

CLINICAL VIGNETTE ABSTRACTS
Poster Session I
Thursday, October 23, 2104

BASIC IBD

4 **AUTOIMMUNE HEMOLYTIC ANEMIA IN A CHILD WITH ULCERATIVE COLITIS.** I. Monteiro, Pediatrics, Rutgers New Jersey Medical School, Newark, New Jersey, UNITED STATES. J. Tamaroff, Rutgers New Jersey Medical School, Newark, New Jersey, UNITED STATES.

Abstract: Autoimmune hemolytic anemia (AIHA) develops in 1.7% of patients with Ulcerative Colitis (UC) at an average age of 50.5 years and onset between colitis and AIHA diagnosis is typically 17 months. Upon review of the literature, one previous case in the pediatric population was reported with both of these diseases. We present a child with history of UC who presented with AIHA and UC exacerbation. Case Report: 11-year-old male with no significant past history was diagnosed with UC 4 months earlier and responded to mesalamine. At presentation his hemoglobin (Hb) was 9.2 and MCV 77. Five days prior to this admission he presented to an outside hospital with fever, headache, vomiting, and pallor and was discharged from ER on acetaminophen. At home, he stopped taking mesalamine and developed bloody stools, weight loss, anorexia, and jaundice. Patient has no history of transfusions or tattoos. Physical exam was significant for delayed capillary refill, icterus and pallor. Pertinent labs: Hb 7.6, reticulocyte 16.5, MCV 92.5, total bilirubin 6.3, direct 0.3, LDH 1127, haptoglobin <10, ESR/CRP 135/6, WBC 18.5. Direct Coombs was positive with warm autoantibody. Hepatitis panel, ANA, dsDNA, G6PD, sickle cell were all negative and folate, B12, anticardiolipin, immunoglobulins, and C3/C4 were all within normal limits. Imaging showed gallbladder sludge and top normal sized spleen of 10.9cm. During admission patient was restarted on mesalamine and bloody stools remitted. Hemoglobin trended from 7.6 to a nadir of 6.9 with reticulocyte count of 35.6. At this time, patient was started on prednisone and Hb improved to 8.6 as did jaundice and pallor. His Hb normalized at 2 weeks post discharge and he was continuing to do well. Discussion: This case indicates that AIHA and UC may also be found in pediatric patients. A patient with UC presenting with anemia is often assumed to have iron-deficient anemia from blood loss or a vitamin-deficient anemia from anorexia and malnutrition. If this is not the case, a full workup for hemolytic anemia should be undertaken. Reported cases in adults required the use of cyclosporine or infliximab for UC associated AIHA but in this case prednisone was adequate. Further study to determine if pediatric hemolytic anemia associated with UC is more amenable to treatment with steroids than in adults is needed to better understand how to treat these patients. Conclusion: Though UC is known to be associated with AIHA in adults, in children, it may be uncommon but, if diagnosed early, could be more amenable to steroid therapy.

5 **ACUTE PSYCHOSIS IN A CHILD WITH CROHN'S DISEASE FOLLOWING INFLIXIMAB THERAPY.** S. Lapsia, J. Difalco, J. Morganstern, M. Choi, A. Chawla, Pediatric Gastroenterology, Stony Brook Children's Hospital, Stony Brook, New York, UNITED STATES.

Introduction: Psychiatric disorders are seen with greater prevalence in Crohn's disease compared to the general population. Anxiety, depression, and panic disorders are well described in the literature and are thought to be associated with the degree of physical morbidity in these patients. Acute psychosis and a schizophrenia-like disorder have been reported in four adults on etanercept, a tumor necrosis factor alpha (TNF- α) antagonist. We describe a case in which infliximab, a chimeric monoclonal antibody against TNF- α , was associated with acute psychosis in a child with Crohn's disease. Case Report: The patient is a 15 year old male who was maintained on 6-mercaptopurine (6-MP) and mesalamine for 3 years. Due to poor weight gain, worsening abdominal/joint pain, and persistent colitis on colonoscopy, 6-MP was discontinued and infliximab therapy was initiated. Four weeks after his third infliximab dose was given, the patient presented to the ER with an acute onset of agitation, confusion, auditory and visual hallucinations, and aggressive behavior warranting anti-psychotic medication. His past medical history was significant for obsessive compulsive disorder (OCD) diagnosed at age 9, for which he took fluoxetine for 3 months. No further psychological concerns were noted since then and he was an honor student. There was no family history of mental disorders. Work-up to rule out organic causes of acute psychosis including urine toxicology, electroencephalogram, and brain MRI were all negative. After a few days of observation, the patient was discharged home on haloperidol. Over the next two weeks, the patient's behavior improved back to baseline. Two months after his last infliximab dose, he was given another dose of infliximab and the following day, he began to exhibit disorganized behavior and combativeness once again. At that time, he was prescribed

olanzapine, an atypical antipsychotic, with good results. Following the second psychotic episode, infliximab was discontinued and 6-MP was resumed along with budesonide to treat his Crohn's disease. One and one half months later, he remains on an olanzapine wean with no evidence of psychosis. Discussion: Research has shown that TNF- α is among the cytokines released in neonatal and post-traumatic brain injury and theoretically, its inhibition should reduce excitotoxic brain injury. However, studies done on injured neonatal rat brains show that TNF- α can have both beneficial and deleterious effects. Although this patient's past history of OCD may have increased his susceptibility to develop psychotic behavior, the temporal relationship between infliximab use and his psychiatric disturbance suggests this medication is a strong etiologic factor for his psychosis. To date, past psychiatric history has not been a consideration towards the use or avoidance of infliximab. Given our case report and the adult experiences with etanercept, further vigilance should be paid towards significant past psychiatric history as well as acute onset of behavioral changes.

6 GASTROINTESTINAL TB: IS IT COMING BACK TO CONFUSE THE DIAGNOSIS OF CROHN'S?. M.V. Mendoza, D. Akerele, A. Shane, G. Tenjarla, Emory University, Atlanta, Georgia, UNITED STATES.

Tuberculosis is a major worldwide health problem where 95% of global cases annually affect impoverished and overcrowded areas with poor sanitation. It is second only to HIV/AIDS as the biggest killer worldwide due to a single infectious agent. In developed countries, its incidence has increased due to immigration, HIV infection, immunosuppressive therapies, and emergence of multi-resistant *M. Tuberculosis* (MTB) strains. While it most commonly affects the lungs, tuberculosis can affect any part of the body, including the abdomen. As such, it can mimic many gastrointestinal diseases, among them Crohn's disease. We present a 16yo Korean-American female with a four month history of diarrhea and unexplained weight loss. Her initial endoscopic evaluation was significant for severe ileocolitis with deep serpiginous ulcers throughout the colon while sparing the sigmoid and rectum. A presumptive diagnosis of Crohn's disease was made. Prior to steroid administration, additional history revealed that she had received BCG and had recently visited Korea. Also, her father had been diagnosed with and completed a six month course of therapy for pulmonary tuberculosis. We withheld steroid therapy and began a workup for intestinal tuberculosis infection. An interferon gamma release (IGRA) assay was sent. Although asymptomatic from a pulmonary standpoint, a chest x-ray demonstrated bilateral apical densities. She underwent a bronchoscopy and repeat upper endoscopy and colonoscopy to obtain sputum, gastric aspirates and intestinal biopsy specimens for acid fast bacilli (AFB) stain and culture. Her IGRA result was positive with appropriate response to positive and negative controls suggesting past exposure to MTB. AFB were detected via stain and culture from cecal tissue biopsies. All health care providers involved in her care prior to diagnosis received a tuberculin skin test, and all returned negative for MTB. Infectious Disease was consulted and patient was started on daily therapy with isoniazid, rifampin, pyrazinamide, and ethambutol for two months followed by 4 months of rifampin and isoniazid, along with pyridoxine to prevent symptoms related to B6 deficiency associated with isoniazid. After two weeks of therapy, our patient reported resolution of her diarrhea and a four pound weight gain. A contact investigation of potentially exposed healthcare workers, family members, and school contacts was performed and her brother and mother had evidence of latent tuberculosis infection while the remainder of the contact investigation was without identification of additional infections. Highlights of this case: Although less common in resource-rich countries, intestinal TB must always be in the differential diagnosis of Crohn's disease, especially in immigrants from countries with high TB prevalence. AFB stain and culture of intestinal biopsies are the mainstay of the diagnostic work up in intestinal TB. Finally, a thorough history is the most important aspect of any diagnostic workup, despite convincing evidence from labs, imaging, and endoscopic evaluation.

CELIAC/EOE/ALLERGIC ENTEROPATHY

23 EOSINOPHILIC GASTROINTESTINAL DISORDER PRESENTING AS SEVERE ANEMIA AND PROTEIN-LOSING ENTEROPATHY AND HYPOGAMMAGLOBULINEMIA BUT WITHOUT GASTROINTESTINAL SYMPTOMS. M. Patel, A. Balan, M. Altaf, OUHSC, Oklahoma City, Oklahoma, UNITED STATES.

Introduction: Eosinophilic gastrointestinal disorders (EGID) are rare disorders that represent eosinophilic infiltration of various areas of the gastrointestinal (GI) tract. One of the more well known manifestations is that of eosinophilic esophagitis which usually presents as dysphagia. Symptoms in EGID other than eosinophilic esophagitis are usually non-specific but always include some GI symptoms. We report a young toddler who presented with severe anemia, hypoalbuminemia, hypogammaglobulinemia, and protein-losing enteropathy (PLE); subsequently diagnosed with EGID. Case Presentation: We report a 15-month-old who initially presented with

severe anemia and hypoalbuminemia. Family denied any gastrointestinal manifestations. Physical exam was unremarkable except for pedal edema. Peripheral smear revealed iron deficiency anemia and eosinophilia. An extensive immunological and infectious workup was negative except Enterovirus and HHV6 viremia; which further led to the diagnosis of hypogammaglobulinemia. These could have contributed to his anemia but did not explain his chronic symptoms. He received multiple Albumin and IVIG infusions over the next few weeks for consistently dropping levels. Urine was negative for protein but fecal Alpha-1 Anti-trypsin was elevated suggestive of PLE and possible etiology for hypogammaglobulinemia. His upper endoscopy and colonoscopy revealed increased eosinophils throughout the GI tract suggestive of EGID; and likely cause of his PLE. Subsequent to this, he was placed on a cow's milk restricted diet and started on oral Prednisone. He had remarkable improvement of his symptoms as well as his hemoglobin and albumin within 3-4 weeks. He has since been weaned off of systemic steroids and is being maintained on a dairy restricted diet and oral Budesonide with complete resolution of anemia and hypoalbuminemia. Discussion: EGID is a rare disorder with less than 300 adult and pediatric cases reported. It is characterized by increased numbers of eosinophils throughout the GI tract. Diagnosis is made based on histological evaluation. Symptoms are usually nonspecific but always include some GI symptoms. Mucosal involvement can lead to protein losing enteropathy, fecal blood loss, and malabsorption. With so few cases reported in the literature, there is no consensus regarding treatment, but include immunosuppression, Azathioprine, or elimination diet. EGID is characterized by a relapsing and remitting course. Our case had EGID causing anemia and PLE leading to hypoalbuminemia. We have been able to maintain this child on cow's milk restricted diet and oral budesonide.

24 PROTEIN LOSING ENTEROPATHY IN A WELL THRIVING INFANT. N. Eltawil, pediatrics, cleveland clinic, Cleveland, Ohio, UNITED STATESR. Gupta, pediatric gastroenterology, cleveland clinic children's hospital, cleveland , Ohio, UNITED STATES.

A previously healthy 15 month old female presented with a 2 week history of bilateral periorbital edema. No history of oliguria/ anuria, new onset allergies, conjunctivitis or acute onset diarrhea. No history of jaundice, fever, breathing difficulty or any new drug intake. The swelling was not present at any other part of body and was most prominent during morning hours. There was history of milk protein allergy for which the child was on hydrolyzed formula till 1 year of age and was switched to regular milk at that time. Since last few months, she was passing 3-5 daily soft stools without any associated blood. She was consuming around 25 ounces of regular milk with other age appropriate foods and was gaining good weight. Her initial work up revealed a serum protein (L) 3.2 g/dL and albumin (L) 1.7 g/dl, hemoglobin (L) 9.3 g/dl, with normal serum transaminases, a normal urinalysis with no evidence of proteinuria, WBC (H) 14.86 with 7.9 % eosinophils. Fecal alpha-1 anti-trypsin level (H) 602 and a normal serum IgE cow's milk allergen test. She underwent an endoscopy and colonoscopy which showed mild active colitis and duodenal eosinophilic mediated injury. She received one albumin infusion, and her cow's milk formula was changed to hydrolyzed formula again. 3 weeks later her serum albumin level was 4.0 g/dL(N), periorbital edema had disappeared and her bowel movements were twice daily. In this case, despite the child gaining adequate weight and having no apparent diarrhea or blood in stool, the child had developed protein losing enteropathy. Cow's milk protein allergy is not a common cause of protein losing enteropathy, especially in children who are thriving well otherwise. Knowledge of this association can help detect these children early before they become hypoalbuminemic and symptomatic.

25 CASE OF LOSS OF RESPONSE TO EOE THERAPY DUE TO DEPLETED MDI MEDICATION.. B. Cunningham, T. Heifert, C. Sullivan, K. O'Meara, M. Goldman, Dept. of Pediatrics, Uniformed Services University, Bethesda, Maryland, UNITED STATESB. Cunningham, T. Heifert, P. Rogers, C. Sullivan, K. O'Meara, M. Goldman, Dept. of Pediatrics, WRNMMC, Bethesda, Maryland, UNITED STATES.

Introduction: Eosinophilic esophagitis (EoE) is a disorder of esophageal mucosal inflammation, characterized by eosinophilic infiltration. EoE can cause a multitude of symptoms from feeding difficulties and poor weight gain in infants to dysphagia and abdominal pain in older patients. In this case, an adolescent male with EoE suddenly redeveloped symptoms of abdominal pain and dysphagia after having been well for 2 months following diagnosis and the initiation of swallowed fluticasone. This case is unique in that nothing changed regarding the patients' diet, medications, family, social life. However, upon further investigation it was found that the patient continued to use the original inhaler he had been prescribed 2 months earlier. The patient denied skipping doses, and maintained that he continued to "taste" the medication with use of his metered dose inhaler (MDI). A literature review

showed no previous case reports of loss of response to EoE therapy due to depleted medication in an MDI. Case Description: A 14-year-old male presented to the pediatric gastroenterology clinic with a two year history of epigastric abdominal pain improved with acid suppression. He denied any dysphagia, weight loss or association with any particular foods; however, large meals were a significant trigger. Physical exam was unremarkable, and laboratory findings demonstrated a mild eosinophilia but normal celiac screen, fecal calprotectin, inflammatory markers and complete metabolic panel. The patient was placed on a proton pump inhibitor (PPI) and had an esophagogastroduodenoscopy) which demonstrated furrowing and trachealization of his esophagus. Biopsy histopathology of his esophagus noted greater than 50 eosinophils / HPF at three biopsied levels of his esophagus. He was diagnosed with EoE and started on a regimen of two 220mcg swallowed fluticasone doses daily administered using an MDI, and his PPI was weaned over the next 2 weeks. His symptoms resolved completely within in 10 days of starting the fluticasone, and he continued to do well until 3 months later when he returned for follow-up and noted return of his previous symptoms. By report the patient had not replaced his MDI because he continued to 'taste' something when he used the inhaler. Given that the typical MDI has 120 metered doses, he had redeveloped symptoms at 150 doses and was likely tasting only propellant for about a month. He was given a new fluticasone MDI, with instructions to replace it after the counter reached 0 or after 120 uses. He was again symptom-free after 2 weeks and continues to do well. Discussion: This case highlights a potentially dangerous situation where patients may assume that the MDI is continuing to provide medication because of the taste or feel of propellant. Patients should be instructed to bring their inhalers to follow-up visits especially if there is an unexpected recurrence of symptoms. This case emphasizes the need for proper, concise instruction regarding the use of MDI's, with particular emphasis on replacement timing.

26 LONG TERM OF BUDESONIDE IN SEROSAL EOSINOPHILIC GASTROENTEROPATHY. C. Rudman, A. Pelissier, M.J. Integlia, H. Shashidhar, New Hampshire Hospital for Children, Elliot Hospital, Manchester, New Hampshire, UNITED STATES.

Case #1 5 year old Caucasian male was admitted with a history of intermittent vomiting for 2 months with a recent onset abdominal distention and diffuse abdominal pain. Passage of mucous per rectum and tenesmus were described in particular. Laboratory evaluation at admission was notable for peripheral eosinophilia and hypoalbuminemia (WCC 28K, Eos 74%, Serum Albumin 2.8). Stool cultures, gram staining, ova, parasite were negative. An abdominal CT scan obtained showed small bowel wall thickening and ascites. An ascitic fluid analysis showed predominant eosinophilis (1760 cell count, 88% eosinophils with SAAG 0.5). Upper GI and sigmoid biopsies were unremarkable with the exception of esophagitis with 20 eos/hpf and basal zone hyperplasia. Prednisone therapy was initiated for a diagnosis of serosal eosinophilic gastroenteropathy and continued for 7mos. Developmental and behavioral issues precluded use of elemental diet. He was switched to budesonide with the lowest dose tolerated 6mg/day, 5x a week and remained on this medication 2 years to date. Several attempts to taper resulted in recurrent symptoms and peripheral eosinophilia. He has required rescue courses of oral prednisone on four occasions. Case #2 10 year old Caucasian male was admitted for two week history of nausea, vomiting, abdominal pain and watery diarrhea with an outside abnormal abdominal CT scan. PMH revealed intermittent vomiting for several months. Laboratory evaluation showed eosinophilia and hypoalbuminemia. (WCC 16K, eos 46%, S. Alb 3.1). Stool cultures, ova and parasites, gram stain as well as inflammatory markers, fecal calprotectin were negative /normal. CT scan showed diffuse small and large bowel wall thickening and ascites. Paracentesis confirmed eosinophilia (1724 with 80% eosinophils and SAAG 0.6). Upper GI and sigmoid biopsies showed >40 eos/hpf in esophagus and microabscess formation and focal intense eosinophilic infiltration in colon and rectum. He was started on prednisone with immediate improvement and tapered to 5mg/day prior to a subsequent switch to budesonide 6 mg daily at 6 weeks. He has remained in remission 10 m on budesonide 6mg qd. Parents are reluctant to attempt elemental diet. Discussion Serosal form of eosinophilic gastroenteropathy is the least common presentation. This diagnosis requires an index of suspicion. Atypical features including intermittent prior symptoms, peripheral eosinophilia, and presence of ascites during an episode of acute enteritis should prompt evaluation for this disease. Peritoneal fluid tap is diagnostic. Periodic exacerbations are invariably accompanied by peripheral eosinophilia with prompt response to oral prednisone. Budesonide, a long and locally acting oral corticosteroid appears to be an effective alternative to oral prednisone in the long term treatment. Very little published data exists to define optimal therapeutic options.

ENDOSCOPY/POTPOURRI

52 PLEXIFORM SCHWANNOMA PRESENTING AS AN UPPER GASTROINTESTINAL BLEED. N.M.

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The most common causes of upper gastrointestinal (UGI) bleeding in children are ulcers, esophagitis, gastritis, and varices. Gastric masses are uncommon in pediatric patients. We present a rare case of an UGI bleed caused by a perigastric mass in a child. A 15 yo female with a two year history of abdominal discomfort presented to the emergency room with two weeks of intermittent melena. Physical examination revealed tachycardia and orthostatic hypotension. Her abdominal and perianal exam were benign. She denied nausea, vomiting, hematemesis, or weight loss. Her hemoglobin was 5.3 g/dL. An esophagogastroduodenoscopy (EGD) revealed an erythematous antrum. Histopathologic examination of biopsies revealed esophagitis, with normal gastric and duodenal mucosa. She was started on a proton pump inhibitor. Three months later, she again presented with melena. A repeat EGD revealed a heaped up and erythematous antrum. The pylorus was intubated with difficulty. Retained food particles in the stomach despite prolonged fasting were visualized again. Histopathologic examination of biopsies revealed esophagitis, nonspecific reactive changes of the gastric antrum, and normal duodenal mucosa. Magnetic resonance imaging revealed circumferential wall thickening of the gastric antrum and pylorus with associated narrowing of the pyloric channel, nodular omental infiltration with thickening and draping over the gastric wall and upper abdomen, and gastrohepatic ligament lymphadenopathy. Histopathologic examination of a CT guided core needle biopsy of the omentum revealed moderately cellular spindle cell tumor with cells arranged in whorls and fascicles with focal palisading and Verocay body formation. Immunohistochemical stains revealed diffuse S100 positivity and low Ki67 proliferative index. Other tumor markers were negative. The patient underwent resection of the tumor. Surgery included a hemigastrectomy and omentectomy with reconstruction using a Bilroth II gastrojejunostomy, without disruption of biliary flow. Histopathologic examination of the resected specimen confirmed a plexiform schwannoma involving the submucosa, muscularis propria, and serosa of the stomach, duodenum, and omentum. Schwannomas are benign slow-growing myelin sheath tumors of neurogenic origin. Plexiform schwannoma is a rare benign variant composed exclusively of Schwann cells arranged in a plexiform pattern, and differs from the typical gastric schwannoma by its multinodular to multilobular composition. Most plexiform schwannomas are solitary, slow growing, asymptomatic skin tumors, and visceral localization is rare. Intramural growth of such tumors can compromise the overlying mucosal blood supply, leading to ulceration with blood loss. In adults, the most common presentation is an UGI bleed. Superficial upper endoscopic biopsies are usually of low diagnostic yield. To our knowledge this is the first case of a plexiform schwannoma in the stomach, duodenum, and omentum and the first case of a plexiform schwannoma presenting as an UGI bleed in a child.

53 GASTROJEJUNAL TUBE ELIMINATED CHRONIC RUMINATION IN DEVELOPMENTALLY DISABLED CHILDREN. A.F. Severio, J. Monagas, R. Noel, P.E. Hyman, Louisiana State University Health Science Center, New Orleans, Louisiana, UNITED STATES. A.F. Severio, J. Monagas, R. Noel, P.E. Hyman, Children's Hospital of New Orleans, New Orleans, Louisiana, UNITED STATES.

Rumination is effortless regurgitation of food, with the subsequent spitting out or re-chewing and re-swallowing the food. In developmentally delayed children rumination is a pleasurable, treatment-resistant, self-stimulating habit. We evaluated and treated two non-ambulatory, non-verbal five year old children, one female with Down's Syndrome and one male with Phalen-McDermind syndrome who ruminated frequently while awake for three years or more. Both mothers of these children were highly motivated to find a solution because the children were not growing and had become social pariahs. We hypothesized that rumination would cease if the stomach stayed empty of food. We intervened by placing gastro-jejunoscopy tubes using the Kimberly-Clark MIC* transgastric jejunal feeding tube gastropexy introducer kit. Then we initiated continuous drip jejunal tube feedings over twelve waking hours to reduce hunger and eliminated the oral diet. In both cases these changes reduced episodes of rumination by > 90%. Both children had rumination episodes of only swallowed saliva and gastric secretions. To

entirely eliminate rumination we gave noxious tasting products, n-acetyl cysteine or cayenne based hot sauce, through the gastrostomy tube every four hours while awake. Within a week of adding the noxious tasting agents, all rumination ceased. After two months of no rumination, one mother restarted oral feeding. There was no recurrence of rumination. Three months after gastro-jejunal tube placement the tube was removed. In one year of follow-up, rumination did not recur. The second mother was pleased with the outcome with jejunal feedings and chose not to resume oral feedings. These results showed that jejunal tube feeding in combination with insertion of unpalatable contents into the stomach may eliminate rumination in developmentally delayed children.

54 ENDOSCOPIC CLOSURE OF A GASTRO CUTANEOUS FISTULA WITH OVER THE SCOPE CLIP WITH ENDOSCOPIC ULTRASOUND GUIDANCE.. M. Haight, C. Huang, R. Gugig, M. Ament, Pediatric Gastroenterology, Children's Hospital Central California, Madera, California, UNITED STATES. M. Haight, R. Gugig, Pediatrics, UCSF-Fresno, Fresno, California, UNITED STATES.

The over-the-scope-clip system (OTSC; Ovesco Endoscopy GmbH, Tuebingen, Germany) is an endoscopic device for the mechanical compression of large areas in the gastrointestinal tract. The OTSC system has been used in adults for hemostasis of postsurgical bleeding and closure of iatrogenic intestinal perforations. Closure of gastrointestinal tract fistulas using this device has also been described in adults. Gastrocutaneous fistulas rarely close spontaneously and are related with sepsis and persistence of symptoms if not closed properly. Their management is often difficult and ineffective with secondary cellulitis at the external drainage site. All previous reports in the literature have been with adult patients. A 15-year-old with acute lymphoblastic leukemia developed a gastrocutaneous fistula from a retroperitoneal abscess that eroded into the greater curvature of the stomach. We present the successful endoscopic closure of the fistula with endoscopic ultrasound guidance using the OTSC system. Fistula closure was successful and documented by imaging and endoscopic examination. Endoscopic application of the OTSC device was safe and effective for the treatment of a gastrocutaneous fistula. This case adds to the literature demonstrating a role for OTSC clip in the closure of gastrocutaneous fistula in pediatric patients. We believe that OTSC application system is a safe and effective endoscopic method for the successful treatment of gastrocutaneous fistulas.

55 A 4-YEAR OLD BOY WITH A COIN IN THE DISTAL ILEUM. A. Alper, M.G. Patel, A.F. Porto, Yale University, New Haven, Connecticut, UNITED STATES. V. Fox, S. Fishman, E. Tracy, Harvard University, Boston, Massachusetts, UNITED STATES.

Introduction: Foreign body ingestion is a common problem in the pediatric population with up to 75% of cases occurring in children below the age of 4 years. Coins account for up to 70% of the cases. Most of them pass spontaneously through the gastrointestinal tract without complications. A coin impaction usually occurs at one of the sites of anatomic constriction in the esophagus. Bowel perforation and obstruction are the most common significant complications reported in adults. Others include bleeding, fistula and abscess formation, zinc deficiency, sideroblastic anemia and nickel-induced dermatitis. There is limited data regarding the clinical manifestations, complications and mode of treatment of small bowel impaction of coins in children. Here we report a rare case of a small bowel obstruction at a site of meckel's diverticulum, secondary to a nickel ingestion. Case report: A 4-year old boy with no significant past medical history, presented with mild abdominal pain and constipation one month after an accidental ingestion of a coin. Physical examination was unremarkable. Radiographic examination with abdominal x-ray showed the radiopaque shadow of a coin at the right lower quadrant. Although the exact location could not be determined, it was presumed to be within the small bowel. He was followed in the clinic for several months and was mostly asymptomatic. Despite extensive bowel clean out attempts, repeated abdominal x-rays showed no significant change in the coin's location. A small bowel follow through exam with contrast material showed partial obstruction of the distal ileum, likely a result of the coin failing to pass through the ileocecal valve. An endoscopic intervention with both a colonoscopy and a retrograde enteroscopy failed to reach the site of obstruction. Finally, a laparoscopy was performed and revealed an impacted coin in a site of a meckel's diverticulum, within one foot of the ileocecal valve. It was surrounded by some knotted loops of bowel secondary to local adhesions. The coin was removed and the diverticulum was resected. Postoperative course was unremarkable. Conclusion: Impaction of foreign bodies at the small bowel is relatively rare and mostly involves smaller objects that have succeeded in passing the pylorus and ligamentum of Treiz. Although considered to be harmless, blunt objects such as coins can be associated with severe morbidity and mortality, especially with prolonged intestinal retention time. Clinical manifestations do not always correlate with distal bowel impaction's

complications. This rare presentation of a partial small bowel obstruction secondary to coin ingestion in an asymptomatic child highlights the importance of earlier rather than later surgical intervention. Furthermore, a small bowel impaction that fails to respond to conservative treatment should also raise a concern for an underlying anatomical pathology.

56 ESOPHAGEAL STENT PLACEMENT AS A THERAPEUTIC OPTION FOR IATROGENIC ESOPHAGEAL PERFORATION. A. Alsafadi, R. Grothe, I. Absah, Department of Pediatric Gastroenterology and Hepatology, Mayo Clinic, Rochester, Minnesota, UNITED STATES. R. Grothe, L.M. Wong Kee Song, I. Absah, Department of Gastroenterology and Hepatology, Mayo Clinic, Rochester, Minnesota, UNITED STATES.

Background: Iatrogenic esophageal perforation (IEP) is a potential adverse event of interventional endoscopy for esophageal stenosis, with an incidence ranging from 1 to 10%. With repeated endoscopic dilatation the risk of perforation, bleeding, and aspiration increases. Management of IEP varies from early surgical intervention to conservative management for small contained perforations. There is paucity of data in the small pediatric population regarding utilization of esophageal stents in managing IEP. Case Report: An 18 month old girl was born with a congenital mid-esophageal stricture that measured 10 mm in length and 6 mm in diameter. The stricture was refractory to multiple balloon dilatations. An attempt at dilation using Savary dilators was complicated by an IEP measuring 1x 3 cm in size. The child remained stable immediately post IEP without evidence of pneumothorax or subcutaneous emphysema. A 16 mm wide x 70 mm long Alimaxx-ES fully covered stent was placed under both endoscopic and fluoroscopic guidance to divert secretions and avoid surgery. The patient was admitted for observation and IV antibiotics. She tolerated the stent well and was discharged after 24 hours on a liquid diet. The stent was removed 2 weeks later with complete healing of the perforation site and a remarkably patent lumen at the site of prior stenosis, measuring 1.2 cm in diameter. Conclusion: In select cases, esophageal stent placement is a feasible and successful alternative to invasive surgery for the management of iatrogenic esophageal perforation in small children.

57 BEWARE OF HOLISTIC TREATMENT DURING ENDOSCOPY!. N. Rawal, A. Safta, S. Blanchard, Pediatric GI, Hepatology and Nutrition, University of Maryland, Baltimore, Maryland, UNITED STATES.

Background: Holistic medicine, including dietary supplements, are increasingly being used in both eastern and western countries, as primary or adjunctive therapy, to treat or provide symptomatic relief for various medical conditions. Patient surveys have reported that 12% of Americans, 12% of Australians, and 4.8% of patients in the UK use herbal remedies. Even though there have been studies supporting the beneficial effects of these medications in multiple conditions, the data remains insufficient to conclude that benefits outweigh the risks. These supplements are not regulated by FDA standards and hence, it is important to recognize the side effects and drug interactions of these herbal remedies. Case Description: We report a case of a 8 year old Indian male with Crohn's disease, who underwent an upper endoscopy and colonoscopy for evaluation of abdominal pain and blood in the stool. At the time of procedure, he was on 50% enteral nutrition therapy for treatment of Crohn's disease, along with aminosalicylate, metronidazole and ciprofloxacin. At the time of the procedure, he had a normal complete blood count (CBC). During endoscopic evaluation, he had excessive bleeding after each biopsy. The number of biopsies was kept to minimum for diagnostic purposes to prevent excessive hemorrhage. Bleeding sites did not require any endoscopic intervention. Such bleeding with biopsies was not noted at his initial diagnostic endoscopy. His CBC remained stable. After the procedure, parents revealed that they were also giving herbal supplements, ginger, garlic and ginseng to treat his colitis. Herbal supplements can have stimulant or depressant effect on the central nervous system (CNS). They are also known to interact with most commonly used drugs in anesthesia and potentially lead to alterations in serum glucose level or increased risk of bleeding. Because of these risk factors, American Society of Anesthesiologists recommends discontinuing all herbal medications 2 to 3 weeks prior to an elective surgical procedure. Several studies have reported on increased risk bleeding with the use of ginger, garlic and ginseng. Ginger is said to have anti-inflammatory properties, but also is a potent inhibitor of thromboxane synthetase enzyme that can prolong bleeding time. Garlic, even though considered beneficial for cardiovascular diseases and diabetes, inhibits platelet aggregation. Ginseng is a CNS stimulant, but also inhibits platelet aggregation, especially when used in combination with NSAIDs and warfarin. In addition to these, alfalfa, feverfew, Echinacea, dong quai root, willow bark, ginger, guarana, horse chestnut and goldenseal are also reported to cause coagulation problems. Gastroenterologists should be aware of these supplements so that we can prevent increased risk of bleeding during endoscopic procedures. Conclusion: With this case report, we intend to

emphasize the importance of including questions about use of herbal supplements, in addition to documenting their current medications. Most patients don't volunteer information about herbal supplements. All herbal supplements should be discontinued 2 to 3 weeks prior to elective endoscopic procedures to prevent complications.

58 POSTPOLYPECTOMY ELECTROCOAGULATION SYNDROME: A PEDIATRIC CASE REPORT. J.R. Rick, D.P. Shover, Gastroenterology, Dayton Children's Hospital, Dayton, Ohio, UNITED STATES; J.R. Rick, D.P. Shover, Pediatrics, Wright State University, Dayton, Ohio, UNITED STATES.

Background: Endoscopic polypectomy is a relatively safe procedure, however, potential life-threatening complications can occur. The most serious are perforation and bleeding. Also, electrocoagulation during polypectomy, can result in postpolypectomy electrocoagulation syndrome (PPCS). First appearing in the medical literature in 1981, it refers to the development of abdominal pain, fever, leukocytosis, and peritoneal inflammation in the absence of frank perforation after polypectomy with electrocoagulation. Only 1 suspected pediatric case could be found. In 1992, a child developed abdominal pain in the absence of perforation after polypectomy. The authors did not label the case as PPCS and patient was treated conservatively. In the adult literature, the reported incidence varies widely with a range of .003 to .01 percent of colonoscopies and 0.5 to 1% of polypectomies. Risk factors for development of PPCS are large (> 2cm) lesions, sessile configuration, location in the right colon and hypertension. Incarceration of normal bowel mucosa by the snare and contralateral mucosal burn are suggested etiologies. Imaging is needed to ensure perforation has not occurred. Treatment is typically conservative with hospitalization, intravenous fluids, antibiotics, and nil per os. If symptoms are mild, outpatient treatment is often used. Roughly, 20% of adult patients with PPCS require hospitalization. Unlike perforation, there is little morbidity. We present a case of pediatric PPCS managed as an outpatient with an uneventful recovery. Case Report: A 15 yr. old female underwent esophagogastroduodenoscopy and colonoscopy for chronic diarrhea. A 1.5 cm pedunculated juvenile polyp was removed from the splenic flexure with snare polypectomy using blended current. There were no immediate complications. Abdominal pain developed and the next day she presented with fever to 102.5 F, periumbilical pain, and emesis. Physical exam showed left upper quadrant tenderness with guarding, but no rebound. Evaluation for another source of fever was negative. Upright and supine abdominal films showed no evidence of perforation. Outpatient management with clear liquids for 24 hours, anti-biotics (metronidazole and ciprofloxacin), and anti-emetics was rendered. Fever and pain resolved in 36 hours and she had an uneventful recovery. Conclusions: We report a case of PPCS in a 15 yr old. Unique aspects to this case are the patient's age, lesion location and polyp configuration. The resultant PPCS was managed as an out-patient. Review of the medical literature could not find an identified case of PPCS in the pediatric population. Possible reasons include under recognition, under reporting, and the relatively infrequent rates of both polypectomy and large sessile polyps in children. Other undetermined factors of the pediatric population may prevent the development of PPCS in children. Further studies regarding PPCS in children are warranted with the aims of increased recognition to avoid unnecessary exploratory laparotomy, guide treatment and understand the factors contributing to its development.

59 SAVORY BOUGINAGE OF ESOPHAGEAL WEBS/STRICTURE OF A PATIENT WITH EPIDERMOLYSIS BULLOSA (EB). E. Grossman, Internal Medicine, Division of Digestive Disease, SUNY Downstate Medical Center, Brooklyn, New York, UNITED STATES; Saghier, R. Nathan, S. Rabinowitz, S. Schwarz, Pediatric Gastroenterology, SUNY Downstate Medical Center, Downstate Children's Hospital, Brooklyn, New York, UNITED STATES.

Introduction EB is a genetically inherited condition associated with esophageal strictures as well as esophageal webs with an increased vulnerability of the esophageal mucosa to perforation. Purpose: To demonstrate that bougie dilatation in EB patients may be a safe and effective alternative therapy for severe esophageal stenosis, when balloon dilatation is not feasible. Case Report: We report an 18-year-old female with EB who presented with food impaction, dysphagia and poor weight gain (BMI = 15). Methods: Barium swallow showed both a partially obstructing esophageal web and a 4mm luminal narrowing at the level of the proximal cervical esophagus. Endoscopy (EGD) demonstrated a web at 16 cm from incisors, which did not permit passage of a 5.9 mm OD endoscope. The working channel in this ultra-thin endoscope (2.0mm diameter) could not accommodate a balloon dilator. Using Savory dilators over a guide wire under fluoroscopic guidance, the esophageal web was gently ruptured and the stenotic segment was serially dilated from 18Fr to 21Fr. Subsequently, the endoscope was able to traverse the dilated esophageal narrowing which revealed additional web remnants at 19cm. After the

procedure, the patient experienced some improvement in her dysphagia. Repeat EGD at one week and three weeks showed a persistent narrowing at 16 cm, which required serial Savary dilations from 21 Fr to 27 Fr (first session) and 27 Fr to 33 Fr (second session). After dilation to 33 Fr, a standard gastroscope was able to traverse the narrowed esophageal segment. Three weeks later, balloon dilations, employing a standard (OD 8.8mm) endoscope, was performed from 10mm to 12 mm (36 Fr). No complications were encountered during these procedures, and the patient reported immediate improvement in dysphagia. Conclusion: In selected EB patients with severe segmental esophageal narrowing precluding balloon dilation, gentle and progressive bouginage under fluoroscopy may be safely performed.

60 SUCCESSFUL NON SURGICAL TREATMENT OF PYLORIC STENOSIS IN A PATIENT WITH TRISOMY 18. L. Diaz Calderon, Medical Education, Miami Children's Hospital, Miami, Florida, UNITED STATES. J. Reeves-Garcia, E. Hernandez, R. Arboleda, R. Gomara, W. Muinos, Pediatric Gastroenterology, Miami Children's Hospital, Miami, Florida, UNITED STATES.

Background: The optimum treatment for infantile hypertrophic pyloric stenosis is the Fredet-Ramstedt pyloromyotomy. Conservative medical treatment is an alternative for some patients who are unsuitable or high risk for surgery with a reported success rate of 88%. We present a 6-week-old female with Trisomy 18 and congenital heart disease with pyloric stenosis who was successfully treated with atropine. Case report: The patient presented with 1-week history of NBNB projectile vomiting. She was malnourished with global developmental delay and severe feeding disorder secondary to marked hypotonia. An ultrasound of the abdomen revealed pyloric stenosis with channel measuring 1.6cm with a thick wall of 0.4mm in transverse dimension. Pyloromyotomy and gastrostomy tube placement were recommended. It was elected not to proceed due to high risk for anesthesia, parental desire to avoid invasive procedures and do-not-resuscitate status given the child's medical prognosis and condition. An oroduodenal tube was placed under fluoroscopic guidance in an attempt to bypass the obstruction. Trophic feeds were started, initially tolerated but rapidly discontinued due to continuous vomiting. Since pyloroplasty was refused and vomiting persisted, it was elected to treat the pyloric stenosis initially with Atropine IV and eventually per feeding tube. Patient was discharged home on oroduodenal tube feedings and atropine. Repeat pyloric ultrasound 5 months later was normal. At 1 year of age, parents agreed to proceed with surgical correction of a large VSD and gastrostomy tube placement. At follow up, the patient has remained well and gaining weight. Conclusion: Laparoscopic pyloromyotomy is the treatment of choice for infantile hypertrophic pyloric stenosis; however, atropine is an effective option for high-risk patients who are not surgical candidates.

61 CASE REPORT OF A CAUSTIC INGESTION. A.M. McClain, T. Hadley, K. Horvath, D. Mehta, Center for Pediatric Digestive Health and Nutrition, Orlando Health, Orlando, Florida, UNITED STATES.

Household cleaning products are known to cause detrimental injuries when ingested and is seen most often in young male children. We present a case of a caustic ingestion leading to life threatening changes in the GI and pulmonary anatomy. The patient was a previously healthy 20 month old male who was found drinking bathroom cleaner. At that time he was coughing, rubbing his tongue and having emesis and bloody oral secretions. Infant was immediately intubated in the ER and was first had bright red and then thin gelatinous heme positive material from the naso-gastric tube. On day two, he underwent endoscopy that showed Grade 3B, non-circumferential burns and sloughing of the esophagus with edema and narrowing at the gastro-esophageal junction. Nasal-jejenum tube was placed during a follow up EGD that showed gray lining of the esophagus and stomach but no strictures or tears. Feeds were started and infant started tolerating a clear liquid diet. On day 12, patient was found to have his NJ tube coiled in the esophagus; then had an acute oropharyngeal bleed and apneic episode. He was found to have a piece of accessory tissue obstructing his airway; he was intubated and emergent EGD showed mucosal sloughing. Later that day a bronchoscopy showed necrosis from the oropharynx to hypopharynx. When a 2.5cm linear tracheo-esophageal fistula was seen, about 1.5cm above the carina, an emergent laparotomy was performed. His stomach was found to be 80% necrosed and perforated during manipulation. The patient underwent removal of the stomach and ligation of distal esophagus at the hiatus. Afterwards he underwent a complicated ICU stay as a second fistula was found at the union of the trachea and the left main stem bronchus, necessitating a Ladd procedure and placement of duodenostomy. No improvement was seen in the two fistulas so he underwent cervical esophagostomy with a 14 French PEG tube. After 2 months in ICU, the patient was discharged with jejunostomy tube feeding and a pharyngostomy tube. Since then he has required esophageal dilations and further surgeries changing his anatomy [see diagram]. He has been having problems gaining weight and

most recently failed an OPMS but there is still hope for him in the future to be able to eat by mouth. Our patient ingested a drain cleaner which contained 34-50% sodium hydroxide [lye], 0-3% potassium hydroxide and 47-68% water. Of the yearly two million reported toxic exposures, about 9% of the cases are due to household cleaning products and esophageal burns account for the most serious injuries and chronic complications; pH below the threshold of 11.5-12.5 causes esophageal injury. Products with bleach have a pH of 9 to 11; these generally cause mild problems; our patient ingested drain cleaner that has a pH of 14. Experiments have shown that a 10% solution of lye needs a minute of contact to produce a deep burn whereas a 30% lye solution can cause transmural necrosis in one second of contact. After this initial necrosis, worsening destruction can happen over a week with inflammation and vascular thrombosis. Perforation is always a concern with initial insult; after 3 weeks the problem is strictures rather than perforation.

62 ENDOSCOPIC DIAGNOSIS OF COLONIC B-CELL LYMPHOMA IN A PEDIATRIC PATIENT. W. Muinos, R. Arboleda, E. Hernandez, R. Gomara, J. Reeves-Garcia, Department of Pediatric Gastroenterology, Miami Children's Hospital, Miami, Florida, UNITED STATES. Pichardo, L. Mullinax, Department of Medical Education, Miami Children's Hospital, Miami, Florida, UNITED STATES. Brathwaite, Department of Pathology, Miami Children's Hospital, Miami, Florida, UNITED STATES.

We describe a case of Colonic B-cell lymphoma in a pediatric patient whose clinical picture was similar to appendicitis; he presented with an acute history of fever accompanied by crampy right lower quadrant pain. No history of weight loss or change in bowel habits. His past medical history included DiGeorge's Syndrome as well as mediastinal B-cell Lymphoma, diagnosed a year earlier. It was thought that the patient was in remission at that time after chemotherapy. An abdominal US revealed bowel wall thickening with lymphadenopathy and the appendix was not visualized. A PET/CT scan of abdomen showed a lesion in right lower quadrant, concerning for recurrent lymphoma. Our patient underwent a diagnostic laparoscopy that showed a large amount matted lymph nodes and the biopsy obtained failed to provide a definite diagnosis other than reactive lymph nodes. Gastroenterology was consulted for a diagnostic colonoscopy to obtain tissue. Pathology from ileocecal region obtained at time of colonoscopy was positive for B-Cell Lymphoma. Colonic B-cell lymphoma is rare in pediatric patients. This is part of the differential diagnosis, especially in respect to this patient's past medical history. We provide high definition as well as narrow band images of the endoscopic appearance of colonic B-Cell Lymphoma. We also demonstrate that via colonoscopy we were able to obtain tissue for a definite diagnosis, when laparoscopy was unable to do so.

63 DYSPHAGIA IN AN ATOPIC 8 YEAR OLD: NOT ALWAYS EOSINOPHILIC ESOPHAGITIS ?. G. Naguib, H. Kader, Pediatric Gastroenterology, University of Maryland, Baltimore, Maryland, UNITED STATES. Shet, Radiology, University of Maryland, Baltimore, Maryland, UNITED STATES. Kaushal, Pediatric Cardiac Surgery, University of Maryland, Baltimore, Maryland, UNITED STATES. Pham, Pediatric Cardiology, University of Maryland, Baltimore, Maryland, UNITED STATES.

We present an 8-year-old female who carries a long-standing clinical diagnosis of gastroesophageal reflux disease (GERD) and asthma. She also has a history of atopic disease and was referred to Pediatric GI by her pulmonologist to exclude eosinophilic esophagitis (EoE). She reports dysphagia to solids occurring mostly with meats and green vegetables as she feels that food would get stuck in her throat when she swallows. Her mother has to cut up her food into small pieces. However, she denies any GER symptoms. She was started on omeprazole 2mg/kg/day by her pulmonologist 6 weeks prior to her GI consultation. After that consultation, she had an UGI that revealed a posterior esophageal impression suggestive of a vascular ring with a left aortic arch. CT of the chest confirmed an aberrant right subclavian artery (ARSA). Endoscopy was done on omeprazole 2mg/kg/day for evaluation of EoE at the request of cardiothoracic surgery to exclude esophageal mucosal disease. Endoscopy showed a posterior esophageal indentation caused by the known ARSA with normal gross and histologic examination. Dysphagia Lusoria is dysphagia secondary to an ARSA that has a retroesophageal course which compresses the posterior esophagus significantly enough to produce this symptom. ARSA usually does not produce symptoms as 60-80% of patients with this are asymptomatic throughout their lifetime. As such, ARSA is generally not considered by cardiologists or cardiothoracic surgeons to be a cause of eating difficulties. However, symptomatic patients require surgical intervention for relief of dysphagia and/or chronic respiratory disease such as chronic respiratory infections and dyspnea. Symptomatic disease is most likely to occur in infancy with spontaneous resolution of symptoms as the infant grows older. Conclusion: When symptomatic dysphagia occurs within the context of

extrinsic esophageal compression causing a partial obstruction and vascular assessment demonstrates this anatomic anomaly, further GI investigative evaluation for EoE is unlikely required in an atopic individual without specific signs and symptoms of GER/GERD. It may also prolong the definitive treatment while subjecting the patient to additional risks: general anesthesia, bleeding, and perforation. In our case, endoscopy did not provide additional diagnostic information. When patients are significantly symptomatic in the context of ARSA, surgical intervention should be performed. Surgical correction resulted in resolution of dysphagia in our patient.

64 UNUSUAL CAUSE OF RECTAL PROLAPSE IN A TEENAGE BOY: GIANT CONDYLOMA ACUMINATUM. G. Naguib, A. Malkani, S. Blanchard, Pediatric Gastroenterology, University of Maryland, Baltimore, Maryland, UNITED STATES. Strauch, Pediatric Surgery, University of Maryland, Baltimore, Maryland, UNITED STATES.

Giant condyloma acuminatum (GCA) is a very rare sexually transmitted disease affecting the ano-genital region. It is a slow growing cauliflower-like tumor, but unlike simple condyloma, it is locally aggressive and destructive with a high recurrence rate. Human papillomavirus, known to cause condyloma acuminata, is also known to induce these tumors. We present a 17-year-old African American male who presented with a large irreducible rectal mass and prolapse for several months duration. He had history of rectal bleeding and prolapse with painful bowel movements. He lost 15 pounds in 3 months as he was limiting his oral intake in order to decrease the number of bowel movements. He engaged in anal intercourse with a same sex partner. Perianal exam revealed a large cauliflower mass. Flexible sigmoidoscopy done under sedation revealed an exophytic mass that measured 8x4 cm. It was completely reduced and perianal exam was completely normal after reduction. He was referred to surgery and underwent transanal circumferential resection of 80-90% of the mass and a temporary colostomy. Complete initial resection was not possible due to proximity to the anoderm and anal sphincter in order to protect sphincter function and to prevent iatrogenic anorectal stenosis. He had his colostomy taken down after 3 months. He was HIV negative and pathologic diagnosis confirmed condyloma acuminata. During the following year, he required 2 more surgeries to complete resection and remove multiple smaller lesions. As the age of sexually active homosexual patients is decreasing, pediatricians and pediatric surgeons should recognize the presentation and treatment options of condyloma acuminata in children. Topical application of podophyllin works well in small lesions but rarely works for GCA. Complete excision is the preferred initial therapy when feasible. Local invasion and recurrence are the major source of morbidity in this disease and are very common. Malignant transformation without metastatic potential is reported in adults.

65 SMALL BOWEL AND COLONIC STRICTURE FORMATION FOLLOWING HEMOLYTIC UREMIC SYNDROME. N. Blondet, UT Southwestern, Dallas, Texas, UNITED STATES. A. Patel, UT Southwestern, Dallas, Texas, UNITED STATES.

