The Leo J. Geppert Award

The Leo J. Geppert Award, given by the Uniformed Services Section of the American Academy of Pediatric, is an annual citation and pursuit for the best paper by a Uniformed Services pediatrician for research in primary pediatric care.

This award was first presented in San Antonio, Texas in March 1997.

The award is named in honor of Dr. Leo J. Geppert for his many contributions to military pediatrics, as the first Chief of Pediatrics at Brooke Army Medical Center, and Chief of the first Department of Defense Pediatrics Residency Program.
Leo J. Geppert
Colonel, Medical Corps, US Army (Ret)

Leo J. Geppert was born on 26 January 1915 in Vermillion, South Dakota. After completing his BA in chemistry at University of South Dakota Medical School, receiving a Masters Degree in Biochemistry in 1937. In 1939, he completed medical school on a full scholarship at Washington University in St. Louis, Missouri, and entered his pediatric internship at St. Louis Children’s Hospital, St. Louis, Missouri. His residency training, completed in 1941, was at St. Louis Children’s Hospital and Johns Hopkins Children’s Hospital in Baltimore, Maryland.

Dr. Geppert was commissioned as a 2LT in the US Army through the ROTC in 1941. His initial assignment was at the Medical Replacement Center, Barkley, Texas as a training officer. During World War II, he was assigned as the Executive Officer and Commander of the 309th Medical Battalion attached to the 84th Infantry Division. This included service during the infamous “Battle of the Bulge.”

As the first Chief of Pediatrics at Brooke Army Hospital from 1946 to 1952, he established the first pediatric service in an Army hospital. From 1953 to 1955 he was Commander to the Tokyo General Dispensary in Tokyo, Japan, and as Theatre Consultants in Pediatrics, Armed Forces of the Far East. In 1955 to 1958, he served as Chief of Pediatrics at Walter Reed Army Hospital.

While there he was involved in the diagnosis and treatment of such dignitaries as President Eisenhower’s grandchildren, Vice President Nixon’s children and the King of Saudi Arabia’s children. Colonel Geppert returned to Brooke Army Medical Center as Chief of Pediatrics in 1958.

He was an unpaid consultant to Santa Rosa Children’s Hospital for a number of years prior to his leaving the Army. In 1964, he retired from the Army and accepted a position as Medical Director of the Santa Rosa Children’s Hospital. He served as an unpaid consultant to Santa Rosa for a number of years prior to his leaving the Army.

In 1968, he went into private practice for one year. He then became a staff physician for the State of Texas in San Antonio, Texas, continuing in this capacity until he was diagnosed with lung cancer in 1979.

During his military career he received many awards and decorations, such as; Combat Medic Badge, Army A commendation Medical (with clusters), Bronze Star for Valor, Legion of Merit, and a Special Award from the Association of Uniformed Pediatricians in 1978.
After a long illness COL Geppert died in San Antonio, Texas, on 8 November 1980. He is buried at Fort Sam Houston National Cemetery.
<table>
<thead>
<tr>
<th>Year</th>
<th>Name</th>
<th>Award Title</th>
</tr>
</thead>
<tbody>
<tr>
<td>1997</td>
<td>CPT Delores M Gries, MC, USA</td>
<td>Evaluation of an Early Discharge Program of Mothers and Infants Following Childbirth in a Military Population</td>
</tr>
<tr>
<td>2002</td>
<td>LTC Mark W Thompson, MC, USA</td>
<td>The Simulated Delivery Room: Teaching Crisis Resource Management Skills to Pediatric Residents</td>
</tr>
<tr>
<td>2003</td>
<td>MAJ William Adelman, MC, USA</td>
<td>Who Sees the Young Women? Providing Comprehensive Teen Women’s Health Through Shared Resources at a Large Military Community Hospital</td>
</tr>
<tr>
<td>1999</td>
<td>LT COL Kent Hymel, USAF, MC</td>
<td>Missed Abusive Head Trauma</td>
</tr>
<tr>
<td>2004</td>
<td>CPT Vinaya Garde, MC, USA</td>
<td>Tertiary Vaccinia in a Breastfeeding Infant</td>
</tr>
<tr>
<td>2000</td>
<td>MAJ Woodson Jones, USAF, MC</td>
<td>Introduction of a Procedures Laboratory for New Pediatric Interns</td>
</tr>
<tr>
<td>2005</td>
<td>Capt Cassandra Burns, MD, USAF, MC</td>
<td>Phytoestrogens as an Exogenous Etiology of Premature Thelarche</td>
</tr>
<tr>
<td>2001</td>
<td>MAJ Woodson Jones, USAF, MC</td>
<td>How Helpful is Pneumatic Otoscopy in Improving Diagnostic Accuracy?</td>
</tr>
<tr>
<td>2006</td>
<td>Case: CAPT Jay Dintaman, MC, USA</td>
<td>Case of Adolescent with Paget-Schroetter Syndrome and Underlying Thrombophilia Due to an Elevated Lipoprotein (A)</td>
</tr>
</tbody>
</table>
| 2007 | Case: Capt Matthew Eberly, USAF, MC | Research: MAJ(P) Victoria Cartwright, MC, USA  
Infant lumbar puncture simulation is valuable for pediatric resident training                                                                 |
<p>|      | Research: MAJ Keith Lemmon, MC, USA | The Effect of Pediatric Resident Participation in a Field Medical Training Program on Knowledge Acquisition and Attitudes about Military Humanitarian Assistance Operations |
|      | MAJ Woodson Jones, USAF, MC | Introduction of a Procedures Laboratory for New Pediatric Interns                                                                          |</p>
<table>
<thead>
<tr>
<th>Year</th>
<th>Case</th>
<th>Research</th>
</tr>
</thead>
</table>
| 2008 | Case: MAJ Taylor Sawyer, MD  
A Unique Case of Pierson syndrome | Research: LT Corrie E Stofcho, MD  
A Partnership with Partnership for Smokefree Families: NMCSD collaborates with the community for successful smoking cessation intervention |
| 2009 | Case: LtCol Lee Williams, USAF, MC  
Deletion of one allele of Chromosome 14q13-14q24 associated with a unique presentation of Brain-Thyroid-Lung Syndrome | Research: LCDR Timothy Wilks, MC, USN  
Experience with Comparative Genomic Hybridization: Pediatricians Peering into Pandora’s Box |
| 2010 | Case: CPT Kara-Marie Hack, MC, USA  
Propanolol: Not just for Cardiovascular Disease Anymore | Research: CPT Kellie Haworth, MC, USA  
Incidence and Severity of Postpartum Depression Among Military Beneficiaries: A Performance Improvement Project |
| 2011 | Case: LCDR Lisa Peterson, MC, USN  
Indolent Osteomyelitis Following a Plantar Puncture Wound in a Immunocompetent Host | Research: Capt Jonathan A. Stering, USAF, MC  
Does the Site of the Vaccination Clinic Effect Infant Vaccination Rates? |
2012 Top 6 Abstract

A rare cause of hypoxemia and stridor in a neonate

A pediatric case of co-trimoxazole-induced drug rash with eosinophilia and systemic systems (DRESS)

An Unusual Presentation of a 14 Year Old Male with Headaches

Breath-Holding Spells and Sleep Apnea in a Child with a Brainstem Glioma

Improvement in Joint Manifestations in a patient with Mucopolysaccharidosis type 1 treated with Etanercept

Gas Gangrene in a Seven Year Old. – An Infectious Disease Emergency

All Abstracts for 2012

ACUTE PANCREATITIS SECONDARY TO LISINOPRIL IN PEDIATRIC PATIENT

Adrenal Insufficiency and Growth Failure Secondary to Inhaled Corticosteroids: A Paradoxical Complication

Anaphylactoid reaction to Multivitamins in Parenteral Nutrition in a Neonate

A Unique Presentation of Crohn’s Disease in a Pediatric Patient with Familial Adenomatous Polyposis

An Unusual Presentation of Duodenal Stenosis in a Patient with Trisomy 21

Auriculotemporal Syndrome with Introduction of Solid Foods

Association between maternal use of the atypical antipsychotic quetiapine and persistent pulmonary hypertension in the newborn

Development of Methemoglobinemia and Hemolytic Anemia Associated with Rasburicase Administration in a Patient with Hyperuricemia and Unknown G6PD Deficiency

E. coli Bacteremia in the setting of Inpatient Fecal Disimpaction
Infant management in the face of maternal Group B Streptococcus sepsis at delivery

Intracranial Bleed in Previously Healthy Infant

Left cerebellar infarction secondary to a spontaneous vertebral artery dissection (sVAD) in a healthy adolescent female

Mistaken Identity: The importance of the genital exam in the general pediatric well-visit

Omphalitis and Evolving Practices in Umbilical Cord Care

Omphalomesenteric Duct Fistula in Infants and a Possible Etiology of this Rare Condition

PRIMARY CILIARY DYSKINESIA IN A NEONATE WITH PERSISTENT HYPOXEMIA TREATED SUCCESSFULLY WITH NASAL SUCTIONING

Severe Hyponatremia Due to Water Birth in a Term Infant of an Uncomplicated Pregnancy

Splenic Abscess in an 8 year old with Trisomy 21

Spontaneous Resolution of Congenital Pulmonary Lymphangiectasia Without Surgical Intervention

Transient Mycobacterium-Avium Complex pulmonary infection in a healthy adolescent

Unusual Presentation of Adrenal Hemorrhage in a Newborn

Using Hair Shaft Analysis to Untangle a Complicated Case

West Nile Encephalitis in a Previously Healthy Child: Evaluation for CCR5 Chemokine Receptor Mutation
Abstract Title: Adrenal Insufficiency and Growth Failure Secondary to Inhaled Corticosteroids: A Paradoxical Complication

Abstract:

Oral steroid use is a well-known cause of Cushing’s syndrome and adrenal axis suppression in children. Far less common is adrenal axis suppression from inhaled steroids, an underappreciated risk of asthma treatment. We report a case of a child who developed symptomatic adrenal axis suppression and growth failure from inhaled corticosteroid use, yet paradoxically did not develop Cushingoid features, leading to a delay in her referral and diagnosis of adrenal insufficiency. The patient is a 10 year old Caucasian female with a history of asthma and allergic rhinitis treated simultaneously with inhaled fluticasone propionate/salmeterol (Advair® HFA 115/21) via valved-holding chamber two puffs twice daily and fluticasone propionate nasal spray (Flonase®) two sprays in each nostril once daily for the past three years. Since 9 years of age she had decreasing linear growth velocity, poor weight gain, and fatigue. She had never taken oral steroids, nor did she have any Cushingoid features. Laboratory evaluation for causes of her poor growth and fatigue showed a random cortisol level that was undetectable with otherwise normal routine screening labs for short stature. Follow-up testing revealed a low baseline ACTH of 8 pg/mL (normal 10 – 60 pg/mL) with no cortisol response (cortisol < 0.2 mcg/dL) to 250 mcg of ACTH. Adrenal antibodies were negative. Her mineralocorticoid axis was unaffected. Given these findings, she was diagnosed with central adrenal insufficiency secondary to suppression from inhaled corticosteroid use. Due to the severity of her symptoms, the patient was started on physiologic steroid replacement with hydrocortisone. Her linear growth improved markedly and her fatigue resolved. The inhaled and intranasal corticosteroids were stopped, with adequate control of her asthma and rhinitis achieved with montelukast (Singulair®) monotherapy. She received six months of oral hydrocortisone treatment followed by a two month taper. A subsequent cortisol level was normal at 32.3 mcg/dL after her taper was completed, indicating recovery of her adrenal axis. Fluticasone propionate, a potent steroid with 18 times dexamethasone’s relative binding affinity for the glucocorticoid receptor, has been reported to cause signs of glucocorticoid excess and adrenal axis suppression in both children and adults. Our patient, however, paradoxically developed symptoms of adrenal insufficiency while still using inhaled corticosteroids. To explain this paradoxical complication, it has been suggested that some patients show differential tissue sensitivity to corticosteroids, and that restriction fragment length polymorphisms of the glucocorticoid receptor affect corticosteroid sensitivity in a tissue-specific manner. For our patient it was likely that her
inhaled corticosteroid dose was sufficient to suppress her more sensitive hypothalamic-pituitary-adrenal (HPA) axis but was not adequate enough to prevent the systemic symptoms of hypoadrenalism, such as fatigue, weight loss and growth failure. In summary, this case emphasizes the need to use the lowest possible dose of inhaled corticosteroid therapy when managing asthma and rhinitis in order to minimize potential adverse effects on the HPA axis. Increased vigilance is needed for early detection of adrenal insufficiency in non-classic presentations.
Abstract Title: Unusual Presentation of Adrenal Hemorrhage in a Newborn

Abstract:

Adrenal hemorrhage in neonates is a rare diagnosis; however, it is not completely uncommon. The newborn in this case with adrenal hemorrhage presented in such a way that a surgical emergency topped the differential diagnosis list. A term female infant was born via spontaneous vaginal delivery to a 25 year-old mother. The pregnancy itself was uneventful; however, the delivery was complicated by the presence of thick meconium and rupture of membranes for 19 hours. APGAR scores were 7 and 8 at 1 and 5 minutes, respectively. Initially the infant transitioned well with the mother, but by 7 hours of life, the infant was noted to be tachypneic to 90 breathes per minute with decreased muscle tone throughout. Although there were no identified maternal infectious risk factors, the infant’s presentation was initially suspicious for sepsis. Therefore, blood cultures were obtained and intravenous ampicillin and gentamicin were started. Upon admission to the neonatal intensive care unit, the infant’s hemoglobin and hematocrit were 10.4 and 30, respectively. This initially raised questions, but the following physical examination finding raised greater concern. During the physical examination, the infant experienced one episode of bilious emesis. At that time, malrotation was suspected and further workup was pursued. An upper GI study suggested a strong suspicion for malrotation and a surgical consultation was requested (image 1). During surgery, malrotation was quickly ruled out and a left adrenal hemorrhage was identified. The abnormal upper GI findings were reported to be secondary to a mass effect on the bowel from the adrenal hematoma. A review of pertinent literature regarding adrenal hemorrhage presentations is presented. The clinical presentation depends on the volume of the bleed. Small bleeds are often asymptomatic; whereas larger bleeds often result in abdominal, flank, or scrotal masses. Adrenal hemorrhages are often found incidentally on radiographic studies. Although adrenal hemorrhages can be identified by abdominal ultrasound, there is documentation of a case presenting as abdominal calcification noted on abdominal radiography. Other clinical findings include a decreasing hematocrit from blood loss or unexplained jaundiced secondary to the reabsorption and breakdown of red blood cells in the hematoma. Although rare, some infants do present with adrenal insufficiency, as this is often a complication of bilateral adrenal hemorrhage. Right sided adrenal hemorrhage is the most common because of its anatomical location between the liver and the spine allowing it to become trapped and therefore hemorrhage. Upon review of the literature, it appears that bilateral adrenal hemorrhages are second most common in frequency, and therefore
the least common are left adrenal hemorrhages. This case demonstrates an unusual presentation of a neonatal adrenal hemorrhage. Although malrotation often presents as bilious emesis, physicians should now add adrenal hemorrhage to the differential diagnosis list of a newborn with this presentation.
Image 1. In this patient, the duodenal-jejunal junction, at the Ligament of Treitz, does not cross midline, as would be expected in a normal upper GI study.
Abstract Title: Transient Mycobacterium-Avium Complex pulmonary infection in a healthy adolescent

