shore University Medical Center, Srividya Naganathan, MD, FAAP, Jersey Shore University Medical Center

Case Presentation: A 5 year old male presented to the Emergency Department (ED) with 3 days of abdominal pain, headache, nausea, vomiting, and increasing lethargy with ataxia. Mom reported a sweet smell to the breath. Parents reported 2 previous admissions for similar symptoms with a negative workup and rapid resolution of symptoms. In the ED, patient had moderate dehydration, diffuse abdominal tenderness, hyporeflexia and weakness of the lower extremities. Lab data revealed bicarbonate level 15, anion gap 20, glucose 57 and ketonuria. Workup of hypoglycemia and acidosis revealed a normal carnitine profile and hormone levels, and significantly elevated 3 OH Butyric Acid in both plasma and urine 13062 (0-4) suggestive of ketotic hypoglycemia. Incidentally, we found borderline elevation of branched chain amino acids (BCAA): Valine 383 (100-300), Leucine 237 (60-230), Isoleucine 170 (3-130). The diagnosis of intermediate variant Maple Syrup Urine Disease (MSUD) was considered. His clinical symptoms, acidosis, and hypoglycemia resolved with hydration.

Discussion: Hypoglycemia has a broad differential including endocrine causes, metabolic disorders, idiopathic and glycogen storage defects. As MSUD is not associated with hypoglycemia, it is likely an incidental secondary diagnosis that may explain the patient’s hyporeflexia and ataxia. MSUD has four variant types including the classic type (75%), intermediate, intermittent, and thiamine dependent. The classic type, usually detected on newborn screen, presents in the neonatal period with encephalopathy leading to coma and death if untreated. In intermittent MSUD, patients have normal development with elevated levels of BCAAs leading to encephalopathy during acute catabolic stress. The elevation of amino acids can be corrected with thiamine in those with thiamine-responsive MSUD. Intermediate MSUD can range from mild to severe based on the branched chain ketoacid dehydrogenase (BCKD) activity. This patient falls into the mild, asymptomatic category with borderline elevation of BCAA in the plasma and no urinary excretion.
Conclusion: Metabolic diseases are a challenging group of disorders that are rare and difficult to identify for the pediatric hospitalist since they present in a myriad of ways and mimic many of the more common inpatient diagnoses. To our knowledge, this is the first reported case of concomitant diagnoses of ketotic hypoglycemia and intermediate MSUD. Hospitalists should maintain a high index of suspicion for co-existing disorders in patients with atypical symptoms not explained by their presenting common diagnosis.

Resources: N/A
Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A126

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**Title:** Fever in a 21 month old: Acute Otitis Media with a Twist  
**Authors:** Yamini Krishnamurthy, M.D., Massachusetts General Hospital for Children  
Jing Ren, M.D., Massachusetts General Hospital for Children, Molly Wolf, M.D., Massachusetts General Hospital for Children

**Case Presentation:** A previously healthy 21-month old presented for evaluation of fussiness and fevers. Two weeks prior to initial presentation, she was diagnosed with acute otitis media (AOM), and treated with 5-days of amoxicillin with improvement. Seven days after initial presentation, she developed fevers to 102F; she was seen in the ED, diagnosed with AOM and discharged with amoxicillin. Three days later, she represented to the ED for persistent fevers and diarrhea. She was having pink, loose stools, which was attributed to amoxicillin. She was given IM ceftriaxone to complete her therapy for AOM and discharged.

Following discharge, the patient developed grossly bloody stools, jaundice, and lethargy with minimal PO intake. She was brought to her pediatrician’s office where laboratory tests revealed WBC 3.21, H/H 8.9/26.9, Plt 36, AST 2044, ALT 835, and LDH 5799. She was referred to the ED.

In the ED, she was febrile to 104.4F with HR 167, BP 93/42, RR 56, and O2 100% on RA. Her exam was notable for fatigue, diffuse lymphadenopathy, a distended abdomen with marked hepatosplenomegaly, and jaundice.

**Discussion:** Further labs were notable for CRP 89.9, D-dimer >100,000, INR 1.4, triglycerides 637 and ferritin 69275. Peripheral blood smear did not show cells consistent with hematologic malignancy; initial infectious work up was unrevealing. She was admitted to the pediatric ICU with a presumptive diagnosis of hemophagocytic lymphohistiocytosis (HLH), and started on treatment with dexamethasone. A bone marrow biopsy was performed, which showed left-shifted myeloid and erythroid precursors, and scattered hemophagocytic histiocytes (Picture 1), and which confirmed the diagnosis. She was initiated on etoposide, in addition to steroids, with clinic improvement within 24-48 hours. Lumbar puncture and brain MRI did not show evidence of CNS involvement. EBV DNA PCR was positive. At time of discharge (12 days after admission), her LFTs, cell counts and ferritin level were normalizing. Her genetic testing was negative.

**Conclusion:** HLH is an uncommon hematologic disorder that can present with non-specific symptoms such as persistent fever, cytopenias, and multiorgan dysfunction that mimic other diseases. Diagnostic criteria for HLH require molecular diagnosis or the presence of 5/8 following signs or symptoms: fever, splenomegaly, cytopenias, hypertriglyceridemia, hemophagocytosis, low or absent NK cell activity, elevated ferritin, and elevated soluble IL-2 receptor. A ferritin level over 10,000 with LFT abnormalities is especially classic. HLH usually presents in the first 18 months of life but can also present in utero and in adults. HLH can be primary (i.e. due to an underlying pre-described genetic disorder), or secondary. HLH is often associated with infections (often viral, of which EBV is most common), autoimmune disease, malignancy, and immunodeficiency. It is helpful to identify potentially treatable underlying disorders, to spare patients from toxic systemic treatment such as stem cell transplants. In acutely ill patients, treatment should not be delayed for immunologic or genetic testing.

Resources: N/A
Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A127

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**Title:** Hepatomegaly, Transaminitis, and Growth Failure in Type 1 Diabetes  
**Authors:** Elizabeth Halvorson, MD. MS., Wake Forest School of Medicine  
Jordan Klein, DO, Wake Forest Baptist Medical Center, Brenner Children's Hospital

**Case Presentation:** 15-year-old male with poorly controlled type 1 diabetes mellitus (DM) and psoriasis presented with hematemesis and chest/abdominal pain. He was hypoglycemic to 39 mg/dL and sleepy on presentation, both of which
normalized with intravenous dextrose. Exam was significant for tenderness to palpation across the upper abdomen, liver edge percussed 5-7 cm below the costal margin, lipodystrophy of the lateral abdomen bilaterally, and Tanner 2 genitalia. Labwork revealed transaminitis (ALT 604 IU/L, AST 712 IU/L) with normal liver synthetic function. Amylase, lipase, CBC, acetaminophen and salicylate level, EKG, troponin and chest X-ray were normal.

Growth chart showed weight and length less than 1st percentile and trending down, BMI less than 3rd percentile. Further work up showed negative/normal ANA, cortisol, adrenal and hepatic autoimmune panel, and hepatitis panel. Hemoglobin A1c was 10.1%. 24 hour urinary copper excretion was elevated; ceruloplasmin was normal. The transaminitis, hepatomegaly, growth failure and delayed puberty led to a presumed diagnosis of Mauriac syndrome.

**Discussion:** Mauriac syndrome is the constellation of heptomegaly, transaminitis, growth failure and delayed puberty in a patient with poorly controlled type 1 DM. Alkaline phosphatase and bilirubin are less often affected, and synthetic liver function is usually intact. Other lab findings may include dyslipidemia, as seen in our patient, and elevated lactate. The current proposed pathophysiology is that the chronic hyperglycemic environment in poorly-controlled patients leads to insulin-independent uptake of glucose by hepatocytes. When insulin is given, glucose is converted to glycogen, resulting in excessive glycogen storage, hepatomegaly, and elevated liver enzymes. Most children with poorly controlled diabetes do not develop severe glycogen deposition in the liver, and a possible genetic contribution has recently been identified. This defect in the gene for glycogen phosphorylase kinase may result in dysfunctional glycojenolysis and could explain the hypoglycemia seen on presentation in this patient.

**Conclusion:** Children with poorly controlled type 1 DM are at risk for end-organ damage, which may manifest as Mauriac syndrome. With improved glycemic control, both the transaminitis and hepatomegaly are reversible, although this may take weeks to years. Liver failure is rare. It is important to consider nonalcoholic fatty liver disease, autoimmune hepatitis, and Wilson’s disease in the differential. Children whose presentation is consistent with Mauriac syndrome may be able to avoid liver biopsy and other invasive testing, so clinician awareness of this disease is important. Additionally, knowledge of the reversibility of this condition is crucial for counseling patients and families to improve diabetes management.

**Resources:**

References


**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A128

**Title:** Influenza A Epiglottitis and Compensatory Pursed-Lip Breathing in an Infant

**Authors:** Shelease O'Bryant, MD, Baylor College of Medicine/Texas Children's Hospital

Brent Mothner, MD, Baylor College of Medicine/Texas Children's Hospital

**Case Presentation:** A healthy 3-month-old infant presented with 2 days of fever, cough, nasal congestion, and increased work of breathing. She had nasal flaring, retractions, stridor, equal and clear breath sounds, clear rhinorrhea, an erythematous oropharynx and pursed lips. White blood count was elevated, urinalysis was negative. Influenza PCR, respiratory syncytial virus PCR, blood culture and urine culture were completed.
Her symptoms waxed and waned with development of tachypnea, neck hyperextension, intermittent stridor and worsened pursed lip breathing. Neck radiography (Figure 1) revealed effacement of the epiglottic vallecula. Respiratory failure necessitated intubation revealing significant airway edema and swollen epiglottis without glottis visualization suggesting a diagnosis of epiglottitis. PCR was positive for influenza A. Oseltamivir was continued in her treatment. Her vital signs improved and increase work of breathing resolved. She was extubated following improvement in epiglottic and aryepiglottic fold edema. She completed her antimicrobial therapy, and was discharged.

**Discussion:** Epiglottitis is inflammation of the supraglottic structures. Since the implementation of Hib vaccine in 1985, there are fewer cases and a general decline in hospital admissions for epiglottitis. Our case illustrates a new causative agent and a physiologic mechanism infrequently seen in infants, pursed lip breathing. In the post-Hib era, epiglottitis has been caused by many pathogens. However, influenza A has never been reported in association with epiglottitis. The lack of association between influenza and epiglottitis could be due to an overall decline in epiglottitis, new causative organisms, or influenza with concurrent bacterial infections. Regardless of the etiology, the lethal complication of epiglottitis is upper airway obstruction. In our patient, pursed lip breathing and neck hyperextension were compensatory mechanisms. Pursed lip breathing is commonly seen in adults with chronic obstructive pulmonary disease to lessen work of breathing. It was unique that our 3-month-old patient was using this mechanism to decrease her work of breathing.

**Conclusion:** Since the initiation of Hib vaccine, epiglottitis is now caused by uncharacteristic pathogens. The current case highlights the occurrence of epiglottitis secondary to influenza A, and how quickly epiglottitis can lead to upper airway obstruction. It underscores the need to identify abnormal and unusual behaviors such as pursed lip breathing in children as a cause of upper airway obstruction since prompt recognition and treatment of upper airway obstruction is critical in preventing morbidity and mortality.

**Resources:**

References:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A129

Title: Labial bruising and fever in a young infant
Authors: Kristin Kalita, MD, Children's Hospital Colorado
Kathleen Hannan, MD, Children's Hospital Colorado, Christina Olson, MD, Children's Hospital Colorado
Case Presentation: A 62 day old girl was referred to the ED from clinic with fever and labial bruising noted at her 2 month well-child check. The mother had noticed vaginal redness and fussiness the night prior, but the baby was otherwise well. There was no history of trauma or sick contacts, and the baby had yet to receive her 2 month vaccines. Her parents were the only caregivers. Past medical history was notable for a 1 month NICU stay following C-section at 31 weeks due to abruption.

On examination, the infant was alert and active. Vital signs were: temperature 38.9C, heart rate 170, and respiratory rate 60. The labia majora were edematous with patchy, non-blanching bluish discoloration extending into the left inguinal area without tenderness or fluctuance (Figure 1). The complete blood count demonstrated 4,800 WBCs (71% neutrophils), and urine Gram stain revealed Gram-negative rods without pyuria. A comprehensive metabolic panel, lipase, skeletal survey and head CT were all reassuring. She was given ceftriaxone after blood and urine were sent for culture and admitted for further management.

Discussion: The differential for the labial discoloration included trauma, lichen sclerosis, and infection (cellulitis, abscess). In our patient, the fever pointed to infection, although there was also initial concern for non-accidental trauma (NAT). The Child Protection Team was consulted and agreed that the exam was concerning for NAT. The blood culture grew Group B streptococcus (GBS) shortly after admission. With continuation of antibiotic therapy, the labial swelling and discoloration quickly resolved without the step-wise evolution of ecchymoses, confirming a diagnosis of GBS cellulitis/bacteremia and lowering suspicion for NAT.

GBS is a known cause of cellulitis in infancy; localization to the face is most frequently seen (1), although cases involving the genital area have also been reported (2,3). A full septic work up should be pursued as there is a high association with invasive disease (90% of cases are associated with bacteremia and 25% with meningitis) (4). In our patient, the cerebrospinal fluid was normal. Additionally, her urine culture grew >100,000 E.coli colonies.

Conclusion: GBS cellulitis of the labia can mimic non-accidental trauma (NAT). Although fever can be a concurrent or coincidental symptom with NAT and NAT must always be considered in pediatrics, fever often indicates infection. It is imperative that pediatric hospitalists be aware that GBS cellulitis can involve the genital area and can appear ecchymotic so that treatment can be instituted promptly and unnecessary studies can be avoided.
Our patient successfully completed 10 days of ceftriaxone to treat GBS and E.coli. She was discharged home with complete resolution of all symptoms and with a very low suspicion for non-accidental trauma.

**Resources:**

Figure 1


**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A130

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**Title:** Plastic Bronchitis: An Innovative and Multidisciplinary Approach  
**Authors:** Janet Strausbaugh, Attending Physician, Division of General Pediatrics, Children's Hospital of Philadelphia  
Yoav Dori, MD, PhD, Children's Hospital of Philadelphia, Maxim Itkin, MD, Children's Hospital of Philadelphia, Jennifer Danzig, MD, Children's Hospital of Philadelphia

**Case Presentation:** We describe a 15 year old female with a 4 year history of intermittent episodes of cough productive of small amounts of bloody and stringy material. She reported symptoms about 2-3 times per week in which she would develop fatigue, malaise and frequent cough. This was relieved once a cast was expectorated. Airway clearance with vest therapy, hypertonic saline and inhaled corticosteroids were ineffective. Bronchoscopy done prior to referral to our institution was therapeutic due to cast removal, but not diagnostic with pathology revealing mucus mixed with alveolar histiocytes and nonspecific eosinophilia; no bacterial were present and culture grew normal flora. The diagnosis of plastic bronchitis due to abnormal lymphatic flow was suspected, and she was referred to the Children’s Hospital of Philadelphia for further diagnostic workup.

**Discussion:** The patient was referred to the Lymphatic Imaging and Intervention Program at our center. She underwent bronchoscopy and then further imaging in which a groin lymph node was accessed percutaneously and MR lymphangiogram was performed. Imaging revealed normal thoracic duct and abnormal bilateral pulmonary lymphatic perfusion. Access to the cisterna chyli was obtained via transabdominal approach and the thoracic duct was cannulated with a microcatheter. Contrast lymphangiogram then confirmed retrograde flow into the pulmonary lymphatic ducts. Embolization of the thoracic duct was then performed with coils and glue. Pathology showed layered proteinaceous material with mucus and variable acute inflammatory cells consistent with a bronchial cast. She recovered on the general pediatrics service with close respiratory monitoring. Her post operative course was notable for fluid responsive tachycardia and abdominal pain. She was discharged on hospital day 5. She stayed on a low fat diet for 2 weeks, and then resumed normal diet and activity.
**Conclusion:** Lymphatic network imaging using groin intranodal MR lymphangiography is a unique technique for imaging the lymphatic system to uncover disorders of abnormal flow. Our center has experience using an approach in which selective catheterization and embolization of abnormal lymphatic ducts can be done in concert with lymphatic imaging. Embolization of abnormal lymphatic channels has been demonstrated to lead to resolution of plastic bronchiitis. On follow up 9 months later, our patient occasionally produces small casts but her other respiratory symptoms and frequent infections have resolved. Disorders of the lymphatic system have a high degree of associated morbidity. The lymphatic program at our institution has pioneered a technique for minimally invasive imaging and intervention that can potentially resolve these disorders. These patients are managed by a collaborative team of general pediatrics hospitalists, cardiologists and interventional radiologists who have developed expertise in the care of disorders of lymphatic flow.

**Resources:**

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A131

**Title:** Recurrent Orange Urine and Abdominal Pain with a rare genetic cause  
**Authors:** Josef Newman, MD, University of Illinois College of Medicine at Peoria  
Bhavana Kandikattu, MD, University of Illinois College of Medicine at Peoria  
**Case Presentation:** 13-year-old male with acute kidney injury (AKI) (Cr = 5.6mg/dL) and hypertension with 18 months of recurrent monthly episodes of back and abdominal pain, dark orange discoloration of urine, difficulty voiding, nausea and anorexia. Pertinent history included anemia empirically treated with iron since childhood and occasional staring spells. 
Exam was unremarkable except for obesity. Differentials included hypercalciuria, porphyria, or metabolic disorders. Laboratory testing revealed an elevated serum uric acid (12.5mg/dL) and fractional excretion of uric acid. Hemoglobin was low (9.8g/dL) with elevated MCV, (9.39 fL) MCH, (31.3pg) and reticulocytes (6.7%). Haptoglobin, iron studies, Folate and Vitamin B12 levels were normal. Renal ultrasound showed no nephrolithiasis. Serum CK, LDH, myoglobin and total porphyrins were normal. Urine studies were positive for myoglobin, with normal random calcium and Ca/Cr ratios. Results were concerning for partial hypoxanthine-guanine-phosphoribosyl-transferase (HPRT) deficiency and the patient was empirically started on Allopurinol. 
**Discussion:** Our patient was managed with intravenous fluids and started on Allopurinol when the uric acid resulted elevated. He responded well to supportive management and was discharged with outpatient genetics follow-up. Given the uric acid levels, whole exome sequencing was sent in suspicion of mutations in HPRT or APRT (Adenine-Phosphoribosyltransferase). It resulted in a de novo substitution (p.S62N) in the PGK1 gene associated with Phosphoglycerate Kinase deficiency.
Phosphoglycerate Kinase 1 deficiency is a rare X-linked recessive disorder characterized by hemolytic anemia, myopathy and neurological dysfunction. Phosphoglycerate Kinase 1 catalyzes the conversion of 1,3-disphosphoglycerate to 3-phosphoglycerate during glycolysis. Clinical presentation includes hemolytic anemia, myopathies and neurological involvement. Patients can express any combination of manifestations. Myopathic involvement can be severe, with reported cases of rhabdomyolysis with subsequent renal failure, and several reports of exercise intolerance with recurrent myoglobinuria.

**Conclusion:** ‘Orange urine’ is an uncommon complaint in Pediatrics. Discoloration of urine with abdominal pain is usually concerning for nephrolithiasis, while orange is very specific for the presence of uric acid. On initial interview the patient could not identify a specific trigger for his episodes. On further discussion he related physical activity the morning or day before the majority of occurrences. Similarly when delving further into neurological history, mother recalled staring spells occurring since the age of 10 for which no investigation was initiated. This history combined with anemia and elevated uric acid prompted further genetic testing. At the present time PGK-1 enzyme activity assays were sent as follow up.

This case illustrates the importance of thorough history taking in pediatric hospital medicine. Patients may omit details that seem inconsequential to them, and connecting the dots often requires deeper probing of seemingly unrelated events. To the best of our knowledge this is the first report of PKG-1 deficiency presenting with elevated serum and urine uric acid.

**Resources:**
Sources:
Scratching Past the Surface: Understanding Bartonella's Spectrum of Disease

Melody Shi, M.D., Duke University Hospital
Juliana Dial, M.D., Duke University Hospital

Case Presentation: 9 yo female presenting with daily fevers for one month, myalgias, abdominal pain and weight loss with recent exposure to cats. Her exam revealed diffuse abdominal tenderness without peritoneal signs or hepatosplenomegaly and diffuse myalgias without joint swelling or warmth. Lab work revealed leukocytosis with bandemia, normocytic anemia, mild thrombocytosis, urinalysis suggestive of urinary tract infection, and profoundly elevated CRP and ESR. X-rays, echocardiogram and abdominal ultrasound were unremarkable. Renal ultrasound revealed mild hydronephrosis. She was started on appropriate antibiotics for presumed pyelonephritis (urine culture grew E. Coli) while evaluating for other causes of fever of unknown origin (FUO) given unexplained abdominal pain and myalgias. Infectious etiologies were sent and later returned positive for B. henselae, IgM 1:400 and IgG 1:2560. Upon discussion with pediatric infectious disease, she was diagnosed with disseminated bartonella given her absence of localized lymphadenopathy. She was treated with azithromycin for one month.

Discussion: Fever of unknown origin (FUO) remains a diagnostic dilemma and includes a wide variety of etiologies [1]. Among cases with an identifiable cause, 40% were infectious, 15% collagen-vascular and 10% oncologic [2]. Of the infectious causes, EBV was most common followed by osteomyelitis and Bartonellosis [3]. Bartonella henselae, the intracellular gram-negative bacilli that causes cat scratch disease (CSD), affects approximately 12,000 patients per year [4]. Cats are the major reservoir and transmit disease to humans via a scratch or saliva. However, prior exposure is not uniformly found, suggesting that such infection should still be considered in FUO. The most commonly recognized presentation consists of regional lymphadenopathy and fevers preceded by a cutaneous lesion. Typical CSD is self-limited and antibiotics are not routinely suggested. However, disseminated infection can have a complicated course with multi-system involvement [5]. This wide spectrum of disease creates a diagnostic challenge as B. henselae can mimic many other diseases and requires high clinical suspicion.

Conclusion: 1. Fever of unknown origin (FUO) is a diagnostic dilemma caused by infectious etiologies in most cases with an identifiable cause.
2. Even though typical CSD is more common, those with FUO and no identifiable source should be tested for bartonella to prevent the morbidity associated with disseminated disease.

Resources:

Works cited:
Title: Stuck in the Middle with Mu-cin: A 5 yo male with ophthalmoplegia and fever  
Authors: Alison Rittenberg, MD, University of North Carolina Children’s Hospital  
Eric Zwemer, MD, University of North Carolina School of Medicine  

Case Presentation: A 5-year-old male with allergic rhinitis presented to the ER with acute onset of right cranial nerve VI palsy and afferent pupillary defect (APD). Mom reported a 5 day history of subjective fever, nasal congestion, and cough. Vital signs were T39.4°C, HR 84, BP 121/71. Exam showed an uncomfortable-appearing boy with right-sided proptosis, orbital edema, visual field deficit, absent right eye abduction, and a sluggishly reactive right pupil measuring 3-4 mm. CT showed an expansile right-sided sphenoid mass with evidence of bony erosion. Differential diagnosis included bacterial sinusitis with orbital cellulitis, fungal infection, mucocele, and tumor. In the OR, a large mass with “peanut butter” consistency and purulent fluid was found in the right sphenoid sinus. Pathology demonstrated septate fungal hyphae and eosinophilic mucus, most consistent with allergic fungal sinusitis. Patient received systemic steroids and antibiotics for superimposed bacterial infection. At discharge, he continued to have a right cranial nerve VI palsy, visual field deficit, and improved but persistent APD.

Discussion: Allergic fungal sinusitis (AFS) is a form of chronic sinusitis due to hypersensitivity response to fungi (typically Aspergillus) in the paranasal sinuses. Atopy is a hallmark of the disease, with the majority of patients reporting a history of allergic rhinitis and/or asthma. The majority of patients are not immunocompromised. Presenting symptoms include nasal obstruction, discharge (at times purulent or dark-colored), headache, and proptosis. If pain is a presenting symptom, it can indicate a superimposed bacterial infection. Cases of visual loss and cranial nerve deficits reported in the literature are rare. The diagnosis of AFS is made based on a detailed history of atopy and symptoms of sinusitis, characteristic CT findings (opacified sinuses and bony erosion), and histologic evidence of eosinophilic mucus with non-invasive fungus. Treatment involves urgent endoscopic resection for orbital decompression and systemic steroids. There are no convincing controlled trials that demonstrate efficacy of antifungals to treat AFS. A high rate of reoccurrence necessitates close follow-up.

Conclusion: AFS occurs in immunocompetent patients and should be on the differential for any pediatric patient with a history of atopy presenting with refractory sinusitis. In rare cases, AFS can present with cranial nerve deficits and vision loss which should prompt immediate imaging and subsequent surgical decompression of the orbit. AFS does not require treatment with antifungal agents, but patients should be treated with steroids to help reduce the risk of recurrence.

Resources:

Patient looking straight ahead:

Patient looking to the left:

Patient looking to the right:

References

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A134

Title: The “Strength” in Numbers
Authors: Colleen Meehan, MD, MPH, Children’s National Health System
Neha Agarwal, MD, Children’s National Health System, Anjna Melwani, MD, Children’s National Health System

Case Presentation: A 21 month old healthy female presented to the Emergency Department with two days of rhinorrhea, poor PO intake and fever. She was diagnosed with acute otitis media and discharged home on amoxicillin. She presented to the ED the next day with continued poor PO, decreased urine output and lethargy. On exam, she was afebrile, slightly tachycardic, mildly tachypneic and minimally responsive to noxious stimuli. There was no evidence of meningismus. The remainder of the exam was within normal limits. Laboratory results were significant for leukocytosis with neutrophil predominance, hyponatremia, hyperkalemia, and elevated transaminases (AST/ALT=2,172/399 u/L). A urinalysis showed large blood and 2-5 RBCs per HPF. She was transferred to a tertiary care facility for admission. After admission, the creatine kinase level returned at 360,000 units/L. She was treated for rhabdomyolysis and improved with rehydration. Genetic testing later revealed a deleterious mutation in the LPIN1 gene (lipin-1 deficiency) which is associated with recurrent episodes of rhabdomyolysis in early childhood.

Discussion: Rhabdomyolysis is caused by infection, trauma, prolonged muscle activity, drugs/toxins, inflammatory myopathies and inherited muscle disorders (metabolic myopathies, muscular dystrophies and congenital myopathies). Our patient’s history and exam did not support many of the above diagnoses as the underlying etiology. Given our patient presented with fever and rhinorrhea, viral myositis leading to rhabdomyolysis was considered. However, infection-induced rhabdomyolysis does not typically lead to creatine kinase levels >100,000 units/L. Given the severity of presentation, molecular genetic studies for metabolic myopathies were sent.

Lipin-1 deficiency is a cause of recurrent rhabdomyolysis in childhood. Children with this disease typically present between 15 months and 7 years of age with episodes of severe rhabdomyolysis triggered by fever or mild illness, exercise, fasting or medications. These patients are at risk of rapid decompensation and cardiac arrest. The mainstay of treatment is supportive care with aggressive hydration and correction of electrolyte abnormalities.

Conclusion: Pediatric health care professionals should consider rhabdomyolysis, even in the absence of muscle pain or weakness, when labs are notable for elevated transaminases, particularly if the AST is significantly more elevated than the ALT, or if blood is noted on urinalysis, particularly if there is a discrepancy between the amount of blood and the number of red blood cells noted per high power field. A significantly elevated creatine kinase level in a previously healthy young child should prompt practitioners to test for metabolic myopathies. Furthermore, metabolic myopathies often go undetected at birth, and manifest acutely in the setting of a minor viral illness – if not recognized early, these patients are at risk for rapid decompensation. Thus, early recognition and aggressive hydration on presentation to the Emergency Department are critical for patients with lipin-1 deficiency.

Resources:
Figure 1: The Differential Diagnosis: Elevated Creatine Kinase

Myopathies
- Muscular dystrophies
- Inflammatory myopathies
- Metabolic myopathies
- Endocrine myopathies
- Congenital myopathies
- Channelopathies
- Drug/toxin-induced

Neuropathies
- Guillain-Barre syndrome
- CIDP

Motor
- Spinal muscular atrophy
- ALS

Neuron Disease
- Postpolio syndrome

Other
- Trauma
- Surgery
- Viral illness
- Strenuous exercise
- Malignant hyperthermia
- Seizures

Figure 2: Metabolic Myopathies

References:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A135

Title: The Curious Case of the 8 Year Old Female with Hypoxemia

Authors: Jason Zamkoff, MD, Children’s Hospital Colorado
Rene Roy, MD, Children’s Hospital Colorado

Case Presentation: Previously healthy 8 yr old female admitted for 6 months of fatigue and exercise intolerance. Evaluation by PCP 1 month prior showed normal exam, with SpO2 87% on RA. 2 weeks prior she was seen in ED for a rash where her SpO2 on RA was 83% with no respiratory distress. CXR was normal and she was sent home with 0.5L O2 via N/C at night. Flovent and Singulair prescribed with no improvement. On admission she c/o headache, blurry vision and dizziness when walking up stairs. Her home SpO2 measured 82%. Patient was taken to ED where O2 was given at 2L/min with resolution of symptoms and increased SpO2 to 98%. EKG and CXR were normal. She was admitted for further evaluation. Family History noncontributory. Patient lives at 6000 ft above sea level. Other ROS negative. Admission Vitals: T 36.9; HR 97; BP 107/76; RR 22; SpO2 94% on 2L O2 N/C EXAM: Well appearing. HEENT exam normal. Normal work of breath on 2L O2 via N/C, lungs clear bilaterally. No murmurs, pulses and cap refill normal. Abdominal exam benign, no hepatomegaly. No skin lesions or clubbing. Neuro exam normal.
**Discussion:** DDX for patient’s hypoxemia: 1) Hypoventilation 2) Low Inspired O2 Tension 3) V/Q Mismatch 4) R→L Shunting 5) Diffusion Limitation 6) Decreased O2 Carrying Capacity 7) Inaccurate SpO2  

**Hospital Course:** Hospital Day (HD) 1: Pulmonary and Cardiology consults obtained. HCT 46. Viral PCR negative. VBG 7.39/32/68/-3 showed no chronic hypoventilation. HBG electrophoresis, carboxyhemoglobin and methemoglobin normal, showed no decreased O2 carrying capacity. Urinalysis normal, showed no overt vasculitis. Echo obtained was technically challenging, read as normal. HD 3: Bubble echo showed intrapulmonary R→L shunt, with contrast noted in left atrium 3 cardiac cycles after injection. HD 4: CT angiogram showed 1.2 cm arteriovenous malformation (AVM) in lower left lung base (see images). Genetics consulted, mutation identified in Endoglin gene, confirming diagnosis of Hereditary Hemorrhagic Telangiectasia. HD 5: Brain MRI angiogram showed no AVM. HD 6: Pt discharged clinically stable on 1.5 Liters O2. Pt later returned for angiogram (see images) and coil embolization of AVM.

**Conclusion:** Hypoxemia in Denver and other regions of altitude is commonly found in our inpatient population, usually associated with respiratory distress and an infectious etiology. Our patient demonstrated hypoxemia without respiratory distress, presenting a more varied and interesting differential diagnosis. She was eventually diagnosed with a pulmonary arteriovenous malformation, which was subsequently embolized with resolution of her hypoxemia. Final conclusions: 1) Pulmonary arteriovenous malformation (AVM) is a rare but known cause of hypoxemia in the pediatric population. 2) Bubble echo is the diagnostic tool of choice to detect the presence of a pulmonary AVM. 3) The majority (58%) of patients with Hereditary Hemorrhagic Telangiectasia (HHT) with have a pulmonary AVM.

**Resources:**
Coronal View of CT Angiogram demonstrating the abnormal communication between the Pulmonary Artery and Pulmonary Vein.

Coronal View of CT Angiogram demonstrating the ectatic Pulmonary Vein secondary to abnormal communication with the arterial system via the AVM.
Case Presentation: A 3-month old infant presented to the ED for the 3rd time since birth after a brief episode of irregular breathing. After eating, he tried to cry and breathe, but could not inhale and had roaming eye movements. He was at baseline 3 minutes after suctioning. Mother reported motor delays and more tachypnea than in siblings. A ROS was otherwise normal.

Of note, the patient was referred to the ED by his pediatrician at 5 days of life for increased work of breathing. He arrived with a normal exam but desaturated with a feed. He was admitted with a BRUE. He had no further desaturation and fed well. EKG and CXR were unremarkable.

At 2 months, the infant also presented with congestion, cough, and panting. His exam normalized after suction. He was discharged with bronchiolitis from the ED.

Exam on the 3rd presentation revealed subcostal retractions with transmitted upper airway sounds, diminished on the left. He had a bell-shaped chest, diffuse hypotonia and absent Moro, Babinski and plantar grasp.

CXR showed collapse of the left lung with mediastinal shift. He was admitted again.

Discussion: A clinical practice guideline published in 2016 recommended replacing the term “Apparent Life Threatening Event” with “Brief Resolved Unexplained Event” (BRUE) for patients <1 year who present with change in color, respiration, tone and/or altered responsiveness, are well-appearing on presentation, and have no explanation after history and exam. Infants >2 months of age and ≥ 45 weeks corrected gestational age, with a single event lasting <1 minute, and never requiring CPR by a trained medical provider may be considered low risk, thereby avoiding unnecessary workup and hospital admission.

Each of the patient’s presentations could have been described as ALTE but only the first as BRUE. On the 2nd and 3rd presentation the patient remained symptomatic. Instead of describing subsequent presentations as BRUE, the team could focus on the poor mucus clearance, difficulty feeding, and hypotonia. Genetic testing during hospitalization revealed Spinal Muscular Atrophy Type 1.

Conclusion: This case illustrates that the new clinical practice guidelines surrounding the phenomenon now known as BRUE may not only identify infants at low risk for a recurring event or serious underlying disorder, but may also help avoid complacency in patients at higher risk and target continuing symptoms in those who do not have a fully resolved exam.

In this case, SMA Type 1 was diagnosed 3 months earlier than average. Despite this genetic condition resulting in extensive and progressive muscle weakness and leading to feeding and respiratory difficulties, it is commonly missed due to variations in symptomatology and severity, as well as visits to multiple pediatric subspecialists to rule out other
illnesses. By making the diagnosis early, the team could initiate quality-of-life interventions that avoided future complications.

**Resources:**

References:


**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A137

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**Title:** What's Hiding in the Dirt?

**Authors:** Lindsey Hastings, Medical Student, University of Kentucky
Alan Hall, MD, University of Kentucky, Elizabeth Seelbach, MD, University of Kentucky

**Case Presentation:** A 2 year-old girl from southeastern Kentucky presented with pallor in the setting of excessive milk consumption. She often played outdoors barefoot with several dogs and had been eating chalk, dirt, sand, and pieces of coal over the past year. She had no history of asthma, seasonal allergies, or eczema.

On exam, she was pale-appearing with conjunctival pallor but had no lymphadenopathy, organomegaly, or a rash. White blood cell count was 37.8 x 10⁹/L (with 71% eosinophils) and hemoglobin 7.7 g/dL (MCV 53 fl and RDW of 22%) with iron studies consistent with iron deficiency. LDH and uric acid were normal. A peripheral smear corroborated the above findings. Stool was negative for ova and parasites.

With pending serologies, she received a 5-day course of albendazole for Toxocara with iron supplementation and follow-up with infectious disease. She missed her initial appointment, and when seen she had stopped taking iron and again had pica with unchanged labs. Antibodies returned positive for both Strongyloides and Toxocara, and she was re-treated with albendazole plus ivermectin.

**Discussion:** Anemia and leukocytosis are common findings encountered by a pediatric hospitalist. In this case, the work-up of iron deficiency secondary to excessive cow's milk consumption revealed a severe eosinophilia that required the consideration of a broader differential including atopic and parasitic diseases, drug reactions, and malignancies. Her history of frequent outdoor play with dogs, pica, and eosinophilia raised suspicion for a parasitic disease. Even prior to testing, the confirmed diagnoses seemed to be the most probable explanation for her eosinophilia given that up to 14% of the population in the United States is seropositive for Toxocara, Strongyloidiasis is endemic in the southeastern United States, and both are transmitted through soil.

**Conclusion:** This case demonstrates the value of a thorough history (especially environmental) in the evaluation of eosinophilia. Appropriate treatment of parasitic infections (albendazole is the drug of choice for toxocariasis but is second to ivermectin for strongyloidiasis) is important, but it must also be emphasized that the treatment of iron deficiency and subsequent resolution of pica are key to preventing re-infections with certain parasites. Additionally, follow-up is vital particularly when medication compliance and access are uncertain.

**Resources:**

References, What's Hiding in the Dirt?


**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A138
Title: A Surprising Case of Raccoon Eyes in a Pediatric Patient

Authors: Paul Madera, MD, Nicklaus Children’s Hospital
Christie De La Vega, MD, Nicklaus Children's Hospital, Melissa Clemente, MD, Nicklaus Children's Hospital

Case Presentation: A 3 yo nonverbal autistic boy with 1 month of limping progressing to refusal to bear weight. No preceding trauma, recent fever or illness.

On exam, periorbital edema noted bilaterally. He had gingival swelling, but no bleeding. Joints had no erythema/swelling. Passive range of motion was full. Supported gait was slow, wide-based.

CBC, CMP, ESR, CRP, PT/INR, PTT, CPK, uric acid all unremarkable. LDH mildly elevated. RPP positive for Rhino/Enterovirus.

Bone scan showed increased uptake about distal femurs/proximal tibiae, suggesting metabolic bone disease.

Detailed dietary history uncovered marked food selectivity. Additional labs obtained, but interim worsening of periorbital swelling developed. Imaging was done to investigate a possible neuroblastoma.

Orbital MRI showed right extracanal mass and left periorbital soft tissue swelling. CT, US revealed no intrathoracic/abdominal mass. VMA/HVA levels were normal.

Vitamin D, PTH were low normal. Vitamin C resulted critically low. Diagnosis of scurvy was made. Patient was started on high-dose Vitamin C with significant improvement.

Discussion: Vitamin C is an essential water-soluble cofactor in the synthesis/fortification of collagen and connective tissue. Critical deficiencies can lead to vascular purpura, poor wound healing, follicular keratosis, and bleeding gums. Children are susceptible to developing severe musculoskeletal symptoms. A still-maturing association between the periosseum and surface of the cortex accommodates fluid from ruptured synovial blood vessels. Hemarthrosis/muscular hematomas preclude arthralgia and myalgia in 80% of patients with scurvy.

Manifest scurvy refers to the classical symptoms. Latent scurvy (fatigue, irritability, dull body aches) is the more common presentation, but tends to a more challenging diagnosis and is consequently underreported.

Pertinent to our case, children with autism spectrum disorder are estimated to have a fivefold increase in odds of having a feeding problem compared to typically developing children. Here, we describe an autistic child with an almost exclusive diet of Gatorade and banana pudding, leading to the development of scurvy.

Conclusion: Scurvy in developed countries is rare, and even less common in pediatrics. Children at highest risk are those with restricted diets.

Confounding our case was the periorbital ecchymosis. In a patient with no prior trauma presenting with “raccoon eyes”, the concern for metastatic neuroblastoma was immediately heightened. No primary lesion was identified and catecholamine metabolites were within reference range.

These findings support expanding the narrow differential diagnosis of “raccoon eyes” to include scurvy in pediatric patients with feeding difficulties.

Resources:

References
Title: A Teenager with Pain in the Neck

Authors: Heidi Greening, DO, Advocate Children’s Hospital - Park Ridge

Case Presentation: D. S. is an 18yo previously healthy male who presented after having six days of sore throat, anterior neck pain and high grade fever. He was previously evaluated and RST and Mono were negative, but was started on azithromycin and decadron. On DOA, he presented to the ER where labs showed a WBC of 21, CRP of 26, and a creatinine of 1.69. Upon admission, he had SIRS and CTX and Clindamycin were empirically started. Procalcitonin >100. CMV IGG/IGM were positive. DIC panel with elevated DDimer, PT/PTT. HIV and EBV negative. His blood culture became positive for Fusobacterium necrophorum. CT Neck showed phlegmon and right sided LAD and compression of his IJV. CT Chest showed pulmonary septic emboli. He required surgical drainage of the neck abscess. Dopplers showed a 7cm superficial basilic thrombophlebitis. Hematology was consulted and testing for hypercoagulability was positive for lupus anticoagulant. He was started on lovenox BID to complete a several week course. He was discharged home to complete clindamycin TID for treatment of Lemierre’s syndrome.

Discussion: Lemierre’s syndrome is a life-threatening, but rare complication of oropharyngeal infections. The suggested diagnostic criteria involve 1.) History of oral pharyngeal infection, 2.) Findings of thrombophlebitis (typical of the internal jugular vein) 3.) Isolation of the pathogen from culture, all of which our patient in the vignette demonstrates. Our patient was positive for CMV likely has the preceding oral infection allowing Fusobacterium to locally migrate. Our patient also demonstrates how this “forgotten disease” can be easily missed in the early stages with many providers diagnosing strep throat or mononucleosis, which in fact our patient did have. Thrombosis in Lemierre’s is likely multifactorial with underlying genetic prothrombotic propensity, increased factor VIII activity, antibody production, and vascular inflammation. While treatment with anticoagulation is debated, most cases with septic emboli do require prolonged anticoagulation. Antibiotic guidelines are not clear, but most Fusobacterium are treated with Beta-lactamase resistant antibiotics.

Conclusion: Lemierre’s disease is a serious, oropharyngeal infection with septic thrombophlebitis and emboli which can be missed in its early stages. There should be a high index of suspicion in toxic appearing children with pharyngitis. Prompt diagnosis and treatment with antibiotics, fluids and anticoagulants can be lifesaving.
Title: Atypical Case of Salicylate Toxicity with a Focus on Non-Aspirin Sources
Authors: Haig Aintablian, MD Candidate, University of Arizona - College of Medicine Phoenix
Sami Kabbara, MD Candidate, University of Arizona - College of Medicine Phoenix, Edith Allen, MD, Phoenix Children’s Hospital

Case Presentation: Our patient is a 17 month old, fully vaccinated, Hispanic male with difficulty breathing and dehydration. His symptoms began with 6 vomiting episodes before presenting for medical evaluation one day later. At the time of evaluation, he was normothermic at 36.8°C, heart rate was 130, and respiratory rate was 28. On exam, he had tachypnea and hyperpnea. His laboratory analysis revealed WBC count of 19,300 with 74% neutrophils, platelet count of 443,000, and significant carboxyhemoglobinemia and methemoglobinemia. His venous blood gas analysis showed pH of 7.36, pCO2 of 17mmHg, bicarbonate of 9mmol/L, and base deficit of 15.5mmol/L. His electrolytes showed ionized calcium of 7.3mEq/L, sodium of 141mEq/L, and potassium of 3.0mEq/L. His glucose was 68mg/dL. A blood culture revealed no organisms, chest X-ray revealed reactive airway disease versus a viral process, and respiratory PCR was negative. After significant investigation, toxic salicylate levels of 51.1mg/dL were discovered, much to the surprise of his concerned mother, who stated that she had locked away the aspirin long ago.
**Discussion:** In 2013, more than 33,000 cases of salicylate exposure were reported. Many of the diagnoses in unobserved ingestion come from salicylates' pathognomonic mixed respiratory alkalosis and metabolic acidosis. Aspirin is not the only source of salicylate; various other sources such as salicylic laxatives, scents, and herbal remedies exist. Studies of oil of wintergreen, a common herb found in many scented wax products, ointments, and flavoring agents, have shown that it can be equivalent to twenty-two 325mg adult aspirin tablets. Sources of salicylates in a common household are more prevalent than just an aspirin bottle, and are well-established. However, the association between salicylate toxicity and carboxyhemoglobinemia and methemoglobinemia has never before been described. Certain salicylates such as p-aminosalicylic acid have the biochemical potential to oxidize the iron containing moiety in hemoglobin in vitro, however this has never been established in vivo. Similarly, carboxyhemoglobinemia has never been mentioned in a case of salicylate toxicity.

