

# Clinical Vignette Abstracts

## NASPGHAN 2017

Poster Session I  
Thursday, November 2, 2017  
5:00pm – 7:00pm

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**1 SOLITARY RECTAL ULCER SYNDROME IN A TEENAGER REQUIRING MULTIPLE BLOOD TRANSFUSIONS.** A.M. McClain, J. Pohl, R.A. Patel, A. Lowichik, K. Jensen, Pediatric Gastroenterology, Hepatology & Nutrition, University of Utah, Salt Lake City, Utah, UNITED STATES.

A previously healthy 17-year-old female presented to an outside hospital after two days of bloody diarrhea, stooling urgency and rectal bleeding. She was found to have a hemoglobin of 6 g/dL and received a blood transfusion. Over the next 8 months, similar symptoms occurred four more times, requiring two more blood transfusions. The patient was referred to our GI clinic and underwent an extensive evaluation including upper endoscopy, colonoscopy on two separate occasions, Meckel's scan, capsule endoscopy and CT angiography. On initial colonoscopy she had a sessile appearing lesion on the left wall of the rectum. Pathology of the rectal lesion showed replacement of lamina propria by collagen and smooth muscle bundles and distortion of the crypts, consistent with solitary rectal ulcer syndrome [SRUS]. She was started on a bowel program. Due to continued bleeding, CT angiography was done and showed nonspecific thickening of the left lateral sidewall of rectum with increased vascularity. Repeat colonoscopy seven months later demonstrated a deep rectal ulcer on the left side of rectum; pathology showed hyperplastic epithelium and crypt distortion again with fibrosis in the lamina propria. Surgical consultation believed this large, worsening ulcer was the cause of recurrent bleeding. Surgical resection at that time was felt to be extremely challenging and a more intensive laxative bowel program recommended with surgical follow up for possible defecography. SRUS has been well described in the adult literature with several theories to explain the pathophysiology. Pediatric cases of SRUS are rare with less than 200 reported cases in previous case reports. Diagnosis usually is through endoscopic visualization and biopsy and treatment ranges from conservative treatment with laxatives and biofeedback to surgical resection, rectopexy or fecal diversion. Our case is unusual in that the patient had recurrent anemia requiring blood transfusions. Most literature regarding SRUS report infrequent need for blood transfusion.

**2 OBSCURE GI BLEEDING SECONDARY TO GASTRIC HETEROTOPIA IN A CHILD WITH SEVERE IRON DEFICIENCY ANEMIA.** A.A. Shah, A. Kastl, B. Godwin, P. Mamula, K. Fiorino, Gastroenterology, Hepatology, & Nutrition, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES.

Introduction: Ectopic gastric mucosa, which can be either congenital or acquired, has been reported throughout the entire alimentary tract. It is most commonly encountered in the form of either Meckel's diverticulae or duplication cysts and is typically symptomatic leading to obstruction, intussusception, and gastrointestinal bleeding. Case: A 7 year old girl with history of neonatal small bowel obstruction secondary to a jejunal web s/p partial jejunal resection with tapering jejunoplasty and primary reanastomosis presented to the ED with three weeks of headaches, shortness of breath, and lethargy. Upon evaluation, her heart rate was 214 beats per minute with EKG showing supraventricular tachycardia, and she was found to have severe iron deficiency anemia with Hgb 2.3 g/dl, reticulocyte count 0.2%, MCV 59 fL, and iron 11 mcg/dL. Patient stabilized after pRBC transfusions, which also resolved her SVT. She then underwent testing with CXR, FOBT, stool studies, and laboratory studies including viral testing all of which were unremarkable and was discharged home on iron supplementation without a conclusive diagnosis. However, since her anemia persisted, she underwent a thorough GI evaluation. Her initial UGI with small bowel follow-through, EGD, and colonoscopy were completely normal. Push enteroscopy visualized edema, ulceration, friability, and exudates within the jejunum with biopsies consistent with fragments of small bowel mucosa intermixed with extensive fields of gastric epithelium but no ulcers. MRE demonstrated small bowel thickening in the proximal jejunum. Video capsule endoscopy visualized abnormal jejunal mucosa with edema, fissuring, and fresh blood seen. Meckel's scan showed ectopic gastric mucosa throughout the abdomen suggestive of multiple enteric duplication cysts. After another hospitalization for symptomatic anemia, our patient underwent exploratory laparotomy with intra-operative push enteroscopy culminating in resection of 25 cm of jejunum with primary reanastomosis. Pathology illustrated multifocal polypoid gastric heterotopia with reactive gastropathy

and serosa with focal areas of hemorrhage. Since resection over one year ago, she has been asymptomatic with stable hemoglobin and no other GI complaints. Conclusion: The differential diagnosis of anemia in school-aged children is broad but includes multiple gastrointestinal etiologies of blood loss. Ectopic gastric mucosa such as Meckel's diverticulum or duplication cysts must be considered, particularly if initial evaluation is non-diagnostic, and in rare cases, such as this instance, may affect an extensive portion of the alimentary tract. Once identified, surgical treatment may be indicated, especially if symptomatic.

**6 TREATMENT OF PORTAL VENOUS GAS EMBOLISM WITH HYPERBARIC OXYGEN AFTER ACCIDENTAL INGESTION OF HYDROGEN PEROXIDE: A CASE REPORT.** S. Ali, Pediatric Gastroenterology, Stanford Children's Health, Walnut Creek, California, UNITED STATES. J. Davis, Hyperbaric Medicine, John Muir Medical Center, Walnut Creek, California, UNITED STATES. Yeh, K. Haas, Pediatric Gastroenterology, Stanford University, Stanford, California, UNITED STATES.

4 year old female with no prior medical history was brought in by her parents to the emergency department with complaint of vomiting after an accidental ingestion of concentrated hydrogen peroxide (35% H<sub>2</sub>O<sub>2</sub>). Within a few minutes after ingestion patient had non bloody non bilious emesis. She was taken to the emergency department (ED) where an urgent CT scan with contrast was completed that showed marked portal venous air throughout the liver and pneumatosis (figure 1) and hyperemia of the gastric wall. Also had scattered foci of air in the submucosal layer of the lower esophageal sphincter (LES) and at gastroesophageal junction to about 3cm above the LES. No evidence of perforation. CT of head showed no evidence of hemorrhage or infarction. Patient had 2 episodes of hematemesis in the ED. She was alert and oriented and well-appearing. Vital signs were normal. Abdomen on exam was soft and non-tender. Oral pharynx with two small lesions on the posterior hard palate. Neurologic exam was normal. Laboratory data included complete blood count and comprehensive metabolic panel that were unremarkable. The patient was transferred to the closest hospital with an available hyperbaric oxygen (HBO) chamber for treatment of the portal venous air emboli. Patient underwent emergent HBO treatment followed by three subsequent HBO treatments. Upper GI study showed no evidence of esophageal and gastric abnormality. Abdominal Ultrasound on second day of hospitalization showed complete resolution of the portal venous air. Patient did well during the treatment with symptom resolution. Diet was advanced slowly and was discharged home on day 7 on continued acid blockade therapy. Conclusion: It is well known that hydrogen peroxide ingestion can cause gas embolism. This is a case report of the successful use of hyperbaric oxygen therapy to treat portal venous gas embolism caused by hydrogen peroxide ingestion in a pediatric patient. Hyperbaric oxygen therapy can be considered for the treatment of symptomatic hydrogen peroxide ingestion.

**7 GASTRIC HYPERPLASTIC POLYPS CAUSING UPPER GASTROINTESTINAL BLEED POST CARDIAC TRANSPLANT.** A.E. Ferguson, D.S. Vitale, P. Putnam, S. Kocoshis, K. El-Chammas, Gastroenterology, Cincinnati Children's Hospital, Cincinnati, Ohio, UNITED STATES. K.E. Bove, Pathology, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, UNITED STATES.

Background: Hyperplastic gastric polyps are a rare complication in post-transplant patients. Significant bleeding caused by multiple gastric hyperplastic polyps, however, has not been reported in pediatric post-transplant patients. A case series published in 2002 described acquired hyperplastic polyps in solid organ transplant recipients, but most patients were asymptomatic or presented with mild symptoms such as nausea. Our case highlights the unusual occurrence of multiple gastric hyperplastic polyps causing an upper GI bleed in a post cardiac transplant patient, reviews the histopathologic findings of these polyps, and discusses the gaps in knowledge open for future research. Case Presentation: A 15 year-old male with history of acute myeloid leukemia (AML) in remission, chemotherapy-induced cardiomyopathy status post cardiac transplant 5 years prior, renal insufficiency, pulmonary hypertension, and autism presented to the emergency room with acute onset of hematemesis. He denied blood per rectum or melena. He had no fevers or upper respiratory symptoms, and he was not on anticoagulation. His medications included prednisone and cellcept for immunosuppression, and daily prevacid. Patient had a previous endoscopy 3 years prior for failure to thrive that identified multiple hyperplastic gastric polyps of various sizes, two of which were excised. Labs indicated hemoglobin of 7.5, decreased from 10.0 one-month prior to presentation. Upper endoscopy identified a large clot burden in stomach and distal esophagus with multiple pedunculated polyps in the stomach and at the gastro-esophageal junction (GEJ). Nine polyps between less than 5 mm and 2.5 cm in size were removed with hot snare. Other than the identified polyp at GE junction, the esophageal lining was normal in appearance. The remainder of gastric mucosa was with a few small

polyps that were not removed. The duodenal mucosa was normal in appearance. The patient was admitted to the CICU where he ultimately did well without further bleeding. Discussion: Our patient is the first pediatric case report of multiple hyperplastic polyps post-transplant causing a significant bleed. Since hyperplastic polyps can, albeit rarely, show malignant potential, it is important to be aware of this issue in our post solid organ transplant patients. Although one, small retrospective study found no histological or clinical differences between the polyps of transplanted patients compared to non-transplanted patients, this is the only study published to date. Our case highlights the need for further investigation of the natural history and development of hyperplastic polyps in the post solid organ transplant population.

**9 PENNY FOR YOUR THOUGHTS: A COIN IN THE STOMACH - WHY DID IT GET STUCK?** A. Shakir, Pediatrics Gastroenterology, Hepatology and Nutrition, University of Oklahoma Health Science Center, Oklahoma City, Oklahoma, UNITED STATES.

A well-appearing 11-year-old with Down syndrome presented to the ED with back pain. The patient denied any symptoms of abdominal pain, early satiety, or vomiting. The physical exam and initial laboratory work-up were negative. A KUB showed levoscoliosis and a radio-opaque foreign body in the area of the stomach (image 1). An elective EGD was performed the next day which revealed a stomach full of food contents, a dilated pylorus, and a disintegrated penny in the first part of the duodenum. An UGI series performed after the EGD showed a duodenal stricture (image 2). The stricture was relieved by surgery electively. Children with Down syndrome are more likely to have congenital intestinal anomalies, however they usually present in the first few weeks of life. This case highlights a unique, delayed presentation of duodenal stricture in an 11-year-old with Down syndrome with an incidental finding of a radio-opaque foreign body.

**10 INCREASING NUMBER OF BIOPSIES INCREASES CHANCES TO ESTABLISH ACCURATE DIAGNOSIS IN PEDIATRIC ADENOCARCINOMA.** B. Younes, Gastroenterology Hepatology and Nutrition, Children's Hospital Orange County, Orange, California, UNITED STATES.

Increasing number of biopsies increases chances to establish accurate diagnosis in pediatric adenocarcinoma BY B.S. Younes, A.K. Sassoon, J. Ho, K. Grant, M.H. Katz Division of pediatric gastroenterology, hepatology and Nutrition Children's hospital of Orange County Background Adenocarcinoma of the colon and rectum is the most common cancer of the GI tract and carries the second highest cancer mortality rate. Approximately 150,000 new cases are diagnosed annually in the United States; less than 1% of which (approximately 80 cases per year) occur in patients younger than 20 years of age. (1) In children, colorectal cancer is the second most common cancer of the alimentary tract after liver tumors; the incidence of colorectal cancer is 1.3-2 cases per million population. Most patients present during the second decade of life. The youngest patient ever reported was 9 months of age. Children are more likely than adults to have advanced-stage at presentation, as well as unfavorable tumor histology (mucinous), which confers them a poor outcome. Six to eight biopsies are recommended to establish the diagnosis of malignant lesion (2,4). Choi et al in most recently published study in the Journal of Korean medical science suggested 4 biopsies from the affected area are enough to establish the diagnosis of colonic adenocarcinoma. They reasoned that more biopsies increased risk of bleeding, prolonged procedure time and increased work load on pathologist. (3) Approach Figure-1A seventeen year old girl previously healthy developed sudden onset of abdominal pain and vomiting after a trip to Brazil. CT scan of the abdomen showed colonic thickening, edema suggesting colitis. Initially she passed loose stools, but eventually developed obstructive picture with persistent bilious emesis, abdominal distension and no stooling. Barium enema (Figure 1) showed bowel obstruction at the sigmoid level. The first colonoscopy was performed and showed an obstructive lesion (figure 3), the colonoscope could not be advanced beyond that point, 4 biopsies were taken from the affected sigmoid area. Biopsies showed normal looking colonic mucosa. (Figure 2,4) A few days later, a second colonoscopy was performed with 8 biopsies from the affected lesion of the sigmoid colon and showed mucinous adenocarcinoma of the colon (figure 5,6) The obstructive lesion was resected during surgery (figure 6) and colostomy was performed Histological findings: Primary Tumor Site: Sigmoid colon Mucinous adenocarcinoma (figure 5,6,7,8) Histologic Grade: Low-grade (well differentiated to moderately differentiated) Mucinous tumor component: Percentage of Mucinous Tumor: 60% Immunohistochemistry Studies for Mismatch Repair Proteins: MLH1, MSH2, MSH6, PMS2 Figure -2 Genetics: Genetic testing was performed and revealed a heterozygous mutation in the MUTYH gene and thus the patient was technically a CARRIER for the condition, rather than affected. However, review of the medical literature has revealed evidence for increased risk of colorectal cancer in patients with

heterozygous MUTYH mutations compared to the general population. More extensive genetic testing for the family is underway. Conclusion Endoscopic biopsy is the gold standard to confirm the histopathologic diagnosis of colon cancer. The number of biopsies needed for the diagnosis is not established, although 6-8 biopsies are usually recommended (2,4). Our first colonoscopy with only four biopsies from the lesions failed to confirm the diagnosis. The repeat colonoscopy with 8 biopsies confirmed the presence of colonic adenocarcinoma. We suggest that increasing the number of biopsies to eight taken from the affected lesion will increase the likelihood of establishing the diagnosis of colonic adenocarcinoma, thus decreasing the need for repeat colonoscopies without increasing the risks of the procedure or the workload for the pathologist.

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4) Lal N, Bhasin DK, Malik AK, Gupta NM, Singh K, Mehta SK. (4) Optimal number of biopsy specimens in the diagnosis of carcinoma of the oesophagus. *Gut.* 1992;33:724-726

**11 COLONIC CARCINOID TUMOR.** B. Kuhn, Pediatric Gastroenterology & Nutrition, Geisinger Health System, Danville, Pennsylvania, UNITED STATES. J. Ramdas, Pediatric Hematology & Oncology, Geisinger Health System, Danville, Pennsylvania, UNITED STATES.

Intro Carcinoid Tumors are well-differentiated neuroendocrine tumors. They are generally found in the digestive tract and lungs. The most common location in the digestive tract is the appendix. In the United States, most recent estimates indicate Carcinoid Tumor incidence and prevalence approaching 5 per 100,000.

Case A 15-year old female presented for consultation regarding 6 months of progressive abdominal pain and diarrhea. Her symptoms were characterized as 8-12 urgent loose brown stools. She had rare night time awakening with abdominal pain and urge to defecate. Fecal soiling occurred if without access to a restroom. No overt blood or mucous in the stool. Her abdominal pain was bilateral lower quadrant cramping. Review of symptoms: negative for fevers, vomiting, or weight loss. No extra intestinal inflammatory bowel disease manifestations. No previous medication. Physical examination was notable for bilateral lower quadrant tenderness to palpation. Hemocult was negative on digital rectal examination.

Initial Work-Up: NORMAL Blood: CBC/diff, ESR, CRP, CMP, Lipase, TTG IgA, Total IgA, TSH, Free T4  
Stool: Comprehensive Stool Culture, Ova & Parasite, Giardia/Cryptosporidium, C. Difficile.  
Urine: Urinalysis, Urine Culture, bHCG.  
Imaging: Complete Abdominal Ultrasound. Upper Endoscopy with Biopsies: NORMAL.  
Colonoscopy with Biopsies and Polypectomy: 5 mm rectal pedunculated polyp (image 1). Resection was completed by electrocautery snare technique (image 2). Pathology: CARCINOID TUMOR with clear margins. Depth: invading the lamina propria. Synaptophysin Immunostain: positive for tumor cells. Mitotic Rate < 1 per 10 HPF. Ki-67 Immunostain Index < 2% (Proliferation Rate). No cytologic atypia. Findings qualify this tumor as a low grade (Grade 1) Neuroendocrine Tumor: CARCINOID.

Follow-Up Evaluation: 2 weeks after resection 24-hour Urine 5-HIAA and Serum Chromogranin A (Carcinoid Markers) = NORMAL. CBC/diff and ESR = NORMAL. CT Abd/Pelvis with Oral and IV Contrast: Right 1.4 cm Adnexal Mass (ultimately determined to be Follicular Cyst). Otherwise, no lymphadenopathy or other organ lesions. Follow-up pelvic ultrasound: resolved follicular cyst.

Long Term Follow-Up: 6 months after resection Serum Chromogranin A (Carcinoid Marker) = NORMAL. CBC/diff and ESR = NORMAL. Complete Abdominal Ultrasound and Pelvic Ultrasound = NORMAL. Upper Endoscopy and Colonoscopy with Biopsies = NORMAL.

Discussion Carcinoid Tumors are rare neuroendocrine tumors typically found in the appendix. Extra-appendiceal colonic tumors are generally found in the right colon (70%) or rectum. Colonic Carcinoid Tumors most often present with nonspecific abdominal symptoms, such as abdominal pain/cramping and intermittent diarrhea; however, some may be asymptomatic. Based on published Colonic Carcinoid Tumor lymph node metastasis rates, our patient possesses a 4% likelihood of metastasis in that her tumor was < 1 cm and located in the submucosa only. Endoscopic resection of Colonic Carcinoid Tumors < 1 cm in size, confined to the mucosa, with clear margins, no cytological atypia, low mitotic rate, and low cell proliferation rate (Ki-67) may not require additional work-up. However, persistent symptoms or clinical concern, may prompt further work-up as indicated. This patient did well with post-resection monitoring of Carcinoid Tumor Markers (Serum Chromogranin A and/or 24-hour Urine 5-HIAA), with overall reduction in abdominal pain and diarrhea.

**12 ENDOSCOPIC FOREIGN BODY REMOVAL WITH AN EXTERNAL DEFIBRILLATOR MAGNET.** B. Waddell, S. Deivanayagam, Pediatrics, Peyton Manning Childrens Hospital, Carmel, Indiana, UNITED STATES.

Introduction: Foreign body ingestion in pediatrics is a challenging clinical problem particularly if the object ingested is sharp, a battery, or magnetic. Removing these objects via Endoscopy is often the treatment of choice if still in the esophagus or stomach but can be quite challenging if the stomach is full of food and other contents. To address this problem we report two successful magnetic foreign body removals by applying a defibrillator magnet to the outside of the abdominal wall. This causes magnetic objects to be drawn toward the anterior portion of the stomach, away from other stomach contents, and then visualized and removed. Case report 1: A 12-year-old previously healthy female presented to the ED after accidental ingestion of a metal nail 5 hours earlier. Radiographic examination demonstrated a sharp radiopaque object in the stomach. She was taken promptly for EGD removal. After introducing the endoscope into the stomach, the field of view was obscured by food. Despite multiple attempts at irrigation and suctioning the metallic object could not be visualized. C-Arm Fluoroscopy confirmed the nail in the stomach. An external defibrillator magnet, typically used to turn off pacemakers, was then applied to the outside of the abdominal wall. The metallic nail was then drawn away from the food mixture toward the magnet where it was visualized and removed with a Roth Basket. She had no procedural complications and was discharged home. Case Report 2: A 6-year-old previously healthy female presented to the ED after accidental ingestion of a button battery approximately 3 hours prior. Physical exam was unremarkable. Radiographic examination confirmed a small, round radiopaque object in the stomach and she was taken for EGD removal. The battery was embedded in a thick chymous mixture and could not be visualized. An external defibrillator magnet was applied to the abdominal wall and drew the foreign body out of the chymus mixture towards the antrum of the stomach. The battery was then visualized and removed with a Roth basket. There were no procedural complications. Conclusion: Removing ingested foreign bodies from the stomach is dependent upon direct visualization, which can be hindered by the presence of food. Here, we present a novel approach to magnetic foreign body removal by using an external defibrillator magnet, readily available in hospitals, to aid in the visualization, manipulation, and removal of magnetic foreign bodies.

**15 COLONOSCOPY ASSISTED CECOSTOMY TUBE REPLACEMENT TO SALVAGE LOST CECOSTOMY TRACT ACCESS IN CHILDREN.** C. Dike, R. Rahhal, Pediatrics, University of Iowa, Iowa City, Iowa, UNITED STATES.

INTRODUCTION Children with spinal defects and anorectal malformations are predisposed to chronic constipation with or without fecal incontinence. Medical management with oral laxatives often fails. A cecostomy tube for antegrade enemas is usually required to address defecation problems. Once surgically placed, cecostomy tubes require changes by percutaneous approach, occasionally with fluoroscopic guidance however this may be unsuccessful requiring repeat laparoscopy or open surgery to re-establish the cecostomy tract. Some adult studies have reported good outcomes with colonoscopy assisted cecostomy tube placement but only few studies have been done in children. The role of colonoscopy assistance to salvage lost cecostomy access in children who fail percutaneous replacement is not well described. We hypothesize that a colonoscopy assisted approach is safe and effective to re-establish lost cecostomy access in children. METHODS A retrospective cohort study of the methods, success and complication rates and durability associated with cecostomy tube replacement in children were studied from May 2000 to May 2017 with focus on the colonoscopy assisted approach. RESULTS Our preliminary data identified 6 colonoscopy assisted cecostomy tube replacement procedures in 5 patients (average age 10.3 years, average weight 28.3 kg, 67% female gender). All patients had underlying myelomeningocele with neurogenic bowel. The most common reason for using colonoscopy assistance is failed percutaneous cecostomy tube replacement. The colonoscopy assisted approach was successful in all cases without documented complications. CONCLUSION Colonoscopy assisted cecostomy tube replacement is safe and effective in re-establishing lost cecostomy access in children after failed attempts with percutaneous or fluoroscopic guided approaches.

**17 APPENDICITIS: A POSSIBLE COMPLICATION OF COLONOSCOPY.** C. LeBlanc, A. Russell, Pediatric Gastroenterology, Vanderbilt University Medical Center, Nashville, Tennessee, UNITED STATES. Wang, Vanderbilt University, Nashville, Tennessee, UNITED STATES.

Introduction: Colonoscopy is a common and safe procedure performed in the pediatric population. Some of the most common indications include abdominal pain, hematochezia and diarrhea.<sup>1</sup> The complication rate is low and has been estimated at 1.1%.<sup>2</sup> Here we present a case of appendicitis in a pediatric patient within 24 hours of colonoscopy. Case Report: A 13-year-old male with a history of autism spectrum disorder (ASD) and chronic abdominal pain presented to our pediatric gastroenterology clinic in June 2016 for evaluation of

his pain. Previous work-up included a normal CBC, BMP, CRP and CT abdomen. At the GI clinic visit, family stated that his pain was daily in the right lower quadrant (RLQ). It was made worse with meals, sometimes caused nausea and vomiting, and occasionally woke him up from sleep at night. He recently developed loose stools with mucous. He denied any other concerning symptoms, and was initially managed conservatively; however, he was scheduled for EGD and colonoscopy 4 months after his presentation due to persistent pain. Both scopes were grossly normal, and he was discharged home after recovery from sedation and tolerating food by mouth. The following day, he passed a bloody bowel movement and developed severe RLQ pain with nausea. His mother brought him to the ED where he was found to have an elevated white blood cell count (11.2, ref 3.2-10.2) and CRP (26.9, ref 0.1-1.0). A CT abdomen showed acute appendicitis without evidence of perforation. He was admitted to the general surgery team who performed an appendectomy. He was discharged home on post-op day 1 after tolerating a regular diet and pain well controlled.

**Discussion:** Colonoscopy is a low-risk procedure in the pediatric population, with an immediate complication rate estimated at 1.1%.<sup>2</sup> Over half of the complications (56.8%) are related to the GI tract with gastrointestinal bleeding being the most common.<sup>2</sup> Acute appendicitis following colonoscopy is a very rare complication reported in the adult literature with an estimated incidence of 0.038%.<sup>3</sup> To date, only 37 cases of post-colonoscopy appendicitis have been described since first reported in 1988, and all cases have occurred in adults.<sup>4,5</sup> Over half the cases (68.4%) showed symptoms within the 24 hours following the procedure, suggesting an etiological relationship.<sup>5</sup> Acute appendicitis after colonoscopy has been attributed to several mechanisms including: preexisting subclinical appendicitis, introduction of feces into the appendix, trauma to the appendix by the colonoscope, or insufflation of the appendix during the procedure.<sup>6</sup> Patients with post-colonoscopy appendicitis can present with RLQ pain, diffuse abdominal pain, and peritonitis.<sup>3,7</sup> In this case, the patient presented with severe RLQ pain and nausea within 24 hours of his scope. As the rate of pediatric appendicitis in children age 10-14 is low (169.6 cases/100,000), it seems likely that these two events were related in this patient.<sup>8</sup> However, the exact cause of his appendicitis is unclear. The appendix was not intubated during the procedure making trauma, over-insufflation or introduction of a fecalith unlikely, and clinical exam prior to the scope was not concerning for a pre-existing appendicitis. This first reported pediatric case of appendicitis following colonoscopy indicates that clinicians must be mindful of this complication.

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**18 "JINGLE ALL THE WAY!"- FOREIGN BODIES EMBEDDED WITHIN THE ESOPHAGEAL MUCOSA DURING THE HOLIDAY SEASON.** C. Maier, M.S. Rosario, Internal medicine and pediatrics, University of South Florida, Tampa, Florida, UNITED STATES. V. Falconer, S. Son, University of Florida, Gainesville, Florida, UNITED STATES. E.K. Swan, A. Wilsey, Florida State University, Tallahassee, Florida, UNITED STATES. Wilsey, Pediatrics, University of South Florida, Tampa, Florida, UNITED STATES. Wilsey, Pediatric Gastroenterology, Johns Hopkins All Children's Hospital, St. Petersburg, Florida, UNITED STATES.