Shiga toxin-producing *Escherichia coli* (E. coli) is known to be one of the main causes of hemolytic uremic syndrome (HUS) in previously healthy children. Of the secondary complications of HUS, colonic stricture is relatively rare. We report the case of a 2 y/o female that developed HUS following a diarrheal illness secondary to E. coli O157:H7. The patient was originally admitted at another facility due to bloody diarrhea. During her admission she developed renal failure requiring hemodialysis, and was subsequently discharged home with normalized renal function and no gastrointestinal symptoms. One month following discharge, she was readmitted due to bloody diarrhea, and was found to have *Clostridium difficile* colitis, treated with Metronidazole. Despite treatment, she continued to have intermittent abdominal pain and bloody stools, and was subsequently evaluated at our center, 2 months after original diarrheal illness. On presentation, abdominal X-ray revealed a large stool burden, for which she underwent an inpatient clean-out. During clean-out, she persistently had abdominal distension, abdominal pain and bloody stools. She underwent colonoscopy for further evaluation. During colonoscopy, a sigmoid stricture was visualized and subsequent contrast study revealed a focal sigmoid colon stricture. She underwent exploratory laparotomy for stricture resection, and during this examination a large inflammatory mass was visualized, involving the distal ileum and the sigmoid colon. Two short-segment strictures were resected, one in the sigmoid colon and one in the distal ileum, with primary anastomosis. Her postoperative course was uneventful with complete resolution of abdominal pain, constipation, and bloody stools. Review of literature revealed 16 cases of intestinal strictures following HUS, all of them involving the left-sided colon and none involving the small bowel. Clinicians caring for patients with history of HUS should be vigilant for symptoms of gastrointestinal obstruction, such as abdominal pain or constipation, and consider the possibility of small bowel and colonic stricture as the cause of such symptoms.

Case: A 5week old infant with persistent vomiting, poor weight gain and hoarseness since birth was subsequently diagnosed with reflux and milk protein allergy. Despite aggressive treatment, she remained symptomatic. She underwent rigid bronchoscopy and laryngoscopy that revealed reflux, subglottic stenosis, laryngomalacia and a red-purple mass at the esophageal introitus. This was suspicious for an esophageal duplication cyst. CT scan of the esophagus was normal but with an incidental finding of a gastric duplication cyst. Patient underwent laparotomy with resection of the cyst, which was adhered to the posterior gastric wall and distal tail of the pancreas. She developed stridor postoperatively and was evaluated by cardiology for possible tracheal ring. Work-up was negative for a tracheal ring, but an echocardiogram revealed a small PDA. Sonographic imaging of her right kidney showed mild fullness in the right collecting system with an extra-renal pelvis. Because of her multi-system anomalies, she was referred to genetics for further evaluation. Chromosomal microarray identified triple X syndrome. Discussion: Trisomy X, though normal at birth, is associated with several phenotypic variations. An incidence of 1 in 1000, only 10% of these patients have clinical manifestations. Patients are known to have tall stature, developmental delays, neurocognitive deficits, and psychological manifestations. Case reports have identified several cardiac and more commonly genitourinary malformations. Patients with trisomy X commonly have chronic abdominal pain and constipation. Rare cases describe esophageal, duodenal and jejunal atresias, anorectal malformations, cloacal exstrophy and omphalocele. To date, this is the only case that describes a gastric duplication cyst associated with triple X syndrome. It is possible to be unrelated and not a direct etiologic relationship. Further research needs to be performed to identify whether this is a causal relationship. However, once the diagnosis has been established for triple X syndrome, complete imaging studies should be performed to assess for possible structural abnormalities.

Salmonella is the most commonly identified cause of foodborne illness in the US. It occurs after ingestion of contaminated food products of animal origin (eggs, dairy products, poultry or ground meat). Along with Yersinia and campylobacter, Salmonella is one of the organisms reported to cause infectious ileocolitis in a few reported case series like DiLauro et al and Ngu et al. A 10 year old male presented with two days history of fever, vomiting, diarrhea, and right-sided abdominal pain. He has a past medical history of a few episodes of intermittent abdominal pain in the last year which was treated as constipation. His initial lab work showed WBC 14,000/uL with left shift, Hgb 12.8 g/dl, ESR 16 mm/hr, albumin 3.1 g/dl, and CRP 10.3 mg/dl. His CT abdomen showed abnormal circumferential thickening of terminal ileum and cecum with no abnormal inflammatory changes in the mesentery. The stool cultures and stool C.diff toxin PCR were negative at 48hrs. On day 3 of presentation, the EGD and colonoscopy showed exudates and multiple aphthous ulcers involving rectum, descending colon, transverse colon, and linear ulcers in the terminal ileum. We obtained biopsies from above abnormal areas which showed chronic active colitis and ileitis with no granulomas. Subsequently, his stool studies were positive for heavy growth of *Salmonella* sp. An 18 year old with known diagnosis of poorly controlled Type 1 diabetes due to non-compliance presented with a 2 week history of right-sided abdominal pain and vomiting. There was no history of fever, diarrhea, weight loss, melena, nor hematochezia. At admission, the labs revealed Hgb 10.2 g/dl, WBC 6,000/uL, albumin 2.7 g/dl, ESR 50 mm/hr, CRP 5.7 mg/dl. The CT scan showed moderate bowel wall thickening and surrounding inflammatory changes involving the terminal ileum and ascending colon. Stool cultures and stool C.diff toxin PCR were negative at 48hrs. Ileocecal biopsies showed acute ileitis and mild to moderate active colitis. Terminal ileitis found on endoscopy is most often Crohn's disease. However, it is known that infectious etiologies such as *Salmonella* should be considered. In the reported cases, the infectious causes were considered, though returned positive several days later. These cases highlight the need to not only send cultures, but consider waiting several days for results in some clinical scenarios. Given the clinical presentation of these two patients, repeat laboratory tests are required at a minimum to confirm that there is no inflammatory bowel disease after the acute infection. Moreover, future studies should elucidate if there is a positive association between infectious ileocolitis and Crohn's disease.

GERD/MOTILITY/FUNCTIONAL DISORDERS

86 SCLEROSING MESENTERITIS IN AN ADOLESCENT. S. Plasencia, R. Thomas, Medicine/Pediatrics, OU Tulsa School of Community Medicine, Tulsa, Oklahoma, UNITED STATES.

Sclerosing Mesenteritis is a rare inflammatory disorder of unknown etiology which involves the mesentery of the sigmoid and small intestine. It has been observed and described throughout medical literature by differing terminology due to its nonspecific presentation and variable degree of mesenteric lipoid fibro necrosis in middle aged patients. Often it is discovered incidentally by imaging studies conducted on patients presenting with a discrete palpable abdominal mass or nonspecific abdominal pain. Commonly the abdominal mass necessitates biopsy to rule out neoplasia. It has been suggested that hypodense zones along the periphery of the mass are characteristic of Sclerosing Mesenteritis. Complete removal is usually unnecessary as this disorder is commonly self limiting. The problem in question arises in cases which are not self limiting. Standards of care do not exist for this disease process which consequently leads to inconsistent medical management. A 19 year old female with a history of appendicular rupture and subsequent abscess formation at the age of 13. Following recovery from this event she experienced recurrent abdominal pain, constipation and vomiting spells. Multiple investigative surgical procedures were performed the following two years culminating in a diverting loop ileostomy complicated by functional/mechanical obstruction necessitating end ileostomy and mucus fistula with some ileal resection. Biopsies of mesentery and rectum were sent to pathology and found to be negative for neoplasia but identified peculiar lipoid necrosis and granulation tissue formation. A diagnosis of exclusion, Sclerosing Mesenteritis was designated as the most likely source. Our patient was then started on steroid therapy. Symptoms ultimately improved significantly when she was started on Azathioprine. Sclerosing Mesenteritis is a part of a spectrum of fibrotic and inflammatory disorders afflicting the mesentery. The obscurity of this disorder has been evaluated by only a few large case series. Due to its ambiguous presentation, nonuniform nomenclature, and lack of concrete treatment guidelines, practitioner strategies often amount to trial and error. Our case highlighted this point unfortunately. It is seldom observed in the pediatric population, owing to the lower percentage of mesenteric fat. This factor likely contributed to the circuitous treatment delivered as the treating specialists likely had limited experience with this disease process. We encountered our patient years after many corrective surgeries and medical regimens. She had attempted multiple different immunomodulator therapies including steroids. Azathioprine ultimately diminished abdominal pain and bloating symptoms. These symptoms were likely due to the mass effect observed in Sclerosing Mesenteritis. As the mesentery increases in size the bowel and vasculature become compressed leading to obstruction and rectal bleeding respectively. Our patient had numerous abdominal surgeries secondary to bowel obstructions and it is difficult to say if these interventions could have been avoided had she been treated with tailored immunosuppressive therapy sooner.

87 USE OF THE CONSTIPATION ACTION PLAN FOR MANAGEMENT OF CHRONIC CONSTIPATION. A. Hildreth, J. Rosen, Pediatric Gastroenterology, Hepatology, and Nutrition, Children's Mercy Kansas City, Kansas City, Missouri, UNITED STATES.

Background: Constipation is the most common digestive complaint among children, with studies showing prevalence as high as 29.6% and accounting for up to 25% of pediatric gastroenterology referrals. Constipation is a disorder that requires personalized management recommendations that consider age, development, symptom severity, and personal or social treatment barriers. Management guidelines are useful for the medical provider, but acute changes in symptom severity may require modification of therapy by the patient or caregiver to prevent emergency room (ER) encounters or hospitalization. We present a case of a 16 year old female with developmental delay and chronic constipation. The patient and her family had difficulty managing symptom exacerbation and were not satisfied with telephone consultation. Methods: A Constipation Action Plan (CAP) was created for the patient and family to follow at home. The CAP was modeled after an Asthma Action Plan as both disorders may require use of controller medications, knowledge of clinical identifiers defining exacerbation, and ability to alter therapy in the home setting based on symptom-severity. The patient was given a written CAP, with Green Zone as her baseline maintenance regimen, Yellow Zone for mild exacerbation, and Red Zone for severe exacerbation. We qualitatively reviewed her clinical course and compared the number of ER visits, clinic visits, and telephone calls that were primarily regarding constipation in the six months before and after initiation of her plan. Results: The patient had one ER and two clinic visits both before and after plan establishment. However, the number of telephone calls made for exacerbation of symptoms and recommendations on treatment decreased by 83% (from

six to one). She also experienced improved bladder control, with less nighttime awakening and decreased urine residual. The patient's behavior also improved, with increased quantity and quality of speech and better eye contact. All of these changes were attributed by her mother to improved pattern of defecation and decreased frequency and severity of symptom exacerbation. Qualitatively, her mother expressed a "huge improvement in quality of life" at their most recent follow up. She consistently cited the CAP as a useful tool that made her more comfortable managing her child's symptoms. Discussion: This case presents a novel strategy for management of chronic constipation. The CAP empowered a caregiver to confidently direct home therapy in the setting of acute symptom change. It decreased the number of healthcare encounters while improving patient symptoms and quality of life. Next steps for the CAP include initiation of a quality improvement cycle within a small cohort, and formal assessment of patient and healthcare provider satisfaction with the tool.

88 ESOPHAGEAL PAPILOMA: A RARE CAUSE OF DYSPHAGIA. S. Singhal, R.H. Alkhoury, A. Chapa, R.D. Baker, S.S. Baker, Women and children's hospital, University at buffalo, Amherst, New York, UNITED STATES. INTRODUCTION: Esophageal squamous papilloma (ESP) is a benign epithelial lesion typically seen in adults. We present a 2 year old boy with recurrent airway obstruction and dysphagia due to multiple squamous papillomas. CASE PRESENTATION: A 2 year old mediterranean boy, presented with noisy breathing. A nasopharyngeal endoscopy revealed multiple papillomas in the pharynx. They were excised with the microdebrider. Eight months later, he presented to our center with difficulty breathing and new onset dysphagia. Esophagogram revealed multiple filling defects throughout the esophagus. The patient underwent Esophagogastroduodenoscopy (EGD) which showed multiple esophageal papillomas. There were too many to remove, but biopsies were consistent with squamous papillomas. DNA probe analysis by insitu hybridization was positive for Human Papilloma virus (HPV) low risk types (6, 11, 42, 43, and 44). An uvulolaryngoscopy was also performed. Three papilloma from the uvula, larynx and hypopharynx were excised using the microdebrider. His respiratory symptoms improved after the excision of airway papillomas. Dysphagia improved over a three months period and a repeat EGD revealed fewer esophageal papillomas. His mother reported a history of multiple skin warts on her extremities and breasts while breast feeding. She did not have genital HPV on screening prior to delivery. Mother noticed spontaneous resolution of her warts over few months. DISCUSSION: ESP is a benign epithelial lesion. The incidence of the ESP in children is not known. The pathogenesis appears to be related to an underlying inflammatory process caused by gastroesophageal reflux, trauma, chemical irritants or, as in our this patient, HPV virus. Patients with ESP are usually asymptomatic; however dysphagia is a rare presenting symptom.

89 BEZOAR IN A PEDIATRIC ONCOLOGY PATIENT TREATED WITH COCA-COLA. A. Virojanapa, S. Naramore, Pediatrics, Penn State Hershey Medical Center, Hershey, Pennsylvania, UNITED STATESM. Bell, Pediatric Hematology and Oncology, Penn State Hershey Medical Center, Hershey, Pennsylvania, UNITED STATESP. Jhaveri, Pediatric Gastroenterology, Penn State Hershey Medical Center, Hershey, Pennsylvania, UNITED STATES. Introduction: Phytobezoars are the most common type of bezoar found in patients with dysmotility. Patients with oncologic disorders are at risk for intestinal dysmotility because of adverse effects of medications, poor fluid and dietary intake, and immobility. We describe an adolescent female with T-cell lymphoblastic lymphoma who was found to have a phytobezoar that was successfully treated with Coca-Cola. Case Description: A sixteen-year-old female with a history of T-cell lymphoblastic lymphoma diagnosed in October 2005 and treatment-related myelodysplastic syndrome status-post stem-cell transplant was admitted to our oncology service due to dehydration in January 2014. She had persistent nausea, early satiety, and periumbilical pain for two weeks. The day prior to admission she developed multiple episodes of nonbloody, nonbilious emesis. An endoscopic evaluation demonstrated the presence of a large gastric phytobezoar. The phytobezoar was effectively dissolved with three liters of Coca-Cola delivered via nasogastric tube¹. Polyethylene glycol with electrolytes was administered to assist with motility and the removal of any remnants of the phytobezoar. A swallow study obtained after Coca-Cola therapy showed no filling defects within the stomach or duodenum. She was seen for follow-up at one and five weeks after discharge. Her abdominal pain and emesis resolved and her nausea was significantly improved. Discussion: In our patient, the formation of a phytobezoar was secondary to the synergistic effect of multiple risk factors. Opioids, the most commonly prescribed medication for pain related to the treatment of cancer, increase the release of inhibitory neurotransmitters and affect neuronal excitability causing increased antral and pyloric tone and decreased tone of the gastric fundus². Many oncology patients receive H₂ receptor antagonists and proton pump inhibitors for prophylaxis against reactive gastropathy that

can be seen with corticosteroids given in conjunction with chemotherapy. A decreased oral intake or a high fiber diet can increase the susceptibility for the formation of phytobezoars. We believe that these factors of hypochlorhydria, decreased oral intake, pain medications, chemotherapy and a sedentary hospitalization played a role in the development of our patient's bezoar and are often overlooked in hospitalized patients in similar settings. To our knowledge, this is the first report describing the formation of any bezoar in a pediatric oncology patient and the first report demonstrating the resolution of a bezoar after Coca-Cola administration in any pediatric patient. Although bezoars are rare in the pediatric population, it is important for clinicians to remain cognizant of their formation in susceptible individuals in order to prevent a delay in diagnosis. 1. Ladas, S. D., et al. Systematic review: Coca-Cola can effectively dissolve gastric phytobezoars as a first-line treatment." *Aliment Pharmacol Ther.* 37.2 (2013): 169-173. 2. Sobczak, Marta, et al. "Physiology, signaling, and pharmacology of opioid receptors and their ligands in the gastrointestinal tract: current concepts and future perspectives." *J Gastroenterol.* 49.1 (2014): 24-45."

90 PNEUMATOSIS INTESTINALIS CAUSED BY CLOSTRIDIUM DIFFICILE IN A PATIENT WITH ANTI-NMDA-R AUTOIMMUNE ENCEPHALITIS. M. Middelhof, R. Abell, Pediatric Gastroenterology and Nutrition, University of Rochester, Rochester, New York, UNITED STATES.

We present a case of a 14-month old female patient with a history of anti-NMDA-R autoimmune post-HSV autoimmune encephalitis, who presented with abdominal distention, increased stool output and feeding intolerance. She had been on high dose methyl prednisone and intravenous immunoglobulin for her encephalitis. However, at her presentation, her labs revealed WBC of 7.5 Thou/uL, Hgb of 8.2 g/dL, Hct of 25% , and Plts of 449 Thou/uL. Due to increasing abdominal distention, an abdominal x-ray was performed which revealed pneumatosis intestinalis. Stool studies were positive for *Clostridium*>*difficile* toxin B. The pneumatosis intestinalis and diarrhea subsequently resolved with bowel rest, intravenous fluids, and oral metronidazole treatment. Pneumatosis intestinalis is defined as the presence of gas within the bowel wall. It has been associated as a complication of necrotizing enterocolitis in the neonatal period, however after the first year of life, pneumatosis intestinalis is rare as seen by scant case reports in the literature. Pneumatosis intestinalis has been described in Inflammatory Bowel Disease, Leukemia, post-bone marrow transplant, trauma, bowel obstruction, congenital heart disease, obstructive pulmonary disease as well as several gastrointestinal infections. Although the finding is rare, if pneumatosis intestinalis is identified on x-ray outside of the first year of life, screening for c.diff should be considered as conservative non-surgical management of this infection can lead to symptom resolution.

91 A UNIQUE CASE OF CONSTIPATION TREATMENT WITH MILK OF MOLASSES ENEMAS GONE AWRY. C. Perez, S.S. Kulkarni, Pediatrics, Miami Children's, Miami, Florida, UNITED STATES. J. Reeves-Garcia, W. Muinos, Gastroenterology, Miami Children's Hospital , Miami, Florida, UNITED STATES.

Enemas are routinely used for the management of constipation. Complications associated with enemas include metabolic derangements, arrhythmias, transient bacteremia, perforation, and allergic reactions. Milk of Molasses (MoM) enema has been shown to be an effective and safe therapy for constipation in a recent observational and randomized control study. We present the case of 5-year old female with Currarino's triad (sacral defect, presacral mass, and GI malformations), recurrent UTIs, multiple abdominal surgeries, and an appendicostomy for management of recalcitrant constipation. The patient's constipation maintenance regimen involved daily antegrade MoM enemas. Days preceding admission, the patient was requiring increased appendicostomy irrigations. After a MoM enema, patient was admitted to the pediatric intensive care unit (PICU) with emesis, tremors, altered mental status and hypotension. Patient had two prior admissions to PICU with similar symptoms. KUB and abdominal US confirmed fecal impaction. Laboratory analysis was significant for leukocytosis, hyperglycemia, and metabolic acidosis. The patient was treated with intravenous fluid resuscitation, antibiotics, rectal enemas and appendicostomy irrigations. Patient returned to neurological baseline. MoM was the suspected culprit of presenting symptoms and it was recommended to avoid such enemas on discharge. There has been no reoccurrence of these episodes. A case series by Walker et al reported five pediatric patients with cardiovascular compromise after MoM enemas, parallel to ours. This case raises awareness and illustrates the potential life-threatening harm of MoM in patients with complex medical histories.

92 A UNIQUE CASE OF H. PYLORI INFECTION IN A 13 YEAR OLD MALE WITH COLONIC INTERPOSITION AND ACUTE VOMITING. J. Reeves-Garcia, E. Hernandez, R. Arboleda, R. Gomara, W. Muinos,

Pediatric Gastroenterology, Miami Children's Hospital, Miami, Florida, UNITED STATES. Diaz Calderon, C. Perez, Medical Education, Miami Children's Hospital, Miami, Florida, UNITED STATES.

Background: Total esophagectomy with simultaneous colonic interposition has been used in pediatric patients with recurrent or recalcitrant strictures secondary to caustic ingestion. Complications of this procedure include anastomotic site strictures, leaks, bezoars, graft disease and gastrocolonic reflux. There are no reports in the literature of colonic interposition associated with H. Pylori infection. Case Report: We present a 13 year-old male with history of colonic interposition secondary to lye ingestion at 18 months of age. The patient was in his usual state of health, until he developed severe vomiting, nighttime regurgitation, abdominal pain and significant halitosis. On exam, patient had a non-acute abdomen and work up was initiated to rule out anatomical versus infectious etiologies. Upper gastrointestinal series demonstrated slow flow of contrast through interposed colon without stenosis at anastomotic site. H. Pylori blood titers were positive, but stool antigen was negative. Upper endoscopy demonstrated an intact colonic interposition and a diffuse area of severe chronic gastritis at fundus, greater and lower curvatures of stomach. Urease test and histology were positive for H. Pylori. Patient was treated with triple therapy – amoxicillin, proton pump inhibitor, and metronidazole for two weeks. At follow up, symptoms had resolved with confirmed test of cure. Discussion: This case elucidates a simple explanation for the patient's dyspepsia in the context of complicated medical history and anatomy. Helicobacter Pylori infection may present with recurrent vomiting in patients that have undergone surgical procedures, therefore testing is indicated when the etiology of the symptoms is not clear.

93 INFANT WITH CHOLELITHIASIS ASSOCIATED WITH PSEUDOHYPO-ALDOSTERONISM (PHA). V. Sood, M. Brown, Division of Pediatric Gastroenterology and Nutrition, Golisano Children's Hospital, Rochester, New York, UNITED STATES.

We report a 13 week old female infant who presented with persistent vomiting and failure to thrive, plus intermittent dehydration with electrolyte imbalances, and gall stones who was eventually diagnosed with Pseudohypo-aldosteronism (PHA). She was born at term following an uneventful delivery with unremarkable prenatal history. She was initially evaluated for gastroesophageal reflux, seborrhea, atopic dermatitis and poor weight gain. She was unable to tolerate cow's milk, soy or semi-elemental formulas with continued poor weight gain. With a working diagnosis of milk protein intolerance; she was switched to elemental formula with some improvement in her vomiting. Abdominal ultrasound to rule out hypertrophic pyloric stenosis revealed findings of incidental gall stones. She underwent further work up for gastroesophageal reflux including upper GI series, gastric emptying study which were normal. She was subsequently hospitalized for persistent vomiting, poor oral intake, severe weight loss and dehydration. On presentation her laboratory workup was significant for hyponatremia, hyperkalemia, metabolic acidosis, anemia and thrombocytopenia and elevated BUN and serum creatinine. Her liver enzymes were mildly elevated with normal bilirubin levels. Newborn screening for inborn errors of metabolism, hypothyroidism and sweat chloride testing was normal. Pseudohypo-aldosteronism was suspected based on the recurrent hyponatremia and hyperkalemia with subsequent testing showing a markedly elevated serum aldosterone and plasma renin activity. ACTH and cortisol levels were normal. Sodium chloride supplementation was started resulting in remarkable improvement in her oral intake with decrease in vomiting symptoms and weight gain. She continues to be asymptomatic and is doing well. Notably, her father was taking salt tablets. Discussion:Cholelithiasis is increasingly recognized in neonates and infancy due to widespread use of ultrasound as a routine procedure. The association of Pseudohypo-aldosteronism and Cholelithiasis has been reported rarely. Findings of gall stones in neonatal period/ infancy in the setting of vomiting, hyponatremia and poor weight gain should prompt further work up for PHA.

94 GASTRO-ESOPHAGEAL JUNCTION OBSTRUCTION IN SETTING OF EOSINOPHILIC ESOPHAGITIS IN A CHILD. T. Ciecieręga, R. Zarnegar, C. Crawford, NYP-Weill Cornell Medical College, New York, New York, UNITED STATES. M. Sultan, Makassed Hospital, Alquds Medical College, Jerusalem , PALESTINE, STATE OF.

Introduction: Dysphagia can present with gagging, coughing or painful swallowing. Its causes are broad including anatomical anomalies, eosinophilic esophagitis (EoE), psychogenic factors and esophageal dysmotility. Prompt diagnosis and treatment are crucial in these patients. We present a case of a 13 year old male with dysphagia caused by co-existing etiologies. Case Description: A 13 year old male with new onset dysphagia and diagnosis of achalasia based on barium esophagogram has been referred to our center. Recently he complained of worsening dysphagia, odynophagia, burning in the esophagus and postprandial nausea. Barium esophagogram showed

dilated esophagus with bird's beak appearance. Upper endoscopy was diagnostic for EoE (40 eosinophils/high power field at multiple levels). Esophageal manometry was consistent with Gastro-Esophageal Junction (GEJ) obstruction with normal peristalsis in setting of poorly relaxing lower esophageal sphincter. He was treated with swallowed steroids and acid suppression medications. Repeat endoscopy showed almost complete resolution of EoE. Repeat esophageal manometry continues to show GEJ obstruction. He is continuing his treatment.

Discussion: While EoE is becoming common diagnosis in children, true esophageal dysmotility is still rare. Their co-occurrence is even less frequent with only few reports thus far. Manometry is crucial in diagnosing specific esophageal dysmotilities as their treatments differ. While esophageal achalasia requires endoscopic or surgical treatment, GEJ obstruction in setting of appropriately treated EoE might not. It important to consider rare causes of dysphagia including esophageal achalasia and GEJ obstruction in children with chronic dysphagia in setting of EoE.

95 CASE OF TWO CHILDREN WITH EARLY ONSET OF MITOCHONDRIAL NEUROGASTROINTESTINAL ENCEPHALOPATHY SYNDROME IN CHILDREN.. T. Ciecierrega, C. Crawford, R. Zarnegar, NYP-Weill Cornell Medical College, New York, New York, UNITED STATESM. Sultan, Makassed Hospital, Alquds Medical College, Jerulasem, PALESTINE, STATE OF.

Introduction: Mitochondrial Neurogastrointestinal Encephalopathy (MNGIE) Syndrome is a rare multisystemic disorder. It usually presents in teenager years with gastrointestinal dysmotility (dysphagia, pseudoobstruction, nausea, vomiting) and neurological complications (ptosis, peripheral neuropathy, others). Specific mutations causing MNGIE have been identified. We report on two, 4 and 5 year old siblings with early presentation of MNGIE Syndrome. Case Series: Patient 1 was born full term without complications. At 40 days of life she developed poor sucking, recurrent vomiting, hypotonia and poor weight gain. Over years she required multiple admissions for dehydration. She presented to our hospital at age of 5 years with recurrent vomiting, dysphagia, hypotonia, poor growth, chronic constipation and lack of cognitive development. Labs were normal. UGI study showed dysmotility of esophagus. Genetics revealed mutation in G145R on TYMP gene (both parents were heterozygous). Patient 2 presented at age of four years with recurrent vomiting, severe nausea, diffuse abdominal pain, abdominal distension, poor appetite, weight loss and episodes of generalized weakness and fatigability. Birth and perinatal history were normal. Developmental milestones were achieved. Parents were relatives. Labs, stool and urine studies were normal. Upper GI series and endoscopy were normal except for dilated duodenal bulb. Genetic testing revealed mutation in G145R on TYMP gene on exon 3 (both parents carriers) diagnostic of MNGIE Syndrome Discussion: Mitochondrial Neurogastrointestinal Encephalopathy Syndrome is rare multisystemic disorder. The earliest reported age of onset is five months; onset is usually between the first and fifth decades Syndromes causes multisystemic dysfunctions with gastrointestinal system being most severely affected. Specific mutations have been identified. We report two siblings with very early disease onset. It is important to consider MNGIE Syndrome in any child with complex neuro-gastrointestinal symptoms presenting at any age.

96 AGANGLIONOSIS: IS IT ALWAYS HIRSCHSPRUNG'S?. M.V. Mendoza, B. McElhanon, Pediatric Gastroenterology, Hepatology, and Nutrition, Emory University, Atlanta, Georgia, UNITED STATESJ. Garza, Children's Healthcare of Atlanta at Scottish Rite, Atlanta, Georgia, UNITED STATES.

Hirschsprung's disease is the most common congenital gastrointestinal motility disorder in children, and should be considered in patients with constipation refractory to medical therapy. Occurring in 1:5,000 births and affecting males in a 4:1 ratio as compared with females, it results from a failure of neural crest-derived cells to colonize affected gut regions. As such, it is histologically characterized by aganglionosis and hypertrophied nerve trunks in rectal biopsies from the affected segment. When the diagnosis is in doubt, acetylcholinesterase (ACHE) staining can be used, with increased ACHe expression present in hypertrophied nerve fibers. We describe a 4yo male who presents with an 18 month history of constipation. Symptoms started when he began toilet training at 2.5yo. After failing outpatient management with polyethylene glycol and enemas, he presented to the emergency room with recurrent fecal impaction. After an episode of feculent emesis, he was admitted to the hospital, started on high doses of PEG3350 with electrolytes for disimpaction, and received additional workup for organic causes of constipation. Manual disimpaction with a strip rectal biopsy were subsequently performed by our surgery colleagues. Histology revealed aganglionosis, but also showed absence of hypertrophic nerve fibers. Subsequent ACHe stain was negative. For additional evaluation, calretinin immunostain returned negative. Recto-anal inhibitory reflex (RAIR) was present on anorectalmanometry. Our investigation provided data that both supported

(no ganglion cells, calretinin negative) and negated (lack of hypertrophic nerve trunks, AChE negative, presence of RAIR). In the context of our patient's acute symptom onset, we felt these findings were most consistent with an acquired hypoganglionosis. Taguchi et.al characterized the histologic findings from intestinal biopsies in a cohort of similar patients and compared them to those diagnosed with congenital hypoganglionosis syndromes (i.e. Hirschsprung's disease). In congenital hypoganglionosis, the number and size of the ganglia were small at birth. Over time, the cell size increased, but their overall numbers remained the same. Thus, the dysmotility symptoms did not improve over time. In the acquired form, the ganglion cells were normal in size and number at birth, but demonstrated degeneration over time. These patients presented with dysmotility symptoms later in life, and responded well to resection of the affected segment. This case illustrates the following important points: In those instances where functional constipation is most likely, cases that are refractory to medical therapy deserve additional diagnostic workup for congenital dysmotility syndromes. Although controversial, a diagnosis of acquired hypoganglionosis must be considered when the symptoms of constipation present acutely later in life and histopathology is non-diagnostic for congenital hypoganglionosis. Finally, additional work must be done to further characterize and determine the etiology of acquired hypoganglionosis.

117 EPSTEIN-BARR VIRUS-ASSOCIATED SMOOTH MUSCLE TUMOR FOLLOWING SOLID ORGAN TRANSPLANTATION IN A CHILD. J. Jossen, J. Chu, H. Hotchkiss, B. Wistinghausen, K. Iyer, M. Magid, R. Arnon, Mount Sinai Medical Center, New York, New York, UNITED STATES. Kamath, S. Roayaie, Hofstra-North Shore-LIJ School of Medicine, Manhasset, New York, UNITED STATES.

EBV-associated smooth muscle tumors are rare, but can occur in immunosuppressed individuals, especially children who have undergone solid organ transplantation. The location and behavior of these tumors vary widely and consequently there is no uniform approach to treatment. Previously described approaches include surgical resection, reduced immunosuppression, and antiviral therapy. Here, we describe the case of an 11-year-old boy who developed multiple Epstein-Barr virus-associated smooth muscle tumors in the liver 22 months following kidney transplant for end stage renal disease secondary to posterior urethral valves and reflux nephropathy. In order to completely remove all of the tumors, our patient required a right hepatic lobe resection. Two months prior to resection, the patient underwent portal vein embolization to induce hypertrophy of the unaffected lobe to help ensure adequate liver function post-resection, an approach not previously described in children. This patient then underwent successful surgical resection and was subsequently switched to sirolimus for ongoing immunosuppression, as recent evidence in the adult population suggests the anti-neoplastic effects of sirolimus may be beneficial in treating patients with EBV-smooth muscle tumors while maintaining immunosuppression for the transplanted organ. Now 16 months post-resection, the patient has not had any tumor recurrence and his kidney graft and liver function remain normal. Our case highlights a new approach to a rare and heterogeneous set of tumors in solid organ transplant recipients.

118 ATYPICAL HEMOLYTIC UREMIC SYNDROME DUE TO TACROLIMUS IN A CHILD POST LIVER TRANSPLANTATION. N. Eltawil, K. Radhakrishnan, N. Alkhouri, C. Quintini, B. Eghtesad, pediatrics, cleveland clinic, Cleveland, Ohio, UNITED STATES.

Two-year-old female with history of biliary atresia s/p failed Kasai at 3 months of age, s/p cadaveric left lateral segment liver transplant recipient EBV-/CMV+, donor EBV+/CMV- and roux-en-Y hepatojejunostomy at 8 months of age. Her post-transplant course was complicated with CMV re-activation, high EBV titers, and cryptosporidium infection. She was on daily tacrolimus for immunosuppression (IS). At 20 months of age, she was found to have transaminitis, a liver biopsy revealed acute cellular rejection, and she was subsequently admitted for treatment of acute rejection of her transplanted liver. She was asymptomatic, she had a normal liver vascular ultrasound, normal CT scan with no evidence of PTLN, labs showed: AST (H) 194, ALT (H) 108, BUN (N) 17, Cr (N) 0.27, tacrolimus level (N) 10.8, alkaline phosphatase (H) 1265, GGT (H) 393, hgb 10.2, platelets (N) 344 and CMV titers (H) 2,000 copies, and EBV titers (H) 281,344 copies. A normal MRCP ruled out any biliary obstructive process. She received high dose IV steroids, and IV ganciclovir. On day 9 of hospitalization, she had an acute drop of hgb to 5.6 g/dL, platelet count of 51 (L), her BUN 48 (H) and creatinine 0.65 (H) levels had tripled from admission, haptoglobin <20 (L), LDH 972 (H), Retic 9.3% (H), normal ADAMTS 13 level, and schizocytes were present on peripheral blood smear. Her clinical picture and laboratory findings were consistent with atypical hemolytic uremic syndrome (aHUS), due to tacrolimus. Tacrolimus was discontinued and cyclosporine was started for IS. Within 48 hours, there was an improvement in AST 57, ALT 44, BUN 24 (N), creatinine 0.41 (N), and platelet

count 122. She required a total of 3 PRBC transfusions, after which her hgb stabilized. Our patient exhibited acute hemolytic anemia, thrombocytopenia and acute kidney injury consistent with aHUS which reversed shortly after switching tacrolimus to cyclosporine. aHUS has mostly been described in post-renal transplant patients on tacrolimus therapy. aHUS should be included in the differential diagnosis of renal function deterioration in patients on tacrolimus post-organ transplantation.

LIVER

119 TYPE-2 AUTOIMMUNE HEPATITIS ASSOCIATED WITH POSITIVE ANTI-MITOCHONDRIAL ANTIBODIES IN A THREE-YEAR-OLD CHILD: AN OVERLAP SYNDROME?. S. Mullin, S. Malas, A. Bitar, Pediatric Gastroenterology & Nutrition, Michigan State University, Lansing, Michigan, UNITED STATESR. Rabah, Pediatric and Perinatal Pathology, University of Michigan Health System, Ann Arbor, Michigan, UNITED STATES.

Background: Autoimmune hepatitis (AIH), primary sclerosing cholangitis (PSC), and primary biliary cirrhosis (PBC) are different types of autoimmune liver diseases. AIH is divided into type-1 and type-2 based on the autoantibody profiles. While PBC is almost exclusively seen in adults, type-2 AIH occurs mostly in children. Although a precise clinical and pathological definition is still missing, an overlap syndrome is described as the association of AIH with PBC or PSC. AIH-PBC is the most common form of overlap syndrome in adults followed by AIH-PSC. In the pediatric literature, there are many reported cases of AIH-PSC overlap syndrome but only one case report with findings suggestive of both type-2 AIH and PBC, in which, the patient responded to immunosuppressive treatment. Case presentation: We report a case of type-2 AIH associated with PBC-specific anti-mitochondrial antibodies (AMA) in a 3-year-old male. The clinical, laboratory and histological features of the child are given in detail. Our patient responded very well to treatment with immunosuppressant agents. One year after diagnosis, our patient remains stable on a low dose of Prednisone and Azathioprine with ALT and AST levels < 2 ULN and no signs of relapse. Conclusion: This is the second pediatric case report of type-2 AIH associated with positive PBC-specific AMA. It is the first case to be reported in a male child. While treatment with Ursodeoxycholic Acid (UDCA) + immune suppression is recommended in adults with AIH-PBC overlap syndrome, immunosuppressant therapy alone seems to be effective in treating the same condition in pediatric patients. Our report adds new data to this still unclear entity.

Laboratory values. Prednisone started at diagnosis and Azathioprine added 6 weeks after diagnosis.

Abbreviations: nt, Not tested; AST, Aspartate Transaminase; ALT, Alanine Transaminase; Alk Phos, Alkaline Phosphatase; GGT, Gamma-Glutamyl Transferase; INR, International Normalized Ratio; Anti-LKM, Anti-liver-kidney microsome antibodies; AMA, Antimitoc

120 TOPIRAMATE INDUCED ACUTE LIVER FAILURE IN A PEDIATRIC PATIENT. J. Cordova, R. Azzam, Pediatric Gastroenterology, Hepatology and Nutrition, The University of Chicago, Chicago, Illinois, UNITED STATESM. Tsien, Internal Medicine, The University of Chicago, Chicago, Illinois, UNITED STATESL. Zhao, J. Hart, Pathology, The University of Chicago, Chicago, Illinois, UNITED STATESA. Qadir, Pritzker School of Medicine, University of Chicago, Chicago, Illinois, UNITED STATES.

The patient is an 11-year-old male with history of cerebral palsy, developmental delay, seizure disorder, and tracheostomy/ gastrostomy-dependence who presented with increased somnolence and three days of diarrhea. Initial laboratory results revealed AST 5666U/L, ALT 7890U/L, GGT 243U/L, bilirubin 5.9mg/dL, INR 10.8 and ammonia 1350mcg/dL; consistent with acute liver failure (ALF). His home medications, since infancy, included topiramate and phenobarbital for seizures and diazepam and baclofen for spasticity. Both serum antiepileptic blood levels were within their respective therapeutic range, although the topiramate level of 21.2ug/ml (normal range 2-25ug/ml) was significantly elevated from his previous levels of 5-7ug/ml. The patient had never had a topiramate level over 7 and the timing of his draw was essentially a trough. In addition to ALF, he was acidotic (pH 7.1, lactate 18mmol/L); in acute renal failure (creatinine 2.2mg/dL) and had signs of hypovolemic shock. On admission, he was unresponsive with grade IV hepatic encephalopathy. A CT head showed no intracranial hemorrhage or cerebral edema. An electroencephalogram was negative for seizures. Topiramate was held given his ALF, but phenobarbital, baclofen, and diazepam were continued. Work up for ALF included: negative serum viral studies (Hep A viral Ab IgM, Hep B core Ab IgM, Hep B surf Ag, Hep C virus Ab, Coxsackie A Ab panel, Coxsackie B Ab panel, Echovirus Ab panel, HSV 1 & 2 IgG/IgM, HSV PCR, Parvovirus B19 Ab, EBV capsid Ag/Ab IgM and IgG, CMV and EBV PCR, HHV6); negative urine toxicology screen; undetectable acetaminophen level; negative

autoimmune workup (ANA, smooth muscle antibody, liver/kidney microsome) and negative tests for genetic causes of his ALF (normal ceruloplasmin, elevated ferritin but negative iron stains on biopsy). A respiratory viral panel from his nares was positive for enterovirus/rhinovirus and adenovirus. A liver ultrasound/doppler showed good hepatic flow and minimal fatty infiltration. LFT's, ammonia, and coagulopathy rapidly improved with resuscitation and fresh frozen plasma. On day 4 of admission, a transjugular liver biopsy was completed. Pathology showed severe acute hepatitis with no necrosis but severe lobular disarray and panlobular inflammatory cell infiltrates with microvesicular steatosis involving 50% of the liver parenchyma consistent with drug-induced liver injury (DILI). Although the adenovirus PCR from his nares was positive, blood PCR and liver biopsy adenoviral stain were negative. Given the pathology and the clinical improvement after discontinuation of topiramate, the patient's ALF and hyperammonemia were deemed a consequence of topiramate toxicity.

Labortatory Values

Patient's laboratory values throughout hospitalization. Included are ALT, AST (U/L), Total bilirubin (mg/dL), INR and Ammonia (mcg/dL)

121 ADENOMYOMATOSIS OF THE GALLBLADDER IN AN INFANT WITH BECKWITH WIEDEMANN SYNDROME. C. Jarasvaraparn, E.M. Mcdonough, Division of Pediatric Gastroenterology, Hepatology and Nutrition, Arkansas Children's Hospital, Little Rock, Arkansas, UNITED STATES. A. Zarate, K.A. Bosanko, J. Vengoechea, Divison of Genetics, Arkansas Children's Hospital, Little Rock, Arkansas, UNITED STATES.

Adenomyotosis of the gallbladder (ADMG) is a benign acquired condition with not fully understood pathogenesis that, when symptomatic, presents as right upper quadrant abdominal pain due to cholelithiasis and inflammation. It is mainly diagnosed by imaging and is postulated that stones and chronic inflammation secondary to ADMG may lead to dysplastic changes and cancer. We report the case of a female infant with clinical diagnosis of Beckwith Wiedemann syndrome (BWS), a common overgrowth disorder of highly variable clinical presentation, in which ADMG was found on her routine liver ultrasound surveillance. This represents the first report of concurrent BWS and ADMG. To our knowledge, this is also the youngest patient reported with this abnormality. Given the lack of objective evidence of complications related to ADMG, our patient has not needed further invasive images done and continues to be monitored clinically. The long term implications of this diagnosis in the context of an underlying tumor predisposition syndrome remain unknown.

Table 1. Adenomyomatosis of gallbladder in children

122 RAPIDLY PROGRESSIVE FIBROSING NONALCOHOLIC STEATOHEPATITIS IN A 6 YEAR-OLD WITH PANHYPOPITUITARISM. S. Ibrahim, D. Freese, Pediatric gastroenterology and hepatology, Mayo Clinic, Rochester, Minnesota, UNITED STATES.

Panhypopituitarism with rapidly fibrosing nonalcoholic steatohepatitis (NASH) is a well-recognized condition in adults where growth hormone replacement halts disease progression and prevents liver transplantation. Here we report a 6 year-old with panhypopituitarism secondary to anaplastic astrocytoma, who presented 3 years after his astrocytoma diagnosis with hyperlipidemia, impaired glucose tolerance, a BMI above the 97th percentile for age, AST of 398 & ALT 476. Percutaneous liver biopsy showed moderately active steatohepatitis, and stage 3-4/4 fibrosis. Patient was not receiving growth hormone replacement therapy based on a normal bone age, a height on the 95th percentile for age and a normal IGF-1, IGFBP-3 levels. Patient's tumor nature and the relatively short interval from diagnosis put him at high risk for recurrence, and given the concern that growth hormone might be mutagenic and might increase the risk for secondary neoplasms; growth hormone supplementation was not pursued. Patient was started on vitamin E 800 IU per day, and ursodeoxycholic acid 10 mg/kg/day, in addition to a diet and exercise regimen, and close follow up. At 1 year follow up patient AST was down to 107, ALT down to 169 and patient was doing well clinically. Patients with hypothalamic/pituitary dysfunction are at risk for rapidly progressive NASH with cirrhosis which is an important implications for their work-up and management, growth hormone supplementation has been shown to improve NASH in these patients, nevertheless the decision to initiate therapy need to be individualized and based on risk benefit assessment.

123 ALPERS-HUTTENLOCHER SYNDROME: AN UNCOMMON CAUSE OF ACUTE LIVER FAILURE. D.S. Say, J. Gold, M. Sivagnanam, University of California, San Diego, San Diego, California, UNITED STATES. D.S. Say, J. Gold, M. Sivagnanam, Rady Children's Hospital, San Diego, San Diego, California, UNITED STATES.

A 4-year-old female with a history of epilepsy, failure to thrive, and developmental regression was hospitalized for evaluation of altered mental status and anorexia in the context of recent initiation of an anti-epileptic drug, divalproex sodium. Physical exam was significant for scleral icterus, jaundice, bruising, and abdominal tenderness, worrisome for valproic acid toxicity. Initial laboratory studies revealed hyperbilirubinemia, transaminitis, marked coagulopathy, and sub-therapeutic valproic acid levels. As liver transplantation was considered, diagnostic studies demonstrated a mutation in the mitochondrial DNA replicase, polymerase gamma (POLG), suggestive of a diagnosis of Alpers-Huttenlocher syndrome (AHS). A uniformly fatal disease, AHS is a rare mitochondrial cerebrohepatopathy characterized by a triad of resistant epilepsy, liver impairment, and progressive developmental regression. Though exposure to valproic acid exacerbates liver dysfunction in patients with AHS, liver failure is part of the defining natural history of this illness and occurs without valproic acid exposure. This case underscores the importance of maintaining a strong clinical suspicion for mitochondrial and metabolic causes of acute liver failure, particularly when considering the possibility of liver transplantation.

124 DISSEMINATED LANGERHANS HISTIOCYTOSIS PRESENTING AS CHOLESTATIC JAUNDICE. R.

Kapoor, S. Sachdeva, R. Yadav, P. Paul, Vardhman Mahavir Medical College & Safdarjung Hospital, New Delhi, INDIA. Loizides, Pediatric Gastroenterology, Children's Hospital at Montefiore, Bronx, New York, UNITED STATES. Loizides, Pediatrics, Albert Einstein College of Medicine, Bronx, New York, UNITED STATES.

Introduction: Langerhan Cell Histiosytosis (LCH) is a disorder associated with infiltration of Langerhan cells into various organs. Cholestasis in LCH may result from lymph nodes obstructing the porta hepatis, or autoimmune biliary diseases which are associated with LCH. In this report, we describe a case of type II LCH with obstructive cholestasis and pulmonary involvement, no lymphadenopathy at the porta, but with microscopic periportal infiltration of LCH cells. Case: A 2 year old boy presented with a one month history of fever, jaundice, acholic stool and generalized pruritus as well as a 6 month history of progressive abdominal distention, right ear discharge and seborrheic dermatitis. On admission to the hospital he was febrile and deeply jaundiced. Numerous scaly white papular lesions were noted on his scalp. Examination of abdomen revealed smooth, firm, non-tender hepatosplenomegaly, with a liver span of 10 cm and spleen 3 cm below the left costal margin. No generalized lymphadenopathy was appreciated. Laboratory findings included anemia, elevated aspartate transaminase, alanine transaminase, alkaline phosphatase, gamma-glutamyl transferase and total and direct bilirubin. Computed tomography of the chest showed nodules in bilateral lung fields exhibiting ground glass opacity with no zonal predominance and cystic changes in the left upper lobe. Computed tomography of the abdomen detected moderate hepatosplenomegaly and periportal hypodensities extending along the portal radicals. There was no lymphadenopathy in the area adjacent to the porta hepatis. Skeletal survey exhibited multiple osteolytic lesions in areas of the parietal skull and in the right iliac crest. On skin biopsy, an infiltrate of large cells with abundant cytoplasm, convoluted to reniform nuclei was appreciated. Biopsy of the liver revealed periportal infiltration of cells that stained positive for CD1a and S100, consistent with the diagnosis of Type II LCH. Chemotherapy was initiated and he showed progressive improvement. DISCUSSION: Langerhan cells are antigen-presenting cells of the skin and mucous membranes that have their origin from bone marrow. The cells have characteristic Birbeck granules and are present in all layers of epidermis, most commonly in stratum spinosum. More than two-thirds of cases have single system disease with bone and skin most commonly involved. However, a wide spectrum of multisystem involvement has been seen including the liver. Needle biopsy of our patient's liver revealed involvement of the periportal triad consistent with type II LCH. Although the literature on LCH is extensive and is most often recognized only at post mortem examination in patients with multi-organ lesions, predominant involvement of the liver is rarely described and usually a result of certain pathological conditions such as primary sclerosing cholangitis, primary biliary cirrhosis, or obstruction secondary to enlargement of lymph node adjacent to the porta hepatis. Health practioners must therefore be cognizant that LCH can present as cholestatic jaundice without significant porta hepatis lymphadenopathy or as the result of direct autoimmunoreactivity involving the biliary tree.

125 ALANINE AMINOTRANSFERASE VALUES IN OBESE CHILDREN WITH NONALCOHOLIC FATTY LIVER

DISEASE DIAGNOSED WITH LIVER ULTRASONOGRAPHY. N. Eltawil, pediatrics, cleveland clinic, Cleveland, Ohio, UNITED STATES. Alkhouri, J. Moses, pediatric gastroenterology, cleveland clinic children's hospital, cleveland, Ohio, UNITED STATES.

Background: No consensus exists on the best screening strategy for nonalcoholic fatty liver disease (NAFLD) in children. The AAP recommends using ALT in overweight children (BMI 85-94% for age and gender) with risk factors in the history or physical examination and in all obese children. Recent European guidelines suggest performing a liver ultrasound (US) in addition to liver enzymes in all overweight and obese children over the age of 3 years. We aimed to investigate ALT values in children with evidence of NAFLD on US. Methods: Consecutive overweight and obese children and adolescents (6-18 years) seen at the Preventive Cardiology and Metabolic Clinic were included. Each child underwent a liver US and had ALT level measured at the time of their first clinic visit. Two cutoff values for abnormal ALT were used: 1. Our laboratory cutoff of 45 U/L. 2. Sex-specific cutoff points recently proposed by Schwimmer et al (>25.8 U/L for boys and >22.1 U/L for girls). Other etiologies of chronic liver disease were ruled out in children with evidence of fatty infiltration of the liver. Results: 187 overweight/obese children were seen in our clinic between 2011 and 2013. NAFLD was present on US in 91/187 (48.6%) (9 children with BMI between 85-94%, 21 with BMI between 95-98%, and 61 with BMI ≥99%). The majority were Caucasian (68%), 20% were Hispanic, 6% were Black and 6% were other races. One or more components of the metabolic syndrome were present in 77%. Mean ALT was 29.52 ± 13.43. Mean ALT was significantly higher in Hispanics compared to Caucasians (45.4 ± 30.4 vs. 24.3 ± 11.2, p < 0.005). High ALT using our laboratory cutoff of 45 U/L was present in only 16/91 patients (17.6%). Using lower cutoff values improved the detection of NAFLD but was only present in 39/91 (42.8%) of children. Conclusion: Using ALT values as the sole screening strategy for NAFLD in obese children will miss a substantial number of patients with fatty liver disease. We suggest using a combination of ALT and US to screen for NAFLD and determine the need for further testing and more aggressive interventions.