**Conclusion:** While aspirin may be the most common source of salicylate toxicity, many other sources have been described in the literature. These sources may present with symptoms typical of salicylate intoxication such as a mixed respiratory alkalosis and metabolic acidosis, hyperpnea, and tachypnea, and may also have signs and symptoms that distinguish them from aspirin. Our patient had a salicylate toxicity that was masked by a significant leukocytosis, electrolyte abnormalities, and CXR findings. The presence of methemoglobinemia and carboxyhemoglobinemia, unreported in a case of salicylate toxicity, further led to confusion about his diagnosis. These findings, as well as the concern for the source of aspirin by the patient's mother, all suggest the possibility that this was a salicylate toxicity not due to aspirin, but to another toxin. Our patient's case draws attention to these alternate sources of salicylate toxicity, as well as the clinical and laboratory findings that differentiate them from a simple aspirin toxicity.

**Resources:**

**References:**


**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A141
Title: Hit by a MAC Truck: Disseminated infection in a previously healthy 6yo

Authors: Marta King, MD, MED, Saint Louis University School of Medicine
Amelia Bray-Aschenbrenner, MD, St. Louis University, Edwin Anderson, MD, St. Louis University, Alan Knutsen, MD, St. Louis University, Deepika Bhatla, MD, St. Louis University

Case Presentation: A 6yo girl with history of cervical Mycobacterium avium-intracellulare (MAI) lymphadenitis status post bilateral lymph node excision (Figure 1) 8mo prior, was admitted with two weeks of intermittent high fevers, abdominal pain, and weight loss. She was otherwise healthy with no recent travel, no TB or cat exposure, and no sick contacts. She was afebrile but ill appearing with tender cervical and post-auricular lymphadenopathy, exquisite abdominal tenderness, and splenomegaly. Labs showed elevated inflammatory markers, LDH, and microcytic anemia (Table 1). Abdominal CT confirmed splenomegaly and revealed intraabdominal lymphadenopathy (Figure 2). Differential diagnosis included disseminated MAI (DMAI) with concurrent HIV or other immunodeficiency, cat scratch disease, TB, EBV, CMV, malignancy including leukemia/lymphoma, and hemophagocytic lymphohistiocytosis. Clarithromycin and rifampin were started due to suspicion for DMAI while other infectious, malignant, and inflammatory etiologies were investigated. Blood, bone marrow, and stool cultures grew MAI, confirming the diagnosis.

Discussion: MAI is a non-tuberculous mycobacterium of low virulence found throughout the environment and typically causing lymphadenitis in young children. Disseminated infections are rare and seen primarily in severely immunocompromised individuals, particularly those with AIDS. DMAI is characterized by fever, abdominal pain, weight loss, anemia, elevated LDH and alkaline phosphatase. Diagnosis is confirmed by positive cultures from two sterile, noncontiguous sites. Disseminated infections can be fatal if treatment is inadequate or delayed.

Conclusion: The patient was treated with clarithromycin, rifampin, ethambutol, amikacin and ciprofloxacin and discharged upon clinical improvement. Interferon gamma infusions and clofazimine (an orphan drug used for treatment of leprosy) were added at 3 weeks and 5 months after discharge, respectively. Extensive immunologic workup was non-diagnostic. Whole exome sequencing revealed a novel mutation in a gene involved in regulating the cellular response to infections, supporting the suspicion for a previously undescribed underlying immunodeficiency. Despite two years of aggressive therapy, blood cultures remain positive for MAI. The patient is currently awaiting bone marrow transplant for definitive treatment. Deciphering variants of common diagnoses from new, rare conditions is challenging, as not every patient follows the anticipated course of even the most “bread and butter” illness. Clinicians therefore must maintain a broad differential in the face of atypical or uncommon presentations in order to treat the mundane without missing the extraordinary, potentially with fatal consequences.

Resources:
Bibliography


Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A142

Title: NOT AS SIMPLEX AS IT APPEARS: CHEST PAIN AND ODYNOPHAGIA IN A HEALTHY BOY

Authors: RAPHAEL STURM, MD, Nicklaus Children's Hospital

Case Presentation: A 10 year-old healthy male presented to the ED with a five day history of fever and sharp, retrosternal chest pain with swallowing. He was unable to tolerate PO and reported no relief with omeprazole or pepto-bismol. He denied nausea, vomiting, recent trauma, cough, sick contacts or rash, including any oral lesions. EKG, CBC and BMP were all normal. A rapid strep test was negative. A CXR revealed a possible mid-thoracic compression fracture and he was admitted for further evaluation. An MRI spine revealed no compression fracture. An esophogram was within normal limits. On third day of admission, the patient developed scattered vesicular lesions on his arms and feet. Endoscopy was performed due to persistence of symptoms which revealed severe striped esophagitis with ulceration throughout the esophagus. Biopsy results and immunohistochemical staining were positive for HSV. Cultures of the skin lesions were positive for HSV-1. The patient was started on acyclovir with clinical improvement of symptoms within two days. He was discharged home to complete ten day course of acyclovir.

Discussion: Herpes simplex virus esophagitis (HSVE) has rarely been reported in immunocompetent children. There is a male predominance of 3:1. The most common symptoms of HSVE are acute onset odynophagia, fever and retrosternal chest pain. Interestingly, oropharyngeal lesions are uncommon. Many cases go unrecognized with symptoms attributed to other self-resolving causes. Other infectious causes of esophagitis include Candida, especially in patients with history of chronic inhaled steroid use. CMV, VZV, HIV and bacterial causes remain possibilities, but are usually found in immunocompromised hosts. Non-infectious causes of esophagitis include GERD, corrosive ingestion and eosinophilic esophagitis. Endoscopy with biopsy is warranted to differentiate the possible diagnoses. Endoscopy findings of HSVE are non-specific and include friable mucosa, with erythema and ulcers. Immunohistochemistry and viral cultures are the most sensitive markers for diagnosis. While treatment is mainly symptomatic and disease is usually self-limited, Acyclovir can be used to decrease length and severity of symptoms.

Conclusion: Esophagitis is a cause of pediatric chest pain and HSV esophagitis (HSVE) should be suspected in an otherwise healthy patient with acute onset odynophagia, chest pain and fevers with no other obvious cause for esophagitis. Endoscopy with biopsy, immunohistochemistry and cultures will confirm the diagnosis and symptomatic treatment is recommended. Immune workup is probably not necessary in otherwise well appearing patients with no concerning history and no other signs of immunosuppression.

Resources:
REFERENCES
Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A143
**Title:** Recollections of the Past: Recurrent Anti-NMDAR Encephalitis

**Authors:** Alyssa Tilly, MD, UNC Department of Pediatrics  
Ashley Sutton, MD, University of North Carolina-Chapel Hill, Sara Sanders, MD, UNC Department of Pediatrics

**Case Presentation:** 17 year-old-male with a history of anti-NMDAR encephalitis three years prior presented with two days of confusion, agitation and insomnia. Associated symptoms included anorexia, blunted affect, slow movements and, though awake, refusal to respond or engage with others. Family also reported apparent hallucinations and associated aggression including biting. Neurologic examination with non-verbal patient, intermittent ability to attend and follow commands, hypertonic extremities, 3+ patellar reflexes and ankle clonus bilaterally. MRI Brain was normal and CSF analysis confirmed anti-NMDAR antibodies. EEG showed diffuse slowing but no seizures. Paraneoplastic work-up was negative. Prior presentation in Mexico was associated with seizure and protracted neurologic impairment which eventually resolved after treatment with steroids and IVIG. Initial management with pulse-dose steroids and IVIG resulted in minimal clinical improvement, thus the patient was treated with Rituximab. Patient received monthly infusions and after three months had returned to baseline.

**Discussion:** Anti-NMDAR encephalitis is a well-described syndrome of autoimmune encephalitis and one of the most common etiologies of non-infectious encephalitis in children. Though possible, anti-NMDAR encephalitis less likely to be paraneoplastic in children and males (1, 2, 3). Presentation classically includes neuropsychiatric symptoms such as agitation, psychosis, behavior and personality changes as well as memory loss, chorea and hyperkinetic movements, and coma. Studies may be normal other than CSF anti-NMDAR antibody detection, though studies may also reveal CSF lymphocytic pleocytosis and elevated protein and various MRI abnormalities (3,4). Up to 25% of patients may experience a relapse of symptoms, often months or years after initial presentation as in this case (5). Relapse may be more likely to occur in patients who do not receive immunotherapy at first presentation and who do not have an associated tumor. Relapsed symptoms may differ from prior and classical presentation (2, 5).

**Conclusion:** Early diagnosis and initiation of immunotherapy leads to decreased levels of anti-NMDAR antibodies. This aids in reducing the risk of relapse, as relapses have been reported in 12-25% of patients. Risk for relapse include lack of immunotherapy and may also be related to absence of tumor, although there is limited information regarding risk for relapses overall. Clinical presentations of relapses often differ from initial symptoms or classic presentations of anti-NMDAR encephalitis (6). Relapses have been described as occurring up to 15 years later with significant recovery between initial and repeat presentations (3, 6). Early treatment, milder disease, and malignancy resection (when applicable) come with a more favorable prognosis (4, 6). This case is unique as relapse is less often described in the pediatric patient and the patient did receive immunomodulator therapy with return to baseline and thus demonstrates the need for clinical suspicion for relapse in cases where patients with a history of anti-NMDAR encephalitis present with concerning neurologic symptoms.

**Resources:**
3. Bale Jr, J. Virus and Immune-Mediated Encephalitides; Epidemiology, Diagnosis, Treatment, Prevention. Pediatric Neurology. 2015; 53; 3-12

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A144
Title: Severe HIV disease in an antibody-negative infant in Lilongwe, Malawi

Authors: Andrea Dean, MD, Baylor College of Medicine
Allyson Mckenney, MD, Baylor College of Medicine-Children’s Foundation Malawi

Case Presentation: A 4-month old, breastfed infant presented with fever and difficulty breathing to Kamuzu Central Hospital in Lilongwe, Malawi. The infant’s mother was HIV negative in second trimester and in labor.

On exam: SpO2 85%, RR 80-90, HR180. He was alert, small-for-age, in respiratory distress with retractions, nasal flaring and had a nonfocal lung exam without wheezes or rales. He had oral thrush and dry mucous membranes but normal skin turgor and capillary refill. Radiography was unavailable. The mother’s rapid HIV antibody testing was positive, the infant’s was negative and his HIV DNA PCR was sent to lab.

Oral thrush and PCP pneumonia were clinically diagnosed. He was treated with oxygen, SMZ-TMP, steroids, ceftriaxone and fluconazole. He did not meet World Health Organization (WHO) criteria for presumptive diagnosis of severe HIV disease due negative antibody testing, but empiric ART was initiated while awaiting result of the DNA PCR, which was positive.

He developed BCG-lymphadenitis at 3 weeks consistent with immune reconstitution due to late-stage immunosuppression prior to ART.

Discussion: WHO prioritizes ART for HIV-infected infants due to high mortality(1,2) but diagnosis is complicated as circulating maternal IgG renders rapid antibody tests nonspecific and resource-limited settings lack prompt virological testing. WHO outlines criteria for presumptive diagnosis of severe HIV disease and recommends urgent ART after diagnosis(2,3).This case reveals diagnostic dilemmas posed by infants vertically infected with HIV after late maternal incident infection i.e. mother’s acquisition of HIV during pregnancy or breastfeeding.Unlike the limited acute infection seen in adults, this infant became severely ill from HIV before mounting his own antibody response and did not have transplacentally inherited maternal IgG to elicit a positive antibody test.Pathophysiological factors of acute maternal infection, like high viral load and lack of maternal antibody protection(4,5), likely accelerate disease progression in all infants infected in this manner and, therefore, cases of HIV-infected, antibody-negative infants with late-stage disease may be more common than recognized

Conclusion: Hospitalists working in sub-Saharan Africa must be equipped not only to care for HIV-infected and exposed patients, but also have a strong understanding of HIV pathophysiology and epidemiology to interpret and adapt WHO and country-specific ART guidelines for individual cases. As prevention of mother-to-child transmission optimization leads to overall decline in new pediatric HIV infections, an increasing percentage will be attributable to late maternal incident infection, which studies show is common across Sub-Saharan Africa (4,5).As demonstrated here, HIV disease in infants infected in this manner are at risk of going undetected and untreated based on current guidelines. Therefore, mortality will approach 100%. In settings with high rates of late maternal incident infection, there must be a high degree of suspicion for HIV disease in all ill children as empiric ART may be lifesaving. Furthermore, until point-of-care virological testing is widely available, the WHO criteria for presumptive diagnosis of severe HIV disease should be expanded to include any HIV exposure.

Resources:
Thyrotoxicosis Presenting as Intermittent Weakness with Hypokalemia

**Authors:** Abby Werner, MD, Levine Children's Hospital at Carolinas Medical Center

**Case Presentation:** 15-year-old healthy male presenting with acute onset of bilateral leg weakness. He awoke suddenly at 3am unable to move his legs or shoulders. Arm weakness resolved quickly but he needed to drag himself downstairs, noting pain in hips and thighs with mild shoulder soreness. History revealed 3 similar but less severe episodes which resolved spontaneously in a few hours, not in setting of exercise or illness. ROS otherwise negative except for 10 pound unintentional weight loss. Strength had already started recovering during EMS transport. ED’s exam demonstrated 4/5 strength in bilateral hip flexors and 4+/5 flexor and extensor at the knees, with intact reflexes. Labs revealed normal CBC and UA, ESR 12, K 2.6, and CPK 389. He received IVFs and 40mEq oral K, then transferred to tertiary center. All symptoms had resolved on arrival. The next morning he was again hypokalemic to 2.4, with low TSH and elevated FT4, and intermittently tachycardic and hypertensive. Endocrinology diagnosed Thyrotoxic Periodic Paralysis from Graves’ disease. Symptoms resolved with methimazole and propranolol.

**Discussion:** Hypokalemic periodic paralysis is a well-known phenomenon in Asian populations, with 10-20x higher incidence than in non-Asian countries. It can be the presenting sign of thyrotoxicosis. The triad of acute hypokalemia, muscle paralysis, and hyperthyroidism is an endocrinologic emergency given potential cardiopulmonary decompensation. It classically presents in young adult males (ages 20s-40s) with recurrent transient muscle weakness, often after a heavy meal or in the early morning.
Initial hypokalemia results from over-activation of Na+/K+ ATPase channels in skeletal muscle due to thyroxine, catecholamines via beta-adrenergic effects and downstream cAMP transcription, insulin, and androgens. Na+/K+ ATPase activity is 80% higher in TPP patients than in thyrotoxic patients who do not develop TPP. Several mutations in outflow potassium channels contribute to this phenomenon.

Treatment requires immediate stabilization of hypokalemia with transition to, and maintenance of, a euthyroid state. Non-selective beta blockers block the hyperadrenergic activity and prevent recurrent paralysis.

**Conclusion:** In cases of acute onset weakness or paralysis, especially in young Asian males, consider thyrotoxic periodic paralysis in the differential. Symptoms can range from very mild to complete flaccid paralysis. It can often be the presenting sign in a thyrotoxic state. Definitive management requires maintenance of euthyroid state, addition of propranolol and a low carbohydrate diet.

**Resources:**


**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A146

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**Title:** Hypoxia: thinking outside the oxygen tank  
**Authors:** Elizabeth Hillebrand, MD, Mayo Clinic  
Carley Udland, MD, Mayo Clinic, Maria Eguiguren Jimenez, MD, Mayo Clinic, Erin Knoebel, MD, MPH, Mayo Clinic  

**Case Presentation:** A 19 month old boy presents with a 1 week history of fever, rhinorrhea and cough. Vaccines are up to date including the influenza vaccine. Past medical history is notable for a lower respiratory tract infection with wheezing and hypoxia 5 months ago for which he was treated with a course of steroids and albuterol.

Physical examination: O2 saturation 75%, respiratory rate 24. He is alert, active, with no respiratory distress. Lung exam is notable for scattered wheezes.

Laboratory/radiology results: hemoglobin 14.0, parainfluenza virus 1 positive. Chest x-ray shows airspace opacity in the left upper lung laterally and right perihilar bronchial wall thickening, similar to x-ray 5 months ago.

Treatment was initiated with continuous high flow nasal cannula O2, albuterol, amoxicillin and prednisolone. Despite titration up to 10L high flow nasal cannula with 100% FiO2, O2 saturations remained in the upper 80’s. After no improvement in hypoxia, a chest CT scan with IV contrast was performed and revealed a large pulmonary arteriovenous malformation in the left upper lobe.

**Discussion:** Pulmonary arteriovenous malformations (PAVMs) are structurally abnormal configurations of vessels that provide direct communication between the pulmonary and systemic circulation and create an anatomic “right to left” shunt. PAVMs generally present after puberty but may be present during childhood. Clinical features of PAVMs can vary widely but the most striking clinical finding is often the presence of asymptomatic hypoxemia due to right-to-left shunting. Hemorrhage of the thin-walled structures is rare but a major concern.

The most common cause of PAVMs is the hereditary hemorrhagic telangiectasia (HHT). HHT affects ~1-5 in 8,000 people and is transmitted as an autosomal dominant trait. Disease manifestations are not present at birth but develop with increasing age. Arteriovenous malformations and telangiectasias develop in multiple sites including nasal, mucocutaneous, pulmonary, hepatic, gastrointestinal and cerebrovascular beds. Patients with HHT typically present with recurrent nosebleeds, iron deficiency anemia and/or complications of previously silent AVMs.

**Conclusion:** Further history revealed evidence of telangiectasias and epistaxis in the maternal lineage. The patient was found to have a heterozygous mutation in exon 3 of the ACVRL1 gene indicating hereditary hemorrhagic telangiectasia.
type 2 known eponymously as Osler-Weber-Rendu syndrome. A brain MRI revealed no concerns for cerebrovascular AVM.

The patient underwent catheterization with placement of 2 large vascular plugs into the largest contributing feeding vessel. At one month follow-up, he continued to have persistent flow through the arteriovenous malformation, but oxygen saturations were 95% on room air. It was explained to the family that the patient is at risk for profound hypoxia with viral infections due to increased pulmonary vascular resistance in the normal lung parenchyma resulting in increased right to left shunting in the AVM. Patient may require further transcatheter procedures on an as-needed basis.

Resources:

References:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A147

Title: 13-year-old with Fever After Foreign Travel – A Harbinger for Infection?
Authors: Colleen Mathis, M.D., University of Michigan
Ashley Dehudy, M.D., University of Michigan

Case Presentation: An adolescent female presented with 2 months of dyspnea, malaise, fever, and loose stools beginning 2 weeks after a trip to Mexico. Fevers were initially sporadic and breathing problems were exertional. She also developed headache and loose stools. She had fevers for 7 days until remitting with Amoxicillin. Headache then began to awaken her from sleep; daily fever also returned.

In the Emergency Department, vital signs were T 36.8°C, BP 133/81 mm Hg, HR 145 beats/minute, and RR 36 breaths/minute. She appeared non-toxic with bitemporal tenderness. Labs were notable for WBC 16.2 K/uL, Hgb 10.9 g/dL, CRP 3.7 mg/dL, and BMP within normal limits. Blood culture was pending; UA was negative.

On repeat exam, she had fever, headache, and new onset word-finding difficulty. Brain MRI was unrevealing and CSF studies were negative. Thyroid studies resulted with TSH <0.01 mIU/L and elevated T3 and T4. Pediatric Endocrinology was consulted for thyrotoxicosis. The patient was started on propranolol and later methimazole with resolution of symptoms. Imaging was consistent with Graves' disease.

Discussion: Hyperthyroidism in children and adolescents is relatively rare, occurring in 0.1-3/100,000 children; 95% of these patients are diagnosed with Graves' disease[1]. Graves' disease involves the production of autoantibodies which stimulate the thyroid stimulating hormone receptor. This causes uninhibited production of thyroid hormones resulting clinically in increased T3 and T4 levels; TSH levels are suppressed [1, 2]. Signs and symptoms of hyperthyroidism are similar in children and adolescents to those in adults. However, there is often a considerable delay in diagnosis ranging from five months in pubertal children to eight months in prepubertal children[3]. Presentation often involves weight loss, frequent stools, fatigue, palpitations, dyspnea, nervousness, emotional lability, and warm, moist skin.

Conclusion: Although many fevers of unknown origin (FUO) are ultimately infectious in nature, accounting for more than 50% of pediatric cases, it is important to recognize potential causes other than infection [4]. These include oncologic, autoimmune, endocrine, inflammatory, neurologic, or developmental conditions which are important to recall in developing a differential diagnosis for FUO [4]. This case represents a rare presentation of thyrotoxicosis in a pediatric patient. It highlights the importance of recognizing heuristic bias; even with preceding international travel, fever is not always due to an infectious cause. Thyroid studies should be considered in patients with fever of unknown origin depending on associated symptoms.

Resources:

References

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A148
Title: 2 year-old with vaginal bleeding: A precocious puzzle

Authors: Emily Albrecht, DO, University of Texas at Austin - Dell Children’s Medical Center
Jorge Ganem, MD, University of Texas at Austin Dell Medical School

Case Presentation: A 2 year-old female, just arrived to the United States from Afghanistan as a refugee with her family, is brought to care by her parents for worsening vaginal bleeding. The vaginal bleeding began when the patient was 1 year of age and occurs every 30-45 days. Father reports the patient was receiving monthly intramuscular injections to “control” the bleeding. Exam revealed an alert, non-verbal patient in no distress. Frontal bossing with coarse facial features, and hypertelorism. A small mass was palpable in the RLQ. Tanner stage I breast and genital development with no signs of adrenarche. The external genitalia were normal female, without clitoromegaly or lesions, no active bleeding. No rash or skin lesions. Generalized hypotonia. An abdominal and pelvic US revealed a menarchal uterus with enlarged ovaries bilaterally with complex cystic lesions. Radiographic bone age survey revealed delayed bone age. LH <0.005 mIU/mL (low), FSH 8.8 mIU/mL (normal mid-cycle level), high TSH >300 mcIU/mL and low FT4 0.18 ng/dL. Brain MRI revealed a large pituitary mass with suprasellar extension.

Discussion: The diagnosis is isosexual precocity secondary to Van Wyk-Grumbach syndrome (VWGS). VWGS is defined as the constellation of hypothyroidism, delayed bone age, and isosexual peripheral precocious puberty. Though the exact mechanism of precocious puberty in VWGS is unknown, it is believed to stem from overlap or lack of specificity in the feedback regulation of multiple hormones. Although the most common cause of juvenile hypothyroidism in patients with VWGS is autoimmune thyroiditis, it is likely our patient had undiagnosed congenital hypothyroidism. The medical team initially considered McCune Albright syndrome (MAS) as the diagnosis. Both conditions are associated with pituitary masses and isosexual precocity with a menarchal uterus and ovarian cysts. Serum hormone levels and radiographic bone age can be helpful in differentiating the two. In VWGS, the pituitary adenoma is associated with elevated TSH, depressed free T4, and pre-pubertal LH with delayed bone age. Treatment of VWGS is thyroid hormone replacement. Oophorectomy or adenoma excision is not part of routine management.

Conclusion: Although Van Wyk-Grumbach syndrome is uncommon, the chief complaint of vaginal bleeding in children is a common presentation requiring a broad differential including hormonal and non-hormonal causes. Narrowed to hormonal causes, both precocious puberty and malignancy must be considered. Peripheral precocity itself carries a broad differential including ovarian cysts, ovarian tumors, primary hypothyroidism, exogenous sex steroids, adrenal pathology, and McCune Albright Syndrome. A common initial work-up can guide the physician to the correct diagnosis when a broad differential is considered. This work up should include MRI of the brain, pelvic ultrasound, bone age, serum LH, FSH, TSH, T4, and estradiol and testosterone levels. In the case of our patient, a broad initial workup revealed the diagnosis and definitive therapy was initiated promptly. Prompt diagnosis is essential as patients do not require therapy to delay the onset of puberty upon correction of hypothyroidism. Symptoms of precocity regress and patients enter puberty at an appropriate age with thyroxine replacement.

Resources:
References
Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A149
Case Presentation: A 3-week-old former 39-week male presented to the Emergency Department (ED) with a rash. One-week prior, he developed purple papules on his forehead, which spread to the abdomen and inguinal area. He had otherwise been afebrile, gaining weight, and developing normally. He was born to a gravida 3 para 3 mother with prenatal care since the 1st trimester and no complications with pregnancy or delivery. He passed his hearing screen, had two normal newborn screens, and mother’s serologies were negative. He had two healthy older siblings and no sick contacts, travel history, or exposures. In the ED he was afebrile with stable vital signs and a normal exam except for ecchymotic, purpuric papules and macules on the scalp, forehead, trunk, and inguinal area. He was admitted to the pediatric hospital medicine team for further work-up. Labs were notable for normocytic anemia (13.0 g/dL) and elevated LDH (2014 IU/L). Cerebral spinal fluid, liver function and coagulation studies were all normal. A biopsy of the skin lesions demonstrated myeloid blast tumor cells diagnostic for myeloid sarcoma.

Discussion: Although rare, myeloid sarcoma and acute leukemia should be considered for a neonate with a “blueberry muffin” appearing rash. It is important to consider a wide differential including infectious causes, as each potential diagnosis can cause severe and long-term sequelae if left untreated.

Conclusion: A “blueberry muffin” rash in a neonate is a challenging presentation with a large differential, including infectious etiologies (TORCH infections), blood dyscrasias, malignancy, and skin disorders, among others. A multidisciplinary team may be required to diagnose a patient with this presentation, and ultimately a biopsy revealed the diagnosis in this case. Notably, the blood counts and peripheral smear were normal except for a mild anemia, and it is important to recognize that normal blood counts do not rule out leukemia. Myeloid sarcoma is a rare condition of immature myeloid cells that form a tumor in an extramedullary site and can occur de novo or concurrently with acute myeloid leukemia (AML) (1). It occurs in only 11% of patients with AML when considering all extramedullary sites (2). The rest of his workup revealed central nervous system involvement and his bone marrow biopsy showed 43% leukemic blasts, confirming a diagnosis of AML with cutaneous myeloid sarcoma. He went into complete remission following two cycles of chemotherapy.

Resources:

Figures 1 and 2:
References:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A150

Title: A Case to Make Your "Hair Stand on End"

Authors: Divya Lakhaney, MD, New York-Presbyterian Morgan Stanley Children's Hospital, Columbia University Medical Center
Teresa McCann, MD, New York-Presbyterian Morgan Stanley Children's Hospital, Columbia University Medical Center

Case Presentation: A healthy 9 month old girl presents with irritability and inability to bear weight on her left leg for 3 weeks and fever to 38.2°C for 1 day. Her mother also noted a bump on her head following minor trauma. On exam, she was febrile and tachycardic with unilateral parietal soft tissue swelling, truncal hypotonia, increased lower extremity tone, and refusal to bear weight on her leg. She had a normocytic anemia with Hgb 8g/dL and RDW 17%. Inflammatory markers were elevated with a normal WBC count. Hip and leg x-rays obtained for her inability to bear weight were normal. A CT scan of her head to evaluate the swelling revealed a “hair on end” appearance of the calvarium, suggestive of marrow hyperplasia, prompting a hematologic work up. An MRI of the spine obtained due to her neurologic exam showed abnormal marrow signal and retroperitoneal lymph nodes suspicious for metastatic disease. An abdominal CT scan revealed a retroperitoneal mass consistent with neuroblastoma. The diagnosis was confirmed by biopsy with osseous metastases on PET scan. She underwent chemotherapy and tumor resection.

Discussion: Neuroblastoma is the most common solid extracranial tumor of childhood. It can arise anywhere throughout the sympathetic nervous system. Presentation reflects the location of the primary tumor and metastatic disease. Signs and symptoms include abdominal mass, fever, weight loss, bone pain, anemia, hypertension, and paraneoplastic syndromes. Metastatic disease usually involves the bone, bone marrow, skin and liver. This case was interesting for the “hair on end” appearance of the calvarium, usually indicative of marrow hyperplasia accompanying chronic hemolytic disease, although this is atypical in children under five. In our patient, this radiographic finding resulted from hyperplasia in the setting of metastatic disease. If a “hair on end” appearance of the calvarium is noted in a child below five, metastatic disease, particularly from neuroblastoma, a tumor that commonly metastasizes to the skull, must be considered. Seemingly unrelated findings in this case confounded our patient’s clinical picture and led to delay in diagnosis as the team evaluated her symptoms individually.

Conclusion: Neuroblastoma can have a wide range of presenting features depending upon the site of the primary tumor and extent of metastatic disease. Because of its varied presentation, neuroblastoma should be considered in patients
with seemingly unrelated signs and symptoms. A high index of suspicion is required to allow for prompt diagnosis and management. When presented with a patient with a variety of disparate symptoms, a single diagnosis should be sought that accounts for all clinical features rather than attributing a different diagnosis to each.

Resources:

References
Title: An Eye for an Eye, a Tooth for a Tooth
Authors: Samantha Cappetto, MD, University of Kentucky
Alan Hall, MD, University of Kentucky

Case Presentation: "A 14 year-old boy presented with headaches, emesis, and a 20 pound weight loss. A month prior, he had a fever of 1 week duration; he was treated with 5 days of amoxicillin for otitis media and then completed 10 days of cefdinir due to persistent symptoms. Abduction of his left eye was limited with bilateral optic disk margins blurred. Multiple dental carries were seen. He had a left tympanic membrane defect with drainage of purulent material. The mastoid processes had no erythema, tenderness, or warmth. White blood cell count, erythrocyte sedimentation rate, and C-reactive protein were normal. A brain MRI showed a left transverse and sigmoid dural venous sinus thrombosis extending to the jugular foramen. A CT scan also showed otitis media with erosion into the left incus and petrous temporal bone with left mastoiditis. He underwent a mastoidectomy, tympanostomy tube placement, drainage of a Bezold abscess, and extraction of 6 teeth. Surgical cultures grew Enterobacter aerogenes, Streptococcus anginosus, and Pseudomonas putida with 6 weeks of cefepime and metronidazole started."

Discussion: "Acute otitis media and its complications are commonly encountered by the pediatric hospitalist. This case describes a patient with chronic suppurative otitis media leading to a polymicrobial infection with multiple uncommon complications, including mastoiditis, Gradenigo's syndrome, dural venous sinus thrombosis, and a Bezold abscess with possible contribution of multiple dental caries. Despite the extent of the infection, inflammatory markers were normal at presentation. Gradenigo's syndrome has been defined as the triad of suppurative otitis media, facial pain along the trigeminal nerve (lacking in this case), and abducens nerve palsy secondary to infectious spread to the petrous apex (confirmed on CT in this case). Either direct extension of the infection or adjacent inflammation of the temporal bone likely led to a thrombophlebitis and hypercoagulable state leading to this patient’s sinus venous thrombosis. A Bezold abscess is a deep neck abscess that likely formed after infectious erosion of the mastoid process and spread between the digastric and sternocleidomastoid muscles" 

Conclusion: This case demonstrates that a common diagnosis (otitis media) can have many very serious complications that have become increasingly rare since the since the advent of antibiotics and vaccines. Early recognition of these complications is extremely important given their potential grave consequences. In addition, is important to not overlook other potential causes for infection, like odontogenic sources, when exploring reasons for persistent or worsening symptoms in the face of treatment failure. While, this case should not prompt providers to over-treat otitis media, it should make us mindful that serious complications can occur from these “simple” infections.

Resources:
References


Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A152

Title: An Unsuspected Pathogen Causing Acute Pyelonephritis in an 8-year old Male

Authors: Rohan Patel, MD, Morehouse School of Medicine

Case Presentation: We present an 8-year old male who presented to the ER with a history of fever of one day. Associated symptoms included decreased oral intake, non-bilious, non-bloody vomiting, as well as urinary urgency. There was no cough, congestion, rhinorrhea, body ache, difficulty breathing, or dysuria. He developed diarrhea while in the ER. Physical exam was unremarkable, with exception of fever and tachycardia. Initial studies were concerning for leukocytosis on CBC and elevated CRP. Urine analysis showed leukocyte esterase and nitrates, as well as an elevated WBC. Blood and urine cultures were sent. During his hospitalization, his urine and blood cultures grew gram-positive cocci. He remained febrile despite appropriate antibiotic coverage. An ultrasound and CT of the abdomen were obtained due to concerns for abscess formation - both confirmed only pyelonephritis of the right kidney. An echocardiogram was also obtained and was negative for endocarditis. The child defervesced with improved diarrhea and vomiting. He was discharged home to continue oral antibiotics.

Discussion: The pathogen in our patient’s blood and urine was identified as Staphylococcus lugdunensis. S. lugdunensis is a gram-positive, coagulase-negative organism first isolated in 1988 [2]. Although it is a common skin flora organism, it has been noted as a possible human pathogen. Its ability to colonize and avoid host defense responses [1] sets it apart from other coagulase-negative flora. It has been reported in a wide variety of infections including skin and soft tissue infections, urinary tract infections, and endocarditis [5]. It is known that S. lugdunensis is a common cause of clinically significant infections in adults [3]. However, what is not clear is the impact on and prevalence in the pediatric population as only sporadic cases have been reported [3,4]. We present this case due to our patient's prolonged febrile illness and the unusual pathogen recovered.

Conclusion: Staphylococcus lugdunensis is rare but can cause serious infection in the pediatric population [3]. Although the organism has been identified to have a propensity for infections involving mechanical hardware as well as immunocompromised patients, it is unclear how this organism may affect the pediatric population. Interestingly, our patient did not have any mechanical hardware, had no history of trauma, and had no significant past medical history, including no previous history of pyelonephritis. Our patient did not have any anatomical abnormalities on ultrasound or CT scan of the abdomen. He did not have a VCUG during his hospital stay. This organism can behave more like Staphylococcus aureus and has the potential to cause serious invasive infection in children [4]. As modern medicine advances, it is expected to see the prevalence of this organism to increase. This case reminded us that "classic" pyelonephritis may end up not being so "classic" after all.

Resources:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A153
Title: Drama or Trauma: A Two-Year-Old's Puzzling Secret

Authors: Stephanie Vander-Plas, M.D., Texas Tech University Health Sciences Center
Ngozi Eboh, M.D., Texas Tech University Health Sciences Center

Case Presentation: A 2-year-old female presented due to the inability to bear weight on her lower extremities. She visited her pediatrician on the previous day, where she was found to have normal development and received her Hepatitis A vaccine. Her mother denied any symptoms of recent illness. That evening, she complained of bilateral leg pain, which persisted the next morning when she refused to walk. A CBC, CMP, CRP, ESR and MRI of the lumbar spine were done prior to transfer and were normal. CSF studies, urinalysis and creatine kinase were done on arrival and were normal. Her condition rapidly progressed to include truncal ataxia, nystagmus and severe upper extremity weakness without deep tendon reflexes. An MRI of the brain and spine was then obtained which showed abnormal enhancement along the nerve roots of the caudal equina. Nerve conduction studies showed decreased response of the proximal nerves. She was given IVIG, based on a diagnosis of Guillain-Barre syndrome. The patient remained hospitalized for 6 days with gradual improvement and was transferred to a rehab facility.

Discussion: Our differential diagnosis included infectious and metabolic etiologies, with Guillain-Barre syndrome (GBS) being the highest on our differential. GBS presents as an acute ascending paralysis, often provoked by a preceding illness, causing demyelination of peripheral nerves. Incidence ranges from 0.3 to 1.3 cases per 100,000 in patients less than 18 years, with males being 1.5 times more likely to be affected than females. Treatment includes supportive care and CR monitoring, as well as IVIG for rapidly progressing cases. Steroids are ineffective. Though we did not consider vaccination as a likely etiology for her case, we did report her case to the Vaccine Adverse Event Reporting System due to the recent administration of her Hepatitis A vaccine and the lack of upper respiratory or gastrointestinal symptoms. We also encouraged the family to continue administration of her vaccines and to discuss this incident with her pediatrician at her next well visit.

Conclusion: In patients that present with lower extremity pain, with weakness and inability to walk, GBS should be placed at the top of the differential. While mild cases may simply need observation and supportive care, early diagnosis and treatment of rapidly progressive cases is critical to prevent respiratory compromise. This case posed a challenge for our team due to the severe progression with no obvious stimulus. GBS has been reported, with rare incidence, as a possible adverse effect of certain vaccines, including those against H1N1 influenza, rabies, tetanus and meningococcus. A study in 2012 regarding the incidence of GBS with hepatitis vaccines, found it to be similar to what is seen in the general population. Though vaccination is an unlikely etiology for these cases, we should care for the emotional state of a family after events such as these, especially considering the current social climate concerning vaccinations. Simultaneously, it is imperative to encourage families that current research shows the unmistakeable benefits of vaccination outweigh those rare and unlikely risks.

Resources:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A154
Title: Not Always Sepsis: A TERrible Diagnosis for an Irritable Infant

Authors: Bhavana Kandikattu, MD, University of Illinois College of Medicine at Peoria
Shane Rainey, DO, University of Illinois College of Medicine at Peoria

Case Presentation: A 4-week-old term female presented with irritability, poor feeding, and ‘floppiness’ that started one week prior to admission, progressing to somnolence and decreased movement in her upper extremities. She was afebrile with normal vitals. On exam, she was extremely irritable and cried with attempts to move her head and arms, with decreased spontaneous movements in her right arm. She had poor suck, Moro and grasp reflexes. The remainder of her exam was unremarkable. A full septic workup and antibiotics were initiated. CSF was grossly xanthochromic with 132 white blood cells and a normal differential. All cultures were negative. She progressively developed decreased lower extremity movements and reflexes. MRI of the brain and spine revealed a hemorrhagic, intramedullary spinal cord mass from C4-T5, which pathology confirmed to be a congenital immature teratoma. The mass was surgically resected she was discharged home to complete physical therapy. Repeat MRI after 3 months showed no recurrence, and her neurologic status had improved.

Discussion: An irritable neonate can present the clinician with a diagnostic challenge, as the differential diagnosis is broad. We are taught to think about, in no particular order: sepsis, ductal dependent congenital cardiac lesions, galactosemia, and non-accidental trauma, to name a few. In this case, our patient’s symptoms were due to a compressive spinal cord lesion. Although teratomas are relatively common solid tumors in children, presentation within the central nervous system is an exceedingly unusual. They are thought to be due to abnormal differentiation of fetal germ cells arising from the yolk sac. Abnormal migration of these cells during the prenatal period can lead to seeding in extra gonadal locations, most commonly in the sacrococcygeal region, but also in the brain and spinal column. These tumors are usually treated with surgical resection; although, studies have shown positive results from treatment with concurrent chemotherapy. Fortunately, our patient did not require chemotherapy as the tumor was completely surgically resected.

Conclusion: There is an extensive differential diagnosis in infants presenting with irritability and non-specific signs and the initial evaluation tends to focus on sepsis. However, space occupying lesions in the spinal cord should also be considered. Although uncommon, central nervous system teratomas need to be on the differential diagnosis list, especially in patients with abnormal neurologic findings. In our case, the ‘red flags’ that pointed to a lesion in the spinal cord were her progressive weakness and decreasing bilateral lower extremity deep tendon reflexes, each occurring after admission to the hospital. Examining an infant’s neurological status is difficult, especially when it is rapidly changing. This case illustrates the importance of diligent physical examination and repeated patient assessments to look for changing clinical status and evolution of symptoms.

Resources:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A155
**Title:** Rhadomyosarcoma Misdiagnosed as an Abscess  
**Authors:** Natalie Evans, MD, Our Lady of the Lake Regional Medical Center  
Mikki Bouquet, MD, Our Lady of the Lake Regional Medical Center, Rachel Bernard, DO, Our Lady of the Lake Regional Medical Center  

**Case Presentation:** A 9 year-old female presented to the emergency department (ED) after presumed failed outpatient treatment of a peritonsillar abscess. The patient presented to her pediatrician two days prior with symptoms of left sided odynophagia and otalagia with associated left tonsillar enlargement on exam. After worsening pain and new onset hoarseness despite compliance with antibiotics, she presented to the ED. Lab work was negative. Computed Topography (CT) of the neck with contrast was read as a left peritonsillar abscess. She was admitted for IV antibiotics and otolaryngology (ENT) was consulted. Upon reviewing the CT, ENT noted an ill-defined large retropharyngeal mass with associated bony displacement and erosion more concerning for malignancy. Pathology from the CT guided needle biopsy showed a small blue cell tumor consistent with an embryonal rhabdomyosarcoma. Oncology work-up was negative and the patient was transported to St Jude for treatment.

**Discussion:** Differential diagnosis of a retropharyngeal mass in the pediatric patient is broad including both infectious and non-infectious etiologies. Previous case studies with similar misdiagnosis of head/neck malignancies have been published. Head/neck malignancies are often misdiagnosed as common infectious or benign conditions such as retropharyngeal abscess, peritonsillar abscess, acute tonsillitis, and congenital lesions to name a few. Misdiagnosis is also related to lack of imaging or lack of consideration for the possibility of malignancy on imaging. This results in unnecessary therapy. Some warning signs of malignancy are persistent enlargement or no decrease in size of mass and failure to respond to antibiotic therapy. Our patient presented with these warning signs. Delayed diagnosis can delay treatment and affect prognosis.

**Conclusion:** Rhabdomyosarcoma of the neck can present in a similar manner to a retropharyngeal abscess. Both infectious and non-infectious differentials should be considered. Being aware of warning signs and utilizing imaging when appropriate can lead to the correct diagnosis and treatment.

**Resources:**

References:

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A156

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**Title:** What a Pain in the Neck!  
**Authors:** SangHee Kim, MD, FAAP, Ann & Robert H. Lurie Children's Hospital of Chicago  
Zarina Dohadwala, MD, FAAP, Ann & Robert H. Lurie Children's Hospital of Chicago  

**Case Presentation:** A 9 year old female with history of benign horizontal nystagmus presented with 4 days of acute, sudden onset neck and chest pain. On day 2, she developed dizziness upon standing followed by syncope. She was taken to an emergency department, where an ECG was unremarkable and she was discharged home. The neck and chest pain persisted, and she started walking stooped over with her neck arched. No headache, bowel or bladder changes, vision changes, paresthesias, or weakness. She returned to the ED where she was febrile, though no reported
fevers at home. Physical exam was notable for meningismus, decreased neck range of motion due to pain (neck held in slight flexion), and no tenderness to palpation of her spine. Nystagmus was unchanged from baseline, and the remainder of her neurological exam was normal. CSF was obtained and did not show pleocytosis. CT neck and CXR showed no retropharyngeal abscess or pneumonia, respectively. CBC was remarkable for slight leukocytosis with predominant neutrophils.