**Background:** Foreign bodies (FB) caught in the esophagus can lead to mucosal ulcerations, infections and other severe complications. Prior to the availability of endoscopy, morbidity and mortality for ingestion of sharp objects were 35% and 26% respectively. Guidelines for several categories of objects, including sharp ones, dictate whether emergent removal or close follow up is warranted. Ultimately, endoscopic removal occurs in only 20% of foreign body ingestion (FBI) cases. We present and discuss management of two cases of relatively sharp holiday foreign bodies embedded within the esophageal mucosa.

**Case Summaries:** First is a 30-month-old male who presented with three days of fever, cough, and rhinorrhea. His symptoms progressed to include sternal pain and refusal of solids, but not liquids. In the emergency department (ED), he was noted to have slight drooling on exam and chest X-rays

revealed a foreign body retained in the esophagus. Endoscopy with a GIF-160 scope revealed a jingle bell embedded within the esophageal mucosa. Vigorous manipulation was required to dislodge the jingle bell during endoscopic removal. However, minimal tearing was observed upon re-examination. Follow up esophogram showed no evidence of perforation. The patient was discharged the same day after tolerating oral intake. Second is a 10-month-old male who presented after choking in his play pen followed by significant drooling. A chest X-ray in the ED revealed a foreign body at the level of the thoracic inlet. ENT evaluation in the OR with rigid bronchoscopy identified a metal Christmas ornament hanger 1 cm distal to the opening of the esophagus. Unfortunately, the metal prongs of the ornament were fixed within the esophageal tissue. The mucosa was impinged by the metal top of the hanger, even after several attempts at removal with a rigid scope. Gastroenterology was emergently consulted for intraoperative assistance. The foreign body was forcefully dislodged off the esophageal mucosa, which prompted advancement into the stomach. The hanger was re-oriented, allowing the apex of the spring load to be grasped and pulled out in a retrograde fashion. The patient remained intubated post-procedure due to upper airway edema and swelling of the site. He was extubated successfully the next day and follow up esophogram with contrast showed no evidence of perforation. He was discharged later that evening. Discussion: We present two cases of FBI that both resulted in embedment of the objects in the esophageal mucosa. These cases highlight both the importance of emergent endoscopic removal and the challenges of endoscopy when a FB becomes embedded within the esophageal tissue. FBI may be asymptomatic in up to 50% of cases. However, impacted FBs can have complication rates of 18%, with esophageal perforation rates varying from 2 to 15%. Therefore, emergent investigation is warranted for patients in whom FB ingestion is suspected. This precaution prevents morbidity and mortality related to esophageal complications.

**21 THERAPEUTIC ERCP WITH DIRECT CHOLANGIOSCOPY IN A 23 MONTH OLD FEMALE WITH TYPE IV-A CHOLEDOCHAL CYST.** D. Freestone, A.S. Huang Pacheco, PEDGI, Nebraska Medical Center, Omaha, Nebraska, UNITED STATES. Schafer, Midwest GI, Omaha, Nebraska, UNITED STATES. Freestone, A.S. Huang Pacheco, M. Schafer, Children's Hospital of Omaha, Omaha, Nebraska, UNITED STATES.

Introduction: Choledochal Cysts (CC) are rare congenital biliary duct abnormalities. There are several different types. Type IV is when there is extra- and at least one intrahepatic cystic upstream dilation. There is an association with congenital heart defects. Cyst formation is likely related to anomalous pancreaticobiliary union (APBDU). Jaundice is present when APBDU allows reflux of bile and pancreatic secretions leading to stone formation. Diagnosis is made with abdominal US and MRCP. ERCP can play a role in identifying extent of ductal involvement and provide some therapeutic options, such as stone retraction, sphincterotomy, and stent placement. Direct cholangioscopy allows for direct visualization of dilated bile ducts to assess for stones, strictures, and other causes, and is not often available in pediatric patients due to their relative narrow bile duct caliber compared to adults. Definitive management of CC is typically surgical with the exception of type III cysts (choledochalcele). Complications of CC include cholangitis, liver failure, and malignancy. Case Report: A 23 month old female with history of surgically repaired VSD as an infant, otherwise in good health presented to Children's Hospital of Omaha with a 2 week history of jaundice, increasing fatigue, abdominal pain, and poor appetite. Labs were significant for conjugated bilirubinemia of 3.4, elevated alkaline phosphatase 1357, AST 585 and ALT 858. Elevated GGT 939, and elevated lipase to 6363. Abdominal US showed intra- and extrahepatic dilation with common bile duct of 10 mm without evidence of stone. MRCP also showed intra- and extrahepatic bile duct dilation, with APBDU formation and a filling defect in the distal common bile duct near the ampulla. ERCP was performed revealing a large stone obstructing the common bile duct at the ampulla. Sphincterotomy was performed and a large biliary stone was removed. Repeat cholangiogram showed continued areas of narrowing. Cholangioscopy was performed revealing no dilated bile ducts without evidence of stricture or additional stones and otherwise normal bile duct appearance. Finally a biliary duct stent was placed. ERCP was tolerated very well. Repeat labs the following day showed resolution of hyperbilirubinemia and elevated lipase, and significant improvement of in transaminases. Comments: There is an association with CC and congenital heart defect. Dilated bile duct allowed for safe use of through the scope direct cholangioscopy. To our knowledge, this is the first case of direct cholangioscopy in a 23 month old patient. Findings were most consistent with type IVa choledochal cyst. Repeat abdominal US vs MRCP will reevaluate for persistent intrahepatic ductal dilation which will differentiate type IVa and Ic choledochal cyst. Definitive management for type IVa CC is different in that in addition to extrahepatic bile duct resection, regions of liver with intrahepatic bile duct dilation also need to be resected to minimize malignant potential.

### **23 JUVENILE POLYP ARISING FROM THE APPENDICEAL MUCOSA FOLLOWING INVERSION-LIGATION**

**APPENDECTOMY.** D. Sankepalli, A. Khalili, T. Sferra, Pediatric Gastroenterology, University Hospitals Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES. Barksdale, Pediatric Surgery, University Hospitals Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES.

Introduction: Colonic polyps during childhood infrequently occur within the cecum and rarely involve appendiceal orifice. We cared for a child who underwent an inversion appendectomy during a Ladd's procedure and later presented with hematochezia from a juvenile polyp involving the appendiceal stump. Case Report: A 2 year-old female with prior history of malrotation as an infant for which she underwent a Ladd's procedure presented with abdominal pain and hematochezia. As an infant she had a history of cow's milk protein intolerance (CMPI), Gastroesophageal reflux and intermittent emesis. An initial upper GI study demonstrated duodenum inversum, however malrotation could not be excluded. The upper GI study was repeated 3 months later and had findings concerning for malrotation of the small bowel. At 7 months of age, she underwent diagnostic laparoscopy with Ladd's procedure and inversion-ligation appendectomy. After surgery, she continued to be followed by pediatric gastroenterology. She had intermittent diarrhea that improved on a lactose-free and juice-restricted diet. She was otherwise well until she developed abdominal pain and persistent hematochezia. At colonoscopy, she was found to have a pedunculated polyp within the cecum at the location of appendiceal orifice. It was believed to represent a polyp arising from the inverted appendix that did not undergo complete necrosis following the original surgery. The polyp was resected by hot snare. Histopathology of the tissue was consistent with a juvenile polyp. Her symptoms resolved following the polypectomy. Discussion: Lower gastrointestinal bleeding is a common presentation of colonic polyps in children. In the pediatric population, polyps usually have hamartomatous histological appearance without adenomatous dysplasia. The terms used to describe such polyps are juvenile, retentive and inflammatory polyp. The expectation following the inversion-ligation appendectomy is that the appendix undergoes necrosis and is sloughed into the colonic lumen. However, retention of the appendix or a small stump can occur. This is usually asymptomatic but can serve as a lead point for intussusception. Also, it can pose a diagnostic challenge if visualized during radiologic imaging or during colonoscopy, especially if the previous procedure is unrecognized. In this case, a juvenile polyp developed within the appendiceal mucosa. Also in our case, it is speculative, but possible that the child's abdominal pain was due to intermittent intussusception as the pain resolved following the polypectomy. We are not aware of another reported case of a juvenile polyp involving an inverted appendix.

### **24 A RARE FINDING IN AN OTHERWISE ROUTINE COLONOSCOPY: SOLITARY GANGLIONEUROMA OF THE**

**GASTROINTESTINAL TRACT.** D. Do, Pediatrics, University Hospitals / Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES. Sankararaman, J. Moses, V. Baez-Socorro, Pediatric Gastroenterology, University Hospitals / Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES. Saab, Pathology, University Hospitals / Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES.

Ganglioneuromas are slow growing well-differentiated, benign tumors arising from peripheral nerves. They are composed of Schwann cells and ganglion cells. When they arise in the gastrointestinal tract (GIT), they are predominantly found in the descending colon. Case Report: A 17-year-old otherwise healthy female presented with burning epigastric pain that was exacerbated by eating and cramping lower abdominal pain that improved with defecation. She had daily bowel movements that were loosely formed and non-bloody. At presentation, she had been receiving esomeprazole for 2 weeks without significant improvement in symptoms. Physical examination was significant for epigastric tenderness. Laboratory studies were normal, including a complete blood count, complete metabolic panel, celiac serology, and C-reactive protein. Due to her persistent abdominal pain, esophagogastroduodenoscopy (EGD) and colonoscopy were performed. The EGD was normal. The colonoscopy was significant for two small, non-bleeding, pedunculated polyps in the sigmoid and descending colon. Both were less than 0.5 cm and were removed with a hot snare. Biopsies were taken with a cold forceps. The polyp in the sigmoid colon was an inflammatory polyp, consisting of granulation tissue and acute and chronic inflammation. The polyp in the descending colon was a ganglioneuroma with numerous ganglion cells and S100 positive background Schwann cells. We felt this polyp was incidental and likely not contributing to her presenting symptoms. Conclusion: Ganglioneuromas in the GIT are relatively uncommon. They occur in two general settings: sporadically as isolated lesions or syndromically as multiple lesions. They can occur anywhere along the GIT from the stomach to the anus as well as the pancreas and gallbladder. The majority are found in the descending colon. They are endoscopically indistinguishable from hyperplastic or adenomatous polyps. Histologically, there is dilation

and distortion of overlying colonic crypts with lamina propria expansion by admixed ganglion and Schwann cells. Schwann cells can be highlighted by *S100* immunohistochemical staining. *S100* is a family of calcium binding proteins; antibodies to *S100* will identify Schwann cells as well as other cells including melanocytes and Langerhans cells. There are three subtypes of GIT ganglioneuromas. Solitary polypoid ganglioneuromas are the most common type; other subtypes include ganglioneuromatous polyposis and diffuse ganglioneuromatosis. Ganglioneuromatosis is associated with NF-1, Cowden disease and MEN2B. Patients with solitary GIT ganglioneuromas are often asymptomatic, but depending on the size and location, they can present with abdominal pain, constipation, ileus, bleeding and obstruction. Due to the relative rarity of this entity, there are no current data or recommendations on follow up and screening for specific syndromes. Our patient will be screened for other GIT ganglioneuromas with a capsule study and magnetic resonance enterography and undergo a medical genetic evaluation.

**25 ESOPHAGEAL ADENOCARCINOMA IN A MALE TEENAGER.** D. Rivera-Nieves, K. Bittar, Y. Smadi, Pediatric Gastroenterology, UF/Arnold Palmer Hospital, Orlando, Florida, UNITED STATES. Levy, Pediatric Hematology/Oncology, Arnold Palmer Hospital/Orlando Health, Orlando, Florida, UNITED STATES.

Case: A previously healthy 15-year-old male presented with a 4 week history of fatigue, pallor, dysphagia, and weight loss. His hemoglobin was found to be 4.4 mg/dL. Further evaluation revealed iron deficiency anemia with a very low ferritin level, positive Hem occult, and normal CMP. Abdominal ultrasound revealed hepatosplenomegaly with possible lymph node enlargement in the retroperitoneal space. An abdominal CT scan showed a large portal vein thrombosis extending into the portal splenic confluence, gastric varices, and splenomegaly. An upper endoscopy to assess for esophageal varices revealed a friable 10 cm mass from the distal esophagus to the gastric cardia, causing near complete obstruction. An endoscopic ultrasound classified it as a stage 4 adenocarcinoma. Biopsy results confirmed the diagnosis and HER-2 FISH analysis was positive. A whole body PET scan demonstrated uptake at the distal esophagus and multiple areas of liver metastasis, and what was originally thought to be a portal vein thrombosis was found to be tumor mass. The patient underwent a jejunal tube and port placement, and was started on an individualized FOLFOX and trastuzumab therapy regimen with enteral feedings. Discussion: While esophageal tumors are the sixth leading cause of death in the world, with highest prevalence between 35 to 64 years of age, they are extremely rare in children. However, malignant esophageal neoplasms should be considered when there is a presenting history of dysphagia and other symptoms such as weight loss and anemia. In the literature, a total of approximately twenty combined cases of esophageal squamous cell carcinoma (SCC) and adenocarcinoma (AC) in patients < 21 years of age have been reported. A mean age of 16 years and male predominance are features of AC. It has been noted that 84% of AC tumors originate in the distal esophagus and gastroesophageal junction. Metastasis has been reported in 68% of patients with AC at presentation, contrasting with 30% in those with SCC. Barrett's esophagus has been reported in one third of the cases. Other pre-existing conditions such as hiatal hernia, repaired esophageal atresia, history of foreign body ingestion, smoking, spinal palsy, and obesity are present in two thirds of the cases. Interestingly, our patient did not have pre-existing conditions or Barrett's esophagus, but did have a family history of cancer, which has been strongly correlated with esophageal cancer in a study of subjects < 30 years of age. The median survival time for pediatric AC, regardless of treatment modality, is reported to be 9 months. Conclusion: Esophageal cancer is rare in children but should be considered anytime there is a short history of dysphagia in association with other constitutional signs and symptoms.

**26 A RARE CAUSE OF RECURRENT LOWER GASTROINTESTINAL BLEEDING IN AN ADOLESCENT BOY: FOCAL NODULAR LYMPHOID HYPERPLASIA.** G.H. Ustundag, Y.D. Soysal, Pediatric Gastroenterology, Hepatology and Nutrition, Bulent Ecevit University, Zonguldak, TURKEY. E. Piskin, Pediatric Intensive Care Unit, Bulent Ecevit University, Zonguldak, TURKEY. Z. Ornek, Pediatrics, Bulent Ecevit University, Zonguldak, TURKEY.

Introduction: Nodular lymphoid hyperplasia (NLH) of the gastrointestinal (GI) tract is characterized by the presence of multiple small nodules, between 2 and 10 mm in diameter. It may occur in all age groups, but primarily in children under ten years of age. It has been divided to diffuse and focal forms, mainly involving the terminal ileum, the rectum and the colon. Although it can be detected in healthy children, several clinical conditions such as refractory constipation, viral infections, food allergy, giardiasis and immunodeficient states has been linked to NLH. It is also considered a risk factor for intestinal lymphoma. Patients are usually asymptomatic; however, severe cases with intestinal obstruction and bleeding have been reported. Case Presentation: A 14-years-old male

presented with recurrent episodes of massive lower GI bleeding for the past six months. This last episode started with watery diarrhea evolving to melena in several hours. On physical examination, he looked pale and had signs of orthostatic hypotension. The bowel sounds were hyperactive, and the abdomen was nontender without organomegaly. Laboratory tests revealed normochromic normocytic anemia (Hgb: 9,3 mg/dl Hct: % 28,6 MCV: 90,3 fL) consistent with acute blood loss. White blood cell and platelet counts, coagulation tests, erythrocyte sedimentation rate, biochemical and immunological workup were all within normal ranges. Food allergy panel was negative. Stool samples contained fresh blood, and microscopically no leucocytes or parasites were detected. Stool antigen tests for Rotavirus and Adenovirus were negative, and stool cultures were sterile. Technetium-99m-pertechnetate scintigraphy for Meckel's diverticulum was negative. Esophagogastroduodenoscopy was normal. Ileocolonoscopy demonstrated numerous small polyps only in the terminal ileum. The surface of the terminal ileum was covered with brownish blood but no active bleeding or oozing site was located. The histopathological diagnoses of the tissue samples obtained from terminal ileum were chronic nonspecific inflammation and edema with focal active inflammation zones, and benign hyperplastic lymphoid follicles were detected. No endoscopic intervention was done, as the bleeding resolved spontaneously. As far as we learned from previous reports in the literature, these patients may benefit from 5-amino-salicylic acid treatment. The patient was discharged with oral iron supplementation and 30 mg/kg/day of mesalazine treatment. During the six months of follow up he remains to be asymptomatic. Conclusion: Pediatric NLH generally has a benign course with spontaneous regression; however, attention should be paid to patients with severe or recurrent episodes of GI bleeding. After excluding other frequent causes of GI bleeding, and a thorough laboratory work up for an underlying infectious, immunological or allergic disease must be concluded. The effect of mesalazine treatment in such patients remains to be proved.

**29 3 YEAR OLD MALE WITH RECURRENT EROSIIVE ESOPHAGITIS, SEVERE ANEMIA, MELENA AND HYPOALBUMENIA IMPROVED AFTER NISSEN FUNDOPLICATION.** A. Kahlon, I. Novak, P. Costa, M. Carobene, J. Thompson, A. Loizides, Pediatric Gastroenterology and nutrition, Children's hospital at Montefiore , BRONX, New York, UNITED STATES. Background: Gastroesophageal reflux disease (GERD) is a common referral to pediatric gastroenterologists. Symptoms in preschool children range from recurrent vomiting, regurgitation or substernal pain . The majority of cases respond to acid blockade. Some cases may require surgical intervention with a fundoplication. We present a case of a patient with profoundly severe anemia, melena and hypoalbuminemia attributed to GERD induced erosive esophagitis (EE). Case discussion: A 3 year old male with history of intermittent vomiting, regurgitation and GERD presented to the ED with pallor, melena, periorbital and pedal edema. On physical exam he was pale, malnourished, with a weight of 10.3 kg (<1 percentile) and positive stool occult blood. His initial hemoglobin (Hgb) was 3g/dL, albumin 2.3, total protein 4.3, platelets 717, iron 7 with normal inflammatory markers. Stool calprotectin was mildly elevated. Initial upper and lower gastrointestinal endoscopy showed severe erosive esophagitis with normal stomach, duodenum, colon and terminal ileum. Pathology displayed ulcerated epithelium with inflammatory exudates, rare eosinophils and no granulomas. CMV, HSV and fungal stains were negative. Due to the severe anemia, melena, chronic occult blood loss with no etiology and hypoalbuminemia he had an extensive work up including MRI with angiography, Meckel's scan, tagged RBC scan and capsule endoscopy. Workup for immunologic abnormalities and other causes of anemia were negative. Stool malabsorption studies were unremarkable. He was started on Elecare for suspected dietary protein enteropathy. TPN was added to supplement his nutrition as he could not maintain weight and hydration. This was later discontinued due to central venous catheter infection. A 24 hour pH probe study showed pH <4 more than 68% of time. He was started on high dose IV proton pump inhibitors (PPI), H2 blockers and sucralfate. Repeat endoscopies after acid suppression showed some improvement in his EE. However, he continued to have frequent melenotic episodes resulting in low Hgb and hypoalbuminemia corresponding with abnormal pH probe studies despite adequate acid suppression. He underwent a Nissen fundoplication and gastrostomy placement. His symptoms improved and repeat endoscopy revealed marked improvement with of EE and healed ulcers. His albumin level and Hgb have stabilized and he has not required any further transfusions. Discussion: It is not usual to have EE associated with hypoalbuminemia and severe anemia due to chronic blood loss. With the advent of PPIs most cases respond to medical management. Our patient failed medical management but had a dramatic improvement with resolution of his EE, anemia and hypoalbuminemia after fundoplication. This case illustrates that severe EE caused by GERD can not only mimic severe anemia and melena but also protein losing enteropathy.

**35 COLLAGENOUS GASTRITIS: A CASE REPORT AND REVIEW OF LITERATURE.** C. Cartagena De Jesus, R. Jain, M. Demant, L. Nicholls, H. Hassan, Pediatrics, University of Arizona, Tucson, Arizona, UNITED STATES.

Collagenous gastritis is a rare disease with only 60 cases reported in the English literature since it was first reported in 1989. The disease is characterized by the subepithelial deposition of collagen bands as well as infiltration of inflammatory mononuclear cells in the lamina propria. So far, no effective therapeutic modality has been described. We are reporting a 17-year-old girl with collagenous gastritis who presented to the emergency department with a history of progressive fatigue, exertional dyspnea, epigastric pain and was found to have severe microcytic anemia. An esophagogastroduodenoscopy was performed and showed nodularity in the stomach associated with erythema and erosions most marked in the prepyloric area. Gastric mucosal biopsy showed thickened subepithelial collagen band measuring 50µm with entrapped superficial mucosal capillaries and sloughing of epithelium. A trichrome stain highlighted the collagen band, establishing the diagnosis criteria of collagenous gastritis. The patient was started on high-dose oral proton pump inhibitor twice daily and iron supplementation. Seven weeks later repeated biopsies showed disappearance of collagen deposition in the gastric mucosa. Furthermore, the patient clinically improved with resolution of symptoms and anemia. Conclusion: Collagenous gastritis is a rare disorder with a variable clinical course. To date there is no established therapeutic strategy to treat patients with collagenous gastritis. Though previous cases have shown no response to high dose PPI and iron supplementation, our case report suggests these may serve as supportive measures in the case of remission of the disease.

**36 EOSINOPHILIC ESOPHAGITIS IN A CHILD WITH LYMPHANGIOENDOTHELIOMATOSIS.** C. Wallace, Pediatrics, University of Alabama at Birmingham, Birmingham, Alabama, UNITED STATES.

Abstract: Multifocal lymphangioendotheliomatosis with thrombocytopenia (MLT), also known as cutaneovisceral angiomas with thrombocytopenia, is an extremely rare congenital vascular disorder. Eosinophilic Esophagitis (EoE) is an inflammatory esophageal disease characterized by esophageal dysfunction and esophageal eosinophilia in which other causes have been excluded. Upon review of the literature, no known previous cases are reported in patients diagnosed with both of these diseases. We present a child with MLT diagnosed at birth who developed EoE at age 4 years. Case Report: The case patient is a female who was born at term without prenatal complications, found at birth to have multiple cutaneous hemangiomas. She was transferred from an outside hospital at 4 days of life to our facility due to massive gastrointestinal (GI) bleeding with hematemesis and hematochezia along with precipitous drop in both hematocrit and platelets. TORCH infection and bacterial sepsis workup were negative, and she was eventually diagnosed with MLT by skin biopsy. The neonatal intensive care hospitalization was complicated by extensive GI bleeding requiring intravenous Octreotide and Carafate, along with frequent blood product transfusions. Following initial stabilization, she was discharged home after one month of supportive care. She continued to have frequent follow up in GI clinic for persistent GI bleeding. At 8 months old, the patient underwent sigmoidoscopy that revealed friable mucosa and multiple hemangiomas which bled easily. Esophagogastroduodenoscopy (EGD) also revealed multiple hemangiomas in the stomach with no active bleeding and a normal-appearing esophagus. She continued to have close GI follow up, and, as she aged, she had less frequent GI bleeding. A repeat surveillance EGD at 4 years old showed gross furrowing of the esophagus with biopsies revealing 47/hpf intraepithelial eosinophils in the distal esophagus, 34/hpf in the proximal esophagus. There was no eosinophil clustering or microabscess formation seen. At that time, the patient continued to have occasional bloody stools, but her family denied any problems with feeding, dysphagia, weight loss, or other gastroesophageal reflux (GER)-like symptoms. She was started on Budesonide slurry for EoE treatment. Around 9 months later, the patient developed worsening symptoms of dysphagia and her family admitted intermittent compliance with home Budesonide. A repeat EGD showed elevated number of eosinophils, sclerosis in the lamina propria, eosinophil clustering, and eosinophil microabscesses. Compliance with Budesonide was subsequently reestablished, and the patient has since had follow up EGD with normal biopsies. Discussion: This case describes a patient with two seemingly unrelated disorders affecting the GI tract. From what is currently known about MLT, there appears to be no pathophysiological correlation between the congenital cutaneous and visceral vascular malformations and the eventual diagnosis of EoE. A literature review revealed no known cases of both MLT and EoE in either the pediatric or adult populations, and it is unknown what impact these concurrent diagnoses will have on disease progression as this patient ages.

**38 COUNTRY, CRITTERS, AND CAMO: CHRONIC ABDOMINAL PAIN AS A PRESENTATION OF CHILDREN WITH ALPHA-GAL ALLERGY.** C.A. McKinney, Pediatrics, University of Virginia Children's Hospital, Charlottesville, Virginia, UNITED STATES. C. McGowan, Allergy/Clinical Immunology, University of Virginia, Charlottesville, Virginia, UNITED STATES. H. Barnes, Pediatric Gastroenterology, Hepatology, and Nutrition, University of Virginia Children's Hospital, Charlottesville, Virginia, UNITED STATES.

Background Mammalian Meat Allergy or "Alpha-gal" is a novel IgE mediated antibody response to the carbohydrate moiety galactose- $\alpha$ -1,3-galactose. This relatively rare tick-borne allergic disorder is an increasingly described etiology of delayed urticarial, angioedema, and anaphylactic reactions in both adult and pediatric populations. However, to our knowledge, it has never been described in association with chronic abdominal complaints. Objective We aim to highlight a cohort of 8 pediatric patients at our institution (8 Males, 14.5yrs) who presented between 2012-2017 with chronic abdominal complaints and were found to have positive testing for IgE specific to galactose- $\alpha$ -1,3-galactose (2.75 kU/L; Total IgE 394 IU/mL). These patients presented with predominantly epigastric and periumbilical pain, nausea, and emesis as their symptomatology and all had comprehensive evaluations that included endoscopy and laboratory studies that were otherwise unrevealing. Discussion and Conclusion Alpha-gal may represent a new etiology for unexplained chronic abdominal complaints. We recommend considering alpha-gal in the workup for chronic abdominal symptomatology of unclear etiology, especially in the context of the correct geographic location and relevant history of potential tick bite exposure.