Patient demographics

126 SPONTANEOUS RESOLUTION OF HEPATIC ARTERY THROMBOSIS IN TWO PEDIATRIC LIVER TRANSPLANT PATIENTS.. E.B. Rand, J.W. Wen, K.M. Loomes, H.C. Lin, Fred & Suzanne Biesecker Liver Center, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES. A.M. Banc-Husu, E.B. Rand, J.W. Wen, K.M. Loomes, H.C. Lin, Department of Pediatrics, Perelman School of Medicine at the University of Pennsylvania, Philadelphia, Pennsylvania, UNITED STATES.

Hepatic artery thrombosis (HAT) is a serious complication after liver transplantation. The incidence of HAT in children is 7- 25%, and the risk is increased with prolonged ischemia time, small recipient and donor size, or small artery diameter. HAT often leads to acute and devastating graft injury, resulting in death or urgent retransplantation. This is the first report of spontaneous resolution of HAT in pediatric liver transplant patients on enoxaparin (LMWH) therapy. Case 1: A 26-month-old boy presented with acute liver failure of indeterminate etiology and underwent emergent transplantation with a live donor left lateral segment graft. The extrahepatic arterial anastomosis required surgical repositioning on POD0 when he was noted to have markedly elevated liver enzymes and failure to identify arterial flow on ultrasound (US). He was started on LMWH 1 mg/kg/dose twice daily and aspirin (ASA) 81 mg daily given the thrombotic risk following hepatic artery manipulation. A CT angiogram on POD8 demonstrated poor visualization of the intrahepatic artery. Liver biopsy on POD11 performed for elevated liver enzymes showed findings concerning for ischemia. Follow-up US on POD18 demonstrated absent flow in the hepatic artery, and was confirmed by a celiac angiogram. As liver synthetic function worsened, he was relisted for transplantation on POD34; LMWH and ASA were stopped given the need for retransplantation. However, liver enzymes spontaneously improved and POD79 US showed patent arterial flow, confirmed by CT angiogram with no stenosis or thrombosis of the donor hepatic artery. Case 2: A 15-year-old boy with ulcerative colitis and autoimmune hepatitis-primary sclerosing cholangitis overlap received a whole deceased donor liver transplant for progressive cirrhosis. On POD11, he became febrile and US demonstrated absent hepatic arterial flow, confirmed by a CT angiogram and a celiac angiogram. He was started on LMWH 1 mg/kg/dose twice daily on POD18 for a line-associated clot for a 6-month treatment course. Liver function was stable throughout his course. Follow-up CT angiogram on POD126 showed greater enhancement of the hepatic artery lumen consistent with continued recanalization of the donor artery. LMWH was subsequently stopped. HAT can complicate pediatric liver transplants and treatment includes thrombectomy or retransplantation. The cases presented show spontaneous resolution of HAT while on LMWH, which has been used predominantly for line-associated clots and pulmonary emboli with clot resolution. These are the first two cases reporting spontaneous recanalization following HAT in the absence of surgical intervention.

COMBINED LIVING DONOR LIVER AND HEMATOPOIETIC STEM CELL TRANSPLANTATION FOR ADVANCED HEPATOCELLULAR CARCINOMA IN A CHILD. J. Picoraro, M. Martinez, S. Lobritto, N. Ovchinsky, Pediatric Gastroenterology, Hepatology and Nutrition, Columbia University Medical Center, New York, New York, UNITED STATES. Satwani, Pediatric Hematology, Oncology and Stem Cell Transplantation, Columbia University Medical Center, New York, New York, UNITED STATES. Kato, Department of Surgery, Columbia University Medical Center, New York, New York, UNITED STATES.

We report a case of a haplo-identical paternal CD34-selected hematopoietic stem cell transplant (HSCT) following a living donor liver transplant (LDLT) for a 12 year old boy with advanced hepatocellular carcinoma (HCC). Graft versus tumor (GVT) effect can be induced by allogeneic bone marrow transplant (BMT) in patients with hematologic malignancies and has also been observed in patients with solid tumors such as metastatic breast carcinoma and metastatic renal cell carcinoma. There have been cases reported in adults with advanced primary liver cancers who received cadaveric liver transplantation followed by HSCT (one from the same cadaveric donor and others from HLA-matched unrelated donor). We performed the combined transplant from the same donor in an attempt to induce graft tolerance and possible GVT effect. The patient received a paternal left lobe LDLT and had an uncomplicated post-transplant course. His immunosuppression included tacrolimus and a prednisone taper. One month following LDLT, he received a haplo-identical paternal CD34-selected HSCT with reduced-intensity conditioning (busulfan, fludarabine) and extensive T-cell depletion (alemtuzumab). Engraftment was demonstrated on Day+18, and chimerism analyses are shown in Table 1. Alpha-fetoprotein level decreased from 104,000 ng/ml pre-LDLT to 3.7 ng/ml on Day+30. Patient expired on Day+36 secondary to a multi-system organ failure and his autopsy demonstrated disseminated toxoplasmosis. He had a positive IgG toxoplasma antibody prior to HSCT and received prophylaxis but was not routinely monitored for toxoplasmosis post-transplant. We report a first case of combined same-donor LDLT and HSCT for a primary liver tumor. We achieved mixed donor-recipient hematopoietic chimerism without any significant acute graft-versus-host-disease (GvHD). Large unresectable hepatic tumors have a high risk of relapse despite liver transplantation. Allogeneic marrow grafting may provide a potent GVT effect and a possibility to induce tolerance to the liver allograft, permitting reduction, if not elimination, of immunosuppressive therapy. Haplo-identical donors may offer sustained engraftment and low incidence of GvHD but also carry a significant risk of opportunistic infections.

Donor Chimerism

PANCREAS

ANNULAR PANCREAS PRESENTING WITH HEMATEMESIS IN AN 18 YEAR OLD MALE WITH DEVELOPMENTAL DISABILITY. R. Sabe, A.N. Aktay, Pediatric Gastroenterology, Hepatology & Nutrition, UH Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES.

Annular pancreas is a rare condition that may present at any age. Symptoms range from abdominal pain to intestinal obstruction. Diagnosis is usually via imaging studies in symptomatic patients. It can also be found as an incidental finding in asymptomatic patients undergoing imaging for another indication. Patients who are unable to verbalize their symptoms may be diagnosed later and may have higher morbidity. Our case involves a developmentally disabled male who presented with hematemesis at the age of 18 years. Our patient was mainly gastrostomy tube fed with minimal oral pureed food, and no solid food in his diet which probably delayed his presentation and diagnosis. An 18 year old male with a history of mental retardation, congenital hydrocephalus with a ventriculo-peritoneal (VP) shunt placement, septo-optic dysplasia, and laparoscopic gastrostomy and Nissen fundoplication who presented with vomiting that progressed to hematemesis. At 11 years of age during the laparoscopic procedures he was found to have multiple adhesions that were attributed to the presence of a VP shunt. The pancreas was not visualized during the procedure. 4 weeks into his acute vomiting and hematemesis episode an upper GI study demonstrated a thickened gastric antrum and narrowing of the proximal duodenum. His laboratory work up was significant for anemia and hypoalbuminemia. He was later admitted to the hospital and underwent upper endoscopy that revealed severe hemorrhagic gastritis and duodenitis. The duodenal bulb and proximal duodenum were dilated and the second portion of the duodenum was narrowed. Biopsies showed acute and chronic inflammation. They were negative for HSV, CMV, *H. pylori*, and viral cultures. An abdominal CT scan showed an annular pancreas. He was kept NPO and placed on an intravenous acid suppression medication and total parenteral nutrition with the goal of reducing inflammation. Repeat upper endoscopy 4 weeks later revealed the same findings. He underwent an exploratory laparotomy with duodenoduodenostomy. Enteral nutrition was

restarted slowly and he tolerated advancement to full calories. Although annular pancreas is a rare condition, it should be considered in patients who present with signs and symptoms of obstruction. We want to increase awareness of possible late presentation of annular pancreas in patients who are unable to verbalize their symptoms such as developmentally disabled patients. Delayed diagnosis may result in gastrointestinal tract inflammation and nutritional deficiencies. Treatment is usually via bypass surgery in patients who are symptomatic. Resection of the annulus should be avoided due to increased risk of morbidity.

153 PHANTOM POST-PANCREATITIS ABDOMINAL PAIN AND THE CHALLENGES OF PAIN MANAGEMENT IN PANCREATITIS. Z. Sellers, M. Garcia-Careaga, Stanford University, Palo Alto, California, UNITED STATES.

A morbidly obese 15-year-old male with a history of a single bout of acute pancreatitis has unrelenting abdominal pain. In spite of an extensive negative work up, that includes a CT, MRI, MRCP, ERCP, endoscopic ultrasound, and normalization of biochemical markers, he continues to experience severe abdominal pain. Post-acute pancreatitis neuropathy was strongly considered. During his endoscopic ultrasound an attempt to do a celiac plexus block failed due to his morbid obesity. He was initially discharged on gabapentin, oxycontin, and clonazepam with outpatient follow-up by the pediatric pain service. He has also been followed by pediatric gastroenterology and psychology for pain-associated anxiety. Over the course of a year, his pharmacological regimen developed into simultaneous use of gabapentin, celecoxib, escitalopram, amitriptyline, clonazepam, oxycodone, ondansetron, pancreatic lipase, sucralfate, omeprazole, polyethylene glycol, and docusate. Non-pharmacologic interventions included low-fat diet, acupuncture, physical therapy, and biofeedback. All of these interventions provided no pain relief with multiple, severe, stabbing, epigastric abdominal pain episodes per day with nausea and intermittent vomiting. These symptoms, coupled with anxiety and sleep deprivation, prevented him from attending school. Subsequently, he developed opioid-induced constipation and urinary retention. Due to these complications, he underwent a bilateral rectus sheath block with transverse abdominis plane trigger point injections of ropivacaine. He developed numbness of the appropriate areas, but experienced no pain relief. As a result, his pain was determined to be more psychological rather than physiological and his pain is currently "managed" on dilaudid, duloxetine, and polyethylene glycol with frequent psychology and physical therapy sessions. Despite this, he continues to have daily severe abdominal pain with only 2-3 hours of sleep per night and remains on independent study for school. Discussion: Acute pancreatitis affects 3.6-13.2/100,000 pediatric patients, with abdominal pain resulting in significant morbidity. Using this unusual case of unrelenting abdominal pain, we review current practices for pancreatitis pain management, including a discussion of common side effects. Acute and acute recurrent pancreatitis is commonly treated with opiates, while chronic pancreatitis pain often requires a combination of analgesics and neuromodulators. Research into the somatic and cerebral neural basis of pancreatitis pain provides insight into potential novel pain management strategies.

154 PANCREATITIS IN THE SETTING OF SPINK1 AND CTSC MUTATIONS COMPLICATED BY PANCREATIC DUCT LEAK AND PANCREATICOPLEURAL FISTULA. N.J. Hoffman, E. Semeao, P. Mamula, Gastroenterology, Hepatology, and Nutrition, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES. Anupindi, Radiology, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES. Adzick, General, Thoracic, and Fetal Surgery, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES. Kaimakliotis, Gastroenterology, Hospital of The University of Pennsylvania, Philadelphia, Pennsylvania, UNITED STATES.

A 7 year old previously healthy female presented to an outside institution with abdominal pain and vomiting. A CT scan was concerning for appendicitis and she underwent appendectomy. She had continued symptoms and 1 week post-operatively her amylase and lipase were noted to be elevated. She was diagnosed with acute pancreatitis. After a period of bowel rest, she was unable to tolerate a diet by mouth and was started on enteral nutrition via nasojejunal tube. An MRCP revealed an irregular and diffusely dilated main pancreatic duct suggestive of chronic pancreatitis and could not rule out a distal pancreatic duct stricture. Pain and emesis continued, and she was still unable to tolerate enteral nutrition. Given that previous imaging could not rule out a pancreatic duct stricture, she underwent ERCP which revealed calcifications and a dilated and tortuous pancreatic duct. A pancreatic duct stent was placed. Her post-ERCP course was significant for SIRS response and the development of large intra-abdominal fluid collections as well as a left pleural effusion. Diagnostic and therapeutic thoracentesis was performed, and the pleural fluid demonstrated elevated lipase concerning for pancreatic fluid. Repeat ERCP demonstrated pancreatic

duct leak and a pancreaticopleural fistula. This was managed by removing and replacing her pancreatic duct stent and continued pancreatic rest and total parenteral nutrition. Her fluid collections slowly resolved over weeks and eventually she was again able to tolerate enteral nutrition. After several weeks in place, her pancreatic duct stent was removed endoscopically. During her prolonged hospitalization, genetic testing revealed heterozygosity for both a pathologic *SPINK1* mutation as well as a pathologic *CTRC* mutation. She was only able to tolerate enteral nutrition for a short time before she again developed pain, vomiting, and elevated lipase. Given her prolonged course and chronic pancreatic changes, she underwent pancreaticojejunostomy (Puestow procedure). Genetic predisposition should be considered in cases of chronic and recurrent pancreatitis. Pancreaticopleural fistula is a rare complication of pancreatitis. Surgical intervention should be considered in complicated cases of pancreatitis refractory to medical management.

155 GIRLS, WORMS AND GOATS! A PEDIATRIC CASE OF ASCARIS LUMBRICOIDES PANCREATITIS. A.S. Huang, B. Infantino, Pediatric Gastroenterology, UNMC, Omaha, Nebraska, UNITED STATES. Johnson, Family Medicine, UNMC, Omaha, Nebraska, UNITED STATES. J. Pham, Pediatrics, UNMC, Omaha, Nebraska, UNITED STATES.

Ascariasis is one of the most common helminthic infections in the world. Approximately 1 billion people are believed to be infected. In the United States it's considered an uncommon disease. Incidence increases in areas of inadequate sanitation and low socioeconomic status. Children are the most affected age group. Most commonly the infection presents as an enteritis, but may also have pulmonary involvement and even intestinal obstruction. Pancreatitis secondary to *Ascaris* has been reported in endemic countries, but there are no reported pediatric cases of pancreatitis secondary to *Ascaris lumbricoides* in the USA. We present a case report of a 7 year old female with pancreatitis with no evidence of obvious obstructive disease documented by imaging including CT and ultrasound. She had an incidental finding of a worm coming out of her mouth, and once this was recovered, the symptomatology and pancreatic enzyme levels resolved. The patient lives on a farm and has had contact with various animals, including goats, hogs, cows, sheep, and a dog. The worm was studied and was determined to be *Ascaris lumbricoides*. The patient and family members were treated with albendazole and are currently asymptomatic.

156 PANCREATIC CYSTOSIS: AN UNCOMMON COMPLICATION OF CYSTIC FIBROSIS. C.A. Reynolds, J. Daniel, Gastroenterology, Children's Mercy Hospital, Kansas City, Missouri, UNITED STATES.

Pancreatic cystosis is a relatively rare finding in cystic fibrosis. The more common abnormalities seen in cystic fibrosis include partial or complete fibrofatty replacement of pancreatic tissue and pancreatic atrophy without fatty replacement. In various case reviews, the incidence of cystosis was highly variable, at most 8% in a small case series of 23 patients and none found in a series of 109 patients. It is infrequently mentioned in case reports in the literature. It is believed that the pancreatic cystic changes seen in cystic fibrosis patients are due to impaired drainage of the ducts but with maintenance of functional secretory capacity of the exocrine pancreas. Case reports note that treatment options include expectant management, surgical drainage or pancreatectomy. We present a case of pancreatic cystosis in a 12 year old male with cystic fibrosis. He presented to our GI clinic with complaints of abdominal pain. His physical exam was significant for right upper quadrant and epigastric tenderness. Laboratory studies demonstrated slight leukocytosis, all other labs, including pancreatic enzymes, were within normal limits. Subsequent ultrasound, CT, and MRCP demonstrated numerous pancreatic cysts, the largest of which was found in the head of the pancreas and measured 6.1 cm x 6.1 cm x 5.6 cm. There was some mass effect on outside structures from this large cyst, but no duodenal obstruction. Other smaller cysts ranged from 2 mm to 14 mm. This case demonstrates an atypical cause of abdominal pain in cystic fibrosis. Early recognition of pancreatic cystosis may improve quality of life in patients and discussion of cases such as this in the literature may facilitate medical decision making regarding treatment.

157 AUTOIMMUNE PANCREATITIS MASQUERADING AS CHOLESTATIC HEPATITIS: A PEDIATRIC CASE SERIES. V. Okwu, S.A. Patel, R. Gupta, N. Alkhouri, K. Radhakrishnan, V. Hupertz, Pediatric Gastroenterology, Cleveland Clinic Children's Hospital, Cleveland, Ohio, UNITED STATES.

Autoimmune pancreatitis (AIP) is a rare but increasingly recognized form of pancreatitis that can present as cholestatic hepatitis. This disease largely affects adults with only a few cases reported in children. We describe two pediatric patients with AIP who presented with cholestatic hepatitis. Case 1: A 13-year-old boy presented with a 2-

week history of abdominal pain and bloody diarrhea. His physical exam was unremarkable but his labs revealed WBC 13.7 k/uL (70% neutrophils), hemoglobin 9.5 g/kL, platelets 835 k/uL, AST 154 U/L, ALT 249 U/L, Total bilirubin 12 mg/dL, Direct bilirubin 6 mg/dL, amylase 456 U/L, lipase 771 U/L, and GGT 177 U/L. Abdominal CT was significant for a prominent pancreas, intra- and extra-hepatic biliary ductal dilatation. Abdominal ultrasound showed dilatation of the intrahepatic and common bile ducts and enlarged pancreas with multiple periportal and peripancreatic lymph nodes. MRCP was concerning for a lobulated mass encasing the porta hepatis for which the patient underwent laparoscopy with multiple biopsies taken of lymph nodes and the retroperitoneal mass. Lymph node biopsies revealed lymphoid hyperplasia; the mass biopsy showed acute and chronic pancreatitis. Further testing was remarkable for positive ANA, positive AMA and an elevated total IgG (IgG1 elevated with mildly elevated IgG3 and IgG4). Liver biopsy showed chronic hepatitis with bridging fibrosis. These findings prompted the initiation of steroids for AIP resulting in a marked improvement in abdominal pain. Amylase and lipase returned to normal and ALT/AST improved to 170/76. Case 2: A 15-year old male presented with a 2 week history of generalized jaundice, abdominal pain and diarrhea. His physical exam was unremarkable but his labs revealed WBC 8.8 k/uL (60% neutrophils), hemoglobin 11.1 g/kL, platelets 444 k/uL, AST 399 U/L, ALT 621 U/L, Total bilirubin 11.2 mg/dL, Direct bilirubin 7 mg/dL, amylase 100 U/L, lipase 125 U/L, and GGT 537 U/L. Abdominal US showed hepatosplenomegaly and dilatation of the common bile duct. MRCP showed decreased signal with patchy enhancement noted throughout the pancreas with peri-pancreatic fluid. There were also findings of mild intra-hepatic and extra-hepatic biliary duct dilatation and mild beading within the intrahepatic ducts. Further testing was remarkable for normal ANA and IgG subclass levels. Liver biopsy showed liver parenchyma with panlobular hepatitis with overlapping changes of downstream obstruction. These findings prompted the initiation of steroids for AIP with subsequent improvement in his labs. Amylase and lipase returned to normal and ALT/AST improved to 225/91. Conclusion: This case series illustrates the fact that although autoimmune pancreatitis is rare in children it can present with a cholestatic picture. Recognition of the varied clinical presentation of autoimmune pancreatitis is critical to the timely institution of appropriate therapy to prevent further complications.

Poster Session II
Friday, October 24, 2014

BASIC IBD

169 CAP POLYPOSIS MISTAKEN FOR ULCERATIVE COLITIS IN AN 8 YEAR OLD. D. Walkiewicz, K.A. Matkowskyj, Pediatrics, UW School of Medicine and Public Health, Madison, Wisconsin, UNITED STATES. Introduction: Cap polyposis is a rare and frequently unrecognized condition in children. Symptoms are bloody or mucoid diarrhea, abdominal pain and tenesmus. Presentation and histopathology can be misdiagnosed as ulcerative colitis. Case: 8 year old girl with chromosome 16p11.2 deletion has been seen in GI clinic for constipation. She presented to the ED with fever, lethargy, mucoid/bloody diarrhea and abdominal pain. Her physical exam was remarkable for abdominal tenderness and distention. Blood work revealed anemia, thrombocytopenia, hypoalbuminemia, elevated ESR, CRP and high serum Epstein - Barr virus (EBV) PCR. CT scan of her abdomen showed diffused inflammation in her colon. Colonoscopy was performed and showed erythematous mucosa with many pseudopolyp in the descending and rectosigmoid colon. Scattered pseudopolyps were seen in the right colon. Histopathology revealed patchy active colitis with negative in situ hybridization for EBV encoded DNA (EBER). The patient was treated with Gancyclovir/ Vangancyclovir and Sulfasalazine. Bloody stools continued. She was placed on prednisone and mesalamine with some improvement of symptoms. She continued to have elevated EBV PCR but anemia, thrombocytopenia, hypoalbuminemia, ESR and CRP normalized. Colonoscopy 4 months later showed persistence of polypoid lesions with presence of whitish coating on some of the lesions with focal active colitis in colonic mucosa between polyps. EBER staining showed rare positive lymphocytes within the lamina propria with negative EBER staining in the epithelial cells. The patient was diagnosed with ulcerative colitis and coexisting EBV infection. Mesalamine and steroids were started with resolution of diarrhea. Colonoscopy 3 months later showed multiple, well-formed pedunculated and sessile polyps with a well-demarcated white coating on the top in the rectosigmoid area with normal looking mucosa between polyps. The remainder of mucosa looked normal. Colon biopsies were negative for colitis. The polyp fragment revealed inflamed colonic mucosa with tortuous, elongated crypts attenuated towards the mucosal surface, inflammation in lamina propria and fibrinopurulent exudate/granulation tissue resting at the top the polyp, raising the possibility of Cap polyposis.

Conclusion: Cap polyposis is a rare condition that can mimic inflammatory bowel disease. Etiology is unclear, but abnormal bowel motility and straining have been proposed as possible triggers. Although rare, it can occur in children and should be considered in differential diagnosis in pediatric patients with abdominal pain, bloody diarrhea and tenesmus. It is not clear at this time if the patient's chronic EBV infection played a role in her GI symptoms. References Kini GP, Murray I, Champion-Young J et al. Cap polyposis mistaken for Crohn's disease: Case report and review of literature. *Journal of Crohn's and Colitis* (2013) 7, e108-e111 Aggarwal R, Gupta P, Chopra P et al. Rectal cap polyposis masquerading as ulcerative colitis with pseudopolyps and presenting as chronic diarrhea. *The Saudi Journal of Gastroenterology*. Volume 19; Number 4. Shabaan 1434. July 2013.

170 AN UNCOMMON NEUROLOGIC SYNDROME IN A PATIENT WITH INFLAMMATORY BOWEL DISEASE: A CASE REPORT. A. Ghazi-Askar, J. O'Connor, Department of Pediatrics, Section of Pediatric Gastroenterology, Hepatology, and Nutrition, University of Oklahoma Health Sciences Center, Oklahoma City, Oklahoma, UNITED STATES.

Introduction Inflammatory bowel disease (IBD) can be associated with many neurologic complications such as peripheral neuropathies, myopathies, cerebrovascular events and vasculitis. We present the case of a 12 year-old female with known long standing IBD who presents with severe, sudden-onset headaches. Initially thought to be due to small vessel vasculitis, our patient was diagnosed with reversible cerebral vasoconstrictive syndrome, a rarely described entity in the pediatric population. Case Presentation 12 year-old female with history of indeterminate colitis was hospitalized with tracheitis and experienced sudden-onset severe headaches and hypertension after receiving IV injection of diphenhydramine. Initial laboratory work up was nondiagnostic. Head MRI revealed scattered subtle areas of increased FLAIR signal without definitive postcontrast leptomeningeal or parenchymal enhancement. CT angiography of the brain revealed multifocal areas of vascular abnormality that were suspicious for a vasculitis. Physical examination, including neurologic exam, was normal. She continued to have headache and nausea, but no emesis. Blood and CSF studies were negative for inflammatory markers and for infectious, rheumatologic, autoimmune and metabolic causes of recurrent headache. She was diagnosed with reversible cerebral vasoconstrictive syndrome (RCVS). She was started on scheduled calcium channel blocker therapy and was discharged home with no more episodes of severe headaches upon follow up several weeks later. Discussion Reversible cerebral vasoconstrictive syndrome (RCVS) is an entity characterized by severe headaches, with or without other acute neurological symptoms, and diffuse segmental constriction of cerebral arteries due to transient disturbances of cerebral vascular tone. Rarely reported in the pediatric population, RCVS is characterized in adult literature as occurring spontaneously, associated with medications, or in the post-partum period. Treatment of RCVS is with calcium channel blockers as opposed to that of vasculitis associated with IBD, which is treated with anti-inflammatory medications. Thus, it is important to consider RCVS when evaluating patients with IBD who present with severe headaches and abnormal vascular imaging.

171 SOLITARY PERINEAL-VULVAR ULCER SECONDARY TO BEHÇET'S DISEASE IN A PEDIATRIC PATIENT SUCCESSFULLY TREATED WITH INFLIXIMAB. C. Pichardo, M. Pereira, Department of Medical Education, Miami Children's Hospital, Miami, Florida, UNITED STATESR. Gomara, R. Arboleda, W. Muinos, E. Hernandez, J. Reeves-Garcia, Department of Pediatric Gastroenterology, Miami Children's Hospital, Miami, Florida, UNITED STATESC. Brathwaite, Department of Pathology, Miami Children's Hospital, Miami, Florida, UNITED STATES.

We describe a case of a 14 year old female presenting with large perineal ulcers for 3 years and a 10 pound weight loss. Patient had developed a mass on vaginal area at age 11 years with a negative work up for sexually transmitted infections. She was treated locally with antibiotic and the mass resolved, but the perineal ulcers persisted and worsened. Patient developed severe genital pain, unable to participate in activities of daily living. Physical exam was remarkable for a grade III gray ulcerated lesions in vulvar area with an eschar measuring 5x3 inches without any drainage or odor. Due to concerns of possible Crohn's disease with perineal involvement the gastroenterology service was consulted. Vulvar biopsies obtained were not conclusive and pyoderma gangrenosum was considered. Work up for sexually transmitted infections was negative. Esophagogastroduodenoscopy and colonoscopy showed mild gastritis with no intestinal involvement and no findings of Inflammatory Bowel Disease on computerized tomography or biopsies. C-ANCA, P-ANCA serology and HLA B27 were negative, cultures showed no growth and ophthalmologic exam was normal. Patient was started on Infliximab achieving weight gain and near complete healing of the perineum. Patient continued treatment with Anti-Tumor Necrosis Factor (Infliximab) at a dose of 5mg/kg Intravenous infusion every 2-3 months for 3 years. She

did well and the treatment was stopped, followed by recurrence of perineal and a new onset of oral ulcers. A pathology test became positive and a diagnosis of Behçet's was made. She was restarted on Infliximab with resolution of symptoms. Behçet's disease is rare entity in the pediatric population. Perineal lesions could resemble Crohn's disease. It is important to remember that the differential diagnosis of perianal lesions in patients with weight loss, mouth and perineal ulcers with positive pathology include pyoderma gangrenosum and Behçet's disease. More important is to know the dramatic resolution of lesions with the use of Anti-Tumor Necrosis Factor medications like in this case. The patient eventually fulfilled criteria for Behçet's with the development of oral ulcers and the positive pathology test. No evidence of Inflammatory Bowel Disease was seen in this child, but she responded dramatically to our treatment with resolution of all the lesions described. We provide dramatic images of the lesions in this child and post treatment resolution.

CELIAC/EOE/ALLERGIC ENTEROPATHY

186 IPEX-LIKE SYNDROME WITH PREDOMINANTLY ENTEROPATHIC PRESENTATION IN A 10 YR OLD MALE. H. Bhatt, Pediatrics, University of Arizona, Tucson, Arizona, UNITED STATESM. Daines, Pulmonology/Allergy-Immunology, University of Arizona, Tucson, Arizona, UNITED STATESA. Har, Gastroenterology and Nutrition, University of Arizona, Tucson, Arizona, UNITED STATES.

Background: IPEX syndrome is a rare syndrome resulting from defects in regulatory T cells with underlying FOXP3 gene involvement and characterized by immune dysregulation, polyendocrinopathy, enteropathy and x-linked inheritance. The FOXP3 gene is located at Xp11.23q13.3 and produces FOXP3 protein or scurfin. FOXP3 protein is critical for the development of naturally occurring regulatory T cells and its absence results in autoimmunity and inflammation. Other genes, including STAT1, CD25, STAT5B, and ITCH, that directly or indirectly affect the regulatory T-cell functioning can result in syndromes that are phenotypically similar to IPEX syndrome but lacking the characteristic FOXP3 gene mutation. Case: A 10 year old boy with known history of left-sided multicystic dysplastic kidney disease, Hashimoto Thyroiditis, Type I diabetes mellitus, and bilateral eye cataracts presented with multiple episodes of severe electrolyte abnormalities. Initial preceding symptoms of cough with post-tussive emesis with minor episodes of watery bowel movements lead to increasing diarrhea, protein losing enteropathy, autoimmune hepatitis, portal hypertension, and ultimately severe malnutrition necessitating total parenteral nutrition with complete bowel rest. Genetic testing for FOXP3 mutations was negative. Endoscopy was significant for intraepithelial lymphocytosis, goblet cell depletion, and villous blunting in the duodenum; acute and chronic inflammation with focal intestinal metaplasia in the stomach consistent with chronic atrophic gastritis. Liver biopsy revealed autoimmune hepatitis. Skin punch biopsy showed lichenoid dermatitis. With continued electrolyte abnormalities, hypoalbuminemia, and coagulopathy, a trial of immunosuppression with Sirolimus was started. His relapses have since become less frequent and severe. Conclusion: Patients lacking a FOXP3 gene mutation, but have a constellation of symptoms resembling IPEX syndrome, may respond to treatment modalities for IPEX including immunosuppression and possibly allogeneic bone marrow transplant.

187 PEDIATRIC CELIAC DISEASE PRESENTING AS ACUTE INTESTINAL PSEUDO-OBSTRUCTION. T.A. Altepeter, S. Shaffer, Pediatric Gastroenterology, Nemours Al Dupont Hospital for Children, Wilmington, Delaware, UNITED STATESA.J. Sloane, Pediatrics, Nemours Al Dupont Hospital for Children, Wilmington, Delaware, UNITED STATES.

Case: A previously healthy 16 month-old female presented with constipation, abdominal distention, irritability and poor oral intake. She had symptoms of gastroenteritis 2 weeks prior. The patient failed to respond to bowel cleanout and ultimately was admitted to the hospital with presumed post-infectious ileus. On examination she appeared fussy, pale and uncomfortable. Her abdomen was soft with marked distention, hypoactive bowel sounds, and diffuse tenderness to palpation. An obstruction series demonstrated dilated loops of small bowel and scattered air-fluid levels in both the small and large bowel, consistent with ileus. A computed tomography scan of the abdomen identified distended loops of small bowel without evidence of mechanical bowel obstruction or mass. She was trialed on large volume enemas, erythromycin and senna to stimulate gut motility, with minimal change. Celiac serologies were found to be positive, and subsequent duodenal biopsies demonstrated active duodenitis and villus blunting, consistent with celiac disease. She improved dramatically with introduction of gluten free diet, and has remained well with no further symptoms of dysmotility in followup. Discussion: This case represents a severe and unusual presentation of celiac disease in a young child. There are rare published case

reports suggesting a paralytic ileus / pseudo-obstructive presentation of celiac disease in the literature, and only one in a pediatric patient. The only reported pediatric case involved an 8 year-old female presenting with acute abdominal distention and pain. In contrast to our patient, she had acute pseudo obstruction affecting only the colon (Ogilvie's syndrome). Abnormalities in GI motility are well described in celiac disease. These can range from delayed gastric emptying and delayed colonic transit time to accelerated colonic transit time. Such abnormalities may contribute to the symptom variability seen in celiac disease, ranging from bloating, constipation and abdominal distention, to frank diarrhea. The mechanism underlying altered motility in celiac disease is not fully understood. Low levels of postprandial cholecystokinin (CCK) have been documented in untreated celiac disease. This may delay transit by decreasing excretion of bile, which in turn results in decreased absorption of fat, which will slow GI motility. Alternatively, neurotensin (a peptide associated with decreased gastric motility), was demonstrated to be elevated in untreated celiac patients with gastroparesis. In general, intestinal inflammation has been shown to cause dysmotility. This may be mediated in part by direct alterations in smooth muscle contractility affected by pro-inflammatory cytokines (such as IL-6, TGF- β , TNF- α) and by alterations in expression of calcium channels. Paralytic ileus and pseudo-obstruction are very challenging medical conditions to treat. Treatments range from supportive care with decompression and TPN, to medical therapies such as neostigmine infusion, and active interventions such as decompression via colonoscopy.

188 HALF A HETERODIMER IS ENOUGH TO INCREASE CELIAC DISEASE SUSCEPTIBILITY. M.M. Leonard, P. Cureton, M. McInnis, H.S. Winter, A. Fasano, Pediatric Gastroenterology and Nutrition, Massachusetts General Hospital for Children, Boston, Massachusetts, UNITED STATES. P. Cureton, School of Medicine, University of Maryland, Baltimore, Maryland, UNITED STATES.

Celiac disease is an autoimmune enteropathy that occurs in genetically predisposed individuals who have ingested gliadin; a protein in wheat, rye, and barley. More than 95% of individuals have the human leukocyte antigen (HLA) DQ2 or DQ8 haplotype which contributes to the pathogenesis of the disease. A six year old boy presented with six weeks of abdominal pain, nausea, vomiting, diarrhea, and weight loss. Outpatient serology and stool studies were normal including IgA (117mg/dl) and IgA tissue transglutaminase (tTG). IgG tTG which was slightly elevated at 16.8 (normal <15). An abdominal CT was normal. An upper endoscopy with biopsies of the duodenum was significant for variable villous blunting and increased epithelial lymphocytes. HLA DQ2 and DQ8 testing was sent to confirm the diagnosis and returned negative. The patient underwent testing for autoimmune enteropathy which also returned negative. Subsequent genetic testing revealed that the patient carried half of the at risk DQ2 heterodimer DQA1*05. The patient was placed on a gluten free diet with resolution of symptoms and normalization of intestinal villous architecture one year after initiation of the gluten free diet. Conclusion: Diagnosis of celiac disease in a patient without HLA DQ2 and/or DQ8 is exceptionally rare. Half of the HLA DQ2 heterodimer even in absence of positive serology is sufficient to increase susceptibility and should be looked for in complex cases to confirm a diagnosis.

189 CASE REPORT OF BUDESONIDE INITIATED MAINTENANCE OF EOSINOPHILIC ESOPHAGITIS PERSISTING AFTER WEANING OFF STEROID.. N. unnikrishnan, Department of Pediatrics, Downstate Childrens Hospital, Brooklyn, New York, UNITED STATES. F.J. Windemuller, J. Xu, S. Schwarz, S.S. Rabinowitz, Department of Gastroenterology, Downstate Childrens Hospital, Brooklyn, New York, UNITED STATES. R. Gupta, Pathology, Downstate Medical Center, Brooklyn, New York, UNITED STATES. E. Grossman, Division of Digestive Diseases, Downstate Medical Center, Brooklyn, New York, UNITED STATES.

Background : Eosinophilic Esophagitis (EoE) is a chronic inflammatory disease that remodels the esophagus with an unclear long term prognosis. While topical steroid treatment is an acceptable mode of therapy, the potential for long term sequelae and the criteria for weaning are unknown. Preliminary work has suggested that endoscopic ultrasound (EUS) can supplement histology to provide insight into EoE progression. Case report: An 16 y/o male with EoE was seen for a second opinion for progressive dysphagia. An upper endoscopy (egd) confirmed active EoE (see table 12/11) with increased eosinophils in the mid and distal esophagus. EUS demonstrated a thickened esophageal wall with a total wall thickness (TWT) measured at 2.9 and 2.6mm (see table 12/11) . He experienced clinical improvement after initiation of dietary restriction (based on allergy testing) and proton pump inhibitor (PPI) therapy but repeat egd documented active EoE. EUS continued to demonstrate increased TWT (see table 6/12). As he continued to have eos and only had minimal compliance with dietary restriction (min), he was started on swallowed budesonide (BDS) 2 g per day. He had an excellent clinical response to topical steroids with

resolution of the dysphagia. Serial egd and EUS confirmed that the eosinophilia had resolved, minimal inflammation was seen on biopsy and the thickness of the esophagus was decreased (see table 1/13). After a year of BDS therapy, egd showed a normal esophagus. Biopsies were also normal and TWT had reached normal values (see table 7/13). A decision was made to perform a six month trial off of steroids. He continued with the same dietary restriction and PPI therapy. After six months he continued to be asymptomatic. Endoscopic biopsies were normal without eosinophils and EUS continued to show a normal TWT (see table 12/13). Conclusions: 1.This case demonstrates that EUS can participate in EoE clinical decision making. 2.As previously described, BDS can result in remission of EoE and reversal of TWT. 3.In this patient, maintenance continued even after weaning of steroids. This management strategy would be parallel to the therapeutic approach often employed in adolescents and adults with ulcerative colitis.

Chronology of EoE features as a function of therapy

Eos- Eosinophils/high power field, BDS- Budesonide, TWT- total wall thickness, Min- minimal diet restriction

CLINICAL/TRANSLATIONAL IBD

206 MULTI-SYSTEM THROMBOSIS HERALDING ULCERATIVE COLITIS. R. Ahlawat, K. Buzzi, T. Weinstein, M.J. Pettei, J. Markowitz, Pediatric Gastroenterology, NSLIJ, Glen Oaks, New York, UNITED STATES. Although rare, Ulcerative colitis (UC) is associated with an increased risk of thrombosis. Venous thrombosis is more common than arterial thrombosis, and occurs most commonly in cerebral (54.3%), followed by extremity (26%), abdominal (13%) and pulmonary (3.3%) veins. We present a rare case of an adolescent female, who presented with cerebral thrombosis and developed multiple thrombi involving the portal, superior mesenteric, splenic and iliac veins, and was subsequently diagnosed with UC. A 14 year old previously healthy female presented with a 2 week history of bloody diarrhea, non-bilious, non-bloody emesis and a 7 pound weight loss. Two days later she developed left hemiparesis, and was transferred to our institution. Brain MRI and MR venography revealed a venous sinus thrombosis with associated hemorrhagic infarction. Laboratory evaluation was significant for Hct of 27.2, platelets 104,000, ESR 10, CRP 65 and albumin 2.4. PT of 19, INR 1.6 and PTT 81. Stool Clostridium difficile PCR was positive and diarrhea improved with oral vancomycin. Anticoagulation therapy with heparin was initiated. Thrombophilia workup including antithrombin III, factor VIII, thrombin time, fibrinogen, homocysteine and protein S free assay was normal except low protein C antigen and functional assay. She then developed altered mental status. An emergent head CT showed diffuse right-side cerebral edema with midline shift, and a decompressive craniotomy was performed. Ten days after admission she had worsening bloody diarrhea with abdominal distension. Abdominal MRI and MR venogram revealed thrombosis of the portal, superior mesenteric and splenic veins. Anticoagulation therapy was escalated. Empiric high dose IV steroids were started due to concern for Catastrophic Anti-phospholipid Antibody Syndrome (CAPS). Four days later she had profuse lower GI bleeding requiring multiple blood transfusions. A flexible sigmoidoscopy was performed showing diffuse inflammation. Biopsies revealed chronic active colitis, without evidence of CMV. EGD was normal. Infliximab (IFX) (5mg/kg/dose) therapy was initiated with decreased bleeding. 1 week later, as anticoagulation therapy was escalated, she again had significant lower GI bleeding requiring blood transfusions. Abdominal MR enterography and venogram showed pancolitis and new left common iliac vein thrombosis. IFX was increased to 10mg/kg with stabilization of bleeding. IBD serology returned positive for p-ANCA consistent with UC. Six weeks after admission, she continued to do well and was discharged home on therapeutic enoxaparin, steroids and IFX. Steroids were tapered and she has remained in remission with combination therapy on IFX and 6-MP. Thromboembolic complications are one of the more feared and rare extraintestinal manifestations of IBD. It usually occurs in children with known IBD during periods of active inflammation. This patient's rapid development of multiple thrombi in several venous systems prior to the diagnosis of IBD is particularly unusual. The diagnosis of IBD needs to be entertained in any pediatric patient who presents with multiple thrombi. Consideration of early therapy with biologics may be important for preventing major morbidity and mortality.

207 UTILITY OF MAGNETIC RESONANCE ENTEROGRAPHY FOR DETECTING EXTRA LUMINAL PATHOLOGIES IN PEDIATRIC INFLAMMATORY DISEASE PATIENTS. A. Anani, J. Moses, E. Collyer, M.J. Wyneski, pediatric gastroenterology, cleveland clinic, Cleveland, Ohio, UNITED STATES. Small bowel imaging at diagnosis for inflammatory bowel disease (IBD) is considered standard of care to establish extent of disease. Use of magnetic resonance enterography (MRE) is becoming the recommended modality due to

higher sensitivity and specificity and no exposure to radiation. Cross sectional nature of the study also allows for additional information of extra-luminal pathologies which are sometimes clinically relevant and lead to changes in medical or surgical management. The sensitivity of MRE is well established, but very little is published about extra luminal pathologies that may be discovered. We present two cases of IBD patients who had MREs at diagnosis and were found to have significant extra-luminal findings. Case 1: 16yo female with a family history of Crohn's disease presented with abdominal pain and hematochezia. Upper endoscopy (EGD) and colonoscopy showed active ulceration of the ileum. Pathology showed chronic inflammation in the terminal ileum and cecum. MRE of the abdomen showed features suggestive of inflammatory predominant Crohn's disease. No MRE features of stricture, bowel obstruction, or abdominal collections. She was noted to have atrophic left kidney with parenchymal scarring and dilated collecting system with mild right upper pole calyectasis concerning for reflux nephropathy. A consult was placed to nephrology due to the finding. Case 2: 17yo female with past medical history of ulcerative proctitis (UC) was seen as a second opinion for poor response to therapy. Repeat EGD and colonoscopy showed ulcerated mucosa 10 cm from the anal verge. Biopsies showed normal upper, normal terminal ileum, but patchy active colitis in rectum. MRE of the abdomen/pelvis showed no evidence of small bowel Crohns but did have two incidental findings: 1) incidental patulous cerebrospinal fluid filled distal spinal canal of the sacrococcygeal region and 2) avascular necrosis (AVN) in left femoral head. She subsequently had surgical repair for both findings. She was switched to steroid sparing regimen of MTX due to finding of AVN. She did not have good control of her UC and subsequent EGD/Colonoscopy showed progression of her disease to a pancolitis. Her therapy was escalated to infliximab, and she has done well to date. Literature review shows little published cases of incidental findings on MRE in pediatrics but there is ample evidence to the utility of MRE as the imaging modality for IBD in pediatric patients. It has the advantages of being more sensitive than a small bowel (SB) series and less radiation than a computed tomography and SB study. The cross sectional nature of the MRE also allows for finding extra- luminal pathology. In their study, Jensen et al found extra-intestinal findings not related to CD in 25% of the patients. In another study by Herfarth et al, 12% of the patients had findings that were of major clinical significance. The most common major finding was abscess. Our cases add to the available literature and support the utility of the MRE in the management of IBD in pediatrics.

208

EARLY ONSET IBD IN A PATIENT WITH A SINGLE GENE MUTATION FOR FAMILIAL

MEDITERRANEAN FEVER. K. Blin, Residency Program , Nationwide Children's Hospital , Columbus, Ohio, UNITED STATESK. Watson, D. Jacob, S. Kim, Gastroenterology, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATESR. Abdul Aziz, Rheumatology, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES.

Abstract H.Z. is a former full term 5 year old male of Middle Eastern background who presented with a ten day history of fevers and diarrhea. He was breast fed the first 5 months of life and had normal growth and development. He then began to have daily emesis which coincided with initiation of formula and pureed foods. At 7 months of age, he began to have diarrhea and frequent fevers. His growth started to plateau and he was started on Neocate and pureed foods via nasogastric tube, and later G-tube. He underwent an extensive evaluation, including celiac serologies, genetic testing for CF and tests for HIV, chronic granulomatous disease and numerous infectious etiologies; all of which were negative. Abnormal tests included decreased CD3/ CD4 levels, one gene mutation for Familial Mediterranean Fever, and allergy to cow's milk protein. Upper endoscopies and colonoscopies were significant for duodenitis and lymphocytic colitis. The patient received oral budesonide and human IgG, which exacerbated his diarrhea and fevers. He was evaluated at a major children's hospital in the U.S. when he turned 3.5 years old. Immunological and infectious testing was repeated. In addition, TRAPS and other periodic fever syndromes were ruled out. The patient's symptoms persisted, resulting in poor growth and developmental delay. When episodes of worsening fevers and diarrhea were treated with antibiotics, he would improve for a few weeks but symptoms always recurred. Colchicine was started for FMF and infliximab for colitis. Per parental report, he initially improved on infliximab. He was seen at our hospital in 2013. All testing repeated at our hospital was negative, including tuberculosis, stool studies and HIV. Evaluations for IPEX FOXP-3 gene and oxidative burst assay were negative; IgA levels were decreased. Upper endoscopy and colonoscopy showed severe pancolitis sparing his terminal ileum, indicating disease progression. His pathology was most consistent with Crohn's disease and IV corticosteroids were initiated without improvement. Colchicine was continued and infliximab was restarted at a higher dose and he gradually improved. Infliximab did have to be stopped secondary to persistent tachycardia and mild left heart dilation and he was then started on enteral therapy. Discussion When infants present with colitis, allergic and infectious etiologies tend to be highest on the differential given their

prevalence. However, in patients with family history or from areas endemic to FMF, the possibility for genetic influence should be considered earlier. IBD etiology is thought to be multi-factorial, with genetics playing a larger role in patients with very-early onset. Kuloglu et al documented an 8 month old with Crohn's disease with FMF unresponsive to steroids until colchicine was added. Recent immunology reports show a substantial portion of patients with clinical FMF only have one MEFV mutation and respond to colchicine. As we learn more about FMF, it is likely we will find more associations with IBD. This could have important implications on treatment of young IBD patients, using colchicine and other treatments in addition to steroid and immunosuppressant therapy.

209 NASAL SEPTAL PERFORATION, A RARE EXTRA-INTESTINAL MANIFESTATION OF CROHN'S DISEASE: A CASE REPORT IN A TEENAGER AT CHILDREN'S HOSPITAL OF NEW JERSEY. C.R. Dike, J. Sinha, M. Ton, F. Sunaryo, Pediatric Gastroenterology, Barnabas health/CHONJ, Newark, New Jersey, UNITED STATESJ. Kumta, St. Barnabas Hospital, Livingston, New Jersey, UNITED STATES.

<p style=margin-bottom: 0in">Multi-organ extra-intestinal manifestations are present in Crohn's disease. Findings such as aphthous ulcers, episcleritis, arthritis, pyoderma gangrenosum, erythema nodosum and DVT are more commonly described. Nasal involvement such as epistaxis, nasal mucosal inflammation and nasal septal perforations are however rare. Only a few cases of nasal septal perforation have been reported in association with Crohn's disease. We report a case of a 17 year old Caucasian male, JQ, who denied any history of smoking or illicit drug use, referred to our facility with a 2 month history of abdominal pain, bloody diarrhea, vomiting, mouth ulcers, dysphagia and a 25lb weight loss. He was admitted at another hospital at the onset of his illness for fever , mouth sores and decreased PO intake and was treated with intravenous fluids He was re-hospitalized a week later with symptoms of facial rash, mouth ulcers, persistent fevers and dysphagia. During that admission, he was treated with and discharged on antibiotics, antifungals and antivirals. Subsequently, he developed abdominal pain, diarrhea with nocturnal stools and vomiting at which point he was referred to our facility. Physical examination at presentation was positive for a malnourished teenager with left lower quadrant tenderness and a healing ulcer on the hard palate. On day 4 of hospitalization, he developed erythema and mild irritation of his nares. ENT evaluation revealed an anterior septal perforation (1x1.2cm in size) with significant scabbing and crusting around the perforation with no active bleeding. Laboratory investigation at presentation was positive for anemia and electrolyte disarray including hypokalemia, hypomagnesemia, hypocalcemia and hypoalbuminemia. Further work-up also revealed increased anti-saccharomyces cerevisiae antibody IgG and positive fecal calprotectin. Limited sigmoidoscopy was performed due to significant inflammation which revealed large deep ulcerations in the distal rectum and sigmoid colon. Upper endoscopy grossly showed erythema in the gastric body with prominent gastric rugae. Pathology revealed edematous colonic mucosa with architectural distortion, predominant superficial inflammation and no granulomas. He was started on steroids after the colonoscopy and his symptoms improved. He was also started on TPN via a PICC line to maximize nutrition and subsequently developed a DVT at the PICC line site. He was discharged on steroid taper, mesalamine and lovenox . His symptoms had greatly improved on his follow up visit 1 month later and thrombosis resolved with anticoagulation.</p>

210 SUCCESSFUL MEDICAL MANAGEMENT OF DUODENAL STRICTURE DUE TO CROHN'S DISEASE. E.J. Foglio, M.G. Patel, U.P. Phatak, D.S. Pashankar, Pediatric Gastroenterology, Yale University, New Haven, Connecticut, UNITED STATES.