Discussion: The differential diagnosis of neck stiffness includes trauma (such as fracture, subluxation, hematoma, or muscular injury), infection (such as meningitis, deep neck space abscess, epidural abscess, lymphadenitis, vertebral osteomyelitis, or discitis), space-occupying lesions (such as tumor, cyst, vascular malformation, or Chiari malformation), demyelinating disease, or torticollis. There was possible head and neck trauma with her syncopal episode, but this happened after her symptoms started. She did not have any paresthesias, changes in bowel or bladder continence, or weakness, making spinal cord lesions less likely. Her fever made infectious causes the most likely diagnosis. Meningitis was less likely due to the absence of CSF pleocytosis. There was concern for a retropharyngeal or peritonsillar abscess, but imaging did not support these diagnoses. She did not have any physical exam findings for lymphadenitis or myositis.

Conclusion: Her blood culture grew staphylococcus aureus within 24 hours (eventually found to be methicillin-resistant). MRI of her spine showed signal changes and enhancement throughout the T2 vertebral body, consistent with vertebral osteomyelitis, with an adjacent sub-periosteal abscess. She had initially been placed on broad spectrum meningitic antibiotics, which were narrowed to IV Clindamycin based on culture and sensitivity. Neurosurgery was consulted for possible drainage of sub-periosteal abscess, but since she did not have neurological deficits, the decision was made to treat her medically. Her hospital course was complicated by bilateral pneumonia with parapneumonic effusion, thought to be secondary to seeding from bacteremia. Once fevers, respiratory status and neck pain had resolved, she was discharged home to complete a 6 week course of antibiotics. Vertebral osteomyelitis should be included in the differential of a patient with neck pain and fever without CSF pleocytosis, especially in patients with methicillin-resistant staph aureus bacteremia.

Resources:
References:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A157

Title: CMV Associated Duodenal Ulcer in an Immunocompetent Pediatric Patient
Authors: Rachel Bernard, DO, Our Lady of the Lake Regional Medical Center
          Emily Klepper, MD, Our Lady of the Lake Regional Medical Center, Julia Kuznetsova, DO, Our Lady of the Lake Children's Hospital, Ghanim Aljomah, MD, Our Lady of the Lake Children's Hospital
Case Presentation: A 13 month old Vietnamese female with atopic dermatitis presented to the Emergency Department with a febrile seizure. She was incidentally admitted for poor weight gain and with an initial work up remarkable only for an ALT of 48. The patient was found to have an oral aversion requiring nasogastric tube feeds for adequate caloric intake. She continued to fail to gain weight and underwent an EGD that revealed a duodenal ulcer. She was started on sucralfate and omeprazole. Repeat lab work demonstrated a pronounced transaminitis. Biopsies from the EGD showed viral inclusion bodies consistent with CMV duodenitis. Immune deficiency was eliminated from the differential diagnosis due to a negative HIV PCR and normal IgA and IgG levels. Her IgE levels were mildly elevated, most likely due to her atopy. An initial CMV viral load of 1080 IU/mL was trended and resolved after treatment with IV gancyclovir and oral valacyclovir. Liver enzymes also normalized. The patient was discharged home demonstrating adequate weight gain via gastrostomy tube feeds after CMV and ulcer treatment were initiated.
Discussion: CMV is a congenital or acquired herpes virus that affects all races and ages. CMV is one of the most common congenital viral infections with characteristic symptoms that include microcephaly, jaundice, hepatosplenomegaly, and rash. Infants with congenital CMV are at risk for developing hearing, vision, and developmental complications. CMV in an immunocompetent older pediatric patient can present with malaise, fever, and sweats. However, most infected with the virus are typically unaware of the diagnosis as they are asymptomatic. In our patient, the concerning symptoms included
failure to thrive and oral aversion which are not specifically characteristic or indicative of CMV infection. Although CMV can affect the entire body, it is well associated with ulcers of the GI tract, particularly the stomach. Duodenal involvement is rare and has been seen in only a handful of adults. It is well known that CMV can cause serious complications for people with weakened immune systems and pregnant women. It is unusual to find immunocompetent patients with symptomatic gastrointestinal CMV disease.

**Conclusion:** Overall, this report describes a rare case and presentation of CMV infection of the duodenum in an immunocompetent pediatric patient. We consider the clinical presentation and course discussed in this case report to be useful for pediatricians due to the vast number of patients presenting with feeding difficulties and poor growth. This presentation advocates for pediatricians to include infectious etiology in their differential diagnosis for failure to thrive. We hope that this case can lead to an earlier diagnosis and management of this treatable condition.

**Resources:**

References:

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A158

**Title:** Food For Thought: Feed A Fever?

**Authors:** Gayatri Madduri, MD, UCSF
Bradley Monash, MD, UCSF

**Case Presentation:** 13-month-old male presented with six weeks of fever and rhinorrhea with regression of motor milestones. He was status-post ablation of posterior urethral valves and took urinary tract infection prophylaxis. He had exclusively breast-fed since birth with appropriate weight gain. He was febrile and tachycardic. He had no hepatosplenomegaly. He sat upright with head titubation. Patellar reflexes were brisk. When held vertically, he stepped with his toes but could not stand. White blood cell count was 3.9 x 10^9/L with absolute neutrophil count of 290. Hemoglobin was 5.3 g/dL with mean corpuscular volume of 90 fl and reticulocyte count of 16.3 x 10^9/L. Platelet count was 45,000/mm3. Urine and blood cultures were negative. Peripheral blood smear showed megaloblasts. On further history, mother reported six years of strict vegetarianism. Further testing revealed elevated MMA and homocysteine levels confirming vitamin B12 and folate deficiencies. He was treated with intramuscular B12, followed by oral B12 and folate supplementation. He demonstrated resolution of fevers and pancytopenia.

**Discussion:** Symptoms of upper respiratory tract infection are exceedingly common in pediatrics, and may coexist with clues to more serious disorders. Fever and pancytopenia may occur with viral infections, drug reactions (e.g., TMP/SMX) or oncologic processes. In this case, megaloblastic anemia and neuromyelopathy signaled B12 deficiency. Inadequate maternal intake (e.g., strict vegetarianism) may lead to B12 deficiency in breastfed children. While B12 and folate are important in converting homocysteine to methionine, only B12 is required to convert MMA to succinyl-CoA. Thus, elevated MMA levels are more specific to B12 deficiency. Folate supplementation without B12 may correct anemia but not other symptoms of B12 deficiency. B12 aids in the production of tetrahydrofolate, which is important in the production of nucleic acids. Severe pancytopenia may stem from impaired DNA synthesis affecting rapid turnover of
hematopoetic cells. B12 is a co-factor in myelin formation, and deficiency can cause symptoms of subacute combined degeneration of the spinal cord, which may be only partially reversible.

**Conclusion:** --Pancytopenia warrants consideration of viral suppression, drug reaction, oncologic process, or nutritional deficiency.
--Maternal dietary practices impact the nutritional status of breastfed children.
--Clinical manifestations of B12 deficiency may include general symptoms (fatigue, irritability, failure to thrive), neurologic symptoms (developmental delay/regression, ataxia, abnormal proprioception), hematologic symptoms (pallor, petechiae, prolonged infections), or other features (glossitis or skin hyperpigmentation).

**Resources:** N/A

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A159

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**Title:** Getting to the heart of the matter

**Authors:** Rebecca Hartog, MD, University of Pittsburgh Medical Center

**Case Presentation:** A 16 year old girl with history of obesity presented for evaluation of pericardial effusion and unintentional 25-pound weight loss. 6 weeks prior to admission, she had a febrile illness with rhinorrhea, cough, diarrhea, and malaise. She defervesced but then had 1 month of chest pain, early satiety, emesis, diarrhea, weight loss. She had 1 mo of L wrist pain after minor trauma and 1 year of intermittent R knee pain. An abdominal CT showed a 12mm pericardial effusion, prompting her referral. Her exam was notable for BMI 98%, pulse 120s, reproducible chest wall pain and mild edema on the dorsum of her L hand. Labs revealed microcytic anemia, low iron, low albumin, ESR 117, CRP 9.09, elevated TSH 11.5 with normal free T4, mildly low C4, and normal ferritin/comprehensive metabolic panel/celiac studies/fecal occult blood/calprotectin /C3. EKG showed NSR, echo showed trace pericardial fluid and normal function. CTA showed persistent moderate pericardial effusion, no pulmonary emboli, and few mildly prominent mediastinal lymph nodes.

**Discussion:** "Several diagnoses were entertained including inflammatory bowel disease, malignancy, post-viral thyroiditis, and rheumatologic disease but she didn’t meet diagnostic criteria for any specific disease. She remained clinically stable so was discharged home. After discharge, pending test results demonstrated positive ANA 1:320 (homogenous), anti-Ro (SS-A) antibodies, anti-dsDNA antibodies, atypical ANCA, thyroglobulin antibody and thyroid peroxidase antibodies. She was started on hydroxychloroquine and steroids for Systemic Lupus Erythematosus (SLE) and synthroid for Hashimoto Thyroiditis (HT). Her initial pericardial effusion was ultimately found to be a marker of her underlying disease rather than the primary diagnosis. Her acute weight loss, multiple somatic symptoms, and abnormal labs prompted an extensive work-up, which led to her diagnosis of SLE and HT. We suspect that our patient’s acute weight loss, vomiting, diarrhea, and fevers were the result of Hashitoxicosis that was resolving and becoming hypothyroidism by the time of diagnosis."

**Conclusion:** Up to 60% of patients with SLE have a pericardial effusion, which is one of the diagnostic criteria for SLE. SLE is also associated with high rates of autoimmune thyroid disease and elevated anti-thyroglobulin levels. In one series (Antonelli et al 2010), the odds ratio for subclinical hypothyroidism for female patients with SLE was 4.5. In contrast, patients with autoimmune thyroid disease do not have higher rates of SLE diagnosis. The majority (80%) of pediatric patients diagnosed with HT are asymptomatic at diagnosis, with 6-14% presenting with “Hashitoxicosis,” which is marked by unregulated thyroid hormone release during autoimmune-mediated destruction of the thyroid gland. Pericardial effusion can occur in up to 25% of patients with hypothyroidism but is very rare in hyperthyroidism. Interestingly, there is a case report of a Hashitoxicosis with pericardial effusion. It is possible that our patient’s effusion was attributable to both her SLE and HT.

**Resources:**

**References**
Title: If the Ear Keeps Bleeding, Keep Thinking

Authors: Tyson Tidwell, DO, University of Texas at Austin Dell Medical School
Peter Gilbreath, MD, University of Texas at Austin Dell Medical School, Jorge Ganem, MD, University of Texas at Austin Dell Medical School

Case Presentation: 10-year-old male with autism and chronic suppurative otitis presents with a 3-week history of bilateral facial pain and bloody ear discharge. Exam significant for severe bilateral parotitis and suppurative, hemorrhagic otitis. After minimal improvement on systemic antibiotics, patient underwent bilateral parotidectomy. The procedure was complicated by post-operative sepsis and pan sinusitis, multiple surgical washouts and prolonged antimicrobial therapy. No pathogen was isolated despite extensive Infectious Disease workup for common and uncommon organisms. Differential diagnosis expanded to include rheumatologic conditions such as sarcoidosis and vasculitides. Workup including ANA, ANCA and ACE was not diagnostic. After some clinical improvement, patient was discharged home. He returned with new onset right hemiplegia and found to have a left basal ganglia hemorrhage. Further workup with imaging studies showed multiple non-calcified lung nodules as well as several heterogeneous renal lesions. Biopsy of renal lesions was positive for severe necrotizing granulomatous inflammation.

Discussion: Although GPA can affect patients of all ages, it is most common in adults. Most estimates place the incidence in children at less than one case/million person-years. A review of the medical literature reveals it can present rapidly or over a period of months. Typically reported ENT manifestations include nasal crusting, sinusitis, otitis media, otalgia, otorrhea, persistent rhinorrhea, purulent/bloody nasal discharge, oral and/or nasal ulcers, and polychondritis. Chest radiograph findings can be variable including nodules, patchy or diffuse opacities, fleeting pulmonary infiltrates, and hilar adenopathy. Glomerulonephritis on presentation is uncommon, but most patients develop it within 2 years of disease onset. Parotid gland pathology in GPA is very uncommon. Approximately 92 percent of patients with either GPA have a positive ANCA, depending upon severity of disease. Proteinase 3 (PR3-ANCA) is more commonly associated with ENT disease and can be positive in 80-90% of patients with GPA while Myeloperoxidase (MPO-ANCA) is more commonly associated with Microscopic Polyangitis.

Conclusion: The triad of upper respiratory findings, lower respiratory findings and necrotizing granulomas is consistent with the diagnosis of Granulomatosis with Polyangitis (GPA). Although initial serology for ANCA levels was not diagnostic, further follow up testing revealed MPO-ANCA was negative, but PR3-ANCA was positive, confirming the diagnosis. He was treated with IV corticosteroid pulse followed by a prolonged oral corticosteroid course. He also received infusions of IV rituximab. The patient responded well to treatment, hemiplegia improved with intense rehabilitation. This case was challenging due to the unusual presentation of GPA. The initial diagnostic and therapeutic focus was of chronic infectious etiologies such as AFB or resistant sinus or oral flora. Although non-infectious causes were considered and worked up, it was not until a very serious complication occurred that the diagnosis became clear. Prior to the cerebrovascular accident, the patient had not exhibited obvious lower respiratory or renal complications.

Resources:
Citations:
Title: When FAST is slow: delay to diagnosis of stroke in a toddler  
Authors: Stephanie Ryan, MD, MPH, University of Vermont Children's Hospital  
Karen Leonard, MD, UVM Children's Hospital  
Case Presentation: An 18 month old boy presented with acute neurological deficits in the context of 10 months of failure to gain new developmental milestones and loss of gross motor skills. He had been developing normally until 9 months of age when he lost the ability to pull to stand. From 9 to 18 months of age he had progressive hypertonia in his lower extremities, developed clonus of his ankles and began having difficulty sitting without assistance. At 18 months of age his PCP referred him to the ED for two days of refusal to use his right arm, left sided facial droop and new difficulty chewing and swallowing. On hospital day 2, he had a head MRI which was notable for multiple areas of ischemic stroke. He had a normal cardiac echo and an equivocal hypercoagulability workup, so he was discharged home on aspirin. One month later he was found to have multiple new ischemic infarcts, so was switched to Etanercept. He continued to have strokes on Etanercept, so whole genome sequencing was done, and he was diagnosed with Aicardi Goutieres syndrome, a progressive leukodystrophy.  
Discussion: The incidence of stroke in children is 1-13/100,000 children/year. About 50% of childhood strokes are ischemic and 50% are hemorrhagic. Of those children who have ischemic stroke, 50% have a significant family history or risk factors for stroke, 25% are considered idiopathic, and 20% will be diagnosed with a stroke mimic such as ADEM or seizures. Of strokes where an etiology is identified, the primary causes are an arteriopathy (i.e. arterial dissection), a prothrombotic condition (protein C or S deficiency, antiphospholipid antibody syndrome, elevated lipoprotein a) or an infection (VZV). There is often a delay in diagnosis due to the variation in clinical presentation, the rarity of stroke in children and the lack of validated bedside stroke scales in children. Workup for suspected stroke should include brain MRI, MRA/MRV, cardiac echo, coagulation labs, and a thorough family history. The acute management is supportive care and aspirin 3-5mg/kg/day. Thrombolysis with tPA or mechanical thrombectomy are not currently recommended in children but are under study.  
Conclusion: It can be difficult to distinguish strokes from stroke mimics in children, which can lead to a delay in diagnosis. Head imaging, ideally an MRI/MRA, should be obtained in any child in whom stroke is suspected. Children who have strokes need a thorough workup to identify an etiology as the highest risk of recurrence is in children who have identified risk factors for strokes.  
Resources:
Case Presentation: An 11 year old male with a past history of headaches and a family history of Marfan Syndrome was admitted for fever, dyspnea, acute bilateral leg weakness, and palmar rash after a cross country camping trip. He was ill appearing, thin, febrile, and hypoxic (SpO2 87%) on arrival. Cardiac exam was notable for a loud S2 and digital clubbing. A full neurologic exam was normal and his leg weakness had resolved. Skin exam was notable for a maculopapular rash on extremities and palms.

He was started on broad spectrum antibiotics. Acute coxsackie virus titers were positive. CT-angiogram showed right atrial (RA) enlargement, right ventricular hypertrophy (RVH), and dilation of the main pulmonary artery (PA). An echocardiogram showed a large, posterior, peri-membranous ventricular septal defect (VSD), with right to left shunting. He was started on inhaled nitric oxide (iNO), and underwent cardiac catheterization, which confirmed a large VSD, elevated PA pressure, increased pulmonary vascular resistance (PVR) to 20 Wood units, and an ultimate diagnosis of Eisenmenger Syndrome (ES).

Discussion: ES is a relatively rare adolescent diagnosis. Undiagnosed or non-repaired truncus arteriosus, VSD, and ASD are the most common lesions to progress to ES in the pediatric population. The diagnosis is difficult to make clinically, as the majority of patients who have already progressed to ES will not have a murmur on presentation. Diagnosis is confirmed with cardiac catheterization, demonstrating elevated PVR, right heart, and PA pressures. Oxygen, iNO,
phosphodiesterase inhibitors, vasodilators, and endothelin cell antagonists are the mainstays of treatment. If surgical repair of the CHD is an option, it is recommend, but the patient will likely continue to experience symptoms of pulmonary hypertension, and thus may ultimately need heart/lung transplant. Our case of undiagnosed CHD was complicated by a family history of Marfan Syndrome and a recent extensive travel history. Our patient's physical exam and CT imaging lacked classic Marfanoid features and genetic testing was negative. His thin body habitus and failure to thrive were better explained by his undiagnosed CHD.

**Conclusion:** The symptoms of failure to thrive, weakness, clubbing, and hypoxia should warrant a workup for congenital heart disease, even in the absence of a heart murmur. Annual medical follow ups can increase the probability of detecting underlying CHD, and decrease the probability of its progression to Eisenmenger syndrome (ES). In our case, acute viral infection likely caused increased cardiac stress, leading to his decompensation and ultimate diagnosis.

**Resources:**

*Figure 1:* CT angiogram with PE protocol (axial and oblique views) demonstrating significantly enlarged main pulmonary artery, right atrial enlargement, right ventricular hypertrophy, and dilation of the aortic root in a patient with Eisenmenger Syndrome.

*Figure 2:* Echocardiogram with agitated saline "bubble study" with right to left shunting across a large, posterior, peri-membranous, VSD.

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References:


**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A163
Case Presentation: An 11 yo pre-menarchal girl with recent diagnosis of functional constipation presented with worsening abdominal pain, decreased stool frequency, dysuria, hesitancy, and acute urinary retention. Abdominal pain and constipation initially developed 3mo prior and persisted despite scheduled miralax, resulting in school absences and frequent medical visits. She was otherwise healthy with good diet, no stressors and no concerns for abuse. Abdomen was distended with positive bowel sounds, bilateral lower quadrant tenderness with guarding and palpable stool. An enema elicited large stool yet urinary retention persisted. Bladder scan showed distended bladder and catheterization produced ~1.5L urine. Patient was admitted with diagnoses of severe constipation and failure of outpatient therapy. Pain and urinary hesitancy persisted in spite of GI cleanout. A pelvic ultrasound (Figure 1) showed markedly distended vagina with fluid extending to the cervix confirming diagnosis of imperforate hymen with hematocolpos.

Discussion: Imperforate hymen, a rare diagnosis occurring in approximately 1 in 2000 girls, is the most common congenital cause of genital outflow obstruction in females and can remain undetected until onset of menses. Hematocolpos is the vaginal distention with menstrual blood which induces pressure on bladder, urethra and bowel resulting in constipation and urinary retention. Several similar case studies in the literature demonstrate that patients who show symptoms other than cyclic abdominal pain seem to reach final diagnosis with greater delay. Hematocolpos can easily be missed when the diagnosis is not considered and a genitourinary exam not performed. A retrospective database study found that in girls younger than 8 yrs, imperforate hymen was found incidentally in 90% of cases while 100% of older girls were already symptomatic at presentation similar to the patient in our case. Our patient had a total of 5 ER visits and 2 hospitalizations before final diagnosis was made, resulting in significant financial and social burden for the family and healthcare system.

Conclusion: An external genitourinary exam following the pelvic ultrasound revealed a tense membrane tender to light palpation encompassing the introitus (Figure 2). Gynecology was consulted and pt underwent a hymenectomy with complete resolution of symptoms approximately 3 months following initial onset. Functional constipation is ubiquitous and hospitalization for failed outpatient therapy common. A diagnosis of imperforate hymen and hematocolpos should be considered in pre-menarchal girls presenting with abdominal pain, especially while cyclical and accompanied by constipation and urinary retention. A complete physical exam, including an external genitourinary exam, as part of routine clinical practice and reassessment of the differential diagnosis are essential in avoiding delay in diagnosis and management.

Resources:

Figure 1. Pelvic ultrasound showing vaginal distention (hematocolpos); sagittal (left), transverse (right)
References

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A164

Title: A rare cause of upper airway obstruction in an infant
Authors: Mark Corden, MD, Children's Hospital Los Angeles
Case Presentation: A 6-week-old female was admitted with respiratory distress after 3 weeks of nasal congestion and progressive dyspnea. She was choking with feeds and had chronic eye discharge. Stridor associated with cyanosis prompted referral to our facility. Perinatal history was remarkable for complete lack of prenatal care and unattended home birth. On exam, she was malnourished (weight < 3%ile), with a gasping respiratory pattern, intermittent inspiratory stridor, and significant retractions. Lungs were clear. Left eye discharge without conjunctival injection also was noted. After racemic epinephrine nebulization and IV dexamethasone, the patient's respiratory status stabilized and stridor resolved. Neck and chest radiographs demonstrated increased central bronchovascular markings with hazy perihilar density. B pertussis PCR and respiratory viral panel were negative. Evaluation by otolaryngology showed normal anatomy. The patient gradually improved with supportive care. On the day of discharge, direct fluorescent antibody (DFA) test of nasal aspirate came back positive for Chlamydia trachomatis.

Discussion: C trachomatis is an obligate intracellular organism that is acquired by ~50% of infants born vaginally to infected mothers. Respiratory tract infection with C trachomatis may be asymptomatic. Our patient exhibited the hallmark features of nasal congestion, onset of symptoms before 8 weeks of age, presentation at 4 to 11 weeks of age, gradually worsening symptoms, absence of fever, and presence of conjunctivitis. The patient’s radiographic abnormalities were also consistent with chlamydial pneumonia (Figure 1). Other findings (not seen in our patient) include staccato cough, rales, and eosinophilia. Her respiratory difficulties on presentation were likely related to severe
Additionally, the chronicity and progressive severity of her symptoms had limited the patient’s ability to feed, contributing to her failure to thrive. Untreated, the infection may linger for weeks, and long-term sequelae in pulmonary function have been observed. Our patient was treated with three days of azithromycin, further perinatal infectious work-up was pursued, and the mother was referred for care.

**Conclusion:** Infectious processes are a common cause of acute upper airway obstruction and respiratory distress in infants. However, our case represents an unusual etiology. In fact, it is rare for *C. trachomatis* infection to manifest as acute upper airway obstruction. Maintaining a broad differential diagnosis while stabilizing patients and pursuing their work-ups may help capture more elusive and treatable causes, such as *C. trachomatis*. The patient's good clinical response to standard management of her airway obstruction may have contributed to a delay in definitive diagnosis. In retrospect, the chronicity of her symptoms, absence of fever, and lack of prenatal care were important clues in clinching the diagnosis.

**Resources:**

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A165

**Title:** An unusual case of lower extremity swelling

**Authors:** Jonathan Uniat, MD, Children's Hospital of Los Angeles
Patricia Castillo, MD, Children's Hospital of Los Angeles

**Case Presentation:** A 7-month-old, ex-28 week preemie of twin gestation, with chronic lung disease requiring supplemental oxygen at night, bilateral hydroceles, and history of inguinal hernia repair presents to the emergency department with one week of right leg swelling. It began in the right thigh and now involves the entire leg. Parents deny fever, increased work of breathing, increased oxygen requirement, vomiting, diarrhea, abdominal swelling, skin color changes, and decreased leg movement.

Initial vital signs are normal. He is well appearing and interactive during the examination. His lung, cardiac, and abdominal exams are normal. Genitourinary exam shows bilateral hydroceles that transilluminate with bilateral palpable testes. Lower extremities have normal strength and neurological exam. Left leg circumference at mid-thigh is 24cm, knee is 19cm, and mid-calf is 17cm. Right leg circumference at mid-thigh is 27cm, knee is 22cm, and mid-calf is 19cm. Complete blood count and coagulation studies are normal. Right lower extremity ultrasound shows no thrombus in arterial or venous systems.

**Discussion:** The differential for lower extremity swelling includes vascular congestion secondary to obstruction of venous flow, vascular anomalies, lymphedema, hemihypertrophy, tumor, or post-traumatic swelling. Vascular ultrasound of the left leg is obtained for comparison and shows a nonocclusive thrombus in the left femoral vein. In order to further evaluate proximal vasculature, a MRV with contrast of the abdomen and pelvis is completed and shows...
a large right scrotal hydrocele with extension into the pelvis exerting a mass effect on right iliac vessels. Urology was consulted and recommended surgical intervention due to the size of abdominoscrotal hydrocele and the unlikely chance of spontaneous resolution.

**Conclusion:** Abdominoscrotal hydroceles are a rare subset of pediatric hydroceles. The formation is unclear, but leading theories include abdominal extension of scrotal hydroceles, high obliteration or persistence of processus vaginalis allowing cephalad development of an abdominoscrotal hydrocele. These are usually diagnosed by abdominal ultrasound, but can also be diagnosed by CT or MRI. Common sequelae of abdominoscrotal hydrocele include lower extremity edema, hydronephrosis, testicular torsion or dysmorphic testes, and hemorrhage. Indications for surgery include persistence of hydrocele, concern for development of complications, or cosmetic reasons.

In our patient, there was initial concern for proximal thrombus given incidental finding of left sided thrombus and evaluation continued with MRV. Once the abdominoscrotal hydrocele was identified, our urology team was involved and recommended surgical correction. As the patient had a previous urologist due to previously diagnosed bilateral hydroceles, the family deferred surgery at time of hospitalization.

**Resources:**

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A166

**Title:** Autoimmune Channelopathy Presents as Catecholamine Excess

**Authors:** Amy Stepp, MD, Barbara Bush Children’s Hospital at Maine Medical Center
Christin Folker, , Barbara Bush Children's Hospital at Maine Medical Center, Thomas Reynolds, DO, Barbara Bush Children's Hospital at Maine Medical Center, Marie Tanzer, MD, Barbara Bush Children's Hospital at Maine Medical Center, Jennifer Hayman, MD, Barbara Bush Children's Hospital at Maine Medical Center, Leah Mallory, MD, Barbara Bush Children's Hospital at Maine Medical Center

**Case Presentation:** A 12-year-old boy presented with 9 months of panic attacks, global pain, anorexia and 30 lb weight loss. During this time, he was diagnosed with depression, anxiety and ADHD unresponsive to medication or behavior interventions. He was admitted to two hospitals for evaluation with discharge diagnoses of somatic pain disorder and hypertension.

We admitted the patient for failure to thrive, then noted persistent tachycardia and hypertension. A broad differential of chronic pain and sympathetic overdrive was considered. Urine metanephrines/catecholamines and plasma metanephrines were high. Workup for pheochromocytoma/paraganglioma was negative.

On hospital day 26, a serum paraneoplastic panel showed autoantibodies to voltage-gated potassium channels. IVIG (2 g/kg) was given over three days with marked improvement after the first dose. He was discharged one week later with
minimal pain, resolved tachycardia, normal plasma metanephrines and good appetite. Two months later, he had gained 26 lbs, antihypertensive therapy was stopped, and repeat serum paraneoplastic panel was negative.

**Discussion:** Voltage-gated potassium channel (VGKC) antibodies are associated with a range of neurological presentations, including central, peripheral and autonomic dysfunction. This is the first description of VGKC autoantibodies associated with hypertension and tachycardia in the setting of elevated catecholamines. Etiology of these antibodies is unclear, and the specificity of the VGKC for specific disease states remains controversial. Given rapid resolution of all clinical symptoms and resolution of the serum antibodies after immunotherapy, it is likely that the VGKC autoantibodies were pathogenic in our patient.

**Conclusion:** We present an unusual case of VGKC autoimmunity in a 12-year-old boy with elevated catecholamines, hypertension, tachycardia, generalized pain, anorexia, and failure to thrive. VGKC antibodies should be considered as part of the testing in patients presenting with elevated metanephrines and catecholamine secretion, as well as those with chronic pain and other unexplained neurologic symptoms.

**Resources:**

Supporting Documentation for Clinical Conundrum: Autoimmune Channelopathy Presents as Catecholamine Excess

**References**


**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A167

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**Title:** Here's Winking at you Kid  
**Authors:** Melanie Rudnick, MD, CHLA  
Robyn Kuroki, MD, CHLA, Pia Pannaraj, MD, CHLA, Gabriela Moriel, MD, CHLA  

**Case Presentation:** 5 week old full term female presents with 2 days of inability to close right eye and form a smile on right. She had a neck rash that was improving, rhinorrhea and diarrhea which self-resolved last week. No fevers, emesis, altered mental status, or travel history. Both parents had cold sores 1 week ago, and brother had cold symptoms. On exam, she was well appearing with no right nasolabial fold. When crying, she could not close her right eyelid or create a grimace on the right. Tongue was midline, gag present, and primitive reflexes intact. A flesh-colored papular rash was noted in neck. She had normal CBC, CRP, and electrolytes, but positive respiratory syncytial virus nasopharyngeal swab. Lumbar puncture was unsuccessful. Acyclovir (20mg/kg q8h) was started empirically for possible herpes simplex virus. Brain MRI was normal. A spinal hematoma developed at the site of the lumbar puncture and remained present up to 18 days later. No CSF fluid was obtained, and she completed 28 days of IV acyclovir. Facial paralysis started to improve by day 5 of acyclovir and fully resolved by day 19.
Discussion: Facial nerve palsy is rare in infants, with only a handful of cases reported in the literature. To our knowledge, there is only 1 reported case of Bell's palsy in a child younger than our patient. Most cases are idiopathic but infectious etiologies are the most common cause if one is identified. For viral etiologies, it is unclear if due to direct viral neuropathy or secondary ischemic neuropathy. Most patients undergo an infectious disease workup, however, opinions vary on the utility of workup in children. Our patient was positive for RSV but without RSV isolated in the CSF, it is unclear if this was the cause of our patient's symptoms. RSV has not yet been reported as a cause of facial nerve paralysis. Treatment is also debated. Steroids have been shown to be of benefit in adults, but evidence is lacking in pediatric patients. Additionally there is no proven benefit of empiric antiviral therapy in children. Due to the lack of a CSF specimen we could not rule out HSV infection and treated for a full course of IV acyclovir. She improved, however facial paralysis often self-resolves

Conclusion: Facial nerve paralysis is rare in the pediatric population, especially infants. Little consensus exists on diagnostic work up and treatment of these patients and should be considered on a case by case basis. Given that HSV infection is on the differential, a complete investigation, including CSF, may be warranted. The differential for facial palsy, particularly in infants, includes many diagnoses (malignancy, encephalitis, congenital malformations) that are prudent to recognize and evaluate for, if clinical suspicion is present.

Resources:

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A168

Title: Not Your Typical Diaper Rash
Authors: Jocelyn Schiller, M.D., University of Michigan Medical School
Jennifer Jehnsen, B.S., University of Michigan Medical School, Shelby Lemke, M.D., University of Michigan Medical School, Patricia Keefer, M.D., University of Michigan Medical School
Case Presentation: A 5-week old boy presented with diaper rash and fever. Initially, his diffuse papular buttock rash was treated with barrier cream then nystatin. Later he developed many large, well-demarcated pustules, so he was started on cephalexin. Three weeks after onset of rash, he developed a fever, so he was admitted. He was well-appearing with diffuse buttock papules and pustules. Inflammatory markers were elevated; blood, urine, and CSF cultures were negative. He was diagnosed with bacterial folliculitis, so started vancomycin and ceftriaxone. Wound culture grew Serratia, Klebsiella, and Enterococcus which were thought to be contaminants. His rash and fever improved, so he was discharged on clindamycin. 3 days later, he was readmitted for worsening rash. Biopsy suggested a neutrophilic dermatosis which is often associated with malignancy, but work-up was negative. Subsequent wound culture grew Serratia. Given the association between Serratia infections and chronic granulomatous disease (CGD), further testing was sent and diagnosis of CGD was made.

Discussion: Initial therapy for this boy was targeted at treating folliculitis, a commonly considered diagnosis in the setting of a pustular rash. However, the rash persisted in the setting of appropriate antibiotic therapy. His significant
leukocytosis and inflammatory markers suggested another process might better explain the clinical picture. A
dihydrorhodamine flow cytometric phorbol myristate acetate test showed a negative oxidative burst in response to
stimulation suggestive of CGD. While Serratia marcescens bone and soft tissue infections are not a rare presentation of
CGD in infants, isolated and localized superficial infection is uncommon. Infants more commonly present with
disseminated infection including infections in the lung parenchyma, intra-abdominal abscesses, and deep soft-tissue
abscesses of the limbs. Our patient did not show signs or symptoms of disseminated infection or deep tissue abscesses.
Knowing the association between cutaneous Serratia infections and chronic granulomatous disease was key in the final
diagnosis.

**Conclusion:** This was an unusual presentation of chronic granulomatous disease in an infant. Although CGD is a relatively
rare disease, it should be considered when infants present with multiple abscesses unresponsive to conventional
antibiotic therapy, even when they are localized. Prompt diagnosis is important to begin prophylactic antimicrobials to
prevent serious disseminated infections.

**Resources:**

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A169

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**Title:** Not Your Typical Temper Tantrum...

**Authors:** Sarah Pradhan, MD, University Hospitals Rainbow Babies and Children's Hospital
Supriya Sharma, MD, University Hospitals Rainbow Babies and Children's Hospital, Allayne Stephans, MD, University
Hospitals Rainbow Babies and Children's Hospital, Jessica Goldstein, MD, University Hospitals Rainbow Babies and
Children's Hospital

**Case Presentation:** A 5 year old male initially diagnosed with post-concussion syndrome presented with 3 weeks of
agression, obsessive-compulsive behavior and nocturnal enuresis. Parents also noted altered sleep patterns and hand
wringing. After 2 negative head CT scans and normal basic labs, he was admitted. Initial examination was notable for
altered sensorium. MRI brain was normal. EEG showed bilateral fronto-temporal seizures, without clinical correlation.
Fosphenytoin therapy was started. Despite a 24-hour seizure free period, behavioral outbursts progressed to dysarthria,
oral dyskinesias, and hand dystonia. EEG showed focal slowing in the bilateral temporal lobes, concerning for focal
cortical dysfunction. Extensive metabolic, endocrine, paraneoplastic and infectious work-up was sent including lumbar
puncture. He clinically improved on empiric IV steroids for presumed autoimmune encephalitis. CSF studies were later
positive for N-methyl-D-aspartate (NMDA) receptor antibodies and oligoclonal bands, confirming a diagnosis of NMDA
encephalitis. He was discharged on a steroid taper and fosphenytoin.

**Discussion:** Anti-NMDA receptor (Anti-NMDAR) encephalitis is an autoimmune mediated paraneoplastic neurologic
syndrome, with 40% of cases diagnosed in patients <18 years of age. Females comprise 80% of pediatric-onset patients,
and can be associated with ovarian teratomas although this is more frequent in adults. Early recognition of
neuropsychiatric symptoms can pose a diagnostic challenge, and may be overlooked or misdiagnosed in young children
as psychiatric illness or abuse which can delay definitive treatment and result in significant morbidity. Although most
children eventually develop dyskinesias and seizures, rapid fluctuations of cognitive function and behaviors are more
characteristic of anti-NMDAR encephalitis. Prognosis is significantly impacted by early treatment with immunotherapy,
and recovery is slow; with resolution of neurologic symptoms often in the reverse order of presentation. Up to 80% of
patients have significant or full recovery, however children appear to have an increased prevalence of long-term neurologic morbidity when compared to other etiologies of encephalitis.

**Conclusion:** This case demonstrates the diagnostic dilemma that may arise when evaluating a child presenting with acute behavioral disturbance, and highlights the step-wise clinical approach to the diagnosis of autoimmune encephalitis.

**Resources:**

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A170

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**Title:** Say it With a Smile  
**Authors:** Sibgha Zaheer, MD, Cohen Children's Medical Center  
Jasmine Lemmons, MD, Cohen Children's Medical Center  

**Case Presentation:** A 13 year-old-boy with autism presents to a community ED with copious gingival bleeding. He had 5 weeks of progressive gum swelling and bleeding, and recent non-traumatic tooth loss. Patient was brought in after a prolonged episode of bleeding. He was found to have Hg of 3.2, with normal PT/PTT/INR. He was given a pRBC transfusion, and then transferred.

Parents also report 5 weeks of bilateral knee swelling and progressive difficulty walking, along with a lower extremity (LE) rash. Patient was treated with PO steroids by pediatrician for a presumed Henoch-Schonlein purpura, which had improved symptoms.

ROS notable for no other sites of bleeding or bruising. FH, SH are unremarkable.

On PE: Vital signs normal. Pertinent findings included:
- hemorrhagic, hypertrophic gingiva w/ active bleeding, halitosis
- petechial rash of bilateral LE , generalized pallor
- bilateral knee swelling

Oral pathology service was consulted; their workup showed:
- dental panoramic scan w/o infiltrates
- gum biopsy w/normal flow cytometry and hemorrhagic granulation tissue with minimal stromal cells

**Discussion:** Scurvy is caused by a deficiency of vitamin C. Although malignancy must remain on the differential diagnosis, nutritional deficiencies should always be considered, especially for patients with developmental or behavioral delays. This patient’s physical exam findings of gingival hypertrophy, perifollicular hemorrhages, and arthralgias were all consistent with a typical manifestation of scurvy. He recently completed a 14 day course of steroids for Henoch-Schonlein Purpura (HSP), which is a common misdiagnosis due to similar dermatologic presentations. Although the clinical findings were enough to make the diagnosis, the biopsy report and Vitamin C levels served as confirmation. Vitamin C is essential for collagen synthesis, connective tissue integrity, and iron absorption. It occurs after about 1 to 3 months of a deficient diet. Treatment consists of oral administration of 100 to 300 mg of Vitamin C daily. Most symptoms typically resolve after a few weeks, but tooth loss is permanent. This patient received oral multivitamins, with a resolution in his symptoms.

**Conclusion:** Further history taking had revealed a diet restricted to pancakes, chicken nuggets, and water. This case highlights the importance of obtaining a dietary history, especially in patients with autism and other developmental or behavioral issues. This was the key to making his diagnosis. Nutritional labs were sent, and notable for a vitamin C level of 0, decreased serum iron, folate, and vitamin A and D levels. Patient was diagnosed with scurvy, and discharged home with oral multivitamins. Patient’s symptoms had significantly improved at 1-month follow-up.

**Resources:**
Silly Rabbit! Ticks are for Kids

**Authors:** Chandni Vaid, MD, Floating Hospital for Children at Tufts Medical Center
Nicole Kalinske, MD, Floating Hospital for Children at Tufts Medical Center

**Case Presentation:** A 6 year old boy presented with 15 days of recurrent fevers with three insect bites on his scalp following a visit to Martha’s Vineyard. His pediatrician saw him 2 weeks prior and started him on 1 week of Bactrim and 3 weeks of Amoxicillin. His fevers did not improve (Tmax 104F), and his scalp lesions were growing while becoming pruritic and more painful with general malaise, headaches and myalgias.

At a community hospital, he was given IV Ceftriaxone with labs significant for elevated inflammatory markers. He was transferred to our tertiary care center where exam revealed two large fluid-filled, painful lesions on the left lower occiput region with bilateral posterior cervical lymphadenopathy. ENT performed an I&D of his two superimposed soft tissue abscesses as confirmed by CT scan. He was started on Ceftriaxone and Vancomycin, then abscess culture grew 2+ oxidase-negative gram negative rods concerning for F. tularensis. Subsequently treated with 5 days of Gentamicin while inpatient and discharged with 7 days of Ciprofloxacin. Upon 2-week follow-up, his lesions and fevers resolved.

**Discussion:** Tularemia is a rare disease endemic throughout most of Europe, Asia, and North America. The clinical presentation of Tularemia is variable depending on the route of transmission. Overall, Tularemia presents in one of six forms and the clinical signs and symptoms depend on route of inoculation: ulceroglandular (cutaneous findings and lymphadenopathy), glandular (lymphadenopathy alone), ocular-glandular (conjunctivitis with lymphadenopathy), pharyngeal (oropharyngeal findings with lymphadenopathy), typhoidal (no localized findings), and pneumatic (pulmonary findings). Most forms begin with a nonspecific and acute febrile illness. Ulceroglandular is the most common form, while pneumatic is the least common although the most severe. Cases have been reported in all states with the exception of Hawaii. The overall case-fatality rate for reported tularemia in the United States is 2%, however primary pneumatic tularemia may have an untreated mortality rate up to 60%. Treatment options include aminoglycosides as well as fluoroquinolones used with caution in the pediatric population.

**Conclusion:** In the evaluation of patients with recurrent fevers and known tick bites, Francisella tularensis and other zoonotic infections should be considered especially with travel to known endemic areas or a history of previous outbreaks. Francisella tularemia is a gram-negative rod-shaped bacterium commonly transmitted by tick or deer fly bite, inhalation of aerosolized bacteria or through contact with contaminated animal products. Failed outpatient antibiotic therapy for fever and abscess formation prompts inpatient evaluation where differential diagnoses should include zoonotic infections, such as Lyme disease, RMSF, tularemia, West Nile, malaria, dengue fever, etc... with particular attention paid to travel history and areas of endemic diseases.

**Resources:**

**Presented at:** Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A172
Title: SLE: Still the great mimicker

Authors: Cherilyn Cecchini, MD, Children's National Medical Center
Bridget Allard, DO, Children's National Health System, Jeremy Kern, MD, Children's National Medical Center

Case Presentation: A 10-year-old female with one year history of intermittent, self-resolving, facial edema presents with anasarca and cough. She is afebrile with stable vitals. Exam reveals facial edema, diminished breath sounds, non-pitting edema of all extremities, diffuse adenopathy, and palpable spleen tip. Initial labs, including CBC/differential, renal function, TSH, ASO titers, and C1 esterase inhibitor function, are unremarkable. UA shows 1+ protein, no blood and 3+ hyaline casts. Her urine Pr:Cr ratio is normal. C3 and C4 levels are very low. CXR shows bilateral pleural effusions. Abdominal ultrasound shows ascites and splenomegaly. She develops dyspnea, fevers, and tachycardia. Echo shows moderate pericardial effusion. She worsens despite lasix and IVIG, requiring chest tubes and pericardial drain. After negative oncologic workup, pulse steroids are started and her condition rapidly improves. Weeks later, she develops increasing proteinuria. A kidney biopsy shows lupus nephritis, confirming the diagnosis of systemic lupus erythematosus.