**40 HYPOALBUMINEMIA AND ANEMIA IN A CHILD WITH EROSIIVE ESOPHAGITIS.** D. Rivera-Nieves, K. Bittar, Y. Smadi, Pediatric Gastroenterology, UF/Arnold Palmer Hospital, Orlando, Florida, UNITED STATES. R. Sutphin, Pediatric Hematology/Oncology, Arnold Palmer Hospital/Orlando Health, Orlando, Florida, UNITED STATES. Case: A 7-year-old previously healthy male presented with a 2-week history of periorbital and ankle edema, 4 Lb weight gain and decreased energy. Serum albumin was 2 mg/dL. Total protein was 3.8 g/dL. Urinalysis was normal. Electrolytes and liver enzymes were normal. CBC was remarkable for mild iron deficiency anemia. Upper and lower endoscopies showed severe erosive exudative pan-esophagitis and a medium sized hiatal hernia. Pathology showed active esophagitis, numerous neutrophils, granulation tissue, ulcer bed, and rare eosinophils. Viral stains were negative. Colonoscopy was unremarkable. He was started on esomeprazole and sucralfate. Extensive evaluation failed to reveal a nutritional, pancreatic, intestinal or renal explanation for his hypoalbuminemia (HA). Infectious work up was negative including EBV, CMV, HIV, parvovirus B19 and stool studies. Stool hemocult was positive. Interestingly, stool alpha-1 antitrypsin was normal, however calprotectin was elevated. He had low serum IgG. An MRE showed normal bowel. An abdominal ultrasound showed ascites. A CT of chest and abdomen showed a thymic cyst and borderline superior mediastinal adenopathy, but otherwise was normal. He then developed worsening anemia and HA requiring several blood and albumin transfusions. A bone marrow biopsy was normal. Repeat endoscopy 3 months later showed persistent severe pan-esophagitis with unchanged pathology. A pH impedance study revealed asymptomatic pathologic reflux. Therefore, he underwent a Nissen fundoplication. Subsequently, he had normalization of albumin, resolution of edema, and returned to normal energy. Repeat endoscopy 3 months later showed significant improvement with resolution of erosions and ulcers. Discussion: We present this interesting case of a child with severe reflux erosive esophagitis presenting with hypoalbuminemia and anemia. Gastroesophageal reflux disease (GERD) has been associated with growth impairment, anemia, and respiratory problems in children. Hypoalbuminemia, anemia, and iron deficiency are commonly associated to inflammatory bowel disease, protein-losing enteropathies, eosinophilic gastroenteritis, poor nutrition, and other GI disorders. It may carry severe complications such as esophageal stricture and GI bleeding, however not hypoalbuminemia. To our knowledge, there has been only one prior case report of hypoalbuminemia in a child with severe erosive esophagitis, in the setting of cystic fibrosis, which responded to PPI and Nissen fundoplication. In our patient, recognition and treatment of severe erosive esophagitis resulted in resolution of hypoalbuminemia and anemia. Conclusion: Severe erosive esophagitis should be considered in the differential diagnosis of children presenting with hypoalbuminemia and anemia, even in the absence of GI symptoms.

**48 ATYPICAL PRESENTATION OF PYLORIC STENOSIS.** A. Chan, E. Prince, Pediatric Gastroenterology, University of Rochester Medical Center, North Chili, New York, UNITED STATES.

Hypertrophic pyloric stenosis is a disorder of unclear etiology characterized by gastric outlet obstructive symptoms due to hypertrophy of the pylorus, typically seen in young infants around 3-6 weeks of age and more commonly in

males and premature infants. Typical symptoms include nonbilious and progressively forceful vomiting described as projectile. Patients were classically described as emaciated or dehydrated with poor weight gain, with hypochloremic, hypokalemic metabolic alkalosis. Ultrasound is used to assess pyloric muscle thickness, pyloric muscle length, and pyloric diameter. Pyloric stenosis is rare after 12 weeks of age. Heritability estimate is about 87% based on a familial aggregation study in Denmark, with monozygotic twins having 200-fold increase in risk. We describe a 7 month old triplet male (Baby B) born prematurely at 29 weeks with prenatal complications of gestational diabetes and hypothyroidism, both treated adequately. The patient presented with persistent nonbilious emesis and was diagnosed with reflux and possible cow milk protein intolerance. The patient was breastfed, and maternal elimination of soy and dairy did not improve the emesis. There was no significant improvement with metoclopramide or a proton pump inhibitor. Despite the emesis, the patient continued to gain weight well with an average of 24 grams/day over three months. Due to persistent vomiting, an upper GI series was done showing a narrow distal antrum and pyloric channel with shouldering of the duodenal bulb. Ultrasound confirmed the pyloric region to be persistently elongated to at least 2.4cm in length, thickened anterior wall at least 3.7mm throughout the exam. At the time of diagnosis, the patient had normal electrolytes. The patient underwent a successful pyloromyotomy. The patient's siblings (Baby A, Baby C) had no emesis to suggest pyloric stenosis. This case highlights the need to consider hypertrophic pyloric stenosis in the differential of persistent reflux and emesis despite a lack of classical symptoms and rarity of the diagnosis in patients of this age group.

**51** *INTESTINAL DUPLICATION CYST MASQUERADING AS COLITIS IN 4 MONTH OLD.* A.M. Murphy, Pediatrics, Inova Children's Hospital, Falls Church, Virginia, UNITED STATES. Honigbaum, N. Sikka, S. Hourigan, Pediatric Gastroenterology, Pediatric Specialists of Virginia, Fairfax, Virginia, UNITED STATES. Honigbaum, N. Sikka, S. Hourigan, Pediatric Gastroenterology, Inova Children's Hospital, Falls Church, Virginia, UNITED STATES. Jerath, Pediatric Radiology, Inova Children's Hospital, Falls Church, Virginia, UNITED STATES.

Case: A 4 month old full term female presented to the emergency department after three large bloody, painless stools. She had no abdominal pain, fussiness, or recent illness. Hemoglobin was 6.8 g/dL. Gastric lavage was clear fluid. Abdominal ultrasound was normal without evidence of intussusception. A Tc-99m pertechnetate scintigraphy scan for Meckel's diverticulum showed immediate visualization of multiple bowel loops with subsequent excretion and movement, suggestive of colitis; due to this, the presence of a Meckel's diverticulum could not be evaluated. (Figure 1). Infectious stool studies and blood inflammatory markers were normal. Colonoscopy was performed to visualize colitis, however was grossly normal with normal biopsies. Repeat Meckel's scan was still inconclusive and suggestive of colitis. Gastrointestinal bleeding stopped and the patient was discharged for further outpatient work up. The patient returned to the hospital eight days later with new hematochezia. Repeat ultrasound demonstrated a fluid-filled structure with surrounding bowel wall measuring up to 6.4 cm which reflected an enteric duplication cyst. The patient was taken to the OR where 36 cm of a small bowel duplication cyst was removed in addition to 6 cm of ileum where a common wall between cyst and ileum was non-reparable, and the bowel was anastomosed. Discussion: Enteric duplication cysts (EDC) are rare congenital anomalies and as such as often misdiagnosed upon initial presentation. There have been reports in which EDCs mimic other entities – teratoma, ovarian cyst, volvulus and Meckel's diverticulum. They can occur anywhere along the enteral tract, but are divided into foregut, small bowel and large bowel duplication cysts and can be cystic or tubular. The most common EDCs are found in the jejunum followed by the ileum. Typically, EDCs are diagnosed via ultrasound however this was missed in our patient on the first ultrasound possibly due to the cyst being collapsed. About 10% of EDCs contains ectopic gastric mucosa; hence in our case this was detected in the Tc-99m pertechnetate scintigraphy performed and mimicked colitis. This case highlights intestinal duplication cysts can sometimes be detected on Tc-99m pertechnetate scintigraphy and there should be a high index of suspicion when unusual findings are seen.

**52** *CASE SERIES: SUCCESSFUL TREATMENT OF GASTROPARESIS WITH GASTRIC ELECTRICAL STIMULATOR IN CYSTIC FIBROSIS PATIENTS.* R. Garcia-Naveiro, Gastroenterology, Akron Children's Hospital, Akron, Ohio, UNITED STATES. Ponsky, Pediatric Surgery, Akron Children's Hospital, Akron, Ohio, UNITED STATES. A. Mathews, B. Arnold, Medical Education, Akron Children's Hospital, Akron, Ohio, UNITED STATES.

Introduction: Cystic fibrosis (CF) has been shown to be associated with gastrointestinal dysmotility such as reflux, meconium ileus, distal intestinal obstruction syndrome, chronic constipation and gastroparesis (GP) with and without diabetes. Studies have shown increased rates of gastroparesis in these patients, although the mechanism is not well understood. This delay in emptying time can worsen their underlying chronic malnutrition due to

reduced oral caloric intake and interference with oral medication delivery. The digestion of fat can be impacted specifically, as absorption can be affected by how rapidly fat enters the duodenum, which can then correlate with how these patients respond to pancreatic enzyme replacement therapy. A systematic review (Corral, et al) showed an increased rate of gastroparesis in CF patients over the general population, with a frequency of 38%, which seems to increase with age. Gastroparesis, regardless of concurrent illnesses, remains a difficult disorder to treat in not only CF patients, but the general population as well. Traditional treatment with metoclopramide and erythromycin has numerous limitations such as age restrictions and side effects, with most studies showing only about a 50% response rate. Gastric electrical stimulators (GES) have begun to emerge as a possible treatment option in those with gastroparesis refractory to medications and traditional therapies. In our series, we found that CF patients respond particularly well to GES and show that it may be an alternative option to treat their GP.

**Case Description:** In our case series, we describe two CF patients with drastic improvements in symptoms associated with gastroparesis after placement of GES. The first patient is a 13 year-old male with CF, GERD, celiac disease, gastroparesis, status post gastro-jejunal tube with Nissen fundoplication who had 2 years of daily, worsening nausea, bloating, burping, emesis progressing to hematemesis, abdominal pain, decreased oral intake and toleration of tube feeds, as well as a weight loss of about 10 pounds. Following placement, he was able to tolerate oral and tube feeds, had an increase in appetite, without emesis and with significant improvement in nausea, with a weight gain of 5 pounds over 6 months. The second patient is a 21 year-old female with CF, gastroparesis secondary to CF-related diabetes, history of recurrent distal intestinal obstruction syndrome, osteopenia and CF related cirrhosis. Her gastric emptying study was severely abnormal for both solids and liquids. She presented with very similar symptoms as our previous case, although even more severe. She also had an improvement in her nausea, emesis, abdominal pain, and tolerating feeds with resulting weight gain of about 3 pounds in one month. In both cases, a temporary GES was first placed either nasogastrically or inserted into prior gastric tube stoma. They were seen in follow up with improvement of their presenting symptoms, at which point they underwent placement of permanent GES. With significant improvement of their gastric emptying times on Upper GI, patients had improvement of their symptoms and better quality of life.

**Discussion:** Gastroparesis in CF patients is not caused directly by loss of CFTR. Studies have hypothesized that GP is caused by different neurologic reflexes and neurohormonal pathways in CF patients. These reflexes include prolonged ileal inhibition, colonic stasis and abnormal circulating gastrointestinal hormone response to oral stimulus all causing delayed emptying. Gastroparesis can be predicted by chronic constipation, malnutrition and a lower BMI, in patients with and without CF. Specific associations of GP with CF include chronic disease which typically leads to chronic inflammation, as well as chronic use of anticholinergics and opiates which slow emptying time. Gastroparesis remains a difficult condition to treat, despite the innovation of GES, with response rates varying from 50-75%. However, in both CF patients described above, the GES immediately and drastically improved symptoms caused by their delayed gastric emptying, with weight gain. Just as the mechanisms underlying the cause of GP in CF patients are both multifactorial and complex, the explanation for this suspected greater response to GES is unclear but encouraging for the future of treatment in this patient population.

**57 STERCORAL COLITIS - A RARE AND LIFE-THREATENING PRESENTATION OF STOOL-WITHHOLDING BEHAVIOR AND CHRONIC CONSTIPATION.** B.D. Constant, Pediatrics, Children's National Medical Center, Washington, District of Columbia, UNITED STATES. Otero, Radiology, Children's National Medical Center, Washington, District of Columbia, UNITED STATES. Barber, Hospitalist Medicine, Children's National Medical Center, Washington, District of Columbia, UNITED STATES.

Stercoral colitis (SC) is colonic distention and ischemic pressure necrosis, with potential for colonic perforation, caused by fecal impaction. An 11-year-old girl presented with severe chronic constipation due to stool-withholding behavior and was diagnosed with SC. To our knowledge, this is the fifth reported case of stercoral colitis in the pediatric population, and the first due to stool-withholding behavior. She presented with acute on chronic abdominal pain, anorexia, compensated hypovolemic shock, and new onset black stools. She was afebrile and her exam was notable for severe lower abdominal tenderness with guarding. Her labs indicated leukocytosis, elevated inflammatory markers, and prerenal azotemia. Computed tomography of the abdomen demonstrated stercoral colitis of the rectosigmoid colon. She was appropriately resuscitated; however, due to persistent pain, the degree of impaction, and radiographic colitis, she later required manual fecal disimpaction under general anesthesia, which successfully relieved her distal impaction. We were unable to safely relieve her remaining stool burden, demonstrated on abdominal x-ray, by enema and colonic washes due to the

risk of perforation. Because of this, she required sigmoidectomy and diverting colostomy to allow for bowel decompression and healing. Psychology was involved due to a large anxiety component to her withholding. No organic etiology of her constipation was identified and she recovered quickly after surgery. The vast majority of patients diagnosed with SC are elderly, with chronic constipation and comorbidities including neurological impairment, physical inactivity, and opiate use. Fewer than 150 total cases have been reported as of 2015. However, this life-threatening condition with significant potential for morbidity and mortality (32-60%) is likely underestimated and commonly mis-diagnosed, especially when perforation is absent, making it an important, albeit rare, diagnosis in the differential of abdominal pain and chronic constipation.

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**58 SEVERE COLONIC FECAL IMPACTION, STERCORAL COLITIS AND LIVER DISPLACEMENT IN AN ADOLESCENT MALE WITH CHRONIC CONSTIPATION.** B. Sparks, B. Boyle, D. Yacob, Pediatric Gastroenterology, Hepatology and Nutrition, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES. Boyle, D. Yacob, The Ohio State University, Columbus, Ohio, UNITED STATES.

Background: Constipation is a common problem in children. It is mostly functional in nature with less than 5% of cases having an organic etiology. Fecal impaction is common in children who suffer from severe chronic constipation and may be associated with fecal soiling, abdominal distension and abdominal discomfort. Life-threatening complications such as abdominal compartment syndrome and bowel perforation are occasionally encountered. Case history: A sixteen-year-old male presented to the emergency department with complaints of fatigue, abdominal pain, and five loose stools per day with fecal soiling. He has a history of chronic constipation followed by pediatric gastroenterology, but he was lost to follow up and off medications for six years. The details of his stooling history were uncertain to the family. Abdominal plain film revealed an eleven centimeter fecal impaction and marked gaseous sigmoid distension. On admission patient developed acute onset hypotension with positional changes. He developed a fever to 38.3°C and his labs were significant for hyponatremia and lactic acidosis. A blood culture was drawn and he was started on piperacillin/tazobactam and vancomycin. He received four liters of normal saline. Clinical changes required urgent evaluation by the ICU and surgical teams. After improved hemodynamics, patient taken for emergent abdominal CT scan demonstrating fecal impaction within his recto sigmoid colon and adjacent colonic wall thickening concerning for ischemic versus inflammatory changes, but no bowel perforation. Severity of sigmoid distension displaced right hepatic lobe medially. Following serial enemas, patient required manual disimpaction on hospital day two. Post disimpaction KUB demonstrated improved distal stool burden and sigmoid distension. Given improvement patient safely completed bowel clean-out with PEG 3350 via nasogastric tube over three days. Patient discharged home on regimen of 10 mg oral bisacodyl daily and 17 g PEG 3350 twice daily. Outpatient colonic motility evaluation demonstrated severe distal colonic dysmotility and an anorectal manometry with an intact RAIR. Conclusions and inferences: This case demonstrates the risk of serious complications of fecal impaction including perforation, compartment syndrome, and ischemic injury in an otherwise healthy adolescent. Stercoral colitis and perforation are more commonly seen in the elderly population.

1 Our patient's hemodynamic instability may have been due to interruption of venous return secondary to compression by the distended recto sigmoid colon, cytokine release from colonic inflammation, or colonic volvulus.

2,3 While fecal impaction is common and often treated as an outpatient, in severe cases a CT scan is crucial in identifying stercoral colitis and potentially life-threatening sequelae.

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**59** *INTRACTABLE VOMITING AS A LATE COMPLICATION OF A VENTRICULOCHOLECYSTIC SHUNT.* R. Arbizu, N. Kasi, C. Jump, Pediatric Gastroenterology, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. Eklund, Pediatric Radiology, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. Eskandari, Pediatric Neurosurgery, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. D.W. Lapp, Pediatrics, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. J. Chandler, Pediatric Surgery, Greenville Health System Children's Hospital, Greenville, South Carolina, UNITED STATES. E.C. Troup, Pediatric Neurosurgery, Greenville Health System Children's Hospital, Greenville, South Carolina, UNITED STATES.

Management of hydrocephalus is often achieved by shunting excess CSF from ventricles to the abdominal cavity via a ventriculoperitoneal (VP) shunt. In cases where a VP shunt is contraindicated or fails, a ventriculocholecystic (VC) shunt is an alternative method. These shunts are thought to adequately control intracranial pressure, have a low risk for infection, and allow electrolyte reabsorption from the small intestine. The most common indications for placement of a VC shunt include ascites, shunt infection, intra-abdominal pathology causing non-absorption of CSF, and abdominal pseudocysts. Here we report on a case of a 9-year old boy with history of prematurity and congenital hydrocephalus that was managed by a VC shunt placed at 3 years of age after recurrent abdominal pseudocyst formation of his VP shunt. His hydrocephalus was well managed. He presented to gastroenterology with a history of intermittent emesis varying between 2-3 times per month to 2-3 times per day over the previous 2 years. Vomiting was associated with significant abdominal distention, epigastric discomfort, and at times headache. The episodes were not related to meals. In addition, he had weight loss falling from the 35<sup>th</sup> to the 10<sup>th</sup> percentile for BMI. Repeat head MRI imaging failed to show signs of elevated intracranial pressure. Workup revealed a normal upper endoscopy, normal gastric emptying, an abdominal ultrasound with shunt catheter tip within the gallbladder lumen with sludge and a dilated common bile duct measuring 8 mm. He subsequently underwent MRCP which confirmed above findings but also showed a tapering of the extrahepatic biliary tree without obstruction. He failed to respond to proton pump inhibitors, cyproheptadine for cyclic vomiting syndrome, ursodiol for biliary sludge. He ultimately underwent cholecystectomy and placement of ventriculo-pleural shunt. During cholecystectomy, his gallbladder was noted to be distended and atonic, as well as socked in with adhesions, consistent with the diagnosis of biliary dyskinesia. There were extensive adhesions throughout the abdominal cavity precluding conversion to a simple ventriculo-peritoneal shunt. On follow up, the patient's abdominal distention and vomiting episodes are significantly decreased to once per month. Known complications of VC shunts include cholelithiasis, retrograde and descending shunt infections. This case highlights a previously undescribed late complication of a VC shunting.

**61** *TRANSIENT JEJUNO-JEJUNAL INTUSSUSCEPTION IN AN ADOLESCENT PATIENT.* C.S. Vijay, C. Chen, Pediatrics, West Virginia University, Morgantown, West Virginia, UNITED STATES.

A 16-year-old male with a history of Type I diabetes, gastroesophageal reflux disease, and appendectomy was transferred to our facility with a one day history of crampy abdominal pain, diarrhea, and non-bloody vomiting. The patient reported one episode of emesis a week prior to presentation, but otherwise had been asymptomatic until the previous day. He denied any fever, bloody stools, or any other symptoms. At the outside facility, computed tomography (CT) scan was performed and showed jejuno-jejunal intussusception in the left lower quadrant. On admission to our facility, he was alert and appeared to be in no acute distress. His vitals were within normal limits. He had diffuse abdominal tenderness that was worst in the left upper quadrant. His abdomen was soft, and he had audible bowel sounds in all quadrants and no palpable masses. Complete blood count, electrolytes, amylase, lipase, and lactic acid were all within normal limits. An abdominal ultrasound was performed which showed persistent intussusception in the left upper quadrant. Pediatric surgery was then consulted for further management. He subsequently underwent a diagnostic laparoscopy, which showed hyperemic bowel with mild fluid-filled distension of the small bowel and mesenteric adenopathy. There were no signs of intussusception. No adhesions or hernias were found, and his stomach, liver, gallbladder, and spleen all appeared normal. He did well after his surgery, and his abdominal pain, vomiting, and diarrhea all improved during his hospital stay. He had no recurrence of these symptoms following hospital discharge. Discussion Transient jejunal intussusception is a very rare condition, and its pathophysiology is not well understood. Intussusception is generally a disease of infancy, and its presentation in patients of this age is a rare occurrence. While most cases of intussusception in young children are idiopathic, cases in older children and adults usually have an identifiable cause, such as polyps, malignancy, and adhesions. The more classic symptoms found in infants are less consistent among adolescents.

Transient intussusception in this age group may present with an array of non-specific symptoms including episodic abdominal pain, nausea, and vomiting. However, they may be completely asymptomatic and may only be discovered on imaging performed for other reasons. Although our patient did have a history of appendectomy, he did not have any adhesions on laparoscopy and he did not have any other identifiable cause of his transient intussusception. This case also represents a diagnostic challenge, and intussusception should be kept in the differential diagnosis even in older patients.

**62 COEXISTING OF TWO RARE PATHOLOGIES: STOMACH PERFORATION AND ACCESSORY SPLEEN TORSION.** Ç. el, Pediatrics, Mustafa Kemal University, Hatay, TURKEY. M.E. Çelikkaya, A. ATICI, B. Akçora, pediatric surgery, mustafa kemal university, HATAY, TURKEY.

**Introduction:** Gastric perforation secondary to acute gastric dilatation is a rare condition in the pediatric age group. Over-feeding, peptic ulceration, respiratory resuscitation using ambu bag and intestinal volvulus has been blamed for its etiology. Early recognition of the perforation, intestinal decompression with using the nasogastric tube and early surgical intervention may be lifesaving. Only few cases of the torsion of the accessory spleen have been reported in the literature. In this case we report both the gastric perforation and the accessory splenic torsion in the same patient. **Case:** 8 years old girl patient has been admitted to our emergency department complaining of acute abdominal pain and vomiting of 24 hour duration. She has a history of growth retardation and chronic loss of appetite. She suddenly ate too much food in the day prior to her illness. Physical examination revealed findings of disseminated peritonitis and the erect abdominal x ray showed air under diaphragm. Immediate surgical intervention revealed gastric perforation of the posterior wall and necrotic tissue which was debrided and the perforation was primarily sutured. Exploration revealed accessory spleen with torsion for which splenectomy were done. No postoperative complications were recorded and the patient discharged in a stable condition. **Conclusion:** Spontaneous gastric perforation secondary to acute gastric dilatation is rare and the early diagnosis will be lifesaving for the patient. The torsion of the accessory spleen is also very rare and its association with the gastric perforation should be searched for.

**64 A NOVEL TECHNIQUE FOR TEMPORARY GASTRIC ELECTRICAL STIMULATOR LEAD PLACEMENT IN CHILDREN.** D. Jacob, P. Lu, C. Di Lorenzo, Pediatric GI, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES.

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**Background:** Gastric electrical stimulation (GES) is used to treat children and adults with chronic nausea and vomiting refractory to conventional treatment and involves electrical stimulation of the stomach via leads connected to a stimulator. In children, GES often involves an initial trial of temporary GES delivered by leads attached endoscopically for several days to the gastric mucosa while the stimulator remains external to the patient. Clinical response during the temporary GES trial determines whether patients proceed to permanent implantation of the stimulator. Secure attachment of the temporary GES leads is therefore critical. Current practice involves placement of endoclips over the lead and underlying gastric mucosa, but dislodgement of the lead has been a common problem during the trial period. We describe a novel technique for secure endoscopic attachment of temporary GES leads. **Methods:** The Crossover Double Loop (CDiLo) technique is shown in Figure 1. **Case Presentation:** Our case is that of a 13 year old male with chronic epigastric pain and intractable nausea and vomiting that developed after a viral illness. Prior evaluation was remarkable only for a mild delay in gastric emptying. His symptoms were refractory to conventional treatment and he was dependent on transpyloric feeding at the time of our evaluation. The CDiLo technique was used to initiate a temporary GES trial. The lead was inserted intranasally and after clipping to the gastric mucosa was connected to an external stimulator. Over the following week, the patient experienced significant symptomatic improvement and was able to increase his oral intake and decrease the tube feedings. Stimulator impedance was in the desired range at 1 week, suggesting that the lead remained secure. The patient underwent laparoscopic implantation of GES leads and stimulator after a successful 2-week temporary GES trial. Endoscopic inspection of the temporary lead at the time of surgery showed the lead had remained securely attached to the gastric mucosa. The temporary lead was removed by using a snare to enclose the endoclips and applying gentle traction. **Conclusion:** The CDiLo technique is a novel method for secure temporary GES lead placement. Secure lead attachment is critical for an adequate temporary GES trial, which may allow for improved patient selection and treatment outcomes.

**72 JUVENILE SYSTEMIC SCLEROSIS: GASTROINTESTINAL SYMPTOM EVALUATION AND MANAGEMENT OF A RARE PEDIATRIC DISEASE.** N. Kasi, C. Jump, R. Arbizu, Pediatric Gastroenterology, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. X.T. Zhang, E. Antonio, Pediatrics, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES.

A 13-year-old African American female patient presented with a 3-year history of dysphagia, odynophagia, worsening gastroesophageal reflux disease (GERD) symptoms despite acid suppression therapy, postprandial left upper quadrant abdominal pain and bloating. Review of systems revealed fatigue, weight loss, arthralgia, skin induration and paresthesias with discoloration of her fingers upon cold exposure. Physical exam showed diffuse, symmetric skin thickening of perioral region and dorsal aspect of hands. Esophagram revealed absent primary peristalsis. Endoscopy demonstrated diffuse esophageal erythema and white plaques. Esophageal tissue culture grew *Candida glabrata*. Gastric emptying study was abnormal (33% meal retention at 4 hours). Esophageal manometry showed a normal swallow induced upper and lower esophageal sphincter resting pressure and relaxation and absent esophageal body peristalsis consistent with a major disorder of peristalsis (Fig. 1). Antinuclear antibody titer was elevated (ANA; >1:2560). Anti-Scl-70, anti-centromere, anti-RNA polymerase III and lupus-specific antibodies were negative. Patient was diagnosed with Juvenile Systemic Sclerosis (JSSc) based on her clinical symptoms, visceral involvement, and supported by a positive ANA titer. Disease modifying therapy with prednisone and methotrexate was initiated. Organ targeted therapy with erythromycin was added for gastroparesis, fluconazole for *Candida* esophagitis, and acid suppression was optimized for GERD. Patient required a gastrostomy tube placement for supplemental nutrition and medication administration. This case illustrates the evaluation and treatment of a patient with JSSc, an uncommon, chronic, multisystem connective tissue disorder that carries the most significant morbidity of the sclerotic disorders. The estimated incidence of JSSc is 0.27 cases per million per year and occurs 3 times more often in girls. The cause of JSSc is unknown and the pathogenesis is complex encompassing an abnormal immune activation leading to vascular damage and excessive synthesis of extracellular matrix with increased deposition of normal collagen in skin and internal organs, including the gastrointestinal (GI) tract. Raynaud's phenomenon and skin induration are the most common presenting symptoms and GI involvement is reported in nearly 90% of patients. The majority have esophageal dysfunction presenting as dysphagia and complications secondary to dysmotility. The stomach, small bowel, colon and anorectum can be affected and can present as gastroparesis, malabsorption secondary to bacterial overgrowth, pseudo-obstruction, malnutrition, constipation and incontinence. As outlined in our case, radiologic, endoscopic and manometry studies are warranted to guide GI organ targeted therapy including the use of acid suppression for GERD control, prokinetics to enhance motility, diet modification and supplementation to optimize nutrition, and stool softeners and/or laxatives for constipation. Since treatment is mainly symptomatic and focused on organ involvement, a clear understanding of medication indication, mechanism of action and side effects is required. Our patient continues to have close follow up with Gastroenterology, Rheumatology and Nutrition and has demonstrated overall clinical improvement.