Background: Inflammatory type of crohn's disease is common in children compared to the stricturing type, which is uncommon. Specifically, duodenal stricture due to crohn's disease is very rare. We report a case of a teenage boy with a duodenal stricture secondary to crohn's disease. Case: 14-year-old male initially presented with severe and frequent emesis as well as abdominal pain. He was unable to eat and had a thirty pound weight loss over the previous three months. Prior to this, patient was well with no significant past medical history. There was no family history of inflammatory bowel disease. Physical examination was remarkable for soft, non-distended abdomen with diffuse tenderness. Laboratory analysis showed platelets 399 x1000/uL, albumin 3.3g/dL, and with normal hemoglobin, lipase, and inflammatory markers. Upper gastrointestinal series revealed diffuse luminal narrowing of the second portion of the duodenum, causing gastric outlet obstruction. Computed tomography scan confirmed findings of duodenal stricture and also showed thickened terminal ileum. Endoscopic evaluation was remarkable for severe ulceration of the duodenum and ileum as well as multiple aphthous ulcers alternating with normal colonic mucosa. Histology showed duodenitis, ileitis and minimally active chronic colitis. No granulomas were identified. Crohn's disease was diagnosed. He was treated with intravenous steroids , bowel rest and total parental

nutrition for two weeks. Once he was tolerating oral intake without pain and vomiting, patient was discharged home on prednisone taper and Pentasa. Two months later, patient returned with abdominal pain and vomiting as prednisone was tapered off. Surgical options were discussed. However, because of difficult location of stricture and high risk of morbidity, it was decided to try anti-tumor necrosis factor therapy. Patient was then started on infliximab 5mg/kg induction therapy and then 6 weekly maintenance. He improved symptomatically. Reassessment at 18 months showed improvement of stricture radiologically. Five years later, patient has been asymptomatic and growth is satisfactory. He remains on maintenance therapy with infliximab 5mg/kg every 6 weeks. Recent imaging shows persistence of stricture but no obstruction. Conclusion: We report an adolescent male with newly diagnosed crohn's disease manifested by duodenal stricture. Duodenal stricture due to crohn's disease is a very rare finding in children and there is very limited data available. Treatment options in adults usually include strictureplasty or resection but because of location of duodenum, these can be associated with significant morbidity. Our patient has been treated with infliximab for the past 5 years with improvement of stricture and lack of symptoms. This case illustrates the successful use of anti tumor necrosis factor agents in management of duodenal stricture due to crohn's disease.

211 INFLAMMATORY BOWEL DISEASE AND DIVERTICULOSIS IN AN ADOLESCENT WITH DIGEORGE SYNDROME. R. Uy, N. Jacobs, C. Mziray-Andrew, Pediatrics, Southern Illinois University, Springfield, Illinois, UNITED STATES.

DiGeorge syndrome (DGS) has a wide clinical spectrum and its gastrointestinal (GI) manifestations are numerous and varied, however inflammatory bowel disease (IBD) as an association is quite rare, moreover – diverticulosis linked to DGS is unheard of. We present a case of an adolescent with DGS who has IBD with co-occurring colonic diverticula. The patient is a 17 year old Caucasian male with DiGeorge Syndrome who presented with a 3 week history of loose watery diarrhea and hematochezia. Associated symptoms included severe, crampy abdominal pain especially when defecating, anorexia and poor feeding due to mouth/tongue sores, significant weight loss of 29% pre-illness and profound prostration. Physical exam findings showed the patient to be severely cachectic and dehydrated with multiple oral and tongue ulcers. Abdomen was noted to be scaphoid and diffusely tender on palpation, and rectal exam was grossly positive for hemoccult stool. Initial blood work on admission revealed anemia, elevated inflammatory markers and severe electrolyte abnormalities. He was managed with fluid resuscitation, bowel rest and empiric antibiotics. IBD serology 7 came back with multiple nucleotide polymorphism mutations detected. Magnetic Resonance Enterography showed diffusely abnormal colon with thickening, fat stranding and significant fibrosis demonstrating lead-pipe sign from a chronic inflammatory process. Following improvement and stabilization of his clinical status, EGD and colonoscopy with biopsy were performed which showed ulceration in the mid-esophagus, duodenitis, mucopus and pseudopolyps in the entire colon. A surprising finding of diverticula were present throughout the transverse and descending colon. Histologic finding of the biopsies revealed widespread inflammation from esophagus to the rectum. The patient was started on corticosteroid therapy on a long taper however due to breakthrough exacerbations, we started him on Infliximab therapy. GI symptoms associated with DGS include abdominal pain, vomiting, reflux and constipation, the mechanism underlying these symptoms are not well known, although hypotonia is a frequent cofactor. The hypothesis for the pathogenesis of IBD is multifactorial which include the interactions between the intestinal microflora and the mucosa leading to a dysregulated inflammation, intestinal dysbiosis and altered immune responses in a genetically susceptible individual. Diverticulosis is uncommon in the pediatric age group. Prevalence rate is approximately 1-2% under the age of 30. In genetic syndromes like Ehlers-Danlos syndrome and Marfan syndrome, abnormality in the extracellular matrix of the colonic wall including collagen, elastin and proteoglycans predispose them into developing diverticula at an early age. To the best of our knowledge, this is the first reported case of a patient with DGS who has IBD with co-occurring colonic diverticula. Given the high incidence of GI manifestations and disease in patients with genetic syndromes where primary immunodeficiency is a prominent component, clinicians must therefore be prudent in routinely evaluating these patients to prevent potentially irreversible tissue damage.

212 CROHN'S DISEASE PRESENTING AS ALLERGIC ENTEROPATHY IN A 3 YEAR OLD MALE. A. Lakhole, H. Naon, V. Bhardwaj, G. Bultron, C. Lin, Gastroenterology, Hepatology, Nutrition, Children's Hospital Los Angeles, Los Angeles, California, UNITED STATES. A. Lakhole, H. Naon, V. Bhardwaj, G. Bultron, C. Lin, University of Southern California, Los Angeles, California, UNITED STATES.

Case description – This is a 3 y/o male presented with bloody diarrhea and swelling of feet and scrotum. His symptoms initially started when solid foods were introduced around 1 year of age and shortly after that he began to develop diarrhea that progressed to bloody stools. He was exclusively breast fed during infancy. On exam, he appeared pale with distended abdomen, periorbital and pedal edema. On growth chart he was at 34 percentile for weight for age and 17 percentile for height for age. Laboratory tests showed hemoglobin of 5.4 g/dl, low albumin of 2.1 g/dl, elevated white cell count of 27,000/ μ l with 68% neutrophils. Stool studies was positive for clostridium difficile infection. Stool A1AT was elevated to 1.5 mg/g/dry stool (normal <1.0). Stool calprotectin was slightly elevated to 140mcg/g (normal <50). RAST IgE panel for various food tests were negative. Upper and lower endoscopy revealed mild erythema and friability of mucosa with nodularity only in the descending colon and overall appeared normal otherwise. Pathology showed increased plasma cells in the transverse and descending colon without any granulomas or cryptitis. Patient symptoms at this point were most consistent with allergic enteropathy and he was discharged home with a course of prednisone and semi elemental formula (Peptamen Junior). A month later, he was readmitted with similar symptoms. During the course of his illness, he had various systemic infections including IV site cellulitis, right sided pneumonia and tracheitis increasing our suspicion for immunodeficiency disorder. Work up for immune function tests including complement levels, immunoglobulin levels and Nitroblue- Tetrazolium (NBT) test for chronic granulomatous disease were done. The results were equivocal and could not confirm any specific immune disorder. During this admission, he also had colonic intussusception originating at hepatic flexure, which was successfully reduced. Repeat sigmoidoscopy showed increased erythema, edema and friability of mucosa compared to the prior endoscopy. Pathology showed cryptitis, crypt abscesses and crypt architecture distortion in sigmoid colon. Inflammatory bowel disease (IBD) serology panel was positive for Anti OmpC IgA, Anti A4 Fla2 IgG, Anti FlaX IgG suggesting Crohn’s disease. Stool calprotectin at this time was elevated to 1137 mcg/g. Stool A1AT was also elevated to 2.3 mg/g/dry stool. Patient was started on Prednisone and 6 mercaptopurine. At last follow up patient showed improvement in clinical symptoms and good weight gain. Discussion- This is a unique presentation of Crohn’s disease in very young pediatric patient who initially presented as allergic enteropathy and possible immunodeficiency, but repeat sigmoidoscopy and IBD panel suggested Crohn’s disease pattern. The recent data suggest that infantile IBD has become more prevalent and physicians need to be more aware of it. We learn from this interesting case that physicians should have high suspicion for IBD as a differential in very young patients with symptoms suggesting allergic enteropathy or immunodeficiency.

213 CROHN'S DISEASE AND RECURRENT HSV-1 INFECTIONS. P.L. Luna, M. Creecy, O. Almadhoun, Pediatrics, KUMC, Overland park, Kansas, UNITED STATES.

We report the case of a 16-year-old with severe Crohn’s disease (CD) and recurrent HSV-1 infections. The patient was diagnosed with Crohn’s colitis in 2009 and since then his disease has been resistant to many therapies including Infliximab. He has been successfully maintained on a combined therapy of Adalimumab 40 mg q weekly and Methotrexate. The patient’s course was complicated by recurrent C. difficile infections, which were a source of flare up of his disease. Three years after starting current medical regimen, he developed recurrent fevers, mouth sores, and loose stools suggestive for an acute flare up. C. difficile by PCR was negative and colonoscopy revealed active left side colitis. Biopsies were negative for CMV and HSV. Physical exam was also remarkable for multiple aphthous appearing ulcers in the buccal, lingual, and tonsillar areas, each with a grey membrane and surrounding erythema. Viral culture from these lesions was positive for HSV-1 by PCR. By report, the patient’s girlfriend had a history of recurrent mouth sores. The patient was treated with Acyclovir for 14 days with significant clinical improvement. The patient has had recurrent HSV-1 related symptoms for which he received three courses of Acyclovir before he was placed on Vancyclovir prophylaxis. This case report demonstrates the potential for HSV infection in CD and emphasizes the need to consider in the differential diagnosis for IBD flares, especially in patients who do not respond to medical therapy. Social history is a very important part of the medical history and should be even more considered in adolescents.

214 INFLAMMATORY BOWEL DISEASE AND AUTOIMMUNE THYROIDITIS IN A YOUNG WOMAN WITH CONGENITAL CHLORIDE DIARRHEA. S.D. Miller, D. Shores, Pediatrics, Johns Hopkins Medicine, Columbia, Maryland, UNITED STATES.

Congenital chloride diarrhea (CLD) is a rare, autosomal recessive disorder caused by mutations in the SLC26A3 gene, which encodes for a Cl⁻/HCO₃⁻ membrane exchange protein and leads to high output secretory diarrhea. A

Finnish case series suggests an increased prevalence of Inflammatory Bowel Disease (IBD) and elevated ESR in patients with CLD. Here, we report a young woman with CLD who developed IBD and autoimmune thyroiditis. Case description: A 16 year-old female with neonatally diagnosed CLD presented with increased stooling from a baseline of 4-5 liquid stools per day and hematochezia. Physical exam revealed a small perianal skin tag. Initial lab workup showed an elevated TSH of 15 mIU/L, Anti-thyroid peroxidase elevated at 1763 IU/mL, and ESR elevated at 40 mm/hr. Initial colonoscopy showed acute colitis with edematous folds and marked spontaneous hemorrhage. P-ANCA ELISA was elevated at 56 IU/mL. The patient was started on Mesalamine for nonspecific colitis and Levothyroxine for autoimmune thyroiditis. The patient initially had improvement in diarrhea back to baseline, but then developed high output bloody diarrhea. Repeat colonoscopy revealed diffuse colitis and biopsies showed cryptitis throughout the colon with aphthous ulceration in the right colon and crypt abscesses in the descending colon. Repeat ESR was elevated at 75 mm/hr, and the patient required frequent hospitalizations for hypokalemia due to high stool output. The patient was started on 6-mercaptopurine, but continued to have elevated ESR and bloody stools. Infliximab was then added, and she had improvement in diarrhea back to baseline. From a GI standpoint, she has stable symptoms and marked histologic improvement on repeat colonoscopy, but ESR has never completely normalized. Discussion: This patient presumably has two autoimmune/immune dysregulatory diseases, IBD and thyroiditis. The pathophysiology of CLD may predispose to development of autoimmune conditions, particularly of the gut. The Cl-/HCO₃⁻ transporter that is dysfunctional in CLD is also down-regulated by inflammation in IBD, which may suggest an etiologic relationship. Additionally, the Cl-/HCO₃⁻ transporter is involved in sulfate transport. Individuals with CLD have elevated levels of gut sulfate which is metabolized to hydrogen sulfide by gut bacteria, and can be toxic to colonocytes, leading to "leaky gut" and inflammation. This patient has had dramatic improvement with infliximab.

215 DEVELOPMENT OF COLONIC CROHN'S DISEASE AFTER CARDIAC TRANSPLANTATION. G. Noel, E. de Zoeten, Gastroenterology, Children's Hospital Colorado, Denver, Colorado, UNITED STATESB. Diamond, S. Auerbach, Cardiology, Children's Hospital Colorado, Aurora, Colorado, UNITED STATESE. Hoffenberg, Gastroenterology, Children's Hospital Colorado, Aurora, Colorado, UNITED STATESG. Noel, University of Colorado, Aurora, Colorado, UNITED STATES.

A 17 year old male with a history of restrictive cardiomyopathy status post orthotopic heart transplant 6 years prior (2007) presented with recurrent diarrhea, malnutrition, severe dehydration and acute kidney injury. In the first year after transplant, he developed steroid-resistant rejection treated with anti-thymocyte globulin (ATG), IVIG, muronomab-CD3 (OKT3), and total lymphoid irradiation; Mycophenolate mofetil was initiated for maintenance therapy. After diagnosis of post-transplant coronary artery disease in 2009, sirolimus was added. For 5 years prior to presentation, he had intermittent diarrhea associated with C difficile positivity leading to growth failure and repeated admissions for IV rehydration. While on tacrolimus, sirolimus and mycophenolate mofetil, the patient presented with diarrhea that led to severe dehydration and acute kidney injury requiring admission. Stool studies were negative for infectious etiology. CT scan showed thickening of entire colon without lymphadenopathy which was not suggestive of PTLT. Esophagogastroduodenoscopy with biopsy was normal while colonoscopy demonstrated normal ileum and numerous deep and wide ulcers with edema and fibrinous exudate most pronounced in the cecum, transverse colon, and descending colon. Histology demonstrated active colitis with patchy, acute neutrophilic cryptitis and chronic colitis with crypt architectural distortion and Paneth cell metaplasia. Immunohistochemical staining revealed multiple foci of positivity but characteristic CMV viral cytopathic changes were not seen. Treatment for CMV colitis with IV ganciclovir did not improve his diarrhea and weight loss. 3 months later, a repeat colonoscopy showed extensive fibrinous exudates, serpiginous linear ulceration, edema and friability in patchy distribution throughout the colon with normal intervening areas and a normal ileum. Histology showed moderate to severe chronic active colitis with skip lesions and no granulomas. CMV and adenovirus stains were negative. MR enterography showed normal small bowel but pancolonial wall thickening. He was diagnosed with inflammatory bowel disease, likely Crohn's given the presence of skip lesions. He was initiated on infliximab at standard dosing (10 mg/kg/dose) and had a rapid response to treatment. TPN was weaned off within a few weeks, he regained his weight, and he has not had a recurrence of diarrhea. Conclusion: Having reviewed the literature, this is the first pediatric case described of inflammatory bowel disease that developed after cardiac transplantation while on significant immunosuppression. Onset of IBD after solid organ transplantation is rare in the pediatric population but should be considered if infectious etiologies are not identified. Our case suggests that anti-TNF α therapy is effective in post-transplant inflammatory bowel disease.

ENDOSCOPY/POTPOURRI

225 A PEDIATRIC PRESENTATION OF GASTRIC MALT LYMPHOMA. T. Hofmekler, S. Park, J. Mas-Moya, G.M. Batsuli, B. McElhanon, S. Kugathanan, F. Keller, G. Tenjarla, Emory University, Scottdale, Georgia, UNITED STATES. Hofmekler, S. Park, G.M. Batsuli, B. McElhanon, S. Kugathanan, F. Keller, G. Tenjarla, Children's Healthcare of Atlanta, Atlanta, Georgia, UNITED STATES.

Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) is a slow growing B cell lymphoma that is commonly associated with *Helicobacter pylori* (*H. pylori*) infections in adults during their sixth and seventh decade. Here we present the case of a 15 year old girl, recently emigrated from Romania to the United States, who presented with epigastric pain and melena. Further history revealed anemia diagnosed in Romania and confirmed on admission. Her physical exam and vitals showed her to be hemodynamically stable but with epigastric pain on palpation. Endoscopy revealed a large ulcer on the lesser curvature of the stomach. Histology showed the lamina propria to be expanded by a population of mature-appearing lymphocytes and plasma cells. Rare *H. Pylori* like organisms were identified. Immunohistochemical staining revealed plasma cells to be lambda light chain restricted. Clonal rearrangement was detected via PCR for IgH and IgK gene rearrangements. Fluorescence in-situ hybridization (FISH) performed on paraffin embedded tissue did not show a t(11,18), a characteristic chromosomal translocation associated with MALT lymphoma that forms the API2-MALT1 chimeric gene. The patient was treated with Amoxicillin, Metronidazole and Omeprazole for the *H. Pylori* infection. CT scans of the chest, abdomen and pelvis were negative for metastasis. Bone marrow aspirate and core biopsy was negative as well. After completing antibiotic therapy, her melena resolved. She continued to have epigastric pain and was continued on high dose omeprazole. A repeat endoscopy three months after her initial assessment showed gastritis but resolved ulcer. Her Clo test and stool *H. Pylori* antigen test were negative. Histology revealed chronic active gastritis, no *H. Pylori* like organisms, and resolution of the MALT lymphoma. Her epigastric abdominal pain resolved at follow up, but she continues to be followed for resurgence of symptoms and serial stool *H. Pylori* antigen tests. Her family members are being evaluated for *H. Pylori* infection. Discussion: Our patient developed gastric MALT lymphoma without metastasis. FISH did not reveal a t(11, 18) and she was treated exclusively for *H. Pylori*. She responded well to antibiotic treatment and eradication of the *H. Pylori*. This presentation is atypical due to the age of the patient. *H. Pylori* is more prevalent in developing countries, including Eastern Europe. Being from Romania, our patient had a higher predisposition to an *H. Pylori* infection. However, MALT lymphoma has not been shown to be increased in the pediatric patients from countries with higher rates of *H. Pylori*. Conclusion: *H. Pylori* may be a predisposition for MALT lymphoma in certain patients. Here we show that gastric MALT lymphoma may present in a pediatric patient at risk for *H. Pylori* infections. Thus, MALT lymphoma needs to be considered when evaluating pediatric patients for *H. Pylori*.

226 MIMICKER OF ACUTE APPENDICITIS. A. Watts, S.K. Gupta, Riley Hospital for Children at Indiana University Health, Indianapolis, Indiana, UNITED STATES.

Omentum is a fat laden peritoneal remnant of embryological development and is anatomically divided into the greater and lesser omentum. Omental infarction (OI) is a rare cause of acute abdomen in children. It can mimic acute appendicitis, a common entity in childhood. Obesity is a known risk factor associated with development of this condition. With different imaging modalities, this rare diagnosis can be made, thus avoiding unnecessary surgical intervention such as an appendectomy. A trial of supportive medical management and antibiotics should be considered prior to surgery. We are describing a case of omental infarction and a specific radiological finding, a "swirling appearance", associated with it. A previously healthy six year old male was admitted with right lower quadrant (RLQ) pain and fever concerning for acute appendicitis. He had no other associated complaints. Physical examination was significant for tender RLQ with voluntary guarding. No masses were palpable. The initial comprehensive metabolic panel and complete blood count were unremarkable and sedimentation rate was borderline increased at 17mm/hr. Computerized tomography (CT) scan of the abdomen showed normal appearance of appendix but swirl-like inflammatory fat stranding in the RLQ, thus suggesting the diagnosis of omental infarction (OI). He improved on intravenous antibiotics and supportive care, and was discharged home five days later. This case emphasizes that OI should be considered a possible differential diagnosis in the pediatric population presenting with RLQ pain. Additionally, this case gives an insight to the view that obesity is a risk factor for OI and in keeping with epidemic of pediatric obesity, incidence of OI is increasing.

227

PNEUMATOSIS INTESTINALIS AFTER IRINOTECAN CHEMOTHERAPY FOR EWING SARCOMA. M.

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A twenty year old male with skull-based Ewing Sarcoma developed mild diarrhea during chemotherapy with vincristine, temozolomide, and irinotecan. His diarrhea progressively worsened. Stool tested negative for Rotavirus, *C. difficile* and Giardia, but he had positive fecal leukocytes and later was found to be positive for Cryptosporidium. Nitazoxanide was then started, but with increased stool output he received a trial of loperamide and octreotide. Patient then developed emesis that was gastrocult positive, increased diarrhea, and a fever of 38.3 celsius. His abdominal exam showed slight distention and an abdominal x-ray showed possible small bowel obstruction and pneumatosis concerning for bowel ischemia. Broad spectrum antibiotics with anaerobic coverage were initiated. CT abdomen and pelvis confirmed diffuse pneumatosis intestinalis, pneumatosis coli, and gastric pneumatosis with mild proximal small bowel distention. He was kept on bowel rest, and serial imaging was followed and showed gradual improvement of pneumatosis. His diarrhea and abdominal distension were also improved, and the patient gradually advanced to a regular diet and was weaned off of parenteral nutrition. Irinotecan is being used as a novel agent in the treatment of pediatric solid tumors. It acts as a DNA topoisomerase-1 inhibitor. It is well-known that the main dose limiting toxicities of Irinotecan (also known as CPT-11) are neutropenia and late-onset diarrhea. A popular hypothesis for the cause of diarrhea is that irinotecan disrupts tight junctions in the intestine to cause intestinal mucositis leading to diarrhea. Certain risk factors including neutropenia, previous or current infection, and genetic polymorphisms may also predispose a patient to diarrhea. Association between irinotecan and pneumatosis is rare but has been reported in a few cases previously. This case highlights this rare association. Early recognition of this potential association is important when using irinotecan as part of the chemotherapy regimen.

228

INTESTINAL HEMANGIOMA PRESENTING AS RECURRENT RECTAL BLEEDING IN A 6-WEEK-OLD

MALE. G.A. Morris, L. Stratchko, M. Sabri, Pediatrics, Geisinger Health System, Danville, Pennsylvania, UNITED STATES.

Introduction: In infants, gastrointestinal bleeding can be caused by several conditions and many of which require different therapeutic interventions. The purpose of this case report is to shed light on intestinal hemangioma as an uncommon cause of gastrointestinal bleeding in infancy and present its diagnostic challenges. **Case Description:** We present a 6 week old male, born at 35 weeks gestational age and with a history of failure to thrive, rectal bleeding, and anemia (hemoglobin 6.0 g/dL). Multiple imaging studies were not suggestive of intussusception, Hirschsprung's disease or Meckel's diverticulum. He was started on Neocate for presumed allergic colitis, his bowel movements improved and he began gaining weight appropriately. An abdominal ultrasound was significant for counter clockwise rotation of the superior mesenteric vein with associated dilation and surrounding echogenic mesentery. A single contrast barium upper gastrointestinal series revealed anterior positioning of the proximal duodenum but normal location of the ligament of Treitz, making the study indeterminate for malrotation. He was discharged in stable condition, however seven days after discharge was readmitted with recurrent hematochezia, anemia, and irritability. An abdominal MRI was performed and showed an enhancing, infiltrating mesenteric mass with mass effect on the transverse colon and displacement of small bowel. On exploratory laparotomy, diffusely edematous small bowel completely covered by what appeared to be a hemangioma versus arteriovenous malformation was found and biopsied. Pathology revealed benign capillary hemangioma with diffuse GLUT1 positivity and focal areas of extension through the small intestinal wall, reaching peri-intestinal soft tissue. Oral feeds were attempted and resulted in increased melanic stools, he was made nil per os, started on total parenteral nutrition, propranolol 1 milligram per kilogram per dose twice a day, and methylprednisolone 0.5 milligram per kilogram per dose twice a day. Early repeat imaging suggests the lesion is responding to pharmacotherapy with the most recent CT scan showing the mass is improving when compared to initial MRI. **Discussion:** Visceral vascular anomalies are a diagnostic challenge in the pediatric population and it is imperative to make an accurate diagnosis as treatment options are drastically different. This patient's initial diagnostic work up was inconclusive. When imaging and histological evidence were paired, we were able to make the diagnosis and initiate appropriate treatment. This case is unusual because retrospectively, his imaging was suggestive of malrotation, however, an MRI was consistent with an infiltrating mesenteric mass that was confirmed to be an intestinal hemangioma. Early recognition and treatment of this pathology may result in prompt initiation of medical treatment, resolution of symptoms, quicker reintroduction of enteral nutrition, and improved quality of life for the patient and family.

229

NUTCRACKER PHENOMENON PRESENTING AS ABDOMINAL PAIN IN A TEENAGE FEMALE. A.L.

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BACKGROUND: Nutcracker syndrome (NCS) is a rare but increasingly recognized condition that involves compression of the left renal vein (LRV) between the abdominal aorta and the superior mesenteric artery. Pain may occur as the result of venous hypertension and may cause a wide variety of symptoms including headache, abdominal pain, and dysmenorrhea. Hematuria, proteinuria, flank pain, pelvic congestion, and varicoceles may also occur. Young adults have a higher prevalence of NCS due to the rapid increase in height of adolescents leading to the narrowing of the angle between the aorta and SMA. We present the case of a young woman with recurrent, intermittent abdominal pain who was found to have NCS. **Case:** A 17 year-old female presented to the GI clinic with a three month history of recurrent, intermittent severe left sided abdominal pain, anorexia, and nausea. She described her pain as sharp and stabbing, without radiation that worsened with weight lifting and menstruation. Over the preceding three months, she was treated for constipation and a UTI with some improvement in her symptoms. Her abdominal pain recurred leading to an evaluation by pediatric surgery. A CT scan was concerning for an omental infarction. She was subsequently admitted to the GI service where a work up including CBC, BMP, lipase, amylase and hypercoagulation panel were negative. It was felt the omental infarction was not the etiology of her pain. She had two urinalyses: one with large blood and one with trace blood, but both without proteinuria. The hematuria found in the two specimens was believed to be due to menstruation which had recently begun. For the next nine months, she had multiple visits to the ED, adolescent clinic and GI clinic for her persistent intermittent abdominal pain. She was treated for possible endometriosis with an OCP, chronic H. pylori gastritis found on upper endoscopy, and musculoskeletal abdominal pain. Repeat labs and imaging continued to be within normal limits, although an additional UA had trace lysed blood cells with unknown correlation to her menstrual cycle. During another episode of abdominal pain, an abdominal CT angiography was obtained. It showed the left renal vein was nearly occluded due to the narrow angle between the SMA and aorta, consistent with NCS. Following the evaluation by two adult vascular surgeons, the patient underwent surgery and her abdominal pain has resolved. **Discussion:** Severe, intermittent abdominal pain as the primary presenting symptom of NCS is rare without supporting evidence of proteinuria or hematuria outside of menstruation. Up to 16% of children and adolescents have been found to have compression of the LRV as determined by flow acceleration on routine ultrasound, most of which are asymptomatic. This case emphasizes the importance of keeping NCS within the differential for children with abdominal pain due to the clear treatment options available and side effects that may occur secondary to not treating. Failure to diagnose this rare but treatable cause of abdominal pain may lead to complications including weight loss, pelvic congestion, varicoceles, and worsening flank and abdominal pain.

230

UNUSUAL PRESENTATION OF DYSPHAGIA IN A 17 YEAR OLD MALE; HODGKIN'S LYMPHOMA

WITH ESOPHAGEAL COMMUNICATION OR ESOPHAGEAL DUPLICATION CYST AND HODGKIN'S LYMPHOMA. E. Park, U. Udayasankar, Pediatric Radiology, Cleveland clinic, cleveland, Ohio, UNITED STATES. A. Anani, K. Radhakrishnan, pediatric gastroenterology, cleveland clinic, Cleveland, Ohio, UNITED STATES. D. Magnuson, Pediatric Surgery, Cleveland clinic, cleveland, Ohio, UNITED STATES. R. Hanna, Pediatric Hematology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES.

We present an uncommon cause of dysphagia with an esophageal communication complicating its management. 17yo male with 3 month history of epigastric pain, dysphagia and unintentional weight loss seen in outpatient GI practice, was started on a proton pump inhibitor but with no interval improvement. Endoscopy (EGD) revealed esophageal erosions and narrowed lumen. Further work up with a CT and MRI revealed a thick walled cavitary lesion 4.5 x 3 cm predominantly air filled structure communicating with the esophagus in the right posterior mediastinum approximately 5 cm from the gastro-esophageal junction with surrounding inflammatory changes. Blood work was unremarkable except for elevated inflammatory markers and Infectious work up was negative. Repeat EGD was done to obtain tissue to rule out malignancy and placement of nasogastric tube for nutrition but revealed two narrow lumens with a tight stricture. A water soluble esophagogram a well-defined cavity measuring 3.9 x 2.8 cm communicating with the right lateral aspect of the distal esophagus, suggestive of an esophageal duplication cyst. Surgical placement of a percutaneous endoscopic gastrostomy tube revealed an internal umbilical hernia with intra-abdominal lymphadenopathy. Biopsy revealed nodular sclerosing Hodgkin's lymphoma. Bone

marrow aspiration/biopsy was normal and PET/CT-scan showed that the esophageal mass as well as neck and mediastinal lymph nodes were intensely FDG avid- indicating active lymphoma. Some suspicious FDG uptake was seen between the rectus abdominal muscles. Hodgkin's lymphoma was managed with chemotherapy with careful consideration for the possibility of mediastinitis as the mass shrinks. Hematology, Surgical, Radiologic and GI teams were closely involved in his management. We present this interesting case of dysphagia due to Hodgkin's lymphoma with an unusual communication with the esophagus. A review of literature suggests that while dysphagia from mediastinal masses has often been reported, there are very few cases in which there is a communication with esophagus. Mediastinal masses should be considered in the differential for dysphagia in pediatrics. In a patient with dysphagia and weight loss an imaging modality should be considered along with endoscopy. The question still remained; Hodgkin's lymphoma with esophageal communication or esophageal duplication cyst and Hodgkin's lymphoma. Persistence of cyst post chemotherapy will make the diagnosis of esophageal duplication cyst coexisting with the Hodgkin's lymphoma a possibility.

231 NON-CLASSICAL HISTOLOGIC AND GENETIC FINDINGS IN MICROVILLUS INCLUSION DISEASE. A. Sepulveda, Columbia University Medical Center, New York, New York, UNITED STATESJ. Picoraro, E. Lamouse-Smirh, Pediatric Gastroenterology, Hepatology and Nutrition, Columbia University Medical Center, New York, New York, UNITED STATESY. Yilmaz Demirdag, Pediatric Allergy and Immunology, Columbia University Medical Center, New York, New York, UNITED STATESN. Wontakal, J.M. Mitchell, V. Aggarwal, Pathology and Cell Biology, Columbia University Medical Center, New York, New York, UNITED STATESA. Iglesias, Clinical Genetics, Columbia University Medical Center, New York, New York, UNITED STATES.

Microvillus inclusion disease (MVID) is a rare autosomal recessive disease of neonatal onset that causes intractable, life-threatening watery diarrhea curable by small bowel transplant. Mutations in the MYO5B gene are implicated in the etiology of the disease. Histological findings include brush border atrophy, intracytoplasmic microvillus inclusions and apical membrane vesicles. However, variants of these ultrastructural findings have also been described. We present a patient with a clinical presentation consistent with MVID, but with atypical electron microscopic (EM) findings thought to be secondary to unique mutations identified in the MYO5B gene. The patient presented with voluminous diarrhea within the first day of life that resulted in hypovolemic shock. Initiation of total parenteral nutrition (TPN) was required to manage stool output of >50ml/kg/day and resulted in appropriate weight gain. Infectious stool studies were negative. Attempts to advance enteral feeds with an elemental formula were limited by recurrence of hypovolemic dehydration. Duodenal biopsies showed villous atrophy. Chromogranin and Ep-CAM staining confirmed normal neuroendocrine cell expression and no evidence of tufting enteropathy. Presence of apoptotic bodies in epithelium and lamina propria and an absence of lymphocytosplasmic infiltrate prompted an extensive evaluation for immunologic causes, all of which were excluded. EM of intestinal biopsy specimens did not detect the classic intracytoplasmic microvillus inclusions or vesicular bodies attributed to MVID. Many normal small bowel microvilli were present with interspersed enterocytes displaying disordered and poorly formed microvilli. Occasional lateral microvilli were observed between enterocytes. Notably, there was the near absence of and poorly formed colonic microvilli, a finding not previously reported in the literature. The findings persisted on serial biopsies performed at 3 and 6 months of age. To determine a possible genetic etiology, whole exome sequencing was performed in the patient and his parents. Sequencing revealed the patient has compound heterozygous mutations in MYO5B with a paternally inherited nonsense mutation and a putative splice site mutation that is maternally inherited. These variants have not been previously identified, and therefore, may explain the atypical presentation observed in this patient. At 8 months of age he continues to have stool output of 70 ml/kg/day. He is TPN dependent for greater than 95% of nutritional and fluid needs and has displayed normal growth and development. This case report presents a case of MVID with non-classical histologic and genetic findings, which may suggest a clinical-pathologic spectrum of disease in patients with this condition. Importantly, the abnormal colonic findings raise intriguing questions about future options for surgical management of this patient. This case illustrates the importance of careful clinical, pathologic, and genetic evaluation for causes of intractable diarrhea in infants presenting in the first weeks of life.

232 MENETRIER MIMICKER COMPLICATING ULCERATIVE COLITIS. P. Bishop, N. Tipnis, M.J. Nowicki, Pediatric Gastroenterology, Univ Mississippi Medical Center, Jackson, Mississippi, UNITED STATESN.T. Parks, School of Medicine, University of Mississippi Medical Center, Jackson, Mississippi, UNITED STATESC. Subramony,

Pathology, University of Mississippi Medical Center, Jackson, Mississippi, UNITED STATESK. Ragland, M. Steiner, Pediatric Radiology, University of Mississippi Medical Center, Jackson, Mississippi, UNITED STATES.

A 12-year-old girl with a 3-½ year history of hard to control ulcerative colitis (UC) presented with an exacerbation manifested as abdominal pain, arthralgia, vomiting, and bloody diarrhea. She had been receiving infliximab for 2 years and esomeprazole (20 mg/day) for gastritis since diagnosis. Examination showed mild tachycardia, pallor, diffuse abdominal tenderness, and bloody stool on digital exam. Laboratory evaluation showed microcytic anemia, thrombocytosis, and hypoalbuminemia. Antibodies to infliximab were positive. Colonoscopy showed severe pancolitis. EGD showed hypertrophied gastric folds and polypoid lesions. The initial concerns for the EGD findings included anti-TNF α induced lymphoma and Ménétrier disease (MD). A pinch biopsy showed severe gastritis with foveolar hyperplasia. Adalimumab was started and esomeprazole increased (40 mg/day). She developed an adalimumab allergy 4 months after initiation. Repeat panendoscopy prior to colectomy showed no changes in the findings. A snare biopsy of the gastric lesion was consistent with a hyperplastic polyp with proton-pump (PPI) effect. Gastric hyperplastic polyps (HPP) are mimickers of MD, a condition characterized by rugal hypertrophy, protein-losing gastropathy, and hypoalbuminemia. MD can occur spontaneously, result from infection (*Helicobacter pylori* and cytomegalovirus), or be associated with UC. Transforming growth factor (TGF)- α is thought to play a major role in the development of MD through binding to epithelial growth factor receptors and leading to gastric epithelial cell proliferation. MD has been seen complicating ulcerative colitis, related to increased expression of TGF α in UC. Proton-pump inhibitors (PPI) are used to treat MD. Long-term PPI use has been associated with development of fundic gland polyps (FGP) and HPPs, with an increased prevalence in patients using PPIs for greater than 12 months. In patients on PPI maintenance therapy for 2-years, FGP developed in 14% and HPP developed in 9%. The proposed mechanism is that PPIs lead to high gastric pH stimulating gastrin release causing parietal cell hypertrophy with protrusion into the gland lumen, this leads to obstruction and subsequent cyst formation resulting in polyps as the cysts enlarge.

233 CONSERVATIVE MANAGEMENT OF GASTRIC OUTLET OBSTRUCTION IN A PEDIATRIC PATIENT: A CASE REPORT. S. Rohatgi, A. Martinez, F. Ashai-Khan, Pediatric Gastroenterology, Medical College of Wisconsin, Milwaukee, Wisconsin, UNITED STATES.

Introduction: Gastric outlet obstruction (GOO) in pediatric population after the first few weeks of life is uncommon and mostly requires surgical interventions. Our patient presented with an uncommon etiology of pediatric GOO, which was amenable to successful non-surgical advanced endoscopy management. Case: A 3 year old previously healthy female with 3 weeks of nausea, vomiting and abdominal distension that started within 2-3 days of Azithromycin and ibuprofen treatment for an upper respiratory tract infection. She was admitted twice to another hospital for dehydration before being referred to us for management of hypertrophic pyloric stenosis on ultrasound and a complete GOO on esophagogastrogram (UGI) and esophagogastroduodenoscopy (EGD). On transfer, a follow up EGD with pyloric Botox injection was done. Serial pyloric dilation with CRE Wire-Guided Boston Balloon dilator using increasing inflation pressures (ATM) up to 10 with inflated balloon diameter of 10 mm was done under fluoroscopy guidance. After the procedure, she was asymptomatic on PPI and high calorie liquid diet for a week but due to symptom relapse, pyloric dilation up to 12mm was reattempted for complete obstruction. A transparent porous vascular membrane surrounded by granulation tissue was noted beyond the dilated area and duodenal intubation was unsuccessful at this time. Biopsies from the antrum were consistent with drug induced injury. Follow up UGI showed dilated stomach with some contrast in the duodenum. At the repeat EGD a week later, duodenal intubation was successful using XP-160. Due to persistent narrowing, serial pyloric balloon dilations up to 12 mm were repeated. Now, 12 months post dilation, she is asymptomatic on regular diet and off medications. Discussion: Mechanical causes of gastric outlet obstruction are uncommon in pediatric population and largely managed by surgical interventions which may be associated with complications, immediate or remote. Advanced Endoscopic approach is not commonly used for pyloric area in pediatric patients but it is a possible therapeutic option for certain surgical conditions. Salah et al used the standard biliary needle-knife and electrocautery for snare resection and electroincision with serial balloon dilations of the gastric antral web causing GOO avoiding surgical intervention. In our case we were successfully able to use pneumatic dilations and Botox injection to dilate the pylorus and avoid surgery. Further, it is well known that most drug induced injuries in the stomach occur in the antrum due to chemical stasis in the region, pyloric spasm secondary to mucosal damage as well as protective prostanoid inhibitory effect of NSAIDs in antrum. We were able to microscopically confirm that the gastric outlet obstruction was the consequence of drug induced antral injury and subsequent pyloric spasm

caused by NSAIDs and possible Azithromycin leading to GOO and pyloric hypertrophy. Conclusion: GOO, possibly secondary to mucosal injury induced by a combination of NSAIDs and Azithromycin was successfully reduced by advanced therapeutic Endoscopic procedure, obviating the need for surgical intervention.

234 MAJOR LOWER GI BLEEDING IN AN ADOLESCENT BOY DUE TO LONG TERM ASPIRIN USE. I.

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Introduction: Upper GI bleeding and gastric ulceration are common complications of long term use of aspirin. GI bleeding from aspirin is rare in children compared to adults. The association between lower GI bleeding and aspirin is not commonly reported. We report a case of major lower GI bleeding in an adolescent boy who has been on enteric coated aspirin for 10 years. Case report: A 13 year old Caucasian boy presented with 3 days history of painless rectal bleeding. His hemoglobin dropped to 8.8 g/dL from the 13.5 most recent level. Our patient had Kawasaki disease at 3 months of age which resulted in coronary artery aneurysm. For that he was on low dose of enteric coated aspirin (81 mg) for 10 years and Coumadin (5mg) for 2 years. His work up included negative stool studies for enteric pathogens, IBD panel, celiac serology, and Meckel's scan. He underwent an upper endoscopy and ileocolonoscopy which were both normal, except for superficial ileal ulceration. Patient bleeding was attributed to aspirin induced small bowel Ulceration. His GI bleeding stopped after discounting aspirin. His hemoglobin improved to 10.5 g/dl after few days. At a follow up 4 weeks later he had no more GI bleeding and was restarted on Coumadin. Conclusion: Long term use of aspirin can result in small bowel ulceration and major lower GI bleed in pediatric patients. Children on long term aspirin should be watched for this complication.

235 RECTAL POLYP WITH ATYPICAL LYMPHOID INFILTRATE; RECTAL LYMPHOMA OR RECTAL TONSIL.

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Background: The rectal tonsil is a localized reactive proliferation of lymphoid tissue of the rectum that protrudes into its lumen. These rare benign polyps are usually located in the rectum just proximal to the dentate line. The rectal tonsil is also known as the benign lymphoid polyp and can present with intermittent rectal bleeding or pain. On endoscopy, these polyps appear benign in nature and resemble juvenile polyps. On the other hand, their histology mimics a gastrointestinal lymphoma, which is the most common extra nodal location of lymphoma. Diagnosis then requires additional immunohistochemical and genotypic studies for confirmation. The treatment for these polyps is local excision. It is important to become familiar with rectal tonsils and their histology to avoid misdiagnosing them as lymphoma. Case: LW is a well appearing 6 yr old male who presented to his PCP with 4 weeks of constipation and hematochezia. A rectal tear was noted on physical exam and stool softening with polyethylene glycol was initiated. At his follow up, his constipation had resolved but his intermittent hematochezia persisted. The patient underwent EGD & colonoscopy and a 1 cm pedunculated polyp in the distal rectum was excised by snare polypectomy and sent for pathology. The polyp was shown to have atypical lymphoid infiltrate involving the mucosa and the submucosa of the rectum initially concerning for rectal lymphoma. Immunophenotype and molecular studies were then performed which favor a reactive lymphoid hyperplasia. The remainder of the esophageal, gastric, duodenal, colonic, and terminal ileal biopsies showed no diagnostic abnormalities. Conclusion: Rectal polyps are a common cause of rectal bleeding. In most instances, rectal polyps are benign hamartomatous juvenile polyps; however they may also represent the rare and benign rectal tonsil. When the histology of such a polyp is suggestive of lymphoma which is also very rare, it is important to include rectal tonsil in the differential diagnosis. This can help decrease or avoid undue familial anxiety, additional medical testing and medical referrals.

236 NORMALIZED FECAL CALPROTECTIN AFTER SMALL INTESTINAL BACTERIAL OVERGROWTH

TREATMENT IN A RENAL TRANSPLANT PATIENT STATUS POST ILEOCECTOMY. S. Wadera, K. Kordy, E.A. Marcus, Department of Pediatrics, DGSOM at UCLA, UCLA, Los Angeles, California, UNITED STATES. E.A. Marcus, VA Greater Los Angeles Health Care System, Los Angeles, California, UNITED STATES.

Background: Fecal calprotectin is a widely used marker of gastrointestinal inflammation. Small intestinal bacterial overgrowth (SIBO) can present with nonspecific gastrointestinal symptoms of bloating, flatulence, abdominal discomfort, or malabsorption. A few small studies have demonstrated no correlation between fecal calprotectin and presence of SIBO (Montalto et al, 2008; Fundaro et al, 2011). We describe a case where treatment for SIBO did normalize elevated fecal calprotectin. Case Presentation: Our patient is a 7 year old male of mixed Asian-Caucasian descent with a history of congenital nephrotic syndrome, who received living related renal transplant at

age 3, also with history of intussusception at 4 days of life requiring ileocectomy. He initially presented at age 4 with symptoms of soft stool and abnormal stooling patterns, abdominal cramping, and nausea. He was placed on a lactose free diet and mycophenolate was changed to azathioprine. Both interventions improved symptoms. He had a lactulose breath test suggestive of bacterial overgrowth, and a negative lactose breath test. Treatment for bacterial overgrowth was not completed. Patient was lost to follow up until age 7, when he again presented with loose stools and abnormal stooling patterns. There was urgency to the visit due to inconsistent tacrolimus levels. He occasionally experienced cramping, distention, and abdominal pain. Complete work up of loose stools was initiated, including stool pH and reducing substances, alpha-1 antitrypsin, spot fecal fat, stool cells, calprotectin, and lactoferrin. Stool studies were negative for malabsorption. Stool inflammatory cells were absent. Lactoferrin and ESR were elevated. Fecal calprotectin was elevated at 202 $\mu\text{g/g}$ (normal < 50 $\mu\text{g/g}$). Lactulose breath test was positive. Treatment for SIBO was started with a 14 day course of metronidazole 250 mg TID. He experienced complete resolution of abdominal distention and diarrhea. Fecal calprotectin measurement 3.5 weeks after starting treatment revealed normalization to 41 $\mu\text{g/g}$. Discussion Calprotectin is a neutrophil protein found in plasma and stool that is elevated in infectious and inflammatory conditions. This patient had no other identifiable etiology of high calprotectin. Both calprotectin and symptoms improved after SIBO treatment. Patients with intestinal bacterial overgrowth may have subclinical intestinal inflammation. Testing and treatment for bacterial overgrowth should be considered for patients with elevated calprotectin in the proper clinical setting. Repeat levels should be checked after treatment, prior to proceeding with more invasive testing such as endoscopy. Further investigation is needed to better understand this association.

237 FIRST DOCUMENTED CASE REPORT OF EBV GASTROENTERITIS (EBV GE) IN AN IMMUNOCOMPETENT CHILD.. J.S. Di Palma, Gastroenterology, Al DuPont Hospital for Children, Wilmington, Delaware, UNITED STATESK. Conard, Pathology, Al DuPont Hospital for Children, Wilmington, Delaware, UNITED STATES E.T. Davis, C.M. Soprano, Pediatrics, Al DuPont Hospital for Children, Wilmington, Delaware, UNITED STATESZ. Molle Rios, Gastroenterology, Al DuPont Hospital for Children, Wilmington, Delaware, UNITED STATES. Introduction: EBV GE has been described in association with immunodeficiency states, underlying IBD, or GI malignancy. EBV GE in healthy immunocompetent individuals, however, is rare. Only 5 documented cases have been reported in adults, who presented with GI symptoms only after the classical symptoms of EBV infectious mononucleosis (IM). To our knowledge, there are no reported cases of EBV GE in healthy pediatric patients. We present a case of a 3 year old healthy immunocompetent male who presented initially with several weeks of severe vomiting and diarrhea and was ultimately diagnosed with EBV GE. Only after spontaneous resolution of his GI symptoms did typical symptoms of EBV IM appear. This case is unitarily unique with regards to the patient's young age, immunocompetent status, and chronology of symptom presentation. Case: A 3 year old male presented with 3 weeks of non-bloody diarrhea, vomiting, abdominal pain, and fever. ROS was negative for fatigue, cough, sore throat, or headache. PMH was significant for frequent infections raising concern for an underlying immunodeficiency. However immunoglobulins, LEP, vaccine titers, EBV titers, EBNA, and sweat test were normal. On initial PE, VS were normal and no pharyngeal erythema or lymphadenopathy were noted. Abdominal exam was benign, without hepatosplenomegaly. One week after initial presentation, he developed hematochezia, decreased PO intake, dehydration, and weight loss. Workup included CBC, CMP, ESR, CRP, Amylase/Lipase, PT/PTT, TTG IGA/IGG, and immunoglobulins which were all normal. Stool bacterial and viral cultures, O&P, stool pH and reducing substances were all negative/nl. Abdominal U/S was normal. Upper GI Series was concerning for inflammatory changes versus LNH in the TI. EGD and colonoscopy were significant for a small gastric ulcer and gastric erosions. Esophagus, duodenum, TI, and colon were grossly normal. Gastric biopsies showed a mixed cellular infiltrate composed of eosinophils and activated lymphocytes compatible with gastritis. Colonic biopsies were significant for scattered neutrophils in the lamina propria of the cecum and sigmoid colon, compatible with active colitis. Biopsies from the esophagus, duodenum, and TI were normal. In situ hybridization for EBER showed immunoreactivity in the stomach and cecum consistent with EBV GE. CMV and H. pylori stains were negative. Ten days later, GI symptoms spontaneously resolved but were followed by symptoms of high fever, fatigue, and sore throat with exam significant for pharyngeal erythema and cervical lymphadenopathy. At this point, EBV serology and PCR were positive for acute EBV IM. 4 weeks later, fever resolved, throat pain and fatigue improved, and EBV serology and PCR reflected past infection but resolution of acute infection. Conclusion: It is important to keep EBV GE in mind when creating a differential for acute diarrhea, hematochezia, and/or vomiting in immunocompetent pediatric patients; especially those with concomitant IM symptoms. Having a high clinical

suspicion for EBVGE in the setting of IM symptoms could save patients from undergoing invasive procedures, costly workup, and unnecessary treatment.

238 SIROLIMUS RESULTS IN RAPID IMPROVEMENT OF INTESTINAL HEMORRHAGE IN CHILD WITH BLUE RUBBER BLEB NEVUS SYNDROME (BRBNS). J.R. Whitworth, Pediatric Gastroenterology, University of Tennessee, Memphis, Tennessee, UNITED STATES.