Abstract:

Introduction Pulmonary disease due to Mycobacterium Avium-Complex (MAC) is well described in patients with chronic lung disease or immunodeficiency. Despite the ubiquitous presence of MAC in the environment, it is not known to cause respiratory disease in children without risk factors. We report a case of self-resolving pulmonary MAC disease in a previously well teenager without risk factors. Case Report A 14 year-old, previously healthy female, presented to the ER with a complaint of left sided chest pain, intermittent productive cough and mild fatigue. She denied fever, weight loss, or night sweats. Physical exam was unremarkable. A chest radiograph (CXR) obtained during the evaluation showed diffuse bilateral nodular airspace opacities. A computed tomography (CT) scan of the chest was obtained and confirmed the presence of multiple diffuse small pulmonary nodules bilaterally (figure 1). She was admitted to facilitate further evaluation. Laboratory investigation demonstrated an erythrocyte sedimentation rate of 60 mm/hr and a C-reactive protein of 2.9 mg/dL. Complete blood counts, serum chemistries and an angiotensin converting enzyme level were all within normal limits. Serologic evaluation was negative for Aspergillus species, Coccidiodes immitus, Histoplasma capsulatum, and Blastomycyes dermatidis as well as Bartonella henselae, and Bartonella quintana. A tuberculosis skin test was nonreactive. Three separate sputum samples were obtained for culture, and all were negative for fungal or common pathogenic bacterial organisms. However, two out of the three samples obtained on consecutive days, grew mycobacterium avium complex (MAC) approximately two weeks after the sample was obtained. At a follow up evaluation two weeks later, her cough and fatigue persisted. However, a repeat CXR demonstrated a significant decrease in pulmonary nodules. Anti-microbial therapy was not initiated, but laboratory evaluation of her immune system was normal. Repeat sputum cultures did not grow organisms. Her cough resolved two months following initial onset of symptoms. Discussion MAC is the most frequently identified nontuberculous mycobacteria to cause human infection. It can cause a variety of illness from localized lymphadenitis in healthy patients to disseminated multi-system disease in immunocompromised individuals. MAC pulmonary disease was once thought to be largely limited to patients with acquired immunodeficiency syndrome. However, more recently, progressive pulmonary infection has been demonstrated in even immunocompetent adults without bronchiectasis or chronic obstructive pulmonary disease. While lymphadenitis attributable to MAC is common in healthy children, reports of pulmonary disease have been limited to young patients with immunodeficiency or chronic lung disease. Specifically, cystic fibrosis is the most frequently associated risk factor. To our knowledge, there have been no
reports of pulmonary disease attributable to MAC in children without known risk factors. MAC is ubiquitous, and may frequently colonize the respiratory tract without causing disease. Distinguishing true disease from colonization can be challenging. In order to aid clinicians in the distinction, the American Thoracic Society has developed guidelines for the diagnosis of pulmonary MAC disease that places equal importance on clinical, radiographic, and microbiologic criteria. The positive sputum cultures from 2 separate samples at the time of presentation, the multi-nodular appearance on CXR and CT, the productive cough, and the exclusion of other etiologies in the presented case fulfill the diagnostic criteria set forth. It is interesting that MAC was not grown from cultures at the follow-up visits. Furthermore, the radiographic resolution of pulmonary abnormalities correlated with the timing of negative cultures. Therefore, we postulate that this patient’s pulmonary MAC infection spontaneously resolved. This case calls into question how many other low risk children may develop transient pulmonary MAC disease that goes unrecognized or mislabeled because of low clinical suspicion.
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Abstract Title: Left cerebellar infarction secondary to a spontaneous vertebral artery dissection (sVAD) in a healthy adolescent female

Abstract:

The patient is a 17 year old healthy and physically active female who woke up one morning with an occipital headache with associated symptoms of nausea, vomiting, and vertigo. Although she remained alert with appropriate mentation, she was unable to ambulate even with assistance. Upon evaluation in the emergency department, initial CT and MRI imaging showed a moderately large left cerebellar infarction in the territory of the posterior inferior cerebellar artery (PICA). Further imaging was completed with an MR angiogram and revealed a left vertebral artery dissection and narrowing at the C1 level (V3) on the left side, with distal filling of PICA with atretic narrowing of the vertebral artery from the PICA takeoff to the basilar artery. The location of her headache was consistent with the image findings. She was initially managed in the intensive care setting with mannitol and steroids given concern for posterior fossa swelling following the stroke. Nevertheless, her hospital stay was unremarkable, and she was clinically without any neurological deficits. Her headache resolved, motor skills and strength remained intact, and was able to maintain appropriate balance and gait. With significant progress in her clinical status, and in agreement with the pediatric neurology and neurosurgery services, she was started on a daily low-dose aspirin and was well on all outpatient follow-ups. Few data exist about sVAD in the pediatric population. Unlike adults who present with neck pain, pediatric and adolescent patients present with a constellation of symptoms to include headache, emesis, and ataxia. Angiography and MRA are often used, and treatments include antiplatelet or anticoagulation therapy. It is a potentially debilitating cause of stroke although research has shown younger age as an independent predictor of a favorable outcome. There have been several cases reporting an association between routine neck movements in physical activities and the evolution of sVAD. In retrospect, the patient remembered completing several series of sit-ups the night before the onset of her symptoms. There is a possible cause and effect, but cannot be proven at this time.
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Abstract Title: Association between maternal use of the atypical antipsychotic quetiapine and persistent pulmonary hypertension in the newborn.

Abstract:
Atypical antipsychotics are used with increasing frequency in women of child bearing age because of their improved side effect profile compared to typical antipsychotics. General consensus suggests that the maternal benefits these medications provide outweigh the potential fetal and neonatal risks, and risks to the neonate are in the form of extrapyramidal signs and withdrawal. To date, there have been no case reports of atypical antipsychotics and neonatal persistent pulmonary hypertension. We describe an association between atypical antipsychotic use during pregnancy and persistent pulmonary hypertension of the newborn. In this case a woman with bipolar disorder was on quetiapine throughout her pregnancy, increased from 25mg to 150mg, and delivered a term infant with persistent pulmonary hypertension. Because of that history, during her next pregnancy it was decided that she remain on 25 mg of quetiapine throughout. However that infant, delivered at 32 weeks post menstrual age, also had clinical evidence of pulmonary hypertension. We speculate that there may be a dose dependent association between maternal quetiapine use and persistent pulmonary hypertension in the newborn.
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Abstract Title: ACUTE PANCREATITIS SECONDARY TO LISINOPRIL IN PEDIATRIC PATIENT

Abstract:

Multiple medications are associated with a drug-induced pancreatitis and the list of implicated medications continues to grow. This is the first report of a child with acute pancreatitis secondary to lisinopril. She presented to the ER with complaints of significant epigastric abdominal pain, chest pain and vomiting for several hours. Past medical history was significant for obesity (BMI of 30.4), recently diagnosed hypertension and asthma. Current medications included Lisinopril 5 mg which she began taking 2 months prior. Family history was non-contributory. Lab data was suggestive of pancreatitis, hemoconcentration and metabolic acidosis. CT scan abdomen showed acute, severe pancreatitis grade E, CTSI 10. She was admitted to ICU for management of severe pancreatitis. Lipid panels including triglycerides, immunoglobulin subclasses, ANA, viral studies, and blood cultures were obtained and were unrevealing. Pancreatitis gene testing was negative. Abdominal ultrasound was negative for biliary disease. Lisinopril was discontinued. She progressed to multiorgan failure which responded to supportive care and early nasojejunal feeds. She was transferred to medical floor from ICU on day 8 of illness and discharged home one week later. CT scan abdomen prior to discharge revealed pseudocyst. We conclude that her severe pancreatitis was drug-induced since all other causes of pancreatitis were negative. Conclusion: With growing epidemic of obesity and resultant co-morbidities, an increased number of pediatric patients are screened and treated for hypertension. This article emphasizes the risk of pancreatitis associated with Lisinopril not previously reported in pediatric literature.
Abstract Title: Splenic Abscess in an 8 year old with Trisomy 21

Abstract:

Case Description: An eight year old female with Trisomy 21 presented after 9 days of fevers and abdominal pain. Fevers had occurred daily, ranging from 101-102.6. The abdominal pain was initially intermittent and diffuse. She had no nausea, vomiting, nor anorexia and no abnormalities on abdominal exam. She had negative blood cultures on Day 1, 3, and 7 of her illness. She also had normal urinalysis and urine cultures on day 1 and 7. EBV titers were negative. Both chest and abdominal x-rays revealed no abnormalities. Her electrolytes remained within normal limits. Her only risk factor for infection was a 3cm laceration on her forehead three weeks prior to presentation. On day 7 of her illness our patient was started on PO clindamycin when she was found to have two small pustules on her forehead around the healed scar. Her CBC at this time demonstrated WBC count of 13.5 with 75% neutrophils. Her ESR was elevated to 86. On day 8-9 her abdominal pain became localized to the left upper quadrant. At this time she demonstrated mild LUQ tenderness with no rebound or guarding, and no hepatosplenomegaly on exam. Hospital Course: The patient was admitted to the pediatric ward for further evaluation and management. A CT scan of the abdomen revealed a heterogeneous splenic mass consistent with an abscess. She was started on IV clindamycin and Piperacillin/Tazobactam. Within 24hrs of starting parenteral antibiotics her fevers and abdominal pain resolved. She was discharged on HD4 with a plan for continued parenteral antibiotics. After two weeks of therapy her PICC was removed and she was transitioned to PO linezolid and augmentin. The patient did not tolerate the augmentin suspension and was changed to moxifloxacin. She received a total of approximately 6 weeks of broad spectrum therapy. She was followed with serial abdominal ultrasounds which demonstrated gradual resolution of her abscess. Discussion: Splenic abscesses are a rare cause of intra-abdominal pathology in the pediatric population, although incidence does appear to be rising. They are more common in patients who are immunosuppressed. High rates of HIV and increased use of chemotherapeutic drugs likely play a role in increased incidence. Diagnoses have also increased with more sensitive imaging modalities. Splenic abscesses have been caused by many organisms including gram positive, gram negative, anaerobic, AFB, and fungal organisms. Patient population, underlying disease process and co-morbid factors all play a role in identity of the likely causative organism. This case represents a typical presentation of a rare condition. Fever, leukocytosis, and LUQ abdominal pain are the classic triad described in the literature. However, a splenic abscess in an immunocompetent patient with no signs of bacteremia or other infectious etiology is extremely
rare. Hematogenous spread from infective endocarditis is a commonly identified cause of splenic abscess. Our patient has a history of AV-canal defect and PDA, both repaired in infancy. She is therefore at an increased risk of infectious endocarditis, but had no clinical signs or symptoms of disease. Evaluation included a total of 4 negative blood cultures as well as a negative echocardiogram. Our patient had no other identified intra-abdominal pathology to suggest contiguous spread, which has commonly been reported as an alternative mechanism. Infections have also been reported secondarily after significant trauma, or related to hemoglobinopathies such as sickle cell disease. No such risk factors were identified in this case.
West Nile Virus (WNV), a flavivirus transmitted by the Culex mosquito, was first detected in the United States in 1999 and has since spread across the country to become the primary cause of arboviral encephalitis. Approximately 80% of infected individuals are completely asymptomatic, 20% develop a non-specific febrile illness, and <1% present with West Nile neuroinvasive disease (WNND). Most cases of WNND occur in adults or immunocompromised patients. We report a case of West Nile Virus Encephalitis in a previously healthy 2 year old girl who presented to the ER in status epilepticus after 2 days of fever, headache, abdominal pain and several bouts of emesis. During her hospitalization she required ventriculostomy and external ventricular drain placement for increased intracranial pressure and 4 days of pentobarbital-induced coma to break her seizures. Magnetic resonance imaging showed focal abnormalities in the left parietal lobe. Despite resolution of her seizures, she continued to show signs of encephalopathy including aphasia, dyskinetic oropharyngeal movements, lower extremity extensor posturing, extreme muscle weakness and a mild hand intention tremor. Within 1 month of her presentation however she had made a complete recovery. Testing for many possible causes of encephalitis was performed including enterovirus, herpes simplex virus, human herpes virus 6, cytomegalovirus, Epstein-Barr virus, influenza, adenovirus, parainfluenza and respiratory syncytial virus, as well as Borrelia, Mycoplasma and Bartonella. Additional testing for arboviral causes through the CDC showed positive WNV IgG and IgM ELISA from both the child’s serum and CSF, which was confirmed on further testing. The patient’s entire family was tested for evidence of WNV infection, and although he was completely asymptomatic, her father had positive serum WNV IgG. In an effort to uncover the cause of WNND in our previously healthy patient, an evaluation for primary immunodeficiency was completed, which showed normal immunoglobulin levels, normal complement, positive titers to immunizations and isohemagglutinins as well as normal T and B cell subsets and absolute leukocyte count, implying a normally functioning immune system. We further hypothesized that our patient had a mutation in a chemokine receptor that affected her immune system’s ability to clear WNV from her CNS. Approximately 1% of Caucasians in the United States possess a loss of function mutation in the chemokine receptor CCR5 32, which is known to provide resistance to HIV-1 infection. This same receptor is important in combating WNV infection as in mouse models it has been shown to mediate recruitment of leukocytes to the WNV infected CNS. A study of WNV disease in adults showed that neurologic manifestations were more common in those patients who were
homozygous for the mutant CCR5 32 allele. Based on this information we tested our patient for this chemokine receptor mutation, but found her to have the normal wild-type pattern. Although her testing was normal, we believe she may have an underlying immunologic defect placing her at increased risk for WNND, such as a mutation in the chemokine CXCL10 which has been shown to play a significant role in recruiting virus specific T-Cells to clear WNV infection. We are presently pursuing testing for a mutation in this chemokine in our patient. This is the first known report of a child with WNND being tested for the CCR5 receptor mutation. We believe a better understanding of the immunologic mechanisms involved in clearing WNV infection could lead to new treatment options and help identify which populations are at increased risk for WNND.
Abstract Title: Spontaneous Resolution of Congenital Pulmonary Lymphangiectasia Without Surgical Intervention

Abstract:

Introduction: Recent case reports and series show that the diagnosis of severe congenital pulmonary lymphangiectasia (CPL), manifested by antenatal pleural effusions, respiratory distress at birth and hydrops fetalis, is not uniformly fatal. However, the natural disease course and ideal management remain unclear. We present a patient with CPL and hydrops fetalis who survived the neonatal period with spontaneous resolution of clinically significant chylothoraces at six weeks of life, which has not previously been reported. Case: A right sided pleural effusion was identified, in a female fetus, on prenatal ultrasound at 32 weeks gestation. Amnioreduction and in-utero therapeutic thoracentesis was recommended but declined by the mother. Additional follow-up was not done until she presented at 35 weeks gestation at which time an ultrasound revealed bilateral pleural effusions, fetal hydrops and worsening polyhydramnios. A viable female fetus was delivered at 35 4/7 weeks gestation. She was diffusely edematous without gross dysmorphic features. She developed respiratory distress requiring bilateral thoracentesis. The pleural fluid was consistent with a chylous effusion. High resolution computerized tomography was consistent with CPL. Supportive care included mechanical ventilation, bilateral chest tubes, prophylactic antibiotics, immunoglobulin and albumin infusion. She failed to resolve the chylous drainage with a trial of octreotide. Although a pleural-peritoneal shunt was recommended at one month of life, the family refused surgical intervention. Over the next several weeks the effusions spontaneously resolved. The patient recovered and was discharged home. Discussion: Previous reports have concluded that the optimal management of the most severe form of isolated CPL includes both aggressive medical management and surgical intervention. Our case adds to the literature demonstrating that survival is possible with aggressive medical support but without surgical intervention. This was true even in the face of effusions persisting beyond one month of age and a trial of octreotide that appeared to be ineffective.
Abstract Title: Using Hair Shaft Analysis to Untangle a Complicated Case

Abstract:

Introduction: Trichothiodystrophy (TTD) is a rare, autosomal recessive, neurocutaneous disorder of DNA repair with a variable clinical phenotype. A recent review identified only 112 published cases of TTD worldwide; 16% of these patients were identified in the United States. True prevalence may be higher due to changes in diagnostic criterion, the use of multiple names to describe the syndrome over time, and poor physician awareness of this rare disease. Affected patients may present with variable combinations of brittle hair, ichthyosis, developmental delay/disability, recurrent infections and immunodeficiencies, CNS abnormalities, growth retardation and history of maternal pregnancy complications. A wide range of severity has been reported, but overall TTD is associated with significant morbidity and mortality. Coarse, sparse hair is a key physical exam finding in TTD, but can also be seen in Menke's Disease, Argininosuccinic aciduria and other rare neurocutaneous disorders. The demonstration of “tiger tail banding” on hair shaft analysis under polarizing microscopy aides in distinguishing TTD from these rare genetic diagnoses; unfortunately, few Pediatricians employ this simple diagnostic tool. We report two brothers with variable clinical findings who were simultaneously identified to have TTD based on hair shaft analysis. Case Report: Patient A presented as a 2 month old male born at 30weeks EGA with coarse, brittle hair, ichthyosis and mild dysmorphic features. Family history was notable for his 21 month old brother (Patient B) who was concurrently hospitalized due to an atypical viral pneumonia. Patient B’s medical history was notable for prematurity, profound developmental delays, hypotonia, nystagmus, recurrent nonbacterial respiratory infections, ichthyosis with photosensitivity, and severe failure to thrive. Brain MRI was notable for hypomyelination. Despite his complex history, thorough evaluation had not yielded a unifying etiology. Examination showed dysmorphic features with coarse, sparse hair and eyebrows. The similar physical exam findings between the brothers prompted further work up. Patient A was found to have normal myelination on MRI, metaphyseal widening and spurring, a blonde fundus and normal hearing. Patient B was identified to have hypogammaglobulinemia, swallow dysfunction, subclinical cataracts and normal hearing. Both had a normal array CGH and family history was unremarkable. Copper and ceruloplasmin were normal, ruling out Menke’s disease. Dermatology assisted in hair shaft analysis. Light microscopy of both sets of hair showed trichoschisis and flattening of the hair shaft; tiger tail banding indicative of Trichoiodystrophy was seen under polarized microscopy. Supplemental DNA studies have been
sent to the National Institutes of Health and are pending. Social history was notable for upcoming reassignment orders for the patients’ father, with plans to relocate in less than 1 month. Information regarding the new diagnosis was shared with the father’s command and reassignment orders were amended to ensure adequate medical care availability for both patients. Conclusion: TTD is a rare and poorly understood disease with variable presentation that is associated with complex medical findings and significant morbidity and mortality. The above cases highlight the utility of hair shaft analysis when neurocutaneous findings are present. This helpful diagnostic tool is quicker and less expensive than DNA analysis, and should be considered early in any young child presenting with complex multisystem and cutaneous abnormalities. Hair shaft anomalies such as trichorrhexis nodosa, trichorrhexis invaginata, pili torti and tiger tail banding may guide a general Pediatrician to an uncommon diagnosis. Although there is no cure for TTD, early recognition allows for aggressive preventative healthcare, improvements in quality of life through supportive interventions and accurate parental counseling for recurrence risk to future pregnancies. Early recognition of complex genetic disorders is particularly and uniquely important for active duty military families, who are frequently assigned to remote locations where complex medical support is unavailable.
Abstract Title: Development of Methemoglobinemia and Hemolytic Anemia Associated with Rasburicase Administration in a Patient with Hyperuricemia and Unknown G6PD Deficiency

Introduction Methemoglobinemia results from the oxidation of ferrous iron (Fe++) contained within the hemoglobin molecule to the ferric form (Fe+++), which decreases the ability of hemoglobin to transport oxygen and carbon dioxide. Rasburicase, a recombinant urate oxidase, has been documented as causing methemoglobinemia secondary to the production of hydrogen peroxide during the breakdown of urate to allantoin. Methylene blue is administered to patients in order to biochemically reduce methemoglobin and restore oxygen carrying capacity. However, the mechanism of action of methylene blue is disrupted in patients who are glucose-6-phosphate dehydrogenase (G6PD) deficient. We describe a case of methemoglobinemia and hemolytic anemia after administration of rasburicase to a patient with unknown G6PD deficiency for treatment of hyperuricemia secondary to suspected acute leukemia. Case Presentation A 15 y/o African American male with unremarkable past medical history was admitted to the pediatric ward for evaluation of suspected acute leukemia. Family history was negative for G6PD deficiency or other hematologic disorders. Initial complete blood count showed WBC of 67,000, hemoglobin of 6, and platelet count of 9,000. Manual differential had 90% blasts. Screening labs for tumor lysis syndrome revealed an elevated uric acid of 9.9mg/dL. In an effort to prevent kidney injury from urate crystal formation, rasburicase was administered. Three hours after rasburicase was given, the patient was noted to have persistent oxygen saturation in the mid 80s without evidence of respiratory distress. Oxygen saturation was not responsive to supplemental oxygen. An ABG was obtained and demonstrated a methemoglobin level of 19.8%. Methylene blue, 1mg/kg/dose, was ordered for treatment of hypoxemic methemoglobinemia. Despite methylene blue, the patient’s oxygen saturation was slow to improve. 30 hours after methylene blue, oxygen saturation had only improved to 92% and there was evidence of progressing tumor lysis syndrome. The patient was transferred to another center for dialysis. Methemoglobin level at time of transfer remained elevated at 7.1%. During this time, the patient developed evidence of hemolysis based on grossly bloody urine, urinalysis positive for large blood and negative for red blood cells, and elevated bilirubin levels. In addition, the patient required 5 units of packed red blood cells to maintain hemoglobin levels between 6 and 8.4mg/dL. This prompted quantitative testing of G6PD which confirmed a deficiency. Discussion G6PD deficiency is an inherited x-linked recessive mutation common in
African Americans and people of Mediterranean descent. Although rasburicase is indicated for the management of plasma uric acid levels in patients with leukemia, it can cause hemolytic anemia and methemoglobinemia in patients with G6PD deficiency secondary to excess hydrogen peroxide formation during uric acid breakdown. In this case, rasburicase administration resulted in methemoglobinemia which was unable to be adequately treated with methylene blue. The activation of methylene blue is dependent on proper functioning of the hexose monophosphate shunt. G6PD catalyzes the initial step of the shunt and is responsible for the conversion of NADP to NADPH. Individuals with G6PD deficiency have insufficient NADPH available to reduce methylene blue. This results in methylene blue accumulation which further oxidizes hemoglobin. In our patient the additional oxidative stress led to further hemolysis rather than contributing to the methemoglobin reduction. Although G6PD deficiency is a disease that infrequently causes clinical manifestations, prior screening would have been useful in this case. G6PD deficiency should be considered prior to the administration of rasburicase and non-emergent chemotherapy in certain high risk populations. Additionally, methylene blue should be avoided if methemoglobinemia develops in individuals with suspected or known G6PD as methylene blue can lead to worsening methemoglobinemia and hemolysis in this patient population.
Abstract Title: Breath-Holding Spells and Sleep Apnea in a Child with a Brainstem Glioma

Abstract:

Cyanotic breath-holding spells (BHS), even when resulting in loss of consciousness, generalized jerks and opisthotonus, are generally considered benign episodes that can occur in otherwise healthy infants and young children. The recommended management focuses on parental reassurance of the self-limiting nature of this condition. Often not known is that these BHS can rarely be associated with abnormalities of the CNS, including tumors of the brainstem. We report a case of a young child with significant obstructive sleep apnea syndrome and increasingly frequent, severe cyanotic breath-holding spells who was found to have a brainstem glioma by imaging before he manifested any overt neurological signs. The patient is a 4-year-old male initially referred at age 3 years for evaluation of recurrent wheezy respiratory illnesses, including two episodes requiring brief hospitalization. Reassuringly, he did not have cough, wheezing or shortness of breath when well, and his chest x-ray was unremarkable. More concerning, however, were his morbid obesity, chronic snoring, and frequent breath-holding spells since infancy. His parents also expressed concerns of respiratory pauses during sleep. His breath-holding spells were characterized by central cyanosis, stiffening of the body, eyes rolling back and loss of consciousness lasting 5-10 seconds, followed by a dazed appearance upon arousal. These spells were always precipitated by crying and/or anger. The child had no reported headache, nausea, vomiting, gait abnormalities or behavioral changes. His initial physical examination was remarkable for a BMI of 27, moderate tonsillar hypertrophy, normal cardiac and respiratory auscultation, and normal neurological exam. The patient underwent polysomnography, which revealed severe obstructive sleep apnea syndrome (OSAS), with an AHI of 16.9 (including 5 central and 3 mixed apneas) and moderately severe oxygen desaturations. He was admitted for CPAP initiation while awaiting ENT consultation. Repeat evaluation showed mildly elevated systolic blood pressure, normal CBC, ferritin, serum HCO3, TSH, and prolactin levels, and a normal pCO2 on capillary blood gas measurement. Mildly elevated pulmonary artery systolic pressure was noted on echocardiogram, but the right ventricular dimensions were normal. The patient became intolerant of his CPAP therapy but eventually underwent tonsillectomy and adenoidectomy, with improvement of the snoring and restless sleep. Because of his obesity, a repeat polysomnogram was obtained, and it showed no correction of the severe OSAS. He again had several central apneas, and now there was also concern of a generalized spike pattern on the EEG channels interpreted as possible epileptiform activity versus
artifact. Meanwhile, the patient continued to have cyanotic BHS that were actually increasing in frequency – now occurring once or twice daily – over the preceding few months, with described opisthotonus developing while still in the upright posture, followed by syncope. He was also noted on examination to have expiratory breaking in the supine position. At this point, nine months after his initial evaluation, a MRI of the brain was ordered to evaluate for abnormalities of the brainstem as the etiology of the central apneas, unusual respiratory pattern and the progressively frequent and severe cyanotic BHS. That study uncovered a partially exophytic dorsal medullary mass, measuring 2.2 cm in largest dimension and enhancing in a focal nodular pattern suspicious for a brainstem glioma. While the mechanism of cyanotic BHS is not yet fully understood, there have been rare reports of medullary tumors found postmortem in children with severe cyanotic BHS who subsequently had sudden, unexplained death. Additionally, the development of severe BHS and central apneas has been seen after resection of medullary tumors. Therefore, this case serves as a reminder to consider the possibility of a brainstem lesion in a child with progressively frequent and severe cyanotic breath-holding spells, especially when associated with central sleep apneas.
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Abstract Title: PRIMARY CILIARY DYSKINESIA IN A NEONATE WITH PERSISTENT HYPOXEMIA TREATED SUCCESSFULLY WITH NASAL SUCTIONING.

Abstract:

INTRODUCTION Primary Ciliary Dyskinesia (PCD) is a genetic disorder that manifests clinically with chronic sinopulmonary and otologic disease. Despite the majority of patients presenting with respiratory symptoms in the neonatal period, the diagnosis is often delayed until several years of age. We report the case of a 5 day old male who was diagnosed with primary ciliary dyskinesia by electron microscopic evaluation of cilia obtained from nasal brush biopsy. This case also demonstrated the successful treatment of persistent hypoxemia with frequent nasal suctioning.

CASE The patient is an infant male born at term following an uncomplicated pregnancy. Family history was notable for an older brother with Kartagener Syndrome. Pt was noted to have an intermittent loose cough several hours following birth, and transmitted upper airway sounds were occasionally heard. A chest radiograph (CXR) obtained on hospital day 1 was unremarkable. Discharge home was anticipated on hospital day 2. Pulse oximetry was obtained during the discharge physical exam and the patient was noted to be hypoxemic with an oxygen saturation of 88%. Respiratory rate varied between 40 and 55 breaths per minute. There was no evidence of accessory respiratory muscle use. The lung sounds were clear to auscultation. The rest of the physical examination was unremarkable. The infant was admitted to the level 2 nursery and further diagnostic evaluations were obtained. Repeat CXR showed no evidence of pneumonia, respiratory distress syndrome, pneumothorax or other evidence of pulmonary disease. Laboratory studies included a complete blood count, cerebral spinal fluid (CSF) cell count, and venous blood gas within normal limits. Blood and CSF cultures were negative for infectious organisms. An electrocardiogram, hyperoxia test and echocardiogram were within normal limits. Ampicillin and gentamicin were started empirically for a presumed neonatal pneumonia. Antibiotics were discontinued after 6 days following negative culture results and due to a lack of clinical change during the course of antibiotic therapy. Suplemental oxygen was provided via nasal cannula. The patient initially required a flow rate of 2 liters per minute (LPM), however this decreased to 0.5 LPM by hospital day 4. Despite attempts to discontinue supplemental oxygen, the patient did not tolerate multiple attempts to wean to room air. Pediatric pulmonology consultation was obtained on hospital day 5 and a nasal brush biopsy of the inferior turbinate was obtained for electron microscopy (EM) analysis of the cilia. Initiation of nasal suctioning with saline drops and chest physiotherapy (CPT) was recommended. CPT was discontinued after one day due to the lack of clinical response. Nasal
suctioning repeatedly obtained thick nasal secretions. The patient began to tolerate trials of room air immediately following nasal suctioning. The length of time the patient was able to tolerate room air gradually increased as regularly scheduled nasal suctioning was performed. Supplemental oxygen was permanently discontinued 60 hours after nasal suctioning was first initiated, with pulse oximetry recordings consistently above 95% on room air. The pt was discharged home on day of life eleven with instructions to continue nasal suctioning four times daily. Results of the EM studies confirmed the diagnosis of PCD. DISCUSSION We report here the case of a neonate with PCD as the underlying cause of persistent hypoxemia. Rhinosinusitis with thickened nasal secretions is a hallmark of PCD. This case demonstrates that onset of rhinosinus disease can occur soon after birth. The combination of relatively narrow nasal passages along with obligate nasal breathing in neonates likely contributed to nasal airflow obstruction. Persistent hypoxemia was the only significant finding that led to a prolonged hospital course in this patient. Frequent nasal suctioning with saline drops proved to be successful treatment for the persistent hypoxemia.
Abstract Title: Omphalomesenteric Duct Fistula in Infants and a Possible Etiology of this Rare Condition