**73 GOOD OUTCOME OF INTESTINAL VOLVULUS WITHOUT MALROTATION IN A NEWBORN; CASE SERIES.** W. Hasebe, Pediatrics, Komaki City Hospital, Komaki, Aichi, JAPAN. W. Hasebe, Y. Kato, H. Asada, T. Fukasawa, T. Hattori, T. Kubota, Pediatrics, Anjo Kosei Hospital, Anjo, Aichi, JAPAN. S. Makita, Pediatric surgery, Anjo Kosei Hospital, Anjo, Aichi, JAPAN.

Introduction Midgut volvulus is a condition in which the small bowel or proximal colon twists around the superior mesenteric artery. Intestinal volvulus in the neonatal period is one of the most urgent diseases. A typical example involves abnormal bowel rotation, but volvulus without malrotation is relatively rare. Extensive intestinal ischemia from volvulus can result in short bowel syndrome. If the volvulus persists for a long time, it results in infarction of extensive intestine with high morbidity and mortality. Cases We report three cases of intestinal volvulus without malrotation in a newborn that occurred within 6 days of the early neonatal period. Of the 3 patients, 2 were male and one was female. Their gestational age was 34, 35 and 40, respectively. Their birth weight was 2405g, 2430g and 2750g, respectively. The onset was 10 hour after birth, day 2 of life and day 6 of life, respectively. Emergency operation was performed at day 1 of life, day 2 of life and day 6 of life. All the infants had an extended intestinal tract on imaging examination, bilious vomiting, and ascites. Results of ultrasonography and contrast studies were negative for intestinal malrotation abnormalities. However, judging from the ascites and ileus findings, we made a diagnosis of acute abdomen and performed emergency open surgery. In all three cases, we observed ascites and twisting of the intestine with shortening of the mesentery, and the ligament of Treitz was in its normal position.

This, we made a diagnosis of intestinal volvulus without malrotation. Only one of the infants underwent intestinal resection of the stenosis. The two other infants only underwent torsion release without resection of the intestinal tract and had an uneventful postoperative course. Fortunately, all three neonates had good outcome without parenteral nutrition after operation. Discussion In the present cases, although no definite diagnosis could be made in all three patients preoperatively, their lives were saved by performing operation at an early stage. When the pediatrician observes bilious vomiting as the initial response of a newborn, even without a bowel rotation abnormality, the possibility of volvulus without malrotation should be considered sufficiently and a surgeon should be consulted immediately because preoperative diagnosis and deciding whether to perform surgery is difficult. During this time, the presence of ascites and changes in the ileus symptoms are important findings.

**75 DIAGNOSING INFLAMMATORY BOWEL DISEASE IN A SEVERELY ANEMIC JEHOVAH'S WITNESS: ETHICAL AND MEDICAL CONSIDERATIONS.** A. Kahlon, E. Prendaj, M. Zeien-Tarantelli, P. Costa, I. Novak, G. Tomer, A. Loizides, Pediatric Gastroenterology and nutrition, Children's hospital at Montefiore , BRONX, New York, UNITED STATES. Fleischman, Pediatrics , Children's hospital at Montefiore , BRONX, New York, UNITED STATES.

Severe acute anemia is typically managed by blood transfusion. Not all patients allow transfusion due to their faith such as Jehovah's Witnesses (JW). In adolescents who are not legally considered to be adults, an ethical dilemma arises regarding the wishes of the patient, the family and the desires of the caretaker to transfuse prior to medical intervention. The use of erythropoietin (EPO) has been supported for patients who decline blood transfusion by increasing hemoglobin (Hgb) preoperatively. There are limited data on the use of EPO in gastrointestinal bleeding and only one report of the use of EPO in the setting of established Inflammatory Bowel Disease (IBD) in JW. We describe the first case of the use of EPO to improve hemoglobin and diagnose IBD in JW. Case Description A 17 year 11-month-old female presented with a 6-month history of worsening bloody diarrhea, severe abdominal pain and iron deficiency anemia. She reported fatigue, severe restriction in activity and weight loss. She denied extraintestinal manifestations of IBD or exposures for infection. Examination revealed a pale female with abdominal tenderness but no perirectal disease. Favoring ulcerative colitis (UC), the Pediatric Ulcerative Colitis Activity Index (PUCAI) was calculated to be 80. Stool studies for infection were negative. Labs on the first hospital day (HD) are in Table One. Esophagogastroduodenoscopy (EGD) and colonoscopy were recommended, but only after transfusions of packed red blood cells due to concern for further bleeding. Transfusions were declined for religious reasons as she and her family are practicing JW. To improve her Hgb, intravenous (IV) iron sucrose 100 mg was given daily for a total of 8 days. On HD#6, the ethics team convened. Our patient and family were interviewed separately. Consensus with the medical team, her and her family was that while clinicians have beneficence based obligations to their legally minor patients, adolescents can weigh the risks and benefits of multiple options, manipulate hypothetical situations, and have an evolving autonomy; therefore, their values and decisions should be respected. Blood products were withheld. A total amount of 120,000 units of EPO were given over the course of 14 days prior to procedures. While waiting for EPO to raise her Hgb to a suitable level, magnetic resonance enterography was obtained that showed diffuse thickening and enhancement of the colon representing colitis. She continued to have colitis, as well as suboptimal Hgb despite EPO and iron. Therefore, with the presumed diagnosis of UC, IV methylprednisolone 20 mg twice daily (BID) was started to avoid further blood loss. Her symptoms and Hgb improved within a few days. Labs on HD#15 after a week of therapy are shown below. An EGD/Colonoscopy performed on HD#17 revealed moderate colitis from the ascending colon to rectum with a normal appearing terminal ileum, confirmed by histology. Our patient was discharged on HD#19 on Prednisone 20 mg BID and a PUCAI of 15. Conclusion While it may be difficult for caretakers who wish to improve the health of their patients, there are instances where faith related reasons preclude using "standard of care." By means of novel approaches while respecting patient's wishes, such as using EPO to increase Hgb in a patient who is a practicing JW, making a preliminary radiographic diagnosis and initiating therapy prior to definitive testing, both the patient's wishes and treatment can be achieved without compromising either.

**80 USTEKINUMAB USE IN PEDIATRIC INFLAMMATORY BOWEL DISEASE.** O.M. Adeyemo, Pediatrics, William Beaumont Hospital, Royal Oak, Michigan, UNITED STATES. Fatima, Oakland University William Beaumont School of Medicine, Royal Oak, Michigan, UNITED STATES.

Ustekinumab is a human monoclonal antibody that targets the p40 subunit of both interleukin 12 and 23, and inhibits their activity. The drug is currently approved for the treatment of Psoriasis in adult patients. It was approved for moderate to severe Crohn's disease in adult patients in 2016. There is however limited data available

for the off-label use in pediatric inflammatory bowel disease. We aim to describe our experience of Ustekinumab in pediatric patients with Crohn's disease. Method: Retrospective chart review identified 6 patients receiving Ustekinumab for Crohn's disease. Indications for starting Ustekinumab was determined by the primary gastroenterologist. Patients between ages of 14 to 19 with refractory Crohn's disease were treated with Ustekinumab at a dose of 6 mg/kg IV, followed by 90 mg subcutaneous every 8 weeks. Results: All 6 patients with Crohn's had severe disease. The mean age was 16. All patients had previously failed at least 2 biologic agents and steroids. 4 patients were on concurrent steroids at initiation of Ustekinumab. 3 patients were able to be weaned off steroids. 1 patient was also on concurrent Tacrolimus. 2 patients required switching Ustekinumab from every 8 weeks to every 7 weeks. All but one patient had clinical improvement. The one patient without any clinical response had improved labs and fecal calprotectin. All patients remain on Ustekinumab. There were no reported adverse events. Conclusion: Ustekinumab use in selective pediatric patients with severe refractory Crohn's disease has so far proven to be safe and effective. Future studies are required to determine the long-term efficacy and safety in pediatric patients with Crohn's disease.

**82 SUCCESSFUL TREATMENT OF REFRACTORY COLLAGENOUS GASTROENTEROPATHY AND COLITIS WITH METHOTREXATE: A CASE REPORT.** B.C. Beinvogl, M. Verhave, Gastroenterology, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES. J. Goldsmith, Pathology, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES. R. Arumugam, M. Kennedy, Minnesota Gastroenterology, PA, St. Paul, Minnesota, UNITED STATES. Mokalla, Children's Minnesota, Minneapolis, Minnesota, UNITED STATES.

Introduction: Collagenous gastroenteropathy and colitis are exceedingly rare gastrointestinal diseases characterized by increased subepithelial collagen deposition, often with an associated inflammatory infiltrate in the lamina propria. Disease severity depends on the location and extent of gastrointestinal involvement. Symptoms range from mild symptoms of abdominal pain and anemia to life threatening diarrhea and malabsorption. Current treatment is based on anecdotal evidence including case reports only. We describe a 2 year old female with collagenous gastroenteropathy and colitis who was successfully treated with methotrexate. Case presentation: A 2 year old previously healthy female presented with a three month history of diffuse abdominal pain, watery diarrhea, emesis and weight loss. Work-up revealed anemia, peripheral eosinophilia, profound hypoalbuminemia and hypogammaglobulinemia. She required parenteral nutrition for nutritional support. Endoscopic examination showed villous blunting. Histopathology showed increased thickness of subepithelial collagen deposition and a mixed-cellular infiltrate in the lamina propria, consistent with collagenous gastroenteritis, ileitis and colitis. She was started on intravenous Methylprednisolone 1mg/kg twice daily for two weeks, followed by Prednisolone 1mg/kg by mouth twice daily for 2 weeks, followed by a four week taper. She was concurrently started on budesonide capsules and a Pulmicort slurry. She had resolution of diarrhea, normalization of serum albumin, improved enteral intake and was successfully weaned off parenteral nutrition within two weeks after initiation of intravenous steroids. Repeat endoscopic evaluation six weeks later revealed a normal histopathology. She remained on budesonide and Pulmicort slurry after completion of the steroid taper. Within one week of discontinuing steroids, however, she had new onset of abdominal pain, diarrhea, abdominal distension and anorexia. She did not respond to oral steroids and was therefore readmitted for intravenous steroids. She again responded well and completed a 4 week course, followed by a taper. Two weeks into the steroid course, we elected to start weekly intramuscular methotrexate 15mg/m<sup>2</sup> as a steroid sparing agent analogous to maintenance therapy for inflammatory bowel disease, along with daily folate supplementation. She remained asymptomatic off steroids and a repeat endoscopy after six months of treatment with methotrexate revealed normal histopathology. Methotrexate was continued for 12 months and she remained asymptomatic. She continues to remain in remission 3 months after discontinuation of methotrexate. Conclusion: We report the case of a 2 year old female with steroid dependent collagenous gastroenteropathy, who was successfully treated with intramuscular methotrexate as steroid sparing agent. She was treated for one year and remains well off therapy. This is the first report of the successful use of methotrexate in pediatric collagenous gastrointestinal disease. More studies are needed to determine the efficacy and safety of treating refractory pediatric collagenous gastroenteropathy and colitis with methotrexate.

**84 IDIOPATHIC JEJUNOILEITIS IN A PEDIATRIC PATIENT.** B. Ji, K. Murthy, Department of Pediatrics, Sinai Hospital of Baltimore, Baltimore, Maryland, UNITED STATES.

Background: Acute necrotizing enteritis or nonspecific jejunoileitis has been described for over 50 years, mostly in adults. The few pediatric cases are from South and South East Asia. A definite etiology has not been described but infectious causes and hypersensitivity reactions have been mentioned. Case description: 3-year-old previously healthy male presented to an outside hospital with 8 days of worsening abdominal pain and melena. Initial labs revealed elevated inflammatory markers, hypoalbuminemia, and anemia. His workup included EGD, colonoscopy, and Meckel's scan, which were negative. Stool studies, including H. pylori antigen and O&P, were negative. Fecal A1AT and calprotectin levels were 690mg/dL and 352.9mcg/g, indicating protein-losing enteropathy, possibly infectious. Parenteral nutrition, meropenem and metronidazole were initiated. Radiologic studies, including small bowel series and CT abdomen/pelvis, showed thickening of jejunal loops and mesenteric adenopathy, concerning for lymphoma. Patient underwent exploratory laparotomy with lymph node biopsy, which demonstrated moderately inflamed proximal jejunum with diffuse mesenteric lymphadenopathy. Lymph nodes showed only reactive changes. Methylprednisolone was initiated given evidence of jejunitis. Serologies for IBD and celiac disease were negative. Genetics for celiac disease was positive for HLA-DQ2. There was rapid clinical improvement within 24 hours of starting steroids. The patient was discharged home 3 days later on a steroid taper course, metronidazole, and PPI. Since then he has been asymptomatic without recurrence of abdominal pain, melena, fevers, or weight loss. He required a prolonged steroid taper due to iatrogenic adrenal suppression. His repeat small bowel follow-through 3 months after discharge was normal without evidence of jejunitis. Discussion: Nonspecific jejunoileitis is an uncommon, progressive, but predominantly self-limiting inflammation of small bowel without a definite etiology. Clinical presentation is non-specific and may include abdominal pain, emesis, diarrhea, and blood in stool, leading to a large differential diagnosis and work-up. Cultures are most often sterile and plain abdominal radiographs may fail to show evidence of disease, especially early in its course. In our case, contrast study showed significant mesenteric lymphadenopathy as well as thickening of the small bowel, helping to establish diagnosis. In reported cases, conservative treatments consisted of bowel rest and antibiotics. Rapid clinical response to steroid therapy seen in our patient may support an etiology of hypersensitivity reaction.

**86 INPATIENT PEDIATRIC GASTROENTEROLOGY PSYCHOLOGY CONSULTATION-LIAISON SERVICES: INITIAL DEVELOPMENT AND IMPLICATIONS FOR FUTURE DIRECTIONS.** B.L. Gresl, L. Iteld, Gastroenterology, Children's Health, Dallas, Texas, UNITED STATES. L. Gresl, L. Iteld, UTSW Medical Center, Dallas, Texas, UNITED STATES. Objective: While integrating pediatric psychology services is beneficial to meet the biopsychosocial needs of pediatric patients across medical settings, few studies have focused on the use of these specialized services in the inpatient setting. The aim of this study was to describe the development of an inpatient Gastroenterology-specific (GI) psychology consultation-liaison service over the course of the first year of implementation to highlight the rationale, process and utilization of specialized pediatric psychology services in an inpatient medical setting. Methods: Retrospective chart reviews were performed for pediatric patients (ages 1-19 years) admitted to the Gastroenterology service who received a GI Psychology consult order during admission. Demographic, diagnostic and psychosocial data were collected over a 1-year period (May 2016-May 2017) following the development of the GI Psychology consultation-liaison service. Results: Once the GI Psychology consult order was established in the electronic medical record, a total of 123 GI Psychology consults were ordered. Twenty-three consults were canceled and 14 were duplicates. Of the 86 consults completed (age = 12.9 years; 52% female), the top 3 diagnoses coded on admission included IBD (33.7%), functional disorders (29.1%) and constipation (19.7%). The primary indications for a consult were coping with hospitalization and mood. The majority of GI psychology consults were ordered within 24 hours of admission (51.2%). Average length of stay was 7.4 days. Following discharge, 33 (36%) patients followed up with a GI psychologist to receive continued outpatient services. Conclusions: The inpatient setting provides a unique opportunity to optimize patient care by integrating pediatric patients' physical and psychological well-being. This analysis demonstrates practice patterns in Gastroenterology, which can be used to examine the extent of the utilization of pediatric GI psychology services following the first year of development of an inpatient pediatric GI psychology consultation-liaison service. Challenges and implications for future directions are discussed.

**89 VEDOLIZUMAB THERAPY IN MODERATE TO SEVERE PEDIATRIC INFLAMMATORY BOWEL DISEASE.** C.S. Huang, Pediatric Gastroenterology, Valley Children's Hospital, Madera, California, UNITED STATES. BACKGROUND: Vedolizumab is effective for inducing and maintaining remission in adults with inflammatory bowel disease (IBD); however, there is limited pediatric data. This case series aimed to describe the clinical response to

vedolizumab in refractory pediatric IBD. **METHODS:** Disease activity indices, clinical response, concomitant medication use, and adverse events were retrospectively measured over 12 children with refractory IBD who had failed anti-tumor necrosis factor therapy and subsequently initiated vedolizumab therapy. **RESULTS:** Twelve subjects, 2 with Crohn's disease and 10 with Ulcerative Colitis, received vedolizumab. Clinical response was observed in 2/2 (100%) of Crohn's patients and 6/10 (60%) of ulcerative colitis patients of the evaluable subjects at week 14. Before induction, 11/12 (91%) participants were treated with systemic corticosteroids, as compared with 4/12 (33%) subjects at 14 weeks. Steroid-free remission was seen in 0/14 (0%) subjects at 6 weeks and 8/12 (66%) at 14 weeks. There was significant improvement in serum Hemoglobin, platelet count, ESR and C-reactive protein by week 14. There were no infusion reactions. Vedolizumab was discontinued in 3 patients because of severe colitis, requiring colectomy. Three out of 12 (25%) showed endoscopic remission at week 14-20. An average Vedolizumab level of 12-8 was observed at 6-8 wks maintenance dose. **CONCLUSIONS:** There is limited experience with vedolizumab therapy in pediatric IBD. There seems to be a marked number of subjects with clinical response in the first 6 to 14 weeks. No adverse events were observed while using vedolizumab. This study is limited by small sample size. A larger prospective study is warranted.

**90 CONTINUING INFLIXIMAB IN THE SETTING OF DISSEMINATED HISTOPLASMOSIS.** C.B. Cox, J. Whitworth, Pediatrics, University of Tennessee Health Science Center, Memphis, Tennessee, UNITED STATES. J. Whitworth, Pediatric Gastroenterology, Le Bonheur Children's Hospital, Memphis, Tennessee, UNITED STATES. Infliximab is a monoclonal antibody targeting TNF-alpha. It was approved in 1998 for the treatment of moderate to severe Crohn's disease, but its use increases risk for opportunistic infections. The Adverse Event Reporting System received 207 reported cases of infliximab related histoplasmosis as of April 2008. This was likely underreported. A paucity of literature exists describing medication management of infliximab therapy in the setting of disseminated histoplasmosis, but some have recommended its discontinuation. We describe the case of a 14-year-old female with steroid dependent Crohn's colitis who was unable to attain symptomatic or endoscopic remission despite mesalamine, azathioprine and prednisone. Six months after her diagnosis, infliximab 5 mg then 10 mg/kg was added to her regimen. The patient had relocated to the Mississippi River valley between the time of diagnosis and initiation of anti-TNF-alpha therapy. Six months after beginning infliximab she became ill with weight loss, malaise, dyspnea and fever, suggestive of opportunistic infection. Due to worsening pleuritic symptoms, bronchoalveolar lavage was performed. Both giemsa staining and serum and urine antigen testing confirmed a diagnosis of disseminated histoplasmosis, and the patient was treated with amphotericin-B and itraconazole. Subsequently, the azathioprine dose was reduced, infliximab was continued using serum level to guide dosing and oral itraconazole was continued. The patient experienced improvement in serial blood histoplasma-antigen levels as well as concomitant resolution of symptoms related to both disseminated histoplasmosis and Crohn's disease. To the author's knowledge, this is the first reported case of successful simultaneous treatment.

**91 GASTROINTESTINAL NEUROECTODERMAL TUMOR MIMICKING IBD.** C. Scherer, A. Koral, Pediatric Gastroenterology and Hepatology, Yale, New Haven, Connecticut, UNITED STATES. **Introduction:** Abdominal pain, weight loss, elevation of inflammatory markers, and anemia are common in the presentation of inflammatory bowel disease. However, these symptoms can also be due to gastrointestinal neuroectodermal tumor (GNET). GNET is an extremely rare intestinal tumor, most often presenting with signs of obstruction. We present a case of a 17-year-old male who was diagnosed with GNET after a suspected neoplasm was identified on MRE. **Case Report:** A 17-year-old male with chronic iron deficiency anemia presented with a 2-month history of abdominal pain and a 5 kg weight loss. The abdominal pain was episodic and severe, associated with emesis and nocturnal awakenings. Stooling patterns varied from constipation to diarrhea. Labs showed Hgb 12 g/dL, albumin 4 g/dL, CRP 48.2 mg/dL, ESR 26 mm/hr, and fecal calprotectin 851 mcg/g. A complete abdominal ultrasound did not show any inflammation, collections, or masses. Upper and lower endoscopy were visually normal and biopsies showed mild chronic gastritis. MRE showed an eccentric focal segment of thickened jejunal wall with 15 cm of proximal small bowel dilation, consistent with obstruction and concerning for a neoplastic process. During laparotomy an enlarged and firm lesion was identified in the small bowel with multiple enlarged lymph nodes in the surrounding mesentery. A small-bowel resection with primary anastomosis was performed. The tumor was identified as malignant GNET, also referred to as clear cell sarcoma-like gastrointestinal tumor. GNET is a highly malignant neoplasm. The resection had clean margins and a total of 16 lymph nodes were removed, none of which were malignant. PET scan and chest CT were negative for metastasis, and he has not

undergone any further treatment. After surgery, his symptoms of abdominal pain and emesis resolved. He has returned to regular activity. Discussion: GNET is an extremely rare gastrointestinal tumor, with less than 50 case reports published. GNET typically occurs in young to middle-aged adults, usually presenting with abdominal pain and intestinal obstruction. The tumor is highly malignant and associated with an overall poor prognosis. Diagnosis may be delayed due to the rare occurrence of the tumor. This case demonstrates the importance of continuing clinical investigation when faced with severe symptoms and an unclear diagnosis. GNET should be considered in patients presenting with symptoms suggestive of intestinal obstruction; imaging and surgery should not be delayed as the neoplasm is highly malignant.

**92 UNUSUAL PRESENTATION OF CUTANEOUS CROHN'S IN PEDIATRICS.** D. Hong, Pediatric Gastroenterology, Medstar Georgetown University Hospital, Washington, District of Columbia, UNITED STATES. C. Frelinger, Georgetown University Medical School, Washington, District of Columbia, UNITED STATES.

Introduction: Atypical presentations of Crohn's disease in pediatric patients have been well described without GI symptoms. However, presentation with vaginal and gluteal ulcers have been rarely described in the pediatric population for Crohn's Disease. Skin disorders are commonly seen in inflammatory bowel disease and represent a well-known extraintestinal complication. Skin disorders commonly seen in IBD are erythema nodosum, pyoderma gangrenosum, enterocutaneous fistula, skin tags, aphthous stomatitis. Here we present a teenager who presented with painful ulcers as her initial symptom for her Crohn's Disease. Case Report: 15 year old Africa-American female was initially admitted to the hospital for sudden onset of "painful rash" involving labia and gluteal folds for 1 week. They were ulcers varied in size, painful to touch, multiple (up to 4), some deep and containing purulent drainage with edematous borders. The patient denied history of sexual activity or trauma, no genitourinary symptoms, no arthralgia, GI symptoms or systemic concerns for growth, fever, or fatigue. Initial laboratory work up was significant only for markedly elevated ESR and CRP levels at presentation, thought to be related to an infectious process. STI screening, ANA, viral cultures/PCR for infections were negative. Magnetic resonance imaging studies of abdomen and pelvis were performed due to deep ulcerations involving gluteal folds and purulent nature, concerning for deeper tissue connection. The images did not show any gastrointestinal involvement but did show superficial sinus tract to the inferior coccyx. Patient was readmitted after several months for recurrence of the ulcers involving bilateral labia and gluteal lesions that were similarly painful and purulent. Similar repeat studies for infectious etiology were negative as during the initial presentation. The growth chart revealed her weight and height plateaued over two-year period without any gain. Esophagogastroduodenoscopy (EGD) and colonoscopy as well as a skin biopsy were performed due to growth concerns as well as the waxing and waning features of the ulcers. EGD and colonoscopy showed chronic inflammation of the esophagus, stomach and right colon with architectural distortion with negative tissue cultures. Her skin biopsy showed neutrophil rich dermal infiltrate with multinucleated giant cells and ulcerations. She was started on infliximab and topical tacrolimus with marked improvement and resolution of all her ulcers after 4 months of therapy. Discussion: In some cases, non-granulomatous skin disorders can occur as a reaction to the intestinal disease such as neutrophilic dermatosis. Although this patient later demonstrated hallmarks of inflammatory bowel disease involving poor growth, abdominal pain, diarrhea, her primary presenting symptom was cutaneous Crohn's disease. Skin manifestations of inflammatory bowel disease occur up to 40% of patients and can vary widely, can parallel intestinal disease involvement but also occur independently of any gastrointestinal symptoms and signs of inflammation. Conclusion: Given our case report with atypical skin findings in newly diagnosed Crohn's disease in a teenager, there should be higher index of suspicion and vigilance paid towards skin findings to suspect inflammatory bowel disease.

**93 LIVER ABSCESS IN A PEDIATRIC PATIENT WITH ULCERATIVE COLITIS.** D.F. Castillo, R. Caicedo, V. Gopalareddy, Pediatrics, Carolinas Medical Center, Charlotte, North Carolina, UNITED STATES.

Hepatic abscesses are a known but rare extra-intestinal manifestation of ulcerative colitis (UC), with only ten cases described in the literature. We present a 16-year old patient with steroid-dependent UC who developed liver abscesses. This is to our knowledge the first report of a pediatric UC patient with this complication. He presented with fevers, right upper quadrant pain, and jaundice a few weeks after being treated with IV corticosteroids and completing an induction regimen of infliximab. He had elevated alkaline phosphatase, transaminases, bilirubin levels, and C-reactive protein. Imaging revealed multiple abscesses in the right hepatic lobe as well as right portal vein thrombosis and diffuse colonic wall thickening. Abscesses were drained multiple times and cultures were polymicrobial. His course was complicated by septic shock soon after surgical drainage and an extended ICU stay,

requiring parenteral nutrition. He improved with IV antibiotics, and he transitioned to exclusive enteral nutrition therapy after approximately a month. By six months post-admission, the fluid collections had entirely resolved. Over this period of time, he did not show any symptoms of an ulcerative colitis flare while remaining off immunosuppressive medications. We discuss what risk factors may have predisposed our patient to develop hepatic abscesses at such an early age, as well as discussing the diagnostic evaluation, management, and outcomes. We also speculate on why his UC has remained in remission.