Blue rubber bleb nevus syndrome is a rare condition characterized by venous malformations of the skin and particularly problematic within the gastrointestinal tract. Chronic anemia and dependence on transfusion of red blood cells is typical and major gastrointestinal hemorrhage is the primary cause of mortality. There is no established standard of care and previous therapies have relied upon surgical excision, sclerotherapy, octreotide, interferon, corticosteroids, cyclophosphamide, or propranolol. There is early but growing evidence that the effect of sirolimus on angiogenesis and cell growth is effective and safe in treating children with various types of vascular anomalies including BRBNS. We report the third case in literature and the youngest patient to respond to sirolimus for chronic anemia and recurrent gastrointestinal hemorrhage. Our patient was diagnosed with BRBNS at birth, started on iron therapy for chronic anemia by 13 months, started serial sclerotherapy for a large cutaneous lesion at 14 months, and began having major lower gastrointestinal bleeding requiring serial transfusions by 22 months of age. In addition to skin lesions her evaluation revealed many small vascular lesions in the stomach and several large lesions in the duodenum and colon by endoscopy. Whole body MRI revealed innumerable lesions within skin and soft tissue in the neck, tongue, buttocks, small and large intestine. At 25 months old, sirolimus 0.8 mg/m²/dose twice daily was begun resulting in immediate control of bleeding and she has not required RBC transfusions now 3 months into therapy which she is tolerating without adverse reaction. There has been visual improvement of skin lesions with complete regression of some lesions. This case presents a rapid response to sirolimus in a young child with gastrointestinal hemorrhage from BRBNS. Image 1-photo of skin lesions prior to therapy Image 2-photo of skin lesions on therapy Image 3-photo of intestinal lesions by endoscopy Image 4, 5-MRI of soft tissue and intestinal lesions (Images not attached in abstract submission)

239 ESOPHAGEAL SQUAMOUS PAPILOMAS WITH FOCAL DERMAL HYPOPLASIA. E.A. Pasman, T. Heifert, C.M. Nylund, Pediatrics, Walter Reed National Military Medical Center, Bethesda, Maryland, UNITED STATES. Heifert, C.M. Nylund, Uniformed Services University, Bethesda, Maryland, UNITED STATES.

HA is an 8 year old female with FDH who presented to the pediatric gastroenterology clinic for failure to thrive, refusing gastrostomy tube feeds because of complaints of feeling fullness in her epigastrium, and poor oral intake. She previously had a G-tube placed at 3 weeks of age due to cleft lip and palate. She describes as a sensation of food getting stuck in her throat. She has had no respiratory problems. She has had a normal upper gastrointestinal contrast study 2 years previously. On exam she was very thin and emaciated with asymmetric and dysmorphic facial features and thin hair. She had multiple scars on her face consistent with her cleft lip repair. On her body she has multiple striae of hypoplastic skin. She had a II/VI systolic murmur. Her lungs were clear. She had normoactive bowel sounds; her abdomen was soft, non-tender with no organomegaly and a low profile balloon gastrostomy tube in place. Her evaluation included an esophagram which showed filling defects in the distal esophagus with delayed clearance of contrast from the esophagus. A nuclear medicine esophageal transit study which demonstrated 19.4 % residual at 30 minutes consistent with delayed esophageal clearance. On endoscopy she was noted to have multiple clusters of papillomas in her esophagus. Pathology of the specimen was consistent with a squamous papilloma and she had > 80 eosinophils per high power field (hpf). Subsequent EGD, after being on a proton pump inhibitor for over 6 weeks, demonstrated the squamous papillomas with up to 20 eosinophils/hpf. She was started on ingested fluticasone with no clinical improvement in her dysphagia. Further endoscopy is pending with planned argon plasma coagulation of the papillomas. Discussion: FDH or Goltz syndrome is a rare disorder of defective ectodermal and mesodermal tissue development. The disorder is inherited in an X-linked dominant manner with a female predominance of 9:1. Females are heterozygous or mosaic for mutation in PORCN, affected males are typically mosaic. The primary clinical manifestations of FDH occur due to dysplasia of the connective tissue of the skin and skeletal tissue. The dermal connective tissue is attenuated with thin appearing collagen fibers leading to hypoplastic and atrophic areas of skin. Cleft lip can be present leading to feeding difficulty. Mucocutaneous squamous papillomas have been reported on the mouth, nose, larynx, anus, and genitals. There have been reports of multiple esophageal squamous papillomas in individuals over the age of 30 but never in a child. These patients were described as having chronic dysphagia as did our patient. Our patient

had a previously normal esophagram suggesting she developed the esophageal papillomas between the ages of 6 and 8 years old possibly in response to EoE. In summary we report a case of dysphagia, esophageal dysmotility, EoE and the presence of distal esophageal papillomas in a child with FDH.

GERD/MOTILITY/FUNCTIONAL DISORDERS

266 GASTROINTESTINAL DYSMOTILITY IN AN ADOLESCENT WITH AUTOIMMUNE AUTONOMIC GANGLIONOPATHY. J. Khlevner, Pediatric Gastroenterology, Hepatology, and Nutrition, New York Presbyterian, Morgan Stanley Children's Hospital- Columbia University Medical Center, New York, New York, UNITED STATES. Bailey, Pediatrics, New York Presbyterian, Morgan Stanley Children's Hospital- Columbia University Medical Center, New York, New York, UNITED STATES.

BACKGROUND: Autoimmune autonomic ganglionopathy (AAG) is a disorder due to the development of antibodies against ganglionic nicotinic acetylcholine receptors (AChR) affecting the sympathetic, parasympathetic and enteric nervous systems. AAG is often subacute and is frequently preceded by a viral illness. The condition is linked to many features of autonomic impairment including gastrointestinal dysmotility. The mean age of onset, 61 years, makes the diagnosis and treatment of AAG in children rare and not well described. We present a case of AAG in an adolescent successfully treated with intravenous immunoglobulin (IVIG) and a promotility agent. CASE REPORT: A 15 year old previously healthy male, presented with a five week history of persistent emesis, dehydration, constipation, decreased urine output, significant weight loss, and progressive fatigue. Prior to presentation to our institution, the patient had an extensive diagnostic evaluation including laboratory studies, an upper gastrointestinal series, upper endoscopy, and abdominal ultrasound; all nonrevealing. A gastric emptying scan (GES) showed gastroparesis (half emptying time 246min). He was started on azithromycin with no improvement. The patient was admitted to our institution for further management. Initial vitals were significant for bradycardia (27bpm), hypothermia (33.3 C°), body mass index (15kg/m²). On physical examination he appeared dehydrated, malnourished, and fatigued. He was hydrated, started on nasoduodenal (ND) tube feeds, metoclopramide and cyproheptadine. Abnormal thyroid tests (suggestive of sick euthyroid), bradycardia and an elevated antinuclear antibody prompted consultation with endocrinology, cardiology, rheumatology, and psychiatry; extensive work up was nonrevealing. An eating disorder was ruled out. Head MRI, chest and abdominal radiographs were unremarkable. Repeat GES on promotility agent showed markedly prolonged gastric emptying. Tilt table test showed subtle abnormalities. Paraneoplastic auto- antibody panel resulted in elevated (AChR) antibody (0.17nmol/L) prompting a diagnosis of AAG. The patient was started on IVIG in addition to a promotility agent given lack of clinical improvement. Initial response was gradual allowing removal of ND tube but patient was unable to sustain adequate caloric intake. Patient refused reinsertion of a feeding tube, requiring total parental nutrition (TPN) to optimize calories and help with weight gain. IVIG was discontinued secondary to elevation of liver function tests (this resolved). One month later, off TPN, IVIG and a promotility agent; he maintained weight, regained strength, and tolerated diet without difficulties. Repeat AChR antibody trended down, GES normalized. CONCLUSIONS: AAG is an antibody mediated disease that results in multi-organ involvement including gastrointestinal dysmotility and dysautonomia that may mimic endocrinologic, rheumatologic, paraneoplastic and eating disorders. The diagnosis of AAG with gastrointestinal manifestations is often very difficult to make and requires gastroenterologist to consider a multidisciplinary approach. Treating with IVIG can be considered if symptom guided therapy is not effective.

267 SURGICAL MANAGEMENT OF MEGADUODENUM IN EHLER-DANLOS SYNDROME. N. HIJAZ, J.T. COCJIN, C.A. Friesen, Division of Gastroenterology, CHILDREN MERCY HOSPITAL, Kansas City, Missouri, UNITED STATES.

Ehlers-Danlos syndrome (EDS) an inherited disorder of connective tissue that is distinguished by the triad of skin hyperextensibility, articular hypermobility, and tissue fragility. It has been associated with small intestinal manifestation such as duodenal diverticulum, hemorrhage, perforations, and rare reported cases of megaduodenum associated with malabsorption. In addition, abnormal myogenic activity of the colon has been reported with a suggested link to colonic perforations but dysmotility of the small intestine has not been reported in EDS. Surgery is usually deferred when possible in EDS to help minimize the rate of surgical complications. We report a case of EDS in association with a functional duodenal obstruction with documented abnormal motility in this segment with successful surgical treatment. The patient is a 14 year old female diagnosed clinically with EDS

with joint hypermobility, frequent subluxation, mildly dilated aortic root and easy bruisability. She presented with symptoms consistent with functional dyspepsia including early satiety, postprandial bloating, and epigastric pain. She was treated conservatively with medication and biofeedback-assisted relaxation training. Her initial work up including routine laboratory tests, upper endoscopy, capsule endoscopy and abdominal X-ray series were unremarkable. Her symptoms got progressively worse with vomiting, positional abdominal pain, poor oral intake and weight loss. The upper GI series and computed tomography angiography showed megadoudenum with partial obstruction and evidence of superior mesenteric artery (SMA) like syndrome. With changes in position, contrast would pass and it was associated with a decrease in pain. She underwent surgical repair for SMA syndrome consisting of release of ligament of trietz and doudenopexy, and it was found to have a jejunal infarction which was resected. Despite the multiple corrective surgeries; they were not successful in relieving the obstruction or symptoms. She did not tolerate transpyloric drip feeds and was TPN-dependent for several months. Repeated evaluations including Upper GI series and magnetic resonance imaging showed focal narrowing involving the second and third portion the duodenum with duodenal dilatation and delayed transit of contrast material. An antroduodenal motility (ADM) study demonstrated a complete absence of motility of her duodenum. She underwent resection of a 7 cm length of her duodenum as mapped by the motility study. Pathology revealed hemorrhagic changes but no overt inflammation or abnormalities of the vasculature or neuromuscular system. Following surgery, she has had complete resolution of her symptoms and resumed a regular diet. Here in we report a unique case of EDS presenting with a functional duodenal obstruction with duodenal dysmotility confirmed by ADM. This is the third reported case with megadoudenum, in this case, surgery was successfully performed without any complications. This case illustrates the utility of ADM in the management of functional obstructions.

268 LONG-TERM GASTROINTESTINAL COMPLICATIONS IN CONJOINED TWINS. A. Alper, M.G. Patel, D.S. Pashankar, U.P. Phatak, Yale University, New Haven, Connecticut, UNITED STATES.

Introduction: Conjoined twins can be some of the most challenging patients encountered by pediatric gastroenterologists. The literature has mostly focused on issues related to surgical separation and cardiovascular complications. Data regarding long-term gastrointestinal management is lacking. Here we present a rare case of 14-year-old parapagus conjoined twins with both mutual and individual gastrointestinal symptoms. Case report: Two parapagus conjoined twins were born to a G1P1 29-year old mother. The twins are conjoined from the chest down. They have two heads, two brains, four arms, two sets of lungs and two functional hearts. They have two esophagi, a single liver, single stomach, single spleen, single small bowel, single colon, double rectum and megasigmoid, a single pelvis and two lower extremities. Although initially considered, surgical separation in this clinical setting carried extremely high mortality rates and was disregarded. The twins have had long-standing gastrointestinal problems: Constipation and fecal incontinence: Since infancy the twins had constipation, requiring aggressive therapy including rectal stimulation and oral laxatives. Interestingly only twin A suffers from lower abdominal pain and cramps, which are relieved by bowel movements. Twin B does not experience similar pain. Further studies of the twins demonstrated that they had a huge megacolon and a narrow anus. The refractory constipation required surgical intervention at the age of 4 years, including sigmoid resection, anoplasty and resection of the duplex rectum. Left colon was anastomosed to a single rectum. The surgery did not lead to an immediate improvement of their symptoms. However, with the aid of aggressive medical therapy, diet modifications and scheduled toilet sitting, the twins have now daily bowel movements, mostly non-bloody, and significantly improved incontinence. GERD: The twins share the same stomach but have two esophagi. In the last 4 years twin B suffered from occasional epigastric pain and reflux symptoms, while twin A remained completely asymptomatic. Those symptoms significantly improved with daily use of a proton pump inhibitor, taken only by the symptomatic twin. Discussion: We present here a case of conjoined twins with refractory constipation, fecal incontinence and GERD. Although they share most of their gastrointestinal tract, each twin experiences different symptoms, possibly due to individual somatosensory pathways. While one twin has reflux symptoms, the other twin has lower abdominal pain associated with constipation. Individual treatment may be helpful in controlling those symptoms. Refractory constipation can be a major concern in unseparated conjoined twins, which requires both medical and surgical intervention and diet modifications. Conclusion: We discuss a unique case of long-term gastrointestinal complications in unseparated conjoined twins. Treatment of those complications may be challenging and requires a thorough acquaintance of the anatomical anomaly and a multidisciplinary approach.

This case confirms that gastrointestinal symptoms are not necessarily shared by both twins and may require individual treatments.

269 A NOVEL DUAL DIAGNOSIS: CYSTIC FIBROSIS AND ACHONDROPLASIA. P. Do, Pediatric Pulmonology, Miller Children's Hospital , Long Beach, California, UNITED STATESM.C. Huckaby, Pediatric Residency Program, University of California, Irvine, Children's Hospital of Orange County, Santa Ana, California, UNITED STATESM.R. Allen-Sharpely, School of Medicine, University of California, Irvine, Irvine, California, UNITED STATESH. Abrams, S. Sela, R.K. Mathis, Pediatric Gastroenterology, Miller Children's Hospital, Long Beach, California, UNITED STATESD. Paralusz, Nutrition, Miller Children's Hospital, Long Beach, California, UNITED STATES. Case: A 5 month old female with a history of cystic fibrosis diagnosed on newborn screen, obstructive sleep apnea and suspected achondroplasia presented to the emergency department from genetics clinic for failure to thrive, respiratory symptoms, and lack of consistent follow up. As part of her initial emergency room evaluation, biochemical analyses revealed sodium of 126 mmol/L, potassium of 2.3 mmol/L, chloride of 66 mmol/L, bicarbonate of 43 mmol/L, blood urea nitrogen of 12 mg/L, creatinine of 0.21 mg/dL. She was then admitted for further evaluation and electrolyte stabilization. Additional laboratory studies demonstrated an elevated lipase of 295 U/L, decreased fecal pancreatic elastase level of 35 mcg/g stool, and elevated folate level of >24 ng/mL. The patient was started on pancreatic enzyme replacement therapy. Given her failure to thrive, loose stools, and her elevated folate level, there was also concern for small intestinal bacterial overgrowth (SIBO). The patient was treated with Bactrim and Flagyl for presumed SIBO, and she was scheduled for an EGD and impedance study, to evaluate for reflux in the context of her respiratory symptoms. Testing was deferred due to complications from intubation prior to the procedure which required placement in the intensive care unit. Discussion: Presently, are no reports in the literature of patients with a dual diagnosis of achondroplasia and cystic fibrosis. Our patient's initial presentation of metabolic alkalosis with hyponatremia, although unusual, has been reported in the literature and is most common in infants with cystic fibrosis who are homozygotes for the delta 508 mutation, which is the genetic profile of our patient. Yet some of the associated features of achondroplasia such as midface hypoplasia, upper airway obstruction, restrictive lung disease, increased obesity risk, and increased risk of gastroesophageal reflux may complicate management of both the pulmonary and gastrointestinal manifestations of cystic fibrosis in our patient. Furthermore, fibroblast growth factor receptor 3 (FGFR3), which has exaggerated function in achondroplasia, is expressed in both the gastrointestinal tract and the pancreas; excess FGFR3 signaling in these tissues may have consequences affecting our patient's disease course, however, too little is known to draw any firm conclusions at this time. In otherwise healthy children, SIBO appears to be a frequent cause of chronic digestive symptoms in children less than 2 years old, possibly due to immaturity of factors such as gastric acid secretion, immune system, and bowel motility. CF patients frequently suffer from SIBO, thought in these patients to be caused by the presence of viscous dehydrated GI contents that are predisposed to bacterial growth. In addition to GI dysfunction or short bowel, exocrine pancreatic insufficiency, as seen in our patient is a known predisposing factor to SIBO, but the prevalence of SIBO in infancy has not been reported. Given the lack of reports in the literature, the prognosis for our patient is unclear at this time. Medical management decisions and treatment outcomes of her disease will be valuable to future patients with similar genetic diagnoses.

270 INTRACTABLE VOMITING AND A DIAGNOSIS OF GASTROPARESIS: A FIRST PRESENTATION OF NEUROMYELITIS OPTICA. J.A. Bitong, R. Nagpal, Pediatrics, Advocate Children's Hospital-Oak Lawn, Chicago, Illinois, UNITED STATESN. Mathenia, Neurology, University of Illinois Chicago, Chicago, Illinois, UNITED STATES. BACKGROUND Intractable vomiting is a common clinical dilemma for pediatric gastroenterologists. The gastrointestinal causes of intractable vomiting are numerous and include functional, obstructive, and anatomic etiologies. The neurologic causes may include increased intracranial pressure and brainstem involvement. However, in the face of a positive gastric emptying study in an otherwise negative work-up of gastrointestinal and neurologic pathology, there is a question of whether gastroparesis can be the only answer. The following case report describes intractable vomiting and hiccups as the only presenting symptoms of neuromyelitis optica (NMO). NMO is a rare demyelinating disease affecting the white matter of the CNS. It typically affects the optic nerves, leading to vision loss, and the spinal cord, resulting in numerous neurologic symptoms. Rarely, patients may present with intractable vomiting and hiccups, which may not be immediately identifiable as an underlying neurologic condition. CASE PRESENTATION A 17-year-old previously healthy girl presented with intractable vomiting and hiccups. Physical exam was unremarkable. Anatomic abnormality was excluded with an upper GI, and

distal intestinal pathology ruled out with a CT of the abdomen. Mucosal disease was addressed with an EGD, with normal biopsies. MRI of the brain to rule out intracranial pathology was normal. Short of a diagnosis, the possibility of cyclic vomiting was considered, and the patient was started on amitriptyline, with improvement in symptoms. The patient was discharged home, only to return in weeks with similar symptoms, with emesis 10-20 times a day and worsening hiccups, as well as a new complaint of headache. She was readmitted for evaluation. Clinical exam again showed no abnormalities, with no neurologic deficits. Repeat MRI of the brain was again normal. Intestinal motility disorder was considered, and a gastric emptying study was done, significant for delay. She was started on erythromycin, discharged with the diagnoses of gastroparesis and anxiety disorder, and was referred for further evaluation for intestinal motility disorder at a tertiary facility. The patient presented a month later, reporting 2 weeks of diplopia, gait instability, severe headaches, and a 3-day history of right facial droop and horizontal nystagmus. Repeat MRI suggested acute inflammatory demyelinating disease. A lumbar puncture was performed, negative for any definitive diagnostic information. A positive serum NMO quickly followed, consistent with the patient's clinical picture and a diagnosis of neuromyelitis optica. **DISCUSSION/CONCLUSION** Although well described in neurology literature as possible initial presenting symptoms of NMO, intractable vomiting and hiccups, in the face of a negative neurologic exam, suggest gastrointestinal pathology. However, 12% of patients with NMO initially present with isolated intractable vomiting. Thus, following a negative gastrointestinal work-up, demyelinating disease should be considered as part of the differential diagnosis. Early treatment of neuromyelitis optica may improve gastric emptying while also addressing the CNS effects of the disease.

271 **USE OF ARGON PLASMA COAGULATION FOR THE TREATMENT OF A GASTRIC INLET PATCH IN A PEDIATRIC PATIENT WITH DYSPHAGIA.** O. Castro, B. Barth, L. Chan, UT Southwestern, Arlington, Texas, UNITED STATES. Castro, B. Barth, L. Chan, Children's Medical Center, Dallas, Texas, UNITED STATES.

A gastric inlet patch is an area of ectopic gastric mucosa in the proximal esophagus which has been associated with dysphagia, reflux, ulceration, bleeding, strictures and webs. Gastric inlet patches are generally considered to be benign but can contribute to upper esophageal symptoms in some patients. Argon plasma coagulation, commonly used to ablate bleeding lesions during endoscopy in the pediatric population, is a safe and effective interventional therapy for children with significant symptoms associated with a gastric inlet patch. A 9 year old female presented with a 7 year history of dysphagia and odynophagia with solids. She restricted herself to a soft foods and liquids diet. Recently, she had difficulty gaining weight and was taking dietary supplements to improve caloric intake. She reported the sensation of food getting stuck in her throat. She also had a history of infrequent regurgitation but denied any chest pain, pulmonary symptoms, hematemesis, or change in bowel habits. Her physical exam was normal with the exception of her weight, which fell at the second percentile for her age. She was initially started on esomeprazole 20 mg daily but was not compliant with therapy. An esophagram demonstrated minimal pooling of contrast at the level of T1 concerning for a possible diverticulum. Esophagogastroduodenoscopy was performed and revealed a gastric inlet patch in the proximal esophagus (15 cm from incisors). Biopsies of the inlet patch showed gastric heterotopia with acute and chronic inflammation. Biopsies of the esophagus, stomach, and duodenum showed normal histology. No esophageal diverticula were seen. She was restarted on esomeprazole 20 mg daily with only mild relief of symptoms after 7 months of therapy despite gaining weight. She also underwent a video fluoroscopic swallow study which showed mild oral phase dysphagia and mild behavior feeding disorder. She was enrolled for oral motor and feeding therapy which did not resolve her dysphagia. Three months later, endoscopy was repeated for the purpose of argon plasma coagulation ablation of the gastric inlet patch using setting of 0.5 liters/minutes and 20 watts. The entire lesion was ablated successfully. On subsequent follow up, one week following ablation, the caregiver reported improved symptoms. Ablation of gastric inlet patches with argon plasma coagulation should be considered in pediatric patients with significant esophageal symptoms that fail to improve with medical management.

272 **THE FIRST KOREAN CHILD CASE REPORT OF DIPHYLLOBOTHRIASIS NIHONKAIENSE CONFIRMED BY MITOCHONDRIAL COX1 GENE SEQUENCING.** E. Lee, Y. Go, Pediatrics, Myongji hospital, Goyang-Si, Gyeonggi-Do, KOREA, REPUBLIC OF. Cho, S. Choi, J. Chai, parasitology and tropical medicine, Seoul National University College of Medicine, Seoul, KOREA, REPUBLIC OF.

Diphyllobothriasis is an infection by intestinal tapeworms that use fresh or brackish water fishes as their second intermediate host. In the past, *D. latum*, based on morphological characteristics, had been considered the causative pathogen in most diphyllobothriasis cases in Korea. However, recently, it has become clear that most of

cases that had been reported as *D. latum* infection were, in fact, caused by *D. nihonkaiense* infection in a study using gene analysis of expelled gravid proglottids. *D. latum* and *D. nihonkaiense* are very similar to each other and only genetic identification method can differentiate clearly between the two species. We report the case of a 7-year-old boy who ate slices of three kinds of raw fish 16 days before he visited our outpatient clinic and expelled a tapeworm segment about 1.2 m long. He complained of abdominal pain and watery diarrhea. Nucleotide sequencing of mitochondrial DNA cytochrome c oxidase subunit 1 gene (*cox1*) of discharged proglottids revealed 99.9% (687/688 bp) similarity with *D. nihonkaiense* and only 93.2% (641/688 bp) similarity with *D. latum*. He was treated with a single dose of praziquantel and passed the rest of tapeworm strobila and complained of no more symptoms. It is the first reported case of pediatric diphyllbothriasis in Korea, confirmed using a genetic analysis of the pathogen. The use of morphological features alone to identify these two *Diphyllbothrium* species is insufficient and can lead to misidentification of the tapeworms. A genetic analysis is necessary for an exact diagnosis and preventive control methods through the acquisition of relevant epidemiological information. Nucleotide identity between our specimen and known *Diphyllbothrium* species in GenBank for the mitochondrial cytochrome c oxidase 1 (*cox1*) gene. The unit of each value is base pairs (%).

273 ABDOMINAL COMPARTMENT SYNDROME SECONDARY TO NOROVIRUS INFECTION IN A PEDIATRIC PATIENT WITH PRIOR NISSEN FUNDOPLICATION. S. Lloyd, M. Liu, C. Gause, F. Seifarth, A. DeRoss, Cleveland Clinic Foundation, Cleveland, Ohio, UNITED STATES.

Norovirus is one of the leading causes of viral gastroenteritis, typically manifesting as a self-limiting infection. To date, there have been no reports of abdominal compartment syndrome (ACS) due to Norovirus infection. We present the first such case. A 7 year-old female with a prior Nissen fundoplication was referred to our institution for ACS and fulminant sepsis secondary to an acute Norovirus infection. The patient was successfully treated with emergent decompressive laparotomy, delayed abdominal closure and supportive therapy. The post-operative course was notable for acute hepatitis and pancreatitis, both of which resolved without further intervention. After a prolonged hospital stay, she was discharged home in good condition. This patient developed a rare but potentially fatal illness, with her previous Nissen wrap likely contributing to intra-abdominal hypertension and ACS. Ultimately, clinical vigilance and early surgical intervention led to a favorable outcome.

274 THE USE OF ACID REDUCING MEDICATIONS IN PEDIATRIC PATIENTS POST NISSEN FUNDOPLICATION.. H. Conrad, Div. of Pediatric Gastroenterology, Helen Devos Children's Hospital, Grand Rapids, Michigan, UNITED STATES. A. George, Pediatric Residency, Helen Devos Children's Hospital, Grand Rapids, Michigan, UNITED STATES.

Background and objective - Pediatric gastroesophageal reflux disease is treated with medical management including acid reducing medications (ARM) and if this fails patients undergo surgical therapy, usually Nissen fundoplication. Surgical therapy is often effective. However, patient's without complete response to surgery or in whom fundoplication has broken down may require the addition of ARM. We studied the use of ARM post Nissen fundoplication at our institution to determine if the medications were used appropriately. Methods - Patients under 18 who had undergone Nissen fundoplication were identified from electronic medical records of clinic visits to pediatric gastroenterology, pediatric pulmonology and pediatric neurodevelopmental outpatient clinics from January 1, 2012 through June 30, 2013. All charts were analyzed for ARM use post Nissen. Additionally, randomly selected charts were further analyzed for diagnoses possibly affecting medication use as well as workup post Nissen before medication was restarted. Results - Our study had 566 patients who underwent Nissen fundoplication, and 247 (43.6%) were still on ARMs after surgery. In our patient subset, 52.6% of patients with neurologic impairments (NI) and 63.6% of patients without NI never had their ARM stopped post-Nissen. The relative risk of being on an ARM after having a Nissen was 1.6 ($p=0.02$) in patients with NI. 68.4% and 21.1% of patients with NI had an upper GI and EGD post-Nissen respectively, whereas 45.5% and 18.2% of patients with no NI had the respective testing. Discussion- This QI project suggests that protocols need to be developed for ARM use post-Nissen, especially for patients with NI.

275 SMALL INTESTINAL BACTERIAL OVERGROWTH (SIBO) WITH STREPTOCOCCUS SALIVARIUS AND PARASANGUINIS. T. Hadley, N.H. Patel, T. Mehta, J. Conrad, D.I. Mehta, K. Horvath, Pediatric Gastroenterology, Center for Digestive Health and Nutrition, Arnold Palmer Hospital for Children, Orlando, Florida, UNITED STATES.

A 19 year old female with a history of ADHD, sensory integration disorder, and audio/visual processing deficit had a long standing history of symptoms suggesting peptic acid disease. Three years ago she underwent an upper endoscopy and 48 hour Bravo esophageal pH study that were normal despite her complains of lower chest pain and heartburn like symptoms which are consistent with gastroesophageal reflux disease. She was placed on a proton pump inhibitor, dexlansoprazole, which provided complete resolution of her symptoms. She presented three years after the initial visit with new onset nausea, vomiting, anorexia and weight loss (8 lb). She continued to take dexlansoprazole but with no alleviation of her symptoms. She denied of having any worsening of heartburn like symptom. She had undergone an evaluation by an allergist and was noted to have intolerance to egg, wheat, chicken and pork. Consequently, she had eliminated them from her diet. Her dexlansoprazole dosage was increased to 60 mg BID with no significant improvement. Due to the persistence of her symptoms, she underwent an EGD, which showed significant bilious residuum in her stomach. Her histologies and disaccharidase activities were normal. The duodenal culture obtained through small intestinal brushing revealed 150,000 CFU/ml with growth of *Streptococcus salivarius* and *parasanguinis*. She received a 14 day course of Amoxicillin for treatment of small intestinal bacterial overgrowth (SIBO). Following the completion of her antibiotic therapy her symptoms completely resolved. PPI medication which she had been on for many years was discontinued; she gained weight and she remained asymptomatic. Conclusion: This case report shows that symptoms may be consistent with chronic peptic acid disease and may respond temporarily to a PPI therapy. However, if the symptoms persist or change, an underlying diagnosis of Small Intestinal Bacterial Overgrowth should be considered.

293 CHOLESTASIS AND DUCTOPENIA CAUSED BY SERTRALINE IN ADOLESCENT MALE. M.A. Conrad, H.C. Lin, Division of GI, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES. J. Cui, University of Pennsylvania, Philadelphia, Pennsylvania, UNITED STATES.

Vanishing bile duct syndrome (VBDS) is an acquired progressive destruction and disappearance of interlobular bile ducts causing chronic cholestasis. Medications are a common cause of VBDS. We present a unique case of bile duct paucity in an otherwise healthy teenager with resolution of symptoms following cessation of sertraline. A 15 year old boy with major depressive disorder on sertraline presented with 1 month of jaundice with associated nausea and anorexia. His stools had become progressively pale over the month prior. Labs were significant for total bilirubin of 6 mg/dL, and hepatomegaly was noted and confirmed by ultrasound. Due to worsening nausea that coincided with the rising bilirubin, he had a liver biopsy 2 weeks after initial presentation. Liver biopsy revealed preserved lobular architecture with 14-16 portal tracts notable for rare bile duct profiles and minimal lymphocytic infiltration as well as marked hepatocellular and canalicular cholestasis. He was started on ursodiol with mild improvement in his bilirubin. His cholestasis and fatigue progressed over the next month, and his total bilirubin peaked at 33.7 mg/dL with direct bilirubin 29.2 mg/dL. Magnetic resonance cholangiopancreatogram was normal. He was evaluated for other causes of ductopenia such as Alagille Syndrome, including a normal echocardiogram, no butterfly vertebrae on x-ray, and an unremarkable ophthalmologic exam. JaundiceChip Resequencing Array which tests for genes representing the most common heritable causes of cholestatic liver disease in children revealed a polymorphism in JAG1 with unknown clinical significance. Urine bile acids were not consistent with any bile acid synthesis defect. Anti-smooth muscle antibody was positive with 1:80 titer, but ANA, ANCA, anti-f-actin, anti-liver/kidney microsome antibodies were all negative. After no etiology for ductopenia was established, he was diagnosed with Vanishing Bile Duct Syndrome, and sertraline was discontinued. Within 4 weeks, his bilirubin had improved, and bilirubin normalized by 4 months after discontinuation. He was subsequently started on fluoxetine to treat depression and continued to have normal hepatic function tests. VBDS is characterized by acquired ductopenia due to a variety of insults to the liver, including infection, drugs, toxins, oncologic and immunologic processes. Selective serotonin reuptake inhibitors including sertraline have not previously been identified in association with vanishing bile duct syndrome in the adult or pediatric literature.

294 ADENOVIRUS HEPATIC ABSCESS: A NOVEL CAUSE OF FEVER OF UNKNOWN ORIGIN IN A PEDIATRIC LIVER TRANSPLANT PATIENT. K. Haas, R.O. Castillo, Pediatric Gastroenterology, Hepatology, and Nutrition, Lucile Packard Children's Hospital at Stanford University, Palo Alto, California, UNITED STATES.

Introduction: Adenovirus is a common cause of febrile illness in the general pediatric population. It is a particularly dangerous infection in immunosuppressed post-transplant patients who are at risk for severe disseminated disease. Adenovirus has not been previously described in the literature as a cause for hepatic abscess. Hepatic abscesses are rare in developed nations, and are most often polymicrobial bacterial infections. Here we present a

pediatric liver transplant recipient who presented with two weeks of fever and was found to have a hepatic abscess with adenovirus positive immunohistochemical staining. Case Report: A 10-year-old girl with a history of biliary atresia who underwent liver transplant in 2012 presented to our institution one year after transplant with two weeks of persistent fever, fatigue, abdominal pain, and anorexia. She had no emesis, diarrhea, cough, rashes, or rhinorrhea. She was fully vaccinated pre-transplant, and had no animal exposures or travel outside of the United States. Her post-liver transplant course was complicated by hepatic artery stenosis and an episode of acute rejection one month post-transplant. At the time of admission she was on tacrolimus and sirolimus immunosuppression with supratherapeutic levels. She was noted to have mild leukopenia, elevated CRP, and transaminitis. Initial infectious evaluation included EBV and CMV PCR, stool bacterial culture, *Clostridium difficile* PCR, and ova and parasites that were negative. Serum adenovirus PCR was significantly elevated at 21,700 viral copies. An abdominal MRI revealed a liver lesion 3.7 x 2.5 x 2.4 cm concerning for pyogenic abscess. Interventional radiology performed CT guided drainage of the fluid collection as well as liver biopsy. Liver tissue surrounding the aspirated fluid collection showed hepatic necrosis with positive immunohistochemical staining for adenovirus. The liver biopsy showed portal inflammation with granulomas that were loosely organized, typical of adenoviral infection. Urine adenovirus PCR was positive, stool adenovirus antigen was negative, and respiratory viral PCR testing was negative. Further infectious work-up was performed given the granulomas on liver biopsy and was negative including Hepatitis A, B, and C serology, Bartonella titers, coccidioidomycosis titers, Brucella antibody, Coxiella titers, Histoplasma antigen, Toxoplasmosis antibody, HIV western blot, RPR, and Quantiferon. Bacterial and fungal cultures from serum, urine, and liver fluid aspirate were negative. Liver tissue fungal and acid fast bacilli stains were negative. Our patient remained afebrile with downtrending adenovirus PCR levels after liver abscess aspiration and reduction in immunosuppression levels. Repeat abdominal imaging showed near resolution of the hepatic fluid collection. Conclusion: This case report describes a novel finding of adenovirus hepatic abscess in a pediatric liver transplant patient who presented with fever of unknown origin. Though adenoviral disease and hepatic abscesses are well described in the liver transplant recipient population, we believe this is the first reported case of hepatic abscess due to adenovirus.

295 CONGENITAL ABSENCE OF DUCTUS VENOSUS IN A 2 YEAR OLD: LATE SPONTANEOUS CLOSURE OF A LARGE PRE-HEPATIC UMBILICAL VEIN TO RIGHT ATRIAL SHUNT WITH RESIDUAL INTRAHEPATIC PORTOSYSTEMIC SHUNTS. S. Syed, S. Karpen, M. Vos, R. Romero, Department of Pediatrics, Division of Pediatric Gastroenterology, Hepatology and Nutrition, Emory University School of Medicine, Atlanta, Georgia, UNITED STATES. A. Alazraki, Department of Radiology, Division of Pediatric Radiology and Department of Pediatrics, Emory University School of Medicine, Atlanta, Georgia, UNITED STATES. J. Petit, Department of Pediatrics, Division of Pediatric Cardiology, Emory University School of Medicine, Atlanta, Georgia, UNITED STATES.

The ductus venosus is a fetal vascular channel shunting blood from the left umbilical vein (UV) to the inferior vena cava. This channel bypasses the developing liver and along with the foramen ovale and ductus arteriosus, preferentially shunts oxygenated blood to the fetal brain. Absent Ductus Venosus (ADV) results in variant UV circulations. Extrahepatic pathways can bypass the liver with drainage into the internal iliac vein, the inferior vena cava, renal vein, or directly to the right atrium (RA). Complete or partial absence of the portal vein (PV) can be identified in those ADV associated with extrahepatic shunts. ADV in the absence of other abnormalities can be associated with prolonged survival. We report a 2-year-old female presenting with hypoalbuminemia and edema with a one month history of diarrhea and intermittent fevers. Her past medical history was notable for an atrial septal defect (ASD) and mild pulmonary valve stenosis. Physical examination showed facial and extremity edema but no hepatosplenomegaly. Laboratory investigations revealed mild elevation of her transaminases, hypoalbuminemia and minimal INR elevation (Table 1). Autoimmune, infectious and metabolic work-up was negative. ECHO showed a structurally normal heart. MRI showed a large portosystemic shunt (PSS) traversing from the superior mesenteric vein anterior to the liver and communicating directly with the RA. Temporary balloon occlusion of the shunt at the time of angiography confirmed the presence of hypoplastic intrahepatic PV branches and small, direct intrahepatic PV to hepatic vein communications. Permanent occlusion was deferred at that time due to concerns regarding inducing a sudden and marked increase in PV pressure. She was discharged after resolution of her edema with albumin and furosemide infusions. Six months after her diagnosis, the patient underwent angiography for the first of a planned series of staged occlusions. However, the previous porto-atrial shunt was significantly atretic on the repeat angiogram. There was persistence of small intrahepatic PV to hepatic vein communications in the right lobe. No cardiovascular abnormalities nor encephalopathy has been detected

and the patient continues to be well with no hypoalbuminemia or ascites fifteen months after her initial presentation. This case represents a combination of previously describe features of ADV in one patient: a prehepatic residual umbilical vein to right atrial extrahepatic shunt, and hypoplasia of the main PV with residual intra-hepatic PV to hepatic vein shunts.

Laboratory investigations at presentation and on follow-up

296 PERCUTANEOUS TRANSHEPATIC CHOLANGIOPLASTY: A THERAPEUTIC OPTION IN THE TREATMENT OF INTRAHEPATIC BILIARY STRICTURES IN PEDIATRIC PATIENTS AFTER HEPATOPORTOENTEROSTOMY. M.N. Weidner, K. Schwarz, Pediatric Gastroenterology and Nutrition, Johns Hopkins Hospital, Baltimore, Maryland, UNITED STATES. Mitchell, Interventional Radiology, Johns Hopkins Hospital, Baltimore, Maryland, UNITED STATES. Although hepatoportenterostomy (HP) can effectively restore bile flow in children with inadequate biliary drainage, biliary strictures can occur following successful surgical intervention often leading to biliary cirrhosis and liver transplantation. Non-surgical therapeutic options such as endoscopic dilation are limited after HP due to access difficulties. Percutaneous transhepatic cholangiography with cholangioplasty (PTC-C) can be a valuable tool but successful application post HP has not been reported. Herein, we present three patients status post HP who presented clinically with a history of recurrent ascending cholangitis and intrahepatic biliary stricture formation between 17 to 21 years of age who underwent successful PTC-C. Case 1 is a 23 year old female with biliary atresia who presented at 18 years of age with acute cholangitis. She underwent her first PTC-C at age 21. PTC revealed an irregular intrahepatic ductal system with multiple irregular branches coursing down toward the porta hepatis. No right, left or common hepatic ductal system could be identified. She required 3 PTC-C procedures in total within 1 week to achieve successful drainage of her biliary system. She has not had any further episodes of cholangitis in the 2 years post PTC-C. Case 2 is a 24 year old female with type IV choledochal cyst who presented at age 5 with cholestasis and underwent her first PTC-C at age 17. PTC revealed multiple chronic biliary strictures from the roux-en-y anastomosis. Over the last 8 years, she has had only 4 episodes of ascending cholangitis, but has continued to require a biliary drain on account of recurrent stricture formation. She has received about 50 PTC-C procedures in total. Case 3 is a 17 year old female with biliary atresia who presented for a third opinion after years of recurrent cholangitis. She underwent her first PTC-C at age 17. PTC revealed multiple irregular bile ducts and focal narrowing at the roux limb. She underwent 4 PTC-C procedures which included balloon dilation of the narrowed roux limb anastomosis. She had initial improvement in cholestasis but ultimately underwent liver transplantation due to subsequent episodes of cholangitis. In summary, all 3 patients underwent PTC-C with placement of right sided biliary drain. Although, the number of procedures required varied between patients, all 3 patients had initial improvement in cholestasis following PTC-C. Two patients had a significantly reduced frequency of cholangitis after PTC-C without adverse events. We demonstrate that PTC-C is a safe procedure that can be effective in this pediatric patient population. Moreover, PTC-C may be a therapeutic option that can decrease the overall need for liver transplantation, but further prospective studies are needed.

297 FATAL HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS ASSOCIATED WITH CHRONIC HEPATITIS B AND ACTIVE EPSTEIN BARR VIRUS. M. Middelhof, V. Sood, M. Brown, P. Mohanty, Pediatric Gastroenterology and Nutrition, University of rochester, Rochester, New York, UNITED STATES. We report a 16-year-old male patient with history of Chronic Hepatitis B infection, who presented with fever and hepatosplenomegaly. Laboratory findings were significant for pancytopenia (WBC 0.9 Thou/uL, Hemoglobin 7.7 g/dL, Platelets 14 Thou/uL), elevated transaminases (AST 72 U/L, ALT 95 U/L), coagulopathy (PT 15.8 secs, INR 1.5), hypofibrinogenemia (119 mg/dL), markedly elevated serum ferritin levels (70248 ng/mL) and lactate dehydrogenase (5270 U/L). His bone marrow biopsy showed hypercellular marrow with extensive lymphohistiocytic and plasmacytic infiltrates. Patient was diagnosed with hemophagocytic lymphohistiocytosis (HLH) secondary to Chronic Hepatitis B. Further work up did not reveal any evidence of lymphoma, malignancy or infections. During the course of his hospitalization he tested positive for Epstein Barr virus infection. He was initiated on dexamethasone and Etoposide according to the HLH-94 protocol and Tenofovir for Chronic Hepatitis B infection. Rituximab was added subsequently for active Epstein Barr virus infection. He initially responded well to the treatment however after five weeks of therapy, his clinical symptoms deteriorated and he eventually became refractory to the treatment. Subsequently our patient developed multi-organ dysfunction and died despite maximization of immunosuppressive and supportive treatment. HLH is often associated with infections, especially viral. Common viruses include Epstein-Barr virus, cytomegalovirus, parvovirus, herpes simplex virus, varicella-

zoster virus, measles virus, human herpes virus-8, H1N1 influenza virus, parechovirus, and HIV. To the best of our knowledge this is the first pediatric case report of HLH associated with Hepatitis B and Epstein Barr virus infection.

298 PROFOUND HYPOCALCEMIA SECONDARY TO VITAMIN D DEFICIENCY IN AN INFANT WITH BILIARY ATRESIA. S. Nasiri, B. Kaj, Pediatrics, Einstein Medical Center, Philadelphia, Pennsylvania, UNITED STATES. Pall, Gastroenterology, Hepatology, and Nutrition, St. Christopher's Hospital for Children, Philadelphia, Pennsylvania, UNITED STATES.

Introduction Cholestasis secondary to biliary atresia (BA) is a recognized cause of fat-soluble vitamin (FSV) deficiencies and as a result, infants with BA are routinely supplemented with vitamins A, D, E and K. With BA often presenting early in infancy, it is unusual for sequelae of FSV deficiencies to be present at initial diagnosis. We report a case of profound hypocalcemia with secondary electrocardiographic changes in an infant presenting with cholestasis. The Case A 10 week-old male infant with uncomplicated birth history was brought to his pediatrician with scleral icterus and found to have bilirubin levels of 11.55mg/dL (total) and 8.65mg/dL (direct). Upon referral to the hospital, laboratory investigations revealed profound hypocalcemia with a serum calcium of 6.6mg/dL (ref 9 – 11mg/dL) and ionized calcium 0.83mmol/l (ref 0.95 – 1.5mmol/l), hyperphosphatemia with serum phosphate of 9mg/dL (ref 4 – 6.5mg/dL) and prolonged QTc (495ms) on electrocardiogram (ref \leq 450ms). The infant was placed on continuous cardiac monitoring and promptly initiated on intravenous and oral calcium supplementation. FSV deficiencies were found on second day of admission, with 25-OH-vitamin D <4ng/mL (normal 30-100ng/mL) and vitamin K1 <0.13 ng/mL (normal 0.28-1.78 ng/ml). These deficiencies were corrected with oral ergocalciferol and subcutaneous vitamin K. Ultrasound revealed a “small atretic gall bladder with no visible extrahepatic biliary ducts”. Hepatobiliary iminodiacetic acid (HIDA) scan on sixth day of admission did not reveal excretion of tracer into intestine. Subsequent intraoperative cholangiogram confirmed the diagnosis of BA. Liver biopsy was compatible with BA and demonstrated diffuse bile duct proliferation, bridging fibrosis, cholestasis, periportal inflammation and bile plugging. The patient successfully underwent Kasai hepatopertoenterostomy. Both the endocrinology and cardiology teams were involved in the management of hypocalcemia in this infant. Several doses of IV calcium gluconate supplementation were required to normalize the infant’s serum calcium level and correct the prolonged QTc on ECG. At time of discharge, oral supplementation was continued with calcium carbonate (100mg QID), ADEK (0.5ml daily), and ergocalciferol (4000IU/day). Conclusion Management of FSV deficiencies can be challenging in an infant with cholestasis. The profound hypocalcemia secondary to vitamin D deficiency in our patient, with associated ECG changes, required prompt identification and supplementation. There was need to correct both the FSV deficiency itself and its ensuing downstream effects. Several studies highlight the importance of early detection and appropriate treatment of FSV deficiencies in cholestasis, and the utility of FSV supplementation is widely accepted. It is also important however, to be aggressive in the detection of complications of FSV deficiencies, particularly in patients with significant cholestasis and need for surgical intervention.

299 ACUTE LIVER FAILURE IN AN INFANT WITH MITOCHONDRIAL TRMU GENE MUTATIONS. H.L. Wang, Pathology & Lab Medicine, DGSOM at UCLA, Los Angeles, California, UNITED STATES. Kordy, S. Wadera, C.V. Strier, E.A. Marcus, Pediatrics, DGSOM at UCLA, Los Angeles, California, UNITED STATES. E.A. Marcus, VA Greater Los Angeles Health Care System, Los Angeles, California, UNITED STATES.

Background: Patients with primary mitochondrial hepatopathies generally have progression of disease and high mortality. However, infants who survive the acute liver failure (ALF) due to TRMU gene mutations typically have spontaneous remission without recurrence. Case Presentation: A two month old previously healthy male presents with one month of progressive feeding intolerance, nonbilious emesis, and subsequent poor growth. He developed jaundice, scleral icterus, and abdominal distention ten days before developing hematemesis, and had a recent history of diarrhea. On admission, he was noted to have severe metabolic acidosis with a bicarbonate level of three. He had moderate transaminitis, hyperbilirubinemia, and poor hepatic function including a significant coagulopathy, hypoglycemia, hypoalbuminemia, and factor V and VII deficiency. The patient also had hyperammonemia, hyperlactatemia, high alpha-fetoprotein, and developed ascites. Infectious workup was negative. Main liver histologic features include diffuse microvesicular, small and large droplet steatosis; focal ballooning of degeneration of hepatocytes; and cholestasis. Exome sequencing was diagnostic for homozygous missense mutations of the TRMU and coenzyme Q genes. Discussion: This patient's presentation is typical of the nineteen reported cases of hepatopathy due to homozygous TRMU gene mutation, with the exception of the CoQ

mutation and ascites. Infants who were previously healthy develop irritability, lethargy, poor feeding, and vomiting at 2-4 months of age. They develop hepatomegaly and jaundice. Their biochemical features include severe coagulopathy, hypoalbuminemia, metabolic acidosis, hyperammonemia, and high alpha-fetoprotein. The unusual feature of lactic acidemia directs attention to a possible mitochondrial disorder. TRMU gene encodes a thiouridylase necessary for the maturation of mitochondrial tRNA and affects the accuracy and efficiency of translation. Dietary cysteine might play an important role for normal TRMU activity within the first few months of life. Cysteine desulfurase transfers sulfur from cysteine to the TRMU gene. However, the availability of cysteine in the neonatal period is limited and the activity of cystathionase enzyme is low at birth, increasing slowly in the first 3-4 months of life. Another source of cysteine, metallothionein, drastically decreases in the neonatal period after initial high levels at birth. Reduced dietary intake, especially of cysteine, and intercurrent illness at 1-4 months of age further compromises TRMU activity in these vulnerable patients. For patients who survived the ALF, there has been no reported extrahepatic involvement and no recurrence on long-term follow-up, the oldest being 20 years old in one cohort. Liver function and clinical improvement typically occurs within 3-4 months. There are currently no available curative therapies for mitochondrial hepatopathies. Supportive care with correction of hypoglycemia, acidosis, and hyperammonemia is a mainstay in management of neonatal ALF. Supplementation with cysteine, coenzyme Q, and carnitine could be helpful in supporting mitochondrial functions in the acute phase in those with TRMU deficiency.

300 AUTOIMMUNE HEPATITIS PRESENTING AS ACUTE LIVER FAILURE IN AN INFANT. M.M. Tessier, S. Harpavat, Pediatric Gastroenterology, Baylor College of Medicine/ Texas Children's Hospital, Houston, Texas, UNITED STATES.