Discussion: Differential diagnosis was extremely broad. At presentation, the patient had normal kidney and liver function, no proteinuria/hematuria and no cardiac dysfunction, making nephrotic syndrome, glomerulonephritis (GN) or other renal/liver disease unlikely. Negative ASO titers ruled out post-strep GN. Hereditary angioedema (HAE) was unlikely given low C3, which typically remains intact during HAE flares. Initially, she met many of the newer SLICC lab criteria for lupus: +DAT, low complement and +ANA, but none of the clinical criteria. The anti-dsDNA was positive at a low level and deemed indeterminate. Thus, she did not qualify for lupus at this point. Hypothyroidism causing myxedema and protein-losing enteropathy were ruled out. With her lymphadenopathy and splenomegaly, oncologic process was considered, but ruled out with lymph node biopsy. As her clinical course progressed, the decision was made to credit her effusions as serositis and she subsequently met both lab and clinical SLICC criteria. Diagnosis was confirmed further with kidney biopsy in the setting of later proteinuria.

Conclusion: This is a unique presentation of SLE as well as a diagnostic dilemma that required extensive work-up and follow-up to reach a definitive diagnosis. Consideration for SLE should be given in cases of diffuse anasarca without other specific symptoms, even with initially inconclusive lab results.

Resources:

http://www.rheumtutor.com/2012-slicc-sle-criteria/
Title: We’re All Mixed-up: A case of a crying infant with spontaneous bleeding

Authors: Kevin Patel, Pediatric Resident, Children’s Hospital Los Angeles
Namrata Ahuja, Clinical Instructor, Clinical Fellow, Children’s Hospital Los Angeles, Kira Molas-Torreblanca, Assistant Professor of Pediatrics, Children’s Hospital Los Angeles

Case Presentation: An 85 day old infant with an unremarkable history presented with excessive crying to the ED and was found to be febrile. Urine and blood were sampled, however, three lumbar puncture (LP) attempts were unsuccessful. His CBC and differential was unremarkable other than a hemoglobin of 7.6 and his urinalysis was normal. Upon admission, he was irritable and un-consolable with bleeding around his IV site and fluctuance over his lower sacrum underneath the dressing of his LP. Repeat CBC showed his hemoglobin had dropped to 6.2. He had prolonged aPTT at 132, with normal PT of 11.4. Spinal ultrasound and MRI showed extensive dorsal epidural hematoma extending from mid cervical spine to the sacrum with spinal cord compression. CT head showed no acute intracranial bleed. Thought this evaluation, he retained spontaneous movement of his arms and legs, with preserved bowel and bladder function. Factor levels were sent, and Factor 8 was empirically given. Factor 8 assay returned < 1% which led to diagnosis of hemophilia A.

Discussion: Children with persistent bleeding after an invasive procedure such as LP or IV placement should be evaluated for coagulation disorders. The differential includes disseminated intravascular coagulopathy, von-Willebrand’s disease (vWD), inherited platelet disorders, Factor XI deficiency, Factor XIII deficiency, or acquired coagulation factor inhibitors. Initial screening tests include CBC, PT, PTT, fibrinogen, and peripheral smear. An isolated elevation in PTT suggests abnormality in the intrinsic pathway of coagulation. Mixing study in which the patient’s plasma is mixed 1:1 with plasma containing 100% of factors can help narrow the differential. Our patient’s prolonged aPTT corrected on mixing study which suggested he had either Factor VIII deficiency, Factor IX deficiency, vWD, or Factor XI deficiency. Factor level assays can then be used to identify the specific factor that is deficient. While awaiting results, individual or multiple coagulation factors may be given in consultation with a hematologist to stop ongoing bleeding.

Conclusion: The time sensitive diagnosis of hemophilia is critical given the significant morbidity associated with this diagnosis. Patients are now are diagnosed earlier compared to 50 years ago. Early diagnosis may be made following an episode of spontaneous bleeding such as cephalohematoma, intracranial hemorrhage, easy bruising, or gastrointestinal bleed in the neonatal period. A high index of suspicion should be maintained when a patient (or infant) presents with irritability and notable spontaneous bleeding with non-invasive interventions such as LP, IV placement, circumcision, or heel stick. Initial screening tests including mixing studies if available, followed by specialized testing such as factor levels should be evaluated in these cases. Empiric treatment with blood products such as coagulation factors should be initiated as soon as possible, in consultation with a hematologist.

Resources: N/A

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A174
Title: Worsening ataxia in an adolescent female
Authors: Sarah Marsicek, MD, Johns Hopkins All Children's Hospital
Kelli Baldwin, MD, Johns Hopkins All Children's Hospital

Case Presentation: 17 year-old previously healthy white female presented with 3 weeks of intensifying intention tremor, ataxia, and urinary incontinence, in addition to headaches and blurry vision. Neurologic exam was remarkable for end nystagmus, unilateral numbness over V1 distribution of the trigeminal nerve, afferent pupillary defect of the left eye, and 4/5 strength of right side. Cerebellar testing was significant for intention tremor and dysdiadochokinesia, most pronounced on right side. Marked wide-based gait was also noted. Initial laboratory testing, including complete blood count, thyroid studies, and comprehensive metabolic panel, was unremarkable. MRI showed extensive multifocal homogeneous and ring-enhancing white matter lesions bilaterally of the cerebral hemispheres, basal ganglia, midbrain, and pons. Spinal MRI showed several intramedullary lesions. CSF cytology was unremarkable.

Discussion: The differential was broad given the acuity of the patient’s presentation, abnormal exam, and presence of ring-enhancing lesions on imaging studies. Etiologies include infectious, autoimmune, inflammatory, and neoplastic processes. Infectious causes were thought less likely due to lack of risk factors and systemic signs. Acute demyelinating processes, such as MS and ADEM were also entertained. Uric acid, LDH, RPR, Lyme titers, HIV, ANA, RF, NMO, and NMDA antibodies were negative. Oligoclonal bands were present in the CSF. Methylprednisolone was initiated for presumptive multiple sclerosis. Significant improvement in intention tremor and gait was noted following treatment.

Conclusion: Approximately 10% of MS patients demonstrate multiple ring-enhancing lesions on imaging. Typical presenting signs and symptoms of the most common subtype of multiple sclerosis include nystagmus, ataxia, limb weakness, and urge incontinence, all of which were noted in our patient. In a large observational study, 55% of patients =12 years of age had mono-symptomatic initial presentations, as opposed to our patient who had multiple neurologic symptoms. Though the majority of MS patients present in young adulthood, it remains an important diagnostic consideration in children and adolescents. It is imperative to entertain a broad differential in cases of acute ataxia with ring-enhancing lesions and be diligent in evaluation to ensure a timely diagnosis.

Resources:
References

Presented at: Poster Session #1, Thursday, July 20, 2017, 4:15 - 5:15 p.m., Poster #A175

Title: New Player in Lemierre’s?
Authors: Sherry Gu, MD, Tulane University
Vanessa Carroll, MD, Ochsner

Case Presentation: 5-week old female transferred for neonatal sepsis with initial presentation to outside hospital of fever/fussiness, started on IV ampicillin & cefotaxime 2 days prior. On admission, she was noted to have signs of compensated septic shock and given repeatedly positive MRSA blood culture, IV vancomycin/ceftriaxone were started. Procalcitonin on admit was 22.21 ng/mL. Despite 2 days of clinical improvement, she remained persistently bacteremic, with CRP 139.9 mg/dL, while on therapeutic dosing of antibiotics. On day 4, patient developed posturing and focal seizure activity, prompting transfer back to PICU for intubation and head CT, which revealed cerebral edema and hypopattenuation concerning for ischemia vs embolic etiology. Brain MRI revealed diffuse global cerebral anoxia/edema with non-enhancement in multiple vessels, most notably in the distal left internal jugular vein and involving the left
sigmoid and transverse sinuses, a complex multi-septated left peritonsillar and parapharyngeal abscess, and areas of apical pulmonary infarct, most consistent with Lemierre’s syndrome.

**Discussion:** Septic thrombophlebitis of the internal jugular vein, commonly referred to as Lemierre’s syndrome, arises as a complication of an oropharyngeal infection. This thrombophlebitis frequently results in septic emboli to organs such as the lungs or brain. Suspicion for oropharyngeal infection commonly relies on reported symptoms such as neck pain or odynophagia, difficult to discern in an infant such as our patient, however it should remain in the differential regardless of age. Prompt and appropriate antibiotic therapy remain the mainstay of management in Lemierre’s, with additional questions such as the role of anticoagulation remaining to be unclear. Given subsequent development of hematochezia and bloody oral secretions concerning for DIC, our patient was not anti-coagulated. She continued to have blood cultures positive for MRSA for 10 days despite appropriate IV antibiotic management with vancomycin and linezolid. Investigation for additional etiologies of septic emboli included echocardiogram, which showed no evidence of vegetation, and tonsillar culture, which did also grow MRSA.

**Conclusion:** While the most commonly associated pathogen associated with Lemierre’s syndrome is Fusobacterium necrophorum, an anaerobic gram-negative organism, community-acquired MRSA has been implicated in a number of case reports in the pediatric population.

**Resources:** N/A

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B100

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**Title:** Starting From Scratch

**Authors:** Matthew Le, MD, Cincinnati Children's Hospital
Hunter Wilson, MD, Cincinnati Children's Hospital, Sanyukta Desai, MD, Cincinnati Children's Hospital

**Case Presentation:** A 9-year-old previously healthy female presented with fever, hip and chest pain. The pain began 4 weeks prior to admission in the left hip and was associated with refusal to walk. She lived in a rural area with exposure to horses, dogs, cats, and pigs. On exam, she was febrile to 102 F with mild tenderness of the left hip but with intact range of motion. CBC was normal. CRP and ESR were elevated at 4.8 mg/dL and 47 mm/hr. MRI identified osteomyelitis of the left superior pubic ramus. She was placed on IV nafcillin and transitioned to oral clindamycin after she was afebrile for 48 hours. After discharge, she continued to have intermittent fever and hip pain. Two weeks later she was re-admitted with new onset right chest pain. CRP was 4.3 mg/dL and ESR 89 mm/hr. Repeat MRI showed intraosseous and sub-periosteal fluid collection in the left hip and an abnormal marrow signal of the right 6th rib. Bartonella IgG and IgM titers were 1:512 and 1:256 respectively. The patient was diagnosed with Bartonella henselae osteomyelitis and was successfully treated with 4 weeks of azithromycin.

**Discussion:** Staphylococcus aureus is the most common organism in pediatric acute hematogenous osteomyelitis. However, despite receiving adequate empiric antibiotic therapy, our patient did not demonstrate significant clinical improvement. While her MRI showed local progression and multifocal spread of her lesions, she remained clinically well-appearing with only intermittent pain and fever, which is not consistent with a disseminated S. aureus infection. Indolent presentation of multifocal osteomyelitis has been described in cases of disseminated cat scratch disease. The most commonly reported sites of B. henselae osteomyelitis are the vertebra and pelvic girdle, though infections of the femur, humerus, and carpal bones have also been reported.1 We identified only 5 other reported cases involving infection of the rib.2 Regardless of antimicrobial agent used, B. henselae osteomyelitis has an excellent prognosis in the immunocompetent host, and symptoms typically resolve without chronic complications.

**Conclusion:** For patients with clinical, laboratory and imaging findings consistent with osteomyelitis, empiric antimicrobial therapy with S. aureus coverage should be initiated. However, if a patient is not improving on appropriate therapy, other organisms should be considered. B. henselae should be considered in particular if the course is indolent, if the lesions are multifocal, or if the location includes the pelvic girdle or vertebral bodies. While our patient had known exposure to cats, there have been cases of B. henselae transmission via dogs, presumably transmitted through flea bites. Other manifestations of disseminated cat-scratch disease in immunocompetent hosts can include prolonged fever of unknown origin, with involvement of liver, spleen, eyes or central nervous system.

**Resources:**
Fig #1: Coronal STIR image showing enhancement in the left superior pubic ramus; taken on initial presentation to the hospital.

Fig #2: T2 Coronal image 2 weeks after initial MRI. Increased signaling in the left pubic ramus (white arrow) with 7 mm adjacent soft tissue swelling (grey arrow).
Title: Fever of Unknown Origin - Follow Your Gut!
Authors: Krista Allen, MD, Riley Hospital for Children at Indiana University Health
Angela Dietrich-Kusch, MD, Riley Hospital for Children at Indiana University Health, Amanda Benaderet, MD, MPH, Riley Hospital for Children at Indiana University Health
Case Presentation: A previously healthy 2 year-old female presented with 4 weeks of fever, weight loss, and fatigue. She was initially diagnosed with pneumonia and treated with antibiotics, but fevers continued. She was also found to have microcytic anemia, and was started on iron therapy. On admit at our facility, PE was only notable for weight at the 4th percentile. Lab work revealed anemia and mild elevation of ESR and CRP. Antibiotics were stopped, but fevers persisted. While hospitalized, the patient developed colicky abdominal pain with dark stool, which was attributed to iron therapy by the GI team (fecal immunochemical test was negative). CT of the abdomen was normal. Whole body MRI and MRA showed findings consistent with previous pneumonia. Bone marrow biopsy was unremarkable. Stool studies eventually returned with profound elevation of fecal calprotectin. Upper/lower endoscopy were grossly normal, but biopsy showed focal non-caseating granulomas consistent with early onset Crohn’s disease. The patient’s fevers and other symptoms improved following therapy with prednisolone and azathioprine.
Discussion: Infectious, oncologic, and collagen-vascular diseases together make up more than 90% of fever of unknown origin (FUO) cases with miscellaneous non-infectious and unknown causes making up the rest. Inflammatory bowel
disease (IBD) is an uncommon final diagnosis for FUO, especially in the young pediatric population. The peak incidence of IBD is between 15 and 30 years of age with only 9.9% of patients diagnosed before age 5. Younger age at presentation is more likely to be associated with vague symptoms and unimpressive visual findings on endoscopy, often requiring pathology results to make the diagnosis. This aligns with our patient’s clinical course, including endoscopy without apparent inflammation, but biopsy with non-caseating granulomas. Our patient’s diagnosis was delayed due to lack of diarrhea or typical extra-intestinal symptoms as well as recent pneumonia, which provided anchoring bias. She had an extensive infectious, oncologic, and rheumatologic evaluation before a fecal calprotectin helped reveal the diagnosis.

**Conclusion:** FUO patients are often admitted to the hospitalist service for diagnostic work-up. Determining when to consider IBD as a cause of FUO is challenging. There are only rare case reports of IBD as a cause of FUO in the pediatric literature. When IBD is the cause of FUO, it is often diagnosed months after symptom onset and diagnosis is commonly preceded by extensive infectious, rheumatologic and oncologic workup.

Fecal calprotectin is a marker of neutrophil activity and thus intestinal inflammation. In the adult population, fecal calprotectin is highly sensitive and specific (93% and 96% respectively) in screening for IBD, but less so in the pediatric population, with a sensitivity of 92% and specificity of 76%. While not perfect, it is a non-invasive test for evaluation of IBD that can help determine the need for endoscopy.

While endoscopy may not be routinely warranted in FUO workup, in a patient with abdominal symptoms and weight loss, it is important to consider IBD in the differential. Testing with fecal calprotectin may help guide further evaluation.

**Resources:**


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B102

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**Title:** Ghastly Polyps and Horrible Emboli, an Unusual Case of Venous Thrombosis

**Authors:** Allison Ta, MD, Inova Fairfax Hospital
Fatuma Barqadle, MD, INOVA Fairfax Hospital

**Case Presentation:** A 13 year old female with history of iron deficiency anemia treated with iron presented with 1 week of arm swelling and shortness of breath. She denied inciting injury. Past medical history was insignificant. Family history significant for familial adenomatous polyposis (FAP) however, patient not yet evaluated. No family history of hematologic disease. A left upper extremity ultrasound demonstrated extensive deep vein thrombosis (DVT) and chest CT showed bilateral pulmonary embolism. She was admitted to the intensive care unit for anticoagulation, but found to have microcytic anemia, hemoglobin 5g/dL. A colonoscopy revealed numerous ulcerated polyps (figure) consistent with FAP. She underwent an extensive laboratory evaluation for her thrombophilia as well as imaging for any anatomic abnormality, all of which were negative. At follow up, she completed 9 months of anticoagulation with improvement. She underwent total colectomy, histology negative for malignancy.
**Discussion:** DVTs are rare in the pediatric population with an annual incidence of 0.07-0.14 per 10,000 children\(^5\). Upper extremity DVTs (UEDVTs) are usually provoked and, in pediatrics, occur most often in the setting of a central venous catheter. Other risk factors for DVTs include malignancy, anatomic obstruction and hematologic conditions. Our patient did not have any typical risk factors for DVTs but was in a proinflammatory state because of her underlying FAP. Only two prior case reports have described patients with FAP that developed DVTs\(^1,4\) but data has shown that systemic inflammation modulates thrombotic responses by suppressing fibrinolysis, upregulation of procoagulant, and downregulating anticoagulants which can lead to thrombosis formation\(^6\). Additionally, our patient presented with severe iron deficiency anemia which has been linked to thrombosis formation although the mechanism isn’t clear\(^2\). One study postulated that severe iron deficiency anemia can affect the function of Factor VIII which also modulates thrombosis\(^3\).

**Conclusion:** Deep vein thrombosis although rare can have life threatening and devastating consequences. Pediatric patients should have a careful history to determine the patient’s risk factors and guide necessary laboratory evaluation for thrombophilia. In patients with underlying proinflammatory conditions there should be a high suspicion for thrombosis. We discussed a possible link between iron deficiency anemia and venous thromboembolism in the setting of systemic inflammatory illnesses such as FAP that requires further investigation.

**Resources:**

**References**

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B103

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**Title:** Here an Aneurysm! There an Aneurysm! Everywhere an Aneurysm!

**Authors:** Marta King, MD, MED, Saint Louis University School of Medicine
Samantha Rohe, MD, SLU-Cardinal Glennon Children’s Hospital, Eric Rohe, MS II, Saint Louis University School of Medicine, Kirubahara Vaheesan, MD, Saint Louis University Hospital

**Case Presentation:** 5 mo male with recent refractory Kawasaki disease (KD) only responsive after IVIG x2, infliximab and pulse steroids followed by 2 week taper presented with 2 days of intermittent left upper extremity pallor most pronounced when supine. He remained on high dose ASA. Recent echocardiogram revealed unchanged bilateral coronary artery aneurysms (CAA). No family history of autoimmune or connective tissue disorders, or aneurysms. Pt well appearing, afebrile with normal vital signs and positional decreased left arm perfusion without edema, erythema or cyanosis. Left brachial pulse slightly diminished compared to right. Diagnostic studies significant for overall down trending inflammatory markers (nl wbc, nl plt, CRP: 0.5, ESR: 23) and stable anemia (Hgb 10.9). Upper extremity CT Angiogram (CTA) (figure 1) revealed two left axillary artery aneurysms and a thrombosed aneurysm at the axillary/brachial arterial junction with distal arteries supplied via collaterals. Admitted to the general pediatric service with cardiology, rheumatology, hematology, and interventional radiology consulting.

**Discussion:** Systemic artery aneurysms (SAA) occur in up to 2% of patients with KD and predispose the patient to vascular compromise of the extremities from aneurysmal thrombosis, stenosis or rupture. Furthermore, if affected vessels are cannulated during cardiac catheterization to follow CAA, there is significant risk of injury to those vessels.
Studies on SAA characteristics in KD are limited, but risk factors include young age at initial diagnosis of KD and the presence of CAA. One study found that all patients less than 8 months of age at time of diagnosis had multiple SAA. In addition, all patients with SAA had coronary artery involvement, many of which were associated with severe cardiac sequelae including MI and death. Vessels most commonly affected are the subclavian, axillary, brachial, iliac, and femoral arteries.

**Conclusion:** Our patient underwent unsuccessful catheter directed tPA thrombolysis; however, distal perfusion was maintained by collateral vessels. He was transitioned from heparin to Lovenox in addition to his high dose ASA. Follow up revealed additional SAA in bilateral subclavian, axillary and brachial arteries. At 1 year follow up, patient remains on Lovenox and has transitioned to low dose ASA. SAA are a rare complication of KD. However, their existence places patients at risk for significant morbidity and mortality which may be prevented by early diagnosis and appropriate anticoagulation, as in this case. Full body magnetic resonance angiography (MRA) can detect these abnormalities and may be useful as a screening tool in the high risk patient. In treatment of KD, the pediatric hospitalist should consider MRA screening of patients who are less than 8 months of age at time of diagnosis and any patient with coronary artery involvement, especially if they are to undergo cardiac catheterization.

**Resources:**

References:

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B104

**Title:** Bites, bats, bugs, and persistent high fevers

**Authors:** Marta King, MD, MED, Saint Louis University School of Medicine
Emily Fretz, Third Year Medical Student, B.A., Saint Louis University School of Medicine, Aaron Miller, MD, MSPH, Saint Louis University School of Medicine

**Case Presentation:** 16yo previously healthy male presented to the ED in summer with 6d of high fevers, myalgias, arthralgias, anorexia, fatigue and 1d of headache and emesis. He lives in a wooded area in MO and recalls many insect bites including a promptly removed tick. 1 mo before admission, he hiked across the Midwest and was exposed to bats while caving. The family has a cat and dog. Pt denies animal scratches or bites, sexual activity, IV drug use. His mother,
who did not travel with the patient, had recent myalgias without fever. Family history remarkable for mother with ill-defined immunodeficiency managed with subQ IG.

Exam showed febrile, ill appearing teen with bilateral conjunctivitis, excoriated papules over calves, but no rashes/lymphadenopathy, no meningeal signs, nl neurologic exam. Labs significant for leukopenia (3K, nl differential), thrombocytopenia (90K), hyponatremia (129), and transaminitis (ALT 649, AST 348). Although patient was at risk for numerous conditions based on exposures (Table 1), empiric doxycycline was initiated due to high suspicion for rickettsial illness (Table 2).

**Discussion:** The patient was discharged on hospital day 2 with significant clinical improvement and resolution of symptoms on follow up. Serum PCR returned positive for Ehrlichia chaffeensis. Though incidence varies by location, tickborne illnesses are found across the US. Prolonged (>24 hrs) tick attachment is needed for Lyme disease and borreliosis transmission; time for transmission of other tickborne illnesses is unknown. Most patients do not recall a tick bite. Common presenting symptoms include fever, headache, malaise, myalgias, nausea, vomiting, and diarrhea. Geographic location, unique physical exam and lab findings (Table 2) help in differentiating tickborne illnesses. Rickettsial illnesses include ehrlichiosis, anaplasmosis, and Rocky Mountain Spotted Fever. Features suggesting ehrlichiosis in our pt include home in woods of MO (disease spread by Lone Star tick found in southeastern/southcentral US), conjunctivitis, leukopenia, anemia, hyponatremia, transaminitis. Rash is absent in 40% of children and 70% of adults; ehrlichiosis is fatal in 3% of cases.

**Conclusion:** A thorough exposure history and awareness of typical presenting symptoms and geographic prevalence of various tickborne illnesses is crucial to prompt diagnosis and treatment. Given the significant morbidity and mortality associated with these illnesses, treatment should be based on clinical suspicion and initiated promptly. Hospitalists need to be aware that doxycycline is the recommended therapy for rickettsial disease regardless of patient’s age and that short courses do not cause teeth staining, which is most physicians’ reason for not using doxycycline. A recent study showed that while 80% of US healthcare providers were aware doxycycline was indicated for treatment of Rocky Mountain Spotted Fever, 65% would NOT prescribe it for a suspected case in a young child. Patients and families should also be counseled about preventative measures to avoid future tickborne illnesses including staying on trails, using DEET and permethrin to repel ticks, and bathing and conducting full-body tick check after outdoor exposures to find and remove ticks.

**Resources:**

**Table 1: Exposures/Risk Factors**

<table>
<thead>
<tr>
<th>Exposure</th>
<th>Disease Risk</th>
<th>Likelihood in our patient</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Ticks</strong></td>
<td><strong>Ehrlichiosis</strong></td>
<td>Correct geography — more likely</td>
</tr>
<tr>
<td></td>
<td><strong>Anaplasmosis</strong></td>
<td>Correct geography — more likely</td>
</tr>
<tr>
<td></td>
<td><strong>Rocky Mountain Spotted Fever</strong> (Rockies)</td>
<td>No rash — less likely</td>
</tr>
<tr>
<td></td>
<td><strong>Tularemia</strong></td>
<td>No cough, pleuritic chest pain, lymphadenopathy, skin findings, or lymphopenia—less likely</td>
</tr>
<tr>
<td><strong>Mosquitoes</strong></td>
<td><strong>West Nile Virus Encephalitis</strong></td>
<td>No meningism or neurologic symptoms — less likely</td>
</tr>
<tr>
<td></td>
<td><strong>St. Louis Encephalitis</strong></td>
<td>No meningism or neurologic symptoms — less likely</td>
</tr>
<tr>
<td><strong>Spiders</strong></td>
<td><strong>Brown recluse with systemic infection</strong></td>
<td>No preceding localized disease — less likely</td>
</tr>
<tr>
<td><strong>Tick bite with immediate removal</strong></td>
<td><strong>Tickborne illness</strong></td>
<td>Prolonged (&gt;24 hours) tick attachment typically needed for disease transmission — less likely from this particular bite. But he most likely has had a bite that he did not notice.</td>
</tr>
<tr>
<td><strong>Travel Midwest US</strong></td>
<td><strong>Lyme disease</strong></td>
<td>No history of erythema migrans — less likely</td>
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<tr>
<td></td>
<td><strong>Borreliosis</strong></td>
<td>No hepatitis A/B, or bactemia — less likely</td>
</tr>
<tr>
<td><strong>Caving</strong></td>
<td><strong>Histoplasmosis</strong></td>
<td>A consideration, especially of underlying immunodeficiency though less likely with no respiratory symptoms, palatal ulcers, or sputumology</td>
</tr>
<tr>
<td><strong>Fresh-water exposure</strong></td>
<td><strong>Leptospirosis</strong></td>
<td>No history of significant freshwater exposure — less likely</td>
</tr>
<tr>
<td></td>
<td><strong>Giardiasis</strong></td>
<td>No history of significant freshwater exposure, no diarrhea — less likely</td>
</tr>
<tr>
<td><strong>Mother with recent rashes</strong></td>
<td><strong>Viral syndrome</strong></td>
<td>Possible</td>
</tr>
<tr>
<td></td>
<td><strong>Influenza</strong></td>
<td>No respiratory symptoms — less likely</td>
</tr>
<tr>
<td><strong>Mother with immunodeficiency</strong></td>
<td><strong>Cytomegalovirus or Epstein-Barr Virus</strong></td>
<td>No lymphadenopathy or hepatitis Bmononucleosis — less likely</td>
</tr>
<tr>
<td><strong>Infant presentation of immune deficiency</strong></td>
<td>Any infection could present as more severe than would be assessed for a young male.</td>
<td></td>
</tr>
<tr>
<td><strong>Pastoralia multocida</strong></td>
<td>No history of bites/scratches and no lymphadenopathy — less likely</td>
<td></td>
</tr>
<tr>
<td><strong>Bartonella henselae</strong></td>
<td>No history of bites/scratches and no cellulitis or lymphadenopathy — less likely</td>
<td></td>
</tr>
<tr>
<td><strong>Glanders</strong></td>
<td>No diarrhea — less likely</td>
<td></td>
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<tr>
<td><strong>Known to transfer ticks</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Pet Cat</strong></td>
<td><strong>Pastoralia multocida</strong></td>
<td>No history of bites/scratches and no cellulitis or lymphadenopathy — less likely</td>
</tr>
<tr>
<td><strong>Pet Dog</strong></td>
<td><strong>Pastoralia multocida</strong></td>
<td>No history of bites/scratches and no cellulitis or lymphadenopathy — less likely</td>
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<tr>
<td></td>
<td><strong>Campylobacter</strong></td>
<td>No moody diarrhea — less likely</td>
</tr>
<tr>
<td></td>
<td><strong>Known to transfer ticks</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Teenager</strong></td>
<td><strong>HIV infection</strong></td>
<td>No history of sexual activity or intravenous drug use — less likely</td>
</tr>
</tbody>
</table>

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B105
**Title:** To EBV or not to EBV? What is the Diagnosis?

**Authors:** Clifton Lee, MD, FAAP, SFHM, Children’s Hospital of Richmond at VCU
Megan Coe, MD, Children’s Hospital of Richmond at VCU

**Case Presentation:** A 12 month old healthy boy presented with several days of fever. He was diagnosed with a viral illness by his PCP and discharged home. Three weeks later, he was seen with continued fever, posterior cervical lymphadenopathy, and abdominal distention. Initial laboratory studies revealed pancytopenia, and he was admitted. EBV serology was confirmatory for acute primary infection. He was negative for CMV, HIV, and parvovirus B19 titers. A bone marrow biopsy revealed normocellular bone marrow with no evidence of malignancy. He was transfused and discharged with a diagnosis of infectious mononucleosis and anemia secondary to splenic sequestration. At follow-up appointment nine days later, he continued to have daily fevers, worsened lymphadenopathy, and continued splenomegaly and pancytopenia and readmitted and started on IV ganciclovir. Due to lack of clinical improvement, a lymph node biopsy was performed that confirmed the diagnosis of Langerhans Cell Histiocytosis. He was started on chemotherapy with resolution of fevers and significant improvement of the lymphadenopathy.

**Discussion:** Langerhans Cell Histiocytosis (LCH) is a rare heterogeneous disease characterized by proliferation of immature dendritic cells normally involved in inflammatory responses. This disease can be single or multi-system in nature and can be self-limiting or rapidly progressive. The most commonly affected system is skeletal, accounting for up to 80% of cases. Rarely, at 5-10% of cases, LCH affects the lymph nodes in isolation as was the case in our patient. Due to its rare and varied nature, few studies exist on the etiologic and pathogenic manifestations of this disease. The role of EBV in the pathogenesis of LCH remains controversial with wide disagreement among the few studies in existence. One study determined the presence of EBV as a possible pathogenic contributor in 100% of cases, another in 0% of cases. More studies fall somewhere in between, with the majority of findings hinting at a non-fully defined role of EBV in causing LCH. Whether EBV was actively present within the lymph node biopsied in our patient remains to be seen.

**Conclusion:** Significant lymphadenopathy and splenomegaly in a young infant with positive EBV serology should raise suspicion for alternative diagnoses. Infection with this virus is typically mild or subclinical in infants. A symptomatically significant or persistent presentation is concerning for lympho-proliferation as was seen with our patient. Additionally, a severe EBV infection in infancy should raise concern for immunodeficiency. Although less likely, X-linked immune-proliferative syndrome, in which a dysregulated immune response to EBV manifests in severe and sometimes life-threatening disease, should be tested.

**Resources:**

References


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B106
Title: Persistent Pneumonia: A diagnosis and treatment by bronchoscopy
Authors: Bethany Bartley, MD, Massachusetts General Hospital for Children
Lindsay Carter, MD, Massachusetts General Hospital

Case Presentation: An 11 year old previously healthy boy presented from an outside hospital as transfer with a persistent right upper lobe infiltrate and fever. Fever and cough started two weeks prior, at which time he was treated as an outpatient with Augmentin, followed by the addition of Cefprozil. Ten days into persistent symptoms, he was admitted to an outside hospital and treated with IV Ceftriaxone and oral Azithromycin. Serial chest x-rays showed partial, then complete opacification of the RUL with volume loss demonstrated by elevation of the minor fissure (Image 1). The patient was not hypoxemic, but was notably dyspneic with continued fever and cough. A thorough infectious work-up was negative to date. The patient's history was negative for recent travel, asthma, or recurrent sinopulmonary infections. Bronchoscopy revealed the takeoff of the RUL bronchus just proximal to the carina (Image 2), consistent with a tracheal bronchus (TB); a substantial mucous plug was successfully lavaged from the orifice. The patient subsequently demonstrated swift improvement. Mycoplasma serology was positive.

Discussion: The differential diagnosis of persistent pneumonia in a previously healthy pediatric patient can be broad, including congenital lesions, airway obstruction or anomaly, foreign body aspiration, or resistant organism. Previously asymptomatic congenital lesions to consider include CPAM, or bronchogenic cyst. Alternate causes of extrinsic airway compression include vascular ring/sling, or mass. The prevalence of antibiotic resistance in communities should be reviewed, and foreign body aspiration should be considered, in all cases of persistent symptoms. TB is a rare congenital airway anomaly to keep in mind, especially in cases with RUL symptoms. TB is defined as any airway that arises from the trachea proximal to the carina. It most often occurs with the RUL bronchus and has a reported incidence of 0.1–2% of the general population. This finding is considered anatomically normal in some animals, including pigs, leading some variations to be known as a pig bronchus. TB can predispose patients to poor airway clearance of the RUL, especially in the setting of a lower respiratory infection.

Conclusion: It can be debated even among pediatric pulmonologists if the most appropriate next step in the management of a pediatric patient with persistent pneumonia would be a chest CT scan or bronchoscopy. In this case, bronchoscopy was pursued given its potential ability to not only diagnose, but also treat the underlying etiology. Bronchoscopy additionally offered the advantage of collecting bronchoalveolar lavage samples. A rare anomaly was ultimately discovered, but notably when TB is identified on bronchoscopy it is most often in the setting of recurrent pneumonia, and almost always occurs in the RUL. TB can also be well visualized for diagnosis on chest CT, however some cases may require bronchoscopy to clear obstructive mucous plugs and open distal lung collapse.

Resources:

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B107

Title: A Tough Diagnosis to See, an Infant with Direct Hyperbilirubinemia
Authors: Brian Williams, MD, UCSD/Rady Children's Hospital, San Diego
Jennifer Bracamontes, MD, University of Washington, Seattle Children's Hospital

Case Presentation: A two-month-old term male presented with 3 days of emesis and ‘lifelong’ jaundice. His initial work-up was notable for a total bilirubin of 9.1 and a direct bilirubin of 6.2. Basic lab studies were notable for AST 600, ALT 355. Physical exam showed a well-appearing infant with scleral icterus and full body jaundice. A HIDA scan showed delayed passage of tracer but no biliary atresia. A broad work-up for metabolic, infectious and toxin etiologies was unremarkable. The patient underwent a liver biopsy noting giant cell hepatitis. He was later found to have nystagmus on exam. A fundoscopic exam showed bilateral optic nerve hypoplasia. Subsequent lab studies showed a sodium level of 157. Urine osmolality was inappropriately low. Serum sodium and urine osmolality both responded to DDAVP. Additional pituitary studies were performed and the patient was found to also have growth hormone deficiency. A brain MRI showed bilateral optic nerve hypoplasia but no midline defects. Given the optic nerve hypoplasia and pituitary dysfunction the patient was diagnosed with septo-optic dysplasia.
Discussion: Septo-optic dysplasia is a rare congenital syndrome characterized by optic nerve hypoplasia, midline brain defects, and pituitary dysfunction. The diagnosis is confirmed when 2 of the 3 features are present. It is estimated that septo-optic dysplasia affects 1 in 10,000 newborns. Presentation is quite variable and may include hypoglycemia, failure to thrive, syndactyly, cleft palate, diabetes insipidus, nystagmus, micropenis, and hyperbilirubinemia. Our case presented with direct hyperbilirubinemia caused by giant cell hepatitis with the subsequent discovery of diabetes insipidus. Although the exact mechanism of giant cell hepatitis is unknown, it can be associated with growth hormone deficiency and is treated with hormone replacement.

Conclusion: Cholestasis caused by giant cell hepatitis is an uncommon presentation of septo-optic dysplasia and can mimic various liver diseases. In order to facilitate early diagnosis and minimize morbidity, a high index of suspicion for septo-optic dysplasia should be maintained in the presence of hyperbilirubinemia.

Resources: N/A

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B108

Title: Necrotizing pneumonia in a machinist's daughter

Authors: Bethany Woomer, MD, FAAP, UCSF

Case Presentation: A 14 yo previously healthy Hispanic female presented with pleurisy without cough or fever. After an abnormal CXR, CT revealed a LLL cavitary pneumonia treated with 14-days of levofloxacin. Her symptoms resolved but NSAIDs were given 2 months later for recurrent pleurisy. She returned 2 months later with two weeks of pleurisy accompanied by fever, chills, and productive cough. No exposures, Dad is a machinist. CXR showed pneumonia. When her respiratory status declined on ceftraixone, vancomycin was added. A chest tube was placed for parapneumonic effusion in the setting of LLL necrotizing pneumonia. Infectious workup was negative for bacteria, fungi and TB and she was discharged on Linezolid. Her sputum grew AFB and clarithromycin was added. Three weeks into therapy, the organism was identified as Mycobacterium immunogenenum. She continued to have fevers, night sweats, and fatigue requiring transition to Azithromycin and PICC placement for IV Imipenem and Amikacin. After 4 months of therapy, she is symptom free, but remains on IV antibiotics with continued AFB-positive sputum cultures.

Discussion: Nontuberculous mycobacteria (NTM) are ubiquitous in the environment and while exposure is widespread, disseminated infections and pulmonary disease are uncommon. Since NTM are not reportable, estimates of incidence and prevalence are poor. While transmission is thought to be environmental, there is some evidence that human-to-human transmission may occur. M. immunogenenum is a rapidly growing NTM. It was identified in 2001 and has been isolated from contaminated metalworking fluids and anti-septic solutions. It has been linked with occupational hypersensitivity pneumonitis and with disseminated infections in immunocompromised patients. Largely isolated from adults, it is also found in certain at-risk pediatric groups, such as solid-organ transplants and CF. A study of pediatric NTM infections found that only 13% of children were previously healthy. In the same study all of the patients with M. immunogenenum were immunocompromised. While there have been a few case reports of healthy patients with M. immunogenenum, none have presented as our patient presented with isolated pulmonary disease.

Conclusion: For our patient, it remains to be seen whether there is a link between the father's job as a machinist and his daughter's M. immunogenenum infection, known to be found in contaminated metalworking fluids. In general, there is much we do not know about NTM infections, particularly ideal antibiotic treatment choices and course, which patients are at highest risk to develop infections as opposed to asymptotic colonization, and whether genetic susceptibilities to these organisms exist. We need more research and consistent reporting to make progress on these questions. As Pediatric hospitalist providers, we are likely to see both immunocompromised and competent patients with these infections and need to have a high suspicion for these organisms in our refractory patients.

Resources:

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B109
Title: Opathy vs Itis: An extreme case of gastro
Authors: Jennifer Danzig, MD, Children’s Hospital of Philadelphia
Sarah Sheppard, MD PhD, Children’s Hospital of Philadelphia

Case Presentation: A 5 year old previously healthy girl presented with 3 months of recurrent episodes of diffuse abdominal pain and nonbloody, nonbilious emesis. Her laboratory studies showed hyponatremia to 128 mmol/L (normal range 138-145), hypochloremia to 96 mmol/L (normal range 98-106) and metabolic alkalosis with bicarbonate of 31 mmol/L (normal range 20-26). These abnormalities persisted despite intravenous hydration. She also had marked hypoalbuminemia to 1.6 g/dL (normal range 3.5-5.2) and mild transaminitis. Abdominal ultrasound showed thickened small bowel in the right lower quadrant consistent with ileitis and hepatosplenomegaly with normal liver and spleen echogenicity. Due to persistent emesis upper GI study was performed to further evaluate anatomy and showed asymmetric thickening of the anterolateral antral and fundal gastric folds. This hyperplastic gastric mural thickening raised concern for Ménétrier's disease and subsequent serum CMV testing was positive.

Discussion: Ménétrier's disease is a rare cause of hypertrophic gastropathy in children. Hyperplasia of the gastric folds leads to protein losing enteropathy. In children, it is associated with indolent infectious etiologies, specifically CMV. The pathogenesis is unclear, and the direct effect of CMV infection does not appear to be responsible for the symptoms and treatment with ganciclovir therapy has not been shown to change the clinical course. It is suspected that mucosal damage caused by the infection results in production of an epithelial growth factor leading to local hyperplasia and proliferation of gastric mucosal cells which inhibit gastric acid secretion and increase production of mucus. While the adult form is associated with persistent symptoms and increased risk of carcinoma often requiring gastrectomy, pediatric hypertrophic gastropathy is more indolent and tends to resolve over time. Our patient improved clinically with supportive care in the hospital and demonstrated good weight gain.

Conclusion: Nonspecific symptoms such as vomiting and abdominal pain are common in pediatrics and often due to benign causes. A high degree of suspicion is required to identify children with these symptoms who require further diagnostic workup. Here, history of recurrent symptoms and absence of other typical associated features such as fever or diarrhea raised concern for less common etiologies. In our cost conscious era, we scrutinize the need for further laboratory and imaging studies. Careful attention to the history is necessary to determine the need for further evaluation. The severity of her laboratory abnormalities and imaging were concerning for more severe chronic conditions. The finding of serum CMV positivity led to the diagnosis of Ménétrier's disease, predicting that symptoms would likely resolve, and obviated the need for more invasive testing such as endoscopy. We present a case of Menetrier's disease due to CMV, a severe, self-limited complication of a common pediatric infection.

Resources: N/A
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B110

Title: PAIN IN THE NEC
Authors: Allison Ashford, MD, University of Nebraska Medical Center
Trudie Owens, APRN-NP, Children's Hospital and Medical Center/UNMC, Nate Goodrich, MD, Children's Hospital and Medical Center/UNMC

Case Presentation: A 16-year old German female presented with 1-day history of abdominal pain, nausea and vomiting of brown emesis, worse with positional changes. She was afebrile without changes in her bowel habits. She was admitted and given intravenous fluids, ondansetron, promethazine and diazepam without relief. Abdominal CT was unremarkable. CBC was normal, but emesis was hemoccult positive. One day prior to admission, she returned from a trip to Washington, DC where she developed the brown emesis while on the plane. Her past history was significant for two previous episodes of similar symptoms requiring hospitalization in Germany, but with no clear medical etiology per reports. On physical exam she was ill appearing but afebrile. She had periumbilical tenderness to palpation, no guarding or rigidity and a negative McBurney's. She had no lymphadenopathy, no thyromegaly, or rashes.

Discussion: GI was consulted for the hematemesis. EGD showed distal esophagitis with exudates thought to be secondary to fungal infection, antral inflammation and erythema. No ulcers were identified and the duodenum was normal. Biopsies from the duodenum and proximal esophagus were normal; stomach biopsy had reactive gastropathy, H-pylori negative. Distal esophagus biopsies showed necrotizing esophagitis but were negative for fungus.
CT of neck/chest/abdomen had normal vasculature. Ultrasound of thyroid was without solid nodules. Chest x-ray was negative. ANCAs, CRP, ANA, thyroid function, immunoglobulins, CMV, HHV-6, EBV and HIV were all negative in the evaluation of her necrotizing esophagitis. HSV in the blood was positive. Infectious Disease and Immunology were both consulted. Patient was initiated on empiric piperacillin-tazobactam and vancomycin. Symptoms of feeding intolerance, emesis and abdominal pain continued. She was made NPO and had several days of TPN. She received acyclovir with resolution of abdominal pain and emesis. She transitioned to a soft diet and discharged.

**Conclusion:** This case is instructive because without biopsy, this diagnosis would likely have been missed.

There are 88 reported cases of necrotizing esophagitis in one study with variable risk factors and pathogenesis, and a predominance of older men. The hallmark finding on endoscopy is diffuse circumferential black mucosa in the distal esophagus that stops at the gastroesophageal junction. Up to 70% of patients present with hematemesis and melena. Biopsy is recommended, although not required for diagnosis.