**94** *MESALAMINE-INDUCED MYOPERICARDITIS IN A NEWLY DIAGNOSED ULCERATIVE COLITIS PATIENT.* L. Alrabadi, D. Rosen, Pediatric Gastroenterology and Hepatology, Yale University, Rye Brook, New York, UNITED STATES.

Introduction: Cardiac involvement in IBD can be classified as an extra-intestinal manifestation or as a side effect of medication, most commonly mesalamine or azathioprine. Here we describe a 14-year old male with ulcerative colitis who developed mesalamine-induced myopericarditis and rapidly recovered with cessation of treatment. While mesalamine is generally considered a safe and well tolerated medication, it is important to recognize this rare but potentially fatal complication. Case Report: A 14-year old male presented with a 4-week history of abdominal pain, bloody stools, anorexia and weight loss. Physical exam was unremarkable except for mild abdominal tenderness. Laboratory evaluation showed anemia (Hgb 11.5g/dL) and an elevated erythrocyte sedimentation rate (67 mm/hr). Endoscopic evaluation showed left-sided inflammation extending to the descending colon. Biopsies confirmed a diagnosis of ulcerative colitis and he was started on a 5-ASA medication. Two weeks after initiation of treatment he developed acute chest pain and fever to 103 Fahrenheit. Of note, he had seen his pediatrician the week before for low grade fever and tested positive for influenza. Further evaluation yielded findings of ST-elevation on electrocardiogram concerning for myocarditis. Troponin T level was elevated at 0.46ng/ml (normal <0.01ng/ml). An initial echocardiogram showed the left ventricular free wall to appear thickened representing possible focal edema versus myocarditis. Mesalamine was immediately stopped due to the association with myocarditis. A cardiac MRI showed delayed enhancement of the basal inferolateral and inferior walls extending to the mid ventricle, delayed enhancement of the visceral and parietal pericardium and a moderate pericardial effusion all consistent with acute myopericarditis. He was started on a 3-day course of high dose steroids and given intravenous immunoglobulin therapy. Troponin levels continued to downtrend and a follow-up echocardiogram showed improved left ventricular systolic function and resolution of pericardial effusion. Subsequently, he completed a steroid taper and was started on vedolizumab for management of his ulcerative colitis. Discussion: Mesalamine-based medications are considered first-line treatment for mild ulcerative colitis for both induction and maintenance of remission. These agents are generally well tolerated, although commonly reported side effects include nausea, vomiting, headache, pancreatitis, interstitial nephritis and paradoxical diarrhea. Mesalamine-induced cardiac inflammation is thought to be a hypersensitivity reaction, although the exact mechanism is unknown. Typical symptoms include acute onset of fever, chest pain, and shortness of breath within 14 to 28 days after starting medication. Cessation of mesalamine will often reverse the cardiac complications and most patients will have a full recovery. Due to a high likelihood of recurrence the use of other mesalamine-based agents, including mesalamine enemas and sulfasalazine, is contraindicated. The described patient had a recent history of influenza, and so viral-induced myopericarditis was considered as a possible etiology. However, his rapid recovery after cessation of mesalamine suggests that his symptoms were likely due to the medication. It is important for pediatric gastroenterologists to be aware that while 5-ASAs are generally well tolerated, the association with cardiac inflammation is a rare but potential side effect that needs to be immediately recognized.

**96** *CANNABIS OIL USE BY ADOLESCENTS AND YOUNG ADULTS WITH INFLAMMATORY BOWEL DISEASE.* E.J.

Hoffenberg, Pediatrics-GI, U Colorado Denver, Aurora, Colorado, UNITED STATES. E.J. Hoffenberg, B. Murphy, Children's Hospital Colorado, Aurora, Colorado, UNITED STATES. Mikulich-Gilbertson, S. McWilliams, C. Hopfer, Psychiatry, University of Colorado Denver School of Medicine, Aurora, Colorado, UNITED STATES. Hoffenberg, Preventive Medicine, University of Colorado Denver School of Medicine, Aurora, Colorado, UNITED STATES. Background Cannabis in multiple forms is used for recreational and medical purposes including by patients with IBD. Little is known about frequency and motivations for use of cannabis oil (CO) products. In an ongoing study, about 10% of our adolescent and young adult patients with IBD endorsed use of CO. Here we describe in detail the 9 subjects who reported CO use. Methods Subjects are from a prospective observational study of patients with IBD 13-23 years of age followed at our institution in which approximately 70% of those approached agreed to

enroll. Standardized clinical data were obtained at each clinic visit using ImproveCareNow electronic data capture. Cannabis use patterns and motivations were assessed by validated questionnaires developed for this study. Results Of 97 enrolled participants, 28 endorsed use of any cannabis product including 9 who used CO. Of these 9, 4 had a state-issued medical use card and 5 had a detectable cannabinoid serum level. There was no difference between CO users (n=9) and non users (n= 88) for age (median 16y, range 13-22), gender, race, disease type, physician global assessment of disease activity, weight z-score or pain score. The CO group reported a higher appetite score ( $p=0.03$ ) on a visual analog scale. Median and range frequency of use in the past 30 days was 25 (6-30) days and 30 (6-120) times. Route of use was swallowed 4, sublingual/tincture 4, rectal suppository 1. Ratio of CBD to THC was most commonly 1:1 (n= 3), 1:19 (n=1), unknown (n=3), and 98:1 in suppository (n= 1). Use for medical reasons was reported by 7 of 9, most commonly for relief of abdominal pain" (n=6) and "physical pain" (n=5). Reported impact of CO use is shown in the Table. Discussion This is the first study to describe that about 10% of adolescents and young adults with IBD use CO products. A significant increase in appetite was observed. CO users also perceived favorable improvements in pain, sleep quality, nausea, and appetite. Physicians treating youth with IBD should be aware of CO use for self-perceived medicinal purposes. However, daily or almost daily use of cannabis products is concerning due to reported increased risks for motor vehicle accidents, drug dependence with anxiety, depression and psychotic disorders, and for psychosocial decline. Providers should become comfortable in asking about cannabis and CO use."

**98 REVERSAL OF ANTIBODY FORMATION IN PEDIATRIC INFLAMMATORY BOWEL DISEASE ON BIOLOGIC THERAPY.**  
E. Kang, Case Western Reserve University, Cleveland, Ohio, UNITED STATES. Khalili, J. Splawski, T. Sfera, J. Moses, Division of Gastroenterology, Hepatology and Nutrition, UH Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES.

Background Development of a humoral immune response to anti-tumor necrosis factor (anti-TNF) agents is associated with loss of therapeutic response in patients with inflammatory bowel disease (IBD). Several studies in adults with IBD have demonstrated reversal of immunogenicity is possible with dose intensification, drug interval shortening, and the addition of an immunomodulator. However, in children with IBD and anti-drug antibodies (ADA) to anti-TNF antagonists, there is limited reported information regarding reversal of immunogenicity. Herein, we report four pediatric IBD patients with successful reversal of immunogenicity on an anti-TNF agent. All testing for the presence of antibodies utilized a homogeneous mobility shift assay (HMSA) that can reliably measure ADA even in the presence of detectable anti-TNF levels. Case 1: A 12-year-old male with ileocolonic Crohn's Disease (CD) was induced and maintained on infliximab with oral low dose methotrexate (MTX) due to the severity of his disease. His regimen was discontinued due to disseminated histoplasmosis. He later restarted infliximab without an immunomodulator at a dose interval of every 7 weeks and developed antibodies after his 15<sup>th</sup> infusion. His interval was decreased to every 4-5 weeks but ADA persisted. MTX was restarted and his ADA resolved after his 20<sup>th</sup> infusion. Case 2: A 17-year-old female with pancolonic ulcerative colitis, diagnosed 10 years earlier, was being managed on azathioprine (AZA) and prednisone. With worsening of disease on endoscopy, she was induced and maintained on infliximab. ADA were detected after her 11<sup>th</sup> infliximab infusion. Her dose was increased and subsequently the dose interval was shortened, with continuation of AZA. ADA resolved with dose interval shortening. Case 3: A 15-year-old male with CD of the ileum, stricturing type, diagnosed 6 years earlier, was being managed on AZA. He was hospitalized for a Crohn's exacerbation and started on standard infliximab with low dose oral MTX. He developed antibodies after his 6<sup>th</sup> infusion and was switched to adalimumab, in addition to oral high dose MTX. He initially demonstrated ADA but cleared his antibodies after dose interval of adalimumab was shortened from every two weeks to every week. Case 4: A 13-year-old male with ileocolonic CD, diagnosed 7 months earlier, was being maintained on AZA and steroids. After lack of clinical response, he was induced and maintained on standard infliximab with continuation of his AZA. He developed ADA after his 7<sup>th</sup> infusion. ADA resolved with a dose increase to around 10 mg/kg. Conclusion Loss of response to anti-TNF agents is a major consideration, and reversal of immunogenicity can re-establish response and increase the durability of these agents. We present the specific sequence of interventions used for reversal of immunogenicity as a guide for the practicing gastroenterologist dealing with loss of response due to ADA formation. In line with the findings in adult IBD patients, our results suggest that dose adjustments and/or the addition of an immunomodulator may be effective strategies to reverse immunogenicity in pediatric IBD patients treated with infliximab or adalimumab.

**99 PULMONARY NODULES IN CROHN'S DISEASE: DISTINGUISHING DISEASE MANIFESTATIONS FROM SIDE EFFECTS OF MEDICAL THERAPY.** E. Berg, A.A. Mencin, K.G. Margolis, J. Picoraro, Department of Pediatric Gastroenterology, Hepatology and Nutrition, Children's Hospital of New York - Columbia University Medical Center, New York, New York, UNITED STATES. S. Ayyala, Department of Radiology, Children's Hospital of New York - Columbia University Medical Center, New York, New York, UNITED STATES. H. Eichenfield, Department of Allergy, Immunology and Rheumatology, Children's Hospital of New York - Columbia University Medical Center, New York, New York, UNITED STATES. V.P. Duron, Department of Surgery, Children's Hospital of New York - Columbia University Medical Center, New York, New York, UNITED STATES.

Extraintestinal manifestations of inflammatory bowel disease (IBD) commonly involve the skin, eyes, joints and hepatobiliary system, and pulmonary manifestations have been described with increasing frequency<sup>1</sup>. Pulmonary findings in IBD may influence treatment decisions as the clinician must determine whether the underlying cause is a manifestation of the disease or a secondary outcome of treatment (i.e., a drug reaction or opportunistic infection). Pulmonary biopsy findings most commonly seen in IBD include granulomata, organizing and eosinophilic pneumoniae, and lymphocytic infiltrates<sup>6</sup>. A 12 year old boy with Crohn's disease and polyarticular arthritis without respiratory symptoms, developed incidental pulmonary nodules. Three years earlier he presented to another hospital with abdominal pain, constipation, bloody stools and joint pain and was diagnosed with ulcerative colitis. Following interval improvement over two years with two courses of steroids plus mesalamine and then 6-mercaptopurine and mesalamine as combined maintenance agents, bloody diarrhea and joint pain recurred. Repeat colonoscopy at our institution confirmed a diagnosis of Crohn's disease. Infliximab was initiated, mesalamine was continued, and the patient demonstrated clinical response until five months later when bloody diarrhea and joint pain recurred. Infliximab dosing was increased, budesonide was added, and bloody diarrhea improved; however, he developed acute onset of tinnitus, trismus, and nightly fevers. A head and neck CT confirmed bilateral temporomandibular joint (TMJ) effusions; fluid studies from arthrocentesis were consistent with reactive arthritis. An incidental finding of ground glass opacities and a solitary pulmonary nodule in the superior lung fields was detected. Chest CT showed a peripheral distribution of multiple sub centimeter ground glass nodules throughout both lungs, some with a solid component. The differential diagnosis included pulmonary emboli given the pattern of distribution and increased risk for thrombotic events in IBD although unlikely as the patient had no respiratory distress; tuberculosis given the history of immunosuppression, foreign travel and an indeterminate quantiferon gold; infectious abscess<sup>2</sup>; drug reaction<sup>3</sup> as can be seen with sulfonamides particularly mesalamine; or extra-intestinal manifestation of IBD<sup>1, 4, 6</sup>. Other considerations included fungal infection, parvovirus, foreign body aspiration, and lymphoma. To determine the appropriate management, an accurate diagnosis was essential because an infectious process secondary to immunosuppression would prompt de-escalation of immunosuppression and initiation of antimicrobial therapy whereas a manifestation of his underlying inflammatory disease would lead to escalation of immunosuppression. He underwent video-assisted thoracoscopy for lung biopsy, which demonstrated a single, necrotizing granuloma near the airway and mild, chronic inflammation around several airways, which was most consistent with inflammatory bowel disease of the lungs. AFB and GMS stains were negative, decreasing the likelihood of tuberculosis or fungal infection, respectively. Therapy for his Crohn's disease was escalated to include methotrexate in addition to increased dosing of infliximab and he has since had decreased stool frequency, less abdominal and joint pain, and improved school participation. He has not developed respiratory symptoms on escalated immunosuppressive therapy and repeat pulmonary imaging is pending.

**100 THE ROLE OF VEDOLIZUMAB IN THE TREATMENT OF CROHN'S DISEASE IN THE SETTING OF A PATIENT WITH DIGEORGE SYNDROME: A CASE REPORT.** E.M. Thau, H. Moore, P. Stein, Section of Gastroenterology, Hepatology and Nutrition, St Christopher's Hospital for Children, Philadelphia, Pennsylvania, UNITED STATES.

DiGeorge Syndrome (DGS) is a syndrome that affects multiple organ systems including the gastrointestinal tract (GI) as well as immune function. While DGS has a wide array of clinical manifestations, inflammatory bowel disease (IBD) is rare and infrequently reported on. IBD treatment becomes complicated in the setting of the immunosuppressed patient. An 8 year old boy with a history of DiGeorge syndrome with resultant hypoparathyroidism and hypogammaglobulinemia presented to gastroenterology clinic with failure to thrive as well as longstanding constipation with blood streaked hard stools. The bloody stool resolved with miralax. He tracked along the 3<sup>rd</sup> percentile for weight. Six months later he presented to the emergency room with bloody diarrhea and dehydration with ESR up to 109 mm/hr. Infection was ruled out. An

esophagogastroduodenoscopy (EGD) and colonoscopy were performed which showed gross colonic inflammation and ulceration in the colon and ileum. Biopsy was consistent with the gross appearance, showing cryptitis, crypt abscesses with multinucleated giant cells. Given his status, it was advisable to minimize further immunosuppression. Prednisone and pentasa were started in an effort to induce clinical remission, with the goal of continuation on pentasa for maintenance. He responded well to steroids but on pentasa alone, despite multiple increases in doses, the patient's CD would flare. He failed weaning of his monthly IVIG infusion after he required hospitalization with intubation for pneumonia. As he had ongoing symptoms with intermittent flares requiring steroids, he needed a different treatment regimen. He was induced into remission with prednisone and was started on monotherapy of vedolizumab with the current regimen of 6 mg/kg every 8 weeks. Since initiation of vedolizumab one year ago, the patient has remained in clinical remission. He has been asymptomatic with calprotectin of 30. He has gained weight well over the past year and is currently at the 25% weight for age. This patient demonstrates a challenging case wherein a child had difficulty to treat Crohn's disease in the setting of a primary immunodeficiency. His case was challenging in that typical agents used to treat IBD such as TNF- $\alpha$  inhibitors have strong immunomodulatory effects. How much would adding this type of medication affect a patient that is already immunosuppressed and at high risk for life-threatening infections? Vedolizumab is a newer agent within pediatric IBD and we are still figuring out the best candidates to use it for as well as determine the appropriate timing to compare this agent to others. Vedolizumab inhibits  $\alpha 4\beta 7$  integrin and thus works by inhibiting T-cell mediated lymphocyte migration and ultimately decreases inflammation along the intestinal epithelial tissue. As this medication is selective where it acts, it limits systemic immunomodulation. This patient has negligent B-cell activity, and would be unlikely to develop antibodies against this agent, a consequence that could eventually lead to treatment failure. Ultimately, this case offers further support that vedolizumab is promising as not only an alternative agent to achieve clinical improvement in refractory IBD patients, but it also provides an affective alternative therapy in pediatric patients with primary immunodeficiencies.

**105 CASE EXAMPLE OF AN EMERGING ADULT WITH REFRACTORY CROHN'S DISEASE: MANAGING THE COMPLEX INTERPLAY BETWEEN DISEASE AND PERSONALITY DURING TRANSITION TO ADULT-CENTERED CARE.** H. Person, Pediatrics, The Susan and Leonard Feinstein Inflammatory Bowel Disease Clinical Center at Mount Sinai, New York, New York, UNITED STATES. A. Keefer, Medicine, Gastroenterology, The Susan and Leonard Feinstein Inflammatory Bowel Disease Clinical Center at Mount Sinai, New York, New York, UNITED STATES.

Ms. D is a twenty-three year old woman with refractory small-bowel CD currently well-managed on ustekinumab for the past four years after failing courses of mercaptopurine, adalimumab, infliximab, and certolizumab. She missed extensive amounts of school as a child and had limited exposure to making or maintaining relationships outside of the family unit. This was developmentally impeding, and she struggled to individuate herself from her parents, succeed in educational goals, and assume responsibility for her medical and psychiatric care. She had been followed by psychiatrists since her teens for mood symptoms, receiving different diagnoses, including Major Depressive Disorder, Generalized Anxiety Disorder, and Attention Deficit Hyperactivity Disorder. By age twenty-two, Ms. D reported worsening depression and was enrolled in our IBD Medical Home, GRITT-IBD<sup>TM</sup> for comprehensive care. She reported low mood, irritability, troubles concentrating, low energy, and low appetite. She was enrolled in a weekly psychotherapy program, where she focused on intense feelings of loneliness, impulsivity, and unstable relationships. These symptoms were significantly impairing, and she withdrew from full-time college coursework and became increasingly alienated from her family and other social supports. In probing her transition to adult-centered care, Ms. D reported feeling invalidated by her parents throughout childhood and distressed at the severity of her CD. Her feelings of parental invalidation stemmed from her parents being deeply concerned with her health, yet seeming dismissive of the patient's desires and controlling of her actions. The patient completed a Young Schema Questionnaire, revealing maladaptive schemas in emotional deprivation, abandonment, social isolation, and mistrust. She was given a diagnosis of Borderline Personality Disorder (BPD). BPD is a pattern of maladaptive feelings and behaviors that result in emotion dysregulation, impulsivity, irritability, and unstable relationships. Both genetic and psychosocial factors contribute to its development, including growing up in an invalidating emotional environment and experience of trauma. Given Ms. D's reports of disease-related trauma and being invalidated by her parents because of having CD, this raised the question of whether her complex IBD course predisposed her to developing maladaptive personality traits. While distress and alteration of parent-child relationships may be a result of pediatric IBD, little is known about the development of BPD in IBD. While positive personality characteristics have been associated with improved bowel health, how BPD

may influence disease course and outcome is unclear. Psychotherapy included individual dialectic behavioral therapy. This therapy combines validation of feelings and experiences while emphasizing the development of emotion regulation skills, including interpersonal effectiveness, distress tolerance, mindfulness, and self-care. She was able to create a new narrative around having CD, build commitment to an effective treatment regimen, and learn core self-management skills that she had neglected to learn as a teenager. She was subsequently referred to an intensive personality disorder treatment program. Here we present the case of a twenty-three year old woman with a complex CD course, marked by trauma and parent-child relational conflict, who was newly diagnosed with BPD. Much is to be explored regarding the relationship between personality disorders and IBD, including how parental factors and skills deficits might promote the development of maladaptive personality characteristics and impact transition into young adulthood.

**108** *DYSPHAGIA AND ODYNOPHAGIA: AN UNUSUAL PRESENTATION OF PEDIATRIC CROHN'S DISEASE.* M. Jensen, Pediatrics, Sanford Children's Hospital, Sioux Falls, South Dakota, UNITED STATES.

Introduction: Dysphagia and odynophagia are rare complications of Crohn's disease. While upper gastrointestinal involvement in Crohn's disease is more common in pediatric patients than in adult patients, it still remains uncommon for pediatric patients with Crohn's disease to have esophageal involvement. Symptoms of esophageal involvement can include dysphagia and odynophagia. However, it is much more unusual for esophageal involvement to be the only presenting clinical symptom of Crohn's disease. Esophageal symptoms in Crohn's disease are more likely to be seen in patients with advanced ileocolonic disease. We report a 10 year old girl who presented with dysphagia and odynophagia whose evaluation revealed a new diagnosis of Crohn's disease. Case History: A previously healthy 10 year old girl presented with acute onset of dysphagia and odynophagia for the past couple of months. Prior to her GI evaluation she was placed on a PPI and had slight improvement in symptoms. She denied abdominal pain, diarrhea, constipation, or blood in the stools. She denied nausea, vomiting, change in appetite or weight loss. Of note, her growth charts revealed both a weight and height that had been near the 1<sup>st</sup> percentile for the past 3 years, but prior to that had only been near the 3<sup>rd</sup>-5<sup>th</sup> percentiles. Evaluation included normal labs: CBC, CMP, CRP, ESR, celiac, and thyroid testing. She had a normal Esophagram. Her PPI was increased to a higher dose and her symptoms resolved. Despite resolution of her symptoms, we proceeded with an upper endoscopy and she was found to have 3-5 white spots in her mid-esophagus, but otherwise a visually normal esophagus, stomach, and duodenum. Biopsies of the esophagus revealed chronic and acute inflammation with granuloma formation, and gastritis with focal granuloma formation. She underwent further lab work-up for possible causes including normal Quantiferon, HIV screening, RF, ANA, ANCA, peripheral smear, and ACE. Chest x-ray was clear. CT enterography revealed mild wall thickening in the distal and terminal ileum involving approximately 10 cm length of bowel. She underwent Colonoscopy which revealed ulcerated, congested, and friable mucosa in the terminal ileum and visually normal colon. Biopsies confirmed chronic ileitis with moderate activity. She was diagnosed with Crohn's disease with involvement of the esophagus, stomach, and terminal ileum. Per parental request, she was started on infliximab infusions as therapy. Conclusion: Dysphagia and odynophagia as the only presenting symptoms are a rare presentation of pediatric Crohn's disease. Esophageal manifestations of Crohn's disease are uncommon and are more likely to be seen in pediatric patients with advanced ileocolonic Crohn's disease. Treatment with a PPI can improve the symptoms of dysphagia and odynophagia, but will not treat the underlying cause. Thus, patients must undergo further evaluation of atypical symptoms.

**109** *PANCYTOPENIA ASSOCIATED WITH A NUDT15 GENE VARIANT IN A CHILD TREATED WITH AZATHIOPRINE FOR ULCERATIVE COLITIS.* C. Rajendra, V. Shenoy, Y. Wild, T. Hammoudi, G. Landman, A. Koegel, M. Hermiston, L. Cooke, M.B. Heyman, S.G. Verstraete, Pediatrics, University of California, San Francisco, San Francisco, California, UNITED STATES. Shah, K. Cordora, M. Kinnebrew, Dermatology, University of California, San Francisco, San Francisco, California, UNITED STATES.

The NUDT15 gene variant is associated with intolerance to thiopurines in patients with acute lymphocytic leukemia. We report a child with a NUDT15 gene variant on azathioprine (AZA) for Ulcerative Colitis (UC) who developed severe pancytopenia, brittle nails and alopecia. A 12-year-old Asian girl with a 2.5-year history of ulcerative colitis developed worsening bloody diarrhea and abdominal pain while on maintenance mesalamine medication. Despite initial treatment with budesonide (Uceris), corticosteroid enemas, metronidazole, and probiotics (VSL#3), she had persistent abdominal pain, bloody diarrhea and tenesmus. Lab tests revealed Hgb

11.7g/dL Albumin 3.7g/dL, CRP <1.0, ESR 22, and no other abnormalities. Thiopurine-S-methyltransferase (TPMT) level was sent, and she was started on AZA (0.5mg/kg) with gradual improvement. TPMT level was normal, and WBC was  $7.4 \times 10^9$ /L. She increased AZA to full dose (2.0g/kg) daily. Three weeks after initiation of medication she developed nail thinning; one week later she developed brittle nails and alopecia. Lab work revealed WBC  $1.5 (ANC 0.2) \times 10^9$ /L, Hgb 10.7g/dL and platelets  $79 \times 10^9$ /L. AZA was discontinued. Two days later, lab results included WBC  $2.0 (ANC 0.12) \times 10^9$ /L, Hgb 10.4g/dL and platelets  $78 \times 10^9$ /L. She returned to the ER and was readmitted 1 week later with fever, pharyngitis and rhinorrhea. Lab results revealed WBC  $1.5 (ANC 0.01) \times 10^9$ /L, Hgb 8.2 g/dL and platelets  $28 \times 10^9$ /L. She had tonsillar swelling with exudate, anterior cervical adenopathy and nasal discharge. Tests for viral and bacterial infections were negative. Intravenous cefepime was given. She continued with intermittent fevers, abdominal cramping and pancytopenia for the next 4 days. Antimicrobial coverage was broadened, and UC medications were held, as she had no evidence for a flare. Bone marrow biopsy showed a hypocellular bone marrow, and flow cytometry was reassuring with findings consistent with bone marrow suppression. By day 4, her lowest levels were reached: Hgb 7.9g/dL, WBC  $1.1 (ANC 0.01) \times 10^9$ /L and platelets  $20 \times 10^9$ /L. She received multiple blood and platelet transfusions and was treated with GM-CSF. Fever resolved on Day 5. She was discharged after 10 days of hospitalization. On discharge, labs revealed Hgb 8.1 g/dL, WBC  $3.7 (ANC 0.67) \times 10^9$ /L and platelets  $49 \times 10^9$ /L. NUDT15 gene variant testing sent after considering her clinical setting (presentation, race, drug exposure) was positive for the abnormality. The NUDT15 gene variant is more common in Hispanic and Asian populations. The NUDT15 gene encodes an enzyme that is a negative regulator of thiopurine activation and toxicity. Patients with a NUDT15 gene variant display poor metabolism of thiopurines and associated thiopurine-induced leukopenia. By 7 weeks after discontinuation of AZA, all lab tests in our patient normalized. She continued on mesalamine (Pentasa) for treatment of UC with sustained remission. Supported in part by NIH T32DK007762 (CR) and by the Cystic Fibrosis Foundation Grant (Shenoy 16B0)

**110 HEPATOCELLULAR ADENOMA IN AN ADOLESCENT WITH CROHN'S DISEASE TREATED WITH AZATHIOPRINE AND INFLIXIMAB.** L. Cooke, S.G. Verstraete, M.B. Heyman, S. Rhee, Pediatrics, University of California, San Francisco, San Francisco, California, UNITED STATES. Umetsu, Pathology, University of California, San Francisco, San Francisco, California, UNITED STATES.