Background: Autoimmune hepatitis (AIH) is rare in children, with a reported incidence of 0.6 in 100,000. (Deneau M, Hepatology 2013) The average age of presentation is around 10 years of age. AIH is categorized into two types: Type I is associated with smooth muscle antibodies, with elevated F-actin antibodies, positive ANA and elevated total IgG. Type II is typically a more severe form and is associated with elevated liver-kidney microsomal antibodies. AIH is also associated with APECED (Autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy). AIH is a documented cause of acute liver failure (ALF) in children but is not commonly considered in infants. Here we present a case of type I autoimmune hepatitis presenting as ALF in an infant. Case: A 7 month old female presented with a two week history of scleral icterus. At the beginning of her illness she had rhinorrhea but no other symptoms. She was found to have an AST of 1637 U/L (20-60 U/L), ALT of 788 U/L (6-45U/L), conjugated bilirubin of 12.2 mg/dL (<0.3mg/dL), INR of 2.1 (0.8-1.2) and ammonia of 83 umol/L (15-47umol/L). She was more fussy and sleepy than usual, concerning for grade I encephalopathy. Additionally, she required a continuous glucose infusion of 10 mg/kg/min to maintain euglycemia. With this presentation, she was listed for liver transplant due to acute liver failure. Her work up revealed an elevated IgG, F-actin antibodies, and a positive F-ANA. Her liver biopsy was consistent with AIH demonstrating interface hepatitis and portal inflammation with scattered plasma cells. Due to her young age of presentation for AIH, she was evaluated for APECED and found to have a mutation of unknown significance in the AIRE gene. She was started on methylprednisolone 1mg/kg BID with resulting normalization of her INR, glucose levels, and ammonia. Steroids were weaned over the course of 6 weeks, and she was started on azathioprine for the treatment of AIH. Her transaminases have improved on treatment but still remain elevated. She has been taken off the active transplant list as she continues to do well on azathioprine without other signs of autoimmune, skin or candidal disease over a year later. Discussion: AIH is very rare in infants but should be considered in the evaluation of a child under a year of age with liver failure. AIH is one of the few causes of ALF that can be successfully treated with medication. In a recent adult study, approximately 50% of patients who presented with ALF and AIH avoided transplant when treated with steroids, compared to 0% of patients who were not treated with steroids. (Yeoman AD, J Hepatology 2014) However steroids can cause harm in the pediatric ALF population so careful consideration of risks and benefits must be made. AIH is one of the autoimmune disorders associated with APECED; it usually presents before 20 years of age. APECED is thought to be caused by mutations in the AIRE gene, which is involved in immune regulation. Our patient's AIH may be related to her mutation in the AIRE gene, however since she has not displayed any of the characteristics candidal and skin disease which typically present in infancy, she has not been diagnosed with APECED.

301 HEPATIC FORM OF VERY LONG CHAIN FATTY ACID METABOLISM DISORDER IN A THREE YEAR OLD WITH A NORMAL NEBORN SCREEN.. A. Al-Nimr, T.J. Sferra, L. Konczal, University Hospitals Case Medical Center,

Cleveland, Ohio, UNITED STATES. N. Vora, Pediatrics, Metrohealth Medical Center, Cleveland, Ohio, UNITED STATES. R. Gulati, Pediatric Gastroenterology, Metrohealth Medical Center, Cleveland, Ohio, UNITED STATES. Al-Nimr, R. Gulati, T.J. Sferra, L. Konczal, Case Western Reserve University, Cleveland, Ohio, UNITED STATES. A four year old African American boy with history of failure to thrive, dietary aversions, cow's milk protein intolerance, & mild developmental delay presented to our pediatric gastroenterology clinic for low-grade transaminitis. He was born at term with an uneventful postnatal course. Review of system was negative aside from occasional difficulty arousing him in the morning. Physical exam revealed an underweight African American male but was otherwise normal. Hepatic panel showed slightly elevated AST, ALT & GGT. Plasma amino acids & urine organic acids were normal. Ohio Newborn Screening had been obtained 24 hours after birth. All values were within reference range with low risk levels including Amino acid & Fatty acid profiles. Workup including Citrin levels, Hepatitis A IgM antibody, alpha-1 antitrypsin mutation, Anti-LKM IgG autoantibody, ceruloplasmin & ammonia were all unrevealing. EGD was normal. Liver biopsy revealed micro vesicular panacinar steatosis involving 90% of the parenchyma. PAS staining was negative, no iron deposition noted, & trichrome stain for fibrosis was stage 0. Further workup revealed slightly elevated underglycosylated transferrin, Elevated plasma C 14:1 & C 14:2 acylcarnitines. Based on the presentation & clinical findings, he was suspected to have the hepatic form of VLCAD deficiency. VLCAD gene sequencing was normal. Full fatty acid oxidation gene panel is pending. Very long chain fatty acid disorder is a long chain fatty acid disorder described initially in 1992, secondary to long chain acylcarnitine deficiency. It is a rare autosomal recessive disorder, affects about 1 in 40,000-120,000 newborns. Newborn screening has allowed the early identification of affected patients. Few cases of false negative newborn screens have been reported. These have mainly revolved around prematurity, early neonatal illness or use of IV fluids containing dextrose prior to newborn screen. Three clinical phenotypes of VLCAD deficiency have been reported. severe early onset form presents with cardiac manifestation, hypotonia, hepatomegaly, & intermittent hypoglycemia. The hepatic form usually presents during early childhood with hypoketotic hypoglycemia & hepatomegaly. Intermittent rhabdomyolysis, muscle pain & exercise intolerance have been noted in the later onset form. His liver biopsy findings, blood chemistries, difficulty arousing him in the morning likely secondary to a prolonged fasting state & absence of other clinical findings led us to suspect the phenotype of hepatic VLCAD deficiency. VLCAD deficiency is treated appropriately with carnitine supplementation & low-fat formula with avoidance of fasting state in order to maintain euglycemia. It is essential to establish strict dietary schedule with restriction of dietary fat, using medium chain triglycerides supplementation & if needed cornstarch in diet to prevent hypoglycemia. It is important for the clinician to be aware of & suspect fatty acid disorders in patient with similar clinical presentation, even if the newborn screening is normal. Early referral to a gastroenterology or metabolism specialist may prevent long term complications.

302 THE CHALLENGES OF DIAGNOSING AND FOLLOWING WILSON DISEASE IN THE PRESENCE OF ALPURT SYNDROME. S. Khan, G. Silber, B. Morgenstern, T.A. Miloh, Phoenix Children's Hospital, Scottsdale, Arizona, UNITED STATES.

A 17 year old male diagnosed with Alport syndrome at the age of 5, due to the presence of hematuria, proteinuria, and a positive family history, had 2 years of persistent elevation of his transaminases (ALT 108-120IU/L, AST 64-66IU/L) on routine monitoring. Initially, this transaminitis was thought to be due to medications (Lisinopril and/or Losartan). However, transaminases remained elevated despite medication discontinuation. He also had elevated cholesterol (256mg/dL), elevated triglycerides (270mg/dL), and an abdominal ultrasound showed increased echogenicity of the liver. Further diagnostic workup after referral to pediatric gastroenterology showed a low ceruloplasmin (<10mg/dL), low albumin (3.5g/dL), normal total protein (6.4g/dL), and was negative for other causes of chronic liver disease. Urinary 24 hour excretion of copper was 159mcg. A liver biopsy revealed diffuse bridging fibrosis, microvascular and macrovascular steatosis, and a high concentration of copper (1260mcg/mg dry weight) leading to the diagnosis of Wilson disease. Genetic testing confirmed the presence of two heterozygous Wilson disease mutations, V1262F and M64R. The patient was started on trientine and liver enzymes were trending down. He was transitioned to zinc monotherapy 7 years after diagnosis, and liver enzymes normalized with urine 24 hour copper decreased to 91mcg. Prior to zinc monotherapy, his urinary 24 hour excretion of zinc was high at 4987mcg even though he was not on supplementation. The patient is currently asymptomatic with ALT 57IU/L, AST 31IU/L, normal serum creatinine, and a slightly elevated urine protein to creatinine ratio of 0.26mg/mg. Conclusion: The coexistence of Wilson disease with Alport syndrome has not previously been reported. The diagnosis of Wilson disease and its ongoing monitoring is challenging when associated with an

underlying renal disease such as Alport syndrome. Proteinuria can lead to low ceruloplasmin since it is among serum proteins inappropriately filtered by the damaged glomerulus, and can also lead to increased urinary loss of heavy metals such as zinc and copper. Elevated transaminases may be attributed to dyslipidemia or drug induced hepatotoxicity. The accurate diagnosis of Wilson's disease is essential for targeted therapy and improved prognosis.

303 JAK-2 KINASE RELATED MYELOPROLIFERATIVE DISORDER PRESENTING WITH PORTAL HYPERTENSION AND HEPATIC EOSINOPHILIC INFILTRATES IN A CHILD WITH ULCERATIVE COLITIS. F.W. Jafri, M.A. Bozic, J.P. Molleston, Pediatric Gastroenterology/Hepatology and Nutrition, Riley Hospital for Children, Indianapolis, Indiana, UNITED STATESK.A. Robertson, Pediatric Hematology/Oncology, Riley Hospital for Children, Indianapolis, Indiana, UNITED STATESK.M. Kuboye, Greater Meridian Health Clinic, Meridian, Mississippi, UNITED STATES.

A previously healthy 13-year-old male presented for evaluation of chronic fatigue, nausea and epigastric pain. Initial physical exam was normal. He had anemia (Hgb 11.2g/dL), eosinophilia (35%), positive anti neutrophil cytoplasmic antibody, and elevated erythrocyte sediment rate with normal albumin, liver profile, and platelets. Colonoscopy revealed severe pancolitis; consistent with ulcerative colitis. He was started on corticosteroids and azathioprine, and eventually began infliximab. Over the ensuing months, he developed hepatosplenomegaly with eosinophilia and continued anemia, despite no symptoms or signs of colitis. Abdominal ultrasound and computed tomography revealed normal liver parenchyma and vasculature. Upper endoscopy revealed grade I esophageal varices. MRCP showed mild focal narrowing in the biliary tree with subtle beading suggestive of early primary sclerosing cholangitis. Subsequent liver biopsy demonstrated mild cholangiopathy with no signs of advanced fibrosis; however immature hematopoietic cells of the granulocytic lineage within the sinusoids consistent with myeloid or eosinophilic precursors were seen. The patient then underwent a bone marrow biopsy which revealed dysplastic changes with massive eosinophilia consisting of immature and mature eosinophils along with erythroid hyperplasia, consistent with myeloproliferative disease (MPD). Cytogenetics were abnormal with a clonal translocation t (8; 9) (p22; p24) resulting in constitutive activation of the JAK-2 tyrosine kinase. Treatment with imatinib was started and eventually he underwent an unrelated donor bone marrow transplant to treat his MPD. His post-transplant course was complicated by moderate hepatic dysfunction, variceal bleeding, worsening portal hypertension, and graft vs. host disease involving the skin and gut with eventual resolution of symptoms, including his ulcerative colitis. He does however, continue to have persistent physical and hematologic evidence of portal hypertension. Discussion: This case reports a rare diagnosis of myeloproliferative disease in a pediatric patient who initially presented with findings suggestive of ulcerative colitis (UC). Subsequent findings of persistent anemia, eosinophilia and hepatosplenomegaly despite adequate treatment of his UC resulted in the diagnosis of MPD with a rare form of the JAK2 mutation. The JAK2 mutation likely played a role in this patient's initial presentation of colitis, liver disease, and MPD. Importantly, in the literature JAK2 mutations have been associated with both portal hypertension and inflammatory bowel disease; and should be considered in a child with unexplained portal hypertension in the context of hematologic abnormalities.

Poster Session III
Saturday, October 25, 2014

BASIC IBD

339 SMALL BOWEL OBSTRUCTION AND PNEUMATOSIS INTESTINALIS AS AN UNCOMMON PRESENTATION OF INFLAMMATORY BOWEL DISEASE. A. Behrle, S. Khan, Gastroenterology, Hepatology, and Nutrition, Children's National Health System, Washington, District of Columbia, UNITED STATES.

The clinical presentation of pediatric inflammatory bowel disease (IBD) can vary widely. Because the onset of symptoms can be insidious, occasionally it is difficult to determine the presence of chronicity. In addition, infection can be a confounding factor and delay diagnosis. We report a case of a previously healthy 16-year-old girl who presented with two days of diffuse abdominal pain and vomiting, and one day of hematochezia and fever. Initial laboratory investigation revealed leukocytosis, anemia, hypoalbuminemia, and elevated inflammatory markers. Stool infectious studies revealed the presence of both adenovirus and norovirus. Imaging including abdominal CT revealed dilated small bowel concerning for obstruction, as well as pneumatosis intestinalis and portal venous gas.

Given a clinically benign abdominal exam, she was closely monitored with serial films. Fifteen days after symptom onset, she developed upper extremity swelling and subsequent tachypnea, and was found to have a PICC-associated thrombus and right lower lobe pulmonary embolus. She was initiated on anticoagulation therapy and her respiratory symptoms improved, but she continued to have abdominal pain, vomiting, diarrhea, and fever. A repeat CT 20 days after symptom onset showed persistent small bowel obstruction, and the decision was made to proceed with diagnostic laparoscopy. Because of extensive inflammation, the procedure was converted to an open laparotomy. Operative findings included markedly dilated proximal jejunum and inflamed distal small bowel with areas concerning for necrosis. Fifty-three centimeters of jejunum with areas of stricture as well as creeping fat and extensive pneumatosis were resected. Histology revealed extensive ulceration, transmural mixed inflammation, and hemorrhage, but no granulomas. Her post-operative course was complicated by the development of multiple intraabdominal abscesses and a wound infection requiring antibiotics and the placement of a wound VAC. She continued to have evidence of systemic inflammation, developing a second PICC-associated thrombus despite therapeutic anticoagulation. With continued concern for IBD given lack of improvement, she underwent upper endoscopy and colonoscopy 20 days after surgery. Biopsies revealed terminal ileitis with cryptitis and crypt abscesses, an ulcer in the cecum, and acute colitis in the ascending colon. Noted in the ileum and proximal colon was the presence of architectural distortion suggestive of early chronic change. The patient was initiated on corticosteroids with the presumed diagnosis of Crohn Disease. Subsequently, she had improved abdominal pain, normalized inflammatory markers, hemoglobin, and albumin, and improved ability to tolerate enteral feeds. Unfortunately, as her diet was advanced, she again developed abdominal pain and vomiting. MR enterography showed evidence of a continued stricture at the anastomotic site which necessitated the resumption of nasogastric tube feeds for symptom control. This case not only illustrates an uncommon presentation of IBD, but also prompts further exploration of the role of enteric viruses in its diagnosis.

340 ABDOMINAL ANGIOSTRONGYLIASIS IS A MIMICKER OF NEW ONSET INFLAMMATORY BOWEL DISEASE. M. Panopoulos, N. Channabasappa, J. Park, University of Texas Southwestern, Dallas, Texas, UNITED STATES.

Angiostrongylus costaricensis is a parasitic nematode that usually infects the mesenteric vasculature of rodents but can also infect humans. Abdominal angiostrongyliasis causes severe gastrointestinal symptoms and can mimic new onset inflammatory bowel disease. We report a case of a 4 year old girl from El Salvador admitted with 4 weeks of fever, hepatomegaly, hematochezia and weight loss. Workup included blood work which was notable for anemia, thrombocytosis and lymphocytosis, with prominent eosinophilia. Urinalysis had evidence of hematuria and proteinuria. Gastroenterology nephrology, oncology and infectious disease were consulted. She underwent kidney biopsy which showed acute diffuse proliferative glomerulonephritis. She also underwent bone marrow biopsy which showed eosinophilia. CT of the abdomen showed focal circumferential bowel wall thickening in the ascending colon and small bowel. EGD and colonoscopy were done. Upper endoscopy was unremarkable both grossly and histologically. Colonoscopy did show ulceration in the ascending colon and cecum. Histologic review of the colonic tissue revealed eosinophils, and numerous granulomas containing the egg forms of *angiostrongylus costaricensis*. Interestingly, her earlier workup had included several ova and parasite samples which were negative. She was started on albendazole initially, but this was discontinued prior to discharge. She followed up 2 months later and her hematochezia and hepatomegaly had resolved. She had an increased appetite and was gaining weight well. Review of case reports in the literature reveals abdominal angiostrongyliasis is a largely self-limited disease, although cases of intestinal necrosis have been reported. It is rare, but should be in the differential diagnosis in a patient with new onset IBD symptoms such as hematochezia, anemia and weight loss, especially in the context of travel from South America.

341 ULCERATIVE COLITIS PRESENTING IN AN ESOPHAGEAL INTERPOSITION. N.H. Patel, D.I. Mehta, J.A. Bornstein, Pediatric Gastroenterology, Center for Digestive Health and Nutrition, Arnold Palmer Hospital for Children, Orlando, Florida, UNITED STATES.

This 14 year old white male was born with VACTERL association consisting of pure esophageal atresia; complex congenital heart disease; hemivertebra and a left lateral chest wall deformity; and a low imperforate anus. He underwent multiple surgeries including cervical esophagostomy to provide for a spit fistula, gastrostomy tube placement, and colostomy followed by posterior sagittal anorectoplasty and subsequent colostomy takedown. Over the first few months of life, he had several episodes of aspiration pneumonia. He was ventilator dependent

and underwent tracheostomy placement at 6 months of age. At 9 months of age, a right colonic esophageal interposition was performed due to concern for chronic lung disease from aspiration. Postoperatively, a number of upper endoscopies and esophageal dilations were performed. At age 34 months, he presented with rectal bleeding. Flexible sigmoidoscopy found chronic active proctitis and upper endoscopy found Paneth cell metaplasia in the interposed distal colonic mucosa without active inflammation. At age 4 ½ years, upper endoscopy was normal but repeat sigmoidoscopy showed active colitis without chronic changes. Early inflammatory bowel disease was suspected and he was started on Pentasa. He continued having hematochezia and was treated with prednisone. Due to recurrent hematochezia, he was reevaluated at 5 ½ years of age with the finding of focal acute colitis in the colonic interposition with chronic and minimally active colitis in the native colon. He was continued on 5-ASA therapy and was started on 6-mercaptopurine (6-MP) along with Entocort to try decrease prednisolone. He subsequently developed pancreatitis and 6-MP was discontinued. Throughout this time he was receiving supplemental feeds via his gastrostomy tube. He was steroid dependent. At 7 years of age biopsies of the colonic interposition revealed mild chronic inflammation, and colonoscopy found active chronic colitis consistent with chronic inflammatory bowel disease. He continued to be managed primarily with 5-ASA compounds, intermittent prednisolone, as well as acid suppression. The patient was lost to follow-up until age 9 years at which time he presented with a history of poor growth, 5-7 stools/day and hematochezia. He complained of occasional chest pain but no dysphagia. He had not been on any GI medications for over a year. Upper endoscopy found marked acute and chronic inflammation of the interposed colonic mucosa. He had diffuse chronic active colitis of the native colon with a normal neoterminal ileum. At this time, given our experience with topical steroids for eosinophilic esophagitis, he was treated with swallowed fluticasone and oral Pentasa. He achieved clinical remission and remained clinically well for next few years. Due to poor compliance with fluticasone, he was transitioned to viscous budesonide. After several months on Pentasa and budesonide, repeat endoscopic evaluation was performed at the age of 12 years. At this time, both the colonic interposition and the native colon were normal without any active or chronic inflammation. He has remained well on this therapy with hemocult negative stools, improved growth, and normal laboratory evaluations.

CELIAC/EOE/ALLERGIC ENTEROPATHY

357 MONTELUKAST THERAPY FOR PATIENTS WITH EOSINOPHILIC GASTROINTESTINAL DISORDERS.
S.S. Eng, S.K. Abraham, Pediatrics, Nemours Children's Clinic, Philadelphia, Pennsylvania, UNITED STATES.
Eosinophilic gastro-intestinal conditions continue to be a clinical management challenge. Dietary manipulation, which may be helpful, is cumbersome and difficult to maintain for any length of time. There is a limited repertoire of medications that can be offered to patients with eosinophilic disorders of the GI tract. We present two male patients, ages 11 and 13 years, who carried a diagnosis of eosinophilic esophagitis (EoE), eosinophilic gastro-enteropathy and/or colitis. Both patients initially presented with varying complaints, from dysphagia to bloody diarrhea, and were eventually diagnosed based on histological findings after upper gastrointestinal endoscopy and/or colonoscopy. Both had no abnormal physical exam findings. Patient A was noted to have up to 20-35 eosinophils/hpf in the distal to proximal esophagus and was diagnosed with EoE. Patient B was noted to have >50 eosinophils/hpf with microabscesses throughout his upper gastrointestinal tract, in the terminal ileum and throughout the colon and was diagnosed with EoE, eosinophilic gastro-enteropathy and colitis. Montelukast therapy was added to the therapeutic protocol after both patients failed a trial of proton-pump inhibitor and, in the case of Patient B, a rigorous elimination diet. Patient A had symptomatic and histologic improvement on daily, low-dose of montelukast (5mg) and 40mg of omeprazole approximately 6 months after initiating therapy. His montelukast dose was then increased to 10mg and he subsequently had complete resolution of EoE on repeat endoscopy and biopsy. After minimal improvement on an extensive elimination diet, mesalamine, and steroid therapy, Patient B began a trial of 10mg of montelukast and 40mg pantoprazole daily. Several months after initiating the montelukast and proton-pump inhibitor combination, his repeat endoscopy showed near resolution of EoE with normal variation in gastric eosinophil content. Repeat colonoscopy also showed top normal range of eosinophilia on colon biopsies. In conclusion, both patients showed improvement clinically and near-resolution histologically when montelukast was added to therapy with a proton-pump inhibitor. The cases above illustrate the need for continued research on the beneficial effects of combination medical therapy with a systemic leukotriene receptor antagonist and proton pump inhibitor for cases of gastrointestinal eosinophilic disorders.

358

EOSINOPHILIC GASTROENTEROPATHY PRESENTING WITH HYPOALBUMINEMIA. R.G. Vallina, R. Mittal, Medical Education, Miami Children's Hospital, Miami, Florida, UNITED STATES. A. Baghel, Kasturba Medical College, Manipal, Miami, Florida, UNITED STATES.

Eosinophilic Gastroenteropathy (EG) is a heterogeneous spectrum of diseases including eosinophilic esophagitis, eosinophilic gastritis, and eosinophilic colitis, characterized by patchy or diffuse eosinophilic infiltration of the gut. EG is rare, with only a handful of cases described in the literature. It can present with abdominal pain, bloating, failure to thrive, vomiting, diarrhea, edema and uncommonly with protein and fat malabsorption. Although the role of food allergy has not been as clearly defined in EG as in eosinophilic esophagitis, an improvement has been seen in disease activity with an elemental or elimination diet. Here we describe a unique case of a three year old girl with multiple food allergies who presented with periorbital swelling. The workup revealed significant hypoalbuminemia, anemia, low immunoglobulin levels and high peripheral eosinophilia. The stool study showed high alpha1-antitrypsin level. The patient needed multiple albumin and immunoglobulin transfusions. Further, endoscopic biopsies showed eosinophilic infiltrates in the esophagus, stomach and the duodenum. She was diagnosed with protein-losing enteropathy secondary to EG. Treatment was started with prednisolone and the patient was advised to avoid foods to which she was allergic. The patient improved and serum albumin and immunoglobulin levels trended up to normal.

359

HERPES SIMPLEX VIRUS ESOPHAGITIS IN AN IMMUNOCOMPETENT HOST- A CASE REPORT. T. Sebastian, A.A. Bader, Pediatric Gastroenterology, Hepatology and Nutrition, Childrens National Medical Center, Washington, District of Columbia, UNITED STATES.

Introduction: Herpes Simplex virus esophagitis is a well recognized infection in an immunocompromised host. However it has rarely been described in the immunocompetent host. There have been only a few published case reports in Pediatrics. We report here a case of an immunocompetent patient with Herpes esophagitis who presented to our institution Case: A 22 month old previously healthy boy presented to another hospital with a 5 day history of progressively worsening oral intake, odynophagia, fever and lethargy. On physical exam he appeared dehydrated. No oropharyngeal lesions were appreciated. Labs were remarkable for elevated CRP and bandemia. CT did not reveal any significant abnormalities. ENT performed a bedside fiberoptic examination that showed erythema within the proximal esophagus. He received a single dose of IV Dexamethasone and was started on Clindamycin. Despite this, there was no significant improvement in symptoms. Thus he was transferred to our institution for further evaluation. Following initial assessments, an upper GI series was normal. EGD demonstrated erythema, ulceration, white exudates and furrowing within the entire esophagus. Biopsies were consistent with active esophagitis, and neutrophilic exudates in the mid- and distal esophagus. Immunostain for HSV as well as cultures were positive. Serology for EBV, CMV and HIV were negative. He was started on IV acyclovir, which was later transitioned to PO with subsequent improvement of his symptoms. His oral intake improved. Immunoglobulin levels and a T-lymphocyte panel requested by infectious diseases were normal except for elevated IgE. An EGD repeated after 6 weeks of PPI therapy showed nodularity in the lower esophagus. Biopsy showed chronic inflammation and 20 eosinophils/HPF in mid esophagus. Conclusion: HSV is well known to cause visceral diseases in the immunocompromised host with the esophagus being one of the most common sites of involvement. HSV esophagitis in the immunocompetent host is rare with only a few pediatric cases being reported. Majority of pediatric cases were male. HSV esophagitis should be considered in immunocompetent patients who complain of chest pain and odynophagia with or without herpetic lesions. HSV esophagitis is self-limiting, however early initiation of acyclovir may be benefit treatment of severe odynophagia. However, its role in therapy for HSV esophagitis in the pediatric population has not been fully evaluated. There have been some reported cases HSV esophagitis where the patients developed endoscopic features of eosinophilic esophagitis. The etiology of this phenomenon is unclear at this point and would require further studies.

360

CASE REPORT OF ESOPHAGEAL DYSMOTILITY IN A CHILD WITH EOSINOPHILIC ESOPHAGITIS (EoE). S.K. Palle, G. Tenjarla, Pediatric Gastroenterology, Emory University, Atlanta, Georgia, UNITED STATES. A. Mohammed, Pediatric Gastroenterology, MCGG, Macon, Georgia, UNITED STATES. K. Freedle, Allergy & Immunology, Emory University, Atlanta, Georgia, UNITED STATES.

Clinical presentation of EoE reflects esophageal dysfunction. An association between EoE and altered esophageal motility has been described in small case report series and retrospective studies conducted in adults. We present a case of esophageal dysmotility disorder in a child with EoE. A 13 yrs. old Caucasian male presented to local ER with

severe epigastric pain for several weeks. He underwent EGD with biopsies, which revealed severe proximal and distal esophageal eosinophilia. He had a negative basic food allergy panel and IBD serology. Discharged home on oral Flovent and Nexium. Two weeks later, the patient complained of recurrence of epigastric pain, new onset of chest pain, and a sensation of solid food getting stuck in his chest. His Flovent dose was increased with improvement in his symptoms. One month later, he was readmitted with worsening symptoms and subsequently underwent EGD and colonoscopy, with esophageal biopsies showing no eosinophils and normal colon biopsies. In addition, he had a normal VCE and Chest CT. Esophageal manometric studies revealed a pattern consistent with diffuse esophageal spasm and Nut cracker esophagus. Psychological evaluation did not reveal a functional explanation for his symptoms. Following a normal EKG he was started on a calcium channel blocker and discharged home on Nexium and Flovent twice a day. Subsequently, he did well with this regimen for one month except for developing some adverse effects of the calcium channel blocker, which was slowly weaned off later. With recurrence of symptoms, a few months later, he had a repeat endoscopy with biopsies revealing a reoccurrence of EoE in the distal esophagus (Eos 20 to 50 /HPF). Elavil was added to his regimen to help his depression. Despite these treatments, he continued to have severe pain and lost weight and was readmitted, with unsuccessful attempts to feed enterally, he was placed on TPN for 8 weeks. He had a repeat Esophageal manometric study which still showed Nutcracker esophagus. At the end of 8 weeks of TPN, a 6-food (dairy, soy, eggs, wheat, nuts and fish) elimination diet was implemented without any improvement. So, he received a trial of Botox injection in the LES with significant but transient improvements in his symptoms. Subsequently, he was readmitted and scoped multiple times. He is now 17 yrs. old with persistent symptoms. The patient has been evaluated in 2 other tertiary centers, both of which confirmed Nutcracker esophagus and one of which showed persistent EoE. He came to our tertiary center recently and after having had negative SPT and APT testing for the top 10 foods known to cause EOE, he was placed on an elemental formula delivered via the G tube. Our case concludes that both EoE and esophageal dysmotility contribute towards the symptoms of chest pain and dysphagia even in children, though mechanisms of this association are unclear at this time. Resolution of dysmotility does not appear to coincide with mucosal healing of EoE.

CLINICAL/TRANSLATIONAL IBD

375 CROHN'S DISEASE ASSOCIATED WITH ACUTE LYMPHOPROLIFERATIVE SYNDROME. K. Patrick, E. Nagel, J. Pavlak, H. Conrad, A. Cornelius, I. Hashemi, Helen DeVos Children, Grand Rapids, Michigan, UNITED STATES.

Autoimmune lymphoproliferative syndrome (ALPS) is a disorder of lymphocyte homeostasis characterized by non-malignant chronic lymphoproliferation, autoimmunity, and increased risk of lymphoma. To date, there is no published association between inflammatory bowel disease (IBD) and ALPS. Since ALPS is associated with immune dysregulation, it is reasonable to find an association with other autoimmune conditions and not only autoimmune cytopenias. A 15-year-old male was admitted to the inpatient service for acute on chronic abdominal pain. He was recently diagnosed with Crohn's disease via biopsy showing inflammation in the ileum and intermittently throughout the colon and MRE showing inflammatory thickening in the terminal ileum and cecum. He was initially treated with prednisone at 30 mg and azathioprine at 100 mg daily with resolution of diarrhea but persistent abdominal pain. Two weeks later, he was weaned to 20 mg but was increased to 40 mg in the two weeks following secondary to worsening abdominal pain despite a therapeutic range 6-MP level. On admission, he had left lower quadrant pain with an elevation in CRP to 60.1 mg/L and decrease in hemoglobin to 10.8 g/dL. Due to evidence of failure to induce Crohn's remission with azathioprine and prednisone, starting infliximab was recommended to the family. During that conversation patient's mother revealed a recent diagnosis of T-cell lymphoma and a history of malignant melanoma. In addition, the patient's maternal great-grandfather also had a history of melanoma, T-cell lymphoma, and Waldenstrom's macroglobulinemia. Due to the strong family history of lymphoproliferative conditions, it was recommended to screen the patient for ALPS to help elucidate his risk for lymphoma. Flow cytometric analysis found 5.4% double negative T-cells. This is a marked increase in a normally rare population of T cells (typically <1%) that are alpha beta T-cell receptor (TCR) positive, as well as negative for both CD4 and CD8 coreceptors (double-negative T cells; DNT), which is highly indicative of ALPS. Since the patient's Crohn's disease was still uncontrolled, he was started on infliximab for Crohn's disease as well as a course of monthly IVIG infusion for presumed ALPS with plans to repeat flow cytometry in 6 months as confirmation of his dual diagnosis. This case illustrates a potential association of Crohn's disease and ALPS, which has not previously been described. It

also raises questions for our patient and other patients with IBD concerning the potential risk for lymphoma secondary to either treatment or possible underlying immune dysregulation.

376 METRONIDAZOLE NEUROTOXICITY IN A TEENAGE GIRL WITH CROHN'S DISEASE: THE FIRST PEDIATRIC REPORT OF METRONIDAZOLE-INDUCED ENCEPHALOPATHY. D. Lévesque, Pediatric Gastroenterology and Nutrition, Montreal Children's Hospital, Montreal, Quebec, CANADAA. Ricciuto, Pediatrics, Montreal Children's Hospital, Montreal, Quebec, CANADA.

Background: Although metronidazole is generally regarded as safe by pediatric gastroenterologists, who use it frequently for treating inflammatory bowel disease (IBD), it has been associated with a number of uncommon, but serious, neurotoxic effects. The most widely recognized of these is peripheral neuropathy, but others include cerebellar dysfunction, seizures and encephalopathy. The underlying pathophysiology is unknown. While metronidazole-induced encephalopathy (MIE) has been reported in adults, to our knowledge, there are no previous reports in children. Objectives: To report the first pediatric case of MIE with the goal of sensitizing the medical community, particularly gastroenterologists, to its existence so as to optimize management and, ultimately, patient outcomes. Case Report: A 16-year-old girl, presenting with isolated perianal abscess, was treated with a combination of metronidazole and ciprofloxacin. Mesalamine was initiated shortly thereafter for suspected Crohn's disease, while investigations were underway. Following definitive diagnosis of Crohn's disease, budesonide was added, as well as lansoprazole, the latter for nausea and vomiting attributed to upper gastrointestinal IBD. 8 weeks into antibiotic therapy (cumulative metronidazole dose 84 g), she presented to the emergency complaining of electric shock-like pain in her extremities, speech impairment, visual disturbances, unsteadiness and weakness. The symptoms, intermittent for the preceding two weeks, had improved by the time of assessment and the patient was diagnosed with conversion disorder, despite slurred speech, nystagmus and a positive Romberg being noted on exam. 2.5 weeks later, during assessment for mild knee arthritis, she reported ongoing symptoms, which had been constant for the preceding several days. She was also found to be lethargic, dysarthric and ataxic, and was thus admitted. Head MRI, performed emergently, demonstrated low T1, high T2 signal in the dentate nuclei, red nuclei, ventral medulla, dorsal pons and splenium of the corpus callosum with bilateral distribution and mild mass effect, without restricted diffusion or enhancement. Given the highly suggestive nature of these findings for metronidazole-related central nervous system toxicity, metronidazole was immediately discontinued. Subsequently, there was rapid clinical resolution of the encephalopathy and cerebellar dysfunction and full radiologic normalization was achieved by 8 months. The peripheral neuropathy, however, persisted in a significant, disabling fashion, with ongoing painful paresthesias at 11 months post metronidazole withdrawal, despite multiple medication trials. Conclusions: Despite the widespread use of metronidazole, its neurotoxicity continues to be under-recognized. We present the first pediatric case of MIE, in a teenager receiving metronidazole for perianal Crohn's disease, occurring in association with cerebellar dysfunction, visual complaints and peripheral neuropathy. The delay in diagnosis, potentially negatively impacting patient outcome, emphasizes the urgent need to raise awareness of this important adverse effect.

377 OVERLAPPING IMMUNE DYSREGULATION - DIAGNOSIS AND TREATMENT IN A CASE REPORT OF FAMILIAL MEDITERRANEAN FEVER AND CROHN'S DISEASE. K. Queliza, R. Sanghavi, A. Patel, UT Southwestern Medical Center, Dallas, Texas, UNITED STATES.

Introduction: Crohn's disease (CD) and Familial Mediterranean fever (FMF) are inflammatory disorders both characterized by immune dysregulation. They share similar clinical manifestations including abdominal pain, diarrhea, arthritis and elevated inflammatory markers. The prevalence of CD may be increased in the setting of FMF suggesting genetic commonality between these two disorders. Given the presence of significant pathologic and physiologic overlap, differentiating CD and FMF can be challenging. Case: Our case describes an 11-year-old Caucasian male of non-Mediterranean descent with a past medical history of anemia who was referred to pediatric gastroenterology for evaluation of chronic abdominal pain, fatigue, failure to thrive, elevated inflammatory markers, and hypoalbuminemia. Initial endoscopy revealed mild colitis while capsule endoscopy demonstrated additional findings consistent with CD. The patient was subsequently started on infliximab with initial improvement in symptoms. Two months later, he developed periodic fevers - occurring every 2 weeks and lasting less than 24 hours. These episodes were accompanied by periumbilical pain and fatigue. Repeat endoscopy and capsule demonstrated active CD. He was also found to have infliximab antibodies, thereby prompting initiation of adalimumab. The fevers were thought to be an inflammatory response as a result of uncontrolled small bowel CD.

Despite change in management, the patient continued to have intermittent fevers and abdominal pain prompting hospitalization. Infectious work-up was negative and abdominal CT unremarkable for abscess or fistula formation. Consideration was given to chronic granulomatous disease and periodic fever syndromes and, ultimately, the patient was found to be heterozygous for the G304R variant in the MEFV gene. A diagnosis of FMF was made. The patient was placed on colchicine with subsequent resolution of fevers. Unfortunately, two additional hospitalizations were needed for severe hypoalbuminemia, presumed to stem from chronic inflammation in the setting of inadequately controlled FMF and/or CD. Enteral therapy for small bowel CD was started. Over the course of several months, the patient exhibited improving inflammatory markers, serum albumin and weight gain as well as resolution of periodic periumbilical abdominal pain. Discussion: This is a report of CD refractory to biologic therapy with concurrent FMF in a pediatric patient. Disease activity was eventually controlled with enteral therapy and colchicine. The patient posed a diagnostic dilemma given significant symptomatic overlap between his two diseases (CD and FMF). CD was also resistant to medication therapy further challenging management. This case highlights the importance of considering concomitant FMF in patients with CD who have ongoing periumbilical abdominal pain and unexplained fevers.

378 GRANULOMATOUS GINGIVITIS, A RARE MANIFESTATION OF ORAL CROHN'S DISEASE. E. Shin, C. Cuffari, pediatric gastroenterology, Johns Hopkins Hospital, Baltimore, Maryland, UNITED STATES.

The oral cavity is frequently affected in 30-48% of patients with inflammatory bowel disease.¹ In Crohn's disease (CD), a wide variety of oral manifestations have been described, including buccal mucosal swelling, mucogingivitis and linear ulcerations, as well as nonspecific findings, including angular cheilitis and perioral erythema. Herein, we present a case of isolated granulomatous gingivitis, a rare manifestation in CD in a Caucasian female with severe colonic CD. The patient is now a 22 year old female who initially presented at age 13 with severe angular cheilitis that responded well to nutritional therapy. However, the colonic CD was recalcitrant to combination anti-TNF and methotrexate therapy ultimately requiring a total colectomy at age 19. She then presented several weeks thereafter with new onset gingival swelling. At the time, she was not on systemic therapy. She was subsequently referred to pediatric dermatology and periodontology for assessment and management. The patient described moderate discomfort and occasional gingival bleeding when brushing her teeth. On exam, there were erythematous, non-bleeding papillated plaques on the upper gum line spanning from the left incisor to the right. There was a smaller distribution of similar appearing papules on the lower gingiva. There were no ulcerations or erosions noted. The buccal mucosa, hard and soft palate appeared normal, and there was no swelling of her face or lips. She had an elevated ESR of 42 (N< 25) and CRP of 2.8 (N< 0.5) as well as persistent iron deficiency anemia with Hb 7.7 despite prolonged iron therapy. A biopsy of the gingiva was performed, and showed on histological examination scattered aggregates of non-caseating granulomatous inflammation consisting of lymphocytes, epithelioid histiocytes and multinucleated giant cells. The patient was diagnosed with granulomatous gingivitis associated with CD and received topical clobetasol cream 0.05% therapy. She responded well to the topical therapy, and has had no disease recurrence up to 2 years in follow-up. Moreover, all of her inflammatory markers have normalized. This is the first report of a pediatric patient with CD complicated by granulomatous gingivitis. The patient's case illustrates how this oral manifestation does not parallel intestinal disease activity. Indeed, the patient underwent a complete colectomy prior to her diagnosis of granulomatous gingivitis. It is also interesting to note that the histologic findings of granulomatous gingivitis were indistinguishable from those of the intestinal CD, as was noted in our patient's colonic biopsies. This case is also instructive for pediatrician's in establishing a focused assessment of the oral cavity to include the periodontal and gingival mucosa in patients with CD. Recent studies have shown that both gingivitis and periodontitis are higher in patients with IBD than in healthy matched control². Furthermore, optimal diagnostic and therapeutic strategies should be developed to treat both oral and intestinal inflammation disease since these oral manifestations of CD may not necessarily mimic intestinal disease activity.

379 PEDIATRIC PATIENTS WITH INFLAMMATORY BOWEL DISEASE AND EOSINOPHILIC ESOPHAGITIS: A CASE SERIES. M. Rose, R. Sockolow, A. Solomon, Pediatric Gastroenterology, New York-Presbyterian Hospital/Weill Cornell, New York, New York, UNITED STATES.

Background: Inflammatory bowel disease and eosinophilic esophagitis are each rapidly increasing in incidence in the pediatric population. Eosinophils play a role in both inflammatory bowel disease and eosinophilic esophagitis, and there is also an immunologic component proposed for both conditions. There is a cohort of patients who have

both inflammatory bowel disease and eosinophilic esophagitis, however the incidence and association is unknown (1,2,3). Abstract: Six pediatric patients ages 14.67 to 17.25 years with both inflammatory bowel disease and eosinophilic esophagitis were identified. Five patients were male and one was female. Two patients were diagnosed with Crohn's disease and four patients were diagnosed with ulcerative colitis. The mean age of inflammatory bowel disease diagnosis was 10.1 years. Three patients had pancolitis, one patient had pancolitis with terminal ileal disease, one had isolated terminal ileal disease, and one had disease from the transverse colon to rectum. Two patients required total abdominal colectomy. Other disease complications occurred in two patients and included sinus venous thrombosis and recurrent infections. The mean age of eosinophilic esophagitis diagnosis was 13.2 years. Eosinophil count ranged from 15-30 eosinophils per high-powered field in the proximal, mid and distal esophagus. All patients were treated with inhaled steroids (fluticasone). The inflammatory bowel disease diagnosis preceded the eosinophilic esophagitis diagnosis in all patients except for one who was diagnosed with both conditions concurrently. At the time of eosinophilic esophagitis diagnosis, inflammatory bowel disease was in remission based on PCDAI/PUCAI less than 10 in four patients; PUCAI 30 in one patient with pouchitis; and PUCAI 40 in the patient who was diagnosed with both conditions simultaneously. Family history of either condition was significant in two patients, one with a father with inflammatory bowel disease and the other with two siblings, a mother and maternal aunt with eosinophilic esophagitis. Conclusion: Inflammatory bowel disease and eosinophilic esophagitis are separate disease entities that can occur together. In most cases, inflammatory bowel disease was in remission at the time of eosinophilic esophagitis diagnosis. It remains unclear if there is an overlapping immunologic or genetic component linking these two chronic diseases. 1. Mulder DJ, Hookey LC, Hurlbut DJ, Justinich CJ. Impact of Crohn Disease on Eosinophilic Esophagitis: Evidence for an Altered TH1-TH2 Immune Response. *JPGN*. 2011 Aug; 53(2):213-5. 2. Suttor VP, Chow C, Turner I. Eosinophilic Esophagitis with Crohn's Disease: A New Association or Overlapping Immune-Mediated Enteropathy? *Am J Gastro*. 2009 Mar; 104(3):794-5. 3. Woodruff SA, Masterson JC, Fillon S, Robinson ZD, Furuta GT. Role of Eosinophils in Inflammatory Bowel and Gastrointestinal Diseases. *JPGN*. 2011 June; 52(6):650-61.

380 SINGLE-CENTER EXPERIENCE OF NATALIZUMAB IN YOUNG PATIENTS WITH INFLAMMATORY BOWEL DISEASE. N. Singh, R. Deshpande, M. Check, S. Rabizadeh, M. Dubinsky, Cedars-Sinai Medical Center, Los Angeles, California, UNITED STATES.

Background: Natalizumab (NTZ) is a humanized monoclonal antibody inhibiting lymphocyte migration and prescribed in Crohn's disease (CD) patients failing anti-TNF therapies. Due to the risk of progressive multifocal leukoencephalopathy (PML) in JC virus (JCV) positive patients, NTZ has not been widely used in clinical practice. Emerging evidence from multiple sclerosis suggests that an anti-JCV antibody index of greater than 0.9 is correlated with the risk of PML. Published experience of off-label use of NTZ in pediatric patients is lacking. Given the overlap of the mechanism of vedolizumab and NTZ, it is important to determine whether anti-integrins are an efficacious class in pediatric IBD patients. We aimed to describe the experience of NTZ in pediatric IBD patients at a tertiary care IBD center. Methods: A retrospective chart review was performed in IBD patients less than 21 years of age receiving NTZ therapy prior to March 2014. Charts were reviewed for demographics, surgical history, disease behavior, location and activity. Prior treatments and NTZ treatment information (duration/dose/JCV status and index/adverse events) were examined. Descriptive statistics, including median and range were computed. Results: Ten patients received NTZ between September 2012 and March 2014. Median age at diagnosis was 10.5 (range 7-16) years and median disease duration at NTZ initiation was 71 (range 13-156) months (Table 1). Four patients had prior CD surgery. All patients (10/10) had failed at least one anti-TNF agent (infliximab, adalimumab, or certolizumab pegol), 60% (6/10) had failed at least two anti-TNF agents and 40% (4/10) had failed three anti-TNF therapies. At last follow-up, a median of 6 (range 2-22) infusions per patient were administered. Baseline median C-reactive protein (CRP) level was 13.6 (range <0.1-36.1)mg/dL. Eighty six percent of patients (6/7) had at least a 50% decrease of CRP level by 16 weeks. Forty percent (4/10) of patients were JCV antibody positive at baseline and all four had an anti-JCV antibody index above 0.9 (median 3.36). All JCV antibody negative patients remained negative during NTZ treatment. There were no serious adverse events, and no cases of PML. All NTZ patients have now been transitioned to Vedolizumab. Conclusion: In our experience, NTZ is a safe and efficacious medication in pediatric IBD patients. The delayed decrease in CRP with NTZ initiation is consistent with the estimated time to steady state of 16 to 24 weeks. Given the favorable phase 3 trial safety profile of Vedolizumab, it should be a preferred treatment option to natalizumab in pediatric IBD patients. Baseline Demographics of patients starting Natalizumab

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The clinical presenting features of inflammatory bowel disease (IBD) overlap infectious, autoimmune and, rarely, oncologic processes. We present a case of an adolescent male with persistent fever, abdominal pain, diarrhea and elevated serum inflammatory markers following an appendectomy who was ultimately diagnosed with lymphoma. A 17-year-old young man presented to an outside institution with one day of severe right lower quadrant pain. An appendectomy was performed due to high clinical suspicion for acute appendicitis, later confirmed by pathology. Difficulty mobilizing the thickened terminal ileum during the operation, a strong family history of IBD and stunted growth raised concern for IBD. On postoperative day two, he developed non-bloody diarrhea. Two days later, he developed diffuse abdominal pain and fever thought to be related to peritonitis. Antibiotic therapy was initiated; however, there was lack of clinical response. Stool infectious workup and computed tomography scan were nonrevealing. On further evaluation, noted to have elevated inflammatory markers with normal complete blood count, uric acid and lactate dehydrogenase. Despite broad spectrum antibiotics, high fevers and diarrhea persisted in association with continued elevation of inflammatory markers with signs of malabsorption. The patient was transferred to our center for further management. On admission patient was febrile with an unremarkable physical examination. A colonoscopy was deferred given proximity to appendectomy and a video capsule endoscopy was offered but refused by the patient. Three weeks into his illness, he developed an enlarged nontender cervical node and an acneiform rash that spread from face to trunk forming discrete erythematous papules prompting skin, lymph node and bone marrow biopsies which were consistent with Alk1+ anaplastic large cell lymphoma (ALCL). The patient decompensated and progressed to multi-organ failure with significant elevation of white blood cell count and increase in tumor lysis markers. The patient was started on combination chemotherapy as per Children's Oncology Group ANHL12P1. ALCL represents a fairly well-characterized group of T-cell lymphomas and comprises approximately 3% of adult and 10%-30% of childhood non-Hodgkin's lymphomas. Depending on the extent of a patient's disease burden, initial symptoms may range from isolated skin lesions to fulminant cardio-respiratory and hepatorenal failure. Large cell lymphomas are often associated with constipation, abdominal pain, masses or ascites. Additionally, multiple case reports describe an associated acute abdomen which may be mistaken for appendicitis. Once the diagnosis of ALCL is suspected, rapid initiation of treatment at a tertiary care facility is crucial as this aggressive lymphoma, if left untreated, can lead to massive cytokine storm and multi-system organ failure as exhibited by our patient. This case highlights the importance of considering a broad differential in a patient who presents with signs and symptoms of IBD. Careful consideration of differential diagnosis and the utilization of a multidisciplinary approach can be critical in accurate diagnosis.

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Introduction: Lymphocytic colitis (LC) is a type of microscopic colitis with colonic intra-epithelial lymphocytosis and normal endoscopic appearance. LC is more common in adults, particularly in elderly females with watery diarrhea. It is rarely seen in pediatrics. Although the etiology is unknown, it is associated with autoimmune disorders like celiac disease, Crohn's disease, Type 1 Diabetes Mellitus, autoimmune thyroiditis, CREST syndrome and immune deficiency disorders. Heindl et al is the only documented series where an association has been postulated between LC and thymic hyperplasia (TH). There have been no pediatric cases reported. We present the case of a 12 year old boy with lymphocytic colitis and benign TH. This case further supports the association of thymic hypertrophy with autoimmune conditions such as LC. We recommend that there should be a low threshold to evaluate for such conditions in the presence of extra-intestinal symptoms. Case Description: We report a 12 year old Caucasian male with microscopic colitis, chronic abdominal pain and constipation who returned with symptom recurrence after 1 year of remission on sub-therapeutic dose of mesalamine, 1gm daily, 12.5mg/kg/day. Physical exam showed soft abdomen, with mild tenderness to palpation in periumbilical area. Laboratory work up revealed a normal CBC, LFT and CRP but an elevated ESR. An abdominal sonogram revealed mild splenomegaly. No stool studies were obtained as patient did not have diarrhea. Colonoscopy was done to follow up on the previous findings of

microscopic colitis given his abdominal pain, elevated ESR and concern for progression to IBD as documented in the current LC literature. The colonoscopy showed grossly normal appearing colonic mucosa, with mild lymphonodularity in the descending colon. Biopsies showed intra-epithelial lymphocytosis in the entire colon, without architectural changes or collagen hypertrophy. Upper endoscopy was negative for celiac disease, peptic disease and H. pylori gastritis. The patient was started on a therapeutic dose of sulfasalazine based on the pediatric literature with plan to use budesonide if his symptoms did not improve. ASA therapy, in combination with laxatives for constipation, resulted in resolution of symptoms. A month later, the patient presented with a new onset chest pain. A chest X ray revealed an anterior mediastinal mass with mediastinal widening. Biopsies of this mass revealed benign TH. A full immunological work up, including Immunophenotypic and gene rearrangement studies, was negative for lymphoma. Other causes of thymic hyperplasia in children, such as prior exposure to corticosteroids and chemotherapy use were not present in this case. Conclusion: This report highlights the association of benign thymic hyperplasia with autoimmune conditions like lymphocytic colitis in the pediatric population. This case suggests that unlike adults, diarrhea might not be the presenting complaint in pediatric patients with LC. We recommend having a low threshold in screening for benign TH and other autoimmune conditions like celiac disease, Crohn's disease, Type 1 DM and autoimmune thyroiditis, in pediatric patients with LC and extra-intestinal symptoms.