Abstract:

A previously healthy 12 day-old male infant presented with periodic bleeding at his umbilicus. Initially, umbilical granuloma was diagnosed and treated with silver nitrate applications which failed to result in improvement. The character of the discharge became feculent at 6 weeks of age and a suspected patent omphalomesenteric duct fistula became evident. Surgical repair involved resecting the omphalomesenteric fistula using a stapling device and his postoperative course was uneventful. The second case is of a newborn with a ruptured omphalocele who was taken to the operating room the day after delivery to repair his abdominal wall defect. During the repair, a Meckel’s diverticulum lying within the omphalocele and umbilical stalk was noted and would have been at risk for ligation during cord clamping. This diverticulum was also resected and the abdominal wall was approximated without complication. Discussion: The differential diagnosis for umbilical lesions in the newborn period broadly includes granulation tissue, urachal remnants, and omphalomesenteric duct remnants. Although the first case presented with what appeared to be a simple umbilical stump granuloma, failure to resolve promptly with silver nitrate application or the presence of discharge, especially feculent, should prompt further examination. The etiology of the fistulization in the first infant is adequately demonstrated in the surgical findings in the second newborn. The tethering of the Meckel’s diverticulum to the membrane of the omphalocele indicates that had the abdominal wall been closed, the patent vitelline duct may have resided in the umbilical cord and remained at risk for injury during cord clamping at delivery. This process would allow for a presentation similar to the first infant with failure of the umbilical stump to heal and possible feculent discharge. Omphalomesenteric, or vitelline duct remnants, represent a failure of obliteration of the connection between midgut and yolk sac in the 5th to 9th weeks of gestation. Rare cases have been noted in the literature with some reports indicating a possible spontaneous regression of the patent omphalomesenteric duct fistula to an asymptomatic Meckel’s. Rates of this condition have been reported to be 0.0053%. Regardless of this reported potential regression, once the diagnosis of a fistula is made, prompt initiation of surgical consultation and subsequent resection is important given the opportunity for small-bowel prolapse and potential strangulation. Recognizing a patent omphalomesenteric duct fistula at delivery is not always possible, but any question of abdominal wall defect or umbilical herniation, or minor omphalocele, should prompt the
pediatrician to avoid proximal clamping and resection of the umbilical cord. Subsequent imaging with ultrasound may reveal a communication with the underlying bowel and necessitate early surgical consultation. In addition, the general pediatrician needs to be aware of the possible diagnosis of an omphalomesenteric duct fistula in the patient with an umbilical stump remnant or prolonged granuloma.
Abstract Title: Mistaken Identity: The importance of the genital exam in the general pediatric well-visit

Abstract:

A 3 year and 10 month old female presented to the pediatric endocrine clinic with a chief complaint of clitoral enlargement that was initially noted during infancy. Her mother raised concern at each well-child appointment, but was repeatedly provided with reassurance and counseled that there is significant variation in the size of a normal clitoris. Her genital exam was documented as “normal” at her well-visits over the first year of life. At age two, the patient presented again with the complaint of an enlarged clitoris and a new concern of a “lump” in the left labia that was initially noticed during a diaper change. The lump was described by the pediatrician as a “vulvar ovary” and coded as an “ovarian prolapse.” A referral was placed to urology, but no follow-up with this referral was made. The patient was evaluated multiple times over the following year for upper respiratory infections or ear pain; however, a follow-up genital exam was not documented during these acute visits. At three years of age, the enlarged clitoris and labial mass remained. The child’s behavior was described as increasingly active and aggressive, and in addition, she had developed new-onset, bothersome clitoral erections. The patient was then appropriately referred to pediatric endocrinology. The exam with Walter Reed National Military Medical Center Pediatric Endocrinology revealed bilateral palpable gonads and clitoromegaly. Pelvic ultrasound demonstrated the absence of müllerian structures, a left labial gonad, and a right inguinal gonad. The location of the gonads was consistent with testicles. Her karyotype was 46, XY. Genetic testing was negative for mutations in the SRD5A2 gene (the cause of 5-a reductase deficiency) and also negative for androgen receptor mutations in the NR3C4 gene (the cause of androgen insensitivity syndrome). The historical and clinical features of this child’s genital ambiguity was most consistent with a diagnosis of partial androgen insensitivity syndrome (PAIS), the most common cause of 46, XY Disorder of Sexual Development (DSD). It is not surprising that androgen receptor mutation analysis was negative, given that the sensitivity in the setting of PAIS is estimated around fifty percent. The sensitivity of receptor mutation analysis for complete androgen insensitivity (CAIS) is much higher, ranging from 83-95%, but this patient’s clinical exam is not consistent with the finding of normal external female genitalia in CAIS. The presence of Y-chromosome material and testicles puts her at increased risk for carcinoma in situ and seminoma of the testicle. The risk associated with PAIS for germ cell tumor is as high as 15% with non-scrotal tumors and is unknown in scrotal tumors. This case of 46, XY DSD illustrates the importance of a careful and complete genital exam, not only during the newborn period, but also with all pediatric well-visits. Adequate knowledge of the normal genital anatomy for both male and female pediatric patients is crucial to ensure abnormal findings are not missed. The ramifications of a missed DSD can range from patient and parent frustration with the medical system to mistaken gender assignment, cancer, or even adrenal crisis leading to death. In our pediatric endocrine clinic over the past few months, three patients
have presented outside of the newborn period at ages 3 years, 6 years, and 7 years with the complaint of clitoromegaly that had previously been discussed with their pediatricians. Of these, one patient had 46, XY DSD, one had 46, XX DSD, and the third had normal clitoral measurements. The occurrence of such cases emphasizes the importance of the general pediatrician being responsive to parental concerns and maintaining clinical proficiency in recognizing genital abnormalities in order to make appropriate referrals with careful follow up.
Abstract Title: Anaphylactoid reaction to Multivitamins in Parenteral Nutrition in a Neonate

Abstract:

Case study We report an infant female patient with gastroschisis status post definitive closure who developed urticaria after administration of total parenteral nutrition (TPN). The initial reaction occurred on day of life 15 and she had been on TPN since birth secondary to her gastroschisis. Definitive closure of the abdominal defect had occurred on day of life 5. The anaphylactoid reaction with urticaria developed approximately 30 minutes after the nightly change out of the TPN fluid bag and resolved in less than an hour after stopping the infusion. The urticaria recurred when the administration of the same mixture was attempted the next evening due to a desire to optimize nutrition. The urticaria did not occur when multivitamins, trace elements and minerals were removed from the TPN. Upon challenge of a TPN solution with multivitamin, urticaria recurred. When advanced to full enteric feeding, the patient tolerated oral multivitamins with no allergic reaction. Discussion This case demonstrates that in addition to the previously reported cases of allergic reaction to the multivitamin component in TPN in an adult and a toddler, reactions can also occur in infants. Peripheral or centrally administered parenteral nutrition is an important component of practice in the neonatal intensive care unit as most infants have little nutritional reserve and have a high demand secondary to their growth needs along with their illnesses. Due to known complications of parenteral nutrition including cholestasis, the goal is usually to minimize the length of time of use of parenteral nutrition and maximize enteral nutrition as quickly as possible. At times the disease process of the infant involves the digestive tract and prolonged parenteral nutrition is necessary such as in the case of our infant with gastroschisis repair. Given third trimester deposition of many nutrients it is standard practice to add multivitamins and trace elements to neonatal parenteral nutrition in addition to the dextrose, amino acids and fat emulsion. A review of literature revealed previous case reports for a toddler and an adult with urticarial reactions to presumably the multivitamin component of TPN but no prior report in a neonate. There was a case report of a reaction to an undetermined component in a SGA neonate with no clear etiology but multivitamins possibly implicated. Most reported TPN reactions are related to the lipid component but there are also case reports implicating occult latex exposure. Infants with myelomeningocele have a reported 48% reaction rate to latex but the rate for infants with other disorders is unknown. Given this knowledge a comprehensive review of the materials for preparation of the TPN in our institution was conducted and it was found to be a latex free process. As the infant was also tolerating intravenous fat emulsion with no reaction the treatment team narrowed the source of the reaction to the multivitamin, trace elements or the selenium that had been the last component
added approximately 3 days prior to the first noted reaction. The challenge with the two bag method differing only by multivitamin mixture implicated the multivitamin as being the likely source. Our institution uses Infuvite parenteral multivitamins manufactured by Sandoz Canada (Boucherville, QC) and distributed by Baxter Healthcare Corporation (Deerfield, IL). The oral multivitamin is obtained through Rugby Laboratories (Duluth, GA). There are some differences in components between the two but the exact allergenic substance in the parenteral multivitamin is unknown at this time.
Abstract Title: Infant management in the face of maternal Group B Streptococcus sepsis at delivery

Abstract:

Case study A female infant was delivered at term gestation to a 32 year old G3P4 mother via Cesarean section for non-reassuring fetal heart tones and maternal chorioamnionitis. The infant's mother had presented to triage for decreased fetal movement and was febrile, tachycardic and was ill appearing with a diffusely tender abdomen on physical exam. Prenatal labs were notable for Group B Streptococcus positive with otherwise uncomplicated prenatal course. Due to her ill appearance a blood culture was performed and ampicillin-sulbactam ordered. The fetal monitoring noted tachycardia to 180s with late decelerations and minimal to absent variability. Since she was remote from cervical delivery with non-reassuring fetal information, obstetrics made the decision to proceed with operative delivery and noted that amniotic fluid was grossly cloudy at delivery. The female infant required only standard delivery room resuscitation with Apgar of 8 and 9 with deduction for color. Per our institution protocol the infant was transferred to the NICU to obtain complete blood count and blood culture and to initiate at least 48 hours of treatment with ampicillin and gentamicin. An initial temperature of 38 degrees Celsius was noted on admission and the CBC with manual differential was within normal limits. The infant's blood culture was no growth but the mother's blood culture returned at 24 hours as gram positive cocci that was later identified to be Group B Streptococcus (GBS) bacteremia. As the blood culture had been obtained prior to delivery with time from admission to delivery of 1 hour 40 minutes inadequate for even prophylactic GBS treatment the team made the decision to treat the infant as if she too had a positive blood culture. Due to inability to rule out meningitis with an unsuccessful attempt to attain CSF the decision was made to treat for 14 days with penicillin G as the appropriate treatment course for uncomplicated meningitis. The female infant tolerated the treatment course well and breastfeeding was well established by the time of both infant and mother's mutual hospital discharge. Discussion This case is significant for the paucity of information on infant management in the setting of GBS bacteremia and sepsis in the mother in the perinatal period. The obstetric team treated the mother for 14 days with penicillin using Red Book guidelines written for infant management due to the lack of data for GBS bacteremia treatment in adults. The decision to treat the infant was based on a maternal blood culture obtained shortly before delivery even in the face of negative infant blood cultures. Infant blood cultures when obtained with an adequate volume of blood can be false negative up to 40% of the time. We treated maternal blood culture as equivalent to the infant’s based on the knowledge that the placenta does allow for intra-uterine transmission of other bloodborne infections both bacterial and viral including syphilis and HIV. The hospital protocol is to treat for 48 hours minimum in cases of chorioamnionitis. However we usually stop treatment if infant blood cultures
are negative at 48 hours in an otherwise well appearing infant with no significant other issues including feeding and hyperbilirubinemia. This infant was treated with a full duration of antibiotics, usually 10 days for bacteremia, 14 for uncomplicated meningitis and 21 for complicated meningitis. In addition to the significant rate of false negative blood cultures, neonates can also have meningitis with negative blood culture up to 20% of the time, a point which guided our decision to treat for 14 days for the possibility of uncomplicated meningitis since we did not have CSF available to rule out meningitis.
Abstract Title: Omphalitis and Evolving Practices in Umbilical Cord Care

Abstract:

Neonatal omphalitis is a rare infection in neonates, with an estimated overall incidence of 0.2-0.7% in industrialized nations. It has a notable mortality risk, usually secondary to necrotizing fasciitis. In cases of omphalitis, Staphylococcus aureus is the most frequently reported organism. Despite a variety of studies, the best practice for umbilical cord care has remained elusive. Attempts to decrease bacterial colonization with topical antimicrobials over time may select for resistant organisms or new pathogens. In addition, attempts to keep the cord site clean often lengthen the period to cord separation. Many institutions have begun practicing “dry cord care,” using simple cleaning techniques without topical solutions or antimicrobials. Three months after instituting a policy of dry cord care in a large nursery at a tertiary military medical facility, we report a case of neonatal omphalitis in a previously well 19 day old infant. A 19 day old infant’s parents brought him to the clinic because they had concerns regarding his umbilicus. The umbilical cord had fallen off five days prior to presentation, coinciding with a visit to the pediatrician’s office for a two week newborn visit. One day prior to presentation, the father noticed that the umbilicus and surrounding skin were becoming more erythematous and appeared inflamed. On examination, the 2.5 cm x 3 cm area around the umbilicus was erythematous and the skin was slightly taut with an interior edge of desquamating skin. The umbilicus itself was crusted and contained two discrete areas of subcutaneous purulence at the three and seven o’clock positions from the center. The patient was evaluated for systemic infection, admitted and placed on parenteral clindamycin dosed at 40 mg/kg/day. By hospital day two, the erythema surrounding his umbilicus had decreased. By day five, his umbilicus had nearly returned to normal. He received nine days of parenteral clindamycin, and given his dramatic improvement, completed his tenth day of therapy via oral formulation. While this individual case will not affect our current cord care recommendations, it does underscore the importance of educating parents on what signs and symptoms should prompt immediate evaluation for their infant. In order to establish evidence-based guidelines for cord care practice, multi-center studies should investigate (a) infection rates among various cord care practices, (b) re-colonization rates in various cord practices used after discharge, and (c) the true incidence of omphalitis post-discharge. Omphalitis is an uncommon infection and mandatory reporting of cases would also help track and better define the incidence of this infection across health care systems. Further research is needed before a guideline can be generated for the best umbilical cord care practice.
While water immersion continues to gain popularity as an alternative analgesic option during first stage of labor, it also remains a controversial practice. A recent study could make no recommendations for or against its use, when adverse affects of thermoregulation, respiration, and infection were taken into account. In contrast, recent cases of respiratory distress syndrome, infection, and hypoxic ischemic encephalopathy have been reported. Many case studies have been published reviewing the potential risks to the neonate secondary to water birth, but direct causation of the complications has been difficult to prove. We report the case of a term neonate born in-hospital with immediate hyponatremia requiring prolonged NICU admission after water birth. A 40+6 week healthy female with an uncomplicated pregnancy and no risk factors for sepsis desired immersion birth. Labor and delivery were uncomplicated and performed in a tub; membranes were ruptured for 2.5 hrs. The infant was placed on mom’s chest immediately after birth. At one minute of life she was noted to be pale and cool with bradycardia, so was transferred to a warmer for resuscitation. Neonatal team was called at 3 minutes of life and noted the infant to be pale and cool with poor tone and agonal gasping. Positive pressure ventilation was initiated with improvement in respiration, heart rate and activity. She was then transferred to the NICU, and quickly transitioned to room air. Arterial blood gas was obtained with initial base deficit of 12 and a serum sodium of 126 mmol/L; a repeat sodium was 123 mmol/L within the first hour of life. Antibiotics were started, and with concern for euvolemic hyponatremia, fluids were restricted. Labs drawn during the workup of neonatal hyponatremia included urine sodium <5 mmol/L (30-90), and urine osmolarity of 66 mOsm/kg (50-1200); other normal labs included: WBC, hematocrit, blood cultures, 17-OHPG, renin, vasopressin, uric acid, FT4, TSH and normal newborn screen. Chest and abdominal radiographs were normal, as was a renal-bladder ultrasound. Her physical exam after the initial resuscitation was unremarkable, and at no time did she show signs of seizure activity. Over the next 6 days, her sodium slowly corrected to 132 mmol/L. She was discharged home and remains without sequelae on routine newborn follow-up exams. Currently, the American Academy of Pediatrics recommends water birth only be performed within the context of a randomized control trial with full informed parental consent. Despite this, popularity appears to be growing, fueled by media, celebrity opinion, and increased desire for more parental control of the birthing process. While hyponatremia is one of the assumed possible adverse effects of water births, there have
been no previous cases demonstrating direct relationship. Review of the literature reveals two case reports of hyponatremia following water birth, the first at home and the second report without specifics. To the best of our knowledge this is the first report of a hospital water immersion birth with hyponatremia immediately following birth, with findings consistent with water intoxication and near-drowning. Possible complications of hyponatremia are not trivial and include: sensorineural hearing loss, cerebral palsy, intracranial hemorrhage, and death. Despite an uncomplicated outcome, our patient was subjected to substantial morbidity in the form of prolonged ICU hospital course, extensive laboratory investigation, significant family disruption and distress. This case adds to the growing literature conveying concern and caution for water immersion births. With the increasing popularity of water births, it is the responsibility of the medical community to strongly counsel parents regarding lack of benefit and potential adverse effects to infants born by water birth.
Abstract Title: An Unusual Presentation of Duodenal Stenosis in a Patient with Trisomy 21