The risk of malignancies including colorectal carcinoma and lymphoma in children with Crohn's disease is well known. We report a case of a large hepatocellular adenoma in a teenager on immunosuppression for Crohn's disease. A 16-year-old male post gastroschisis repair had been diagnosed in 2009 with ileocolonic and perianal fistulizing Crohn's disease and had been stable on long term treatment with azathioprine 1.4 mg/kg (since April, 2009) and infliximab 5 mg/kg (since February, 2011). About 6 months prior to admission, on routine surveillance laboratory testing, he was noted to have slightly elevated GGT (28 IU/L; normal range 7-26 IU/L). GGT continued to increase slowly (peak at 42 IU/L on day of admission). Aspartate transaminase (AST), alanine transglutaminase (ALT), alkaline phosphatase, and total and direct bilirubin levels were normal throughout this time. ESR, CRP and ferritin levels were elevated above normal values. The patient maintained a normal pCDAI ( $\leq 10$ ). Due to concerns for development of liver disease (e.g., PSC), abdominal including hepatobiliary ultrasonography revealed hepatosplenomegaly with an 8 centimeter right hepatic mass and normal blood flow by Doppler. He was admitted and had a right hepatectomy. Histology revealed a well-differentiated hepatocellular neoplasm with isolated arterioles, scattered lymphocytic inflammation, and patchy sinusoidal dilatation, with no cytologic atypia. Immunohistochemical stains for CRP and SAA were diffusely positive, consistent with inflammatory variant hepatocellular adenoma. LFABP showed normal intact expression, excluding HNF1-alpha inactivated adenoma. Beta-catenin stain did not show nuclear staining. Glutamine synthetase was negative. Reticulin stain was intact. Serum alpha-fetoprotein obtained after surgery was normal. He continues on azathioprine and Infliximab with no problems. Ongoing monitoring includes liver enzymes every three months and AFP every six months. Large hepatocellular adenomas are uncommon in pediatric patients. While no reports of hepatocellular adenomas associated with Crohn's disease and its treatment are currently in the literature, whether his immunosuppression with azathioprine and/or infliximab contributed to the size of his lesion is unknown.

**112 ISOLATED PERIANAL ABSCESS IN AN ADOLESCENT: IS IT ALWAYS CROHN'S DISEASE.** N. Mendez, J. Markowitz, Pediatric Gastroenterology, Northwell Health, Englewood, New Jersey, UNITED STATES. Navetta-Modrov, A. Jongco III, Allergy & Immunology, Northwell Health, New Hydr Park, New York, UNITED STATES.

Crohn's disease (CD) is a chronic inflammatory bowel disease (IBD) characterized by granulomatous inflammation thought to be caused by an abnormal response of the enteric immune system to normal enteric flora. No curative therapy exists. CD is not associated with systemic infection except in the setting of immunosuppressive therapy. Perianal abscess and fistula are common, and optimally treated with an anti-TNF agent. Chronic granulomatous disease (CGD) is a primary immunodeficiency disorder of phagocytes resulting in impaired killing of bacteria and fungi. Recurrent infections are common and GI tract involvement including the esophagus, stomach, colon and perianal abscess/fistula have been described. Histology is characterized by granulomatous inflammation and abscesses. Bone marrow transplant is curative. Anti-TNF therapy for CGD is contraindicated due to increased morbidity and mortality (Uzel, G. <i>Clin Infect Dis.</i>2010) We report an unusual case of an adolescent presenting with isolated chronic perianal abscess and fistula suggestive of CD, who ultimately was found to have CGD. CASE REPORT: A previously healthy 17 yr old male presented with a perianal abscess leading to an evaluation for IBD. He had no past history of severe, recurrent or chronic infection and no family history of CD or immunologic disease. Abdominal CT revealed a 5.6 cm left ischioanal abscess with no obvious fistula. The patient required incision and drainage (I&D) to control discomfort, but 2 months of continued discomfort and drainage prompted GI evaluation for possible CD. MRE revealed only a perianal fistula and suggestion of distal proctitis. Upper endoscopy/biopsy was normal. Colonoscopy revealed scattered nonspecific aphthous ulcers in the rectum but no inflammation or granulomas were identified histologically. The findings were not considered sufficient to diagnose CD and the patient returned to the surgeon for further management. He underwent a second I&D with seton placement and remained relatively asymptomatic for a few months. An advancement flap procedure was cancelled after a second abscess requiring I&D and antibiotics was noted. Subsequently, the original abscess recurred, requiring another I&D, seton and antibiotics. He returned to GI for consideration of anti-TNF therapy. Additional labs included a dihydrorhodamine (DHR) test that was abnormal, suggesting possible autosomal recessive CGD. Immunology consultation confirmed the diagnosis as genetic testing was positive for the c.75\_76delGT variant in NCF1 gene. The patient required hospitalization for pain control/persistent drainage, ultimately requiring diverting ileostomy. He is currently undergoing evaluation for hematopoietic stem cell transplant. CONCLUSIONS: This case outlines the importance of recognizing that perianal fistulas or abscesses may be the sole finding in some individuals with CGD. Differentiating between CGD and Crohn's disease is critical, as anti-TNF agents are first line therapy for fistulizing CD but contraindicated in CGD due to increased frequency of severe infections and death. CGD should be considered in the differential diagnosis of all patients with isolated perianal disease.

**115 NOVEL MUTATION WITHIN UGT1A1 IN A PATIENT WITH INDIRECT HYPERBILIRUBINEMIA.** A. Chan, W. Elfar, G.N. Bahia, N. Patel, T. Rossi, R. Abell, Pediatric Gastroenterology, University of Rochester Medical Center, North Chili, New York, UNITED STATES. Lakhani, Yale, New Haven, Connecticut, UNITED STATES.

Introduction: Uridine diphosphoglucuronate-glucuronosyltransferase 1A1 (UGT1A1) is an enzyme that is responsible for conjugation of bilirubin to glucuronic acid. A defect in UGT1A1 gene can lead to syndromes of elevated unconjugated bilirubin, such as Gilbert syndrome or Crigler-Najjar (CN) syndrome (Types I or II). CN Type I is the most severe form with complete absence of hepatic bilirubin-uridinediphosphoglucuronate glucuronosyltransferase activity. CN Type II presents with intermediate levels of hyperbilirubinemia, resulting from an incomplete deficiency of hepatic UGT1A1 activity. We describe a case of a novel variant mutation of UGT1A1 that does not meet criteria for diagnosing either CN Type I or II. Case presentation: A 2 year old Sudanese immigrant male presented from Egypt for evaluation of liver transplantation. He was diagnosed early in life with presumed CN Type I due to severity of hyperbilirubinemia and response to phenobarbital. There was a history of consanguinity and maternal hyperthyroidism for which his mother received thiouracil. He had been receiving weekly phototherapy to decrease his hyperbilirubinemia and reduce his risk of kernicterus. One year prior to his arrival to the United States, his parents stopped his phototherapy treatment. Despite lack of medical interventions, he remained asymptomatic with normal development. Upon arrival to the hospital, his total bilirubin was 27mg/dL and 26.8mg/dL of this was indirect. He was treated with intensive phototherapy, phenobarbital and cholestyramine with good response (down to 9.4). Neurologic workup was unremarkable except for abnormal otoacoustic testing on the right ear. Abdominal ultrasound demonstrated patent hepatic and portal vasculature. To confirm the diagnosis, UGT1A1 gene sequencing was done, showing that the patient has a

homozygous mutation in the UGT1A1 gene of uncertain significance. The sequence variant is designated c.824\_826delTTG, which is predicted to result in the in-frame deletion of an amino acid (p.Val275del). This mutation has not previously been reported in literature or public databases. Due to other patients with in-frame deletions in exon 1 of UGT1A1 having significant disease, there is suggestion that this patient's variant is possibly pathogenic, but clinical significance is not entirely certain. The patient is also homozygous in the UGT1A1 gene for c.-41\_-40dupTA, a dinucleotide repeat variation in the A(TA)<sub>n</sub>TAA motif in the promoter region, specifically the A(TA)<sub>7</sub>TAA allele, which has been associated with Gilbert syndrome. However, the frequency of this allele is common, and has been classified as a benign variant. Discussion: Currently, our patient just received a deceased donor liver transplant and we are awaiting further genetic testing and histopathology of the native liver. To our knowledge, this is the first reported case of this particular sequence mutation. Given that it does not fit with a classic presentation of Type I or II, this case report may expand the spectrum of Crigler-Najjar syndrome.

**116 MAURIAC SYNDROME: AN EARLY PRESENTATION.** A. Schosheim, E. Korn, R. Gill, Pediatrics, Saint Peter's University Hospital, New Brunswick, New Jersey, UNITED STATES.

Background: Mauriac Syndrome is a rare complication of uncontrolled insulin-dependent diabetes typically in adolescents and older children. It is a clinical diagnosis with the triad of hepatomegaly, hypertriglyceridemia and short stature. The earliest reported case of Mauriac Syndrome is a 3-year old, making our case the earliest reported case of Mauriac Syndrome. Case Presentation: A 20-month-old female with recently diagnosed type I diabetes presented for evaluation of abdominal distension noticed for one week. The patient's mother denied any vomiting, diarrhea, abdominal pain, jaundice, or fever. She was taking Humalog mixed 75/25 insulin with an HbA1c of 13.2 one-month prior to admission. On physical exam, the patient had short stature and hepatomegaly. Hematology/Oncology, gastroenterology and genetics were consulted for further workup. AFP, ferritin, LDH, alpha1-antitrypsin, Hepatitis B, c-peptide, karyotype and microarray were all unremarkable. Further testing revealed HbA1c of 13.4, triglycerides of 540 mg/dL and AST of 71 IU/L. Abdominal x-ray showed enlarged liver with mild stool burden. Abdominal ultrasound with Doppler showed enlarged echogenic liver (14.58cm) with normal venous flow. Discussion/Conclusion: In a retrospective review, the median age of Mauriac Syndrome was 15.1 years, with median age of diagnosis of diabetes at 10 years. Transaminases were abnormal with a median AST of 76 IU/L and GGT of 71 IU/L. Transaminases followed the trend of the HbA1c. With adequate glycemic control, the features of Mauriac Syndrome may be reversed. Reduction of hepatic enzymes after appropriate glycemic control shows further work-up and biopsy are unnecessary.

**117 A RARE PHENOTYPE OF ALPHA-1-ANTITRYPSIN DEFICIENCY DUE TO PI\*IS IN A NEWBORN WITH LIVER DISEASE.** L. Felipez, E. Hernandez, Pediatric Gastroenterology, Nicklaus Children's Hospital, Miami, Florida, UNITED STATES. C. Perez, Pediatric Gastroenterology, Baylor College of Medicine, Houston, Texas, UNITED STATES. C. Gabriel, Pathology, Nicklaus Children's Hospital, Miami, Florida, UNITED STATES. C. Finegold, Pediatric Pathology, Texas Children's Hospital, Houston, Texas, UNITED STATES. C. Huang, Pediatrics, Nicklaus Children's Hospital, Miami, Florida, UNITED STATES.

Alpha-1-antitrypsin (AAT) deficiency is a common genetic cause of pediatric liver disease. AAT deficiency is a mutation on the SERPINE1 gene. More than 100 variants of the allele have been identified. We present the first reported pediatric case of a rare variant of AAT deficiency due to phenotype PI\*IS. A 2-month-old male with an unremarkable birth history presented with jaundice and hepatomegaly. He had normal growth parameters. His family history was unremarkable. He had unremarkable complete blood count, electrolytes and coagulation, including international normalized ratio. His urinalysis and urine cytomegalovirus culture were normal. Our patient had increasing alanine aminotransferase, aspartate aminotransferase, and alkaline phosphatase. His gamma glutamyl transferase was normal. His fasting liver ultrasound showed a normal sized liver with no mass nor biliary abnormalities. His nuclear medicine hepatobiliary scan demonstrated a delayed radiotracer excretion in the gallbladder and small bowel. The hematoxylin and eosin stain showed giant cell changes and significant macrovesicular and microvesicular steatosis. An excessive abundance of stainable alpha-1-antitrypsin was observed in the hepatocytes. The Periodic acid Schiff stain showed Type 1 diastase-resistant globules in the periportal area of zone 1 hepatocytes. His quantitative alpha-1-antitrypsin level was normal: 104 mg/dL (normal range 83-199 mg/dL). Urine bile acids with Jaundice Chip Resequencing Array that included JAG1, ATP8B1, ABCB11, and ABCB4 were normal. Our patient has compound heterozygosity for a well characterized missense variant c.863A>(p.E288V), designated as PI\*S allele. An infrequent missense variant c.187C>(p.R63C),

designated PI\*I was found in the SERPINA1 gene. He did not have other mutations associated with heritable liver disease. Although both alleles I and S are individually reported as deficiency variants, we did not find a previously reported case of a pediatric patient with liver disease presenting with both of these abnormal alleles. The patient's histology of his liver biopsy was atypical, showing marked abundant stainable alpha-1-antitrypsin in zone 1 hepatocytes associated with steatohepatitis. Despite his transaminase elevation, there was minimal inflammation with mild ductular reaction. The presence of bile in Kupffer cells indicated cholestasis. Our patient was supplemented with liposoluble vitamins, an enriched medium-chain triglycerides infant formula, and ursodeoxycholic acid 10 milligrams per kilogram per dose three times daily. His liver enzymes normalized at 12 months old even after stopping the interventions. His phenotype may be associated with a better clinical outcome possibly due to the S allele producing significantly more polymerization of the abnormal protein only when it is inherited with the Z allele. We can further investigate the prognosis of this phenotype when more patients with PI\*IS are identified.

**122 SYNERGISTIC HETEROZYGOSITY OF MULTIPLE GENES IS ASSOCIATED WITH NEONATAL CHOLESTASIS.** A. Sinha, A. Thavamani, Pediatrics, MetroHealth Medical Center, Parma, Ohio, UNITED STATES. Kabbany, P. Conjeevaram Selvakumar, K. Radhakrishnan, Pediatric Gastroenterology, Cleveland Clinic Foundation, Cleveland, Ohio, UNITED STATES. Gulati, Pediatric Gastroenterology, MetroHealth Medical Center, Cleveland, Ohio, UNITED STATES. Causes of neonatal cholestasis are very diverse and amongst them are several genes that have been implicated in the etiology. In this case report, we describe a 5 months old female infant, a former 29 week preterm baby, who was transferred to Cleveland Clinic Children's with severe cholestasis. Since birth, her course was complicated by multiple infections requiring antibiotics and prolonged course of TPN. Direct hyperbilirubinemia progressively worsened on day 175 of life with total/direct bilirubin of 39.1/25, mild elevation in liver enzymes AST 319 (normal 13-35 U/L), ALT 171 (normal 7-38 U/L), GGT 126 (normal 0-35 U/L) and mild prolongation of INR 1.29 (normal 0.8 - 1.2). Patient underwent extensive work up which excluded biliary atresia, alpha-1- antitrypsin deficiency, infectious etiology, Alagille syndrome and chromosomal aberrations. A genetic cholestasis panel from Emory University was done and showed heterozygosity for variants of the following genes: AKR1D1, ATP8B1/PFIC1, ABCC2 and NPHP1. In the homozygous form, these gene variants are associated with the following diseases which manifest with neonatal cholestasis: congenital bile acid synthesis defect (type 2), progressive familial intrahepatic cholestasis type 1, Dubin-Johnson syndrome and nephronophthosis, nephrotic syndrome, respectively. Patient was started on Ursodiol, Phenobarbital and fat-soluble vitamins. Cholestasis and liver enzymes eventually normalized as patient clinical status stabilized over the next few weeks. To our knowledge, this is the first report of potential role of synergistic heterozygosity in neonatal cholestasis. Although cholestasis in our patient is likely multifactorial and is related to prematurity, prolonged TPN course, and multiple infections, the presence of variants of genes involved in neonatal cholestasis in the heterozygous form highlights the possibility of synergistic heterozygosity as a factor affecting stability of metabolic networks underlying some forms of cholestasis. With the increased use of different genetic cholestasis panels, clinicians will likely be able to further uncover the possible role of synergistic heterozygosity in the etiology of neonatal cholestasis in some patients.

**123 SPONTANEOUS BILIARY PERFORATION PRESENTING AS ABDOMINAL DISTENSION AND JAUNDICE IN AN INFANT: A CASE REPORT.** A.J. Peasley, S.L. Ciciora, Gastroenterology, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES.

Introduction: Spontaneous biliary perforation (SBP) is a rare cause of jaundice in infants, with an estimated incidence of 1.5 in 1 million live births. The presenting signs and symptoms of SBP pose a diagnostic dilemma for gastroenterologists as they can be mistaken for other causes of direct hyperbilirubinemia, such as biliary atresia. The etiology for SBP remains unknown. Early recognition and treatment of SBP yields a good prognosis. The mainstay of treatment is surgery, but controversy exists regarding the optimal surgical modality. We present a case of an infant with abdominal distension and acholic stools who was found to have bilious ascites on paracentesis and ultimately underwent a roux-en-y hepaticojejunostomy. Case Description: A 5 week old girl was admitted to the gastroenterology service for abdominal distension and acholic stools. Laboratory work up on presentation revealed direct hyperbilirubinemia (total bilirubin 5.6 mg/dL, direct bilirubin 4.3 mg/dL), elevated GGT (1159 U/L), and elevated alkaline phosphatase (1022 U/L). ALT and AST were normal (27 U/L and 44 U/L, respectively). Complete blood count was notable for thrombocytosis (platelet count 768,000). PT and PTT were within normal limits. Initial abdominal ultrasound revealed cholelithiasis, a small gallbladder likely related to recent feeding, and

large volume ascites. Spleen and liver appeared normal, without intrahepatic ductal dilation and a normal common bile duct size. Repeat abdominal ultrasound was unable to visualize the common bile duct and a small, contracted gallbladder was noted despite adequate fasting. Doppler evaluation was normal. Diagnostic paracentesis revealed a high bilirubin concentration (36 U/L) in the aspirate. Hepatobiliary nuclear imaging revealed an abnormal focal collection of the tracer at the inferior hepatic margin, corresponding to a well circumscribed lobular fluid collection on ultrasound. In the setting of bilious ascites, these findings were concerning for spontaneous biliary perforation with biloma formation versus obstruction and rupture of a choledochal cyst. Infant underwent an exploratory laparotomy and intraoperative cholangiogram, which revealed a large pseudocyst. Extensive inflammation was seen surrounding the porta hepatis at the level of the common bile duct and further exploration was abandoned. A cholecystostomy tube was placed and a drain was placed in the pseudocyst, with plans to repeat imaging in 1-3 weeks. Direct hyperbilirubinemia improved following drainage. Her course was complicated by persistent biliary fistula and recurrent biloma formation requiring further external drainage. Additionally, she developed poor feeding and poor weight gain requiring nasojunal tube feeds, sodium chloride supplementation, and attempts at bile refeeding. At 10 weeks of age, cholangiogram through the cholecystostomy tube could not identify any connection between the right and left ducts or the intra and extrahepatic ducts. At 3 months of age, she underwent a roux-en-y hepaticojejunostomy with biliary stent placement. Postoperative course was complicated by biliary stent migration and central line associated DVT requiring anti-coagulation therapy. Following surgery, she was maintained on ursodiol and a multivitamin. She developed splenomegaly at 8 months of age and portal vein thrombosis was found on doppler evaluation at 13 months of age. She had an unremarkable work up for coagulopathy. Anti-coagulation was not recommended by hematology. Around 19 months of age, she was hospitalized for an episode of coffee ground emesis during an RSV infection. She remained stable without any further hematemesis or need for intervention. She underwent esophagogastroduodenoscopy at 21 months which did not reveal any significant esophageal or gastric varices. Liver function tests, alpha fetoprotein, and abdominal ultrasound with elastography are being serially monitored. Conclusion: Although rare, SBP is an important cause of jaundice and acholic stools in infants requiring surgical intervention. Most perforations tend to spontaneously heal with decompression of the biliary tree through external drainage. However, similar to our patient, major biliary reconstruction may be needed in cases of persistent biliary fistula formation. Finally, portal vein thrombosis is a described complication following surgical management of SBP, and may follow in the immediate postoperative period.

**126 UTILITY OF DOPPLER ULTRASOUND IN THE EVALUATION OF THE CHOLESTATIC INFANT.** A.G. Goodwin, T. Miloh, S. Harpavat, Pediatric Gastroenterology, Hematology, and Nutrition, Baylor College of Medicine, Houston, Texas, UNITED STATES.

Introduction: In biliary atresia (BA), early diagnosis and treatment correlate with better outcomes. One well-established test in the evaluation of cholestatic infants for BA is the abdominal ultrasound (AUS). AUS can detect signs indicative of (triangular cord sign, abnormal gall bladder, polysplenia) and not indicative of (choledochal cyst) BA. What is less clear is whether AUS should be performed alone, or if Doppler interrogation along with AUS provides any additional benefit in distinguishing BA from other causes of neonatal cholestasis. This case series documents the use of Doppler AUS in the evaluation of neonatal cholestasis to identify congenital portosystemic shunts (CPS) as the potential cause of cholestasis. Case Presentation: We present five infants with elevated conjugated bilirubin (Bc) undergoing evaluation for BA. Age at presentation ranged from 1-12 days of life. Each subject received the same evaluation for BA, including review of newborn screening results (to detect metabolic disorders), protease inhibitor typing (to detect alpha-1-antitrypsin disease), chest radiograph (to identify butterfly vertebrate characteristic of Alagille Syndrome), and AUS with Doppler. In all five subjects, Doppler interrogation identified CPSs. All shunts were intrahepatic, with one type IV intrahepatic shunt (multiple communications between peripheral portal and hepatic veins in several segments); three type II intrahepatic shunts (a localized peripheral shunt in which one or more communications are found in a single hepatic segment); and one type V intrahepatic shunt (patent ductus venosus). Management and Outcome: No patient had elevated ammonia levels. For one patient, the Bc resolved within 3 weeks of life so further evaluation for BA was not performed. Percutaneous cholangiogram was performed in four patients and was normal, which thereby excluded BA. Three patients underwent liver biopsy at the time of cholangiogram, which showed giant cell hepatitis, portal and lobular inflammation, and canalicular bile plugs, but no duct proliferation or portal fibrosis seen in BA. Management was supportive, and CPS closure occurred in all cases between 2 weeks and 9 months of age. Bc normalized in all but

one subject, between 2 weeks and 3 months of age. Conclusion: In this report, we show that AUS with Doppler can help in the evaluation of the cholestatic infant. Our results suggest that finding CPS by Doppler evaluation could streamline diagnostic algorithms for cholestasis, by preventing further invasive testing for BA. They raise the possibility that Doppler AUS (versus AUS alone) should be a standard test in infants undergoing evaluation for neonatal cholestasis. Future studies are needed to confirm that CPS is sufficient to explain elevated Bc in cholestatic infants, as well as to assess the cost-effectiveness of the test.

**127 HYPOGLYCEMIA IN PORTOSYSTEMIC SHUNTS.** B. Kaj, C. Baker, S. Hardy, U. Shah, Pediatric Gastroenterology, Hepatology and Nutrition, Massachusetts General Hospital for Children, Boston, Massachusetts, UNITED STATES. Balza, M. Gee, Pediatric Radiology, Massachusetts General Hospital for Children, Boston, Massachusetts, UNITED STATES. Roumiantsev, Neonatology, Massachusetts General Hospital for Children, Boston, Massachusetts, UNITED STATES.

Background Hypoglycemia is a rare but significant consequence of congenital portosystemic shunts (CPSS). We report two patients with persistent hypoglycemia, and four additional cases of CPSS, highlighting the heterogeneity in clinical manifestation. Methods We performed a retrospective chart review of CPSS, identified over a 10-year period. Results Clinical features varied significantly, as shown in Table 1. Case 1 was a male infant who was critically ill at birth with 1) hypoglycemia (glucose range 21-45mg/dL) requiring correction with IV dextrose boluses, 2) thrombocytopenia and synthetic liver failure, requiring FFP/platelet transfusions and 3) respiratory distress requiring CPAP. Two intrahepatic shunts originating from the left portal vein were identified to the middle and left hepatic veins. Hypoglycemia lasted for 13 days and was managed with IV boluses of D10W and IV dextrose infusion (max. glucose infusion rate 23mg/kg/min). Comprehensive infectious workup was negative. Cardiorespiratory issues and liver failure resolved with supportive care. Case 2 is a 3yo female, diagnosed prenatally with an intrahepatic shunt and patent ductus venosus. She developed recurrent episodes of altered mental status with hypoglycemia (range 30 to 50mg/dL) or hyperammonemia after shunt embolization. Higher glucose load was required for management. Conclusion Hypoglycemia in CPSS has been thought to be driven by hyperinsulinemia and bypass of hepatic metabolism. Higher levels of insulin reach the systemic circulation, leading to hypoglycemia. Prior reports suggest that this hypoglycemia resolves with shunt closure. However, we note cases of hypoglycemia both with and without closure. Further studies are needed to expand understanding in this area.

**128 A CASE OF INDETERMINATE RECURRENT ACUTE LIVER FAILURE UNVEILED.** C. Perez, Z. Khan, B.A. Carter, Gastroenterology, Texas Children's, Houston, Texas, UNITED STATES.