383 METASTATIC CROHN'S DISEASE: NOT JUST A SIMPLE SKIN RASH. R. Sabe, V. Baez-Socorro, T.J. Sferra, Pediatric Gastroenterology, Hepatology & Nutrition, UH Rainbow Babies & Children's Hospital, Cleveland, Ohio, UNITED STATES. Garcia-Naveiro, Division of Pediatric Gastroenterology, Akron Children's Hospital, Akron, Ohio, UNITED STATES.

Introduction: Skin manifestations are the most common extra-intestinal manifestations of Crohn's disease (CD). They can be divided into CD-specific lesions (fissures, fistulae, metastatic CD), reactive conditions (erythema nodosum, pyoderma gangrenosum), associated conditions (vitiligo, palmoplantar pustulosis), nutritional deficiencies (glossitis, acrodermatitis enteropathica), and therapy-related lesions (drug hypersensitivity, toxic erythema, neutrophilic dermatoses). Metastatic CD is characterized by histological features of the disease occurring at sites not contiguous with intestinal granulomas. Case: An 11-year-old male who presented with scrotal and penile swelling was found subsequently to have CD. Ten months prior to the diagnosis of CD, he developed scrotal and penile erythema and edema following a cardiac catheterization for a cardiac arrhythmia. The cutaneous findings initially were attributed to irritation from tape placed during the catheterization and treated with over-the-counter topical medications. After an initial improvement, the edema persisted prompting a dermatology evaluation. A scrotal skin biopsy demonstrated edema of the dermis and numerous foci of non-necrotizing granulomatous inflammation with histiocytes and multi-nucleated giant cells. He was prescribed oral steroids. The cutaneous manifestations improved but did not resolve. Because of pain with bowel movements and occasional rectal bleeding that improved with the steroid therapy, he was referred to Pediatric Gastroenterology. Laboratory testing revealed anemia, hypoalbuminemia, and elevated inflammatory markers. He underwent esophagogastroduodenoscopy and colonoscopy revealing mild-to-moderate, patchy, active chronic inflammation of the esophagus, stomach, duodenum, and right colon. There were no granulomas. He was treated initially with steroids and subsequently with azathioprine and infliximab. His gastrointestinal symptoms resolved and there was improvement but not complete resolution of the genital edema. Discussion: Metastatic CD is a rare complication and the least common skin manifestation of CD. In pediatric patients, it usually precedes the diagnosis of CD. Metastatic CD can involve genital and extra-genital areas of the body. Extra-genital sites include upper and lower extremities, intertiginous areas, trunk, abdomen, and rarely the face. It can present as edema, induration, papules, nodules, plaques, abscesses, or ulcers. The differential diagnosis of metastatic CD of the genitalia includes infections, sarcoidosis, contact dermatitis, acquired lymphangiectasia, vascular malformations, sarcoidosis, and sexual abuse. Skin biopsy should be performed when there is a high suspicion for CD. Treatment for metastatic CD include steroids (topical, systemic, intra-lesional), antibiotics (metronidazole, tetracycline), anti-inflammatories (5-aminosalicylic acid), immunomodulators (6-mercaptopurine, azathioprine), anti-tumor necrosis factor α -antibody, oral zinc supplements, thalidomide, hyperbaric oxygen, and surgical excision. Metastatic CD should be suspected in patients with genital rashes, especially if there are associated gastrointestinal symptoms.

384 AUTOIMMUNE ENTEROPATHY AND HEPATITIS IN PEDIATRIC HEART TRANSPLANT RECIPIENT. J. Quiros, Pediatric Gi & Nutrition, MUSC Children's Hospital, Charleston, South Carolina, UNITED STATES. Butts, A.

Savage, Pediatric Cardiology, MUSC Childrens Hospital, Charleston, South Carolina, UNITED STATES. Hudspeth, Bone Marrow Transplant Program, MUSC Childrens Hospital, Charleston, South Carolina, UNITED STATES. Background: Autoimmune enteropathy is a rare disorder in children, which presents with severe diarrhea and malabsorption, caused by immune mediated damage to intestinal mucosa. Autoimmune enteropathy has been associated with immunodeficiencies as well as dysfunction of T-cell regulators (Tregs). Clinical Course: Our patient was transplanted at 8 months of age (January 2006) after her hemi-fontan operation for hypoplastic left heart syndrome was complicated by prolonged ECMO support. Since her transplant, she has had a multiple rounds of antibiotics for pulmonary infections. In fall of 2011, the patient began experiencing intestinal discomfort, weight loss, anorexia malabsorption, alopecia, and elevations in transaminases. In February 2013, she was admitted with pancreatitis, fluid overload and acute on chronic renal failure precipitating a need for mechanical ventilation. In a subsequent admission for continued gastrointestinal symptoms, an EGD was performed finding significant and diffuse inflammation in the duodenum. Biopsy of the affected areas showed a non-lymphomatous lymphocytic infiltrate by histology and flow cytometry. FoxP3 staining showed only a few scattered Tregs in the infiltrate. Her serum anti-enterocyte antibody returned positive. Prednisone was started and after a twelve week taper she was switched to 6-MP. Her anorexia resolved after high dose steroids and her alopecia has improved. She is currently thriving. Follow-up EGD done 6 weeks after starting prednisone showed improvement of the previously inflamed areas and no new lesions. Conclusion: Autoimmune enteropathy is a rare disorder that can cause significant morbidity. The patient underwent a thymectomy during her infant surgery for congenital heart disease which is associated with decreased circulating T-cells and disturbed T-cell homeostasis, including Tregs function. Repeated exposures to antibiotics has been associated with decreased Tregs in animal models. These abnormalities combined with her immunosuppression due to transplantation, are possible risk factors for her developing autoimmune enteropathy. Pediatric heart transplant recipients with infant cardiac surgery and frequent antibiotic use may be at risk to develop autoimmune enteropathy due to thymectomy and long-term immunosuppression.

ENDOSCOPY/POTPOURRI

395 "GASTRIC OUTLET" PATCH: AN UNUSUAL CAUSE OF RECTAL BLEEDING IN A CHILD". A.K. Williamson, Pathology, NSLIJ Health System, New Hyde Park, New York, UNITED STATES. D. Haller, M. Greifer, Pediatrics, Cohen Children's Medical Center, NSLIJ Health System, New Hyde Park, New York, UNITED STATES. A 5 year-old boy presented to the outpatient pediatric gastroenterology division with intermittent bright red rectal bleeding of one year's duration. He denied constipation, abdominal pain, and diarrhea, and had been growing appropriately along the 75th percentile for both weight and height. His past medical history was significant for constipation as a toddler, for which he had taken Miralax, and family history revealed a maternal grandfather recently diagnosed with colon cancer. A review of systems and physical examination were unremarkable. Bloodwork, including a complete blood count, erythrocyte sedimentation rate, C-reactive protein, and comprehensive metabolic panel, were normal. Despite the use of Miralax over the next few months, he continued to bleed, and a colonoscopy was performed. While the majority of the colon appeared grossly normal, a nodular and friable area of the rectal mucosa was partially visualized immediately proximal to the anal verge as the colonoscope was being withdrawn. Biopsies from that site revealed the presence of non-ulcerated oxyntic gastric mucosa. The presence of *Helicobacter pylori* was not identified. This finding, in addition to the presence of persistent rectal bleeding, prompted further evaluation. An examination-under-anesthesia revealed a 2cm by 3cm serpiginous lesion with the appearance of gastric rugae about 2 cm proximal to the dentate line. The lesion was removed by electrocautery, and the presence of non-ulcerated gastric oxyntic mucosa was confirmed histologically. There have not been any further reports of bleeding since the lesion was removed eighteen months ago. While painless rectal bleeding in an otherwise healthy and thriving child with a normal defecatory pattern is a relatively common indication for evaluation by a pediatric gastroenterologist, the finding of an etiology other than an anal fissure or a polyp is atypical. Gastric heterotopia in the rectum is an unusual cause of rectal bleeding. Ectopic gastric tissue has been recognized in all areas of the digestive tract, most commonly in the esophagus, as well as the biliary tract and gallbladder. The presence of multiple colonic foci have been reported simultaneously in a single patient, and in several cases gastric heterotopia has been found in association with other congenital anomalies. Generally, this displaced gastric mucosa is associated with painless bleeding, but it has also been accompanied by more significant symptoms, such as ulceration, bowel perforation and severe gastrointestinal bleeding. Its etiology has been speculated to be due to either improper regeneration of damaged tissue or

abnormal differentiation in pluripotential embryonic endoderm. Analysis of biopsy specimens typically reveals gastric fundic-type mucosa, and *Helicobacter pylori* infection has been reported. There have been reports of successful resolution of rectal bleeding with the use of H₂-receptor antagonists or proton-pump inhibitors. While malignant transformation has not been identified in rectal lesions, most reported have been locally excised. As pediatric gastroenterologists, we must consider this entity in the differential diagnosis of painless rectal bleeding in children.

396 HOOKWORM INFESTATION: AN UNUSUAL CAUSE OF OBSCURE GASTROINTESTINAL BLEEDING. M. Middelhof, V. Sood, T. ROSSI, R. Abell, Pediatric Gastroenterology and Nutrition, University of Rochester, Rochester, New York, UNITED STATES.

We report a 10 year old male patient with hookworm infestation leading to significant anemia. The patient first presented with hematemesis and a Hct of 24 %. Upper endoscopy demonstrated 2 gastric ulcers but pathology was unremarkable. Patient was treated with a Proton Pump Inhibitor and Carafate and clinically did well. Two months later, the patient again presented, but now with melena and abdominal pain. At this presentation, his Hct was 35% , Platelets were 456 Thou/uL, and MCV was 80fL. Fecal occult blood was positive, but all stool testes were negative, including those for O&P. Repeat upper endoscopy was significant for two antral ulcers. Colonoscopy as well as capsule endoscopy revealed erythema and multiple hookworms in the duodenum and proximal jejunum. The patient was treated with albendazole with resolution of symptoms. Hookworms, including, *Necator americanus* and *Ancylostoma duodenale* infect around 740 million people worldwide and are more widely distributed in Asia and Africa but can also be found in other areas. Gastrointestinal symptoms typically include mild abdominal pain, nausea, vomiting, and anorexia. Hookworm-associated blood loss results from the destruction of capillaries in the intestinal mucosa. The degree of anemia is typically related to the hookworm burden. Internationally, hookworms are an important cause of obscure occult gastrointestinal bleeding and iron-deficiency anemia, but are rare in the United States. Most patients with hookworm infection are asymptomatic or have mild degrees of anemia. Overt gastrointestinal bleeding caused by hookworms as seen in our patient in this country is very rare.

397 AN EXTREMELY RARE TYPE OF GASTRIC ADENOCARCINOMA IN A TEENAGE MALE. D.A. Moya , W.L. Taylor, M. Ammar, C. Cuevas, Y. Al-Tawil, Pediatric Gastroenterology, GI for Kids, PLLC - East Tennessee Children's Hospital , Knoxville, Tennessee, UNITED STATES. S. Corns, S.E. Spiller, Pediatric Hematology/Oncology, East Tennessee Children's Hospital , Knoxville, Tennessee, UNITED STATES.

Gastric adenocarcinoma (GAC) is an extremely rare cancer in children. We present a case of a 15-year-old teenage male, previously healthy, who was diagnosed with a poorly differentiated metastatic signet-ring cell gastric adenocarcinoma. Family history was negative for hereditary gastric and colorectal malignancies. He presented with a 3 week history of abdominal pain and intermittent fever. Two weeks later, he developed abdominal distention and was treated for constipation. Due to persistent abdominal distention, an abdominal CT scan was performed and revealed massive ascites and left pleural effusion with suspected omental caking of carcinomatosis. Macroscopically, upper endoscopy showed esophagitis, nodular gastritis, and a large gastric mass in the fundus invading the gastric wall. Colonoscopy showed coffee ground material in the right colon and nodular changes throughout the colon. Esophageal biopsies showed *Candida* esophagitis, and gastric biopsies confirmed *Helicobacter pylori* gastritis, and an invasive signet-ring cell adenocarcinoma. Colonic biopsies were normal. Abdominal positron emission tomography (PET) scan was consistent with metastatic disease. Paracentesis confirmed signet-cell gastric adenocarcinoma. Additionally, pleural fluid yielded signet-ring cells. Gene sequence analysis of APC, MUTYH, CDH1, TP53, BMPR1A and SMAD4 mutations were negative. Human epidermal growth factor receptor 2 (HER-2/neu) overexpression was negative by fluorescence in situ hybridization (FISH) and immunohistochemical analysis. The patient received neoadjuvant chemotherapy. He was referred to a tertiary cancer center for possible cytoreductive surgery of peritoneal disease followed by hyperthermic intraperitoneal chemotherapy (HIPEC). Discussion: Data is limited in regards of clinical presentation, treatment and outcomes in pediatric gastric adenocarcinoma. It is usually a malignant disease with ominous prognosis. Family history of gastric or colorectal carcinoma and *Helicobacter pylori* infection with the *vasAs1-*, *vacAm1-*, and *cagA*-positive genotypes, have been linked to GAC. Germ line mutations have been reported in patients with hereditary diffuse gastric cancer. Our patient did not have polyposis or a prior history of lymphoma. Additionally, he tested negative for germ line mutations, specifically CDH1, thus it was likely a de novo occurrence adenocarcinoma. For children

affected with GAC, radical surgery is an essential treatment. Perioperative chemotherapy may improve the prognosis of this fatal condition. Therapeutic regimens in children are based on adult oncology experience and remain a significant challenge. Large multi-institutional studies are not possible given the infrequent occurrence of gastric adenocarcinoma in children.

398 MULTIPLE POLYPS - UNUSUAL PRESENTATION OF SOLITARY RECTAL ULCERATIVE SYNDROME IN HOMOSEXUAL TEENAGE PATIENT. D. Hong, A. Malkani, S. Blanchard, Pediatric Gastroenterology , University of Maryland, Baltimore, Maryland, UNITED STATES.

We report 18 year old Caucasian male who presented with painless hematochezia. He had history of genotype 3 hepatitis C infection which was treated 6 years ago and he achieved sustained viral response. There was no constipation, diarrhea, weight loss or any other constitutional symptoms. He was on multiple psychiatric medications for depression. He denied any digital rectal manipulation. He admitted that he preferred same sex partners. He had normal complete blood count and PT/PTT. He had a normal physical exam with no perianal lesions. He underwent colonoscopy which showed multiple sessile polyps and nodules circumferentially in rectum without signs of bleeding. These were all biopsied. The histopathology showed rectal mucosa with distorted and disorganized glands, mucinophages and thickened muscularis mucosa with mild fibrosis. These findings were consistent with solitary rectal ulcer syndrome (SRUS). The pathology was negative for human papillomavirus and other infectious work up process. At his follow up visit, he was asymptomatic as there had been no rectal intercourse since the procedure. SRUS in pediatrics is rare and often related to prolonged excessive straining due to abnormal defecation. It can present with multiple ulcers or a single ulcer, hyperemic mucosa or broad based polyps in the anterolateral surface of rectum. Anal intercourse is a risk factor for chronic mucosal trauma leading to the development of circumferential lesions. In pediatrics, we should recognize this presentation of SRUS in the spectrum of anorectal disorders of homosexual teenagers.

399 ESOPHAGEAL DIVERTICULUM AND HIGH-GRADE ESOPHAGEAL STRICTURE RESPONSIVE TO SERIAL BALLOON DILATION AND NUTRITIONAL THERAPY. K. Law, USF, Tampa, Florida, UNITED STATES. K. Law, All Children's Hospital, Tampa, Florida, UNITED STATES.

Patient is a 16 year old boy with recessive dystrophic epidermolysis bullosa and previous history of esophageal strictures, presenting with one day of dysphagia and hematemesis. He was hemodynamically stable, and gastroenterology was consulted. On day one of admission, endoscopy revealed esophagitis, erythema, and friability of the mucosa of the esophagus. At levels 25 and 27 centimeters there were blood clots attached to the esophageal lumen. A tight esophageal stricture was noted at 35cm, which the Olympus GIF-XP 160 endoscope could not pass. Active bleeding was found at the time, and dilation was held off secondary to the presence of marked esophagitis. Patient tolerated procedure well and started on carafate four times daily to alleviate the discomfort from the noted esophagitis. Overnight patient continued to have epigastric pain. He underwent a second endoscopy two days later. Endoscopy revealed improved esophagitis and no further blood clots. A bifurcation appeared at level 35 cm from the incisors, which was not present on the previous day's endoscopy. Instilling a small amount of water revealed a blind ended sac on the left and a tract passing into the stomach on the right. A CRE™ Wireguided balloon dilator was inserted into the true lumen and expanded to 6 millimeters and then to 7 millimeters. At 8 millimeters, significant resistance was felt, and dilator removed. Upon removal, the true tract leading to the stomach was more dilated than the false tract. He underwent a third endoscopy on his fifth day of admission. Esophagitis was improved. The bifurcation was still present with each lumen appearing the same diameter. Balloon dilator was inserted and dilated to 8 millimeters, 9 millimeters, and 10 millimeters. The fourth endoscopy was performed on his seventh day of admission. The true lumen appeared slightly wider than the false tract. Balloon dilator was inserted and inflated to 10 millimeters and then to 12 millimeters. Some pressure was felt, and there was a small amount of bleeding at dilation site. On hospital day ten, he had his next endoscopy. Balloon dilator was inserted into the true lumen and dilated from 12 millimeters to 13 and then to 15 millimeters. A follow up esophagram was performed that same day revealing the notable esophageal diverticulum and a small amount of mediastinal air, suggesting microscopic esophageal perforation. Patient had thus far been asymptomatic without any history of fever or tachypnea, and patient was treated conservatively with intravenous antibiotics. He was advanced to a clear liquid diet by day eleven of admission and advanced to full liquids by day thirteen of admission, which he tolerated well. Gastrostomy tube was placed on admission day fourteen secondary to patient's malnutrition. In addition to receiving gastrostomy tube feeds, he eventually tolerated a soft diet prior

to his discharge home on day eighteen. He was seen in clinic eight days after discharge without any dysphagia, hematemesis, and tolerating his soft diet well. Three months later endoscopy revealed no residual esophageal stricture and a complete resolution of the esophageal diverticulum.

400 DYSPHAGIA AFTER H-TYPE TRACHEOESOPHAGEAL FISTULA REPAIR. K. Suryawala, C. Marshall, Pediatric Gastroenterology, Oklahoma University- Children's Hospital, Oklahoma City, Oklahoma, UNITED STATES. Introduction: Tracheoesophageal fistula (TEF) without esophageal atresia (H-type TEF) is a rare congenital anomaly and accounts for 4-5% of all congenital tracheoesophageal malformations. Diagnosis can be difficult, but once diagnosed and surgically corrected, it is unusual to have esophageal symptoms or difficulty swallowing. Case report: A term male infant born with birth weight of 2835 gm was discharged home on second day of life. On day three of life, he experienced choking and gagging associated with perioral cyanosis after feeding. These episodes gradually increased occurring with every feed lasting about 20 seconds. On five days of age, patient was admitted for progressively worsening symptoms. The physical examination was unremarkable. Complete blood count, coagulation profiles and blood chemistry were within normal range. Echoencephalogram and echocardiogram were unremarkable. An upper gastrointestinal series with dysphagiagram showed contrast flowing into trachea via TEF. Patient underwent rigid bronchoscopy and TEF repair through cervical approach at two weeks of age. Repeat esophagram five days after surgery showed no evidence of TEF but did reveal a fixed esophageal narrowed segment in the mid esophagus without proximal dilation of esophagus. After TEF repair, patient improved and was discharged home on postoperative day seven. He did well with liquids and soft foods but usually refused to eat solid foods as his diet was advanced based on age. At sixteen months of age he was admitted to the hospital with vomiting and dehydration. An esophagram revealed delayed clearing of contrast due to moderate narrowing in the mid esophagus as well as a filling defect suggesting a food impaction. Esophagoscopy revealed food impaction in mid-esophagus. Dilation was performed with Savory dilators. Despite dilation, patient continues to have dysphagia and vomiting with solid foods. Discussion: We describe a patient with H-TEF and esophageal stenosis. Congenital esophageal stenosis (CES) is a rare anomaly with estimated incidence of 1:25,000 to 50,000 live births. The association of CES in children with TEF is very unusual. The association in this patient highlights importance of considering CES in any patient presenting with dysphagia and/or food impaction. Close attention should also be paid to contrast studies which may reveal CES as described in our case.

401 SEVERE ESOPHAGEAL ULCERATION SECONDARY TO PILL ESOPHAGITIS IN THE SETTING OF EOSINOPHILIC ESOPHAGITIS (EOE).. E. Grossman, Internal Medicine, Division of Digestive Disease, SUNY Downstate Hospital, Brooklyn, New York, UNITED STATES. Saghier, F. Windemuller, J. Xu, S. Schwarz, S.S. Rabinowitz, Pediatric Gastroenterology, SUNY Downstate Medical Center, Downstate Children's Hospital, Brooklyn, New York, UNITED STATES. Gupta, Pathology, SUNY Downstate Hospital, Brooklyn, New York, UNITED STATES. Purpose: To demonstrate that EOE should be considered and evaluated with appropriate biopsies in the setting of unusually large or deep esophageal ulcerations, irrespective of the presumptive etiology. Methods: Case report: 14 year old boy was referred to Pediatric GI clinic after an ER visit for odynophagia and dysphagia of uncertain etiology. The patient's symptoms began shortly after the initiation of doxycycline for acne. He reported a 5 lbs weight loss related to decrease oral intake. He denied any emesis, regurgitation, heartburn, choking, or coughing. Upper endoscopy (EGD) revealed mild trachealization proximal to a long, deep mid-esophageal ulcer (estimated to be 7cm x 0.7cm) that extended into the distal third of the esophagus without esophageal narrowing. Multiple biopsies of the base and the ulcer rim were obtained (for histology and culture). Additional biopsies were taken from non-ulcerated mid and distal esophageal mucosa. Serum was also sent for viral serology. Results: Biopsies from the ulcer revealed extensive acute and chronic inflammation with exudates without any evidence of inclusion bodies. The mid and distal esophagus both had >80eos/hpf, consistent with EOE. Stains for Herpes, CMV and fungal organisms, viral cultures and viral serology were negative. The patient was started on a proton pump inhibitor (PPI) and a restrictive diet (based on allergy testing) with full resolution of his symptoms and improved weight gain. EGD was repeated at 6 weeks and showed substantial improvement with almost full resolution of the ulcer. Repeat biopsies (on PPI) from non-ulcerated mid and distal esophagus were improved but permitted a definitive diagnosis of EOE. Conclusion: Esophageal dysmotility secondary to EOE can exacerbate the damage of doxycycline related pill esophagitis. Uncharacteristically large and deep esophageal ulcer(s) should prompt the endoscopist to consider co-morbidities that could potentially worsen pill esophagitis. Clinicians should routinely provide anticipatory guidance to patients given medications associated with pill esophagitis.

TEETH STRAIGHT, STOMACH IN KNOTS. A.K. Hersch, G. Yanni, M. Klooster, Pediatrics, Loma Linda University Children's Hospital, Irvine, California, UNITED STATES. C. Flannery, M. Oman, Adult Gastroenterology, Loma Linda University Medical Center, Loma Linda, California, UNITED STATES. G. Gollin, Pediatric Surgery, Loma Linda University Medical Center, Loma Linda, California, UNITED STATES.

Background Foreign body (FB) ingestion is a commonly encountered problem in pediatric medicine, affecting approximately 80,000 children each year. Although many FB ingestions are asymptomatic, symptoms can range from dysphagia and abdominal pain to shock and even death. In fact, it has been reported that 1,500 people die annually from FB ingestion in the US. Once FB's enter in the stomach, up to 90% will pass without complications, 10-20% will require endoscopic intervention, and 1% will require surgical removal. Unintentional ingestion of orthodontic appliances is a rare type of FB ingestion, which can present with serious complications. These appliances are often sharp and pointed and it has been reported that objects of this nature account for 1/3 of perforations from FB and perforate in 15-35% of patients. Archwires are metallic and pointed orthodontic appliances which have risk of perforation if ingested. Case description A fifteen-year-old female presented to the Emergency Room with a one-day history of constant, severe, stabbing, non-radiating epigastric and right upper quadrant abdominal pain. Past medical history was negative with the exception of dental braces. Labs were within normal limits, but imaging demonstrated a FB in the distal stomach. The patient denied intentional or unintentional swallowing of a FB.

Esophagogastroduodenoscopy was done and a submucosal curvilinear raised structure was visualized in the stomach antrum. Endoscopic ultrasound demonstrated a submucosal FB with penetration through the muscularis propria layer. In an attempt to remove the FB, tunnel biopsies were done to grasp the object but were unsuccessful. Surgical intervention removed the FB, which was consistent with a 1.5 cm dental archwire. Further patient history revealed dental braces removed one year earlier, without any memory of swallowing an appliance. She recalled that the archwires were changed nearly monthly and may have unknowingly swallowed the archwire. Discussion Unintentional ingestion of orthodontic appliances is rare and can present with serious complications. Unusual in this particular case is that an archwire unintentionally ingested one to three years ago became an acute medical emergency requiring surgical intervention. Lee reported a 15-year-old boy swallowed his archwire while eating a bread roll and Umesan reported a 12-year-old girl swallowed her archwire while having an orthodontic adjustment. Both cases were recognized right away and FB removed without complications. In this case the patient was never aware she had swallowed her archwire and it had completely migrated through the gastric mucosa into the submucosa and into the omentum. Fortunately her diagnosis was not further delayed resulting in possible complications such as peritonitis, sepsis, shock, and death, all known complications of FB perforation. The gastrointestinal tract is generally very resistant to perforation or obstruction following FB ingestion, with the exception of sharp, pointed, and long (> 5 cm) objects."

COLONIC ANGIOECTASIA IN A 13 YEAR OLD ADOLESCENT BOY WITH DYSKERATOSIS CONGENITA ON LONG-TERM ANABOLIC STEROID THERAPY.. E. Tibesar, M.N. Weidner, P. Okolo, C. Cuffari, Johns Hopkins, Baltimore, Maryland, UNITED STATES.

Dyskeratosis congenita (DC) is a rare, progressive congenital disorder caused by mutations in the telomerase gene. Patients will present clinically with a triad of finger and toenail dystrophy, changes in skin pigmentation and oral leukoplakia. The disease can also cause life threatening bone marrow dysfunction that can progress to either myelodysplastic syndrome or leukemia. Although DC can involve the intestinal mucosa, it is not commonly associated with gastrointestinal bleeding. Herein, we present a case of a 13 year old male with Hoyeraal Hreidarsson syndrome, a severe form of DC, who was admitted to the hospital after three separate bouts of painless, bright red rectal bleeding that required transfusions. At the time of his initial presentation, he had been on long-term anabolic steroid therapy for his pancytopenia. An infectious work up was negative. A flexible sigmoidoscopy revealed two small discrete ulcers at the splenic flexure that were not actively bleeding. No intervention was taken at the time of the procedure. Colonic biopsies were negative for either infection or inflammation. At the time of his second episode, he was switched to an amino acid formula based on the presumption of a possible allergic etiology. One week later, he had his third large volume bout of hematochezia ultimately prompting a referral to The Johns Hopkins Hospital for further evaluation. At the time, his platelet count was 22,000/mm³ (normal 150,000-300,000/mm³), hemoglobin 8.1 g/dL (normal 12.4-14.8 g/dL) and a total white blood cell count was 720/mm³ (normal 4500-13,500/mm³). During that evaluation, he also underwent a full

colonoscopy that revealed diffuse angioectasia within the cecum and ascending colon, with five discrete lesions measuring up to 3 cm in diameter. The patient was successfully treated by argon plasma coagulation and placement of hemoclips on the larger lesions. He tolerated the procedure well and has had no repeat episodes of lower GI bleeding. This patient is the first reported case of an adolescent boy with DC and colonic angioectasia presumably due to long-term anabolic steroid use. Although there is an association between cutaneous telangiectasias and long-term anabolic steroid therapy, the gastrointestinal tract is generally not affected. Moreover, vascular lesions of the lower GI tract are rare in pediatrics. We believe that the combination of the child's underlying multi-visceral mucosal disorder and long-term anabolic steroid use led to enhanced fragility of the vascular endothelium and thus angioectasia formation. This case is instructive on the potential risk of angioectasia formation with long-term anabolic steroid use, especially among patients with underlying mucosal disorders. This case also underscores the use of argon laser therapy to treat vascular lesions of the gastrointestinal tract.

404 MECKEL'S DIVERTICULITIS IN AN ADOLESCENT WITH PRIOR HISTORY OF LOWER GASTROINTESTINAL BLEED. H. Al-Atrash, V. Uppal, E. Kutsch, Z. Molle Rios, Nemours du Pont Hospital, Wilmington, Delaware, UNITED STATES.

Case: A 17 yr old male presented with sudden onset of severe lower abdominal pain. One year prior to his presentation he developed unexplained lower gastrointestinal (GI) bleeding with hemodynamic instability and hemoglobin drop from 11.3 to 9.4 gram/dl. His prior workup included a negative Meckel's scan, a normal upper endoscopy/colonoscopy and a normal capsule endoscopy. His bleeding stopped spontaneously and his hemoglobin stabilized. In an attempt to establish a diagnosis, he had another Meckel's scan that was negative. He also had a negative computed tomography (CT) scan of the abdomen with angiography to evaluate for vascular malformations. On his most recent presentation, he experienced sudden onset of severe lower abdominal pain and tenderness on exam. An abdominal CT scan was positive for new ovoid structure measuring 5 X 4.7 cm close to distal ileum suggestive of diverticulitis, which was not seen on prior scans. He underwent surgical exploration and an inflamed and gangrenous was resected. His final pathology confirmed a diagnosis of Meckel's diverticulitis. He did well in his post operative course without further bleeding or pain. Discussion: Meckel's diverticulum is a frequently reported congenital anomaly of the GI tract. It typically occurs in the terminal ileum proximal to ileocecal valve. In most cases, it is asymptomatic. However, some patients may develop painless rectal bleeding, intestinal obstruction, lymphoma, chronic peptic ulceration or Meckel's diverticulitis. Meckel's diverticulitis accounts for 10-20 % of complications related to Meckel's diverticulum and it is more commonly seen in older patients. It usually presents with symptoms similar to an acute appendicitis and failure to establish the diagnosis may lead to perforation, peritonitis and death. Diagnosis of Meckel's diverticulum is often difficult, requiring high index of suspicion, and it can be easily missed. A Technetium-99m (Meckel's scan) is the most common noninvasive form of testing for Meckel's diverticulum but it has a relatively low sensitivity of 80-90% in children and 62.5% in adults. This might account for why our patient had a negative Meckel's scan at initial presentation. Meckel's diverticulum may also be diagnosed by abdominal CT scan, magnetic resonance imaging (MRI) enterography or incidental finding during laparoscopic procedure. Surgical resection, even in asymptomatic patients, is recommended due to high risk of complications.

405 ESOPHAGEOGASTRIC FISTULA: A RARE COMPLICATION OF FUNDOPLICATION. N. Ibrahim, Pediatrics, KUMC, Lenexa, Kansas, UNITED STATES. N. Ibrahim, Gastroenterology, CMH, Kansas City, Missouri, UNITED STATES.

A 13-year-old boy presented to our clinic the first time with epigastric pain, vomiting and dysphagia for 2 years. Patient had a history of Thal fundoplication at 2 months of age for gastroesophageal reflux. Physical exam and laboratory studies were normal. Patient had a normal upper GI series and failed a trial of antacid. An upper endoscopy was then performed and showed a 5 cm long cord like object in the distal esophagus that was thought to be a retained suture. There was surrounding erosions and ulcerations. The object was retrieved without complications. Surgery report for fundoplication was reviewed and reported using a 3-0 Gore-Tex suture to secure the anterior body and fundus of the stomach to the esophagus anteriorly. Patient had resolution of symptoms and lost follow up. 2 years later, he presented again with reflux like symptoms, burning sensation in the chest, trouble swallowing, and sensation of food sticking in his throat. He was limiting his foods to milk and liquids and was avoiding tomato based and greasy foods. Patient underwent a repeat upper endoscopy that showed severe

erosive esophagitis and a fistula formation between distal esophagus and gastric fundus. A CT scan and an esophageogram were performed and confirmed the diagnosis of esophageogastric fistula. Esophageogastric fistula (EG fistula) is a very rare complication, with only 12 previous cases reported in the past. Almost half of the reported cases of EG fistulas had history of fundoplication (Laparoscopic fundoplication in all cases) and most of the patients had history of prolonged gastroesophageal reflux disease. To our knowledge, this is the first case to be reported in a pediatric patient and the first case to be reported after an open fundoplication. The exact mechanism of EG fistula formation is still not well understood because of the rarity of the cases. It is thought that intra-operative subclinical ischemia and necrosis from a tight or deeply placed suture, microscopic perforations from repeated manipulation of the esophagus and endothelial necrosis from esophagitis trigger the formation of the fistulous tracts. There are no established guidelines for management of EG fistulas. Medical management with acid suppression is a reasonable initial approach. Endoscopic clipping has been reported to have high recurrence rate, especially in chronic cases, thus surgical management remains the best permanent option. EG fistula, although rare, should always be suspected in patients with prolonged gastroesophageal reflux symptoms and history of esophageal manipulation, especially fundoplication.

406 DRAMATIC MUCOSAL AND HISTOLOGIC RESPONSE TO INTRALESIONAL CORTICOSTEROID INJECTION FOR SEVERE STRICTURING EOSINOPHILIC ESOPHAGITIS: A CASE REPORT. K. Kovacic, R. Noel, F. Ashai-Khan, D.G. Lerner, Division of Pediatric Gastroenterology, Hepatology and Nutrition, Department of Pediatrics, Medical College of Wisconsin, Milwaukee, Wisconsin, UNITED STATES. Szabo, Division of Pathology, Department of Pediatrics, Medical College of Wisconsin, Milwaukee, Wisconsin, UNITED STATES.

Introduction: Intralesional corticosteroid injection is an effective therapy for esophageal strictures of diverse etiologies. In conjunction with endoscopic dilation, this treatment option is usually reserved for complex strictures due to severe peptic, caustic or radiation injury. Stricture recurrence is common, prompting repeated dilations with risk of complications such as esophageal perforation. Eosinophilic esophagitis (EoE) can present with focal stenosis unresponsive to medical or dietary treatment. We report a case of dramatic response to intrastricture corticosteroid injection in an adolescent with severe EoE. Case: A 17 year-old male with asthma, peanut allergy and dysphagia with solids presented with meat impaction requiring endoscopic removal. Numerous prior food impactions were resolved by vomiting. An 8 cm-long stricture was noted in the mid-esophagus with an esophageal luminal diameter of 7 mm. Histology confirmed EoE, with eosinophilic micro-abscesses, basal cell hyperplasia, intraepithelial edema and up to 70 eosinophils/hpf. There was no improvement with acid suppression and swallowed fluticasone over three months. We performed CRE balloon dilations q month x 4 with successive increases in diameters by increments of 2mm or until evidence of esophageal tear/bleeding. There was minimal improvement after the first dilation. During the second dilation, we injected the narrowest stricture part with triamcinolone, 10 mg in 0.5 ml aliquots, in four quadrants using a 25-gauge sclerotherapy needle. A month later, the mucosa within 3-5 cm of the injections appeared normal endoscopically. The non-injected mucosa continued to manifest typical EoE appearance. Due to clinical improvement, injections were continued during three consecutive balloon dilations in different areas along the length of the stricture. Three months after the first injection, the injected areas show complete resolution of EoE endoscopically and histologically, with a dramatic transition to EoE distally to the injected mucosa. There were no procedure-related complications. Discussion: Injection of corticosteroids into strictures due to EoE may enhance healing and accelerate stricture resolution. Chronic esophageal inflammation results in collagen deposition and formation of scar tissue. Although mechanisms are unclear, corticosteroids may impede collagen deposition and scar formation by preventing cross-linking of collagen. Our findings add to existing literature, which suggests that tissue remodeling is reversible in young patients with EoE. Intralesional corticosteroids with dilations may improve outcomes and decrease the need for further dilations in treatment-refractory focal esophageal stenosis. Prospective studies are needed to investigate this therapy for stricturing EoE.

407 CONCENTRATED HYDROGEN PEROXIDE INGESTION - ROLE OF ADVANCED IMAGING IN MANAGEMENT. D. Hong, S. Zawahir, S. Blanchard, R. Watkins, Pediatric Gastroenterology, University of Maryland, Baltimore, Maryland, UNITED STATES.

A 2 year old Ghanian boy presented with hematemesis, coughing and foaming at the mouth immediately after drinking a solution of 35% hydrogen peroxide mixed with water. He did not complain of mouth or chest pain. His physical exam was benign and labs revealed a mild metabolic acidosis without an elevated anion gap. CT of the

abdomen and pelvis showed two discrete subcentimeter peripheral lucencies in the hepatic dome, which was consistent with portal venous air. The esophagogastroduodenoscopy revealed diffuse, severely erythematous mucosa without bleeding throughout the stomach. Histopathology revealed chronic inflammation, congestion and mild reactive changes. Caustic ingestions are a health hazard despite educational programs, proper labeling and legislation to limit the strength and availability of substances. This patient ingested the hydrogen peroxide as part of a naturopathic health maintenance regimen. This report aims to recognize the importance of imaging as part of the work up to assess portal venous gas along with endoscopic studies. If a patient has ingested a concentrated hydrogen peroxide solution (>10%) or a substantial amount of 3% solution, chest and abdominal imaging from x-rays or CT should be obtained to detect perforation from mechanical distention or to detect portal venous gas. A CT or MRI of the brain may also need to be considered if neurologic symptoms arise.

408 LOW PROFILE SINGLE STEP GASTROSTOMY TUBE PLACEMENT-CASE SERIES OF THREE-YEAR SINGLE CENTER EXPERIENCE.. A.K. Hunter, A. Sicolo, R. Gugig, Children's Hospital Central California, Clovis, California, UNITED STATES.A.K. Hunter, C. Huang, A. Sicolo, R. Gugig, UCSF-Fresno, Fresno, California, UNITED STATES.

Objective: Many approaches are currently available for supplemental nutrition in children from Percutaneous Endoscopic Gastrostomy (PEG) to surgically placed gastrostomy. Here we present a case series of patients who underwent endoscopic MIC-KEY G Introducer Kit placement with a primary low profile balloon gastrostomy tube placement. This procedure utilizes T fasteners for gastropexy with serial dilator used to create an opening for a Mic-Key button. Three doses of antibiotics are provided. Methods: Retrospective chart review of patients who underwent a MIC-KEY G Introducer Kit placement at our institution between 2011 and 2014 was performed. Results: A total of 28 patients were scheduled for a direct gastrostomy placement. Age of the subjects ranged between 9 month and 16 years old. Lowest weight reported was 7.43 kg. Indications included: failure to thrive and feeding difficulty due to congenital heart disease, cerebral palsy, traumatic brain injury, aspiration as well as enteral therapy for Crohn disease and enteral nutrition for epidermolysis bullosa with protein losing enteropathy. All patients underwent an Upper GI study and all had normal anatomy. All patients received nasogastric feeds prior to gastrostomy tube placement. In five patients (17.7%) placement of a primary Mic-Key tube was not possible due to lack of a safe window for placement of T fasteners. Four of those patients underwent PEG placement instead. One patient's body habitus did not allow appropriate transillumination and that patient was referred for a surgical gastrostomy. Remaining 23 (82%) patients underwent an uncomplicated MIC-KEY G Introducer Kit placement. Nineteen patients (68%) had the procedure performed as outpatient followed by a 48 hour hospital stay as per our protocol. Four patients (14.2%) underwent the procedure during an admission for other indications. All patients had a first gastrostomy change performed in the office 10-12 weeks after the initial placement. No major complications have been reported. Conclusions: MIC-KEY G Introducer Kit allows for a safe placement of a low profile device. First tube change does not require sedation, decreasing risks associated with anesthesia. The device does not need to be pulled in place through the esophagus and is a good option in children where potential esophageal trauma is a concern. One incision as compared to laparoscopic placement decreases scarring and does not increase the pain control requirement.

409 GASTROINTESTINAL STROMAL TUMOR CAUSING UPPER GI BLEED IN A TEENAGER—A CASE REPORT. T. Sebastian, A.A. Bader, Gastroenterology, Childrens National Medical Center, Washington, District of Columbia, UNITED STATES.

Introduction: Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors of the gastrointestinal tract and predominantly affect middle aged or older adults. GIST in younger patients is rare and not well characterized. Among young adults (age 20-29) the incidence rate was reported to be 0.06/100,000. GIST may occur sporadically or rarely in association with other tumor syndromes such as neurofibromatosis. Great majority of the patients present with gastrointestinal bleeding, with the stomach being the most common site. Our case report describes a patient who presented to our hospital with upper GI bleeding, which was eventually diagnosed to be secondary to a gastric GIST. Case: A 15-year-old previously healthy male presented to the ED with a 5-day history of black tarry stools, dizziness and fatigue. Physical exam was remarkable for pallor, tachycardia and abnormal orthostatic blood pressures. He also had significant anemia with hemoglobin 3.7, mildly elevated INR, normal transaminases and bilirubin. Patient also reported significant weight loss over last two years. Abdominal ultrasound showed hepatomegaly (17cm) and splenomegaly (14cm) with normal echogenicity and

normal doppler. He was transferred to the PICU for close monitoring and transfused to correct anemia. An EGD showed a mass (~4 cm) was visualized in the antrum. A clot was noticed on the surface of the mass although there was no active bleeding. General surgery was consulted. Subsequently, a CT demonstrated a large mass arising from the anterior wall of the stomach. Two days after, he underwent a laparoscopic partial gastrectomy for excision of the mass from the anterior surface of the stomach. Pathology of the specimen was consistent with GIST. Sections showed nodular proliferation of spindle cells with ill-defined fascicles involving the submucosa and muscularis propria. Overall the mitotic count was < 5/50 HPF. The cells were strongly positive for c-kit immunostain and CD34. Patient had a fairly uneventful postoperative course was discharged. He is currently scheduled to obtain a post-resection CT scan in 3 months. Conclusion: GISTs are very rare in childhood; however increased awareness of their existence is important for early diagnosis, treatment and prevention of metastasis. Clinical presentation often depends on the size and the site of the tumor. GI bleeding from mucosal ulceration is common. Complete surgical resection of the primary lesion is the treatment of choice. Along with endoscopy, CT and MRI are important tools in assessment of primary lesion and detection of metastasis. GIST should be considered in the work up of children who present with GI bleeding after other common causes have been ruled out.

410 BURKITT'S LYMPHOMA: ON THE DIFFERENTIAL FOR CROHN'S DISEASE?. A.K. Shakir, J. Weitzner, V. Goei, Pediatrics, GRU, Augusta, Georgia, UNITED STATES.

Crohn's disease is diagnosed with the presence of GI tract fistulas and abscesses when accompanied by other clinical signs. The occurrence of lymphomas arising after years of immunosuppressive therapy for IBD is well documented in literature. Burkitt's lymphoma (BL), a mature B-cell neoplasm, has the shortest doubling time of all neoplasms affecting humans. Sporadic BL is most commonly an extranodal disease in children and can occur as a mass in the GI tract, often in the ileocecal region. We present the case of a 15 YOWM who presented with unintentional weight loss, diarrhea, mild hepatosplenomegaly, and microcytic anemia. MRI showed several abscesses with fistulous tracts to the small intestine. Crohn's disease was confirmed by biopsies taken during upper endoscopy and colonoscopy. JW was discharged home on azathioprine, antibiotics, and steroids. He presented 3 weeks later with several episodes of bloody diarrhea. Repeat MRI showed new, extensive mesenteric lymphadenopathy; biopsy of the lymph nodes revealed Burkitt's lymphoma. This case highlights an atypical GI presentation of Burkitt's lymphoma in the pediatric population as well as the importance of keeping a broad differential for fistulous diseases in the GI tract.

LIVER

449 BRAF MUTATION IN LANGERHANS CELL HISTIOCYTOSIS WITH ISOLATED LIVER INVOLVEMENT. N. Ovchinsky, M. Martinez, S. Lobritto, Pediatrics, Columbia University Medical Center, Short Hills, New Jersey, UNITED STATES. Kato, Surgery, Columbia University Medical Center, New York, New York, UNITED STATES. Koslowe, S. Halpern, Pediatrics, Goryeb Children's Hospital, Morristown, New Jersey, UNITED STATES.

We report a case of a 4-year-old female referred for evaluation of persistent abdominal pain, abnormal hepatic imaging, elevated liver aminotransferases and inflammatory markers. She had a history of cholecystectomy 2 years prior to current presentation. Her physical exam was notable for anicteric sclera and a firm hepatomegaly palpable 3 cm below the costal margin with no associated splenomegaly or ascites. Abdominal MRI showed an unusual finding of irregular, dilated, tubular, branching fluid-filled lesions in the left hepatic lobe that extended to the porta hepatis and localized to the biliary system. The patient underwent a left hepatectomy and a choledochojejunostomy biliary reconstruction of the right hepatic lobe. Pathology specimens revealed cavernous ectasia of the intrahepatic ducts with extensive lymphohistiocytic infiltration and segmental ductal dilatation. Hilar tissue showed obliterative destruction of the large bile ducts. Immunohistochemical stains of hilar histiocytes were S100+ and CD1a+, diagnostic of Langerhans cell histiocytosis (LCH). Real-time polymerase chain reaction using allele-specific hydrolysis probes on DNA extracted from tissue was positive for BRAF V600E mutation, a kinase-encoding gene in the RAS/RAF/MAPK pathway, further supporting the neoplastic nature of LCH. The patient had a negative work-up for systemic manifestations of LCH. LCH remains a rare pediatric disease that results in abnormal proliferation of histiocytes in various tissues with unpredictable clinical course. LCH with isolated liver involvement is extremely rare. Cases involving the liver are usually multisystemic and carry a poor prognosis. Liver involvement can manifest as biliary cirrhosis since LCH cells are highly selective for biliary epithelium. LCH cells are rarely seen on liver biopsy since primarily the large ducts are involved. LCH-induced biliary cirrhosis may eventually

necessitate liver transplant (LT). Biliary injury can progress even in the absence of active LCH cells. There are no treatment guidelines for solitary liver involvement. LCH has recurred after LT in the transplanted liver with skin lesions, osteolytic bone lesions, and diabetes insipidus. Post transplant lymphoproliferative disease and acute cellular rejection are more frequently observed in LCH patients that undergo LT. Our patient was initially treated with steroids and an immunomodulator but eventually required a LT for recurrent cholangitis and progressive portal hypertension. There is debate whether LCH is a neoplastic disorder or a reactive immune condition. The demonstration of the BRAF mutation supports an oncogenic basis for LCH in this unusual case.

450 DIAGNOSIS OF MUSCULAR DYSTROPHY IN A 6 WEEK OLD WITH JAUNDICE. A. Russell, L. Gillis, Gastroenterology, Hepatology, and Nutrition, Vanderbilt University, Nashville, Tennessee, UNITED STATES. Case: BP is a 6-week-old term male infant who presented with prolonged jaundice initially presumed to be secondary to breastfeeding. His total bilirubin continued to rise, reaching a maximum of 17.8 mg/dL with a conjugated of 0.5 mg/dL at 5 weeks of age. Liver function tests were obtained at that time and were notable for an aspartate aminotransferase (AST) of 141 and alanine aminotransferase (ALT) of 73. He was referred to the pediatric hepatology clinic for evaluation. His physical exam was normal and he was feeding and growing well. A serum creatinine kinase (CK) was obtained and was significantly elevated at 8832 unit/L. Pediatric genetics was contacted and he was admitted for an expedited work up of possible muscular dystrophy, infectious rhabdomyolysis, fatty acid oxidation disorder, or inborn error of metabolism. PCR sequencing confirmed a hemizygous deletion of exons 48-50 on the dystrophin gene, consistent with Duchenne or Becker muscular dystrophy. Discussion: Serum CK, AST, and ALT can all be highly elevated with muscle breakdown and can be useful markers of muscular dystrophy. In fact, the transaminitis in patients with muscular dystrophies is most impressive before the symptoms of muscle disease are present, and so their incidental detection can facilitate early diagnosis¹. To date, the earliest reported case of muscular dystrophy diagnosed through incidental recognition of transaminitis was 10 weeks of age^{2,3}. Therefore, to our knowledge, this is the earliest report of muscular dystrophy detected secondary to abnormal hepatic labs to date. Early evaluation of CK when evaluating transaminitis can suggest the correct diagnosis and minimize otherwise extraneous or invasive testing. References: 1. Wright, M. A., Yang, M. L., Parsons, J. A., Westfall, J. M. & Yee, A. S. Consider muscle disease in children with elevated transaminase. *J Am Board Fam Med* 25, 536–540 (2012). 2. Zamora, S., Adams, C., Butzner, J. D., Machida, H. & Scott, R. B. Elevated aminotransferase activity as an indication of muscular dystrophy: case reports and review of the literature. *Can. J. Gastroenterol.* 10, 389–393 (1996). 3. Veropalumbo, C. et al. Aminotransferases and muscular diseases: a disregarded lesson. Case reports and review of the literature. *J Paediatr Child Health* 48, 886–890 (2012).