Lab findings often due underlying disease include lactic acidosis, hypoalbuminemia, anemia, renal insufficiency and hyperglycemia, but are usually unremarkable. Ischemia and gastric outlet obstruction may be inciting events. Common etiologies are Klebsiella pneumonia, CMV, HSV, candida, fungus, gastric volvulus, and prolonged vomiting.

Mortality is 13-35%, often due to underlying disease. Complications include esophageal perforation or stricture, stenosis, infections and death. This case should alert us of the possibility of necrotizing esophagitis as an etiology of UGI ble

**Resources:**


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B111

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**Title:** Unusual Presentation of Kawasaki Disease Resembling Retropharyngeal Abscess

**Authors:** Lauren Beebe, Resident Physician/MD, Rainbow Babies & Children's Hospital
Erin Frank, Attending Physician/MD, Rainbow Babies & Children's Hospital, Hemangini Bhakta, MD, Rainbow Babies and Children's Hospital, Cleveland Medical Center, Cleveland, Ohio, Kendall Wyllie, Resident Physician/MD, Rainbow Babies & Children's Hospital, Ankita Desai, Attending Physician/MD, Rainbow Babies & Children's Hospital

**Case Presentation:** A 2-year-old previously healthy girl presented to the emergency department (ED) with 2 days of fever, right-sided neck swelling, irritability, and decreased oral intake. On exam, she had right-sided neck swelling and tenderness, scattered papules over neck and back, and bilateral lymphadenopathy. A CT of the neck showed a 5mm right-sided retropharyngeal fluid collection and bilateral lymphadenopathy. Otolaryngology (ENT) was consulted due to concern for a developing retropharyngeal abscess. She was admitted for intravenous (IV) antibiotics and possible surgical drainage. Over the next 24 hours, she developed bilateral non-exudative conjunctivitis, hand and feet edema, fissured lips, and full body polymorphous rash. She continued to be febrile and had elevated C-reactive protein and erythrocyte sedimentation rate; symptoms persisted despite broad-spectrum IV antibiotics. Infectious Diseases (ID) was consulted and Kawasaki disease (KD) was suspected. On day 5 of fever, she met criteria for KD and was treated with intravenous immunoglobulin (IVIG) and high-dose aspirin.

**Discussion:** KD is a systemic vasculitis diagnosed by clinical criteria of fever for >5 days, plus 4 of the following: bulbar conjunctivitis, mucositis, polymorphous rash, extremity changes, and cervical lymphadenopathy (1). Unusual head and neck findings can be barriers to diagnosis, specifically mastoiditis, cervical adenitis, pharyngitis, tonsillitis, and retropharyngeal abscess (RPA) (2). KD presenting as RPA is rare; a literature search via PubMed yields < 15 case reports (3-6). These cases initially presented with fever and lymphadenopathy, retropharyngeal low-density area on CT, and persistence of fevers, lymphadenopathy, irritability and malaise despite antibiotics and surgical drainage. While eventual diagnosis of KD was made and treatment initiated, at least one case reported coronary artery aneurysm at time of
diagnosis (7). Unusual presentation of KD may delay diagnosis and treatment, thus increasing risk for cardiac complications. In our patient, treatment with IVIG and high dose aspirin was initiated on day 5 of fever, with subsequent clinical improvement.

**Conclusion:** Given lack of specific diagnostic testing and fluctuating symptoms, diagnosis of KD relies on high index of clinical suspicion. This case demonstrates the importance of recognizing unusual presentations of KD, and being adaptable regarding a changing diagnosis based on a clinical course. Close observation following initiation of antibiotic therapy, frequent communication with consultants in ID and ENT, and review of similar cases in literature was crucial to our timely diagnosis of KD. This allowed us to avoid unnecessary risk and cost of surgical intervention and exposure to radiation with serial imaging, and decrease risk of cardiac complications. Clinicians should include KD in the differential diagnosis for fever, lymphadenopathy, and retropharyngeal edema. In pediatrics, there are unusual presentations of every condition, and we must be flexible and recognize when a patient’s clinical course leads us down an unexpected path.

**Resources:**


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B112

**Title:** When a rash is more than just a rash

**Authors:** Gokila Pillai, MD, Indiana University
David Rayburn, MD, Indiana University, Andrew Shriner, MD, Indiana University, Department of Pediatrics

**Case Presentation:** A 6yoM presented to the ED for 1 day of bilateral leg pain, fever, altered mental status, and rash. Initial vital signs were T 102.9°F, HR 147, RR 30, BP 101/42, and O2 96%. Once afebrile, he was well appearing but with a
diffuse petechial rash (see figures 1 and 2). Labs were notable for WBC 3.9, platelets 155, PT 18.8, INR 1.65, and CRP 5.5 mg/dL. He received fluids and ceftriaxone prior to transfer for ongoing care.

Upon transfer, he was playful, behaving normally, and without leg pain. His working diagnosis at time of transfer was Hennoch-Schonlein Purpura (HSP).

The following morning, he again endorsed hip and leg pain and difficulty walking. Repeat labs showed a WBC of 43.6 and a worsening coagulopathy (PT 25.4, INR 2.37). Given concern for infection, ceftriaxone and fluids were re-started.

The blood culture came back positive for gram negative diplococci 36 hours after being sent and speciated to Neisseria meningitidis (Group B). He was diagnosed with meningococcemia with purpura fulminans and completed 7 days of IV ceftriaxone. He never developed signs of meningitis.

Discussion: Meningococcemia is a rare diagnosis and part of the spectrum of invasive meningococcal disease (IMD). IMD can manifest as meningitis, meningococcemia, or meningitis plus meningococcemia. Meningitis is the most common presentation of IMD, accounting for 30-60% of cases. Meningococcemia accounts for 20-30% of cases, and 12% of patients have meningitis plus meningococcemia. Isolated meningococcemia classically presents with nonspecific viral symptoms followed by leg pain, cold extremities, and skin pallor or mottling, followed by rapid decompensation.

While the case seems straightforward in hindsight as our patient did present classically, diagnosis was still difficult. Challenges included rarity of the disease, particularly isolated meningococcal septicemia, and our patient’s reassuring exam, which biased away from IMD. A focus on his persistent symptoms of rash and leg pain at time of transfer also increased suspicion for HSP, which likely contributed to diagnosis momentum the following day.

Conclusion: Though uncommon and not endemic in the US, IMD definitely still occurs and should always be on the differential. Timely diagnosis and treatment is critical as even with treatment, mortality still approaches 10-15%. A well appearing exam should not reassure against IMD. Our patient’s first specific symptom of IMD was bilateral leg pain, which we can now appreciate as an early sign of sepsis. Though clinically stable and mentating appropriately, our patient’s leg pain in the context of rash and history of fever with altered mental status serves the most important clue for our patient’s ultimate diagnosis.

Resources:

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B113
Title: Recurrent Back Pain reveals a Rare Inflammatory Condition

Authors: Cara Tillotson, D.O. FAAP, Carilion Children’s
Paul (PJ) Whalen, M.D., Carilion Childrens, Michael Burbridge, D.O.. FAAP, Carilion Childrens

Case Presentation: A 14 year old presented with bilateral lower back pain and intermittent fever. Back pain was acute and progressive, with initial resolution with NSAIDs. Fever was intermittent for approximately two weeks. MRI demonstrated areas of abnormal signal enhancement at L2-L3 and L4, consistent with an inflammation. Workup showed normal white blood cell count and differential. CRP and ESR were elevated slightly at 2.36 (Ref < 1.0) and 40 (0-20) respectively. Blood culture was negative. Past medical history was significant for bilateral pelvic osteomyelitis one-year prior, for which she received prolonged IV antibiotic therapy. Bone biopsy at that time showed inflammation but culture was negative. The history of recurrent osteomyelitis, elevated inflammatory markers, and otherwise negative infectious work up was consistent with a diagnosis of Chronic Recurrent Multifocal Osteomyelitis. The patient improved with NSAID treatment and discharged to follow up with Pediatric Rheumatology, where diagnosis of Chronic Recurrent Multifocal Osteomyelitis was confirmed.

Discussion: Chronic Recurrent Multifocal Osteomyelitis (CRMO) is an autoimmune inflammatory bone condition that presents similarly to osteomyelitis. The condition is rare, though exact incidence is unknown. It is more common in children and adolescents with female to male ratio approximately 2:1. Patients present with bone pain, swelling, and fever and may have pustular rash on the hands and feet. Inflammatory markers are typically elevated. Lesions may be unifocal at initial diagnosis, though multifocal lesions are common. Diagnosis is clinical and may be made after failure to respond to treatment for bacterial osteomyelitis or when bone biopsies are found to be sterile. NSAIDs and corticosteroids are often used in the initial treatment and help with pain, however recurrence is common. Long-term treatment options include sulfasalazine, methotrexate, gamma interferon, tumor necrosis factor alpha blocking therapy, and bisphosphonates. Prognosis varies, with some patients having spontaneous resolution while others progress to complications such as fractures, scoliosis, and vertebral collapse.

Conclusion: Chronic Recurrent Multifocal Osteomyelitis (CRMO) is a rare autoimmune inflammatory bone condition that is commonly mistaken for bacterial osteomyelitis, which may lead to a delay in diagnosis. The condition should be suspected in a child with recurrent osteomyelitis or those who fail to improve with treatment for infectious osteomyelitis. A bone biopsy is typically required to rule out the more common bacterial osteomyelitis. Treatment for CRMO includes NSAIDs, biologic agents, and corticosteroids. Flares of pain are common, though recurrent episodes may decrease in frequency with time. Rheumatology consultation and follow up is recommended. In general, patients with CRMO are able to participate in normal physical activities in between flares, though physical and occupational therapy are helpful to optimize quality of life.

Resources:

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B114
Title: 9 year old farm boy with fevers and a chest mass

Authors: Aleisha Nabower, M.D., University of Nebraska Medical Center
Lisa Siczkowski, M.D., Children’s Hospital and Medical Center, Andrea Green-Hines, M.D., Children’s Hospital and Medical Center, Terri Love, M.D., Children’s Hospital and Medical Center

Case Presentation: A 9yo previously healthy male was admitted with one week of fever (Tmax 104.7), chest and right shoulder pain. A chest x-ray performed 5 days prior demonstrated slight prominence of the perihiphler regions (Fig 1). Azithromycin was initiated for presumed community acquired pneumonia but did not result in improvement of patient’s symptoms. On the day of admission, a chest CT demonstrated a 3.3 x 3.8 cm mediastinal mass anterior to the trachea extending anteriorly to the right pulmonary artery and inferior to the carina. Areas of hypodensity were located centrally representing liquefaction. Mass effect was present on the SVC and trachea. Hilar adenopathy was also present with a 3mm nodule in the lingula and a 10 mm round nodule in the left lower lobe with tree and bud pattern (Fig 2-5). Empiric cefepime, vancomycin, gentamicin and itraconazole were initiated for broad coverage of bacterial and fungal etiologies. Sinus, neck, abdominal, and pelvic CT were significant for splenomegaly and multiple ill-defined subcentimeter splenic hypodensities (Fig 2). Labs are presented in table 1.

Discussion: The differential diagnosis included infectious etiologies: complicated community acquired pneumonia, tularemia, histoplasmosis, cryptococcal pneumonia and tuberculosis as well as oncologic etiologies: Hodgkin’s or non-Hodgkin’s Lymphoma and germ cell tumor were highest on the differential, with PNET also included, although more commonly presenting in the posterior mediastinum1. Due to location of the lesions, biopsy was deemed to be too high risk. An extensive infectious work-up was initially unrevealing. Urine and serum Histoplasma antigens (Ag) were negative but based on imaging and epidemiologic risk factors (Midwest location, outdoor recreational activities), treatment was targeted for histoplasmosis. Based on continued high fevers, histoplasmosis therapy was escalated to liposomal amphotericin B. Histoplasmosis serology was sent and the yeast-phase was markedly positive (1:512). He was discharged on oral itraconazole with significant improvement 1 week post-discharge. He completed 6 weeks of itraconazole at which time he was clinically back to baseline.

Conclusion: The presentation of this patient was complicated by the inability to perform a biopsy and the negative histoplasmosis Ag screening. However, a meta-analysis found that the overall sensitivity for urine and serum histoplasmosis Ag detection was 81.4% (56-100% with the lower rates in acute pulmonary disease) and a specificity of 98.3%. Culture is the gold standard for diagnosis but requires 2-4 weeks. Ag detection tends to be more sensitive in disseminated histoplasmosis, while serology is more sensitive in acute pulmonary histoplasmosis.3 This case highlights the importance of properly selecting histoplasmosis diagnostic tools based on the type of infection present. The diagnosis was ultimately made based on the markedly high titers combined with the clinical improvement on treatment for histoplasmosis.

Resources: References

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B115

Title: A Case of Childhood Cat Scratch Disease causing Osteomyelitis of Skull

Authors: Yaseen Rafee, MD, Hurley Medical Center, Michigan State University
Dustin Miller, MD, Hurley Medical Center, Michigan State University

Case Presentation: 3-year-old Caucasian female who was recently diagnosed and treated for CSD presented to the emergency room for 2-week history of worsening scalp lesion in left frontal region. Skull X-ray revealed focal irregular lucency involving left frontal bone and follow-up CT of the head showed focal soft tissue lesion and destructive process
involving the adjacent bone in the left frontal region. Initial immunologic, hematologic/oncologic, and infectious workup was unremarkable. MRI of head revealed septated scalp swelling with destructive process involving adjacent calvaria with intracranial extension. Aspirate from the lesion was viewed with Warthin Starry stain which showed multiple clusters of organisms. Serum Bartonella henselae antibody showed IgG and IgM titers, and Bartonella henselae PCR from the skull abscess aspirate was positive. The patient was treated for six weeks with Azithromycin and Rifampin.

**Discussion:** Cat Scratch Disease is a self-limiting condition caused by the gram-negative bacillus Bartonella henselae that is generally characterized by regional lymphadenopathy. Although rare, osteomyelitis is a known complication of CSD, with the vertebral column and pelvic girdle being the most common sites. Skull involvement has been reported previously in literature, however it remains exceedingly rare. A literature review (prior to 2007) found 47 reported cases of osteomyelitis due to Bartonella henselae, with 4 of those involving the skull. Since 2007, skull involvement was reported in just 2 of the 14 reported cases we found.

**Conclusion:** Skull involvement of cat scratch disease usually presents with lytic lesions, which poses a diagnostic challenge, as immunologic, neoplastic, and other infectious etiologies must be ruled out.

**Resources:**

**References:**


Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B116

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**Title:** A Tired Teenager: A Case Report

**Authors:** Emily Frierson, MS3, University of Central Florida

**Case Presentation:** An otherwise healthy 14 year old African American male presented with a 48 hour history of lethargy. Two weeks ago, he was in his normal state of health when he came down with ‘flu-like’ symptoms which consisted of a nonproductive cough and general malaise. After 1 week, he was prescribed Azithromycin for persistent symptoms. He subsequently developed nausea, vomiting and a fever 102°F. The next day, the patient complained of a headache and intermittent blurry vision. In the ED, his parents report all symptoms have since resolved with the exception of worsening fatigue, noting the patient had been asleep for nearly 2 days. On exam, patient was responsive to loud commands but quickly fell asleep during tasks. No neck stiffness or meningeal signs, no focal neurological deficits. Pertinent diagnostic findings include Mycoplasma pneumoniae detected on respiratory PCR and CSF pleocytosis. Patient diagnosed with M. pneumoniae associated encephalitis, and treated with steroids. He improved clinically during hospital stay. By discharge, patient was alert, talkative and ambulatory.

**Discussion:** M. pneumoniae is a common respiratory pathogen among school-aged children that has also been associated with a number of neurological manifestations. Of note, the California Encephalitis Project found M. pneumoniae to be the most common cause of unexplained encephalitis in pediatric patients (1). A recent study suggested two distinct pathological mechanisms: one direct infection of the CNS and one indirect, likely immunologic process (2). The patient described in this case report nearly mirrors the presentation associated with an immunologically-mediated CNS disease: presenting with prodromal period of ≥7 days, respiratory manifestations, reactive IgM in acute serum, and detection of M. pneumoniae in the respiratory tract, but not the CSF (2). It is also possible that the Azithromycin treated an acute infection, further supporting an immunologic process. For this reason, we elected to treat with steroids. The patient had improved clinically prior to administering steroids, so it is difficult to say if the subsequent improvement was due to a resolving process or the steroids themselves.

**Conclusion:** Our otherwise healthy 14 year old male presented with a 48 hour history of lethargy after 2 weeks of flu-like symptoms. His decreased level of consciousness for greater than 24 hours, fever over 100.4°F, and CSF pleocytosis suggested encephalitis. M. pneumoniae was detected on respiratory PCR, and as this microbe can also affect the central nervous system, we treated with steroids.
nervous system, it is the most likely etiology. This patient’s presentation was consistent with an immunologically-mediated CNS disease, and we elected to manage with steroids for anti-inflammatory purposes. The prognosis of M. pneumoniae CNS involvement is variable. While our patient experienced complete recovery, adverse sequelae such focal deficits or epilepsy, and death have been described (2). The goal of this report is to further increase awareness of the neurologic manifestations of Mycoplasma, as well as discuss the variable mechanisms and management strategies.

Resources:
References

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B117

Title: Avoiding Tunnel Vision in the Sexually Active Adolescent with Labial Ulcers
Authors: Lauren Beene, Resident Physician/MD, Rainbow Babies & Children's Hospital
Erin Frank, Attending Physician/MD, Rainbow Babies & Children's Hospital, Michael Dell, Attending Physician, Rainbow Babies & Children's Hospital, Hemangini Bhakta, MD, Rainbow Babies and Children's Hospital, Cleveland Medical Center, Cleveland, Ohio

Case Presentation: A 15-year-old sexually active girl with reported history of HSV2 presented with painful labial swelling, ulceration, and urinary retention. Two weeks prior she had oral surgery; then developed fever, malaise, and tonsillitis. Despite prophylactic acyclovir, genital lesions emerged. Exam revealed severe edema of labia minora, large medial surface ulcerations, thick necrosis, and purulent discharge. Given suspicion for recurrent HSV with superinfection, IV acyclovir and antibiotics were started, but ulcerations were slow to heal. HSV PCR was indeterminate; viral culture, HSV2 IgG, HSV1 and 2 IgM, HIV antigen and RNA PCR were negative; HSV1 IgG positive. Records from initial presentation 9 months prior were similar except for previously negative HSV1 IgG. Bacteriodes grew on culture, and vaginal discharge improved with metronidazole. Diagnosis of Lipschutz ulcers was made given lack of acute HSV, and no ocular or oral findings to suggest Bechet’s. EBV serologies were consistent with late acute or prior infection. She was discharged with supportive care and gynecology follow up.

Discussion: Lipschutz ulcers are non-infectious vulvar ulcers historically diagnosed in non-sexually active early adolescent girls. Uncommon, it is characterized by rapid onset of large (>1cm) deep ulcers on the vestibule and inner labia minora, sometimes as symmetrical “kissing ulcers.” Prodromal symptoms including malaise, fever, and sore throat may be reported. While associations with EBV, CMV, Mycoplasma pneumonia, and other viruses and bacteria have been reported, a causal versus temporal relationship is unclear. Atypical presentations in sexually active older women are increasingly recognized, and recurrence has been reported in one third of cases. Diagnosis is primarily by exclusion of HSV and other sexually transmitted infections, and Bechet’s. Management is supportive including wound care, pain management, and reassurance. In our patient, her presentation was consistent with Lipschutz, although complicated by spurious diagnosis of HSV2. Once HSV2 was excluded, her management was changed, with discontinuation of acyclovir for both acute and suppressive therapy.

Conclusion: This case highlights the importance of maintaining a broad differential; in sexually active adolescents with labial ulceration, there is great potential for tunnel vision. The differential for vaginal ulcers includes infectious and noninfectious causes including HSV, HIV, syphilis, Lipschutz, complex aphthosis, Bechet’s, Crohn’s disease, pyoderma gangrenosum, and autoimmune bullous disease. Our patient’s presentation was unusual in severity and phenotype for
recurrent HSV2, however given her self-reported history, she was treated with acyclovir. During admission, numerous care providers were suspicious of a separate etiology, as no vesicular lesions were visualized and her labial ulceration was slow to improve on acyclovir. By setting aside her prior misdiagnosis and our own subconscious biases, we were ultimately able to diagnose Lipschutz and offer appropriate care. As HSV2 has life-long implications, including the potential for detrimental psycho-social impact, it is extremely important to ensure accurate diagnosis in young sexually active adolescents.

**Resources:**


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B118

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Figure 1: Large ulcerations of labia minora medial surface in patient with Lipschutz ulcers. Photo from day 7 of hospitalization.

**References:**


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B118

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**Title:** Diagnostic approach to a 6-year-old with HIV and fever of unknown origin  

**Authors:** Madeleine Matthiesen, MD, Massachusetts General Hospital  

**Case Presentation:** A 6-year-old girl with a history of HIV presented with several months of fevers, anorexia, mouth ulcers, and abdominal distension. She was born in the US but had traveled to Uganda twice, last two years prior. On admission, she was febrile to 104, tachypneic, and tachycardic. Labs were notable for a normal BMP, elevated AST (183), CRP (134), ESR (>140), CK (1683), and low hemoglobin (7.2). Her CD4 count was 495 (CD4% 28 from 14 prior) with a low viral load (24). CT scans showed normal lungs but generalized lymphadenopathy with enlarged heart, liver, spleen, and pulmonary arteries. Viral, bacterial, tuberculosis, and opportunistic infection (OI) testing was negative. Lymph node and bone marrow biopsies were unrevealing. At this point, several serologies were sent and positive, including ANA (1:5120), anti-RNP (372), anti-dsDNA (1:160), and anti-Ro (133). The patient was admitted to the pediatric hospitalist service for ongoing workup of fever of unknown origin (FUO) with ID, oncology, and rheumatology following.

**Discussion:** Given the patient’s age and comorbidities, we initially considered the patient’s fever to be infectious until proven otherwise. After an extensive negative workup and lack of improvement on antibiotics or TB treatment, we broadened our approach.
Biopsies were not consistent with lymphoma. Autoimmune etiologies are often overlooked among younger children and those with HIV, but we strongly considered immune reconstitution inflammatory syndrome (IRIS) and connective tissue diseases (CTDs).

A diagnosis of exclusion, IRIS was considered given the patient’s intermittent medication compliance. She had had a recent drop in her viral load and a large increase in her CD4%, which can be suggestive of IRIS (3); however, the patient’s time course and level of viremia were less consistent with this diagnosis.

Rheumatology diagnosed the patient with CTD and started the patient on high-dose steroids. After weeks of fever and tachycardia, the patient’s vital signs normalized within 24 hours. Her significantly elevated ANA, Ro, and RNP months later confirmed her diagnosis.

**Conclusion:** FUO is one of the most common, long-standing diagnostic dilemmas facing pediatricians, but with several advances in both treatment and diagnostics, the traditional approach to its differential is shifting. While previously infections made up the majority of cases of FUO of all ages, studies have shown that the rate of infections is dropping and autoimmune diagnoses are on the rise (1). Parallel changes have been seen in the HIV-positive world: prior to the HAART era, the incidence of OIs, pneumonias, and bacteremia made infection by far the most common FUO etiology. However with increasing HAART use and the associated improvement in immune status, these causes of FUO have become less common (3). Given this changing landscape for HIV patients, we wonder whether these patients should be considered to be similar to patients without HIV.

**Resources:**
Fever of Unknown Origin Causes by Age. December 1, 2016.

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B119

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**Title:** Unexplained Neonatal Vomiting: the Secret is in the Fat

**Authors:** Laurie Stover, MD, FAAP, UCSD/Rady Children’s Hospital, San Diego
Brian Williams, MD, UCSD/Rady Children’s Hospital, San Diego

**Case Presentation:** GN is an 18 do term baby born to a G4P4 mother with unremarkable prenatal hx who presented with persistent nonbilious vomiting, lethargy & poor feeding. The patient had been admitted DOL 14-18 for vomiting, at which time she had reassuring CBC, CMP & UGI. Her symptoms resolved over several days with bowel rest. On DOL 21 sxvs recurred & she was readmitted. She had no fever, diarrhea or URI sxvs. She was irritable, not consoled by feeding & mother felt she had abdominal pain. Her FH was notable only for high cholesterol. VS were normal but she was irritable & dehydrated. Weight was 2.96 kg (2%) and 6 g above BW. Abdomen was non-distended but was diffusely tender to TTP. Liver was 2.5cm below the RCM. She vomited whenever fed. A full sepsis eval, CMP & KUB were unrevealing. CT scan revealed a moderate amount of free fluid and diffuse small bowel hyperemia. On HD 2, the RN noted her blood appeared lipemic. A triglyceride level was checked & was 4160 mg/dl. Pancreatitis was then suspected as a cause for the symptoms, & a lipase was found to be 3450 U/L.

**Discussion:** More focused FH was obtained. Parents stated that there was a paternal aunt with a history of triglyceride levels of 20,000 mg/dl & chronic pancreatitis. They reported that both paternal and maternal grandparents had elevated triglycerides and chronic pancreatitis. Parents revealed that they were second cousins. They stated that they did not reveal this information as their own lipids had been found to be normal, so they did not believe their children could develop the disease.

Metabolic & GI were consulted. Pt was NPO until sxvs & labs improved & ultimately required a VLF formula (10%) to maintain her at acceptable triglycerides & lipase. Molecular testing confirmed Lipoprotein Lipase Deficiency (LLD). The patient is a homozygote for a mutation in the G215E gene known to cause LPLD & both parents are heterozygotes.
The patient is now 4 yo and is generally healthy & thriving. She maintains baseline levels of triglycerides of ~ 800-1200 mg/dl & of lipase in the 500-1500 U/L range. She had had several bouts of acute pancreatitis, likely due to difficulties with diet maintenance.

**Conclusion:** LPLD is a rare AR disorder that disrupts the normal clearance of chylomicrons from the blood, resulting in elevated triglyceride levels. Several causative mutations in the LPL gene have been identified. Consanguinity is often observed in patients with homozygous familial LPLD. Patients with familial LPLD typically develop symptoms before age 10, with 25% symptomatic in the 1st year of life. The symptoms are usually abdominal pain & poor feeding due to pancreatitis. If left untreated, pancreatitis can become chronic, cause permanent pancreatic insufficiency, & in rare cases, be life-threatening. Patients may also have HSM and cutaneous xanthoma.

This case highlights the importance of considering neonatal pancreatitis in the ddx of young infants & non-verbal patients with non-specific symptoms of poor feeding & vomiting. It also emphasizes the importance of FH in arriving at a diagnosis in select patients. Parents frequently do not reveal sensitive history that they feel is not relevant, & will often not volunteer consanguinity without direct questioning.

**Resources:**

**Figure 1: Diagnostic Course**

**Figure 2: Lipoprotein Lipase Pathway**

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B120

**Title:** When Bones Cause Diagnostic Groans: An Infant with Bony Abnormalities

**Authors:** Sara Duffus, MD, University of North Carolina
Bradly Thrasher, DO, University of North Carolina, Department of Pediatric Endocrinology, Ali Calikoglu, MD, University of North Carolina, Department of Pediatric Endocrinology

**Case Presentation:** A 4-week-old, term male was found to have a hip click bilaterally on exam. Hip films were interpreted as having an abnormal appearance of the proximal femurs concerning for non-accidental trauma (NAT). Subsequent NAT workup included a skeletal survey with cupping and metaphyseal irregularities in the majority of the long bones, in addition to rachitic rosary (figure 1,2). Initial workup for rickets was inconclusive with normal calcium (9.7), normal parathyroid hormone (25), and only mildly elevated phosphorous (5.8). Endocrinology was consulted and was concerned for imaging features atypical of rickets, in particular a moth-eaten appearance of the talus and calcaneus. Further review of initial labs identified a significantly low alkaline phosphatase level (33), raising concern for hypophosphatasia. Confirmatory testing was consistent with hypophosphatasia, including elevated urine phosphoethanolamine and elevated serum vitamin B6 levels. The patient was initiated on enzyme replacement therapy and experienced nearly complete regression of his bony abnormalities.

**Discussion:** Hypophosphatasia (HPP) is a disorder characterized by low serum alkaline phosphatase activity causing defective bone mineralization. Clinical manifestations in infancy include skeletal deformities and fractures progressing to poor weight gain and respiratory difficulties due inadequate chest wall expansion. The differential diagnosis for an infant with fractures and failure to thrive includes non-accidental trauma, inborn errors of metabolism, primary and secondary rickets, and osteogenesis imperfecta. A complete metabolic panel, including serum alkaline phosphatase, is often obtained, however, low alkaline phosphatase is frequently overlooked as a significant value. Low serum alkaline phosphatase should raise suspicion for hypophosphatasia.
Mortality is high in untreated infantile HPP, greater than 50% at 9 months of age. However, enzyme replacement therapy has become available within the last year and has significantly improved outcomes, including prolonged survival, improvement of bony abnormalities, and decreased respiratory complications.

**Conclusion:** Hypophosphatasia should be considered on the differential diagnosis for infants presenting with bony abnormalities and fractures, particularly if there is also failure to thrive. A complete metabolic panel should be considered as part of the workup, with low alkaline phosphatase levels raising concern for hypophosphatasia and prompting further evaluation. Early diagnosis and treatment, as seen in this case, are crucial and can prevent the development of life-threatening complications of the disease.

**Resources:** N/A

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B121

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**Title:** A case of very late-onset GBS: Adenitis as a harbinger of meningitis.

**Authors:** Maria Quidgley-Martin
Anne Fallon, MD, Children's Hospital of Philadelphia

**Case Presentation:** A 107 day old male infant born at 31 weeks, with a PMH significant for a 68 day stay in the NICU for mild RDS and apnea of prematurity, presented with left-sided neck swelling and redness as well as fever. In the ED, a blood culture and labs were obtained which were significant for bandemia and a significantly elevated CRP. He also underwent imaging of his neck including a CT and U/S which revealed only mildly enlarged cervical and supraclavicular lymph nodes. He was admitted to the general pediatrics floor with a suspected diagnosis of lymphadenitis and received a dose of unasyn prior to admission. Nine hours after it was drawn, his blood culture returned positive, ultimately speciating as streptococcus agalactiae. An LP was performed with cell counts that were consistent with bacterial meningitis, for which he was treated with 14 days of IV antibiotics, transitioning from ampicillin & gentamicin to penicillin after sensitivities returned. He was ultimately discharged with a diagnosis of GBS meningitis with his initial presentation of neck swelling representing cellulitis-adenitis.

**Discussion:** Late-onset GBS disease is defined as GBS infection diagnosed between 7 and 89 days of age while very late-onset GBS disease is a rarer condition that occurs in infants >90 days of age (1). Cellulitis-adenitis syndrome is a well-described entity in late-onset GBS infection and is known to be associated with meningitis, but it has only been rarely documented in very late-onset disease (2,3,4). Our patient’s unusual case of very-late onset GBS infection highlights the importance of awareness of cellulitis-adenitis as a potential harbinger of GBS meningitis in young infants. It is especially imperative to consider this diagnosis in infants >30-60 days old who present with fever, as LPs may not be universally performed and a focal source of infection may be mistakenly interpreted as reassuring (2). This diagnosis should be particularly considered in infants with a history of prematurity, such as in this patient, as it is a known risk factor for both late-onset and, perhaps even more so, very late-onset GBS disease (5,6).

**Conclusion:** Cellulitis-adenitis may be the only physical exam finding in infants with GBS meningitis. Awareness of this clinical syndrome is imperative in order to ensure early diagnosis and treatment, given the risk of neurodevelopmental impairment associated with GBS meningitis in young infants (7). A thorough work-up, including an LP, should be considered even in older infants presenting with neck swelling and fever, particularly if there is a history of prematurity.

**Resources:**

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**Supporting Information**

[Image] A case of very late-onset GBS: Adenitis as a harbinger of meningitis. Anne Fallon, MD. Children's Hospital of Philadelphia
References


Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B122

Title: A rare cause of abdominal pain and fever: hepatic abscess

Authors: Ngozi Eboh, M.D., Texas Tech University Health Sciences Center
Tala Porter, M.D., TTUHSC SOM, Fatma Levent, M.D., FAAP, TTUHSC-Lubbock

Case Presentation: A 5-year-old boy presented to our service with severe waxing and waning periumbilical pain of one week duration. He was also febrile up to 103F. Initial findings were WBC count of 12.4, 87% neutrophils, CRP of 181 and abdominal CT revealing multiple hepatic abscesses requiring drainage. Hepatic abscess cultures grew MSSA, and he was started on targeted therapy with Nafcillin. The presentation of hepatic abscess in this young child prompted an extensive immunological workup, to include immunoglobulins, dihydrorhodamine test for Chronic Granulomatous Disease (CGD), and immunization titers for pneumococcal vaccine. Results revealed normal IgM, high/normal IgE and IgG, very low IgA, and no response to the pneumococcal vaccine. DHR test was positive, yielding a diagnosis of CGD. He was discharged with a prolonged course of Nafcillin and close infectious disease and immunology follow up. Following diagnosis of CGD, sulfamethoxazole-trimethoprim and itraconazole prophylaxis was commenced and he was placed on the Allogenic Hematopoietic Stem Cell Transplant (HSCT) List.

Discussion: This case was unique in the etiology for the abdominal pain presentation, which prompted investigation for the underlying condition leading to development of hepatic abscess. Top differentials included immune disorders, particularly CGD. CGD is a primary immunodeficiency disorder of phagocytes (neutrophils, monocytes, macrophages, and eosinophils) resulting from impaired killing of bacteria and fungi. CGD is characterized by multiple bacterial and fungal infections and dysregulated inflammatory response. Hepatic involvement is typically in the form of abscess. Diagnostic tests measure neutrophil superoxide production, with the dihydrohodamine (DHR) test being the most currently used. In North America, most infections are caused by S. aureus, B. cepacia complex, S. marcescens, Nocardia and Aspergillus species. A combination of sulfamethoxazole-trimethoprim, itraconazole and interferon gamma is imperative in preventing morbidity and mortality.

Conclusion: Combining anti-bacterial, antifungal and immunomodulatory therapy is part of the prophylactic regimen for patients with CGD. Sulfamethoxazole-trimethoprim has shown to be effective in decreasing bacterial infections and itraconazole reduces the risk of fungal infections. One retrospective analysis showed that sulfamethoxazole-trimethoprim decreased the risk of serious bacterial infection by about 50%, while those receiving itraconazole prophylaxis did not suffer serious fungal infections as compared to placebo. Although it is unclear the mechanism that interferon gamma affects CGD, when combined with sulfamethoxazole-trimethoprim, the risk of bacterial infection was even further decreased. HSCT is the only known cure. Liver involvement is a significant cause of morbidity and invasive fungal infections remain the leading cause of mortality in patients with CGD. Therefore, it is essential that patients with CGD receive prophylaxis with sulfamethoxazole-trimethoprim, itraconazole and interferon gamma and high index of suspicion maintained for CGD in patients with a similar presentation.
Title: Altered mental status in a healthy child: the cat's out of the bag!

Authors: Mary Terrell, MD, University of North Carolina Department of Pediatrics
Denise Jones, MD, UNC Chapel Hill Medical School, Joanna Hales, MD, UNC Chapel Hill Medical School, Michael Cinoman, MD, Moses Cone Health

Case Presentation: A 6-year-old boy presented to the ED with altered mental status. He was found acutely unconscious at home after returning from his soccer game. Parents denied trauma, ingestions or recent fevers. Initial vital signs were normal. He was intubated for GCS of 6. Physical exam revealed a 3 cm inguinal lymph node for which his pediatrician had started cephalexin for lymphadenitis. LFTs, CBC, lactate, blood gas, urine drug screen and chemistries were unremarkable. CT scan showed mild cerebral edema. LP was performed with an elevated opening pressure but CSF studies were otherwise normal. Parents did endorse exposure to a cat and Bartonella titers were sent. An EEG showed no seizure activity and MRI was normal. He was extubated within 12 hours and was discharged with a normal neurologic exam. However, he was readmitted within 24 hours with seizures and required multiple antiepileptic drugs. He returned to neurologic baseline and was discharged home. CSF Bartonella henselae IgG later resulted positive (1:640) consistent with cat-scratch disease encephalopathy (CSDE).

Discussion: Cat scratch disease (CSD) is caused by Bartonella henselae, a gram-negative bacilli. Cats are the major carrier. Close to 90% of patients with CSD have a history of contact with cats. Lymphadenopathy occurs 1-3 weeks after inoculation and is usually asymmetric involving a single node. Most patients have uncomplicated lymphadenitis as the main presentation, but the spectrum of CSD includes: lymphadenopathy, fever of unknown origin, eye disorders, endocarditis, CSDE and status epilepticus. CSDE occurs in approximately 2% of patients, 46-80% of whom develop seizures. CSF changes are often minimal with little or no pleocytosis and occasionally elevated protein. Brain imaging is often normal and EEG will typically show early changes of diffuse slowing. Diagnosis is made with serologic testing with positive IgG to Bartonella henselae. IgM is often negative as production is brief. There is little evidence that antimicrobials help in CSDE, but are often used. Options include azithromycin, rifampin and doxycycline. Prognosis is good in patients with CSDE with >90% achieving complete recovery.

Conclusion: Most clinicians consider CSD in the differential diagnosis of the wide variety of presentations associated with lymphadenopathy, especially if there is known exposure to cats. This case highlights the importance of considering CSDE when that presentation also includes altered mental status and/or seizures. Diagnosis is made by history in conjunction with a positive Bartonella henselae IgG. In addition, Bartonella henselae IgM titers often result negative, as the acute phase is often missed by the time of presentation. Early consideration of CSDE in these cases may help direct diagnostic testing, treatment and prognosis.
Title: Don't Believe Everything You See: Visual Hallucinations in an 8 year old
Authors: Caroline Spalla, MD, University of Texas at Austin Dell Medical School
Case Presentation: 8-year-old male with spina bifida and shunted hydrocephalus presents to the ER complaining of recurrent visual hallucinations. He states the episodes all follow the same pattern: his face turns visibly red, his face feels warm, and he then develops a headache and feels dizzy. The visual hallucinations are the final symptom and he describes them as seeing bumps on his face and blood on both his hands. One month prior to the onset of symptoms, the patient had a generalized tonic clonic seizure in the setting of fever and acute pyelonephritis. The patient states he "heard voices in his head" prior to the onset of that episode. EEG was read as normal. No anti-epileptic medications were started. The patient is afebrile with normal vital signs. His neurologic exam is at baseline remarkable only for bilateral decreased sensation and strength in lower extremities. He denies active hallucinations. Electrolytes and urine drug screen normal. Shuntogram normal. Urine is positive for nitrites and leukocyte esterase. EEG is markedly abnormal with epileptiform activity in bilateral frontal lobes.

Discussion: We considered a broad differential diagnosis on presentation. Ventriculo-peritoneal shunt malfunction can present with acute mental status changes and behavioral changes. This was ruled out with normal head imaging and no sign of increased intra-cranial pressure on exam. The aura-like events preceding his hallucinations made migraines a possibility. Visual auras associated with migraines are usually simple hallucinations, with typically linear or geometric shapes. Most visual hallucinations in psychiatric illness are complex or formed and can include images of people, animals, and lifelike scenes. Due to the detailed and formed nature of his hallucinations, we considered acute psychosis as a diagnosis, although the patient had no symptoms of depression, mania, anxiety, or delusions. The etiology of frontal lobe epilepsy as well as the response to anti-epileptic medications is still largely unknown. Our patient had significant cognitive delay at baseline. Children with frontal lobe epilepsy typically have impairments in executive functions, impulse control, and attention span.

Conclusion: The diagnosis is complex partial seizures originating in the bilateral frontal lobes. The patient’s formed hallucinations were actually ictal manifestations of his complex partial seizures. Although these types of seizures are unusual, the patient has a higher risk of epilepsy due to his history of spina bifida and shunted hydrocephalus. The EEG results prompted the Neurology team to re-evaluate the patient’s prior EEG, which was previously read as normal. On further review, a few bursts of epileptiform discharges from the bilateral frontal lobes were actually present on the initial EEG. The findings were not impressive initially but were able to be localized as a result of the most recent EEG findings. The patient was loaded with levetiracetam (50mg/kg) and then started on daily levetiracetam (10mg/kg BID). He was also treated for an acute urinary tract infection. He had no additional hallucinations and no seizure activity the remainder of his hospital stay. He was discharged home to follow up as an outpatient with Pediatric Neurology.

Resources: N/A
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B125
macrophage activation and suspected polyclonal lymphocyte activation leading to production of non-specific antibodies felt to have led to misdiagnosis of mycoplasma and/or acute rheumatic fever.

**Discussion:** Subcutaneous panniculitis-like T cell lymphoma presents with painless subcutaneous nodules or plaques. 20% of patients present < 20 years of age. 20% of patients will have an associated autoimmune disorder at presentation. 60% will have systemic B symptoms, 20-30% will have abnormal bone marrow (most commonly hemophagocytosis). 45% will have lab findings of decreased cell lines and/or elevated transaminases. Diagnosis is via deep skin biopsy. Median time to diagnosis is 7 months. Treatment is chemotherapy. Prognosis depends on presence/absence of secondary HLH (46% 5 year survival with, 91% without). Polyclonal antibody activation has been described from various infectious, autoimmune and oncologic triggers. This may in turn lead to the development of autoimmune conditions. Non-specific antibody activation may also make it difficult to interpret serologic studies ordered during diagnostic workup.

**Conclusion:** Evolving symptoms and overlap with other disorders can lead to delay in diagnosis for patients with rare conditions such as subcutaneous panniculitis-like T cell lymphoma. Macrophage activation and polyclonal lymphocyte activation can lead to production of non-specific antibodies and serological findings which may be misleading in diagnostic workup of clinical conundrums.

**Resources:**

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B126

**Title:** Granulomatosis with Polyangitis Presentation with Uncommon Features

**Authors:** Daniel Hershey, M.D., Rady Children’s Hospital

Wen Jiang, MD, University of California, San Diego

**Case Presentation:** A 10 year-old girl had ear tubes placed 1 week prior to admission, after failing 3 different courses of antibiotics over 6 weeks. 5 days prior to admission, she developed fever to 103, throbbing frontal headache, purulent otorrhea and rhinorrhea, intermittent epistaxis, intermittent right mastoid pain, and right-sided facial nerve palsy. 2 weeks prior to admission, her gums were described as purple with red spots, but within a couple days of the ear tube surgery, much of the purple discoloration resolved. At the time of admission, there was still some gingival discoloration and edema(see attached image). Also, the nasal mucosa was noted to be crusted with blood, with friable mucosa. Labs on admission were significant for ESR 63, CRP 21.3 mg/dl, WBC 14 with left shift, and fibrinogen >1400. Otherwise the CBC, CMP, and PT/PTT were normal. A head CT demonstrated right, greater than left, otomastoiditis and pansinusitis. She was admitted for treatment of mastoiditis, but she did not have the typical exam findings.