Ornithine transcarbamylase deficiency (OTCD) is classically an X-linked disorder in which a mitochondrial enzyme, predominantly in the liver, is charged with the detoxification of ammonia. The phenotype is variable, but typically involves hyperammonemia and encephalopathy. Liver dysfunction is under recognized as a complication of OTCD. We describe a presentation of OTCD with recurrent acute liver failure. 10 month old full term female was transferred from OSH with acute liver failure. She presented with acute vomiting, in the absence of fever and diarrhea. Laboratory findings were significant for AST 1021, ALT 1235, Direct bilirubin 0.3, INR 8.5, and Ammonia 121. She was treated with Vitamin K, FFP and lactulose. Her past medical history was significant for abnormal newborn screen and diagnosed with heterozygous of MAT1a mutation. Her development is appropriate for age. On arrival, there was no encephalopathy and her INR corrected with Vit K and FFP. Initially, factor 5 and 7 were low, but corrected prior to discharge. Liver ultrasound was normal. A percutaneous liver biopsy demonstrated mild hepatocyte swelling with rare acidophil bodies, no fibrosis and negative viral studies, including electron microscopy. Infectious workup yielded positive enterovirus in plasma. Pi typing, ceruloplasmin, pyruvic acid, lactate, acylcarnitine profile and autoimmune hepatitis markers were normal. Plasma amino acids were notable for elevated methionine consistent with liver dysfunction. She was discharged with complete resolution of liver dysfunction off lactulose. One month later, she returned with elevated transaminases, coagulopathy, hyperammonemia in the absence of cholestasis. She again received FFP and Vitamin K. Repeat plasma amino acids was unchanged and UOA notable for uracil. Liver dysfunction again resolved and she was discharged on lactulose. Genetics team was involved and sent trio Whole Exome sequence (WES), CMA, and mitochondrial testing prior to discharge. The trio WES revealed a de novo heterozygous c.717G>8 pathogenic variant for OTC. She was started on Na benzoate, citrulline and protein restricted diet. She was evaluated for liver transplantation, but has been listed. Her development continues to be appropriate for age. Although half of pediatric acute liver failure (PALF) is typically indeterminate, accurate diagnosis has major implications on patient outcomes. Approximately 10% of

pediatric acute liver failure are secondary to metabolic disorders, including galactosemia, tyrosinemia, mitochondrial disease, and others. While OTC deficiency is not classically associated with PALF, it is an important consideration. 50% of patients with OTCD are described to have liver injury or dysfunction throughout their clinical history. Reports have suggested severe hepatic dysfunction is associated with more severe phenotype. A suggested mechanism of injury is an ammonia inhibition of hepatic protein synthesis. This case demonstrates that recurrent acute liver failure can be a presentation of OTCD and reminds pediatric gastroenterologists to consider OTCD in setting of hepatic dysfunction or indeterminate ALF. Future investigations are needed to elucidate the pathogenesis and outcomes of OTCD patients with hepatic dysfunction.

**129 DRUG-INDUCED LIVER INJURY IN AN ADOLESCENT FEMALE WITH CYSTIC FIBROSIS RECEIVING LUMACAFTOR/IVACAFTOR AND VORICONAZOLE.** C. Lopez, A. Freeman, Pediatric Gastroenterology, Hepatology, & Nutrition, Emory University, Atlanta, Georgia, UNITED STATES.

Introduction: New therapies in cystic fibrosis (CF) have an increased risk for hepatic side effects as well as significant drug interactions due to either inducing and/or acting as a substrate of cytochrome P450 isoenzymes. Vignette: A 12-year-old female with CF (F508del/F508del) and pancreatic insufficiency presented to her CF provider with 3 weeks of cough and sinus congestion. She subsequently developed worsening shortness of breath and malaise despite treatment with oral trimethoprim-sulfamethoxazole. On exam, she had decreased airflow, crackles, wheezes and a 7-pound weight loss. FEV1 was 39%, 0.94L (baseline 92%, 1.92L). She was admitted to the hospital and laboratory testing revealed an IgE level of 984 and 9% peripheral eosinophilia. Sputum culture was negative but she was clinically diagnosed with allergic bronchopulmonary aspergillosis and treated with steroids and voriconazole. Two months later, her FEV1 was 88%, and she was started on lumacaftor 200mg/ivacaftor 125mg 1 tablet twice a day, as per the recommendations when starting in combination with a CYP3A inhibitor such as voriconazole. After 1 week, the dose was increased to 2 tablets twice a day. Two days after increasing her dose, she developed abdominal pain, vomiting, and severe fatigue. On exam, scleral icterus was noted and labs revealed ALT 322, AST 503, and total bilirubin 9.4. Lumacaftor/ivacaftor was held and she was subsequently admitted for inpatient care at an outside facility. Liver ultrasound showed increased echogenicity in the periportal regions, contracted gallbladder with irregular thick walls, and a small amount of pericholecystic fluid. Labs improved slightly over the next 2 days, and she was discharged home with a presumed diagnosis of passage of a gallstone. After discharge, vomiting continued and lower extremity edema developed and she was readmitted. Voriconazole was discontinued. Labs peaked at total bilirubin 23.8, direct bilirubin 19, albumin 2.2, and INR 3.2. Extensive infectious and autoimmune testing was negative. Liver biopsy showed centrilobular necrosis with plasma cells, cholestasis, and periportal fibrosis with incomplete bridging fibrosis – suggestive of drug-induced liver injury secondary to voriconazole. Liver tests were followed closely as an outpatient and continued to improve. However, 2 months later she was noted to have pancytopenia: WBC 3.9 (ANC 310), hematocrit 25.8, and platelets 18. Bone marrow biopsy showed no blasts with a hypocellular marrow (<10% cellularity), resulting in a diagnosis of severe aplastic anemia. Further testing for infectious and inherited causes of bone marrow failure was negative, resulting in a diagnosis of presumed hepatitis-induced aplastic anemia. Treatment options, including bone-marrow transplant and immunosuppressive therapy were explored. She had a repeat liver biopsy showing stage 3-4 fibrosis with one area of evolving nodularity. Given degree of fibrosis she was deemed a poor candidate for bone marrow transplant. She was therefore started on immunosuppressive therapy with anti-thymocyte globulin, pulse steroids, and cyclosporine. She is currently 3 months post-initiation and showing good response with improving blood cell counts. Conclusion: New therapies for CF have known hepatic side effects and may pose an increased risk for drug-induced liver injury, specifically through cytochrome P450 influenced drug-drug interactions.

**131 PERCUTANEOUS TREATMENT OF HYDATID CYST IN A LIVER TRANSPLANT RECIPIENT WITH DE-NOVO ALLOIMMUNE HEPATITIS.** C. Baker, B. Kaj, S. Hardy, U. Shah, Pediatric Gastroenterology, Hepatology and Nutrition, Mass General Hospital for Children, Boston, Massachusetts, UNITED STATES. Gee, Pediatric Radiology, Mass General Hospital for Children, Boston, Massachusetts, UNITED STATES. Mueller, A. Thabet, Interventional Radiology, Mass General Hospital, Boston, Massachusetts, UNITED STATES. J. Harris, M. Pasternack, Pediatric Infectious Disease, Mass General Hospital for Children, Boston, Massachusetts, UNITED STATES. V. Deshpande, Pathology, Mass General Hospital, Boston, Massachusetts, UNITED STATES. H. Yeh, Transplant Surgery, Mass General Hospital for Children, Boston, Massachusetts, UNITED STATES. Atkins, Pediatric Residency, Mass General Hospital for Children, Boston, Massachusetts, UNITED STATES.

**BACKGROUND:** This is a twelve year old female with biliary atresia status post failed Kasai, underwent deceased donor orthotopic liver transplant at 11 months of age. Post transplant she developed alloimmune hepatitis and was maintained on tacrolimus and intermittent low dose prednisone for immunosuppression. On routine follow up, a complex hepatic cyst was identified on ultrasound, was stable in size for six months, and not associated with any symptoms or biochemical abnormalities. A subsequent acute rise in transaminases prompted biopsy of both the hepatic parenchyma and cyst. The biopsy was consistent with persistent alloimmune hepatitis and identified a chitinous layer suggestive of an echinococcal infection.**OBJECTIVE:** To effectively treat a complex hydatid cyst in a liver transplant recipient with alloimmune hepatitis.**METHODS:** There was a concern given her underlying immunosuppressive state for a disseminated infection. An abdominal and chest CT were negative for additional cysts. An MRCP showed no involvement of the hepatic vasculature or biliary system. She was started on daily albendazole, with the addition of praziquantel 48 hours pre and post a total of three PAIR (puncture, aspirate, injection, re-aspirate) procedures with 100% alcohol. Her immunosuppression was adjusted based on the level of her hepatic enzymes with progressively increasing tacrolimus when appropriate based on trough levels and decreasing prednisone.**RESULTS:** The first aspirate showed hooklets and protoscolices consistent with a viable echinococcus infection. A repeat MRI showed a slight decrease in size of the cyst. A second PAIR procedure five weeks after the initial PAIR showed a decrease in scoleces and no active organisms. A repeat scheduled MRI showed that the cyst was stable. A third PAIR procedure was performed five weeks after the second PAIR and the aspirate showed resolution of infection.**DISCUSSION:** The management of hydatid cysts have been well described in immunocompetent patients. Here we describe treatment of an echinococcal infection via multiple PAIR procedures in a patient who was a previous recipient of an orthotopic transplant liver with co-existent de-novo alloimmune hepatitis. This suggests that PAIR procedures may be a management option for a hydatid cyst in liver transplant recipients.

**134 ACID LIPASE DEFICIENCY: FROM DIAGNOSIS TO THERAPY IN CANADA.** D. Ashok, C. Prasad, S. Colaiacovo, Pediatrics, Western University, London, Ontario, CANADAD. Ashok, C. Prasad, Children's Health Research Institute , London, Ontario, CANADAD. Ashok, C. Prasad, London Health Sciences Centre, London, Ontario, CANADAJ. Walsh, T. Rugar, Pathology & Laboratory Medicine, Western University , London, Ontario, CANADA.

**Background:** Lysosomal Acid Lipase (LAL-D) deficiency is an ultra-rare lysosomal storage disorder. Clinical features in the late-onset form include dyslipidemia (elevated LDL, low HDL), elevated liver enzymes, hepatomegaly, and splenomegaly. This can progress to liver fibrosis and cirrhosis. LAL-D is caused by deficient activity of the LAL enzyme, resulting in the accumulation of cholesteryl esters and triglycerides throughout the body, predominately in the liver, spleen, gastrointestinal tract, and blood vessel walls. Supportive management with lipid-modifying agents, hematopoietic stem cell and liver transplant has been without major success.**Methods and Results:** Over the last 20 years we have had three patients with a confirmed diagnosis of LAL-D. Two adult patients, currently 31 and 40 years old, were diagnosed at 11y and 14 y with LAL activities at about 7% of control mean using fibroblasts and peripheral blood leukocytes after presenting with hepatosplenomegaly. A nine year old girl of French and Welsh background presented with hepatomegaly, elevations in liver enzymes and dyslipidemia. LAL enzyme showed low levels of 13 pmol/hour (normal 80-230) in dried blood spots. She has c.684delT and c.894G>A pathogenic mutations.**Discussion:** The third patient has received Sebelipase Alfa Kanuma<sup>®</sup> intravenous therapy every two weeks at 1 mg/kg. The therapy was started after significant advocacy from the family as the drug is not yet approved for coverage in Canada. She has tolerated the infusions well for the last six months with no side effects. The liver enzymes and lipid profile have normalized. Preliminary results of enzyme replacement therapy appear to be encouraging.

**135 ALPHA-1 ANTITRYPSIN DEFICIENCY PRESENTING AS LATE HEMORRHAGIC DISEASE OF THE NEWBORN.** D.A. Cheung, D. Goldner, L. Smith, Department of Pediatric Gastroenterology, Jackson Memorial Hospital - University of Miami, Miami, Florida, UNITED STATESP. Delgado, C.P. Rojas, Department of Pathology, Jackson Memorial Hospital - University of Miami, Miami, Florida, UNITED STATES.

Hemorrhagic disease of the newborn(HDN) is a bleeding diathesis from Vitamin K deficiency. Late HDN occurs in infants 2-12 weeks old. Cholestasis can lead to Vit K deficiency due to decreased bile excretion and malabsorption of fat soluble vitamins. Without adequate Vit K supplementation, exclusively breastfed infants with cholestasis are at high risk of vit K deficiency bleeding (VKDB)[1].We describe a 6 week old male who presented with HDN. He was born full-term at home. Antenatal and delivery period was unremarkable. He was exclusively breastfed and had

received a dose of oral vit K. He presented at 6 weeks in status epilepticus and was found to have multiple subdural hematomas requiring immediate craniotomy. His exam was significant for jaundice and hepatomegaly. His initial labs showed a significantly elevated PT of 77, INR 9.7, PTT 83.8, hemoglobin 5.5 with normal platelet count. His labs also showed direct hyperbilirubinemia with elevated transaminases. He had received fresh frozen plasma and 3 doses of intravenous Vit K after which his coagulopathy normalized and LFTs improved. His initial diagnosis was Vit K deficiency. Workup included evaluation for biliary atresia, metabolic and infectious etiologies, but workup was non-revealing. With close monthly follow up his cholestasis improved but his GGTP and transaminases did not normalize. Further work up revealed he is homozygous for AAT deficiency (PiZZ). Oral vit K is not recommended in preventing VKDB and is significantly inferior to parenteral vit K in preventing late VKDB. It is not considered adequate prophylaxis in patients with cholestasis, irrespective of underlying etiology or degree of liver injury[1]. It is prudent that all newborns receive parenteral vit K as recommended by the AAP and it is especially important in exclusively breast-fed infants with cholestasis as the risk of bleeding is substantial in this subset. AAT deficiency should be considered in the differential diagnosis of cholestatic infants presenting with intracranial hemorrhage from late VKDB. Reference [1] van Hasselt, Kok, Vorselaars et al. Vitamin K deficiency bleeding in cholestatic infants with alpha-1-antitrypsin deficiency. *Archives of Disease in Childhood - Fetal and Neonatal Edition* 94(6):F456-60

**136 INTRAPULMONARY VASCULAR DILATATIONS IN AN END STAGE LIVER DISEASE PATIENT WITH NORMAL OXYGEN SATURATIONS.** A. Panchoo, Pediatrics, Jackson Memorial Hospital, Miami, Florida, UNITED STATES. D. Rivera Rivera, Pediatrics - Gastroenterology, University of Miami, Miami, Florida, UNITED STATES. J.C. Infante, Radiology - Pediatrics, University of Miami, Miami, Florida, UNITED STATES.

**Introduction:** Hepatopulmonary syndrome (HPS) is a rare condition in children characterized by hypoxemia and intrapulmonary vascular dilatations (IPVDs) in the setting of liver disease. IPVDs are vital to the development of hypoxemia in patients with HPS. However, there is an important subset of patients with liver disease who are noted to have IPVDs without abnormalities in arterial oxygenation. The prevalence of such has been reported to be around 13-47% in the adult population with IPVD, with some literature suggesting that there are similar frequencies noted in children. Therefore, the diagnosis of IPVD in patients with liver disease may be somewhat challenging since there is this subset of patients that never develop abnormal oxygen saturations. **Case report:** We describe the case of a 19 year old female diagnosed with end stage liver disease (ESLD) and treated by the Pediatric Gastroenterology Division at our institution who presented initially to the Emergency Room with a complaint of constipation. She was subsequently admitted because she was noted to have new onset dyspnea on exertion. She described that for the past 6 weeks she has been experiencing shortness of breath on climbing stairs. On presentation her vital signs were stable and normal for age, including the oxygen saturations on room air. Physical examination was significant for left lower quadrant abdominal tenderness and splenomegaly. Abdominal X-ray done on admission was consistent with a large stool burden in the colon. Throughout her entire hospital course, the patient's oxygen saturation remained above 97%. No platypnea or changes in oxygen saturation were noted in the upright, recumbent or standing positions. Due to this new onset dyspnea on exertion, a transthoracic contrast echocardiography was performed, and it revealed the presence of IPVDs. No therapeutic interventions were recommended at that point after been evaluated by our Pediatric Cardiology team. She was discharged following successful bowel clean out. To date, she stable and no new symptoms have been present. Her dyspnea on exertion seems to be sporadic and no other symptoms or changes in oxygen saturation have been observed during her follow up visits. **Discussion:** Although a rare complication, HPS can be seen in pediatric patients with ESLD. Normally, this will include the presence of IPVDs and abnormally low oxygen saturations at room air. In this case, we saw the presence of the IPVDs, but in the setting of normal oxygen saturations. To our knowledge, little research exists regarding natural history and progression to HPS, including morbidity and mortality rates, in this subset of patients. Special consideration on the waiting list for liver transplantation, disease progression and outcomes pre and post-transplant are some of the questions that can be raised with this small group of patients. Also, it seems that relying on oxygen saturations to evaluate these patient might not be as accurate and should probably be combined with a complete history, physical examination and possibly imaging evaluation in order to diagnose these patients. By highlighting this case, we aim to draw awareness to this subgroup of patients and prompt further research in order to better characterize these patients.

**139 GASTROINTESTINAL BLEEDING IN A TEENAGER SECONDARY TO ARTERIOPORTAL FISTULA CAUSING HEMOBILIA AFTER PERCUTANEOUS LIVER BIOPSY.** E. Hilow, L. Feinberg, P. Conjeevaram Selvakumar, Pediatrics, Cleveland Clinic Foundation, Cleveland, Ohio, UNITED STATES. D. Thompson, Radiology, Cleveland Clinic Foundation, Cleveland, Ohio, UNITED STATES.

Background: Hemobilia indicates bleeding into the biliary tree which can occur as a complication of liver biopsy. Although extremely rare, mortality has been reported as high as 25%. Herein we report a case of hemobilia as a complication of liver biopsy and was treated with arterial angiography and embolization. Case: A 20-year old female with history of sickle cell and myelodysplastic syndrome who is 6 months status-post haploidentical hematopoietic stem cell transplant developed hematemesis, hematochezia, and abdominal pain 2 days after ultrasound guided percutaneous liver biopsy. Her transplant course was complicated by chronic graft-versus-host-disease (GVHD) affecting skin and eyes for which she was being treated with steroids and Sirolimus. She was hospitalized for bilateral lower limb pain of unknown origin. She also had elevated liver enzymes during this admission. So she underwent ultrasound (US) guided percutaneous liver biopsy (PLB) which showed secondary iron overload but there was no evidence of inflammation or GVHD. She tolerated the biopsy well, but had worsening abdominal pain and back pain for 2 days later. She then had 2 episodes of hematemesis with frank blood as well as hematochezia associated with tachycardia. Her hemoglobin (Hb) trended down from 11.4 to 10.6 to 9.8 g/dl. She received 1 unit of packed red blood cells with no improvement in Hb. CT scan of abdomen was obtained which was significant for wall thickening of duodenum. She was started on Octreotide to reduce splanchnic blood flow. She had an esophagogastroduodenoscopy (EGD) for further evaluation. EGD was remarkable for patchy erythema in the 2nd portion of the duodenum with no clear ulcers or bleeding vessels. After prolonged inspection blood was noted to be oozing from the ampulla indicating hemobilia. She had an arteriography which showed left hepatic artery to left portal vein fistula which was successfully embolized with a coil. Hematochezia and hematemesis resolved after embolization. Discussion: Hemobilia is the least common hemorrhagic complication after percutaneous liver biopsy with an incidence of 0.059%. It is typically caused by needle injury to portal structures resulting in arteriovenous or arterial bile duct or venous bile duct fistula. Non-traumatic causes in children include ascaris lumbricoides, liver abscess, choledochal cyst, and Von Willebrand Disease. Clinical presentations include abdominal pain, upper gastrointestinal (GI) bleeding, jaundice and pancreatitis. Hemobilia should also be suspected in setting if a post-liver biopsy drop in Hb associated with these symptoms. GI bleeding can start from few days to weeks after biopsy. Diagnosis of hemobilia can be initially made with Doppler ultrasound, contrast-enhanced computed tomography and magnetic resonance imaging. Hepatic angiography is considered the most accurate in diagnosing various fistula formations following liver biopsy. In addition, hepatic angiography is an effective therapeutic modality. Angiographic embolization of active hemobilia is the preferred method of treatment in hemodynamically unstable patients with a success rate of > 95%. This case highlights the importance of being aware of hemobilia as a complication of PLB even with US guidance.

**145 ADENOMYOMATOSIS OF THE GALLBLADDER IN A CHILD: A RARE GALBLADDER FINDING YEARS BEFORE TYPICAL PRESENTATION.** G. Koon, M. Schmalz, Pediatrics and Adolescent Medicine, Western Michigan University Homer Stryker MD School of Medicine, Jackson, Michigan, UNITED STATES. R. Cameron, Pediatric Gastroenterology, Bronson Methodist Children's Hospital, Kalamazoo, Michigan, UNITED STATES.

Adenomyomatosis of the gallbladder (AMG) is an uncommon finding in childhood, with only 5 previously reported cases in the literature. We present the case of an eight-year-old male found to have AMG in addition to his past medical history of ulcerative colitis, autoimmune hepatitis, hereditary spherocytosis as well as cholangitis, which we suspect to be developing primary sclerosing cholangitis. Due to the multitude of diagnoses affecting his biliary tree, each with the ability to induce a pathologic change, routine imaging studies have been performed. It is through this nearly seven-year history of imaging we have seen an evolution of his gallbladder from normal physiology to that of AMG. Upon recognition of AMG, the best treatment option was to pursue cholecystectomy. These lesions have the propensity for malignant change, albeit low, the ability to mask other potentially malignant lesions and it has a known association with the development of cholelithiasis. AMG is most certainly not on the differential of most pediatric gastroenterologists, not only because of its rare occurrence, but also because the mean age of onset is in the fifth decade of life, well beyond our age threshold. Through both our gross and microscopic depictions we describe the characteristic findings of AMG and discuss how we may keep this diagnosis fresh in our minds as we are seeing children with complex biliary disease and multiple problems. Maintaining a

high index of suspicion, even amongst the rarest of diagnoses is critical for providing an accurate diagnosis and treatment recommendations.

**149 LATE-ONSET MPV17-RELATED HEPATOCEREBRAL MITOCHONDRIAL DNA DEPLETION SYNDROME.** D. Taylor, A.A. Shah, H.C. Lin, Pediatrics, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES. Introduction: MPV17-related hepatocerebral mitochondrial DNA depletion syndrome (MDS) is an autosomal recessive disease characterized by profound reductions in mitochondrial DNA (mtDNA) content, which usually leads to liver failure and death before age two. Case: A 16-year-old girl presented with persistent muscle weakness, paresthesia, and primary amenorrhea. Previous neurologic work-up included a nerve conductive study and electromyography, which showed sensory and motor neuropathy. She was noted to have elevated AST and ALT levels of 61 U/L and 65 U/L respectively, and had a liver ultrasound showing hyperechoic liver nodules. Liver biopsy showed macrovesicular steatosis, portal fibrosis and mixed portal inflammatory infiltrate, and an overabundance of mitochondria in hepatocytes on electron microscopy. These histologic features are typical in mitochondriopathies and a genome analysis was performed, which showed the presence of two heterozygous novel unclassified variants in the MPV17 gene (c.191C>T (p.P64L) and c.375+5G>T) and a novel unclassified variant in the x-linked PHKA2 gene. Muscle biopsy showed normal histopathologic findings. The genetic findings support a variant of MPV17 with mild disease phenotype, with isolated liver and little to no skeletal muscle involvement. She was started on a mitochondrial cocktail consisting of Ubiquinol, Riboflavin, B50 complex, vitamin E, alpha-lipoic acid, and leucovorin with symptom improvement. Conclusion: Variants of MPV17 that result in a less severe phenotypes are rare. This disorder could potentially be more common than was previously thought, if patients have been misdiagnosed with other disorders due to the complexity and difficulty in getting to this diagnosis.

**150 BILIARY ATRESIA IN CHILDREN WITH CHROMOSOME 22Q11 ABERRATIONS.** H. Wang, T. Miloh, S. Harpavat, Pediatric Gastroenterology, Baylor College of Medicine, Houston, Texas, UNITED STATES. H. Wang, T. Miloh, S. Harpavat, Gastroenterology, Hepatology and Nutrition, Texas Children's Hospital, Houston, Texas, UNITED STATES. Introduction: Biliary atresia (BA) is a rare life-threatening disease that occurs during infancy. Approximately 10–20% of infants with BA are associated with other congenital anomalies, indicating possible genetic composition of the pathogenesis of BA. Here we reported 3 cases of biliary atresia with chromosomal multiplication representing 3 distinct genotypes of 22q11 aberrations. Case presentation: Case 1, a 4 week old male, presented with conjugated hyperbilirubinemia, found having Emanuel syndrome (defined as t(11;22) (q23;q11.2) translocation) and triplication of 22q11.1 to 11.21. Case 2, a 4 year old female, had diagnosed BA with intraoperative cholangiogram and received Kasai procedure at 2 weeks of life, found having Cat-eye syndrome (defined as trisomy or tetrasomy of pericentromeric region of chromosome 22) with quadruplication of 22q11.1 and triplication of 22q11.1 to 11.21. Case 3, a 9 year old female, diagnosed congenital duodenal atresia and then BA, received Roux-en-Y reconstruction at 2 days of age and Kasai procedure at 8 weeks of age, found having triplication of 22q11.22 inherited from her mom. Management and Outcome: Case 1 developed sepsis and liver failure and died at 2<sup>nd</sup> month of life, autopsy confirmed his BA and overlapping neonatal hemochromatosis. Case 2 received liver transplant at 4 year age due to end stage liver disease, developed significant bile leak and renal impairment, currently doing well with normal hepatic synthetic functions at 6 year age. Case 3 developed significant food protein allergy which had resolved, is currently transplant-free at 10 years of age, but on liver transplant waiting list due to progressive liver fibrosis without hyperbilirubinemia. Discussion: From these BA cases with distinct chromosomal anomalies of 22q11 following a spectrum of clinical phenotypes of early mortality, liver transplant or prolonged transplant free survival, we consider whether genetic over-dosing of 22q11 might contribute to BA's pathogenesis. We discussed possible roles in biliary pathology of genes located at this region including interleukin 17 receptor A (IL17RA), clathrin heavy chain like 1 (CLTCL1), Solute Carrier Family 25 members SLC25 A18/A1, peroxisomal biogenesis factor 26 (PEX26), claudin 5 (CLDN5), and Cell Division Cycle 45 (CDC45).