Abstract:

The association of intestinal obstructive lesions and Down syndrome (trisomy 21) has been well established. Most of these disorders present with symptoms in early infancy. Here, we describe an atypical case of nearly asymptomatic duodenal stenosis. Pt is a 4.5-year-old male with trisomy 21. He was a former 35-week premature infant without significant complications. Throughout his first 11 months of life, he demonstrated normal weight gain. He had no vomiting, no abdominal distention, no significant gastroesophageal reflux, and no feeding intolerance, or pain associated with feeds. Screening labs done at his 12 month well baby examination revealed a normal complete blood count and electrolyte panel. By 14 months of age, however, he began demonstrating a decrease in weight velocity and was referred to pediatric gastroenterology. His parents reported an increase in regurgitation and some rare episodes of emesis but denied any abdominal distention, or choking with feeds. His physical exam was otherwise unremarkable. Routine labs were obtained, his caloric intake was optimized, and he was started on a proton pump inhibitor (PPI). A follow up visit 3 months later confirmed resolution of his reflux symptoms. A trial to wean off the PPI only resulted in a worsening of his reflux symptoms, and consequently, his medication was resumed. Over the following two years, he demonstrated stable growth. During this period, he had no further vomiting, no significant regurgitation, and was tolerating a normal diet. At the age of 4.5 years, approximately 1 year after stopping his PPI, he began demonstrating an increase in “wet burps” shortly after meals. His parents, assuming a return of his acid reflux symptoms, called in a request to renew his PPI prescription. Given that he had not had any recent follow up evaluations, the peds GI provider arranged for an office visit and ordered an upper gastrointestinal (UGI) contrast study. The UGI revealed a severely dilated duodenal bulb with significant narrowing of the 2nd portion of the duodenum consistent with duodenal stenosis. An abdominal CT scan confirmed this finding, as well as ruling out the presence of any abdominal mass or annular pancreas. A pediatric surgeon subsequently performed a duodenoduodenostomy to repair the stenosis. Following the operation, the patient had no further reflux symptoms, and had a notable increase in PO intake. The majority of intestinal obstructions associated with trisomy 21 are identified early in infancy due to the severity of their symptoms. In the few case reports of patients presenting later in childhood with duodenal stenosis, vomiting has universally been the presenting symptom. Our patient had no significant history of vomiting. Furthermore, his intermittent episodes of mild reflux responded promptly to a PPI. Overall, he thrived on a normal diet without any other GI issues, thus it was not unreasonable that he had no previous imaging studies. Down syndrome is a common predisposing...
condition of obstructive intestinal lesions. Recent AAP guideline for the management of Down syndrome reports that 12% have a form of intestinal atresia. Buchin, et al found that 14% of Down syndrome patients admitted to his institution over a 12 year span had a significant GI disorder with duodenal stenosis being not only the most common, but also associated with the highest mortality. Currently, there are no recommendations for routine radiological screening for gastrointestinal abnormalities in patients with trisomy 21. Given the relatively high prevalence of obstructive intestinal lesions in this population, a form of universal screening could be considered. Nonetheless, in the patient with Down syndrome of any age presenting with even mild GI complaints, further evaluation by a pediatric gastroenterologist or with appropriate imaging studies may be useful.
INTRODUCTION: Familial adenomatous polyposis (FAP) is a condition of autosomal dominant inheritance that is characterized by hundreds to thousands of adenomatous colonic polyps. It carries with it a 100% lifetime risk of development of colon cancer. It can be associated with a variety of conditions, such as congenital hypertrophy of the retinal pigment epithelium, desmoid tumors, and hepatoblastomas, among others; it has not, however, been found to be associated with inflammatory bowel disease. We report the case of a 13 year old female with a history of FAP who presented with acute onset abdominal pain, tenesmus, urgent nocturnal stools, diarrhea, and weight loss. Endoscopy demonstrated patchy duodenitis, ileitis, and left sided colitis, consistent with Crohn’s disease with confounding familial adenomatous polyposis. CASE REPORT: 13 year old female with maternal history of familial adenomatous polyposis initially presented to clinic for discussion of genetic testing of the adenomatous polyposis coli (APC) gene, which was positive for an A to G transition at splice site distal to exon 9, three base pairs from the end of exon 9; this mutation is of unclear significance, and is not known to cause FAP, but screening colonoscopy was nonetheless scheduled given maternal history. Initial colonoscopy was limited to flexible sigmoidoscopy, extending proximally to the splenic flexure, secondary to numerous pedunculated and sessile polyps. Multiple polyps were removed, and were found to be histologically consistent with tubular adenomas. A follow up colonoscopy was to be done in the next 3 to 6 months. Our patient presented 3 months later with the complaint of 2 months of gradually worsening abdominal pain, tenesmus, diarrhea, and an 8 pound weight loss. Other family members initially had similar symptoms that self-resolved in 1 to 2 days, but our patient’s symptoms persisted. The patient was admitted to the inpatient pediatric ward. Admission labs were significant for microcytic anemia, thrombocytosis, hypoalbuminemia, and elevated inflammatory markers, with a hemoccult positive stool. CT of the abdomen revealed distal terminal ileal and right colonic thickening. Upper endoscopy showed mild gastritis and aphthous duodenal ulcers. Colonoscopy revealed aphthous ulcers throughout the colon, and skip lesions of erythematous, edematous mucosa with purulent exudates, which were more prominent in the proximal colon. No polyps were visualized. The distal terminal ileum was also noted to be markedly inflamed. An extensive work-up for infectious etiology was negative. Our patient improved significantly with intravenous corticosteroids, metronidazole, and oral and rectal mesalamine. The biopsies were consistent with Crohn’s disease, involving the duodenum, ileum, and colon. Our patient was started on infliximab and has had an excellent
response for over nine months. DISCUSSION: Patients with familial adenomatous polyposis are often asymptomatic, but can present with a variety of symptoms, including abdominal pain, increased bowel motility, and the passage of blood or mucous with stools. Our patient’s presentation could have been secondary to multiple etiologies, including infectious colitis, large polyp burden, intussusception with a polyp serving as a lead-point, inflammatory bowel disease, or an initial presentation of colon cancer. Additionally, given the undeniable risk of patients with FAP developing colon cancer, affected patients invariably require colectomy at some point in their lives. The presence of Crohn’s disease in our patient will unfortunately complicate her future treatment, including the necessary colectomy. This case illustrates the necessity of developing a broad differential diagnosis in evaluating abdominal pain in pediatric patients with familial adenomatous polyposis.
Abstract Title: A pediatric case of co-trimoxazole-induced drug rash with eosinophilia and systemic symptoms (DRESS)

Abstract:

The evaluation and diagnosis of pediatric patients with fever and rash is both common and challenging. Drug rash with eosinophilia and systemic symptoms (DRESS) is a rare but potentially life-threatening hypersensitivity syndrome characterized by a severe cutaneous eruption, hematologic abnormalities, and variable multi-organ involvement. Diagnosis is associated with a 10% mortality. Along with sulfasalazine and allopurinol, it is most commonly associated with anti-epileptic medications and less commonly with antibiotics. DRESS syndrome distinguishes itself from other cutaneous reactions such as Stevens-Johnson Syndrome (SJS) by a variable and prolonged course that despite prompt withdrawal of the inciting agent, requires immediate treatment with systemic corticosteroids, followed by a slow taper. A previously healthy 9-year-old Caucasian female was treated three weeks prior to admission with a two week course of co-trimoxazole for a presumed urinary tract infection. Shortly after completing treatment, she developed intermittent fevers with progressive symptoms to include headache, fatigue, emesis, diarrhea and anorexia. Onset of an erythematous papular rash and peri-orbital edema prompted clinic evaluations and eventual admission to the pediatric intensive care unit secondary to severe hyponatremia. Review of systems was negative for sore throat, conjunctivitis, respiratory symptoms, joint pains, or mental status changes. Additional history revealed no other medication exposures or drug allergies. The hospital course was significant for the onset of multiple organ involvement to include: hepatitis (peak aspartate aminotransferase 815 units/L (normal < 26), alanine aminotransferase 410 units/L (normal < 44)) with hepatomegaly and generalized ascites, severe persistent hyponatremia unassociated with SIADH (sodium 116 mmol/L), pulmonary edema, generalized lymphadenopathy, hemolytic anemia, leukemoid reaction (WBC of 71,000/µL) with atypical lymphocytes and eosinophilia (16%), hypoalbuminemia (1.9 g/dL), and intermittent somnolence. The rash progressed to involve the majority of her body, sparing the oral and genital mucosa. The lesions coalesced to diffuse erythroderma followed by complete desquamation. A punch biopsy revealed a vacuolar interface dermatitis consistent with DRESS syndrome and inconsistent with SJS. Human herpesvirus-6 (HHV-6) PCR was positive, a consistent finding with this diagnosis, while studies for Streptococcal organisms, Mycoplasma, EBV, and CMV were negative. Intravenous corticosteroids were initiated; the patient slowly improved and was discharged after a 12-day hospitalization. She continued on a slow oral steroid taper, although the recurrence of rash necessitated a six month wean. Three months after discontinuing oral
corticosteroid therapy, the patient remained well without obvious sequelae. While DRESS syndrome is described in the dermatology and immunology literature, it remains relatively unknown to the pediatric clinician, particularly in contrast with SJS. Our patient’s histopathology appeared most consistent with an erythema multiforme reaction, consistent with the emerging theory that DRESS syndrome occurs along a continuum of serious cutaneous adverse reactions which may begin with erythema multiforme and progress to SJS or DRESS, depending on the character and degree of inflammation. This case clearly differentiates between DRESS and SJS: the comparatively late onset of symptoms (weeks versus days after exposure), significant leukocytosis and eosinophilia, frequent and severe liver involvement, lack of mucocutaneous involvement, and the association with re-activation of HHV-6 are features unique to DRESS syndrome. While sulfonamide-containing antibiotics are infamously associated with mucocutaneous drug eruptions, physician prescriptions of co-trimoxazole have only increased in the era of community-acquired MRSA. Although these reactions are uncommon alone, their collective potential morbidity and mortality argues for increased physician discretion when choosing therapy for common pediatric conditions such as urinary tract infections and skin infections. In addition, while treatment of both syndromes necessitates prompt drug removal and supportive care, the role of corticosteroids in DRESS syndrome is vital and should be considered early. This distinguishing and possibly life-saving feature highlights the importance of increased primary physician awareness of this disease.
Abstract Title: Intracranial Bleed in Previously Healthy Infant

Abstract:

Vitamin K dependent bleeding is a rare, but potentially fatal and preventable condition. Because it can be seen in infants even months after birth, clinicians must maintain a high degree of suspicion. A 31-day-old male presented to the pediatric clinic for follow up after an emergency department visit the prior morning. Forty-eight hours prior, the patient's parents had noticed that their son had an isolated temperature of 100.5 degrees Fahrenheit (rectal). In the emergency department, the patient was afebrile and well-appearing. He was discharged home with recommended follow up. Over the next 24 hours, the infant was noted to have a change in his normal cry. Parents also noted increased irritability, decreased activity, and poor breastfeeding. The patient's past medical history revealed a term infant male born via spontaneous vaginal delivery at home and was unable to receive vitamin K at birth. Parents later refused vitamin K at the 2 day and 2 week well baby checks. At these visits the infant was noted to have a good latch, appropriate weight gain, and normal neurologic exam. Hearing and both newborn screens were normal. On the day of presentation, infant was noted to be drowsy, but arousable. On physical exam his anterior fontanel was soft, open, and flat, pupils were equally round and reactive to light, and he was moving all extremities equally. Upon review of the patient's history, a limited sepsis work up with blood cultures was initiated and a coagulation panel was drawn. The initial coagulation panel showed an elevated PT and PTT at greater than 190 seconds and 119 seconds respectively, with an elevated fibrinogen, and a normal d-dimer. An emergent head CT scan revealed a large intracranial bleed. Within one hour of presentation, the patient began to show spasticity in his right upper extremity, with a decreased responsiveness to stimuli. The patient was emergently transported to the PICU, where he was loaded with keppra and given 3 percent sodium chloride, as well as 1mg of intravenous vitamin K and 10mL/kg of fresh frozen plasma. A complete factor panel, drawn prior to the administration of the fresh frozen plasma, showed less than 7 percent activity for vitamin K dependent factors II, VII, IX, and X, and normal factor V and VIII. A comprehensive metabolic panel demonstrated normal liver enzymes and kidney function. Upon stabilization, the infant was transferred to a nearby children’s hospital where he further decompensated and required neurosurgical evacuation of the intracranial hemorrhage. The infant was discharged on post-operative day two in improved condition. This case illustrates the potential life-threatening adverse outcomes from late vitamin K dependent bleeding. Although late vitamin K dependent bleeding is rare, it is preventable with a vitamin K injection at birth. Bleeding, often in the form of an intracranial hemorrhage, has been reported as late as six months after birth, but typically presents within the...
first 2-12 weeks of life. While sepsis is the most common reason for altered behaviors in infants of this age, home birth, vitamin K refusal, and breastfeeding are important risk factors to glean from the history of an infant with changed behaviors that may provide clues for vitamin K deficiency. As the number of home births continues to increase, it is the role of every Pediatrician to actively counsel families on the continued importance of early, exogenous vitamin K administration.
Abstract Title: E. coli Bacteremia in the setting of Inpatient Fecal Disimpaction