**Discussion:** Subsequent evaluation demonstrated pulmonary nodules and adenopathy, microscopic hematuria, and strong positive CANCA. Gingival biopsy demonstrated pseudoepitheliomatous hyperplasia, which in this context is nearly pathognomonic for GPA (Granulomatosis with Polyangitis, previously known as Wegener’s Granulomatosis). Once the diagnosis became clear, antibiotics were discontinued, and the patient demonstrated clinical improvement after initiation of systemic steroids.
The key here was that it was difficult to tie gingival petechiae to any one of these infections, and that too many different systems were involved. Gingival petechiae are seen with vasculitis, malignancy, or coagulopathy. Given the inflammatory markers and fever, and normal PT/PTT and platelets, vasculitis is most likely. The dramatic clearing of the purple gingiva described by parents, was later determined to likely be result of steroids that were administered after her surgery.

**Conclusion:** Among children, GPA most commonly affects females in the early teen years. Her involvement of the sinuses, nose, and ears are also very common for GPA initial presentation. She had several symptoms that are quite rare, even for GPA. Gingival petechiae and peripheral neuropathy are each seen in less than 5%, and pulmonary nodules 10% of pediatric GPA presentations. Diagnosis can be challenging, with lung and kidney biopsy not resulting in definitive diagnosis half the time. The cANCA can be negative 20% of the time, and is nonspecific.

GPA is a vasculitis that is very rare in children. It can present with a varied combination of clinical features depending on what organs are affected. Recognition of the association between GPA and gingival petechiae, can help speed the diagnosis.

**Resources:**
Ludici M, Quartier P, Terrier B, et. al. 1,2 Pierre Quartier,3 Benjamin Terrier,1 Luc Mouthon,1 Loïc Guillevin,1 and Xavier Puéchal. Childhood-onset granulomatosis with polyangiitis and microscopic polyangiitis: systematic review and meta-analysis. Orphanet J Rare Dis. 2016; 11: 141.

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B127

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**Title:** How Low Can Na Go? Severe Hyponatremia in a Patient with Failure to Thrive

**Authors:** Jeanine Ronan, MD, MSEd, MS, The Children's Hospital of Philadelphia
Catherine Mezzacappa, MPH, Perelman School of Medicine at the University of Pennsylvania, Eloise Salmon, MD, Children’s Hospital of Philadelphia, George Dalembert, MD, Children’s Hospital of Philadelphia

**Case Presentation:** A 5-month-old Caucasian male is referred to the ED for weight loss in the setting of resolved febrile illness 1 week ago, with subsequent decrease in breastfeeding. At his PMD’s office he was noted to have simultaneously lost growth progress in weight, height, and head circumference for the past 4 months. He was born at term and had a normal newborn screen. He has a history of buried penis and prenatal bilateral hydronephrosis with persistent but resolving right sided fullness. His family history is notable for a 3-year-old brother with unspecified growth and developmental delay. On exam in the ED he is afebrile with pulse 110, BP 112/69, and appears thin and fussy, but otherwise well. Initial labs are notable for sodium 119, potassium 5.4, chloride 83, carbon dioxide 18, BUN 45, and creatinine 0.5, and urine dip significant for large leukocytes and large blood. A renal bladder ultrasound shows right pyelonephritis with megaureter and layering debris. After volume resuscitation, he is started on broad-spectrum antibiotics and is admitted to the pediatric ICU.

**Discussion:** An infant with a history of genital and renal anomalies presents for failure to thrive and is found to have pyelonephritis, profound hyponatremia, and hyperkalemia. His failure to thrive and electrolyte abnormalities may be explained by a single underlying pathology or may be unrelated. His failure to thrive is not well explained by decreased intake or malabsorption by history, indicating a metabolic or increased energy demand etiology. It seems unlikely that his current pyelonephritis alone could be responsible given the chronicity and symmetry of his growth failure. Furthermore, despite his profound hyponatremia, this infant appears well, suggesting chronic sodium losses. Despite his normal newborn screen, this infant’s genitourinary anomalies are initially concerning for rare etiologies of congenital adrenal hyperplasia. A pituitary cause could explain both his electrolyte abnormalities and symmetric growth failure. Finally, a type 4 renal tubular acidosis, aldosterone deficiency or resistance to its actions, could similarly lead to chronic sodium wasting and failure to thrive.

**Conclusion:** Despite the abnormal appearance of his external genitalia, the patient was found to have normal bilateral descended testicles. Additional laboratory studies were obtained. ACTH was within normal limits and cortisol, renin, and
aldosterone were appropriately elevated, ruling out a pituitary process, congenital adrenal hyperplasia, or intrinsic deficiency of aldosterone production. With IV fluid resuscitation his sodium improved to 128 and was maintained at this level after several days of exclusive breastfeeding without fluid supplementation. It was concluded that he suffered from a transient pseudohypoaldosteronism due to an indolent, chronic pyelonephritis resulting in renal tubule insensitivity to aldosterone, and he was discharged with nephrology and urology follow-up, with the expectation that his electrolytes would normalize after the insult of the pyelonephritis was allowed to resolve. A repeat metabolic panel 10 days after discharge showed a serum sodium of 138 and a follow-up VCU confirmed Grade 4 VUR on the right side and Grade 1 VUR on the left.

Resources:
Supplement I. Patient’s growth charts prior to admission

Supplement II. Selection of patient’s additional laboratory results

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<tr>
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Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B128
**Title:** Keeping an Eye Out for a Diagnosis in a Patient with Acute Vision Loss  
**Authors:** Paula Soung, MD, Medical College of Wisconsin  
**Case Presentation:** A previously healthy 14 yo female with history of anxiety presented for acute vision change in the left eye. Ophthalmology diagnosed intra retinal hemorrhages with central whitening, consistent with multiple Roth spots. No associated fever, trauma or other neurologic complaints. On further history, patient had 30lb weight loss attributed to anxiety with recurrent emesis in the mornings prior to school. On admission, vitals significant for BP 142/73. Exam normal except decreased left peripheral vision and acuity. Initial labs notable for: hemoglobin 6.6, MCV 53, mildly elevated CRP 3.7 and ESR 43. MRI brain with nonspecific changes and normal angiography. Echo identified mild left ventricular hypertrophy but normal structure without vegetations. Carotid ultrasound (US) showed moderate wall thickening of the bilateral common carotids. Aorta US and renal US were normal. MRI and MRA of the chest and abdomen revealed wall thickening and enhancement of the ascending, transverse and descending abdominal aorta, consistent with large vessel vasculitis such as Takayasu arteritis.  
**Discussion:** Atraumatic acute vision loss is an uncommon presentation in pediatrics. Evaluation is time sensitive and etiology of acute vision loss can guide further work up. Roth’s spots have been associated with a variety of conditions including subacute bacterial endocarditis, other infections, leukemia, anemia, hypertension, ischemic events, trauma and vasculitis. The finding of Roth spots lead to thorough evaluation revealing unprovoked weight loss, fatigue, anorexia, nausea and vomiting, hypertension, microcytic anemia, and mildly elevated inflammatory markers. Early symptoms of Takayasu arteritis are non-specific and thorough exam for reduction in pulses, differences in blood pressures, or bruits narrows the differential diagnosis. Less invasive procedures such as ultrasound of the proximal aorta, carotids, and brachial arteries may show vessel wall thickening and luminal narrowing. In most cases, the diagnosis is based on clinical features and imaging of the arterial tree by MRI, CT, or angiography with luminal narrowing or occlusion accompanied by thickening of vessel walls.  
**Conclusion:** Retinal hemorrhages, or Roth spots can frequently be the presenting condition for a systemic disease. Visual loss is the most common ocular symptom in Takayasu arteritis and Takayasu retinopathy is well reported. Roth spots as a primary presentation of Takayasu arteritis has not been reported. In this scenario, it remains unclear if the etiology of Roth spots is best attributed to anemia, hypertension, or vasculopathy. However, acute vision changes are uncommon in the pediatric population and should prompt a thorough history and physical exam followed by appropriate evaluation. The presence of constitutional symptoms such as fever, malaise, weight loss, myalgias, abdominal pain, and ischemic symptoms or signs of one or more large arterial stenoses should raise suspicion for Takayasu arteritis. Early diagnosis and treatment of systemic diseases such as Takayasu arteritis can help prevent life altering complications.  
**Resources:**

References  
**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B129

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**Title:** Lupus Enteritis as Initial Presentation of Systemic Lupus Erythematosus  
**Authors:** Peter Roman, MD, Pediatric Resident (PL-2), Hasbro Children’s Hospital  
**Case Presentation:** 16-year-old girl with four days of vomiting and diffuse abdominal pain, without diarrhea or fever. Admitted four months prior with vomiting and epigastric pain. Presumed to have gastritis after a normal KUB and...
abdominal ultrasound, and improved with IVF and PPI. Noted to have leukopenia (2.1), thought related to viral suppression. In the interim, had complete resolution of symptoms. Upon re-presentation, once again had leukopenia (2.9), with normal Lipase, LFTs, UA and HIV. KUB suggestive of SBO given multiple air-fluid levels. Abdominal CT concerning for transgastric internal hernia with sequestered jejunal loops, along with moderate ascites, bowel wall thickening, and trace pleural effusions. Diagnostic laparoscopy revealed diffuse bowel dilation without internal hernia, obstruction or volvulus. Additional history revealed episodic urticaria and intermittent pale hands, and was found to have an ESR (23) and speckled ANA titer of 1:10240. Diagnosis of Lupus confirmed after elevated anti-Smith, anti-SSA, anti-RNP AB. She received pulse dose steroids, and had symptomatic resolution.

Discussion: SLE is a frequently elusive diagnosis that can produce symptoms involving nearly every organ system, with significant patient to patient variation. GI involvement is not uncommon, and has been described to include everything from esophagitis to pancreatitis, to hepatic steatosis. Enteritis is an uncommon presentation of Lupus, and does not appear in the diagnostic criteria proposed by the ACR or SLICC. Few case reports describe initial presentation of SLE with Enteritis, and none appear described amongst the Pediatric population. In this case, while possible small bowel obstruction was on our initial differential based on radiographic findings, Lupus was not considered a diagnostic possibility. Even after her laparoscopy, differential included infectious gastroenteritis, cyclic vomiting syndrome, hereditary angioedema, immunodeficiency and lymphoma. Her leukopenia remained a concerning finding as a SLE lab criteria, and led to additional history describing past hives and likely Reynaud’s phenomenon. ANA thus was obtained, and was found to be wildly positive, leading to diagnosis.

Conclusion: Abdominal pain and vomiting are amongst the most common presenting symptoms to our clinics and hospitals. The relapsing and remitting pattern with varying multiorgan system involvement of Lupus can present significant difficulty in diagnosis. Lupus should be considered as a potential etiology of enteritis, when symptoms do not respond to conservative management, and the patient presents with signs of possible serositis, such as ascites and pleural effusions. In this case, careful history, and re-assessment of differential diagnosis led to the correct diagnosis. IV steroids is the standard of care for initial treatment and demonstrated dramatic improvement in this case.

Resources: N/A

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B130

Title: Pneumonia on Steroids!

Authors: Robyn Kuroki, MD, CHLA
Rory Kretzmer, MD, CHLA, Grant Christman, MD, CHLA

Case Presentation: A 3 year old girl with trisomy 21 and PFO presents with 3 days of fever, cough, and respiratory distress not improving on amoxicillin by Pediatrician. On exam she was febrile, tachycardic, had wheezing with retractions and saturation of 94% on room air. Chest XR showed bibasilar opacities, WBC 15.9, Hgb 8.1, and nasopharyngeal swab +rhinovirus/enterovirus. Ceftriaxone was started for presumed community acquired pneumonia. Repeat hgb was 6.4, and she was transfused with improvement to 9.1. Vancomycin and Azithromycin were added due to continued need for increased oxygen support. Repeat XR showed extensive infiltrates consistent with "multilobar pneumonia". Bronchoalveolar lavage demonstrated frankly bloody fluid from bilateral lower lobes and hemosiderin-laden macrophages. She was diagnosed with idiopathic pulmonary hemosiderosis (IPH) and started on daily IV methylprednisolone. She had a thorascopic right lung biopsy with histology confirming IPH, however required a chest tube for pneumothorax and ultimately pleurodesis. She was discharged in excellent condition 2 months later.

Discussion: IPH results from hemorrhage in the low-pressure alveolar circulation, which is often diffuse and insidious, although rarely can be fulminant. This results in the nonspecific symptoms of malaise, cough, and dyspnea with tachypnea and crackles on exam. Hemoptyisis may be absent. CBC shows microcytic anemia. Chest XR reveals bilateral, diffuse alveolar and interstitial opacities. Work-up centers on evaluation for pulmonary-renal syndromes, coagulation disorders, and cardiovascular pathology. Serologies are sent for autoantibodies, and flexible bronchoscopy with alveolar lavage is performed with staining for hemosiderin-laden macrophages. In patients without cardiovascular pathology or autoimmune serologies, lung biopsy is increasingly recommended as pulmonary capillaritis may be found. Pulmonary capillaritis is suggestive of a systemic vasculitis, which is challenging to treat and tends to recur more frequently than IPH. Initial management for IPH is systemic corticosteroids. 5 year survival may be greater than 80% although some cases can progress to pulmonary fibrosis.
Conclusion: 1. Simple community acquired pneumonia should respond to appropriate antibiotic therapy within 48 hours. If no improvement is noted, differential should be expanded to include complicated pneumonia (empyema, effusion, foreign body, aspiration), other infectious etiologies (atypical bacterial, viral, fungal), vasculitides, and rarer pulmonary diagnoses.
2. The use of the term “pneumonia” in a radiology report can obscure the fact that interstitial and alveolar opacities may also be caused by edema, atelectasis, malignancies, and hemorrhage.
3. Significant acute anemia requiring transfusion should be factored into the differential diagnosis. In patients with respiratory distress, pulmonary infiltrates, and anemia, pulmonary hemorrhage, although rare, should be considered.
4. Diffuse alveolar hemorrhage may be immune-mediated. IPH is a diagnosis of exclusion where pulmonary capillaritis and autoantibodies are absent. Given the type and length of treatment, other diagnoses must be excluded prior to starting treatment.

Resources:

References:

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B131
Title: Salmonella, an unusual pathogen in infantile meningitis?

Authors: Emily Sou, DO, Maimonides Medical Center
Alicia Kodsi, MD, Maimonides Medical Center

Case Presentation: A 7 week old male presented febrile to 103.3°F, HR 188 bpm, and RR 48/min. He was ill-appearing, lethargic, mottled, delayed capillary refill, and with a bulging fontanelle. Patient had one seizure-like episode lasting <30 seconds involving posturing of upper extremities and eye rolling. All cultures were obtained and he was started on Vancomycin and Ceftriaxone. CSF showed: WBC count of 1,000μL, RBC 63μL, protein 252mg/dL, glucose <10mg/dL; Gram stain showed gram negative rods. Non-contrast CT of the head was unremarkable. Patient was started on a dopamine drip following fluid resuscitation and Keppra was started. A vEEG was nonspecifically abnormal and he had no further clinical seizures. CSF culture grew Salmonella Heidelberg susceptible to Ceftriaxone. Repeat lumbar puncture showed clearance of bacteria from the CSF. Blood, urine and stool cultures remained negative. Echo and abdominal ultrasound were unremarkable. Contrast MRI of the brain showed a subdural empyema over the right temporal and inferior frontal lobe. Neurosurgery recommended medical management with serial imaging.

Discussion: To our knowledge this is the first clinical case report of S. Heidelberg meningitis published in the United States since 1976.3 Of 383 CSF isolates of Salmonella reported to the CDC from 1968-1979, 62 cases were S. Heidelberg.4 Infants <1 year of age accounted for more than half of CSF Salmonella isolates with a majority of them in infants <3 months of age. Of 103,420 Salmonella Heidelberg isolates reported to CDC from 1968-2011, 7.7% were from blood, CSF, and joint fluid, but neither the exact number of cases from CSF or from infants was available in the report.5 The majority of Salmonella Heidelberg non-human isolates are reported from chickens.5 While a causal association between the family members’ handling of chicken obtained from a live poultry market was not definitively identified as the source in this case, transmission possibly occurred via a family member’s hands becoming contaminated with Salmonella Heidelberg and then transmission via direct handling of the infant, objects in contact with the infant or formula preparation.

Conclusion: Four to six weeks of intravenous antibiotics are recommended for the treatment of Salmonella meningitis,1 but presence of an intracranial abscess could require a longer duration of treatment. Practitioners should strongly consider brain MRI with contrast in infants with Salmonella meningitis to assess for complications such as intracranial abscess. Long term follow up of children with Salmonella meningitis should include hearing screening as well as close developmental surveillance with referral to appropriate services as necessary.

S. Heidelberg meningitis confers a risk of significant morbidity and must be promptly diagnosed and treated. Unusual pathogens causing meningitis should be considered during initial evaluation of an infant with fever especially in the context of a concerning exposure history. When eliciting an exposure history an emphasis should be placed on animal contacts of both the patient and family members, including associations with live poultry markets.

Resources:

References

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B132
Title: Seizing High-Value for a Seizing Patient  
Authors: Maria Martin, BS, The Ohio State University College of Medicine  
Zach Rossfeld, MD, Nationwide Children’s Hospital, Allison Heacock, MD, Nationwide Children's Hospital  
Case Presentation: A 15 day-old male presented to the ED with seizures. Born vaginally at 37 weeks and discharged in two days, he began to have rhythmic twitching of his right arm lasting 3-5 seconds at 7 days of life. Family history was positive for similar episodes in newborns but by 14 days of life he had seizures every 10 minutes, for up to 1 minute, involving all extremities. Labs showed hypocalcemia, hyperphosphatemia, hypovitaminosis D and high PTH (Table 1). He received calcium gluconate and levetiracetam and was admitted. His CSF was confirmed as sterile, EEG showed excessive cortical irritability without focal abnormality, and imaging ruled out athymia. After receiving multiple doses of calcium gluconate he had no further seizure activity. His mother was asked to have labs drawn and had a low 25-OH vitamin D level with normal calcium, phosphorous, and PTH. He responded to calcium carbonate, cholecalciferol, and calcitriol and was discharged on hospital day 7 to primary care and endocrinology follow-up for hypocalcemic seizures and hypovitaminosis D secondary to maternal hypovitaminosis D.  
Discussion: The human body regulates calcium with fantastic precision. There is a physiologic nadir of 8 to 9 mg/dL at 24 hours of life prior to normalization by two weeks of age. While preterm infants and infants of diabetic mothers frequently have an exaggerated, symptomatic nadir, pathologic hypocalcemia may stem from parathyroid, genetic, or nutritional causes. Short newborn nursery stays may delay diagnosis of hypocalcemia and more infants may present to the hospital setting for evaluation. Establishing the etiology of neonatal hypocalcemia requires a step-wise approach that prioritizes stabilizing the patient, history and physical exam, and a value-conscious workup. In this case, inherited pseudohypoparathyroidism was plausible given family history but neither the patient nor his mother had characteristic metacarpal shortening and lab testing (i.e., cAMP responsiveness, CaSR gene analysis) was not readily available. Maternal labs ultimately established the diagnosis, obviated the need for extensive genetic testing, and allowed for more timely treatment in both the patient and his mother.  
Conclusion: We describe a case of maternal hypovitaminosis D presenting as neonatal hypocalcemia with seizures. The diagnosis was established in measuring maternal vitamin D and electrolyte levels. Maternal hypovitaminosis D is an important cause of neonatal hypocalcemia and should be kept high on the differential. Prior to genetic testing and extensive imaging, teams should consider acquiring maternal labs in the evaluation of neonatal hypocalcemia.  
Resources:  

<table>
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<th>Table 1: Laboratory Results</th>
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<tr>
<td>Corrected Total Calcium (mg/dL)</td>
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<td>Patient Lab Value on Presentation</td>
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<td>Maternal Lab Value</td>
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<td>Reference Range</td>
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Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B133

Title: Tachycardia: Going beyond the Usual Suspects  
Authors: Amanda Rogers, MD, Medical College of Wisconsin  
Anna Schmitz, MD, Medical College of Wisconsin  
Case Presentation: A 35.5 WGA, now 29 day old girl with a history a Sickle Cell Disease (HbSS) presented after an episode of bilateral upper extremity shaking, whole body stiffness, and eyes rolling back. The event lasted less than one minute with immediate return to baseline. Upon presentation to the Emergency Department, she had a normal physical exam aside from tachycardia to 200 bpm. She was admitted for evaluation of a Brief Resolved Unexplained Event (BRUE). Her resting tachycardia persisted despite fluid resuscitation, and a head CT, CBC, electrolytes, infectious studies, ECG, chest x-ray and EEG were all unrevealing. Family history was notable for maternal Grave's disease during
pregnancy. Further evaluation showed hyperthyroidism, and the patient was subsequently diagnosed with neonatal Grave's disease.

**Discussion:** Hospitalists frequently manage infants presenting with unexplained events. In 2016, the AAP released the first guidelines addressing BRUE, formerly known as Apparent Life-Threatening Events (ALTE). A BRUE is defined by an infant with a brief resolved episode involving change in: color, breathing, tone or responsiveness. The guidelines provide a risk-based approach to a BRUE and management recommendations for low-risk cases. When history and exam are consistent with a low-risk BRUE, the guidelines support minimal testing. This case was not low-risk as she had persistent tachycardia, age <60 days and prematurity. A focused evaluation was indicated, revealing the potentially fatal diagnosis of neonatal Grave's disease.

Tachycardia is commonly encountered in the hospital, and most often is due to fever, hypovolemia, anxiety, pain, cardiac pathology or anemia. When common etiologies are excluded, it is pertinent to consider rarer causes. Endocrinopathies including hyperthyroidism can present with elevated heart rates and should be included on the differential for unexplained tachycardia.

**Conclusion:** Hospitalists are often on the frontline for managing patients who are admitted following a BRUE. It is imperative to understand and apply the new AAP BRUE guidelines appropriately to avoid unnecessary and excessive testing in low-risk patients while still identifying higher risk patients who may need further evaluation. A thorough history and physical is imperative in the evaluation of these patients, and any abnormalities should be noted as they may help identify underlying pathology requiring intervention. Persistent tachycardia is an abnormality which merits further evaluation, starting with the most common etiologies. When the usual suspects have been excluded, a comprehensive history and physical exam should be done in the context of a broader differential diagnosis of tachycardia followed by targeted testing and management.

**Resources:**

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B134

**Title:** The Case of the Vanishing Cerebral Aneurysm

**Authors:** Kimberly Monroe, MD, University of Michigan Pediatric Hospital Medicine
Yael Braunschweig, PhD, University of Michigan, Bethany Mohr, MD, FAAP, University of Michigan

**Case Presentation:** A six-week-old boy presented to the hospital with a 1-day history of non-bloody, non-bilious emesis and irritability. On physical exam, the patient was noted to have a bulging anterior fontanelle. Head CT demonstrated bilateral subdural fluid collections and MRA further showed an ischemic infarct in the right temporal lobe and a 3-4 mm basilar artery aneurysm.

Ophthalmologic exam revealed too numerous to count bilateral intra- and pre-retinal hemorrhages extending to the periphery. CTA redemonstrated previous imaging findings including the basilar aneurysm (Figure 1). Neurosurgery recommended a catheterized angiogram to further characterize the basilar aneurysm. Catheterized angiogram, however, revealed no evidence of an aneurysm. Instead, tortuosity of the posterior cerebral arteries including the presence of a vertical loop gave the vessel a bulbous appearance that suggested aneurysm on CTA and MRA.

A workup for hematologic and metabolic causes was negative. Given the patient's ophthalmologic and intracranial injuries, a diagnosis of abusive head trauma was made.

**Discussion:** Abusive head trauma (AHT), is defined by the CDC as “injury to the skull or intracranial contents...due to inflicted blunt impact and/or violent shaking.” 1 Symptoms are typically nonspecific and include vomiting, seizures, and altered mental status. 2 AHT is a diagnosis of exclusion and requires ruling out birth-related trauma, vascular
abnormalities and other genetic disorders. In this case, what appeared to be an aneurysm on imaging raised concern for a genetic or vascular cause of the patient’s presentation.

Catheterized angiography provides high magnification images of up to 0.1-0.2 mm.3 For our case, it distinguished arterial tortuosity from aneurysm and enabled vascular defect to be ruled out. Catheterization is the gold standard diagnostic approach, yet its invasiveness and complication rate remain significant drawbacks.4 The two main alternative approaches are CTA and MRA. Both are minimally invasive options with relatively high specificity and sensitivity. Reported false positive rates, however, range from 0% to 20.5% for CTA and 5.6%-38% for MRA.3,5,6

**Conclusion:** Pediatric intracranial aneurysms are rare, accounting for 0.5%-4.6% of all intracranial aneurysms.7,8 Trauma accounts for between 5-40% of cases, with other causes including infection, cerebrovascular abnormalities, polycystic kidney disease, connective tissue disorders, tuberous sclerosis, and sickle-cell disease.8,9,10,11

Given the ramifications of an AHT diagnosis, it is imperative to exclude all other possible diagnoses. Here this involved the exclusion of vascular defects, birth trauma, bleeding disorders and metabolic conditions. Despite negative lab results, diagnostic uncertainty remained due to the MRA and CTA findings. Ultimately, catheterized angiogram ruled out aneurysm and enabled appropriate planning and care for the patient.

CTA and MRA are increasingly used screening tools for intracranial aneurysm. While these tests provide non-invasive options, practitioners should keep in mind their false positive rates when interpreting findings and determining diagnostic and treatment plans.

**Resources:**

![CT Angiography](image)

**REFERENCES**


Title: The Endless Acidosis

Authors: Jennifer Buehler, MD, St. Louis University, Department of Pediatrics
Marta King, MD, MED, Saint Louis University School of Medicine, Aline Tanios, MD, St. Louis University

Case Presentation: 5 week old, former 36-week SGA male presented with poor feeding, acute onset profuse, watery diarrhea, and decreased urine output. He had no fevers or emesis, and no ill contacts. Vital signs were within normal limits for age. Exam showed pale infant with dry mucous membranes, but was otherwise normal. Birth weight was 1761 grams (1.1%ile), 2070 grams (<0.01%ile) on admission. Diagnostic studies were significant for non-gap metabolic acidosis (bicarbonate 11). Working diagnosis was severe dehydration secondary to viral gastroenteritis. He received 20 ml/kg normal saline then was placed on 1.5x maintenance fluids with D5 1/2NS. 1:1 stool replacement with lactated ringers was initiated, and he remained on ad lib diet. On hospital day (HD) 3, sodium acetate was added to fluids due to worsening metabolic acidosis (bicarbonate 8, see figure). On HD 5, diarrhea and acidosis resolved, and IVF were discontinued. On HD 8, acidosis returned and persisted. Differential diagnosis was reconsidered (see table) and additional studies were sent. Arterial blood gas revealed methemoglobinemia of 30.8.

Discussion: Methemoglobinemia is a rare condition that occurs when the ferrous iron of heme is oxidized to the ferric state, where it is unable to bind oxygen. The remaining ferrous hemes have stronger oxygen affinity, resulting in a shift in the oxygen dissociation curve to the left, decreased oxygen delivery to tissue, and resulting anaerobic and lactic acidosis. Methylene blue decreases the level of methemoglobin by reducing oxidized iron back to the ferrous state, allowing increased oxygen delivery and resolution of acidosis. Methemoglobinemia can be congenital or acquired. The etiology of acquired methemoglobinemia can be endogenous (diarrhea, infection, systemic acidosis) or exogenous (local anesthetics, nitrates). Infants, especially those small for gestational age, may be at increased risk for methemoglobinemia due to lower levels of cytochrome-B5 reductase, which acts in reduction of methemoglobin. In addition, it is easier to oxidize fetal hemoglobin when compared to adult hemoglobin, putting infants less than 9 months at even higher risk.

Conclusion: Methemoglobinemia can be a serious, potentially fatal complication of infection in infants. Our patient was transferred to the PICU and given one dose of methylene blue; methemoglobin and bicarbonate levels normalized over 48 hours (see figure). Clinical features of methemoglobinemia include acidosis that does not respond or frequently rebounds despite fluid resuscitation, and/or cyanosis and hypoxia that does not respond to supplemental oxygen. Other symptoms include irritability, tachycardia, lethargy, and seizures. Rarely, if methemoglobinemia level reaches >70%, death can occur. Methemoglobinemia should be considered when an infant's clinical appearance and degree of acidosis are out of proportion to their history of illness, and when other common diagnoses have been ruled out.
Resources:

**Laboratory Trends**

- **Bicarbonate (mmol/L)**
- **Met Hgb (%)**

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<th>Hospital Day</th>
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**Differential diagnosis of persistent metabolic acidosis**

<table>
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<tr>
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<th>Supporting or conflicting evidence in our patient</th>
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<tr>
<td>Diarrhea</td>
<td>Acidosis continued beyond resolution of symptoms</td>
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<td>Renal Tubular Acidosis</td>
<td>Urine anion gap -78, consistent with diarrheal causes</td>
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<tr>
<td>Inborn Errors of Metabolism</td>
<td>Normal newborn screen</td>
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<tr>
<td>Methemoglobinemia</td>
<td>Level 30.8 on co-oximetry</td>
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Resources


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B136
Title: Trampoline Tachycardia

Authors: Nora Pfaff, MD, Children's Hospital Los Angeles
Kevin Waloff, MD, Children's Hospital Los Angeles/ University of Southern California

Case Presentation: A 13 year old boy with recent URI symptoms presented to the emergency department for syncope after jumping on a trampoline and waking up with chest pain. Workup in the ED was significant for signs of ischemia: rising troponin and EKG showing ST depression in the lateral leads. In context of his URI symptoms, he was diagnosed with myocarditis and was admitted to a tertiary care children’s hospital.

The next day his troponin trended down, but he was found to have prolonged QTc of about 550 with a pattern concerning for LQTS1 (See Figure 1). Prior findings on EKG were absent and the patient’s initial syncopal event was thought to have been due to LQTS. However, echo that same day showed an aberrant left coronary artery arising from the right sinus of Valsalva with an intramural course. This was thought to be the actual cause for the ischemic event, which later led to an acquired prolonged QTc. Cardiac MRI confirmed the diagnosis, and he was taken to the OR for surgical repair due to risk of sudden cardiac death (See Figure 2).

Discussion: The incidence of abnormal origins of coronary arteries is low—less than 1% of births and noted in 0.17% of echocardiogram reports of asymptomatic pediatric patients. The most common variant is a coronary artery arising from the sinus of Valsalva, which is the type found in our patient. The clinical presentation includes chest pain or syncope with exertion, but sudden death can occur. Diagnosis without any signs of ischemia is difficult as physical exam and echocardiogram of asymptomatic patients is typically not revealing.

In our patient, who presented initially with ST segment changes and elevated troponins, it was easy to recognize that he had myocardial ischemia. However, the differential is still somewhat broad and two different potential etiologies were assumed before the correct diagnosis was discovered—bringing to light how nuanced the differences can be between the different etiologies.

Conclusion: The causes of syncope in adolescents are varied ranging from benign diagnoses such as vasovagal syncope to serious cardiac conditions like the ones we considered in our case. It is important to consider all possible causes of cardiac ischemia in young adults, even when prolonged QTc interval is seen on EKG. Prolonged QTc can be a finding that occurs in early transmural ischemia rather than the cause as it was in our case. However, although this interval prolongation may not signify that the patient has congenital LQTS, it does have clinical relevance in the predisposition to development of fatal ventricular tachycardia.

Abnormal coronary origins are a very rare but serious cause of syncope that can also present as sudden cardiac death.

Resources:
Figure 1. The patient’s EKG showing prolonged QTc on the day after admission.

Figure 2. Transesophageal echocardiogram and cardiac MRI demonstrating abnormal origin of the left coronary artery from the right sinus of Valsalva.

References:
Title: A pain in the neck: cervical lymphadenopathy that won’t go away
Authors: Jennifer Lee, MD, New York Presbyterian Morgan Stanley Children's Hospital of New York
Case Presentation: A previously healthy, fully immunized 3 year old girl presents with left submandibular swelling for one month. She was initially treated with Augmentin with no improvement. One month later she returned with worsening swelling. She had intermittent fever and URI symptoms but review of systems was otherwise negative. Socioeconomic risk factors for infection include living in a shelter and having close contact with a parent who is HIV-positive. Exam was remarkable for a 4 cm left submandibular indurated mass that was nontender and minimally mobile without warmth or erythema. Ultrasound showed a conglomeration of markedly enlarged lymph nodes. The child had a normal CBC, mildly elevated CRP, negative Quantiferon and PPD, negative respiratory viral panel, negative HIV, and no evidence of EBV/CMV infections. CXR was normal. CT scan of the neck showed a heterogeneously enhancing soft tissue mass with necrosis. Core biopsy yielded results suspicious for Rosai Dorfman disease (RDD), a diagnosis that had not been previously considered. Subsequent excisional biopsy confirmed the diagnosis.
Discussion: Chronic cervical lymphadenopathy has diverse etiologies, including nontuberculous Mycobacteria, lymphoma and RDD. RDD, or sinus histiocytosis with massive lymphadenopathy, is a disorder characterized by overproduction and accumulation of histiocytes. It is rare with a prevalence of 1:200,000. Incidence is higher in young adults though cases have been reported from ages 1 to 74 years. Younger children are most frequently affected by purely nodal RDD. Disease onset is gradual, with usually 3-6 months between initial presentation and diagnosis. The diagnosis is based on histological findings, including emperipolesis and specific immunostaining. RDD is typically self-limiting and benign but recurrence and dissemination can occur. In our patient, the greatest challenge was choosing a biopsy method that would balance diagnostic yield with procedural risks. In discussion with ENT, interventional radiology, and multiple subspecialists, core biopsy was done initially. When results were consistent with RDD and lymphoma and infection were ruled out, excisional biopsy was performed.
Conclusion: Cervical lymphadenopathy occurs frequently in children. Close follow up is necessary to ensure symptoms respond to treatments for the most common causes. Though rare, the histiocytoses should be considered in the differential diagnosis and pediatricians should be aware of these entities as they can mimic infectious and oncologic causes of cervical lymphadenopathy. Early discussion of these rarer causes of cervical lymphadenopathy with patients and families can help to better set expectations and improve patient satisfaction. Awareness of this rare disorder is also important in guiding multidisciplinary efforts to determine the best approach to obtaining tissue for biopsy, such as fine needle aspiration, core biopsy, or open biopsy. Our patient benefited from a stepwise and collaborative approach to care.
Resources:
Title: A Teenager Requiring Hospitalization For Croup?

Authors: Paula Soung, MD, Medical College of Wisconsin

Case Presentation: A 16 yo previously healthy female presented with difficulty breathing. Symptoms began 2 weeks prior with rhinorrhea, congestion, and cough. Patient diagnosed with sinusitis and started antibiotic course prior to presentation. Over the last few days, patient has had more difficulty with breathing, inability to talk, and congestion. Patient developed difficulty swallowing and now is frequently spitting her secretions out. Denies throat pain and fever. She feels like she cannot take a deep breath. In ED, CXR and XR lateral neck was normal. Given racemic epi x1 and decadron. Patient had desat to 81% and decision made to admit for "croup". On admission, patient on 4LPM by oxymask. Exam notable for breathy phonation, unable to elicit cough, progressive respiratory distress with diminished aeration, with no visible oropharyngeal abnormalities. On neurologic exam, mild R ptosis, impaired EOM, mild peripheral extremity weakness, and decreased 1+ reflexes. ENT performed bedside scope confirming vocal cord paresis with aspiration. Patient transferred to ICU and electively intubated.

Discussion: Many conditions may cause acute respiratory distress in children. The underlying etiology, while commonly within the respiratory system, may lie within other systems and should be considered when a patient does not fit the typical clinical picture. A detailed history and physical exam can help point to the cause. Acute neuromuscular weakness may result in dysfunction of respiratory muscles or inability to protect the airway causing respiratory compromise. For this patient, evaluation after admission included normal MRI brain and spine as well as normal CSF studies. EMG showed evidence of polyneuropathy. Patient received 5 days of IVIG for treatment of Miller Fisher syndrome/pharyngeal-cervical-brachial variant of Guillain-Barre syndrome with improvement in neurologic status. Post discharge, antibody testing returned positive for anti-GQ1b supporting the diagnosis.

Conclusion: The etiologies for acute respiratory distress is expansive with many masqueraders. When acute neurologic deficits are associated with respiratory distress, emergent evaluation is needed. The pharyngeal-cervical-brachial and Miller Fisher syndrome variants of Guillain-Barré syndrome are uncommon in children. Miller Fisher syndrome is characterized by external ophthalmoplegia, ataxia, and muscle weakness with areflexia. The pharyngeal-cervical-brachial variant of GBS is characterized by acute weakness of the oropharyngeal, neck, and shoulder, with swallowing...
dysfunction. This variant of GBS may overlap with Miller Fisher syndrome. Detection of serum IgG antibodies to GQ1b, support the diagnosis of the GBS variants Miller Fisher syndrome, and pharyngeal-cervical brachial variant. Close respiratory monitoring is necessary and risk of respiratory failure is a concern in all patients with Guillain-Barre. However, the variants which have cranial nerve involvement may present with respiratory complications more acutely than the typical Guillain-Barre syndrome with ascending weakness.

Resources:

References

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B139

Title: An Eye Opener: Klebsiella Neonatal Conjunctivitis in a Term Infant
Authors: Mary Terrell, MD, University of North Carolina Department of Pediatrics
Jacob Lohr, MD, University of North Carolina Department of Pediatrics

Case Presentation: A two-day-old term male in the newborn nursery presented with unilateral purulent eye discharge. Vaginal delivery of the infant was uncomplicated without prolonged rupture of membranes, and ocular erythromycin was applied. Mother received adequate prophylaxis for GBS+ status. Standard prenatal labs were otherwise unremarkable. Notably, gonorrhea and chlamydia were negative in both the first and third trimesters and there was no history of HSV.
The neonate was vigorous on exam with normal vital signs for age. The left eye was edematous without spontaneous opening and profuse green discharge was projectile on palpation. The left conjunctiva was markedly injected. An eye swab was sent for bacterial, AFB and fungal cultures as well as N. gonorrhea, C. trachomatis, and HSV PCR. The culture resulted positive for Klebsiella pneumoniae resistant to ampicillin; remaining studies were negative. Blood and urine cultures were obtained and resulted negative. IV cefotaxime was administered prior to discharge on cefdinir and topical tobramycin. Complete resolution was noted on follow up.

Discussion: Neonatal conjunctivitis (NC) is one of the most common conditions of the neonatal period. In addition to ophthalmologic complications including loss of vision, systemic infection can occur.
Neonates with infectious NC are thought to acquire the pathogens vertically regardless of mode of delivery. Ocular prophylaxis at birth has reduced Neisseria gonorrhea NC worldwide, but Chlamydia trachomatis remains prevalent. Other pathogens implicated in NC, though less studied, abound and appear to vary geographically. Klebsiella pneumoniae are Gram negative bacteria found in GI and vaginal flora, and are known surface water contaminants. Though rarely reported in healthy newborns, they are a common cause of hospital-acquired conjunctivitis in the NICU. B-lactam and carbapenem resistance in isolates is an increasing problem and susceptibility testing is important for guiding treatment.
To our knowledge, this is the second reported case of Klebsiella NC presenting in a healthy newborn. The first case occurred locally, which prompts consideration of a geographical predilection.

Conclusion: This case illustrates an unusual pathogen for neonatal conjunctivitis (NC) in a healthy term infant in the newborn nursery. This case reinforces the need to consider Klebsiella pneumoniae and other Gram negative organisms whenever NC is suspected, especially when the maternal history does not suggest Neisseria gonorrhea or Chlamydia trachomatis. While history and timeline of symptoms are helpful in diagnosis, sending a culture with susceptibilities and PCR when available is imperative. It remains important to include empiric gonococcal and chlamydial coverage while awaiting cultures. Given potential for systemic infection, IV antibiotics, at least initially, should be considered for this high-risk population.
Resources:


Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B140

Title: Down in the Dumps: A Case of Persistent Hypoglycemia

Authors: Andrew Wang, DO, University of Texas at Austin Dell Medical School Pediatric Residency
Jorge Ganem, MD, University of Texas at Austin Dell Medical School

Case Presentation: Patient is a 13-month-old girl with a complex medical history brought to the emergency department for recurrent low blood glucose. She was born with bilateral vocal cord paralysis requiring tracheostomy and
gastrostomy with Nissen fundoplication and was diagnosed with epilepsy at four months of age. She was admitted to the hospital one-month prior due to hypoglycemic seizures. After extensive and normal Endocrinology workup, the hypoglycemia resolved and the patient was discharged with a working diagnosis of ketotic hypoglycemia. On day of presentation, home glucose checks were 40mg/dl at home and 44mg/dl without ketosis in the ED. Glucose levels normalized with IV dextrose. Hypoglycemia recurred when bolus enteral feeds were restarted and IV dextrose weaned. Endocrinology workup was repeated and a comprehensive metabolic workup was completed. Results all normal. Persistent hypoglycemia resolved after careful transition to slow, continuous enteral formula feeds and slow withdrawal of IV dextrose. A diagnosis of dumping syndrome was reached after workup excluded other possibilities.

**Discussion:** The differential diagnosis of hypoglycemia is broad. Diagnoses initially considered were prolonged fasting periods with ketotic hypoglycemia; hyperinsulinemia, exogenous hypoglycemic agents, hypothalamic-pituitary-adrenal axis derangements, hyperthyroidism, and inborn errors of metabolism. While hospitalized, the patient had persistent hypoglycemia despite appropriate, supervised nutritional intake of bolus formula feeds. Critical labs taken while the serum glucose was 42mg/dl revealed normal lactate, pyruvate, beta-hydroxy-butyrate, cortisol, insulin, c-peptide and growth hormone. Serum amino acid panel, acyl-carnitine profile and urine organic acid levels were obtained and normal. Furthermore, fructose deficiency testing and ACTH stimulation test were normal with appropriate TSH and FT4 levels. During an episode of acute hypoglycemia, the patient lost vascular access and received a bolus of apple juice via her gastrostomy tube leading to an even lower drop in blood glucose. Noting this and excluding other possible diagnoses, dumping syndrome became a more likely diagnosis.

**Conclusion:** Dumping syndrome is more commonly seen in adults, but has been documented in children who have undergone Nissen fundoplication (NF) though the pathophysiology is not entirely clear. Current understanding in medical literature is that a NF may decrease the size of the stomach fundus accelerating gastric emptying. Unusually large osmotic loads entering the small intestine then stimulates release of vasoactive intestinal peptide, which stimulates the release of insulin, leading to hypoglycemia. Our patient had her first onset of symptomatic hypoglycemia about 12 months post NF. Diagnosis of dumping syndrome can be delayed up to 27 months post NF in some published studies. Since many patients have cognitive deficiencies or are non-verbal, the diagnosis may be under-recognized. Our patient has maintained normal glucose levels with continuous gastric feeds without further episodes of hypoglycemia. Hypoglycemia secondary to dumping syndrome is a diagnosis of exclusion. A thorough endocrinology and metabolic workup, and a keen clinical observation helped us reach this diagnosis.