451 AN UNUSUAL PRESENTATION OF PFIC2: ACUTE SPONTANEOUS SUBDURAL HEMATOMA. A.S. Huang, R.E. Quiros-Tejeira, Pediatric Gastroenterology, UNMC, Omaha, Nebraska, UNITED STATES. Progressive familial intrahepatic cholestasis (PFIC) is a rare disease, usually manifested as a chronic cholestatic disorder that begins during the first six months of life and progresses to cirrhosis within the first decade of life. Its incidence is not well known but estimated to be 0.5 to 1 per 100,000. PFIC is an autosomal recessive disorder. It was initially known as Byler syndrome, named after an Amish farmer from the 18th century. As the genetic bases of this disorder were established, it has been sub-classified. Byler syndrome is now PFIC type 1. PFIC type 2 is probably the most common of all PFICs accounting for about 50% of the patients with this disorder. PFIC 2 is a disorder caused by mutations in FIC2 locus in chromosome 2q24 and also thought to affect the ABCB11 gene. The latter codifies for the expression of the bile salt export pump. PFIC 2 is characterized by persistent neonatal cholestasis that if biopsied, appeared to be microscopically similar to neonatal hepatitis and may progress to cirrhosis. PFIC1 & PFIC2 are classically known to have elevated bile acids and alkaline phosphatase levels but normal GGT level. We are presenting a case report of an unusual presentation of PFIC2 with a spontaneous subdural hematoma. Our patient debuted at 5 months-old with findings of acute on chronic subdural hematomas, coagulopathy, elevated transaminases, direct hyperbilirubinemia and undetectable levels of vitamin K. Because of this, she required an emergent surgical decompression due to significant herniation and midbrain deformation. She was found to have a PT > 100, INR > 10. Initially, it was attributed to severe vitamin K deficiency (Vitamin K level of 0.04 - normal 0.1-2.2). Our patient improved after treating her with Vitamin K IM, multiple PRBC transfusions and plateletpheresis. She was originally diagnosed with congenital vitamin K deficiency yet because of her abnormal liver tests and evidence of pruritus and cholestasis with normal GGT a Jaundice Chip was ordered

and confirmed the diagnosis of PFIC2. A liver biopsy was performed showing findings consistent with cholestasis and possible PFIC. After diagnosing, her vitamin K was changed from IM to PO without any problems. She has remained clinically stable with a normal PT/INR and vitamin K levels. She does not have any overt neurological sequelae from her spontaneous subdural hematoma episode.

452 IGG SUBCLASS 4 SCLEROSING CHOLANGITIS AND RESPONSE TO ORAL VANCOMYCIN. P. Arias, K. Cox, J. Kerner, M. Christofferson, D. Bass, Stanford, Palo Alto, California, UNITED STATES.

IgG subclass 4 sclerosing cholangitis (IAC) is part of a group of immune mediated diagnoses that have in common IgG4 deposition in the tissue of the affected organ as well as elevated IgG4 levels measured in serum. This group of immune mediated disorders obtains clinical remission fairly quickly with the use of systemic steroids. In adults it has been proposed that both autoimmune pancreatitis with IAC are underdiagnosed, and that some cases of Primary Sclerosing Cholangitis are secondary to IgG subclass 4 deposition in both the liver and the bile ducts. A 3 year old female presented to the ER with acute onset of abdominal pain. Her initial laboratory studies showed evidence of pancreatitis as well as elevation of her transaminases (AST 693 and ALT 785) and GGT of 669. Her past medical history was relevant for three episodes of parotitis that improved with the use of antibiotics and steroids. An MRCP showed inflammatory patterns in her pancreas consistent with the diagnosis of pancreatitis as well as stricture and dilation of the extra-hepatic bile ducts, prompting concern for sclerosing cholangitis. Further testing obtained a positive Anti-nuclear Antibody and positive Anti-smooth muscle antibody, as well as an elevated total IgG:1570 (normal is < 1200) and elevated IgG subclass 4: 346 (normal is < 100). Based on the MRCP findings and elevated IgG subclass 4 titers, she was diagnosed with autoimmune pancreatitis. Due to the persistent elevation of her liver numbers, MRCP findings and positive serology, a liver biopsy was obtained. The biopsy showed findings in the liver parenchyma consistent with sclerosing cholangitis. She was started on steroids (2mg/kg) and Ursodeoxycholic acid (Actigall), and her clinical symptoms resolved. Ten days after discharge, she had no additional episodes of abdominal pain; however, both her transaminases and GGT had not normalized with the use of systemic oral steroids. Based on the failure to see a complete normalization of her liver numbers, at ten weeks after diagnosis we decided to start oral vancomycin at 10mg/kg TID, based on previous evidence by Cox et al. that shows clinical and laboratory improvement in patients with PSC after the start of oral vancomycin. In the course of three weeks her liver numbers improved significantly, and we were able to wean her off steroids successfully. Her clinical improvement continued, and she has not had any more episodes of abdominal pain. Oral vancomycin has been shown by Cox et al. to work in the intestinal immune system, in particular in the regulatory T Cells. The results of this intervention in case series and long term follow up of patients with PSC is promising and has shown to be an effective therapy to achieve long term remission of clinically relevant PSC. This case further demonstrates that oral vancomycin may have immune-regulatory properties. Though our patient has been weaned off steroids, the long term efficacy and safety of vancomycin as an alternative maintenance medication in patients with IAC/autoimmune pancreatitis remains a very important question. The dose related effect of vancomycin for achieving remission in these patients has yet to be determined.

453 A RARE CASE OF A RANITIDINE OVERDOSE PRESENTING AS ACUTE LIVER FAILURE.. L. Mullinax, Department of Medical Education, Miami Children's Hospital, Miami, Florida, UNITED STATESL. Caicedo Oquendo, Department of Pediatric Gastroenterology, Miami Children's Hospital, Miami, Florida, UNITED STATES.

Ranitidine is one of the most commonly prescribed medications; numerous scripts are written daily in both the general pediatrician's office as well as in the pediatric gastroenterologist's office for ranitidine. Ranitidine is even more common than one is able to account for as it is available over-the-counter without need of a proper medical evaluation. It can also be recommended in a wide range of dosage amounts during the day or even multiple times a day. We report a case in which ranitidine was unfortunately overdosed by a primary medical doctor in a pediatric patient suffering from gastroesophageal reflux and in a relatively short amount of time, the patient developed acute liver failure secondary to the high dosage. Luckily, the patient was closely monitored was diagnosed in a timely fashion with acute liver failure. With great delight, the patient improved dramatically after ranitidine was discontinued and after the patient was given an N-acetylcysteine infusion. Interestingly, from the subsequent work up afterwards, our patient was diagnosed with Alagile syndrome. This case raises awareness and emphasizes the potential life-threatening harm of Ranitidine, a generally benign medication, particularly in complex patients.

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A 4 week old full term girl was evaluated for failure to thrive, prolonged jaundice and hypoglycemia. The neonatal course was complicated by adrenal insufficiency, septic shock and hypoxic respiratory failure. Her length and weight were at the 3rd centile. She had scleral icterus and wandering nystagmus. Ophthalmologic examination showed bilateral optic nerve hypoplasia. Liver function tests showed elevated transaminases and direct hyperbilirubinemia. Liver biopsy demonstrated mild hepatocellular and canalicular cholestasis. An intraoperative cholangiogram showed normal extrahepatic bile ducts. MRI of the head showed absence of the septum pellucidum and hypoplastic appearance of the optic nerves and chiasm, which is consistent with Septo-optic dysplasia. Because results of the thyrotrophin releasing hormone stimulation test was consistent with hypothalamic hypothyroidism, L-thyroxine was prescribed. Follow up liver function tests showed resolution of cholestasis. However, due to the recurrent hypoglycemia, persistently elevated transaminases and growth delay secondary to growth hormone deficiency, the patient was started on growth hormone replacement. Repeat liver function tests showed significant improvement. Discussion: The association of liver dysfunction with hypopituitarism was first suggested in 1956. The etiology of the liver dysfunction in optic nerve hypoplasia and hypopituitarism is not well understood. It has been suggested that growth hormone and cortisol deficiencies may be responsible for the development of hyperbilirubinemia since these hormones seem to modulate bile acid synthesis and bile flow. Although hypothyroidism may present with prolonged neonatal jaundice, it is generally associated with unconjugated hyperbilirubinemia. Conclusion: The diagnosis of Septo-optic dysplasia must be considered in infants who present for evaluation of cholestatic jaundice, particularly if there is associated hypoglycemia and nystagmus. The liver function tests seem to normalize following hormone replacement.

ALPHA 1 ANTITRYPSIN DEFICIENCY. S. Mani, N. Patel, N. Alkhouri, Pediatrics, Cleveland Clinic Children's, Cleveland, Ohio, UNITED STATES.

Introduction: Alpha1-antitrypsin deficiency (AATD) is the most common genetic cause of liver disease in neonates and children, with the severe phenotype (PiZZ) occurring in approximately 1:3500 births. In infancy, initial presentation is usually associated with neonatal hepatitis or cholestatic jaundice, and rarely intracranial hemorrhage (ICH). Of these infants only 2-3% will experience advanced fibrosis or cirrhosis leading to transplantation in childhood. Case Report: A 6-week old previously healthy male presented to the emergency department (ED) with a 1-day history of increasing lethargy, poor feeding, and persistent vomiting. The patient was born full term via a planned home delivery using a midwife and vitamin K prophylaxis was not given after birth. He was exclusively breast fed, had a normal newborn screen and no history of jaundice, poor feeding, or illness prior to the acute onset of his presenting symptoms. Physical exam revealed jaundice, poor responsiveness, extremely tight hand grasp, ptosis of the left eyelid and a 4mm fixed and dilated pupil. CT scan of the brain revealed a left fronto-temporal intracranial hemorrhage, which required an emergent left-sided decompression hemicraniectomy with evacuation of a left-sided subdural hematoma. Initial lab studies revealed transaminitis and conjugated hyperbilirubinemia with a prolonged PT/INR and PTT. Post-operatively the patient was maintained on TPN and monitored in the ICU. Persistent conjugated hyperbilirubinemia and an ongoing TPN requirement prompted evaluation for cholestatic liver disease. Genetic evaluation for AATD returned positive for the PiZZ phenotype at two months of age. By three months of age the patient developed ascites secondary to portal hypertension, and subsequent liver biopsy revealed extensive fibrosis and cirrhosis of the liver. By six months of age, he developed significant ascites requiring paracentesis, and by seven months he underwent successful orthotopic liver transplant. Discussion: Late hemorrhagic disease of the newborn (HDN) is bleeding which occurs between one week and six months of life due to severe vitamin K deficiency. Infants with cholestatic liver disease may become vitamin K deficient secondary to fat malabsorption or poor intake and may present with late HDN in the form of a life threatening ICH. ICH is not often identified as the presenting symptom of severe vitamin K deficiency secondary to severe AATD as seen in this patient, and is more often associated with cholestasis secondary to biliary atresia or hepatitis. Conclusion: The presence of late HDN presenting with ICH in addition to laboratory evidence suggestive of cholestatic liver disease with coagulopathy should prompt the evaluation of AATD as a potential etiology of Vitamin K deficiency.

Introduction: Drug Rash with Eosinophilia and Systemic Symptoms (DRESS Syndrome) is a potentially life threatening drug induced hypersensitivity reaction characterized by skin rash, fever, leukocytosis with eosinophilia or atypical lymphocytosis, lymphadenopathy and liver or renal dysfunctions. It has most commonly been associated with antiepileptics, allopurinol, sulfonamides and some antibiotics. The symptoms usually occur 2-6 weeks after offending drug is initiated. The incidence of DRESS ranges from 1 in 1000 to 1 in 10,000 exposures. It is quite rare in children. Primary HHV 6 infection or reactivation has been incriminated in the pathogenesis of DRESS syndrome. We report a patient at our institution who developed DRESS syndrome following the use of Bactrim.

Case: A 15 year old previously healthy female presented to the ED with jaundice, abdominal pain and dark urine. She had been on Bactrim for a urinary tract infection 5 days prior to the onset of jaundice. Physical exam revealed icterus, hepatomegaly. Initial lab tests showed significantly elevated transaminases, bilirubin, peripheral eosinophils and INR. Abdominal ultrasound showed gall bladder wall thickening and hepatomegaly. Work up for hepatitis and cholestasis include infectious, metabolic and autoimmune hepatitis was negative. She underwent a percutaneous liver biopsy which showed an acute hepatitic pattern with cholestasis, mild peri portal fibrosis and scattered eosinophils. She was started on Vitamin K and Ursodeoxycholic acid. Since the transaminases and INR started improving she was discharged. The patient returned to the clinic 1 week after discharge with a pruritic maculopapular rash on her arms and legs. Labs done at that visit showed transaminases trending up and persistent cholestasis. An allergic reaction was suspected and the Allergy Department was consulted who agreed that her presentation was consistent with DRESS and recommended starting her on oral steroids. She was on Prednisone for around 3 weeks and had an excellent initial response with a down trending of her transaminases and bilirubin. However once she was tapered off the Prednisone the transaminases started rising again. She underwent a repeat liver biopsy which showed marked inflammation with fibrosis and bile duct proliferation suggestive of progression of the disease. Prednisone was restarted which normalized the liver function tests. After a slow taper of steroids she remains symptom free with normalized labs.

Conclusion: Bactrim associated DRESS syndrome is rare with only few isolated case reports in the literature review. A high index of suspicion for DRESS syndrome must be maintained in patients who are on medications that are known to cause DRESS, specially because the symptoms can be delayed by weeks after initiation of the offending agent. Prompt withdrawal of the drug is imperative. Systemic corticosteroids may be used in case of organ failure or life threatening conditions. A cholestatic pattern tends to have a poor prognosis which can sometimes lead to liver failure and need for liver transplant.

Giant Cell Hepatitis (GCH) in association with Autoimmune Hemolysis Anemia (AIHA) is an uncommon disease of early childhood with unknown pathogenesis and poor response to immunosuppressive therapy and often progresses to fatal liver disease. We report a case of GCH with a Coombs positive AIHA in an 8-month-old boy.

CASE PRESENTATION. An 8 month old boy with acute onset of jaundice following upper respiratory tract infection. He was first child from unrelated parents. There were no complaints of acholic stool, fever, weight loss or gastrointestinal bleeding. In physical examination he had hepatomegaly, petechiae and bruises in limbs. His height and weight were normal. Blood test revealed severe anemia, hemolysis and abnormal liver function test. Serologic evaluation was negative for hepatitis viruses A, B, and C, Epstein-Barr virus, Cytomegalovirus and HIV. Ceruloplasmin and antinuclear antibodies were negative. Liver biopsy showed giant cell transformation of hepatocytes. The diagnosis of GCH with AIHA was made. Treatment started with IV infusion of IVIG (1 gr/kg /day) and methyl prednisone (10 mg/kg/day) for five days, followed by oral prednisone (4 mg/kg/day). After that, hemoglobin level increased and liver enzyme level decreased. He had follow-up in consulting-room. During that period of 6 months, he had a stable evolution and the steroid therapy was decreased. However, he developed 3 upper respiratory tract infections, and he died at 16 month old for pneumonia.

DISCUSSION. Hepatic giant cell transformation is distinctly uncommon after the neonatal period. The association of GCH with Coombs-positive hemolytic anemia seems to be a distinct entity. All patients had disease onset between 6 months and 2 years of age, with manifestations of acute and Coombs positive haemolytic anemia at presentation. Hepatitis occurred within 2 months after the diagnosis of AIHA. The clinical course of GCH with AIHA is aggressive. In most patients there is an initial response to immunosuppressive therapy, but relapses occur commonly, and immunosuppression

with steroids and azathioprine is needed for maintaining remission. Usually, the hemolytic anemia is controlled readily after the initial episode. In a few children with persistent hemolysis, plasmapheresis and splenectomy have been reported to be beneficial. Some patients had a severe, rapid fatal course, which is unresponsive to immunosuppression. Liver failure, severe seizure disorder, unexplained encephalopathy, and severe infections are usually the causes of death. REFERENCES: Gorelik M, Debski R, Frangoul H. Autoimmune hemolytic anemia with giant cell hepatitis: case report and review of the literature. *J. Pediatr Hematol Oncol.* 2004;26(12):837-9. Gehrs, BC, Friedberg RC. Autoimmune hemolytic anemia. *Am J Hematol* 2002;69:258-271. Brichard B, Sokal E, Gosseye S, Buts JP, Gadsseux JF, Cornu G. Coombs-positive giant cell hepatitis of infancy: effect of steroids and azathioprine therapy. *Eur J Pediatr* 1991; 150:314-317. Lau JY, Koukoulis G, Mieli-Vergani G, Portmann BC, Williams R. Syncytial giant-cell hepatitis-a specific disease entity? *J Hepatol* 1992; 15:216-9.

458 A ROLE FOR VANCOMYCIN IN THE TREATMENT OF CRYPTOSPORIDIUM-ASSOCIATED SCLEROSING CHOLANGITIS AND REJECTION POST LIVER TRANSPLANT. R. Fischer, J. Daniel, Gastroenterology, Children's Mercy Hospital, Kansas City, Missouri, UNITED STATESW. Andrews, R. Hendrickson, Pediatric and Transplant Surgery, Children's Mercy Hospital, Kansas City, Missouri, UNITED STATES.

BACKGROUND: Liver transplantation (LTx) is a life-saving measure for many children with severe liver disease. Successful management of patients post-LTx generally requires life-long immunosuppression. Unfortunately, immunosuppressed individuals are at risk of infectious complications, including gastroenteritis due to cryptosporidium and the development of cryptosporidium-associated sclerosing cholangitis (cSC). Herein, we describe 3 cases of cryptosporidium infection in children post-LTx associated with rejection and cSC. And, we describe our use of oral vancomycin for the treatment of cSC in these patients. METHODS: We reviewed the records of patients in our liver transplant program with a known history of cryptosporidium infection complicated by histologic and/or radiographic sclerosing cholangitis. Individual consent for each case was obtained. RESULTS: We identified three LTx patients (two females and one male; aged 4, 6 and 16 years) with a history of cryptosporidium infection complicated by allograft rejection recalcitrant to intravenous methylprednisolone therapy. Treatment for cryptosporidium infections included nitazoxinide/azithromycin, nitazoxinide/paromomycin and nitazoxinide alone. Each patient had histologic and/or radiographic evidence of sclerosing cholangitis not seen prior to cryptosporidium infection. Two patients received thymoglobulin therapy to help manage rejection. One patient that received thymoglobulin went on to recover fully, with histologic resolution of rejection and cSC without the use of vancomycin. In the second patient (4-year-old male), rejection resolved, but cSC persisted, eventually responding to the addition of vancomycin (40 mg/kg/day divided TID) to the patient's medication regimen. In the third patient (16-year-old female), recalcitrant rejection and concomitant cSC were subverted by the early use of oral vancomycin (500 mg TID) with complete resolution of the histologic and radiographic features of each. Thymoglobulin was not required to treat rejection once vancomycin was added. CONCLUSION: Vancomycin is an emerging treatment adjunct for primary sclerosing cholangitis in children. We have successfully used vancomycin to manage cSC and rejection in children following LTx.

459 INFLIXIMAB ASSOCIATED AUTOIMMUNE HEPATITIS IN CHILDREN WITH INFLAMMATORY BOWEL DISEASE. S. Mostamand, S. Schroeder, J. Schenkein, T.A. Miloh, Phoenix Children's Hospital, Phoenix, Arizona, UNITED STATES.

Introduction The liver can be affected by different mechanisms in inflammatory bowel disease (IBD). Infliximab (INX) is commonly used to induce and maintain remission in IBD. We report two cases of children with ulcerative colitis (UC) who had normal liver enzymes (LFTs) prior to INX and diagnosed with autoimmune hepatitis (AIH) after INX induction. Case 1 A 12 year old boy at diagnosis of UC was treated with prednisone, mesalamine and balsalazide. He had worsening UC 2 years later, started purinethol which was poorly tolerated and received INX induction. At induction: ALT 10 IU/L, AST 19 IU/L. After 5 months of INX every 8 weeks: ALT 234 IU/L, AST 184 IU/L, ALP 117 IU/L, bilirubin 0.5 mg/dL, albumin 4.5g/dL and CRP < 1.0 mg/L. Anti-nuclear antibody (ANA), anti-Smooth muscle antibody (SMA), anti-mitochondrial antibody, IgA-endomysial antibody and anti-Liver Kidney Microsomal Antibodies (LKM) were negative. Liver ultrasound was normal. Liver biopsy showed moderate inflammatory infiltrate of mainly CD3 T-lymphocytes, few CD20 B-cells, neutrophils, plasma cells and rare eosinophils. Bile ducts were normal. There was no fibrosis. Within a month of discontinuing INX, LFTs improved with complete normalization by 3 months. His current regimen is balsalazide, probiotic and a Paleo diet with control of UC symptoms. LFTs remain normal. Case 2 A 15 year old female diagnosed with UC at age 3 was lost to follow up for

11 years as parents pursued naturopathic care. At age 14, she was hospitalized with hematochezia and anemia and started on methylprednisolone with INX induction (5mg/kg). Lab work up: ALT 11 IU/L, AST 5 IU/L, GGT 415 IU/L, ALP 108 IU/L, bilirubin 0.2 mg/dL, albumin 2.6 g/dL and INR 1.0. Autoimmune markers were negative (SMA, anti-LKM, and ANA), except for positive double-stranded DNA (dsDNA) 14 IU/ml. MRCP showed intrahepatic biliary dilatation with alternating focal segments of narrowing suggestive of primary sclerosing cholangitis. Ursodiol and budesonide were started in addition to balsalazide. She was refractory to therapy, INX drug level was low with negative INX antibodies and INX infusions were increased to 10mg/kg monthly with a re-induction phase. Ten months after initial INX: ALT 260 IU/L, AST 423 IU/L, GGT 117 IU/L. Liver biopsy showed interface hepatitis with lymphocytes, plasma cells and PMNs, mild fibrosis around bile ducts and stage II-III fibrosis. INX frequency was decreased to every 6 weeks then discontinued. Azathioprine was started and poorly tolerated. LFTs normalized 1 month after discontinuing INX. Post-INX colonoscopy shows severe pancolitis, inflammatory polyp and ileitis. Conclusion To our knowledge, these are the first case reports of INX associated AIH in children with IBD. There are 27 adult cases reported. This condition may present with increased LFTs in the presence of negative autoimmune antibodies or positive dsDNA. A liver biopsy may be required for accurate diagnosis. LFTs normalized after INX discontinuation without need for long term steroids or immunosuppression. The choice of subsequent IBD maintenance drugs may be challenging and other biologics, immunosuppressants or colectomy may need to be considered.

460 FAMILIAL HEMOPHAGOCYtic LYMPHOHISTIOCYTOSIS MIMICKING NEONATAL HEMOCHROMATOSIS. B.E. Vitola, J. Casper, Pediatric Gastroenterology, Hepatology, and Nutrition, Medical College of Wisconsin, Milwaukee, Wisconsin, UNITED STATESM. Thakar, J. Talano, Pediatric Blood and Marrow Transplant, Medical College of Wisconsin, Milwaukee, Wisconsin, UNITED STATES. Szabo, Pediatric Pathology, Medical College of Wisconsin, Milwaukee, Wisconsin, UNITED STATESD. Basel, R. Veith, Genetics, Medical College of Wisconsin, Milwaukee, Wisconsin, UNITED STATES.

We describe a case of suspected neonatal hemochromatosis that was diagnosed in a stillborn infant of an affected sibling who subsequently presented at 2 months of age with hemophagocytic lymphohistiocytosis (HLH). A first time mother gave birth to a 37-week stillborn male with hydrops fetalis. Based on an autopsy of this infant, he was presumptively diagnosed with neonatal hemochromatosis although this could not be confirmed. Due to this history, with a subsequent pregnancy, this mother received intravenous immune globulin starting at 20 weeks prior to this child's birth on a protocol with the aim of preventing alloimmune neonatal hemochromatosis. He was born at 36 weeks gestation via planned cesarean section. He remained in the NICU briefly for mild respiratory distress and neonatal jaundice requiring phototherapy. Evaluation at birth revealed an INR of 1.26, which was consistent with newborn coagulation findings, and his ferritin was in the low 200s; therefore, it was elected to follow him. He was well until he presented at 59 days of life with a 3 day history of fever to a maximum of 102.6°F, sleepiness and poor feeding. A septic workup was initiated in the ED. On admission, his laboratory findings were significant for an AST 280 IU/L, ALT 136 IU/L, albumin 2.5 g/dL, total bilirubin 1.3 mg/dL, PT 17.4 seconds, INR 1.49, PTT 44 seconds, D-dimer 8.96 ug/mL, fibrinogen 242 mg/dL, ferritin 5210 ng/dL, WBC 3 K/uL, Hg 8.3 g/dL, platelets 74 K/uL, ANC 1500. He was also noted to have prominent hepatosplenomegaly. He was intubated for respiratory failure. An MRI of the liver and pancreas was obtained to evaluate for hemochromatosis which revealed low signal intensity throughout the liver on all pulse sequences, particularly well seen on the T2W and FISP images consistent with suspected primary hemochromatosis. However, due to the atypical presentation for neonatal hemochromatosis, further diagnostic studies were obtained. The patient underwent lower lip biopsy to obtain minor salivary gland tissue to look for iron deposition within the minor salivary glands. However the tissue obtained was inadequate. The sIL2r was elevated at 38,872 units/ml, NK cell and B cell counts were zero, but granzyme B was detected. No perforin was detected. Familial HLH was suspected and genetic testing showed a mutation in MUNC 13-4 confirming the diagnosis of HLH. Mother's obstetrical history raised questions about the possibility of familial HLH contributing to the demise of the stillborn sibling. Genetic studies were obtained on the stillborn brother and he was found to have the same mutation as the patient. We reported this case to increase awareness of familial HLH in the differential diagnosis where neonatal hemochromatosis is a concern.

461 EOSINOPHILIC CHOLANGITIS: A CHALLENGING CASE OF AN OBSTRUCTIVE BILIARY MASS. A. Koral, Y. Rivas, U. Khan, G. Tomer, Children's Hospital at Montefiore, Bronx, New York, UNITED STATES.

Eosinophilic cholangitis is a rare biliary tract disorder characterized by a benign liver mass predominantly comprised of eosinophils, sometimes causing biliary obstruction. Less than 30 cases have been reported worldwide. This case report presents a 15 year old adolescent female with asthma who presented with 1 month of intermittent abdominal pain, 5 episodes of non-bilious non-bloody emesis and a 7 pound weight loss. About 1 week prior to presentation she developed pruritus and jaundice. She had no pets, no allergies and no recent travel. She denied taking any medication or a family history of liver disease. On physical exam her vital signs were normal. She was jaundiced with a soft, non-tender abdomen and no hepatosplenomegaly. Her initial labs showed a WBC of 17.7 k/ μ L with 19% eosinophils, a total bilirubin of 11.8 mg/dL with a direct bilirubin of 7.5 mg/dL, an AST of 65 U/L, ALT of 72 U/L and a GGT of 67 U/L. Imaging of her liver by ultrasound and MRCP showed a mass at the bifurcation of the hepatic ducts and dilation of the proximal intrahepatic bile ducts. She underwent an ERCP and cholangioscopy where biopsies were taken and a biliary stent was placed in the left hepatic duct. Several days later her pain and jaundice resolved. Pathology of the mass revealed an eosinophilic infiltrate of the bile ducts. Further work up showed persistent elevation of her WBC and peripheral eosinophilia, so a bone marrow biopsy was done and showed a mild eosinophilia of 10%. Biopsies from an upper endoscopy did not show an eosinophilic infiltrate. She was antigen positive for *Giardia* and she was treated with metronidazole. Antigenic and microscopic studies for other parasites were negative. She was started on high dose oral prednisone and discharged from the hospital. On follow up she was feeling well and had no clinical signs of obstruction. Her eosinophilia had resolved and her total bilirubin had decreased to 0.9 mg/dL and direct bilirubin to 0.5 mg/dL. An ultrasound performed 6 weeks after her initial presentation showed resolution of the mass and no dilation of the intrahepatic bile ducts. This is the first reported case of eosinophilic cholangitis in a pediatric patient in the US. This condition can pose a diagnostic challenge to clinicians because it often mimics malignant disease. In this case, an extensive work up was conducted to rule out other causes of eosinophilia, such as parasites, allergies, and bone marrow malignancies. In most reported cases of eosinophilic cholangitis, a diagnosis is not made until after surgical resection of the mass. In this patient, a multispecialty team was able to diagnose the condition based on endoscopic biopsies and careful workup to rule out other causes of eosinophilic disease. The patient responded well to medical management with corticosteroids that led to resolution of her symptoms and regression of the hepatic mass without the need for invasive surgical resection. Knowledge of this rare condition may prevent other patients with eosinophilic cholangitis from unnecessary invasive procedures.

NUTRITION

482 A NOVEL COMPLICATION OF THE NATIONAL PARENTERAL MICRONUTRIENT SHORTAGES: SUSPECTED COPPER DEFICIENCY INDUCED MYELONEUROPATHY. D. Wendel, C. Bales, M. Mascarenhas, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES.

A 23 month old with short bowel syndrome secondary to total aganglionosis, who was maintained exclusively on parenteral nutrition (PN), was admitted to the hospital with bilateral leg weakness. His mother had noticed some leg shaking upon standing for a few days prior to presentation. On exam, he had an ataxic gait and decreased reflexes in his lower extremities with intact sensation. His evaluation included a CBC with WBC 3.9 g/dL and Hgb 10.5 g/dL, negative blood and CSF cultures, normal spinal/brain MRI, EMG with increased small and short polyphasic motor units, normal thyroid testing, ESR 42 mm/hr, CRP 5 mg/dL, and CK 25 U/L (normal 60-305). The following vitamins/minerals were checked and normal: B6, B12/MMA, vitamin E, carnitine, and zinc. His selenium level had been intermittently low in the past and was undetectable at the time of the admission. Copper levels were reported as <10 ug/dL (normal 75-153) and ceruloplasmin 6 mg/dL (normal 18-27). There had been a shortage of micronutrients, and he had not been receiving copper in his PN during the previous 7 months. Intravenous copper became available during the admission and was added back at a dose of 20 mcg/kg/day. With this change, his copper gradually normalized over the next 4 months. His symptoms and exam showed marked improvement and eventual full recovery that coincided with the copper normalization. The patient's symptoms were determined to be secondary to a copper deficiency induced myeloneuropathy with possible myopathy. This case underscores the importance of adequate micronutrient monitoring and management in the setting of ongoing shortages.

483 UNDIAGNOSED MYASTHENIA GRAVIS UNMASKED BY GENERAL ANESTHESIA IN A PATIENT WITH PERSISTENT WEIGHT LOSS. L.H. Gutierrez, Pediatrics, SUNY Upstate Medical University, Syracuse, New York,

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Introduction: Myasthenia gravis (MG) is an autoimmune disease resulting in destruction of the post-synaptic nicotinic receptors at the neuromuscular junction. Classically, the earliest symptoms of MG are ocular, including ptosis and diplopia. Other less common early symptoms include dysphagia, and fatigable chewing. This case report describes the unusual presentation of MG in a teenager patient who presented for an elective percutaneous endoscopic gastrostomy (PEG) tube placement due to persistent weight loss. The patient underwent the procedure without any complications until the end, when she could not maintain spontaneous ventilation. The patient was transferred from the recovery room to the pediatric intensive care unit (PICU) for further management. Subsequent investigations including a bedside electromyography (EMG) led to the diagnosis of myasthenia gravis. Case report: A 16 year old with a chief complaint of weight loss and gastro esophageal reflux (GER) presented to the pediatric gastroenterology clinic for evaluation. The patient failed treatment with anti-reflux medication which prompted an upper endoscopy, colonoscopy and pH probe study, all of which were normal. The patient's GER subsided but she continued to demonstrate persistent weight loss on subsequent visits to the clinic. The patient continued to lose more weight despite the use of appetite stimulants and nutritional supplements. Therefore, a PEG tube for night feedings was recommended. The patient underwent a PEG tube placement without any complications until it was time to extubate. At this point she developed respiratory failure, which triggered the use of reversal anesthetic agents such as naloxone and flumazenil. While in the recovery room the patient's mental status deteriorated which led to a transfer to the PICU requiring mechanical ventilation. Possible etiologies for the patient's hypoventilation and decreased responsiveness were studied. Further investigations led to a bedside EMG which revealed findings supportive of MG. The patient was finally successfully extubated after treatment with intravenous immunoglobulin, oral prednisone and pyridostigmine. Conclusion: This case highlights the complexity of the differential diagnoses of a patient presenting with persistent weight loss which could be the first presentation of underlying myasthenia gravis

484

A SERIOUS ADVERSE EVENT ASSOCIATED WITH THE CURRENT ADULT INTRAVENOUS

MULTIVITAMIN SHORTAGE: A CASE OF ACUTE THIAMINE DEFICIENCY LEADING TO SEVERE LACTIC ACIDOSIS, HYPERAMMONEMIA, CARDIOVASCULAR COLLAPSE, ACUTE RENAL FAILURE, AND UNNECESSARY EXPLORATORY L. D.M. Barnes, J. Kerner, Pediatric Gastroenterology, Hepatology, and Nutrition, Stanford Lucile Packard Children's Hospital, Union City, California, UNITED STATESD.M. Barnes, United States Navy, Palo Alto, California, UNITED STATESE. Burgener, Pediatrics, Lucile Packard Children's Hospital, Palo Alto, California, UNITED STATESM. Ramsi, Pediatric Intensive Care Unit, Stanford Lucile Packard Children's Hospital, Palo Alto, California, UNITED STATESC. Mowbray, Pharmacy, Stanford Lucile Packard Children's Hospital, Palo Alto, California, UNITED STATES.

Introduction: Thiamine is an essential vitamin, and its deficiency is often considered only in the setting of a malnourished alcoholic patient. In acute thiamine deficiency, the body cannot convert pyruvate to acetyl coenzyme A causing a predominance of anaerobic metabolism then leading to the build up of lactic acid. In the mid-1990s, cases of acute thiamine deficiency were reported in the setting of a multivitamin shortage. Case Report: A 16 year-old male with ulcerative colitis underwent total colectomy at an outside hospital and was started on TPN on admission. By day 32, he developed ataxia, tremors, and hallucinations; over the following 2 days, his bicarbonate dropped to 11 and lactic acid rose to 16. An abdominal CT scan did not show a source of infection. He became hypotensive requiring vasopressors, and was intubated for impending respiratory failure. He was transferred to our intensive care unit. Despite receiving multiple doses of sodium bicarbonate en route, but his pH upon arrival was still extremely low at 6.8. Immediately after arrival, he was taken to the operating room for emergent exploratory surgery where they found well-perfused bowel with no abnormality. He returned to the PICU where he required an insulin drip, many units of blood products, and was placed on continuous venovenous hemodialysis (CVVH) for refractory acidosis with lactate >30. His ammonia had risen to 608 and his creatinine to 2.3. The PICU fellow confirmed he had been receiving TPN at the outside hospital without multivitamin for the previous 31 days. The patient was then treated with 400mg IV thiamine. Two hours after administration, his pH was 7.17 and his bicarbonate 12.1 (up from 3.6). He was given a second dose of 400mg 2 hours later and was then dosed every 4 hours. Within 6 hours, he no longer required vasopressors nor insulin and his pH and bicarbonate had normalized. Within 12 hours, he was taken off of CVVH. His ventilator settings were weaned aggressively, and was extubated the following day. After his sedation was discontinued, his mental status improved. His renal function had not completely recovered, which was felt to be related to hypoperfusion during his decompensation.

He had a brain MRI 3 days later which showed no signs of Wernicke's encephalopathy. A thiamine level drawn prior to his first dose of multivitamin (did not result for 3 days), was 7 nmol/L (normal 8-30). Upon further contact with the transferring hospital, they had no supply of intravenous adult multivitamin. A memorandum was sent to their practitioners notifying them of this shortage. Discussion: This case demonstrates the danger associated with the current multivitamin shortage as well as the lack of education about the severe presentation of acute thiamine deficiency. It is important to consider thiamine deficiency in a TPN dependent patient with severe metabolic acidosis. This case demonstrates preventable morbidity that is a direct result of the current multivitamin shortage.

485 FAILURE TO THRIVE AND HYPOCHOLESTEROLEMIA, A CASE REPORT. P. Freswick, K. Thomsen, M. Linton, Vanderbilt University, Nashville, Tennessee, UNITED STATES.

Case: CD is a 2 year-old male who presented at 3 months old for failure to thrive (3rd percentile) and vomiting. Patient was born term, 2.90 kg, vaginal deliver with normal newborn screen. His vomiting started soon after birth and he did not attain birth weight until after 2 weeks old. Bowel movements described as 1-2 per day watery, non-bloody but guaiac positive. Secondary to symptoms, he was started on omeprazole and transitioned from exclusive breastfeeding to Alimentum and breastfeeding with maternal dairy, soy, and egg avoidance but symptoms persisted. Admission for NG Alimentum demonstrated adequate weight gain. After discharge vomiting and poor weight gain persisted, however, despite transition to NG Neocate. CD was subsequently admitted again at 5 months old for further testing: CMV, EBV, stool tests, sweat chloride where normal, but abdominal ultrasound demonstrated increased liver echogenicity and EGD demonstrated diffuse lipid vacuolation of duodenal epithelial cells. Laboratories as follows: LDL 15 mg/dl, HDL 16 mg/dl, triglycerides 29 mg/dl, cholesterol 37 md/dl, vitamin E < 0.5 mg/dl (normal vitamin A, vitamin D levels), Apolipoprotein B 38 mg/dl, CPK 128 U/L. After treatment with high MCT formula (Monogen), essential fatty acid supplementation, and 100 IU/kg vitamin E, he demonstrated appropriate weight gain and vitamin E level normalization. *SAR1B* genotyping revealed no mutations. Microsomal triglyceridetransfer protein (MTTP) genotyping revealed exon 10 heterozygote aspartic acid to alanine amino acid substitution of unknown clinical significance. Whole genome exome sequencing is in progress. Discussion. Familial hypocholesterolemia disease subtypes, namely abetalipoproteinemia, hypobetalipoproteinemia, and chylomicron retention disease (CRD), may be difficult to differentiate. Our patient clinically fits CRD as his LDL and Apolipoprotein B were decreased but not zero¹ However, all patients with CRD described thus far in the literature have had a *SAR1B* >gene mutation¹⁻³, which our patient does not. Thus, our patient either has a unique presentation of failure to thrive and hypocholesterolemia, or a novel CRD mutation. References 1. Peretti N, Sassolas A, Roy CC, et al. Guidelines for the diagnosis and management of chylomicron retention disease based on a review of the literature and the experience of two centers. Orphanet J Rare Dis. 2010;5(1):24. doi:10.1186/1750-1172-5-24. 2. Peretti N, Roy CC, Sassolas A, et al. Chylomicron retention disease: a long term study of two cohorts. Mol Genet Metab. 2009;97(2):136-142. doi:10.1016/j.ymgme.2009.02.003. 3. Cefalù AB, Calvo PL, Noto D, et al. Variable phenotypic expression of chylomicron retention disease in a kindred carrying a mutation of the Sara2 gene. Metab Clin Exp. 2010;59(4):463-467. doi:10.1016/j.metabol.2009.07.042.

486 RECURRENT PNEUMATOSIS INTESTINALIS IN 2 PEDIATRIC PATIENTS WITH INTESTINAL. J. Lim, T. Mcenaney, R. Fischer, B. Lyman, R. Johnson, Gastroenterology, Childrens Mercy Hospital, Kansas City, Missouri, UNITED STATES. J. Lim, R. Fischer, R. Hendrickson, Pediatrics, University of Missouri in Kansas City, Kansas City, Missouri, UNITED STATES. R. Hendrickson, Surgery, Childrens Mercy Hospital, Kansas City, Missouri, UNITED STATES. Pneumatosis intestinalis (PI) is a radiographic finding of intramural air in the small intestine and is concerning for necrotizing enterocolitis (NEC). There is a paucity of literature regarding medical treatment strategies of PI in infants and children, especially in patients with intestinal failure (IF). We describe 2 cases of recurrent PI in non-neonatal pediatric patients with intestinal failure that occurred during advancement in their oral/enteral feeding treated with enteral metronidazole. The first patient was a 3-month-old male with a diagnosis of gastroschisis. He developed NEC requiring medical and surgical intervention. He subsequently developed recurrent PI. In one of the episodes he was positive for a Norovirus infection. His feeds were advanced slowly and metronidazole was initiated at the same time. After 2 months he was completely enterally fed and the antibiotic was discontinued. He had no further complications. The second patient was a 22-month-old female with a diagnosis of short bowel syndrome secondary to NEC requiring small bowel and colonic resection. She developed recurrent bouts of PI. This was initially managed by withholding enteral nutrition and intravenous antibiotics. Subsequently, metronidazole was initiated while slowly advancing enteral feeding for the next 4 months. She is close to being enterally

autonomous and off antibiotics. Recurrent PI is a concerning problem in the setting of IF. Enteral metronidazole therapy with slow advancement of feeds may be a viable treatment for these patients.

487 THIAMINE DEFICIENCY PRESENTING AS ENCEPHALOPATHY IN A TPN DEPENDENT CHILD. B.M. Roebuck, Pediatrics, University of Oklahoma Health Sciences Center, Oklahoma City, Oklahoma, UNITED STATES. A.C. Andrews, M.I. Steele, Pediatric Gastroenterology, Hepatology and Nutrition, University of Oklahoma Health Sciences Center, Oklahoma City, Oklahoma, UNITED STATES.

Six-year-old female with Short Bowel Syndrome and total parental nutrition (TPN) dependence was admitted with a four day history of emesis, headaches, vision disturbances and motor incoordination. There was no history of fever, upper respiratory symptoms, decreased oral intake or accidental ingestion. Past medical history was significant for resection of distal duodenum to transverse colon secondary to midgut volvulus at three years of age. Physical exam was significant for horizontal nystagmus in lateral gaze bilaterally as well as vertical nystagmus with upgaze and mild dysmetria on finger-to-nose on the left. CBC, CMP and urinalysis on admission were within normal limits. A neurological cause was pursued due to the acuteness of the symptoms. Brain CT and subsequent MRI were performed and results were not consistent with an acute intracranial process. She had a normal opening pressure on lumbar puncture. Cerebral spinal fluid had a normal cell count and bacterial cultures, enterovirus and West Nile Virus PCR were negative. Fatty acid panel was normal and amino acid panel had non-specific elevations consistent with a non-fasting state. Vitamins B12 and E, folate, magnesium, zinc, copper, pyruvic acid, IGF-1, IGFBP-3 and acetylcholine receptor Ab were normal. Initial thiamine level was low at 43.3nmol/L (range: 66.5-200nmol/L). Thiamine supplementation was initiated and within 24 hours, neurological symptoms had resolved. Repeat thiamine level two days later was normal at 98.4nmol/L. She was discharged on 50mg of thiamine once daily for a total of 14 days, then 10mg daily for another month. After the treatment phase, she received every other day TPN multivitamin supplementation for two months (the maximum allowed due to the national shortage of intravenous multivitamins). Once the shortage ended, she resumed daily vitamin infusion. Thiamine is absorbed in the small intestine with the maximal absorption in the jejunum and ileum. Thiamine's half-life is approximately 10 to 20 days and due to limited tissue storage, continuous supplementation is required. Multivitamin solutions added to total parental nutrition become chemically unstable after 48 hours. Proper supplementation is therefore reliant on administration by compliant caregivers. Thiamine deficiency can occur as a complication of TPN if adequate supplementation is not provided. During the late 1990s, there were multiple reports of symptomatic thiamine deficiency among recipients of parenteral nutrition secondary to a widespread national shortage of parenteral multivitamins. To our knowledge, thiamine deficiency in TPN dependent Short Bowel Syndrome has not been reported with the recent multivitamin shortage. It is our aim that our case report will serve as a reminder to clinicians to check micronutrient levels periodically in this patient population.

488 INTRANASAL VITAMIN B12 SUPPLEMENTATION IN A CHILD.. T. Ciecieroga, NYP-Weill Cornell Medical College, New York, New York, UNITED STATES. V.L. Goh, Boston University/ Boston Medical Center, Boston, Massachusetts, UNITED STATES.

Introduction: Vitamin B12 (cobalamine) deficiency is a frequent finding in various pathologies. Standard of care is regular intramuscular injections. These can lead to pain, poor compliance and allergic reactions. There is minimal data on intranasal use of cobalamine in children. Case Description: A 7-year-old male presented with constipation and rectal prolapse. He was born at 24-weeks gestational age and developed necrotizing enterocolitis requiring small bowel resection. He had 2/3 of his jejunum and 8cm ileum intact, including IC valve and the colon. He was weaned off parenteral nutrition at 10 months of age but lost to follow up. He was re-referred to the pediatric gastroenterologist for his constipation. Laboratory assessment revealed low vitamin B12 level <146pg/ml (213-816pg/ml) and elevated methylmalonic acid level (MMA) 1080nmol/L (87-318nmol/L). He was started on cyanocobalamine intramuscular 1mg once monthly. As level stabilized, he was switched to Nascobal 500mcg intranasal every 2 weeks. Three months later, he continues to have a stable level of Vitamin B12 574pg/ml and MMA 117nmol/L. His constipation is as well controlled with minimal polyethylene glycol3350 with no recurrence of rectal prolapse. Discussion: Many patients are affected by vitamin B12 deficiency. Standard of care is intramuscular injection supplementation. Few reports showed that intranasal cobalamine is better tolerated and has good safety profile in adults. There is no such reports in children. We have successfully treated and maintained appropriate levels of vitamin B12 in a child by using intranasal supplementation. Intranasal cobalamine should be considered as an option for use in children with chronic vitamin B12 deficiency.

ASSESSMENT OF VITAMIN B12 STATUS IN PEDIATRIC INTESTINAL FAILURE PATIENTS: IS METHYLMALONIC ACID A RELIABLE BIOMARKER?. B. Tellier, C. Duggan, Division of Gastroenterology, Hepatology and Nutrition, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES. D. Stamm, Department of Surgery, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES. D. Stamm, B. Tellier, C. Duggan, Center for Advanced Intestinal Rehabilitation, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES. Purpose: To describe the relationship between serum methylmalonic acid (MMA) concentrations and exposure to enteral antibiotics in pediatric intestinal failure patients being treated for vitamin B12 deficiency. Methods: A retrospective chart review was performed on 3 children with intestinal failure and a history of terminal ileal resection. All were on stable regimens of parenteral cyanocobalamin as treatment for previous biochemical evidence of vitamin B12 deficiency. Results: Patient 1 was a 7 year old girl with segmental volvulus and ileocolonic anastomosis. Patient 2 was a 4 year old boy with necrotizing enterocolitis and an ileocolonic anastomosis. Patient 3 was a 3 year old girl with necrotizing enterocolitis with a jejunocolonic anastomosis. All had terminal ileum resection, none were receiving acid blockade, and none had clinical evidence of small bowel bacterial overgrowth. All had previously been started on parenteral cyanocobalamin in response to biochemical evidence of vitamin B12 deficiency, with subsequent improvement in biochemical measures of vitamin B12 status. All 3 later developed increased MMA concentrations in the setting of normal serum B12 and total homocysteine (tHcy) concentrations. They were treated with metronidazole (10 mg/kg/dose twice daily for 7 days) and underwent repeat laboratory testing within 2 weeks. Table 1 shows serum markers of vitamin B12 status before and after enteral antibiotics. Abnormal elevations in MMA (with concurrently normal levels of vitamin B12 and tHcy) were noted in each patient, and resolved following metronidazole treatment. Conclusions: MMA may not be a reliable marker of vitamin B12 status in pediatric intestinal failure. The use of enteral antibiotics to reduce gut bacterial load should be considered when evaluating patients with elevated MMA in this population, even in those without overt signs of small bowel bacterial overgrowth.

Serum Markers of Vitamin B12 Status Before and After Treatment with Metronidazole

EPIDEMIOLOGICAL AND CLINICAL CHARACTERISTICS OF CHILDREN WITH NEWLY DIAGNOSED IBD IN THE CENTRAL VALLEY, CA. J. Brock, C. Huang, Pediatrics, UCSF Fresno, Fresno, California, UNITED STATES. R. Gugig, A. Hunter, C. Huang, Pediatric Gastroenterology, Hepatology and Nutrition, Children's Hospital of Central California, Madera, California, UNITED STATES. J. Brock, Pediatrics, Children's Hospital of Central California, Madera, California, UNITED STATES. R. Gugig, University of California, San Francisco, San Francisco, California, UNITED STATES. Background Inflammatory Bowel Disease (IBD) encompasses a group of idiopathic, chronic, relapsing conditions characterized by inflammatory injury to the gastrointestinal tract. Crohn's disease (CD), Ulcerative Colitis (UC), and Indeterminate Colitis (IC) are the major classified types of IBD. The prevalence of early onset IBD has been steadily increasing; approximately 30% of patients with this condition are diagnosed in childhood. Prior studies have demonstrated early onset CD to be twice as common as UC. Increased incidence of UC among Hispanic children has also been reported. To our knowledge this is the first pediatric IBD epidemiological study done in the Central Valley, CA. Children's Hospital Central California (CHCC), located in Madera, CA, is a major referral center for a pediatric population with diverse cultural and ethnic backgrounds. Methods A retrospective chart review of 105 children diagnosed with IBD between 2011 and 2014 was performed. Patients less than 21 years old were included. A spreadsheet coded for standardization of predetermined clinical and demographic parameters allowed for data analysis. Statistical analysis including mean, median, range, standard deviation, percent, 95% confidence interval were calculated. The data was compiled and analyzed using t-test and chi-square analysis. Our goal is to demonstrate the epidemiological characteristics of IBD incidence in the Central Valley, CA as well as interesting data of multicultural diversity that may impact the incidence of IBD in this region. Results Out of 105 patients, 61.9% (65) were found to have UC, 34.3% (36) were found to have CD and 3.8% (4) IC. 55 patients were male (52%) and 50 female (48%). The majority of our patients were of Caucasian descent 54% (56) followed by children of Latino heritage 35% (37) and patients of Asian heritage 11% (12). Our results showed a statistically significant increase of incidence of UC vs CD among Hispanic patients (75% vs 21%). The mean age of diagnosis among all IBD patients at the Central Valley was 11.7 years. 57% of Hispanic children with UC were diagnosed before age 12 versus 25% of children with UC of Caucasian background. Conclusions In the Central Valley, CA, the most common subtype of IBD is UC compared to CD disease (62% vs 34%). The UC phenotype of IBD is twice as common as CD

among the Hispanic patients. Surprisingly, 57% of Latino children with UC were diagnosed before the age of 12 years. Further epidemiological studies will be required to better identify the disease characteristics of IBD in the Central Valley, CA.

IBD TYPE BY ETHNICITY

UC: Ulcerative Colitis, CD: Crohn's Disease, IC: Indeterminate Colitis