Introduction: Constipation is a common clinical problem encountered in pediatric practices. Treatment through well-established algorithms necessitates that disimpaction of the stool burden be performed before initiating maintenance therapy. This can be done orally, rectally, or through a combination of these methods. In certain cases inpatient hospitalization is necessary where therapies such as enemas, suppositories, and/or nasogastric (NG) administration of balanced electrolyte solutions may used alone or in combination to accomplish adequate disimpaction. We describe a case that demonstrates a previously unreported complication of an inpatient bowel cleanout. Case: An 8 year old male with a history of chronic constipation, ADHD, and Asperger Syndrome presented to the pediatrics ward for scheduled inpatient fecal disimpaction after failure of outpatient management. Prior to arriving to the ward, a Gastrograffin enema was performed to rule out distal obstructive processes and evaluate colonic caliber. Results were within normal limits. This study also resulted in evacuation of distal stool, and upon admission a NG tube was placed to enable continuous infusion of polyethylene glycol electrolyte solution as part of a disimpaction protocol. This was continued along with Senna to stimulate motility until the patient’s stool clarified, which took 7 days. Due to concerns of malnutrition due to prolonged lack of enteral nutrition, peripheral parenteral nutrition (PPN) and intralipids (IL) were begun in the afternoon of hospital day (HD) #6. Later that evening the patient’s course was then complicated by development of a fever of 40.4°C. PPN and IL were then stopped due to the possibility of contamination, and cultures of these fluids and the patient’s blood were obtained. The fever quickly resolved thereafter, and the patient remained clinically stable on his disimpaction protocol. The next day his disimpaction was halted as stools clarified, and he was discharged home. The following morning (31 hours from blood draw) the patient’s blood culture grew Gram negative rods (GNR). He was contacted and subsequently readmitted. At the time of this second admission, the patient reported fatigue, malaise, and fever up to 102°F since his discharge. A repeat blood culture and abdominal x-ray showing no signs of free air or megacolon were obtained, empiric coverage for GNRS with ceftriaxone was begun, and the patient quickly defervesced. Later, his initial blood culture was found to have grown Escherichia coli sensitive to ceftriaxone. After his repeat blood culture was negative for 48 hours he was discharged on a 14 day course of oral cefdinir. The repeat blood culture was followed for 5 days with no growth noted. Discussion: Bacteremia in the setting of an inpatient fecal disimpaction is a clinical situation not previously reported. In the case described
here, the source of the patient's E. coli bacteremia was not decisively known. There was initial concern for contamination of the PPN and IL infusates, but both had negative culture results. Contamination of the blood sample sent for culture was considered as well, but seemed unlikely given our patient's new fever at the time of culture. Contamination with stool would also more likely be polymicrobial. It then seemed most probable, given his prolonged fecal disimpaction, that the patient's own GI tract was the source of his infection by bacterial translocation. The physical damage to our patient's intestinal mucosa during his prolonged disimpaction could have led to increased permeability, and E. coli is known to undergo translocation relatively easily. Conclusion: In summary, this case of E. coli bacteremia in a child undergoing inpatient NG fecal disimpaction represents a clinical problem never before reported in the literature. The source of this bacteremia is also suspicious for a novel presentation of bacterial translocation.
Abstract Title: An Unusual Presentation of a 14 Year Old Male with Headaches

Abstract:

Introduction Rathke’s cleft cysts may present with headaches at any age as their only symptom and may be associated with pituitary-hypothalamic-chiasmal derangements. This case involves an adolescent male with changes in the characteristics of his baseline headaches who was diagnosed by MRI with Rathke’s cleft cyst without any further symptoms and was subsequently diagnosed with borderline hypocortisolism. Clinical Course A 13 year old male with history of migraines for 4 years often preceded by an aura of central vision changes and nausea experienced headaches that worsened over the past 6 months. The headaches were located bifrontally and occipitally, often woke him, occurred about four times a week, and were unresponsive to medication. Review of systems was noncontributory, and physical exam was unremarkable including the neurological exam. An MRI revealed a 1.3 cm cystic intrasellar mass. A Rathke’s cleft cyst was considered the most likely etiology. MRI also revealed some mass effect and displacement of normal-appearing pituitary tissue in addition to upward mass effect on the optic chiasm. Extensive endocrine testing revealed a borderline low response to the low dose cortrosyn stimulation test with cortisol values of 10.6 µ g/dL and 14 µ g/dL, respectively, and the remainder of his screening pituitary function labs including IGF-1, testosterone, LH, FSH, prolactin, free T4, and TSH were normal. Neuro-ophthalmology examination was normal, including a visual field exam. Four months after diagnosis, our patient has experienced fewer and less severe headaches on migraine medication. He will be treated with stress doses of hydrocortisone for surgery, persistent vomiting, or significant febrile illnesses. This patient will have serial MRI monitoring in 6 months and then yearly MRI and visual field testing. Pt will also have laboratory surveillance every 4 months. Discussion Rathke’s cleft cyst is a non-neoplastic lesion entrapped by a single layer of epithelium in the sellar or suprasellar regions. It develops from failure in early fetal development of the obliteration of Rathke’s pouch which is tissue that gives rise to segments of the pituitary gland. Although the cysts are often asymptomatic, encountered in 12 to 33% of normal pituitary glands on routine autopsies, symptoms of Rathke’s cleft cysts can be subtle and longstanding or initially present at any age. However, the slow growth rate of the cyst makes the incidence of symptoms increase with aging. There is little documentation in the pediatric literature regarding manifestations and outcomes of Rathke’s cleft cysts. In children with symptomatic presentations, visual loss from compression and diabetes insipidus due to pituitary stalk involvement are the most common symptoms. Less common presentations include variable endocrine insufficiencies including delayed puberty, central precocious puberty, growth retardation, menstrual abnormalities, headaches, and vomiting. In adults, with symptomatic endocrine derangements, hyperprolactinemia is the most common estimated as high as 75%, followed by gonadotropin deficiency, pan- hypopituitarism, hypothyroidism, and hypocortisolism estimated in about 25%. Spontaneous rupture of the cyst can
cause a severe aseptic meningitis, abscess formation, or pituitary apoplexy. This patient presented with headaches, and further evaluation revealed possible hypocortisolism. Headaches from cysts are present in approximately 50% of symptomatic adult patients and reflect either increased intracranial pressure from mass effect of the tumor itself, from obstructive hydrocephalus from tumor compression of the 3rd ventricle, or from meningeal irritation by escaped cyst contents. Magnetic resonance imaging usually is performed for the diagnosis and evaluation of these lesions. Simple drainage and partial excision of the cyst wall is undertaken in some symptomatic lesions usually those causing recurrent visual field defects and hyperprolactinomas. Drainage does not often improve pituitary function. Persistent headaches that wake children from sleep are concerning and physicians should consider neurologic imaging.
Abstract Title: Gas Gangrene in a Seven Year Old. – An Infectious Disease Emergency

Abstract:

Introduction: Clostridial myonecrosis or gas gangrene affects upwards of three thousand adults in the United States annually. It is uncommonly seen in children, with less than fifty cases reported in the medical literature. The following describes a case of gas gangrene in a pediatric patient. Case: A seven year old African-American male, with a past medical history significant for pancytopenia secondary to aplastic anemia with Myelodysplastic Syndrome, presented to the Emergency Department with a day long history of intensifying left calf pain and an eight hour history of worsening fevers. Two days prior to admission, he had had a bone marrow biopsy of his left posterior iliac crest performed without complications. On the day prior to admission, he had been transfused platelets due to a low count, and was found to have an unremarkable examination. His only medications were prophylactic Fluconazole and Aminocaproic Acid as needed. He is allergic to Penicillin, which causes him to develop urticaria. No recent travel or pets in the home. No recent history of leg trauma or insect bites was reported. Admission vital signs were significant for a fever to 39.1 C, a heart rate of 160, and a blood pressure of 119/88. On physical exam he was ill appearing, his left leg flexed in a position of comfort. The site of his bone marrow biopsy did not appear infected. His left lower extremity was extremely tender to palpation, with tense overlying indurated skin and several large bullae. His distal pulses were minimally palpable. Labs drawn included a complete blood count (WBC of 0.79 with 20S 67L and 13M, H/H of 9/25.8 with 26K platelets) as well as a set of blood cultures. Vancomycin, Meropenem, and Clindamycin were started to provide broad spectrum coverage. Radiographs of his left leg showed free air within the muscles of the calf and thigh. An above the knee amputation was emergently performed with necrotic muscle noted to the level of his left upper thigh. He made four trips to a hyperbaric oxygen chamber in the first three days of his illness, and was administered granulocyte colony - stimulating factor (GCSF) and intravenous immunoglobulin (IVIG). His blood cultures were negative but tissue culture from his initial surgery was positive for Clostridium septicum, for which he received a nearly six week course of IV antibiotics. Subsequent operating room debridement revealed no further necrotic muscle or subcutaneous tissue. A split-thickness skin graft was placed over the stump site three weeks after the initial surgery. Patient was discharged home after an almost two month hospital stay. Discussion Clostridium septicum is a large, gram positive spore forming bacilli that is found in the normal gut flora of humans and is ubiquitous in soil. Gas gangrene caused by Clostridium septicum is seen most commonly in patients with underlying hematologic abnormalities, colon cancer and trauma victims, though cases in those previously healthy have been reported. Clinical findings in children typically include pain out of proportion with physical findings early on, with subsequent development of discolored and edematous skin as well as marked tachycardia.
Mainstays of treatment are antibiotics and debridement, though hyperbaric oxygen, GCSF and IVIG have been used as adjunctive therapy. The disease is rapidly fatal without treatment. Conclusion Gas gangrene in pediatric patients, while rarely reported, should be considered in patients whose pain is out of proportion with physical exam findings – especially if free air is seen on radiographic studies. It is a true medical emergency - prompt initiation of appropriate antibiotics and surgical consultation for debridement is critical if the patient is to have any chance of survival.
Abstract Title: Auriculotemporal Syndrome with Introduction of Solid Foods

Abstract:

Introduction: Auriculotemporal Syndrome, otherwise known as Frey Syndrome or Gustatory Flushing Syndrome, is a rare condition in infants and toddlers. It is often confused for a food allergy as it manifests during introduction of solid foods. Typical presentation is flushing and/or hyperhydrosis along the auriculotemporal nerve when eating food and which resolves almost immediately once the gustatory stimulus ceases. Case Report: We present a 7 month old previously healthy male in clinic with his PCM with “feeding problems”. Patient had previously been eating baby foods for 2.5 months with little to no concerns. However, for the past 1 to 2 weeks, he has developed a red blotchy rash under his eyes and down the sides of his face [Left side greater than Right side] that begins halfway through a feeding session. The rash persists until eating stops and then promptly disappears. Throughout feeding, he is in no distress and appears to be non-pruritic. There are no other associated lesions to include urticaria abberentelsewhere on patient’s body. He has no difficulties breathing. There is no direct contact with the food in this distribution and there is no erythema peri-orally or in locations where there is direct food contact. The erythema only occurs with solid foods but does not occur with the 4 8 oz bottles of formula he takes daily. Over this same time period he has also developed a concurrent solid food aversion where he will take food in his mouth and then gag and spit it back up. He is still taking his daily formula. FHx: Mother is a very picky eater who often gags and vomits with foods with many textures of foods. Father’s side of family with significant atopy history and food allergies. PMH: none Physical: HR: 123, T: 98.3 °F, HT: 74 cm [90%], Weight: 9.8 kg [78%], Pulse Oximetry: 100% on Room Air, BMI: 17.9 General appearance: Well-appearing, alert, well nourished, well hydrated, active male in no acute distress. HEENT: Ear normal bilaterally. Nares/nose normal. Oral cavity and oral pharynx normal. Neck: Normal with no cervical lymphadenopathy. Lungs: Clear to auscultation. Cardiovascular system: Regular, rate and rhythm with no murmurs heard Abdomen: Nontender to palpation with no masses appreciated. Skin: Face is normal without any lesions at this time in clinic with no food introduction in clinic. No other rashes or lesions present. Discussion: Auriculotemporal Syndrome is an uncommon condition in infants. Typically it is associated with introduction of solid foods without a good source of etiology. In adults, this condition is connected to trauma of the parotid glands with resultant aberrant nerve healing. While it has been proposed forceps delivery at
birth as a cause of trauma, there are an equal number of cases in literature in which delivery did not require forceps usage. The flushing in response to gustatory stimuli resolves on its own over time without intervention. Auriculotemporal Syndrome is important to keep in the differential of patients with rash and no respiratory distress symptoms with introduction of solid foods because work up and sensitization testing with an immunologist is expensive and can be painful.
Abstract Title: A rare cause of hypoxemia and stridor in a neonate

Abstract:

Intro The differential for hypoxemia and stridor is extensive, but in the newborn period congenital anomalies must be considered. There are many known congenital anomalies associated with hypoxemia and stridor, including both those that are intrinsic and extrinsic to the airway. Ectopic thymic tissue has been documented in the literature as part of the differential for a neck mass, but we present the first clinically significant case of ectopic thymic tissue causing hypoxemia and stridor. Furthermore, this mass was not appreciable on physical exam. Following excision there was complete resolution of all respiratory symptoms. Case Presentation A 23-day-old otherwise healthy male presented to the emergency room on day of life 23 because of maternal concern for increased work of breathing and difficulty feeding. Patient was afebrile and had an otherwise normal exam with no focal signs of infection. In the ED he was placed on continuous pulse oximetry where a desaturation was observed to the high 80s, lasting less than a minute and resolving with blowby oxygen. Patient was admitted to the pediatric ward for observation pending the results of a sepsis evaluation. Antibiotics were discontinued after 48 hours as there was no growth observed in blood, urine, and cerebral spinal fluid cultures. Further work-up included flexible laryngoscopy and MRI which revealed a right-sided parapharyngeal mass. Infant was taken to OR for complete excision, at which time the mass had to be freed from cranial nerve XI, internal jugular vein, internal and external carotid arteries, and cranial nerve XII, with the pathologic diagnosis of thymic tissue. Discussion During the sixth week of development, the thymus develops from the third pharyngeal pouch. During the sixth to eighth week, it travels through the neck to the superior mediastinum. It is during this descent that remnant tissue can be left. This tissue is usually asymptomatic, only increasing in size in response to an infection or vaccination. It is in this evaluation of an enlarging neck mass that ectopic thymic tissue is found. Literature review suggests <100 cases of ectopic thymus found in the evaluation of an asymptomatic mass, however this is the first reported case of ectopic thymus being found in the evaluation of hypoxemia and stridor with no palpable mass. Ectopic thymus is generally only a postoperative diagnosis. At time of biopsy, it is impossible to differentiate between a thymic mass with cystic components versus a thymoma with cystic degeneration. Since thymic masses are generally well-encapsulated, a complete excision once the mass is freed from surrounding neurovasculature is done with otherwise little complication. Once the diagnosis is made after removal of the complete mass, there have been no reported cases of
recurrence. And similarly to our patient, excision is curative with resolution of respiratory symptoms. Conclusion When evaluating an infant for hypoxemia and stridor that cannot be explained by pulmonary or cardiovascular causes, it is important to include ectopic thymus as part of the differential even without a mass on physical exam.
Abstract Title: Improvement in Joint Manifestations in a patient with Mucopolysaccharidosis type 1 treated with Etanercept