**Resources:**

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B141

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**Title:** Mediastinal mystery: a rare cause of respiratory distress

**Authors:** Kathryn Kyler, MD, Children's Mercy Hospital
Molly Krager, MD, Children's Mercy Hospital

**Case Presentation:** A healthy 8 month old boy presented to the ER in with stridor and hypoxia, which quickly progressed to respiratory failure requiring intubation. CT of his neck/chest showed a large complex prevertebral fluid collection extending from base of skull to carina with mass effect on pharyngeal airway. IR placed a drain, which produced purulent fluid. He was initially treated with broad spectrum antibiotics, then narrowed to ampicillin when cultures grew GAS. His drain was removed 2 days prior to discharge and he was transitioned to oral amoxicillin for a planned 3 week course. The next day, he returned to the ER with respiratory distress. CT showed reaccumulation of the fluid collection.
He underwent repeat drain placement, which produced chylous fluid (TG 1,406) with negative cultures. Lymphangiogram showed connection between the thoracic duct and fluid collection, confirming diagnosis of thoracic duct cyst. He was placed on a low fat diet. He ultimately underwent sclerotherapy using doxycycline. He was advanced to regular diet without reaccumulation of fluid and drain was removed.

**Discussion:** Thoracic duct cysts are a very rare diagnosis in general. When they do occur, it is most often in the neck region rather than the mediastinum. There are few case reports of mediastinal thoracic duct cysts published only on adults. The pathogenesis of thoracic duct cysts is thought to be either congenital or degenerative. They are benign lesions that typically have a good prognosis following excision or sclerotherapy. This patient’s initial presentation of an infected posterior mediastinal/retropharyngeal fluid collection seemed most consistent with a large retropharyngeal abscess, especially considering the fluid culture was positive for GAS. Once he re-presented with similar symptoms and a different fluid analysis, the differential was broadened and eventually his ultimate diagnosis became more clear.

**Conclusion:** This case serves as a good reminder to keep a broad differential diagnosis, especially when admitting a patient with a common disease process with atypical features. In this patient, the remarkable size and extent of the fluid collection was unusual for a retropharyngeal abscess. The possibility of an underlying congenital malformation was discussed during the initial admission and a plan was made for follow up imaging; however, the rapid reaccumulation of fluid was unexpected. The final diagnosis in this case of a thoracic duct cyst is extremely rare in children.

**Resources:**

CT neck/chest images (4):
Citations:

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B142
Title: Pancreatitis, hematuria and abdominal pain: Only time will tell.

Authors: Marcos Mestre, MD, Nicklaus Children's Hospital
Vaishali Dayalan, MD, Nicklaus Children's Hospital, Justin Jeter, MD, Nicklaus Childrens Hospital

Case Presentation: A 4 year old girl presented with emesis and lower back pain. Her urine was tea colored with blood clots. She denied any fevers. A CBC, BMP and coagulation profile were normal. A UA showed moderate leukocyte esterase with 210 RBC/46 WBC. An abdominal US showed bowel wall thickening with mesenteric lymphadenopathy and a renal US showed left urothelial thickening. Bacterial and viral urine cultures were negative. Complements including C3 and C4 were normal. She then developed worsening abdominal pain with recurrence of emesis and an amylase and lipase were elevated. An US of the pancreas showed a swollen pancreas and US of the gallbladder showed no obstructing stone. MRCP demonstrated focal pancreatitis and edema of the duodenum. The mural thickening of the duodenum was suggested to be secondary to vasculitis. That evening, the patient developed palpable purpura on the lower extremities with significant joint pain. She was started on steroids with significant improvement. Skin biopsy was consistent with HSP with leukocytoclastic infiltrates with IgA deposition on immunofluorescence.

Discussion: Henoch-Schönlein Purpura (HSP) is the most common form of systemic vasculitis in children. It is usually a limited vasculitis which presents between 3 and 15 yrs of age. Most cases of HSP are preceded by an URI but other types of infections, vaccinations and insect bites have been implicated as possible triggers. As per the EULAR/PRINTO/PRES classification, diagnostic criteria for HSP include a mandatory purpura or petechiae with lower limb predominance without thrombocytopenia or coagulopathy and with any of the following additional symptoms: arthritis/arthralgia (acute), abdominal pain (diffuse, acute), renal disease (proteinuria/hematuria) or leukocytoclastic vasculitis or proliferative glomerulonephritis with predominant IgA deposition. The most common GI manifestations of HSP include mild nausea, vomiting, abdominal pain and transient paralytic ileus to more significant presentations including GI bleeding, intussusception, bowel ischemia, and necrosis. Rare episodes of HSP are associated with pancreatitis, gallbladder involvement and protein losing enteropathy.

Conclusion: This is a case of HSP with an atypical presentation. Our patient presented with abdominal pain due to pancreatitis as one of the initial manifestations along with hematuria. These symptoms preceded the classic purpuric rash. A search of the literature showed that pancreatitis associated with HSP has rarely been reported. The reported cases are mostly in adults and for those involving children, none occurred in the Unites States (China4, Turkey5, England6, Israel7). We believe this is the first reported case in the US where a child had pancreatitis as a presenting feature of HSP. The mechanism of the pancreatitis was thought to be due to the mural thickening of the duodenum secondary to the vasculitis which then caused an obstruction of the ampulla with secondary pancreatitis. The initial hematuria is also unusual as a presenting manifestation of HSP. On follow up, the child had not developed any renal involvement after being treated with a steroid taper. In cases such as these which are a diagnostic challenge, time will be your friend in leading to a diagnosis.

Resources:

References

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B143
Title: Salmonella Septic Joint Suggests Underlying Immunodeficiency
Authors: Unikora Yang, MD, Children's Hospital Los Angeles, Keck School of Medicine
Ronen Zipkin, MD, Division of Hospital Medicine, Children's Hospital Los Angeles, Keck School of Medicine
Case Presentation: A 2yo boy presented with refusal to bear weight, pain over left malleolus, and intermittent fever. One month prior, he was found to be pancytopenic. A bone marrow biopsy had 3% B-cell lymphoblasts but given recovery of cell counts and a positive EBV serology, he was diagnosed with acquired aplastic anemia and discharged. Labs were: WBC 8, normal ANC, Hgb 10.7, platelets 232, CRP 4.8, ESR 74. He was started on clindamycin and cefazolin, switched to ampicillin after a joint aspirate grew pan-susceptible Salmonella non-typhi. Further questioning revealed a pet turtle. Despite antibiotics, he remained febrile and developed diffuse bone pain and neutropenia. Repeat MRI showed osteomyelitis of the left tibia and right wrist with marrow enhancement. Antibiotics were broadened to ceftriaxone. Repeat bone marrow biopsy had 8% aberrant B-cell lymphoblasts, which was repeated two weeks later and had 61% lymphoblasts, consistent with ALL. Induction chemotherapy was initiated and well tolerated.
Discussion: In the setting of invasive Salmonella infection, immunocompromised states must be considered. There are only three cases of pediatric leukemia and Salmonella septic arthritis in the literature.(1-3)
Conclusion: We present a case of Salmonella septic arthritis and osteomyelitis in the setting of an evolving ALL. Nontyphoid Salmonella is an uncommon cause of septic arthritis and osteomyelitis in the setting of an evolving ALL. Non-typhoid Salmonella is an uncommon cause of septic arthritis and osteomyelitis in the setting of possible immunocompromised state, including sickle cell disease and cancer. ALL has a varied initial presentation. Transient bone marrow aplasia is a rare prodrome, occurring in 1-2% of cases.(4,5) The first biopsy did not meet diagnostic criteria, but the small population of lymphoblasts from that biopsy was able to proliferate clonally within a few months. Patients with Salmonella infection may present with leukopenia or neutropenia, potentially delaying the diagnosis of leukemia.
Resources:
References
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B144

Title: The Proof is in the Pudding
Authors: Alanna Boyajian, MD, Children's Hospital of Pittsburgh
Sylvia Choi, MD, FAAP, Children's Hospital of Pittsburgh
Case Presentation: A 15-year-old male with a history of autism was admitted for management of severe anemia, found during work-up for progressive weakness, pallor, and mood changes. He had extreme food selectivity, consuming only whole milk and butterscotch pudding. He was a thin, globally delayed male with decreased strength and hyperreflexia. Labs revealed a hemoglobin of 4.7, low MCV, low total iron, low iron saturation, high TIBC, high transferrin, normal reticulocytes, and mild neutropenia. He had normal B1, B3, B9, B12, zinc, methylmalonic acid, and homocysteine levels. Levels of vitamin C, D, E, and K were low. He was transfused and received IV iron. A gastrostomy tube was recommended but not pursued. Despite outpatient iron supplementation, his hemoglobin remained around 9 with mild neutropenia. He also suffered neuroregression, becoming non-ambulatory and incontinent. Further work-up by neurology and genetics found low copper and ceruloplasmin levels. After copper repletion, his anemia and neutropenia resolved, and he has regained some neurologic function with aggressive rehabilitation.
Discussion: Copper is an essential trace element and required as a cofactor in many enzymatic reactions. Its deficiency can cause reversible anemia, leukopenia with neutropenia, and neurodegenerative symptoms. Patients can present with myelopathies and peripheral neuropathies. It is rare to see copper deficiency from diet alone as an average diet usually
provides sufficient supplies. Our patient’s severely selective diet caused him to develop significant nutritional deficiencies. While his initial presentation seemed consistent with iron deficiency anemia, his persistent hematologic and progressive neurologic abnormalities were ultimately felt to be secondary to copper deficiency. His hematologic derangements responded well to supplementation with both iron and copper. Unfortunately, although he has regained some neurologic function, the neurologic manifestations of copper deficiency are often irreversible.

**Conclusion:** When treating iron deficiency anemia, it is important to evaluate the response to iron therapy, because persistently low hemoglobin levels despite therapy may indicate occult diagnoses. Severe copper deficiency is uncommon and can mimic more common diseases, including iron deficiency anemia. It is important to identify and treat copper deficiency to prevent neurologic deterioration. We hope that this case will help clinicians continue to consider other causes anemia until a clear diagnosis of iron deficiency is confirmed by response to treatment.

**Resources:** N/A

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B145

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**Title:** Treating with Tea: Severe Hyperphosphatemia and Hypocalcemia in an Infant  
**Authors:** Tyler Severance, MD, Riley Hospital for Children  
Melinda Chen, MD, Riley Hospital for Children  

**Case Presentation:** A 6-week-old Hispanic infant presents to the ED with worsening NBN emesis and diarrhea for 24 hours. Patient also had increased fussiness, looser stools, and tactile temperatures. Due to poor PO, mom gave two servings of tea (3oz Chamomile, 3oz homegrown spearmint). As patient did not improve, mom brought him to the ED. In the ED, the patient was stable and tolerating PO. Physical exam was unremarkable. Due to recent tea ingestion, a BMP, Mag, and Phos were obtained showing Ca of 4.3 and Phos of 21.7. Lab dilution confirmed sample. Additional workup was sent and an EKG showed a QTc of 507. Calcium Gluconate, Calcitriol, and oral Calcium Carbonate were given and patient was admitted. Recheck in 6 hours revealed a Phosphorus of 8.4 and a Calcium of 4.3. Of note, the repeat labs were sent after receiving fluid resuscitation, but no further treatment.

The hospital course was otherwise unremarkable with normalization of Ca and urine electrolytes. Follow up PTH and 25 OH Vitamin D were appropriate, while the 1, 25 OH Vitamin D was elevated.

**Discussion:** The etiology of hypocalcemia and hyperphosphatemia typically centers on abnormalities of the Parathyroid axis. At this age, several etiologies should be considered. These include genetic causes such as primary hyperparathyroidism or DiGeorge Syndrome causing parathyroid defects, as well as low magnesium impairing PTH secretion. However, in the absence of physical exam findings or further laboratory abnormalities, these etiologies were less likely. Additional consideration was made for an external phosphate load, which would bind and reduce serum calcium. With appropriate levels of Vitamin D labs, and urine studies indicative of markedly elevated urine phosphate excretion, the diagnosis of exogenous phosphorus administration was made. This was further supported by the abrupt resolution of both the serum phosphorus concentration and urine phosphorus excretion with only fluid resuscitation. The true underlying source was not found, although the tea samples were considered the most likely cause.

**Conclusion:** Pediatric hospitalists should be vigilant in patients who receive supplemental therapies at home. In addition, when such supplementation is suspected – particularly in infants - a rapid workup should be performed, as underlying electrolyte abnormalities can prove significant. In this case, hypocalcemia was manifesting with cardiac rhythm disturbances requiring immediate intervention.

**Resources:** N/A

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B146

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**Title:** It’s all in her head  
**Authors:** John Darby, M.D., Baylor College of Medicine  
Lauren Hess, MD, Baylor College of Medicine  

**Case Presentation:** A 2-year-old F presents to the ER for poor weight gain. At 8 months of life, the patient was noted to stop gaining weight despite normal appetite and diet. The child’s mother denied any preceding illness, injury, or change in eating habits and there was no vomiting, diarrhea, fevers, fatigue, or recurrent infections. She was born term (wt 2.7
kg 8th%) and had normal growth velocity until 8 months. Her past medical history was significant for a parasitic infection treated with an unknown medication at 18 months of age.

In the ER, her weight was 7.6kg (<1%) and the physical exam revealed muscle wasting, cachexia and quivering lateral eye movements with near focus (video available). Initial labs including chemistry, LFTs, CBC, ESR/CRP, TFTs, amylase, lipase, ferritin and uric acid were all normal. The patient underwent an inpatient calorie count and demonstrated adequate intake but insufficient weight gain.

Due to severe failure to thrive and nystagmus a brain MRI was obtained which revealed a hypothalamic tumor confirming a diagnosis of diencephalic syndrome.

**Discussion:** Failure to thrive is a condition in which energy intake is insufficient for energy expenditure. It can have many etiologies including: inadequate intake, inadequate appetite, poor absorption, increased losses or increased metabolic demand. A rare but well described cause of failure to thrive is diencephalic syndrome. The syndrome is characterized by poor weight gain, often associated with normal linear growth, increased or normal appetite, and hyperactivity secondary to an intracranial tumor often located in the 3rd ventricle. Patients with diencephalic syndrome can present with signs of intracranial pressure but often have no neurological symptoms. In one case series, 55% of patient presented with nystagmus and 68% with vomiting.

The mechanism for severe growth disturbances in diencephalic syndrome is still unclear but studies have revealed elevated growth hormone levels suggesting acquired partial growth hormone resistance.

**Conclusion:** Although diencephalic syndrome is a rare cause of failure to thrive it should be considered in patients with preserved linear growth and normal intake who present with new neurological abnormalities such as nystagmus.

**Resources:**

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*Figure showing growth charts and tables from CDC.*
References:

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B147

Title: 14 year old with headache and syncope: Neurologic problem or broken heart?

Authors: Kimmy Hart, DO, Sanford School of Medicine
Alex Schaller, DO, Le Bonheur, William Waltz, MD, PhD, FAAP, FACC, Sanford School of Medicine

Case Presentation: A 14yo previously healthy female developed stiff neck, back pain and fatigue after a softball tournament. The next day she had a fever and a brief syncopal event. Initial WBC was 21000 and she was given course of azithromycin and a dose of ceftriaxone two days later. During the following 2 weeks she had daily fevers and developed bilateral conjunctivitis. Labs at that time included WBC 12000, CRP 163, and blood culture drawn which were negative. On exam she was non-toxic appearing with bilaterally injected eyes, tender neck musculature but no meningeal signs, and had a low pitched 2/6 systolic ejection murmur. Echocardiogram significant for a quadricuspid aortic valve with a 7mm vegetation and she was started on vancomycin, ceftriaxone, and gentamicin with improvement in symptoms. Further history revealed that one day prior to symptoms developing she had a routine dental cleaning. Treatment included 2 weeks IV gentamicin and 8 weeks of ceftriaxone. Vegetation was shorter and thinner on echocardiogram at 8 weeks and at 6 months had resolved.

Discussion: Infectious endocarditis (IE) is a bacterial infection found on the inner lining of the heart, the endocardium. Patients with abnormal cardiac anatomy, especially artificial heart valves or disruption of the native tissue are at risk. The incidence is 0.05-0.12 cases per 10002. Dental procedures continue to be the most common precipitating event leading to recommendation that antibiotic prophylaxis prior to dental procedures be provided for those with highest risk.

The most common organisms are Staphylococcus aureus, Enterococcus and streptococci viridans. IE typically presents with fevers, malaise, arthralgias and leads to cardiac arrhythmias, new murmurs and congestive heart failure. Physical findings such as splinter hemorrhages and Osler nodes are rarely present in children. Negative blood cultures are seen in 8%-36% of clinically diagnosed IE; often due to recent antibiotics or infection with bacterium that do not grow well in vitro. The frequency of quadricuspid aortic valves (QAV) is less than 0.001%. There are adult case studies identifying an association with IE and QAV.

Conclusion: This case illustrates the importance of a broad differential, optimizing diagnostic modalities, and providing appropriate interventions. Analyzing and reviewing this case is important because it identifies two rare diagnoses. Had an echocardiogram not been obtained, the patient may have been inadequately treated and developed increased risk for complications such as cardiac dysfunction or emboli from the vegetation. The antibiotics without an identified...
illness at the beginning of her course, might have contributed to the negative blood cultures making the diagnosis and treatment more difficult. This is the first pediatric case in the literature of endocarditis involving a quadricuspid aortic valve. This anomaly is not specifically addressed in the current guidelines, but in this case screening for family members was offered. Given her history of endocarditis, she will require antibiotic prophylaxis and the importance of good dental hygiene was stressed; however, the role of prophylaxis in a patient with a quadricuspid aortic valve without history of IE is unclear at this time.

Resources:


Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B148

Title: A 17 Year Old with Altered Mental Status: All Worked Up?
Authors: Maxine Ames, MD, Jefferson/duPont Hospital for Children Josephine Elia, MD, DuPont Hospital for Children, Hunter Wernick, DO, Jefferson/duPont Hospital for Children, George Datto, MD, DuPont Hospital for Children

Case Presentation: A 17 year old male with history of neonatal HSV infection with resultant cortical blindness, and developmental delay presented with altered mental status (AMS) for 1 month. The patient had had 3 inpatient evaluations for AMS since the onset. The patient had significant regression from his baseline, was having slowing of his movements and speech and had decreased responsiveness. At his first admission, MRI brain and LP were normal; HSV PCR and autoimmune encephalitis evaluation were negative. EEGs at each admission were normal. On presentation his skin was notably greasy; he was minimally interactive, had muscle rigidity, facial grimacing, manneristic movements of his hands, and catalepsy (keeping his arms elevated and torso raised off of the bed). CBC, CMP, UA and UDS were
normal. Triggered by the catalepsy and rigidity on exam, particularly in the context of an inconclusive and extensive prior evaluation, catatonia emerged as the leading diagnosis and treatment with lorazepam was initiated. The patient significantly improved by hospital day 3 and was back to baseline at discharge.

**Discussion:** Catatonia represents a state of unresponsiveness when a patient appears to be awake. There are a number of symptoms which characterize catatonia, including for example grimacing, waxy flexibility and mutism; three or more are required to formally confirm the diagnosis. Physical exam findings of rapid pulse, excessive oiliness of skin, or drooling are also often identified. Catatonia may be idiopathic or result from a variety of medical, neurologic or psychiatric conditions. It is often missed or attributed to other conditions such as schizophrenia or status epilepticus. This case highlights the importance of keeping catatonia in the differential for AMS because treatment is often effective. In this patient with a significant neurologic history, catatonia was not initially considered.

**Conclusion:** Catatonia is significantly better characterized in the adult literature. It is not rare and is increasingly being recognized in children. Once identified, catatonia can usually be successfully treated with benzodiazepines, whereas antipsychotics and other medications may exacerbate it. Catatonia, if untreated, may have serious complications. Although pediatric data is scarce, in adults it is associated with substantial morbidity and mortality. Providers must consider catatonia as a cause of change in mental status, regardless of medical, neurological or neuropsychiatric presentation, particularly so when a neurologic evaluation is otherwise unrevealing.

**Resources:**


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B149

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**Title:** A Breathtaking Case

**Authors:** Kristin Schneider, MD, Cincinnati Children's Hospital Medical Center
Traci Hail, APRN, Cincinnati Children's Hospital Medical Center, Shivani Patel, DO, Cincinnati Children's Hospital Medical Center

**Case Presentation:** A previously healthy 16 year old male was seen for cough. He was seen by his PCP 1.5 weeks prior at the onset and had 5 days of azithromycin without improvement. He was again seen by his PCP with worsening cough, shortness of breath, tactile fevers, and chest pain where he had decreased aeration over his right upper lobe, oxygen saturations in the 80s, heart rate to 120, and a respiratory rate of 20. He was sent to the ED where his saturations improved with nasal cannula oxygen. A chest x-ray showed an opacity in the right upper lobe and he was admitted for treatment of presumed community acquired pneumonia. Over 24 hours he improved and was discharged home. One day later he returned to the ED after acutely developing worsening difficulty breathing and cough. In the ED he was tachypneic, tachycardic, and unable to lie flat due to shortness of breath. He had decreased breath sounds at the right upper lobe and a normal blood pressure. A chest x-ray was unchanged. A CTA of his chest showed right upper lobe opacities suggestive of infection or inflammation and multiple pulmonary emboli.

**Discussion:** Much of the diagnostic and treatment information about pediatric pulmonary embolism is extrapolated from adult studies due to the rare occurrence in pediatrics. Studies show an estimated incidence of 0.14-0.9 per 100,000 children in the community and 8.6-57 per 100,00 hospitalized children. While an uncommon pediatric diagnosis, pulmonary embolism should remain in the differential, especially when patients deviate from the expected course of their presumed diagnosis. When a first time diagnosis of PE is made, evaluation for underlying disorders should be done.

**Conclusion:** Pulmonary embolism is often clinically silent or presents with symptoms similar to other diseases. In teenagers, the most common symptom is pleuritic chest pain. Pulmonary angiography is the gold standard for diagnosis; tests such as spiral CT are becoming the first choice because they can be performed quickly, visualize emboli, and identify other intrathoracic disorders. Treatment of PE in children is guided by clinical condition. Those with unstable hemodynamics are candidates for thrombolysis while hemodynamically stable patients receive anticoagulation. In children with PE, idiopathic thrombi are uncommon; the majority of patients had an underlying risk factor (such as a
central line, thrombophilic abnormality, or nephrotic syndrome). Our patient was started on a heparin drip and admitted to the PICU. He underwent an echo which had no evidence of heart strain. Further studies showed a non-occlusive thrombus within the infrahepatic IVC with extension into both renal veins. Additional work up revealed a diagnosis of idiopathic membranous glomerulonephritis.

**Resources:**

**References**


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B150

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**Title:** A Small Problem

**Authors:** Anisha Mazloom, MD, Stanford Pediatrics Residency Program
Nivedita Srinivas, MD, Stanford University School of Medicine

**Case Presentation:** An 8 month old ex-term female was admitted for evaluation of a BRUE. She had a prior BRUE at 1 week of life characterized by apnea and unresponsiveness. Prenatal ultrasound and cardiac evaluation were reportedly negative. She was well until the day of admission when she had 3 episodes of NBNB emesis after a feed, followed by 30 seconds of apnea and unresponsiveness. By ED arrival she was at neurologic baseline and fed without difficulty. Vital signs were normal but weight was <1st percentile for age. Physical exam was unremarkable aside from her thin appearance. CBC, CMP, UA, and EKG were normal. A CXR was preliminarily read as normal (Image 1). She was admitted to the pediatric hospitalist service for observation but had no further events. However, the radiologist called the following morning to report an abnormality on the initial CXR, prompting a 2-view study that was suggestive of a diaphragmatic hernia (Images 2,3). CT chest and abdomen confirmed an anterior left diaphragmatic defect with hiatal and Morgagni hernias. She underwent laparoscopic repair and did well subsequently.

**Discussion:** Consider congenital diaphragmatic hernia in children presenting with multiple unexplained BRUEs, poor feeding, and a history of failure to thrive. Late presentation is uncommon, but should still be considered even in children who have had a normal prenatal ultrasound.

**Conclusion:** The repeated BRUEs and FTT in this case were suspicious for underlying pathology, despite a normal prenatal ultrasound and a prior negative BRUE evaluation. Ultimately, the key to this patient’s diagnosis was a chest x-ray which was initially read as normal. The incidence of late presenting CDH has been reported to be 5-25% and generally carries a good prognosis [1]. Left posterolateral hernias are the most common hernia presenting outside the neonatal period with a 2:1 male to female ratio [1]. Anterior congenital diaphragmatic hernia through the foramen of Morgagni is particularly rare, making up 3-5% of all types of congenital diaphragmatic hernias [2]. Morgagni hernias have been known to remain asymptomatic until adulthood, and often present with non-specific symptoms including recurrent chest infections, gastrointestinal symptoms, and failure to thrive [2,3]. On CXR, Morgagni hernias may appear as a fatty mass in the right cardiophrenic angle or displaced curvilinear omental vessels may be visible [4]. Laparoscopic repair is considered to be safe and effective in children [2].

**Resources:**

**References**

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B151

Title: A Proctored Exam
Authors: Jennifer Fuchs, MD, University of Texas Southwestern

Case Presentation: A 14 year old girl presents with 2 weeks of watery diarrhea, crampy abdominal pain and vomiting. Prior ER visit for fever of 103.4F was notable for elevated inflammatory markers and hepatomegaly and small bowel enteritis on abdominal CT.

Exam reveals a thin, dehydrated patient with diffusely tender abdomen, normal rectal and external genitourinary exam with unintentional 10 pound weight loss over 2 weeks. Labwork shows ESR 93, CRP 4.6, hemoglobin 10.9, and albumin 2.9 with negative stool blood.

Given concern for Crohn’s disease, she has an endoscopy, colonoscopy and MR enterography that show no small bowel inflammation. Symptoms persist and she insists on wearing diapers due to stool leakage, urgency and tenesmus. Despite initial denial of sexual activity, she later admits to genital contact. Full gynecologic exam reveals left adnexal tenderness and urine gonorrhea and chlamydia result as positive. Final diagnosis is made: proctitis from gonorrhea/chlamydia with abdominal pain from pelvic inflammatory disease (PID) and hepatomegaly due to Fitz-Hugh Curtis syndrome.

Discussion: “-” Sexually transmitted gastrointestinal syndromes include proctitis, proctocolitis, and enteritis. “-” Proctitis can be caused by N. gonorrhoeae, C. trachomatis, T. pallidum, or herpes simplex virus and involves inflammation of the distal 10-12 cm of the rectum, causing anorectal pain, tenesmus, or rectal discharge. Proctocolitis can be caused by C. trachomatis and involves both rectal and colonic inflammation, leading to diarrhea, abdominal cramping, and positive fecal leukocytes. “-” Recent studies show up to 25% of chlamydia cases and 19% of gonorrhea cases in woman are rectal-only infections. “-” Standard therapy includes one dose of intramuscular ceftriaxone plus one week of doxycycline therapy. Patients with herpes proctitis are treated like those with genital herpes. “-”PID is a bacterial infection of the upper parts of the female reproductive system. Recent CDC guidelines recommend 2 weeks of metronidazole therapy (in addition to traditional ceftriaxone IM dose and 2 weeks of doxycycline) for better anaerobic coverage and bacterial vaginosis treatment.

Conclusion: “-” Although anemia, hypoalbuminemia, and elevated inflammatory markers are classic findings of IBD, symptoms are typically more long-standing than 2 weeks. “-” Sexually transmitted diseases may present with gastrointestinal findings or symptoms and should be considered in the differential diagnosis for diarrhea and crampy abdominal pain in any sexually active patient. “-” Currently, the CDC only recommends routine STI rectal screening for sexually active men who have sex with other men. However, rectal swabs with nucleic acid amplification tests should be performed for detection in select cases given significant rates of rectal-only infection.

Resources: N/A

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B152

Title: Anorexia - an uncommon presentation for vaginal agenesis
Authors: Rachel Miller, DO, East Tennessee State University
Fazila Zaidi, MD, East Tennessee State University, Nathaniel Justice, MD, East Tennessee State University

Case Presentation: An 11 y.o. female is admitted with four months of abdominal pain and weight loss. She experiences early satiety. Abdominal pain, nausea and emesis occur with meals. Food and drink cause similar symptoms, and she has restricted intake to avoid discomfort. She is premenarchal, but had an episode of spotting four months ago.

On exam, she is frail-appearing with a palpable mass in the left lower quadrant. A normal introitus with an SMR of IV is noted. Laboratory work-up for common etiologies is unrevealing. An ultrasound reveals an enlarged uterus with debris and a hemorrhagic cyst in the left adnexa. An MRI confirms hematometrium and left-sided hematosalpinx; the vaginal canal is not distended.

These findings yield a diagnosis of vaginal agenesis. The patient’s symptoms are attributed to the mass effect caused by the uterine retention of menstrual products. In consultation with a pediatric gynecologist, she is referred for
percutaneous drainage of these collections. Leuprolide is started to suppress menses to prevent the reaccumulation of menstrual products while awaiting surgery.

**Discussion:** Vaginal agenesis is the second most common cause of primary amenorrhea. It results from in utero failure of differentiation of the Müllerian duct. Affected individuals have normal growth and development of secondary sexual characteristics. They commonly present with primary amenorrhea or sexual dysfunction, and examination reveals an absent vagina or a short, blind-ended pouch. Most individuals have a remnant of the Müllerian duct that lacks endometrial tissue.

This case introduces a unique presentation for vaginal agenesis. Though individuals with functional endometrium may experience episodic pain, the typical Müllerian remnant is too small to produce a mass effect that causes anorexia. This patient’s weight loss was so anomalous that further work-up to exclude an ovarian tumor was recommended by her gynecologist. This patient’s reproductive tract has also differentiated to a greater extent than is typically seen. While interventions for vaginal agenesis are usually aimed at restoring sexual function, her gynecologist is hopeful for an intervention that may allow reproduction.

**Conclusion:** Abdominal pain and weight loss are frequently encountered complaints in the care of adolescents. While gastrointestinal pathologies, endocrinopathies and eating disorders are commonly considered, this case illustrates that gynecologic disorders must also be considered in a female with secondary sexual characteristics. The key to this patient’s diagnosis was the discordance between her sexual maturity rating and absence of menarche.

**Resources:**

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B153

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**Title:** Ending a Resident’s Perception: Edification by Enterovirus

**Authors:** Vanessa McFadden, MD, PhD, Medical College of Wisconsin
Alyssa Stephany, MD, Medical College of Wisconsin, Danita Hahn, MD, Medical College of Wisconsin

**Case Presentation:** 6 day old full term male presented with one day of fever and irritability. Birth history also significant for maternal fever during delivery (with sick contact just prior to delivery) and 48 hours of antibiotic therapy prior to discharge from the nursery. Exam only significant for fever and irritability. Given concern for meningitis, workup included blood, urine and cerebrospinal fluid (CSF) cultures along with herpes simplex virus (HSV), influenza and enterovirus testing. Urinalysis was reassuring. CSF studies demonstrated no organisms on gram stain, elevated protein (234 mg/dL), normal glucose; total nucleated and red blood cells could not be determined due to specimen clotting. Initial blood work demonstrated thrombocytopenia (47,000/uL), mildly elevated transaminases (AST 160 iU/L and ALT 61 iU/L), and mildly elevated prothrombin time (16.2 s). Empiric cefotaxime, ampicillin and acyclovir were started. Irritability improved, however, he required multiple platelet transfusions (nadir of 14,000/uL). At 48 hours, blood, urine and CSF cultures along with HSV studies were negative.

**Discussion:** Enterovirus PCR was positive from CSF, nasopharyngeal, and rectal samples, and antimicrobials were discontinued. On day 3 of hospitalization, he rapidly deteriorated developing hypoxia then signs of shock. He required intubation and significant inotropic support including milrinone due to cardiac involvement with moderately diminished left ventricle systolic function and dilated left atrium on echocardiogram. The constellation of maternal fever during delivery, age at presentation, persistent thrombocytopenia, hepatitis, coagulopathy, and development of heart failure from acute myocarditis established the diagnosis of vertically transmitted enterovirus. Due to severity of condition and potential benefits, he was given 2 doses of intravenous immunoglobulin (IVIG). Ultimately, respiratory and inotropic support were weaned off, enterovirus viral loads down trended, cardiac function improved with ejection fraction of 43% prior to discharge, and follow up outpatient echocardiogram had normal ventricular function.

**Conclusion:** Differential for an ill appearing febrile neonate includes more than bacterial or HSV infection. Recognize the spectrum of neonatal enterovirus disease. Hospitalists often encounter enterovirus infections, which are usually relatively innocuous, but keen clinical acumen and a high degree of suspicion is needed to detect disseminated vertically transmitted enterovirus that can be life-threatening. And although, no specific treatment exists, awareness through anticipation of complications leads to timely supportive care at facilities with appropriate level resources to potentially minimize adverse outcomes. Neonatal enterovirus infection, specifically when vertically transmitted, has a high risk for
morbidity and mortality. Enterovirus in the newborn can cause a range of disease with severe disease consisting of pneumonia, hepatitis, coagulopathy, myocarditis, meningitis and sepsis. Frequent symptoms include fever, irritability, anorexia and rash. Severe disease is associated with maternal illness prior to or at delivery, illness onset within the first few days of life and prematurity.

Resources:
References:

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B154

Title: Pulmonary Histoplasmosis causing Cardiac Tamponade
Authors: Ann-Marie Tantoco, MD, Nationwide Children's Hospital
Case Presentation: A 16 year old Liberian male presented with acute chest pain and hypotension. He was recently hospitalized for pleuritic chest pain at another hospital and was found to have calcified mediastinal adenopathy. No diagnosis was made and since symptoms improved, he was discharged with a scheduled outpatient bronchoscopy. He moved from Liberia 4 years ago. He previously received the BCG vaccine and was healthy. He was afebrile with BP 84/44, HR 101, RR 16, and 99% O2 saturation on room air. Pulsus paradoxus was present with 10-12 mmHg respiratory variation. He was somnolent and had distant heart sounds without murmur. Lungs were clear. TTE showed a large circumferential pericardial effusion. Emergent pericardiocentesis removed 900 ml of fluid. BP returned to normal. Fluid testing was undiagnostic. Results from his former hospitalization showed serum detection of Histoplasma antibody ID M band and Histoplasma antibody CF titer of 1:128. He was given amphotericin B for 7 days then itraconazole with colchicine and ibuprofen. No effusion was present when seen in follow up two months later.
Discussion: This illustrates a case of pulmonary histoplasmosis causing pericarditis with a large inflammatory pericardial effusion resulting in cardiac tamponade. Pericarditis is caused by an inflammatory response to Histoplasma capsulatum since pericardial fluid is usually culture negative. Patients can recover with anti-inflammatory medications alone such as nonsteroidal anti-inflammatory agents or steroids. In hemodynamic instability occurs, the pericardial fluid must be drained.
Conclusion: Pulmonary histoplasmosis should be considered in those who present with mediastinal lymphadenopathy and pericarditis. An inflammatory reaction by the fungus can result in a large, culture negative pericardial effusion.
Resources:
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B155

Title: Salmonella isn’t Hip: Osteomyelitis in a Healthy Adolescent Female
Authors: James Odum, MD, Children's Mercy Hospital
Kayla Heller, MD, CMH
Case Presentation: 12 yo female presents with acute-onset severe back and left leg pain to an ED. Initial laboratory work-up showed WBC of 9.21 x 103 mcL, CRP < 0.5 mg/dL, and ESR of 22 mm/hr. Plain films of the lumbar spine and hips were normal. MRI of hip did not show any evidence of trauma, infection, or inflammation. She had mild limp on gait assessment and pain with active left hip movement. Her neurological exam was normal. She was admitted for pain management and serial exams.
Patient was started on scheduled IV pain medications. During the evening of HD 1, she began having fevers. UA demonstrated possible urinary tract infection (UTI), so she was started on IV ceftriaxone. On HD 3, she had repeat hip MRI which revealed evidence of left sacroiliitis with surrounding osteomyelitis. Her blood culture from HD2 was positive for Salmonella species, Group D. Her urine culture grew multiple flora.
Hospital course was also complicated by septic shock requiring transfer to the PICU for fluid resuscitation. In total, she completed 6 weeks of antibiotics for Salmonella bacteremia and osteomyelitis.
Discussion: Salmonella species cause osteomyelitis infrequently in an immunocompetent patient. In resource poor areas, retrospective studies have found a strong association with underlying an immunodeficiency or linked to malaria transmission secondary to damaged intestinal epithelium. Our patient was tested for an underlying immunodeficiency per NIH recommendations, and results returned negative. Our patient’s initial presentation was suggestive of possible osteomyelitis, but her laboratory and radiographic studies did not initially support this diagnosis. She presented many clinical conundrums throughout her course, including normal initial MRI of hip, negative blood cultures at presentation, development of septic shock after receiving five doses of appropriate antibiotics, and culture proven bacteremia of a non-classical organism in an immunocompetent patient. Diagnosis was achieved thorough serial exams, trending of laboratory and radiographic studies, and communication with a patient who was old enough to articulate the progression of her symptoms.

Conclusion: Acute changes in a child should warrant thoughtful consideration and evaluation. The importance of a detailed history and physical examination was outlined in this case. This patient was originally considered suitable for discharge with routine follow-up from the ER, but her symptoms did not align with a benign diagnosis. The benefit of inpatient admission is that it allows for serial examinations and further discussions that may elicit important details. Often, time and attention to detail will lead to a better approach to evaluation and management in a non-critically ill patient.

Although Salmonella osteomyelitis is not an association we memorize for healthy immunocompetent children, this case demonstrates the need to keep a broad differential when clinical conundrums present themselves.

Resources:
Copy of article: Unusual Manifestations of Histoplasmosis in Childhood Geoffrey A. Weinberg, MD, Martin B. Kleiman, MD, Jay L. Grosfeld, MD, Thomas R. Weber, MD, and Lawrence J. Wheat, MD
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B156

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Title: What's up with the Hematoma?
Authors: Nicole Washington, MD, The Children's Hospital of Philadelphia

Case Presentation: A 6 year-old healthy male presented with 3 days of abdominal pain and NBNB emesis. Review of systems was negative for fever, trauma, dysuria, or hematuria. Vital signs were normal, except for an elevated BP of 129/69. His exam was normal, except for mild periumbilical/RLQ abdominal tenderness. Labs were as follows: WBC 20, Hgb 12.2, Platelets 325, CRP 18.4, normal BMP and normal coagulation studies. RLQ US: appendix not visualized, but right kidney appeared markedly abnormal. Follow-up abdominal CT scan showed a normal appendix and a large subcapsular hematoma surrounding the right kidney. Consulted Nephrology who suspected trauma, tumor, or bleeding diathesis. Bleeding diathesis was unlikely in the setting of normal platelet count/coagulation studies. There was no evidence of non-accidental or accidental trauma. MRI showed a right hemorrhagic renal mass surrounded by large subcapsular hematoma. Patient underwent right total nephrectomy (renal capsule intact). Pathology confirmed Wilms Tumor. He completed post-operative chemotherapy with no major complications.

Discussion: The most common presentation of Wilms tumor is an asymptomatic abdominal mass in a healthy child less than 4-5 years old. Our patient was slightly older, presented with abdominal pain and a subcapsular hematoma, illustrating an atypical presentation of Wilms tumor. While specimens of renal malignancy can demonstrate hemorrhage, spontaneous formation of a subcapsular hematoma from renal tumor is rarely seen. There are only a few case reports in the literature, mostly in adults and mostly associated with Wilms tumor. Diagnosis and treatment are critical in Wilms tumor, especially in this case. Hemorrhage can progress rapidly and extend into the peritoneal cavity leading to tumor cell spillage. This complication changes the stage of disease and alters the patient’s prognosis. In our case, the tumor was resected in time and no such complication occurred; underscoring the importance of knowing and recognizing the association of renal subcapsular hematoma with Wilms tumor.

Conclusion: Early diagnosis and treatment are critical in Wilms tumor, especially in this case. Hemorrhage can progress rapidly and extend into the peritoneal cavity leading to tumor cell spillage. This complication changes the stage of disease and alters the patient’s prognosis. In our case, the tumor was resected in time and no such complication occurred; underscoring the importance of knowing and recognizing the association of renal subcapsular hematoma with Wilms tumor.

Resources:
Copy of Article
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B157
Title: Disseminated Neisseria Gonorrhea Imitating Lupus in an 18 Year Old Female  
Authors: Avideh Rashed, Doctor of Medicine, Upstate Golisano Children's Hospital  
Case Presentation: 18 year old sexually active female with history (hx) of systemic lupus erythematosus (SLE) non-compliant with medication presented with a 2 day hx of bilateral (b/l) leg pain, right (R) elbow pain, and fevers. Patient (pt) was seen in the ED immediately prior to admission with similar symptoms (sx). Given these sx and response to prednisone pt was diagnosed with SLE flare (SF). On current admission pt was febrile and tachycardioc. Physical exam (PE) revealed b/l knee swelling with effusion, limited extension of R elbow, and ulcers on the tongue consistent with vasculitis. Labs revealed normal C3 and C4 complement levels and elevated ESR and CRP. Rheumatology recommended treating with Solu-Medrol for likely SF. Hospital day (HD) 1 pt continued to spike fevers without improvement on PE. Solu-Medrol was stopped, blood cultures (BC) were drawn, and pt was started on Ceftriaxone and Vancomycin for possible sepsis. On HD 3 BC grew N. gonorrhea. Pt was only continued on Ceftriaxone and given one dose of Azithromycin for concomitant C. trachomatis. Sx improved and repeat BC was negative.

Discussion: Here we report a case of disseminated gonococcal infection (DGI) in a poorly compliant sexually active teenage patient with a past medical history of SLE. Given the patient’s similar signs and symptoms, negative infectious work up on prior ED visits, and previous response to steroids the most likely diagnosis was a lupus flare. Upon further work-up the patient was correctly diagnosed with DGI. There have been other case reports that describe the tendency of SLE patients to develop DGI; however, the susceptibility to N. gonorrhea was due to complement deficiencies in those patients [1,2]. What makes this case unique is that the patient was not immunocompromised as she was not compliant with her SLE medications and had normal C3,C4 complement levels at the time of dissemination. These findings and patients presenting symptoms made this case especially difficult to diagnose as DGI. Thus it is always important to consider the sexual history of patients with SLE and to always do an STD panel in sexually active patients with SLE.

Conclusion: It is always difficult to distinguish SLE flares from infectious etiologies, as SLE is known to be the “great imitator” however, in sexually active teenage patients with SLE it is especially important to always consider disseminated Neisseria gonorrhea in the differential as it can mimic a SLE flare.

Resources:
References

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B158

Title: Gastric pneumatosis and portal venous gas: Suspect etiology in adolescent  
Authors: Manju Korattiyil, MD, Children's National Health System  
Bridget Huysman, MPH, George Washington University School of Medicine and Health Sciences, Sonal Kalburgi, DO, MSHS, Children's National Health System  
Case Presentation: A 17 year old African American male presented to an outside hospital after 1 day of crampy abdominal pain, distention, nausea, diarrhea, and nonbloody, nonbilious emesis, without fever. On exam, he was non-toxic appearing with diffuse pain greatest at the RLQ, and voluntary guarding. Labs showed normal CBC and CMP and elevated CRP. Abdominal CT with contrast revealed a dilated stomach with gastric pneumatosis, swirling of mesenteric vessels, and hepatic portal venous gas concerning for midgut volvulus. Appendix appeared normal, and lymphadenopathy was noted.

Upon transfer, Pediatric Surgery recommended broad spectrum antibiotics. He remained NPO and an NG tube was placed to suction. UGI study confirmed no evidence of obstruction or structural anomaly. He was treated supportively with intravenous fluids, proton pump inhibitor, bowel rest, and pain control. Abdominal distention and pain improved, and his diet was advanced over the next 72 hours. Blood and stool cultures subsequently remained negative. Stool studies were positive for norovirus, rotavirus, and adenovirus.

Discussion: Gastric pneumatosis is an uncommon radiographic finding in adolescents. Three proposed physiological theories include: 1)gastric emphysema from increased intraluminal pressure from mucosal injury; 2) emphysematous...
gastritis from bacterial gas production by direct invasion; and 3) pulmonary gas from extravasation of intrathoracic gas. Hepatic portal venous gas is proposed to occur similarly.

In our patient’s case, the etiology for gastric pneumatosis and portal venous gas was likely the identified pathogen norovirus. Rotavirus is thought to only disturb the intestinal lining and not the gastric mucosa, resulting in pneumatosis intestinalis. Adenovirus is also a less likely etiology, as asymptomatic carriage in the stool can be positive for up to 18 months. Norovirus causes mucosal injury via blunting and broadening of villi, intracellular edema, and apoptosis of enterocytes. Delayed gastric emptying and inflammation of the pyloric junction are thought to induce emesis, resulting in increased intraluminal pressure in the setting of mucosal permeability leading to gastric emphysema.

**Conclusion:** Our case describes a rare adolescent case of gastric pneumatosis with hepatic portal venous gas, most likely due to viral etiology that was successfully treated with medical management. This has not been previously described in the literature for this age group. It is likely this was a benign finding on imaging that became evident due to early detection and better imaging modalities, and not necessarily representing the same life threatening prognosis as pneumatosis intestinalis in necrotizing enterocolitis.

**Resources:**

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B159

**Title:** Headache with a Hint of Lyme: A Juicy Case of Pediatric Meningitis  
**Authors:** Alyssa Stephany, MD, Medical College of Wisconsin  
Bailey Hutchison, BS, Medical College of Wisconsin, Rachel Weigert, MD, Medical College of Wisconsin  
**Case Presentation:** 7-year-old female presented in July with two days of fever and neck pain to a hospital in Wisconsin. Family endorsed two weeks of morning headaches (partially relieved with ibuprofen), occasional emesis, but no vision changes, rash, sick contacts, travel, head trauma, or known tick exposure. Exam revealed nuchal rigidity; no papilledema/neurologic deficits. CBC revealed thrombocytopenia. Lumbar puncture (LP) showed increased protein (111 mg/dL), normal glucose, 9 RBCs and 2518 WBCs (98% neutrophil). Vancomycin, ceftriaxone, and doxycycline were started. CSF culture and studies for enterovirus, HSV, and CMV were negative. More testing ensued -- brain MRI negative; Ehrlichia, Anaplasma, and Babesia negative; repeat LP with elevated opening pressure, negative BioFire, but improving WBCs/protein and negative cytology. Serum Lyme antibodies were positive (2.07 ISR). Confirmatory immunoblot was anti-Lyme IgM positive (two of three bands) but IgG negative (one of ten bands), suggesting early disseminated Lyme. Thus ceftriaxone was continued with complete symptom resolution.
Discussion: In this case, initial suspicion was high for enterovirus, a common summer meningitis agent. Immediate diagnosis was also confounded by thrombocytopenia and CSF findings that are not typically those found in Lyme meningitis. When enterovirus and other viral etiologies were ruled out, the differential was expanded, but due to the endemic area, season, and the "risk of disease," Lyme meningitis remained a potential etiology, so Lyme-directed therapy was continued. Follow-up testing showed IgG reactivity to only one borrelial protein, without IgM reactivity, indicating probable recent B. burgdorferi infection per CDC criteria. This was further supported by complete symptom resolution after 14 days of Lyme-directed therapy.

Conclusion: Hospitalists in Lyme-endemic areas must keep Lyme in their differential for meningitis when there is pleocytosis and intracranial hypertension, even with lack of rash, known tick exposure, or classic CSF findings. Literature even suggests that like our patient, Lyme can present with only persistent headaches and elevated opening pressure. While serologic testing can indicated probable Lyme, sometimes these results cannot establish a diagnosis alone if the results are only suggestive and not conclusive, as in our patient. Thus, hospitalists must remain vigilant regarding the presentation for Lyme, remember to consider it in the differential, and treat for it when it is suspected.

Resources: N/A

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B160

Title: It can't always be bronchiolitis: A 2-week-old infant with tachypnea
Authors: Eric Zwemer, MD, University of North Carolina School of Medicine
Laura Parente, MD, MPH, University of North Carolina Children's Hospital, Suresh Nagappan, MD, MSPH, Moses Cone Hospital, University of North Carolina Children's Hospital

Case Presentation: A 2-week-old girl presented to the ED with 3 days of worsening tachypnea. Parents denied fever, cough, rhinorrhea, or feeding difficulty. History notable for normal prenatal ultrasounds, birth at 40w1d, normal pulse oximetry screening, and appropriate regaining of birth weight. Her pediatrician had evaluated tachypnea twice and started anti-reflux medicine. ED workup included normal CXR and negative RSV test and she was admitted for presumed bronchiolitis. Exam showed an afebrile infant with HR 150, RR 66, BP 90/67, and O2 saturation 100%. She was in mild respiratory distress with supraclavicular retractions, clear lungs, II/VI systolic ejection murmur at the LSB, and palpable femoral pulses. ECHO showed VSD with inadequate visualization of aortic arch. Right upper and lower extremity BPs were 113/68 and 70/58 respectively. Pre/post ductal saturations were 100%/99%. Repeat ECHO showed juxtaductal coarctation of aorta with moderately decreased left ventricular systolic function and no patent ductus arteriosus (PDA). She underwent immediate surgical repair with discharge 3 days later.

Discussion: Coarctation of the aorta (CoA) is the fifth most common congenital cardiac lesion but typically does not cause problems in utero due to bypass of coarctation by PDA. Newborns with CoA may initially be asymptomatic but start to manifest symptoms at 2-5 days of life with PDA closure. Initial manifestations include murmur, decreased/absent femoral pulses, tachypnea, and feeding difficulties. Varying degrees of heart failure can occur based on severity of coarctation, and patients can quickly deteriorate with shock and circulatory collapse. Untreated CoA has significant early mortality, but surgical repair after early diagnosis has excellent outcomes. Screening methods include prenatal ultrasound, newborn physical exam, and pulse oximetry screening, but these methods miss many infants with CoA, as was the case for this patient. Pulse oximetry in particular can miss duct-dependent lesions as it is typically performed before duct closure, and neonates with CoA are at the highest risk of being missed. Studies show over half of neonates with CoA are discharged undiagnosed from the nursery.

Conclusion: Pediatricians should recognize that CoA is the cardiac lesion most likely to be missed by initial newborn exam and pulse oximetry screening. Careful physical exam including cardiac exam, palpation of femoral pulses, and four-extremity blood pressure measurement remain fundamental in diagnosis. This is true not just in the nursery, but also in the pediatrician's office at follow up visits since these infants often present later as the PDA closes. CoA should be considered in infants 1-2 weeks of age who present with tachypnea or murmur even without more overt signs of heart failure.

Resources:
References:
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B161

Title: Why won't this heal?
Authors: Ilyssa Scheinbach, Hospitalist Physician/DO, Maimonides Infants and Children's Hospital
Christina Gagliardo, Pediatric Infectious Disease Physician/MD, Maimonides Infants and Children's Hospital
Case Presentation: A 14 year old girl presented with a non-healing wound on the lateral aspect of her lower leg. It began 6 months prior as a small wound but became larger and ulcerated. She was seen by a dermatologist who administered triamcinolone injections and prescribed oral steroids for a diagnosis of pyoderma gangrenosum though no biopsy had

Figure 1: Echocardiogram of this patient showing juxta ductal coarctation of the aorta, with a peak pressure gradient by Doppler of 35 mmHg in the descending aorta.

Figure 2: Time of diagnosis for individual cardiac malformations, ranked according to probability of pre-discharge diagnosis. From Wren, et al.
been performed. Given lack of improvement, she was referred to pediatric surgery for excision. Social history revealed she moved from China 2 years prior and had been living in Brooklyn, New York. Physical exam was significant only for a 3 by 2 centimeter ulcerated lesion (Fig 1). She underwent surgical excision and debridement (Fig 2). Tissue was sent for pathology, bacterial and fungal cultures. Pathology showed an ulcerative lesion (Fig 3) and granulomas (Fig 4) with AFB-positive staining organisms (Fig 5). Pediatric Infectious Disease was consulted who requested a mycobacterial culture. Approximately 3 weeks later, mycobacteria growth indicator tube broth culture grew a mycobacterium species, later identified as Mycobacterium marinum.

Discussion: M. marinum is a salt and fresh water environmental bacterium that is found after abrasions or introduction of an open wound into environments containing the bacteria. Typical presentation involves skin lesions localized to the site of injury. Diagnosis is often dependent on a detailed exposure history as well as histology and smears of the lesion. Stains for AFB may be negative and the accuracy of culture is variable. Treatment is often started empirically if clinical suspicion is high given the difficulties in confirming the diagnosis. The isolation of M. marinum in our patient was fortuitous as she had no exposure risk. Of interest, around the same time she presented, the NYC Department of Health and Mental Hygiene released an alert describing an outbreak of M. marinum skin and soft tissue infections. The outbreak was in Chinese-speaking communities and all patients had handled live or raw fish and seafood, and infections were localized to patients’ hands and arms. While our patient resided in one of the Chinese-speaking communities, she denied any exposure.

Conclusion: While the puzzle regarding the cause of her non-healing wound was solved, the exact exposure to M. marinum remains a mystery. Clinicians should have a high index of suspicion for atypical mycobacterial infections when evaluating chronic, non-healing wounds.

Resources:
References:
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B162

Title: 9 year old with hyponatremia: Some side effects actually happen
Authors: Luz Castellanos, MD, Dell Children’s Medical Center of Central Texas
Sara Gibson, MD, Dell Children’s Medical Center of Central Texas, Kirsten Nieto, MD, Dell Children's Medical Center of Central Texas

Case Presentation: A 9 year-old male with severe allergies and asthma, on intranasal and medium dose inhaled corticosteroids, presented with one day of fever, headache, abdominal pain, emesis, and anuria. On exam, drowsy with low-grade fever, normal heart rate, normal blood pressure, and right lower quadrant tenderness. CBC and CSF labs normal, sodium 128 mmol/L, potassium 3.0 mmol/L, chloride 92 mmol/L, serum osmolality 270, urine osmolality 1066. Abdominal ultrasound normal. Received fluid bolus and started on D5NS +20KCl/100ml. Next morning, mental status worsened and progressed to generalized seizures. Stat labs revealed: sodium 122 mEq/L, S osmolality 253, cortisol 5.8 mcg/dL, aldosterone 3.3 ng/dL (nl 4-44), ACTH 23 pg/mL (nl). Stat CT with diffuse cerebral edema. Transferred to ICU and treated with mannitol until sodium corrected with hypertonic saline. High and low dose cosyntropin stimulation test showed inappropriate cortisol response. CT abdomen with atrophic adrenal glands. Diagnosed with adrenal insufficiency and started on hydrocortisone and fludrocortisone with return to baseline.
Discussion: Use of inhaled and intranasal corticosteroids is common in pediatric practices; however, the development of adrenal insufficiency from the systemic suppressive effects of inhaled corticosteroids is not often considered in the differential for a hyponatremic patient. Ahmet reviewed studies looking at adrenal suppression (AS) as a complication of inhaled corticosteroids and found more than 60 cases of AS with almost all children being treated with =500 µg/day of fluticasone, but AS was also seen with >1000 µg/day of beclomethasone and budesonide. Our patient’s total dose of fluticasone was 600ug/day. Donaldson et al published a case report two siblings who presented with acute hyponatremia and cerebral edema. Both used inhaled fluticasone in doses between 500-2000 µg/day for asthma and had adrenal suppression. The mechanism was believed to be low cortisol resulting in high ADH levels, more free water retention, and thus hyponatremia. Our patient’s hyponatremia was likely due to both aldosterone deficiency and elevated ADH levels in the setting of low cortisol.

Conclusion: While SIADH and acute hyponatremic dehydration from free water replacement of losses are more common etiologies, it is important to include AS in the differential for a patient with hyponatremia. In this child, primary adrenal insufficiency was also considered given symptoms of mineralocorticoid deficiency with hyponatremia, volume depletion, and salt cravings but this was ruled out. Asthma is the most common chronic condition of the young and is a major cause for admissions. Inhaled corticosteroids (ICSs) are the most efficacious preventive therapy for persistent asthma and have minimal systemic effects in low to medium doses. High or prolonged systemic absorption of ICS can result in hypothalamic-pituitary-adrenal axis suppression and decreased cortisol production in response to physiologic stress. Systemic absorption and likelihood of AS increases from use of ICSs with: greater oral bioavailability, larger particle size, suboptimal inhaler technique, absence of spacer, and younger age. A good history including detailed medication history is essential for proper diagnosis.

Resources: N/A
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B163

Title: A disappearing act – where did my blood vessels go?
Authors: Yu-Lin Lee, Internal Medicine-Pediatrics, MD, Duke University Hospital

Case Presentation: A healthy 4-year-old boy was incidentally found to be hypertensive during evaluation for a right inguinal hernia repair. Repeat outpatient visits continued to show elevated pressures (130-140/90s). Labs including CBC, electrolytes, BUN, Cr renin, aldosterone, plasma/urine catecholamines, and thyroid studies were normal. An echocardiogram showed trace aortic valve insufficiency. He was started on Lisinopril, but developed pressures as high as 170s/100s and was admitted. Otherwise, he has been asymptomatic. On admission, Lisinopril was initially held due to concern for renal artery stenosis; he was switched to Amlodipine. On exam, he had strong, palpable and symmetric radial, brachial, femoral and dorsalis pedis pulses and no abdominal bruits. An abdominal CT angiogram showed diminutive suprarenal aorta and absent infrarenal abdominal aorta below the L3 vertebral body, multiple enlarged abdominal wall and lumbar collaterals supplying the pelvis, diminutive renal arteries with the proximal right renal artery not visualized. These findings were suggestive of mid-aortic syndrome.

Discussion: Mid-aortic syndrome (MAS) is an uncommon clinical condition caused by segmental narrowing of the abdominal or distal descending thoracic aorta. Luminal compromise may vary from minimal to highly significant. MAS can be congenital (related to developmental anomalies in the maturation of the aorta) or acquired (secondary to neurofibromatosis, fibromuscular dysplasia, retroperitoneal fibrosis, mucopolysaccharidosis, giant cell arteritides or acquired insults in utero). MAS often manifests in early adulthood (average age of 20 years) with dizziness, palpitations, fatigability, headaches, blurred vision, abdominal pain, and intermittent claudication. In children, there is a delay in diagnosis because they are often asymptomatic, blood pressures are not frequently measured and high values are generally dismissed as inaccurate. The natural progression of MAS includes left ventricular hypertrophy, cardiomegaly, cerebrovascular accidents, coronary artery disease, myocardial infarction, heart failure, intracranial hemorrhage, aortic rupture, and renal function impairment/failure.

Conclusion: Mid-aortic syndrome is a rare cause of hypertension but is important because it can lead to early morbidity and mortality. While medical management with anti-hypertensives is the initial therapy, it is important to keep in mind that most patients ultimately need surgical intervention.

Resources:
References:

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B164

**Title:** Osteitis Pubis – A Rare Confounding Diagnosis in Young Athletes  
**Authors:** Kendra Mitchell, MD, Children's Hospital Los Angeles  
Lauren Rissman, MD, Children's Hospital Los Angeles  

**Case Presentation:** An active, previously healthy 12-year-old male presented with 1 month of progressively worsening groin pain. Laboratory studies were notable for elevated ESR and CRP, but with a normal WBC. MRI revealed bony inflammation, consistent with osteomyelitis and adductor myositis. Deep bone biopsy was performed and IV clindamycin was initiated to treat osteomyelitis while awaiting biopsy results. The patient showed no improvement on antibiotics; both his pain and elevated inflammatory markers remained unchanged. When the bone biopsy results returned with no neutrophils and with negative cultures, and repeat MRI revealed no change on antibiotics, we were forced to revisit our diagnosis. Further history revealed that the patient was not merely an active boy who liked sports, but an adolescent soccer player training for a national team. Given this new history of intense athleticism, and failure to improve on antibiotics, his diagnosis was changed from osteomyelitis to osteitis pubis. Antibiotics were discontinued, and he was discharged home on a regimen of rest and NSAIDs.

**Discussion:** Given a presentation of progressively worsening leg pain, elevated inflammatory markers, and bony inflammation on MRI, osteomyelitis is the most likely diagnosis; however, in an intensely athletic adolescent, a diagnosis of osteitis pubis should be considered.

Osteitis pubis is inflammation of the symphysis pubis and surrounding muscles due to exertional stress, causing hip pain and an elevation of inflammatory markers. Although a rare diagnosis, osteitis pubis is most commonly described in young male athletes who play soccer and football. Incidence has been described in up to 7% of the young athletic population. Diagnosis is based on history and physical. Classically, patients with osteitis pubis have limited unilateral or bilateral hip flexion due to pain. Patients may describe radiation to the abdomen or groin, as well. Laboratory data commonly reveal elevated inflammatory markers. Medical management includes rest, NSAIDs, and non-pharmacologic therapies such as massage.

**Conclusion:** 1) If osteomyelitis does not improve clinically or radiographically within 48 hours of initiating antibiotic therapy, the differential diagnosis should be expanded. 2) Osteitis pubis should be considered in athletes with hip or groin pain. 3) Osteitis pubis and osteomyelitis can be difficult to distinguish due to similar clinical, laboratory and radiographic findings. (See Image)

**Resources:**
Title: An Unusual Case of Persistent Hypernatremia
Authors: David Rayburn, MD, Indiana University
Daniel Hinds, MD, Indiana University, Department of Pediatrics, Andrew Shriner, MD, Indiana University, Department of Pediatrics
Case Presentation: A two and a half year old male with history of repaired cleft palate and double aortic arch was admitted to the PICU obtunded with a serum sodium of 172mmol/L. Prior to this, the patient had one day of URI symptoms and decreased PO intake without vomiting, diarrhea, or excessive sweating. Initial labs showed an anion gap metabolic acidosis, pre-renal AKI, and rhinovirus on RVP. After two days of IV fluids, his mental status improved and sodium levels trended down to 160mmol/L. Despite high sodium levels, he still had no interest in PO fluids. Persistent hypernatremia with aggressive IV hydration on the floor prompted Nephrology and Endocrinology consults. Testing showed an HPA axis, renal function, and CNS structure without pathology. A head MRI showed hypogenesis of the corpus callosum, though no pituitary abnormalities. Free water boluses through NG tube decreased his sodium to 135mmol/L over the next two days. To find his baseline sodium, NG fluids were discontinued and an oral fluid goal of 20 oz/day was set. His sodium slowly stabilized to 153mmol/L by discharge.
Discussion: Most commonly, hypernatremia occurs through a dysregulation of the body’s free water, manifesting through high serum sodium. Specifically in pediatrics, it is often due to an inability to replace GI water losses during an acute illness. Large baseline insensible losses through skin and an inability to communicate thirst predispose young children to hypernatremia. Typically, free water losses are corrected by increased posterior pituitary ADH, which decreases renal free water excretion. Additionally, active thirst centers increase desire for free water intake. For any case of hypernatremia, a broad differential must include free water loss in acute illness with inadequate intake, central and nephrogenic diabetes insipidus, dysfuction of renin-aldosterone pathways, excess salt intake, and hypodipsic hypernatremia. Correct diagnosis is of critical importance to avoid future occurrences and complications such as seizure, brain shrinkage, coma, and death. One must also be careful in treating hypernatremia since aggressive correction can lead to cerebral edema and death.
Conclusion: This case highlights the importance of a broad differential diagnosis when approaching cases of hypernatremia. Although very uncommon and with only a handful of case reports in the pediatric literature, the diagnosis of hypodipsic hypernatremia was made for this patient. After examination of potential irregularities in the HPA axis, urine concentration, and CNS lesions that could affect sodium regulation centers, no etiology could be found, posing a unique diagnostic dilemma. A history of disinterest in drinking fluids at baseline and continued poor PO fluid intake despite dangerously high serum sodium levels demonstrates the hypodipsic nature of the illness. Additionally, an offsetting of the patient’s osmotic regulation centers seen through high baseline sodium levels, predisposed the patient to dangerous increases in sodium with minor acute illnesses. This combination of factors led to PICU admission after only 24 hours of decreased PO intake and URI symptoms. For these patients, if a PO fluid goal fails to correct baseline hypernatremia, NG or G-tube placement may be indicated.
Resources:

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B165
Title: Do You Have An Eye for N. Meningitidis? Two Cases of Neonatal Conjunctivitis

Authors: Shira Pedan, MD, The Floating Hospital for Children at Tufts Medical Center
Yamini Sharma, MD, The Floating Hospital for Children at Tufts Medical Center, Dan Hale, MD, The Floating Hospital for Children at Tufts Medical Center

Case Presentation: Patient one is an 8 day old baby boy admitted with bilateral purulent eye discharge and inability to open his eyes due to eyelid swelling. His exam was otherwise unremarkable. After a full septic work-up including eye swabs for routine culture and Gonorrhea/Chlamydia, the patient was started on Cefotaxime IV, Azithromycin PO, Acyclovir IV, and Polytrim and Viroptic eye drops. His laboratory work-up revealed Neisseria meningitidis that grew on the routine eye culture. Infectious Disease and Ophthalmology were consulted. Cefotaxime alone was continued to complete a 7 day course with full resolution of symptoms.

Patient two is a 3 day old baby girl who developed mild bilateral eye discharge without eyelid swelling during her birth hospitalization. Her exam was otherwise unremarkable. A routine culture of the eye discharge was sent and revealed N. meningitidis. Prior to the result, she completed 4 days of erythromycin eye ointment with resolution of symptoms. Infectious disease was consulted. A CBC was negative and she given Cefotaxime until the blood culture was negative for 48 hours.

Discussion: Neonatal conjunctivitis due to N. meningitidis is extremely rare — exact incidence is unknown. Close contact with a meningococcal carrier (as in our cases) and ocular trauma are risk factors for primary meningococcal conjunctivitis (PMC). Prophylactic eye ointments given at birth may protect against conjunctivitis due to Chlamydia trachomatis and Neisseria gonorrhoeae (more common pathogens), but not against N. meningitidis. PMC typically manifests as purulent discharge analogous to N. gonorrhoeae. Corneal ulcers, keratitis, subconjunctival hemorrhage, and iritis are the commonest complications. The most feared complication is dissemination, with an associated mortality of 10-15% — more likely to occur if only topical medication is used. Thus, IV antibiotics is often administered though duration of therapy is not well established. According to the CDC, chemoprophylaxis is not recommended for close contacts of patients with N. meningitidis in non-sterile sites, like the conjunctiva. Yet, the mothers in our cases were treated due to the close proximity during routine care of infants.

Conclusion: Though acute conjunctivitis is a common pediatric diagnosis, there are few reported cases where N.meningitidis is the culprit, with even fewer as a cause of neonatal conjunctivitis. As seen in our patients, there can be a great degree of variability in presentation. Given the potential for serious eye complications as well as dissemination, it is important to establish an individualized plan of care for patients depending on the severity of symptom presentation. This may include consultation with Infectious Disease and Ophthalmology.

Resources:

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B167
Title: Horse, horse, zebra? When sepsis is really a rare tumor

Authors: Megan Derrer, MD, Children's Hospital of Illinois/University of Illinois College of Medicine at Peoria
Matthew Davis, MD, Children's Hospital of Illinois/University of Illinois College of Medicine at Peoria, Trina Croland, MD, FAAP, Children's Hospital of Illinois/University of Illinois College of Medicine at Peoria

Case Presentation: A previously healthy 27 day old, full term female presented to the ED for poor feeding and irritability. Parents noted fussiness with being held that progressed to intermittent inconsolable crying, as well as progressive loss of reflexes and tone, especially in her upper extremities. Mother was GBS positive but otherwise had a normal pregnancy. Significant findings on physical exam were fussiness with movement, dry mucus membranes, tachycardia without murmur, poor tone, lack of grasp, rooting, and Moro reflexes. Fontanelle and head circumference were normal. Meningitis was initially suspected; LP performed in the ED yielded little xanthochromic fluid before turning grossly bloody. CT head was negative. On admission, upper extremity weakness was felt to be worse than lower, and MRI of the head demonstrated abnormal signal and expansion of the cervical cord. Further MRI studies revealed an expansile heterogenous intramedullary mass from the lower medulla to T12 with areas of hemorrhage. Gross total resection was performed; pathology was consistent with grade 1 immature teratoma.

Discussion: Differential diagnosis was broad, including infection, trauma, inflammatory, and tumor. In this patient with no infectious symptoms, imaging should have been obtained prior to lumbar puncture in the ED. CT and MRI brain were obtained prior to repeating the lumbar puncture, which revealed bloody fluid and elevated protein. The patient went on to have full lab and imaging workup for malignancy after MRI spine, which was significant only for LDH 352 U/L (125-220). Alpha fetoprotein was measured after resection to assist in therapeutic decisions, and was markedly elevated at 113.1 ng/mL (0-8.8). Her post-op course was complicated by hypertension, which required nicardipine drip, but she otherwise did very well. She regained some spontaneous movement of her extremities. The decision was made to monitor with AFP, beta HCG, and total spine MRI based on literature search. AFP and beta HCG are now within normal limits for age, scans have been negative for residual disease, and she continues to regain movement with ongoing therapy.

Conclusion: At 27 days old, our patient is one of the youngest documented cases of an intramedullary immature teratoma. Although this type of tumor is very uncommon, it should remain on the list of differential diagnoses when dealing with neurological deficits in the absence of infectious symptoms.

Resources:
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B168

Title: Not your typical obstruction

Authors: Nancy Clemens, MD, Cincinnati Children’s Hospital Medical Center

Case Presentation: A 17 month old male with a history of severe constipation presented with one day history of fever and abdominal distention. He was febrile to 39.1°C and physical exam was significant for abdominal distention with diffuse tenderness to palpation but no peritoneal signs. He had no recent illness, no vomiting or diarrhea. Laboratory studies were notable for a white blood cell count of 26.5 mmol/L, hemoglobin of 8.8 gm/dl, and creatinine of 2.16 mg/dl. Urinalysis showed blood and large leukocyte esterase. Blood and urine cultures grew streptococcus mitis. The positive cultures prompted a renal ultrasound which demonstrated marked bilateral hydronephrosis, hydroureter, and bladder distention. Further history revealed a weak and dribbling urinary stream since birth. A voiding cystourethrogram showed posterior urethral valves (PUV).

The S. mitis urosepsis with treated with a 14 day course of antibiotics. His PUV were removed via endoscopic ablation. Six weeks after diagnosis, his creatinine improved to 0.84 mg/dl consistent with stage III chronic kidney disease.

Discussion: This patient presented with urosepsis and AKI superimposed on a chronic obstructive uropathy caused by PUV. Further history revealed that his constipation was diagnosed at 10 months of age with painful hard formed stools and overflow diarrhea, which had persisted despite treatment with miralax. This constipation was likely due to obstruction from bladder distention secondary to PUV.

PUV occur in 1 of 5,000 live male births, and are observed in 0.003% of fetal ultrasound screenings. About half of the cases are found on prenatal ultrasound. Outside of the neonatal period, the most common presenting symptoms are voiding complaints, such as straining to void, dysuria or polyuria, followed by UTI and failure to thrive. The gold standard for diagnosis is the voiding cystourethrogram (VCUG) which demonstrates a dilated and elongated posterior urethra. Treatment is endoscopic removal of the obstructing tissue. Even with timely surgery, upto 30% of patients develop chronic kidney disease, possibly due to morphologic bladder changes leading to ongoing dysfunctional emptying.

Conclusion: In male infants who have a significant history of abdominal distention and constipation, it is important to consider bladder distention as a source of distal obstruction. Clinicians should screen patients with severe or early onset constipation with questions about urinary habits and urinary stream. When there is concern for bladder obstruction, renal and bladder ultrasound is an appropriate initial study, which might demonstrate a thickened bladder wall, hydroureter, hydronephrosis or dysplastic kidneys. If posterior urethral valves are suspected, obtaining a VCUG and referral to pediatric urologist are appropriate next steps.

Given the ongoing dysfunctional emptying and risk for renal insufficiency in patients with PUV, a general pediatrician should closely monitor growth, blood pressure and voiding habits in these patients. Serum creatinine, urinalysis and renal ultrasound should be followed regularly. Families should be educated on avoiding nephrotoxic drugs, and followed by pediatric nephrologist when appropriate.

Resources:
Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B169

Title: Retropharyngeal Abscess Presenting With Hidden Lemierre’s Syndrome

Authors: Daniel Hershey, M.D., Rady Children’s Hospital
Nicole Herrick, BS, University of California, San Diego

Case Presentation: A 21 month-old female presented with 5 days of hoarseness and sore throat, and one day of progressive work of breathing, stridor, and agitation. She became unresponsive in the ED requiring CPR, epinephrine, and intubation. CT with contrast revealed retropharyngeal abscess (RPA), multifocal pneumonia, and bilateral pleural effusions. Chest tubes were placed, the pleural fluid grew C. albicans and S. anginosus. Repeat CT revealed a hypodensity in the right internal jugular (IJ) vein, confirmed via neck ultrasound to be a thrombosis. She slowly improved on ceftriaxone, clindamycin, and fluconazole. Heparin was started upon discovery of the clot, and she was discharged with 3 months of enoxaparin.

Over the next year, she continued to have infections requiring antibiotics, including eyelid abscess, oral thrush while off antibiotics, and infected seborrheic dermatitis. She was hospitalized for pneumonia at age 3, requiring 5 days of IV ceftriaxone and clindamycin. An immunodeficiency work up was initiated, which demonstrated poor Ab response to pneumococcal and tetanus vaccines.

Discussion: RPA is an infection in the retropharyngeal lymph nodes, which drain the mucosa of the upper airway. Clinical manifestations of RPA vary greatly and commonly include fever, irritability, and torticollis. Respiratory findings, if present, include sore throat, stridor, and respiratory distress.

Lemierre’s syndrome is direct extension of oropharyngeal flora into the IJ, resulting in septic thrombophlebitis, that can shower emboli to the lungs. It is a known, but rare, complication of RPA.

This patient’s presentation was unique in that she initially complained of nonspecific hoarseness and sore throat. She did not become febrile, or develop respiratory distress, until 5 days into her illness. The multifocal pneumonia present at admission indicates that she likely had already developed Lemierre’s syndrome, despite this not being apparent on initial imaging.

This patient’s history of severe infections with atypical organisms points to the need for an immunodeficiency evaluation, which in the hospital setting can be initiated by CBC, quantitative Ig’s, and vaccine titers.

Conclusion: While this is an extreme example, RPA can be associated with severe complications. In this case, airway obstruction and IJ thrombophlebitis combined with near-catastrophic effect. Given the non-specific clinical manifestations of RPA, and the rapidity with which patients can deteriorate, clinicians should be diligent about ruling out RPA in young patients presenting with stridor or respiratory distress. Notably, this patient had airway compromise after only 1 day of stridor.

Multifocal pneumonia is not characteristic of RPA, and should point the clinician towards septic thrombophlebitis. Clinicians should maintain a low threshold for neck ultrasound, because Lemierre’s syndrome, especially in its early stages, can be missed on CT.
Many patients with Lemierre’s syndrome develop thrombosis secondary to infection and do not have an underlying coagulopathy. Thus, there is debate if anticoagulation is necessary at all, and if so for what duration. Despite clot resolution at 3 weeks post discharge, this patient was continued on enoxaparin for 3 months.

**Resources:**

**References:**


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B170

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**Title:** Shed some Light  
**Authors:** Omar Shakeel, PGY-3, Children's Healthcare of Atlanta  
Ann Beach, MD, Children's Healthcare of Atlanta  

**Case Presentation:** A 8 year female presented with recurrent painful and swollen hand and feet that started while at Six Flags over Georgia. This had occurred twice before specifically after outdoor exposure and was associated with a burning sensation. On exam the swelling was most obvious across dorsum of her bilateral hands and feet. The swelling did not improve after benadryl or epinephrine. Initial workup including RF, ANA, CBC-d, BMP, UA, RVP, and CRP revealed leukocytosis but was otherwise reassuring. Given concern for angioedema, Allergy and Immunology was consulted and complement level were obtained and found to be normal. By hospital day 2, an erythematous nonblanching rash had appeared over the dorsum of her bilateral hands and lateral aspects of her feet. The rash continued to evolve and formed purpuric macules coalescing into patches.

**Discussion:** The differential for nonblistering skin lesions is extensive and includes polymorphous light eruption (PMLE), juvenile spring eruption, solar urticarial, drug induced photosensitivity, sunburn, Erythropoietic Protoporphyria (EPP), and X-linked Protoporphyria (XLP). Dermatology was consulted and performed a punch biopsy on our patient. Total and free erythrocyte protoporphyrin as well as fecal protoporphyrin levels were obtained and found to be quite elevated—consistent with Erythropoietic Protoporphoria (EPP). Pathology results were also consistent with EPP. The patient was discharged and given instructions on strict sun protection with zinc based sunscreen and prescribed beta carotene.

**Conclusion:** Porphyrias are rare metabolic disorders that occur in the synthesis of heme. The porphyrin precursors that accumulate can affect the skin (cutaneous porphyrias), neuroviscera (acute porphyrias), or combined. EPP is the third most common porphyria and the most common in childhood. EPP is an autosomal dominant disorder due to deficiency of the enzyme ferrochelatase (FECH) causing protoporphyrin to accumulate in tissues of the body. Patients typically present with painful blistering of skin after exposure to sunlight. The initial screening test is to obtain a total erythrocyte protoporphyrin and if elevated obtain metal-free (elevated in EPP) and zinc protoporphyrin levels. Urine and fecal porphyrin testing is not typically required for diagnosis. Unfortunately, there is no cure for EPP. Protection or avoidance from sunlight is required throughout life and beta carotene may increase tolerance to sunlight. Investigational therapies are currently underway to provide a better quality of life for patients with EPP.

**Resources:**  
Wang HC, Yousef E. Erythropoietic Protoporphoria Masquerading as Angioedema in a 4-YearOld Female. Allergy, Asthma, and Clinical Immunology: Official Journal of the Canadian Society of Allergy and Clinical Immunology. 2006; 2(1):20-23.  

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B171
Title: Six-month Old with Hypoglycemia and Metabolic Acidosis  
Authors: Elizabeth Darnell, MD, PGY-2, University of North Carolina  
Case Presentation: A 6-month-old exclusively breastfed boy presented to an outside hospital with hypoglycemia, lactic acidosis, and tachypnea in the setting of refusal to take a bottle. Past medical history included transfer to the NICU at 2 days of life for compensated anion gap metabolic acidosis, which improved with dextrose containing fluids. Physical exam was notable for tachypnea, increased work of breathing, tachycardia, and hepatomegaly. Labs were significant for hypoglycemia and an anion gap metabolic acidosis (glucose of <20 mg/dl, pH of 6.9, lactate of 15.7 mmol/L). He was started on dextrose containing fluids and transferred to our facility after 2 days.

On transfer, he was euglycemic with lactate of 8.9 mmol/L, pH of 7.34, and lactate:pyruvate ratio of 19.7. Physical exam was significant only for hepatomegaly. He had asymptomatic hypoglycemia overnight (glucose as low as 12 mg/dl). A liver ultrasound showed an enlarged echogenic liver, suggesting an infiltrative process. Findings were consistent with a glycogen storage disease. Genetic testing confirmed glycogen storage disease type Ia.

Discussion: Glycogen storage disease (GSD) type I is an autosomal recessive disorder caused by a deficiency in the activity of glucose-6-phosphatase (G6Pase) and its transporter complex. Fasting hypoglycemia is the hallmark of this disease. G6Pase affects the terminal step of gluconeogenesis, which leads to the accumulation of glycogen. The liver, kidney, and intestines are primarily affected, leading to hepatomegaly and nephromegaly. In addition, hyperlipidemia, hyperuricemia, and lactic acidemia are characteristic metabolic changes.

The mainstay of management for GSD I is dietary therapy aimed at maintaining euglycemia, such as tube feedings and slowly digested carbohydrates. Dietary therapy allows avoidance of early symptomatic signs of disease; however, it does not change the chronic complications. Labs and imaging are regularly monitored to assess development of complications, including nephrolithiasis, gout, chronic kidney disease, hepatocellular adenomas, anemia, and low bone density. Liver transplantation is a last resort, but can be curative.

Conclusion: While glycogen storage diseases are rare metabolic disorders, it is important to keep on the differential of infants who present with tachypnea, increased work of breathing, hypoglycemia, and metabolic acidosis. Newborn screens pick up several inborn error of metabolism, but some can be missed. Diagnosis is important to decrease episodes of hypoglycemia and acidosis and its sequelae. Since fasting even 3-4 hours can cause hypoglycemia, night-time feeding is important to avoid hypoglycemia due to overnight fasting. It is important for pediatric hospitalists to make this diagnosis in the inpatient setting to help guide outpatient management and regular screening for complications.

Resources: N/A

Presented at: Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B172

Title: Uncommonly Common: The Dishonest Urinalysis  
Authors: Justen Aprile, MD, Penn State Hershey Medical Center  
Case Presentation: We describe the case of a previously healthy 4yo female referred to our institution because of multiple renal infarcts on CT obtained as part of her workup for fevers, vomiting, and abdominal pain. She was well until 5 days prior to presentation when she developed fevers, cough, sore throat, abdominal pain, and emesis. Vitals revealed a temperature of 37.1° C, HR 150, RR 26, BP 101/64. Patient complained of abdominal pain, but distraction yielded benign palpation. She had a 1/6 SEM vs holosystolic murmur at the left sternal border without stigmata of endocarditis. Laboratory testing revealed mild leukocytosis of 14. BMP was normal. Urine was positive for small ketones and blood. Abdominal CT revealed multiple wedge-shaped lesions in the left kidney suspicious for renal infarctions, striated left nephrogram with the possibility of pyelonephritis, and multiple pericecal lymph nodes consistent with mesenteric adenitis. Echocardiogram, CXR, renal ultrasound, and ultrasound of the jugular and subclavian vessels were normal.

Discussion: Unilateral, striated nephrograms with wedge lesions takes on a limited differential: acute pyelonephritis, ureteric obstruction, contusion, renal vein thrombosis[1]. Unilateral renal infarcts are quite rare and a review of the literature yielded predominately adult literature and sparse pediatric case studies. The differential diagnosis for renal infarctions includes septic emboli, embolic events secondary to cardiac pathology, hypercoagulability, and vasculitis. Consultation with Rheumatology and Heme/Onc lead to exploration of some of the listed differential diagnoses. Coagulation studies were in the upper limit of normal, fibrinogen was 613, D-Dimer 1.6, LDH 1,143, CRP 24.55, and ESR.
Evaluation for common, inheritable thrombophilia was negative. Blood and throat cultures were negative. ID was consulted for persistent fevers and tachycardia. Ceftriaxone, vancomycin, and metronidazole were initiated because of concern that infarcts represented septic emboli, potentially from Lemierre’s disease given sore throat.

**Conclusion:** On day 5 of hospitalization, urine culture grew > 100K pan-sensitive E Coli. A transient bacteremia with hematogenous seeding of the kidney and early abscess formation may have accounted for her presentation. Striated nephrograms with wedge shaped renal lesions can be secondary to acute pyelonephritis, but typically with supportive findings on urinalysis. Rare case reports supported her hypercoagulable and rheumatologic workup. Kuzmanovska et al describe a 9yo male with flank pain found to have renal infarcts. Vasculitis was suspected and SLE was diagnosed with positive antiphospholipid antibodies[2]. Tsugawa et al discuss a 13yo male with known SLE without antiphospholipid antibodies presenting with abdominal pain, hypertension, and fever found to have a left renal artery thrombosis[3].

With her urine culture, additional workup was held and patient was discharged on PO antibiotics with an improved trajectory. This case represents a unique presentation of acute pyelonephritis with a misleading urinalysis leading to the workup of hypercoaguable and rheumatologic etiologies.

**Resources:**

**Bibliography**


**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B173

**Title:** When the patient looks worse than the labs...a 3 year old with leg pain

**Authors:** Nirupma Sharma, MD, Medical college of Georgia
Kathleen Mahoney, MD, Medical College of Georgia at Augusta University

**Case Presentation:** 3 year-old previously healthy Caucasian female presents with 9 week history of right lower extremity (RLE) pain in the posterior right thigh and right popliteal fossa. Initially, she also complained of left lower extremity pain that resolved after a few days, but she denies pain, erythema, or swelling of other joints or muscles. She has had no fever. The pain is episodic and not relieved by acetaminophen, ibuprofen, ice, or heat; it occasionally wakes her from sleep. RLE radiographs, bone scintigraphy, and inflammatory markers were normal in previous work-up. Symptoms have worsened over the past month and now include refusal to walk for the past two weeks, decreased oral intake with 1kg weight loss, and intermittent rash. Exam reveals a child writhing in pain without focal musculoskeletal findings. Work-up this admission shows leukocytosis with mildly elevated inflammatory markers. While awaiting MR imaging, she develops fever, urinary retention, and she loses her anal wink reflex.

**Discussion:** MRI of pelvis showed a large sacral mass originating in S1-S2 vertebrae with large nonenhancing regions concerning for infection versus less likely necrotic tumor. MRI of thoracic, lumbar, and sacral spine showed extension along the lumbosacral plexus with encasement of the L5 and S1 nerve roots radiologically favored to be neuroblastoma or sacral teratoma. Preliminary biopsy showed small round blue cell tumor. Final biopsy results showed Ewing sarcoma; in addition, FISH analysis was performed and found abnormal rearrangement of the EWSR1 gene that is commonly associated with Ewing sarcoma. Repeat bone scan showed sacral activity and left lower extremity activity concerning for metastasis; MRI did not show LLE involvement. The patient was placed on induction chemotherapy including vincristine, ifosfamide, and etoposide per Children’s Oncology Group Ewing sarcoma regimen. Her urinary retention has improved and she is walking.

**Conclusion:** The differential diagnosis of a limping child is broad and includes congenital, traumatic, infectious, rheumatologic, and neoplastic etiologies (developmental dysplasia of the hip, fracture, dislocation, septic arthritis, acute and chronic osteomyelitis, pyomyositis, discitis, transient synovitis, juvenile idiopathic arthritis, bony tumors like Ewing sarcoma and osteosarcoma, leukemia).
Bone scintigraphy is often recommended as a sensitive albeit less specific test to evaluate extremity pain. It can detect osteomyelitis very early in the disease course. In patients presenting with back pain, bone scintigraphy has a reported 98% negative predictive value. The key in this clinical case is the persistence and worsening of clinical symptoms; these should prompt additional work-up including imaging.

Physical exam of children with lower extremity pain should include assessment of lumbosacral nerve roots including anal wink and saddle area sensation. Development of cauda equina syndrome should prompt emergent work-up.

**Resources:** N/A

**Presented at:** Poster Session #2, Saturday, July 22, 2017, 5:30 - 6:30 p.m., Poster #B174