Abstract:

Introduction: Mucopolysaccharidosis type I (MPS I) is a prototypic lysosomal storage disease with a spectrum of disease severity. Diagnosis of MPS I is based on demonstration of deficient activity of lysosomal enzyme α-L-iduronidase, and/or identifying mutations in the IDUA gene. We present a patient recently diagnosed with MPS I who had improvement in her dysostosis multiplex joint symptoms when being treated with etanercept for a presumed diagnosis of polyarticular Juvenile Rheumatoid Arthritis (JRA). Case: K.G. is a 12 year old female who had been diagnosed at age 7 with Noonan syndrome, based on her history of cardiac valve abnormalities, short stature, and suggestive facial features. Sequencing of Noonan gene panel identified a variant in the SOS1 gene which was felt to confirm her diagnosis. At 7 years old she was noted to have decreased range of motion in her shoulders bilaterally and stiffness in a number of other joints. She was diagnosed with JRA for which she was treated with a combination of medication, including etanercept, and physical therapy. When she presented to the NMCSD genetics clinic at age 9, an alternative explanation for her features was pursued because even the combination of Noonan and JRA did not explain her phenotype. An arthritis survey showed small humeral and femoral heads, developmental abnormalities of her bilateral clavicles, and arthritic changes throughout that were not consistent with her diagnoses. Additionally, the patient’s unaffected mother carried the same SOS1 variant, and it has since been shown to be a common benign polymorphism in Hispanic individuals. The patient was lost to follow up for 2 years during which time she developed bilateral knee contractures and worsening claw deformity of her hands. She underwent steroid injections in her fingers to help with mobility and had both legs casted for the contractures and discontinued the etanercept due to maternal concerns about the JRA diagnosis being incorrect. When the patient began experiencing pain and inflammation in her knees, the etanercept was restarted with significant improvement in her knee pain. Improvement in her cervical spine extension and range of motion was also noted. The patient returned to genetics clinic at age 12 with coarsening of her facial features, macroglossia, hepatomegaly and no significant increase in height. These new features suggested a storage disorder and MPS type I was confirmed through biochemical and molecular testing. Discussion: Current treatment for MPS I includes enzyme replacement therapy...
with laronidase, hematopoietic stem cell transplantation for severe MPS I, low impact physical therapy, surgical management for contractures and decompression for nerve entrapments. Animal studies suggest that dermatan sulfate is an endotoxin-like molecule that incites an inflammatory response via the tumor necrosis factor pathway. Etanercept is a competitive inhibitor of TNF-α. Our patient, due to the misdiagnoses of JRA, was treated with etanercept for her joint swelling and pain but had a clinical improvement reported by the patient, her parents and her physicians. To our knowledge, etanercept has never been reported to treat skeletal manifestations of MPS I. The benefit to this patient with etanercept suggests a new area of research and treatment for MPS associated dysostosis multiplex.
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Abstract Title: Improving Teamwork in the NICU: The Effects of TeamSTEPPS® Training

Abstract:

Introduction: Teamwork is essential to optimal patient care and patient safety in the NICU. Prior research has shown that participation in teamwork training can improve performance and patient safety in the clinical environment. TeamSTEPPS® (Team Strategies and Tools to Enhance Performance & Patient Safety) is a teamwork training course developed by the Department or Defense and the Agency for Healthcare Research and Quality. The TeamSTEPPS® curriculum focuses on five essential areas of teamwork including; team structure, leadership, situational monitoring, mutual support, and communication. TeamSTEPPS® is widely considered to be the national standard for medical team training. TeamSTEPPS® training has recently been mandated at all Army Medical Centers. Here we report the changes in the teamwork attitudes, knowledge and skills of our NICU staff after participation in TeamSTEPPS® training. Methods: Physicians, nurses and respiratory therapists at Tripler Army Medical Center participated in a TeamSTEPPS® training course. Measures of teamwork attitudes, knowledge and skills were obtained before and after the TeamSTEPPS® course using a prospective pretest-posttest design. Teamwork attitudes were evaluated using the TeamSTEPPS® Teamwork Attitudes Questionnaire (TAQ). Teamwork knowledge was evaluated using the TeamSTEPPS® Learning Benchmarks (LB). Teamwork skills were evaluated via medical simulation. The simulation involved a neonatal resuscitation scenario and used an event-based approach to training, wherein physicians were directed to request an incorrect dose of epinephrine and nurses were directed to provide slow and ineffective compressions. Teamwork during the simulations was measured using the TeamSTEPPS® Team Performance Observation Tool, a behaviorally anchored rating scale that rated teamwork skills on a scale from 1 (very poor) to 5 (excellent). The willingness of the nurses to speak up and challenge the incorrect medication dose and resuscitation leader’s correction of ineffective compressions were recorded. Results: Forty-two participants completed the training, including 29 nurses, 10 physicians (4 attendings, 6 fellows) and three respiratory therapists. Attitudes towards teamwork on the TAQ improved significantly from the pretest to the posttest (P <0.001). Participant teamwork knowledge scores on the LB improved significantly after the TeamSTEPPS® course (pretest 86.8% vs. posttest 92.6%; P <0.001). Observed teamwork behaviors in the simulation exercise improved significantly after the TeamSTEPPS® training. Improvements were seen in team structure (pretest 2.5 vs. posttest 4.2; P <0.001), leadership (pretest 2.6 vs. posttest 4.4; P <0.001), situation...
monitoring (pretest 2.5 vs. posttest 4.3; P <0.001), mutual support (pretest 2.9 vs. posttest 4.3; P <0.001) and communication (pretest 3.0 vs. posttest 4.4; P <0.001). The incorrect dose of epinephrine was challenged by the nurses in 38.4% of simulations before the training and in 76.9% of simulations after the training (P= 0.063). Before the training, fellow’s ordering an incorrect dose were challenged 55% of the time but no attending neonatologists were challenged (OR = 11). After the training, fellows were challenged 77% of the time and attendings were challenged 75% of the time (OR = 1.1). Inadequate compressions were corrected by the resuscitation leader 61.5% of the time before the training and 84.6% of the time after the training (P=0.248). Discussion: We were able to demonstrate significant improvements in the teamwork attitudes, knowledge and skills of our NICU staff after participation in TeamSTEPPS® training. The odds of a nurses speaking up and challenging an attending’s incorrect order also increased. To our knowledge this is the first report of learning outcomes after TeamSTEPPS® training in the NICU. We feel these short term learning outcomes are important and will translate to improved teamwork within our unit. To fully evaluate the effectiveness of the TeamSTEPPS® training we plan to monitor clinical outcomes in our NICU as part of our continuous quality improvement efforts.
Abstract Title: Bundled Clinic-based Intervention Improved 2-dose Flu Vaccine Compliance

Abstract:

Background: The burden of influenza is significant; 250-500,000 deaths worldwide occur annually as a result of this virus. It is especially problematic in the pediatric population. Review of US data from the past 30 years reveals an average of approximately 100 deaths per year in patients less than 19 years old. Thus, the Advisory Committee on Immunization Practices recommends annual influenza vaccination for all children aged 6 months through 18 years of age. Children from 6 months through 8 years who have not previously been immunized against influenza are specifically advised to receive a 2-dose vaccine series during their first season of influenza vaccination. Prior research has demonstrated that while two doses of vaccine decreased the number of office visits for influenza-like illness (ILI), one dose of vaccine had no effect on visits for ILI. Average national compliance with the 2-dose recommendation is less than ideal. A Colorado retrospective analysis estimated overall compliance at 37%. A 2006 assessment of an outpatient population found rates varying between 29-54% in children 6-23 months old, while older children, 2-8 years, had even poorer compliance (12-24%). Methods: The aim of this project was to determine if clinic-based interventions could improve observance of the 2-dose flu vaccine recommendation for patients less than 2 years of age. The bundled intervention was three-fold and included provider education, reminder cards, and phone call reminders. The provider education session included physicians and reiterated the flu vaccination recommendations for all children, with specific emphasis on the 2-dose recommendation and its merits. The reminder cards were given to patients’ families at the time of their initial vaccination indicating the need for a second immunization in one month’s time. Finally, families were called at least 1 month from administration of their initial flu vaccine to remind them of the need for a second dose. The E-immune immunization database was used to analyze 2-dose vaccine compliance rates for patients up to 2 years old who received their first flu vaccine from October 2010–February 2011. Results: Our clinic population’s compliance initially mirrored the national population at 37.5%. There was a statistically significant improvement in the 2-dose flu vaccine compliance rate after implementation of the bundled intervention (53.8%, p<0.005). 8% of children did NOT receive a phone reminder (due to wrong or unlisted number) and therefore received only a 2-part bundle. The overall rate of compliance in those patients was similar to that of patients prior to the implementation of the bundle. This suggests that the phone call was the most influential aspect of the bundle. Conclusion: This process improvement project demonstrated that a
A clinic-based intervention comprised of physician education, reminder notes, and phone follow-up resulted in a statistically significant improvement in the 2-dose flu vaccine compliance rate for children less than 2 years of age. Data suggests that the reminder phone call had the highest yield of all components of the intervention though further research is required to definitively assess the impact of each individual element.
Abstract Title: Screening for Vitamin D Deficiency in Children with Suspected Non-Accidental Fracture

Abstract:

Introduction: Debate exists regarding the need for routine measurement of vitamin D levels in evaluating a child with suspected non-accidental fracture. Prior studies have shown that serum vitamin D levels predict fracture risk only if rickets is clinically suspected, as the risk of fractures due to vitamin D deficiency is unlikely without other clinical signs of rickets. Other studies have demonstrated that the differences between rickets and fractures from abuse can be subtle, and both diagnoses can co-exist. Given the increasing incidence of vitamin D deficiency in breastfed children and the medical legal implications of an inappropriate diagnosis of child abuse, some providers at our institution have measured vitamin D levels in children with a suspected non-accidental fracture. Our study evaluates these vitamin D levels and compares them to their provider’s clinical suspicion for vitamin D deficiency. Objective: To determine if routine screening of vitamin D levels after suspected non-accidental fracture detects vitamin D deficiency and changes clinical outcomes. Methods: After IRB approval, we retrospectively reviewed all skeletal surveys performed at Tripler Army Medical Center (TAMC) in the last 10 years and selected the children who were evaluated for suspected non-accidental fracture. We determined if a 25-hydroxyvitamin D [25(OH)D] level was requested for these patients and characterized their provider’s clinical suspicion for vitamin D deficiency as high or low. Per the 2010 Institute of Medicine Report and 2011 Endocrine Society Guidelines, we defined vitamin D deficiency as a 25(OH)D level of less than 20 ng/mL. Finally, we calculated the prevalence of children with low 25(OH)D levels whose providers had low clinical suspicion for vitamin D deficiency during their evaluation. Results: 396 children had skeletal surveys done at TAMC from November 2000 through July 2011. 99 were performed after identifying a suspected non-accidental fracture. Of these patients, 11 children ages 1 to 7 months had 25(OH)D levels requested. For children whose providers had a low pre-test suspicion for vitamin D deficiency, the prevalence of vitamin D deficiency was 12.5% (95% binomial CI 0.003-0.524, 1 of 8 cases [Table 1]). These results indicate that at least 1 out of every 300 children evaluated for non-accidental fracture could have vitamin D deficiency despite a low clinical suspicion by their provider, although the actual rate is likely much higher given that we found 1 in 8 cases. The child we identified with a low Vitamin D level whose provider had no suspicion for
rickets was treated with Ergocalciferol (Drisdol®) but continued to be evaluated for abuse.
Conclusion: Routine screening of vitamin D levels after non-accidental fracture may detect vitamin D deficiency in children for whom there is low clinical suspicion. The clinical relevance of these low vitamin D levels is not clear. As our population receives greater than average sun exposure, the rate of deficiency in children with suspected non-accidental fracture may be greater in other areas.
Table 1. 25-hydroxyvitamin D levels and clinical suspicion for vitamin D deficiency.

<table>
<thead>
<tr>
<th>Age</th>
<th>Fractures</th>
<th>Clinical Suspicion</th>
<th>25(OH)D</th>
</tr>
</thead>
<tbody>
<tr>
<td>8 months</td>
<td>Humeral Torus</td>
<td>Low</td>
<td>40</td>
</tr>
<tr>
<td>7 months</td>
<td>Posterior Ribs</td>
<td>Low</td>
<td>73</td>
</tr>
<tr>
<td>3 months</td>
<td>Multiple Ribs, Tibial Diaphysis, Multiple CMLs</td>
<td>Low</td>
<td>QNS *</td>
</tr>
<tr>
<td>10 weeks</td>
<td>Complex Skull, Multiple Ribs, Ulnar Metadiaphysis, Radial Torus</td>
<td>Low</td>
<td>33</td>
</tr>
<tr>
<td>6 weeks</td>
<td>Femoral Diaphysis, Multiple CMLs</td>
<td>Low</td>
<td>&lt; 4</td>
</tr>
<tr>
<td>2 months</td>
<td>Anterolateral 6th rib</td>
<td>Low</td>
<td>52</td>
</tr>
<tr>
<td>2 months</td>
<td>Multiple Ribs, Radial and Ulnar Diaphyses, Ulnar Torus, Multiple CMLs</td>
<td>Low</td>
<td>24</td>
</tr>
<tr>
<td>1 month</td>
<td>Tibial Diaphysis, Tibia CML</td>
<td>Low</td>
<td>40</td>
</tr>
<tr>
<td>7 months</td>
<td>Femoral Diaphysis</td>
<td>High</td>
<td>31</td>
</tr>
<tr>
<td>3 months</td>
<td>Femoral, Humeral, and Ulnar Diaphyses</td>
<td>High</td>
<td>19</td>
</tr>
<tr>
<td>10 weeks</td>
<td>Diffuse Metaphyseal Changes</td>
<td>High</td>
<td>&lt; 5</td>
</tr>
</tbody>
</table>

* No evidence of rickets on remainder of labs. No further workup done.
Abstract Title: The Asynchronous Local/Overseas Hospital Academic (ALOHA) system: A Tele-Education Platform for Graduate Medical Education

Abstract:

Introduction: It is increasingly challenging to meet Graduate Medical Educational (GME) requirements due to a combination of geographic, regulatory, and educational factors. The ACGME’s (Accreditation Council for Graduate Medical Education) resident physician work hour restrictions and military deployments of teaching staff have severely limited educational opportunities, leading to significant variation in trainee experience as well as competency in primary and subspecialty education. To address these constraints, our goal was to create a tele-educational portal within our established asynchronous telemedicine system to meet our institution's ACGME competency-based training requirements. Methods: Our Asynchronous Local/Overseas Hospital Academic (ALOHA) system provides educational content, lectures, morning report, reading materials, quizzes in an asynchronous format for use by our pediatric trainees. This approach ensures a core curriculum is available to all trainees and also provides Residency Program Directors with a measure of accountability and competence. The ALOHA system is secure, HIPAA-compliant, and simply requires an internet connection and standard browser to run on any device. The ALOHA system is based on the Sakai course management software which provides the infrastructure for the virtual learning environment. Sakai is a widely accepted, open-access educational platform used at over 160 institutions worldwide chosen after review of other course management systems for a variety of reasons including open source architecture with a large number of customizable features, ease of deployment with current DoD software, and no cost or ongoing financial obligation. To capture our didactic lecture activities we created a simple recording system that captures audio and digital video input that automatically compresses and transfers the data to the ALOHA database. Rather than purchasing costly new video recording hardware for each conference room our software-based approach leverages our existing equipment in each conference room (microphone, computer, and overhead projector), thereby minimizing cost. The ALOHA system reduces demands on clinical teaching staff who otherwise must provide duplicate educational activities for both day and night resident physicians and also provides lectures from subspecialists who are unavailable due to deployments. This ensures that resident physicians on night shift or on a rotation at an outside facility can access these valuable educational materials at anytime. The General Pediatrics ALOHA course was initiated in April, 2009 and currently has 115 users with approximately 100 site visits per week. This
course routinely captures our daily morning report and lecture series, providing an instant curriculum for our night shift resident teams. In addition there are currently four other ALOHA courses including Primary Pediatric Care, Pediatric Cardiology, Mother Baby Nursery, Neonatology, and Neonatology Fellowship. Results: The system has proven to be user friendly and presents only a small learning curve for course administrators (staff) to start up and run their own ALOHA page. Our current ALOHA system provides residents and staff with online readings, downloadable lectures, online resources, interactive websites, and printable PDF files. This system has aided in our ongoing practice to meet Graduate Medical Educational (GME) requirements. Conclusion: The ALOHA system has become a valuable educational resource for our Pediatric Residency training program. The asynchronous approach and extensive system functionality allows us to leverage our limited resources while simultaneously meeting ACGME training requirements. Sustainability of this system is supported by its ease of use, automated data handling, and open-source architecture. We anticipate that the ALOHA system will serve as a model for other military and civilian residency training programs. This platform could also be utilized in the future for staff physicians to earn continuing medical education (CME) credits.
Abstract Title: “Lights, Camera, Action: Increasing the Use and Documentation of Asthma Action Plans in a Pediatric Outpatient Setting”

Abstract:

Background: Asthma is the most common chronic illness of childhood. Despite improvements in therapy there continues to be increasing morbidity associated with this illness. In 2002, the National Heart Lung and Blood Institute published “The National Asthma Education and Prevention Program Expert Panel Report Three,” a summary of the most recent national guidelines for asthma care. Although the guidelines exist, and have been shown to decrease morbidity and mortality for asthmatics, there is poor compliance with these guidelines by medical providers. In order to assess how our clinic measured up to the national guidelines of asthma care, we identified one aspect of recommended management, the use of Asthma Action Plans (AAP), and sought to assess our clinic’s compliance. Aim: Our aim was to identify the current use and documentation of AAP in our clinic and to improve the compliance of our clinicians with the national guideline. Hypothesis: We hypothesized that our clinic’s compliance would be poor and that we could improve our clinic’s compliance through a multifactorial intervention. Methods: We performed a chart review of all patients seen in our clinic during the months of February and March of 2010, for an acute or routine asthma visit. We assessed each chart for documentation of the creation or discussion of an AAP. We assessed each chart for documentation of the creation or discussion of an AAP. During the month of January 2011 we implemented a multifactorial intervention to improve compliance. First, we posted flyers in the clinic’s waiting area and exam rooms to remind parents to discuss AAP during their child’s visit. In addition, we placed colored copies of AAP in brightly colored folders in patient exam rooms to make the AAP more accessible to providers. We conducted two morning-report teaching sessions on how providers could use our electronic medical record to document discussion and distribution of the AAP. Finally, we sent an email summarizing the interventions and step by step instruction on electronic medical record documentation. After these interventions we then performed another chart review in February and March of 2011 to assess the impact. Results: In February and March 2010, 56 patient charts were identified as having a diagnosis of Asthma. Nine of the 56 (16%) charts identified had documentation of either discussion or creation of an AAP. In February and March of 2011, after our intervention, we identified 43 charts with the diagnosis of Asthma and found that 16 (37%), \( \chi^2=5.758; P <0.02 \) of the charts identified had documentation of either discussion or creation of an AAP. Conclusion: We concluded that with a multi factorial approach, we were able to nearly double compliance with
national treatment guidelines for asthma care regarding AAPs to 37%. The current national average of compliance with AAP usage is estimated at 33%. These intervention strategies can continue to be utilized in our clinic but can also be applied to other aspects of asthma care in which our clinic could improve upon. We will continue to explore additional means to further increase our clinic’s compliance with the national asthma guidelines.
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Abstract Title: The Primary Care Behavioral Health Initiative (PCBHI) : A Pilot Program to Improve Access to Behavioral Health Care Services on the Front Lines of Operation Iraqi Freedom

Abstract:

Introduction: The provision of optimized behavioral health care in the deployed environment continues to be impeded by many barriers. In this primary care driven pilot program the unit surgeon, physician assistant and all 25 combat medics in a battalion sized unit were trained in basic behavioral health competencies. The US Army has one of the most robust and well resourced behavioral health systems in the world. However, hundreds of Soldiers commit suicide each year and tens of thousands of Soldiers live with undiagnosed and untreated behavioral health issues that impact the combat effectiveness of their units as well as their individual long term health and quality of life. An alternative approach that was piloted in Iraq during OIF 09-10 leverages the existing health care personnel available within a battalion sized unit – it’s Surgeon, Physician Assistant and the battalion medics. Program Description: The pilot PCBHI program consists of the following components: --An awareness campaign advertising the availability of behavioral and emotional health care services within the framework of existing primary care access points (line medics, squadron aid station). --Five one hour blocks of training for all medics focusing on the prevalence of behavioral health issues among Soldiers and their Families, how to establish a trusting and professional relationship with Soldiers, how to conduct a strengths based psychosocial interview including screening questions on the status of a Soldier's emotional and behavioral health, more in depth screening for common behavioral health issues, decreasing stigma, resources available to Soldiers and Families, and a cumulative practical exercise. --Adapting aid station policies so that most Soldiers who present to a medic or provider receive, within the confines of a trusting professional relationship, psychosocial screening for emotional and behavioral health issues. --Conducting a leader professional development session for all Squadron E-7s and above to establish leadership buy in and awareness of the existence of the pilot program. --Eliciting from soldiers how they view their Family is adapting to their deployment and providing resources to support their Family. --Close coordination with the Squadron Chaplain and Brigade Behavioral Health Officer and direct referral to these resources when warranted. Results: Figure # 1 – Data showing the number of Soldiers receiving behavioral health diagnoses through the PCBHI mechanism in the 1-14 CAV operational environment from 1 Aug 2009 to 15 June 2010. Numbers represent initial diagnosis visit only. Two behavioral health evacuations for Soldiers with suicidal
ideation did occur during this time period. PCBHI Rollout Across USD-North: The 1-14 CAV Squadron’s PCBHI was presented to the USD-N medical planning conference in Dec 2009. A mandate was generated during the conference to develop a train the trainer program based on an enhanced version of the 1-14 CAV’s PCBHI that could be delivered to battalion physician assistants and senior medics and trained down to all medics at the battalion levels across the USD-North’s operational environment. Conclusion: In all 17 physician assistants and 81 medics were trained from April through June 2010 at main hubs across USD-N including FOB Warhorse, FOB Warrior, FOB Marez, FOB Sykes, and COS Speicher. The PCBHI effectively transcends many of the barriers that exist by significantly enhancing Soldier’s access to behavioral health prevention, early identification, and treatment services by existing unit level medical professionals.
<table>
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<tr>
<th>Diagnosis</th>
<th>Number Seen</th>
<th>1-14 CAV</th>
<th>Attachments</th>
<th>Tenant Units - Caldwell</th>
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</thead>
<tbody>
<tr>
<td>Deprssion</td>
<td>28</td>
<td>23</td>
<td>2</td>
<td>3</td>
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<tr>
<td>Anxiety</td>
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<td>25</td>
<td>2</td>
<td>6</td>
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<td>PTSD</td>
<td>5</td>
<td>2</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Sleep Disorder</td>
<td>20</td>
<td>12</td>
<td>8</td>
<td></td>
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<tr>
<td>Attention Deficit Disorder</td>
<td>6</td>
<td>6</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adjustment Disorder</td>
<td>17</td>
<td>14</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Anger Management</td>
<td>5</td>
<td>4</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Combat Operational Stress Reaction 2</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>116</strong></td>
<td><strong>88</strong></td>
<td><strong>6</strong></td>
<td><strong>22</strong></td>
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</tbody>
</table>
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Abstract Title: Development and Distribution of the Military Youth Deployment Support Video Program (MYDSVP)

Abstract:

Introduction: The Military Youth Deployment Support Video Program (MYDSVP) consists of two developmentally appropriate videos that were funded and developed by the American Academy of Pediatrics and the United States Army Medical Command (MEDCOM). Development and Funding of the MYDSVP: Military Youth Coping with Separation: When Family Members Deploy and Mr. Poe and Friends Discuss Family Reunion After Deployment were originally developed and funded by the Uniformed Services West Chapter of the American Academy of Pediatrics. An initial grant of $20,000 was awarded to the Chapter which was used to make a pilot of the program. MEDCOM approved and funded the project with an additional $30,000. MEDCOM then funded an initial reproduction and distribution of the program in 2007. The cost to reproduce and distribute an initial 100,000 copies of the program was $96,000. Reproduction of the videos was done by Premiere Media Solutions. All shipping costs were handled by the Department of Defense through their contract with Military One Source. The first 100,000 copies were completely distributed by Nov 2007. A second set of 100,000 copies was approved by MEDCOM and funded with another $96,000 for reproduction and distribution in Jan 2009. In March 2010 the 200,000th and final copy of the program was distributed. Program Description: In Military Youth Coping With Separation Cameron, a teen whose family experienced deployment, guides viewers through candid interviews with other military youth. The interviews capture true feelings and coping strategies of military youth. Teens interviewed advise their peers to listen to the deployed parent rather than the media or to avoid watching the news completely. They also encouraged others facing the same issues to speak to someone. Recommended sources of support are family members, friends or trusted adults, such as a teacher or their doctor. In Mr. Poe and Friends cartoon characters talk about deployment issues. The animated host, Mr. Poe, interacts with families at the airport as they await the return of their deployed loved ones. The video features the voices of real military children, parents, and youth serving professionals who have experienced deployment. Both videos are available for online viewing on the American Academy of Pediatrics Deployment Support Web site at www.aap.org/sections/unifserv/deployment/index.htm. Results: • A recent analysis of the program's distribution data shows the following utilization patterns: 61% Army, 12% Air Force, 11% DOD Civilian, 10% Navy, 4% Marine Corps, 2% other • Component – 67% Active Duty, 15 % National Guard, 12% Reserve, 6% other • The program has been requested by families and agencies from every state in the country plus Guam, Puerto Rico, the Virgin Islands and multiple foreign countries via APO/FPO addresses • Top states utilizing the program include TX – 13.5%,
CA-8.4%, NY-5.9%, WA- 5.8%, VA-4.9%, NC-4.6%, AE (Armed Forces Europe, Canada, Middle East, Africa) – 3.4% The MYDSVP been recommended on the Army Behavioral Health website, is supported as a best practices within the Army Family Covenant operations guide, was officially endorsed by the American Academy of Pediatrics through its committee on Psychosocial Aspects of Child and Family Health, forms a critical component of the Army’s Battlemind program for families, and was distributed by request to all families attending the National Military Family Association’s Operation Purple Camps during the summer of 2009. Conclusion: The MYDSVP was developed on the front lines by pediatricians to support the healthy emotional and behavioral development of military children and adolescents during potentially difficult times in their lives. The program helps to decrease feelings of stigma and isolation in military children while sensitizing the larger American community to military child and adolescent culture and support needs.
Abstract Title: Development of the Military Child and Adolescent Center of Excellence (MCA CoE)

Abstract:

Introduction: When warriors are assured that their family is being cared for by the community they are fighting to protect, they are able to more clearly focus on the critical combat and sustainment operations they are performing far from home. It is likely that children who grow up in a well functioning and emotionally connected military family are more likely to consider military service as a viable career choice. Military youth live the concept of service and sacrifice every bit as much as their military parent. They are expected to take on more advanced family roles when family members are away. They may live in environment where a parent is absent – either physically or emotionally. For all of the reasons, the Military Child and Adolescent Center of Excellence (MCA CoE) was established in July of 2008. Background: The MCA CoE was established to address the child and adolescent specific mandates issued within the the Army Family Covenant, the DoD Mental Health Task Force Recommendations, the American Psychological Association Task Force Recommendations and the American Academy of Pediatrics Prioritized Legislative Recommendations for military children. Events Leading to the Establishment of the MCA CoE: - March 2005 - American Academy of Pediatrics Healthy People 2010 Chapter Grant of $20,000 awarded to the Uniformed Services West Chapter of the AAP to explore "Deployment Effects on Child and Adolescent Mental Health" - March 2006 - Development of the AAP Deployment Support Website www.aap.org/sections/uniformedservices/deployment/index.html Establishment of the CoE: In 2008 as a direct result of collaboration between uniformed pediatricians and child and adolescent behavioral health specialists, the United States Army Medical Command approved and funded the establishment of the Military Child and Adolescent Center of Excellence (MCA CoE). The vision of the Center was to ensure the optimal physical, emotional, and behavioral well-being of military children and adolescents worldwide through comprehensive, youth-centered, strengths-based program development and provision of technical assistance to military youth-serving professionals of all disciplines. Initial Budget for the MCA CoE: $3.5 Million – appropriated for facility acquisition, personal, travel and coordination with other agencies, distribution of the Military Youth Deployment Support Video Program, and development of next generation support programs. Workproducts of the MCA CoE: Two national Summits on Military Children in June 2008 - Military Child and Adolescent Behavioral Health and Well-Being During Wartime and Beyond and June 2009 - Frontline Responders: Coming Together for Military Youth – work products available at http://www.mamc.amedd.army.mil/MilitaryChildSummit/SummitPresentations.htm Major Divisions of the MCA CoE: School Based Health Initiative Division – a combination of the best features of the Army’s primary care and behavioral school based health programs. The final program will provide
for the primary health care needs of military dependent children and adolescents while incorporating heavy behavioral and emotional health screening and resilience based strategies unique to the stresses that military children face. Program Development Division- development of phase II of the Military Youth Deployment Support Video Program – focusing on children/adolescents whose parents have physical/emotional injuries along with other military child and adolescent unique programming. Program Analysis Division - evaluation of the School Based Health Initiative, MYDSVP Evaluation – Phase I and II, Assembly of Best Practices/Best Value Programs for Army children and adolescents, management of a military child/adolescent specific research agenda, and consultation to other institutions to ensure inclusion of military child/adolescent sensitive subject matter expertise in their projects. Conclusion: After many years of advocacy work by both uniformed pediatricians and behavioral health specialists, the Military Child and Adolescent Center of Excellence was established in July 2008. The Center has gone on to become the Army’s Child, Adolescent, and Family Behavioral Health Office (CAF BHO).