**154 LATE ONSET EBV-ASSOCIATED POST TRANSPLANT LYMPHOPROLIFERATIVE DISORDER.** R. Josyabhatla, I. Monteiro, Pediatrics, Rutgers New Jersey Medical School, Newark, New Jersey, UNITED STATES. Sharma, Rutgers Cancer Institute of New Jersey, New Brunswick, New Jersey, UNITED STATES. Introduction: Post transplant lymphoproliferative disorder (PTLD) is one of the most common post transplant malignancies seen with both solid organ transplantation (SOT) and allogeneic hematopoietic stem cell

transplantation. Most cases occur within the first year post-transplant. We present a case of PTLD occurring more than 10 years post transplant. Case report: 15 year-old female, who underwent orthotopic liver transplant for tyrosinemia at age 4, presented with acute-onset fever and worsening fatigue. She was maintained on tacrolimus for immunosuppression. On presentation, she was noted to have pancytopenia - WBC 3300 (lymphocyte predominant), Hemoglobin 9.8, Platelets 81,000. Liver enzymes were normal. ESR, LDH and uric acid were elevated. Abdominal ultrasound revealed enlarged mesenteric and retroperitoneal lymph nodes - indicating infectious vs neoplastic process. CT scan confirmed these findings and also revealed mild ascites and splenomegaly with low density lesions. Viral work up showed elevated EBV IgG. Quantitative EBV PCR was positive. Biopsy of abdominal lymph nodes confirmed diagnosis of PTLD with positive EBV staining. Tacrolimus was discontinued and she was treated with combination of prednisone, rituximab and cyclophosphamide. Routine PET scan a year later showed an incidental finding of a sacral lesion. CT-guided biopsy revealed poorly differentiated malignant neoplasm. Serial PET scans showed the lesion was stable. A year later she developed back pain. Repeat biopsy of the lesion was negative for infection and malignancy. 6 months later she developed progressive leg pain, MRI showed progressive disease involving the sacrum and an epidural lesion in the lumbosacral region. Biopsy revealed Hodgkin type lymphoma. She received 4-cycles of chemotherapy and radiation. A PET scan after 2-cycles showed complete resolution of the lesion. She is now doing well for the last 2 years. Discussion: PTLD is a well known post transplant complication commonly occurring within one year after transplant, and is most often due to EBV infection of B-cells. Higher immunosuppression, increases the risk of developing PTLD, with an earlier onset. PTLD has variable presentation and often mimics benign processes. It is important to screen post transplant patients for complications. Qualitative EBV PCR is not a reliable marker for diagnosis or follow up and hence it is important to monitor high risk patients with quantitative EBV PCR. Serial monitoring of EBV PCR allows early detection of EBV infection, which can then be managed by decreasing immunosuppression and treatment with intravenous ganciclovir. Clinical suspicion of PTLD should immediately prompt histopathological confirmation. Homeostatic chemokine sCXCL13, plays a crucial role in B-cell homing and lymphoid organ development, and is found to be higher in PTLD as compared to SOT recipients with EBV reactivation and may be a readily available surrogate marker for diagnosis and monitoring of response to cytoreductive treatment. Conclusion: To prevent morbidity and mortality from EBV-associated PTLD, a high index of suspicion should be maintained, irrespective of time since transplant as seen in our patient who presented more than 10 years post transplant.

**159 NEONATAL CHOLESTASIS IN AN INFANT WITH DOWN SYNDROME.** M. Kabbany, P. Conjeevaram Selvakumar, K. Radhakrishnan, Department of Pediatric Gastroenterology and Hepatology, Cleveland Clinic Foundation, Cleveland, Ohio, UNITED STATES. Conley, Pediatric Institute, Cleveland Clinic Foundation, Cleveland, Ohio, UNITED STATES.

Patients with Down Syndrome have higher prevalence of neonatal cholestasis. In this report we present a 2 month old male with Down syndrome and perimembranous VSD who presented to Cleveland Clinic Children's Hospital for a second opinion for cholestasis and elevated liver enzymes. His initial labs showed total bilirubin of 18.7 mg/dL (Normal 0-1.5 mg/dL), direct bilirubin 13.8 mg/dL (Normal 0-0.4 mg/dL), AST 681 U/L (Normal 0-60 U/L), ALT 548 U/L (Normal 5-50 U/L) and GGT 78 U/L (Normal 0-50 U/L). Patient was a full term baby with uncomplicated prenatal course. He was born vaginally and stayed in the neonatal ICU for respiratory distress for 16 days. He did not need intubation or TPN. On day of life 5, direct hyperbilirubinemia was noted. Abdominal US showed normal common bile duct and gallbladder. He was discharged home on Ursodiol and ADEK. At the outside hospital, further evaluation revealed normal thyroid function, A1 antitrypsin and negative CMV and newborn screen. Bilirubin and liver enzymes continued to rise except GGT by day 25. Total/direct bilirubin 14/10, AST 144, ALT 96, GGT 90, and INR 1.16. Patient underwent a liver biopsy on DOL 26 which showed paucity of bile ducts and marked diffuse giant cell transformation. Liver enzymes progressively increased on day 61 of life with AST/ALT (>750/686) and total bilirubin 18.7. At Cleveland Clinic Children's, patient underwent intraoperative cholangiogram and liver biopsy on day of life 69 which didn't show biliary atresia. Biopsy showed normal structure with severe cholestasis and extensive giant cell transformation of hepatocytes. Stains were negative for viral inclusions. Genetic cholestasis panel done Emory University showed heterozygosity for two gene variants SLC27A5 and TRMU. Those gene variants are associated with Bile acid amidation defect and transient infantile liver failure, respectively. Patient was continued on Ursodiol and Aquadeks and later his cholestasis slowly improved and resolved by 7 months of age. Few case series have reported that the incidence of neonatal cholestasis among Down syndrome infants is 0.11-3.9% reflecting 10-100 fold increase compared to the general population. The etiology of cholestasis in this patient

population is still not fully understood. Few potential mechanisms include paucity of bile ducts, immature transport function in the biliary canaliculi and smaller bile acid pool size. Besides that, there might be a role of synergistic heterozygosity in our patient. Overall the prognosis of neonatal cholestasis is benign especially in Down syndrome patients without bone marrow involvement. In conclusion, pediatric hepatologists should be aware of the increased incidence of neonatal cholestasis among Down Syndrome infants.

**161 A RARE CAUSE OF CHEST PAIN IN A HEALTHY TEENAGER: HSV ESOPHAGITIS.** B. Alabd Alrazzak, M. Shah, B. Mouser, A. Marshburn, A.Z. Solaiman, Pediatrics , UT Health Science Center Houston, Houston, Texas, UNITED STATES.

**Case presentation** A 17-year-old male with no past medical history presented to the emergency department with 2 days of fevers and severe, sharp chest pain. Extensive workup including complete blood count (CBC), complete metabolic profile (CMP), D-dimer, cardiac enzymes, chest x-ray, CT angiography, and electrocardiogram was normal except white blood cell (WBC) count was  $12.9 \times 10^3/\mu\text{L}$ . Upon further review, the patient's pain was exacerbated by eating and drinking. Exam was only significant for sinus tachycardia and mild pain on epigastric palpation. Upper GI series was normal. He was started on IV pantoprazole, PO sucralfate, and IV hydromorphone which provided minimal relief. Patient ultimately underwent esophagogastroduodenoscopy (EGD) with biopsies, which showed severe pan-esophagitis (Figure 1). Biopsy results were positive for herpes simplex virus (HSV) on staining. Patient's girlfriend reported a history of frequent cold sores. HIV testing was negative. Patient received IV acyclovir which provided immediate relief, and he was discharged home to complete 7 days of oral valacyclovir. **Discussion** HSV as a cause of esophagitis has been extensively described in immunocompromised hosts in whom this condition can be fatal (1, 2). While rare in immunocompetent patients, there are now a handful of case reports of severe HSV esophagitis in normal healthy patients (1-9). A review published in 2000 by Ramanathan *et al* described 38 healthy patients, both adult and pediatric, who were diagnosed with the condition. There was a 3:1 male predominance overall, which increased to 90% in the pediatric population. The typical patient was a young (less than 40 years old in 78% and less than 18 years old in 24% of cases), healthy male presenting with acute odynophagia, dysphagia, or heartburn with or without prodromal symptoms (fever, pharyngitis, respiratory symptoms) or oral lesions (1). As was the case with our patient, patients with esophagitis frequently present with retrosternal "chest pain" (1, 7, 8). Esophagitis should be considered for all patients presenting with the triad of chest pain, odynophagia, and fever, as early recognition can prevent broad cardiopulmonary workups (9). **Conclusion** HSV esophagitis should be kept in the differential diagnosis for patients presenting with acute onset of fever, heartburn, retrosternal "chest pain", odynophagia or dysphagia, though it does not uniformly present with classic oral lesions like gingivostomatitis or coupled to a known exposure to a HSV-positive contact. Prior to initiation of broad and potentially unnecessary cardiac workups in the emergency department, healthy adolescents presenting with "chest pain" should be screened for symptoms of esophagitis. While common in the immunocompromised patient, HSV esophagitis is now increasingly being described in immunocompetent patients as well. Prompt diagnosis and treatment with antivirals such as acyclovir or valacyclovir typically results in complete resolution in otherwise healthy patients with HSV esophagitis.

**164 ROTAVIRUS AS A CAUSE OF ATYPICAL HEMOLYTIC UREMIC SYNDROME IN A PATIENT WITH SHORT BOWEL SYNDROME.** A. Jazayeri, S. Kinberg, Pediatric Gastroenterology, Columbia University, New York, New York, UNITED STATES. Z. Henen, Pediatrics , St Joseph University, New York, New York, UNITED STATES.

**Abstract** Hemolytic Uremic Syndrome (HUS) is a thrombotic microangiopathy characterized by the triad of hemolytic anemia, thrombocytopenia, and acute renal failure. It is typically seen following a diarrheal illness. In 90% of HUS cases, Shiga toxin is secreted from *Escherichia coli* (*E. coli*) O157-H7 or *Shigella Dysenteriae* type 1, causing endothelial damage to the glomerulus and precipitating a robust activation of the complement cascade. This represents approximately 90% of all HUS diagnosis. However, there is a small percentage of patients that develop HUS in the absence of Shiga toxin. These atypical presentations of HUS are thought to be related to congenital deficiencies to regulatory proteins of the complement cascade that become activated by other illnesses. We present a case of a patient who developed HUS after a rotavirus infection that then progressed to HUS. **Case report:** A 2 year old two-year-old female with a history of short bowel syndrome, chronic lung disease and , developmental delays, and short bowel syndrome presented to our emergency department (ED) with a 2 day history of profuse watery non-bloody diarrhea. Upon presentation to the ER ED, she was afebrile and had mild tachycardia. On physical exam, the patient had dry mucous membranes and a delayed

capillary refill. Her initial laboratory tests were significant for a work showed a CBC with an elevated white blood cell count WBCs of  $23,700.7 \times 10^3/\mu\text{L}$ , hemoglobin of 11.5 g/dL, and platelets count of  $792,000 \times 10^3/\mu\text{L}$ . Her initial sodium was 122 mmol/L [137-145 mmol/L], Her Blood Urea Nitrogen was 23mg/dL [3-14 mg/dL], and Creatinine was 1.2mg/dL [0.4-0.7 mg/dL]. Stool infectious studies were sent and an abdominal x-ray was ordered that showed evidence of pneumatosis intestinalis. The patient was made NPO, started on antibiotics, and admitted to the pediatric intensive care unit (PICU). In the PICU, the patient continued to have profuse watery non-bloody diarrhea. A respiratory viral panel came back positive for rhinovirus/enterovirus, a stool polymerase chain reaction (PCR) came back positive for rotavirus, and a blood culture was positive for streptococcus mitis. Stool PCR was negative for E. coli and Shigella. Her creatinine had increased from baseline 0.2mg/dL to 1.2mg/dL. Over the following week, she developed progressive oliguria with increasing elevated creatinine (max 3.67 mg/dL). Her Hgb dropped from 11 to 5.1 g/dL and her platelet counts dropped from 31,000 to  $792 \times 10^3/\mu\text{L}$ . She received multiple packed red blood cell transfusions. The patient was diagnosed with atypical HUS caused by rotavirus. Nephrology was consulted and hemodialysis was started. The patient was placed on hemodialysis. Hematology was consulted and a peripheral smear was performed that showed schistocytes, a reticulocyte count was elevated to 9.4%, a lactate dehydrogenase (LDH) an LDH was elevated to 2293 U/L, and a coombs test was negative. Testing for ADAMTS-13 Activity, complement C3 and complement C4 were within normal limits. Urine was negative for streptococcus pneumoniae antigen. A diagnosis of atypical HUS caused by rotavirus was diagnosed. After three days of hemodialysis, the patient's urine output improved and dialysis was discontinued. The patient's hemoglobin, and platelets and creatinine began to recover and the pneumatosis intestinalis resolved. The patient was restarted on feeds and was transferred to the floor for ongoing care. Discussion: There is growing literature about atypical HUS and its possible causes. The prevailing theory is that it stems from a complement dysregulation triggered either through medications, infections, or an autoimmune process that may be predisposed through genetic mutations of Factor H, Factor B, Factor I, or CD46. This mutation leads to decreased proteolytic inactivation of C3b thus leading to amplification of the alternative pathway. Ultimately, this activation of the complement pathway leads to damage of red blood cells, platelets, and endothelial cells. HUS due to rotavirus is a rare complication with only one case reported in the literature to date. This instance is the second reported case of isolated rotavirus-induced HUS. Rotavirus infection and is something that should be considered when looking for atypical triggers of HUS. Furthermore, clinicians should be aware of HUS as a possible complication of rotavirus infection and should monitor hemoglobin, platelet count and renal function in these patients.

**166** *CAMPYLOBACTER JEJUNI INDUCED INTUSSUSCEPTION RECURRENCE WITHIN A 30 HOUR PERIOD.* A.O. Walker, T.M. Attard, Pediatric Gastroenterology, Children's Mercy Hospital, Kansas City, Missouri, UNITED STATES. Introduction Intussusception of the small bowel represents the most common etiology for intestinal obstruction in early childhood. It represents telescoping of the small bowel potentially resulting in ischemia and perforation, and it is recognized as potentially recurrent in up to 10% of cases. The presence of an identifiable mass lesion that provides a lead point is understood as a risk factor for recurrence. Signs and symptoms of intussusception include acute intermittent abdominal pain, and bloody stools. Campylobacter jejuni is a gram-negative, pathogenic bacterium that causes gastroenteritis and has been known to cause intussusception. Some studies have recognized the risk of intussusception recurrence with relation to different microbial pathogens, although there have been no reported cases of Campylobacter causing recurrent intussusception within a 30 hour period. Case We report a previously healthy 2 year old male who presented to the emergency department with a 7 day history of diarrhea, fever, a three day history of bloody stools and spasmodic abdominal pain that worsened acutely on the day of presentation. At that time, the parent also noted one self-limited episode of rectal prolapse. The patient was seen on three occasions in an emergency department, and at the first visit, he was sent home with a stool kit for stool study investigation, as he was unable to produce a bowel movement. On final return, an abdominal ultrasound reported short segment intussusception in the right lower quadrant. The patient subsequently developed recurrent ileocolic intussusception (3 recurrences total within 30 hours). All three episodes were successfully reduced via barostatic enema. For suspected lead point, he underwent exploratory laparoscopy but none was identified. After exploratory laparoscopy, his parent developed bloody stools. This prompted stool collection from our patient for investigation of infectious etiologies. Stool studies were positive for Campylobacter jejuni and he was subsequently treated with Azithromycin. Discussion Small bowel obstruction from intussusception can complicate several luminal processes that result in bowel wall thickening or mass acting as a lead point. While

infectious etiologies are a recognized risk factor for intussusception, this is the first report of recurrent intussusception associated with *Campylobacter jejuni* enteritis. In this case, stool collection for studies was unsuccessful until his parent presented with similar symptoms. Despite there being no recommendations currently for stool studies in the evaluation of intussusception in society guidelines, the authors would speculate that stool studies for enteric pathogens should be routinely obtained in children presenting with intussusception and treated wherever identified, potentially impacting the risk of recurrence. Current treatment recommendations for bacterial enteritis secondary to *Campylobacter jejuni* include oral rehydration, as it is self-limited in the majority of cases. For complicated cases, treatment includes use of Macrolide antibiotics.

**169** *FECAL MICROBIOTA TRANSPLANTATION IN THE SOLID ORGAN TRANSPLANT COMMUNITY: A GROWING EXPERIENCE.* D.M. Barnes, Pediatrics, Naval Medical Center San Diego, San Diego, California, UNITED STATES. Park, Pediatrics, Stanford University, Palo Alto, California, UNITED STATES.

Introduction:Fecal Microbiota Transplantation (FMT) is a treatment used to repopulate the gut microbial population in an effort to treat a disease caused by dysbiosis or a derangement of the microbial make-up. Recent randomized clinical trials have well supported the role of this therapy in recurrent *Clostridium difficile* infection (CDI) in the immune competent host. Though the experience with FMT for CDI in the literature has grown vastly in the past 5 years, the experience with performing FMT on the immunosuppressed patient has been just recently started being defined. Case Descriptions:Case 1: A 16 year old girl status post orthotopic liver transplantation on immunosuppressive monotherapy with Tacrolimus presented with multiply recurrent CDI. Case 2: A 3 year old male child status post orthotopic heart transplantation on dual immunosuppression with Tacrolimus and Mycophenylate presented with multiply recurrent CDI. Case 3: A 17 year old boy status post orthotopic renal transplantation on triple immunosuppression with Tacrolimus and Mycophenylate as well as low dose Prednisone (6mg, 0.1mg/kg/day) presented with multiply recurrent CDI. Patients 1 and 2 received FMT preparations colonoscopically and patient 3 received the preparation via preexisting gastrostomy tube with inoculum from a meticulously screened donor via our collaboration with OpenBiome stool bank. Patients' diarrhea resolved within 4-6 days post-transplant and all patients remained asymptomatic at 10 weeks post-transplant with no recurrences of CDI. Patient 1 developed transient constipation during the first week post-transplant, which resolved spontaneously. None of the patients experienced any other adverse effect. Discussion:This series helps to grow the collective experience of FMT in the immunosuppressed patient and, in particular, in those status post solid organ transplantation. These cases begin to characterize the safety profile in this setting and are critical given the roadblocks to accomplishing randomized clinical trials in such a group due to the small numbers and heterogeneity.

**175** *CLASSIFICATION OF FEEDING DISORDERS IN PATIENTS REFERRED TO AN INTERDISCIPLINARY NUTRITION AND FEEDING TEAM.* A. Kaistha, A. Goldhaber, T. Tan, Pediatrics / Division of GI,Nutrition., NYU Langone Medical Center, NY, New York, UNITED STATES.

Abstract Type: Clinical Vignette- PosterIntroduction Previous work has sought to outline classification of feeding difficulties based on symptomology, etiology, and mode of treatment. It has been surmised that more complex feeding problems require referral to and management by a multidisciplinary team (McComish, et al., 2016). This poster seeks to confirm this as we outline the etiology of pediatric feeding problems referred to an interdisciplinary nutrition and feeding team at a large academic medical center in the New York metro area, demonstrating appropriateness for joint management and follow up by a pediatric gastroenterologist (GI), registered dietitian (RD), and speech language pathologist (SLP).MethodsData over a period of 6 months was reviewed to determine if referrals to the interdisciplinary team program were appropriate and warranted for simultaneous management by all team members. Patient problems were divided into the following etiological categories: medical (e.g., anatomical, physiologic, organic causes), developmental (e.g., oral motor, oral sensory), behavioral or caregiver mismanagement. Patients were then categorized into those requiring follow up with the team and those requiring follow up with one discipline alone (e.g., RD or SLP only). We surmise that by classifying the type of feeding disorder, level of complexity is established and appropriate means for management and follow-up are determined.ResultsReview of 44 patients referred to the program revealed that 77% of patients met criteria for all 3 categories; medical, developmental and behavioral/caregiver mismanagement. This demonstrates that majority of feeding disorders are multifactorial. Furthermore, 77% of the "all categories" patients were determined to require regular follow up with the team. This differs from the patients only meeting criteria for 1

category, more often determined to need follow up with 1 discipline only. Additionally, a majority of patients who presented with an underlying primary medical diagnosis which was not of gastrointestinal etiology (e.g., cardiac, craniofacial) demonstrated "secondary" GI diagnoses such as recurrent vomiting, Gastroesophageal reflux disease (GERD), or constipation. Conclusion This review demonstrates the multifactorial and complex nature of feeding disorders, often with overlapping elements of medical, developmental and behavioral concerns. Additionally, categorization of feeding difficulties can help clinicians determine referral criteria to interdisciplinary programs and management pathways as etiology of feeding problems in the individual patient are better defined. References: Kerzner, B., Milan, K., MacLean, W., Berall, G., Stuart, S., Chatoor, I. (2015). A practical approach to classifying and managing feeding difficulties. *Pediatrics*, 135, 344. DOI: 10.1542/peds.2014-1630. McComish, C., Brackett, K., Kelly, M., Hall, C., Wallace, S., Powell, V. (Jul/Aug 2016). Interdisciplinary feeding team: a medical, motor, behavioral approach to complex feeding problems. *MCN, The American Journal of Maternal/Child Nursing*, 41(4), 230-236. DOI:10.1097/NMC.000000000000252. Rybak, A. (2015). Organic and nonorganic feeding disorders. *Annals of Nutrition & Metabolism*, 66(5), 16-22. DOI: 10.1159/000381373.

**176 CHOLECALCIFEROL VERSUS ERGOCALCIFEROL SUPPLEMENTATION FOR TREATING VITAMIN D DEFICIENCY IN CHILDREN WITH INFLAMMATORY BOWEL DISEASE.** A. Lundin, Clinical Nutrition, UCSF Benioff Children's Hospital Oakland, Oakland, California, UNITED STATES.

Background Stoss therapy, or high doses of vitamin D, has found to be a safe and effective for treating vitamin D deficiency in pediatric patients with diagnosed inflammatory bowel disease (IBD).<sup>1</sup> Providing a very high dose of cholecalciferol (vitamin D3) remarkably elevates the patient's vitamin 25-hydroxy D level suggesting sufficient stores that would last for several months. However, at our institution, due to ordering errors, varying insurance coverage, and pharmacy substitutions, some patients received ergocalciferol (vitamin D2) for stoss therapy versus cholecalciferol. Both ergocalciferol and cholecalciferol are commonly found in vitamin D supplements, however cholecalciferol has a greater affinity for vitamin D-binding protein and a longer half-life in the body.<sup>2</sup> In this clinical vignette, we retrospectively examined if using ergocalciferol supplementation for stoss therapy provided comparable results when compared to when cholecalciferol supplementation was utilized. Methods Eight patients with IBD and deficient vitamin 25-hydroxy D levels (<20 ng/mL) were given stoss therapy according to established guidelines (Table 1).<sup>1</sup> A retrospective chart review of the electronic medical record was conducted to see if cholecalciferol or ergocalciferol was prescribed. All children had vitamin 25-hydroxy D levels in the deficient range (<20 ng/mL), 3 had Crohn's disease (CD), 2 had ulcerative colitis (UC), and 3 had indeterminate colitis (IC). The patients' ages ranged from 12 to 20 years with an average age of 15.6 years. Pharmacies were called to verify the prescription was filled and inquire if a substitution was made. Six patients received cholecalciferol and 2 received ergocalciferol. Within one month of taking the prescribed dose, vitamin 25-hydroxy D levels were measured again to determine response to the therapy. A t-test was performed with laboratory findings and a <math>p</math>-value was calculated to determine statistical significance between the two groups. Results Our preliminary results found that patients who received cholecalciferol supplementation had a significant (<math>p</math> <0.001) positive response to stoss therapy when compared to patients who received ergocalciferol supplementation. Conclusions In our small population of patients, our data suggests that stoss therapy using cholecalciferol supplementation is more effective at replenishing vitamin D levels in children with IBD compared to when ergocalciferol supplementation was used. Based on our results, we will encourage practitioners prescribing stoss therapy in this patient population to prescribe cholecalciferol and instruct pharmacies to not substitute with ergocalciferol. 1. Shepherd D. *JPGN* Oct 2015. Pazirandeh S. Up to Date. 2016

**182 MANGANESE TOXICITY IN TODDLER DEPENDENT ON PARENTERAL NUTRITION.** A. Fifi, Pediatric Gastroenterology, University of Miami, Miami, Florida, UNITED STATES.

Manganese (Mn) is a trace element used as a cofactor for many of important enzymes. However clinical implications of inadequate Mn intake are not well described, of greater clinical significance is manganese toxicity, which has been reported in instances of parenteral use of trace elements in nutritional formulations. Case A 17-month old male underwent a multivisceral transplant for intestinal failure secondary to gastroschisis and parenteral associated liver disease was dependent on parenteral nutrition (PN) since birth. Shortly after transplant he presented with constant abnormal tics of unknown etiology, associated with tremors and irritability. He had issues with feeding intolerance and rejection that resulted in continued administration of PN post-transplant. Neurology was consulted and he underwent an EEG to rule out seizures, which was negative. His medications were

reviewed and concern was raised over use of adult trace elements daily and possibility of manganese (Mn) toxicity. Levels had been sent in one month prior but manganese was not resulted so it was resent. An MRI was performed which was significant for a "Hyperintense T1 signal within the globus pallidi bilaterally, which may be related to the patient's history of PN or patient's known liver disease (manganese deposition)." Simultaneously Manganese level was found to be 11.6. Trace elements were immediately discontinued in his PN and Mn level fell to normal (Mn=1.0) three weeks later. Follow up MRI two months later showed resolution of hyperintense signaling in globus and over next several months his symptoms resolved and he made a complete neurological recovery.

Discussion Mn is absorbed in the small intestines and while the absorption process is slow, the total absorption rate is high - about 40%. Iron deficiency is well known to increase absorption of Mn from gut. Daily requirements for Mn increase with age, from as little as 0.5mg at birth to 5mg in adults. Also, Mn has been shown to cross the blood-brain barrier, with the rate of penetration in animal experiments being 4 times higher in neonates than in adults. The primary targets of manganese toxicity are the brain and central nervous system. Mn has been shown to be deposited in certain regions of the brain, with T1 weighted images on MRI scan, showing typically bilateral symmetrically increased signal intensity in the globus pallidus and subthalamic nuclei that can be reversible after Mn supplementation is discontinued. However prolonged exposure to high concentrations can be associated with permanent damage, with symptoms of impaired neurological and neuromuscular control, mental and emotional disturbances, muscle stiffness, lack of coordination, tremors, difficulties with breathing or swallowing, and other neuromuscular problems. Infants and children on PN, either solely or as an adjunct to enteral feeds are especially at risk for multiple reasons. Mn administered parenterally by passes gut thus daily requirements are lower in these patients. In addition often times children on PN have some degree of liver impairment or cholestasis and this can further add to risk of development of Mn toxicity as Mn excretion in bile is impaired. The chronically ill child on long term PN is usually iron deficient and thus putting them at even further risk of toxicity because iron deficiency has been shown to increase Mn accumulation in brain. Finally, in children also taking some diet by mouth, iron deficient meals typical in infants and children, cause increased absorption of Mn in gut because Mn and Fe compete for same receptor for their uptake. Mn is found in considerably higher concentrations in formulas than human milk. Since Mn toxicity is a real danger to patients receiving PN and its deficiency is still not well described some authors argue against supplementing with Mn. However current available preparations of trace elements for administration in PN contain Mn so the prescriber of PN would have to order each element individually which is more tedious and sometimes difficult due to recurrent shortages in these elements individually. It is thus vitally important to follow levels of trace elements intermittently. It is our practice to check them every 3 months if changes are made to PN or every 6 months if stable.

**185** *A LIFELONG JOURNEY OF TWO BROTHERS WITH GENETIC SUCRASE-ISOMALTASE DEFICIENCY (GSID) AND EOSINOPHILIC ESOPHAGITIS (EOE)*. A. Boney, QOL Medical, LLC, Vero Beach, Florida, UNITED STATES. McCarthy, Kindrdfood, Boston, Massachusetts, UNITED STATES.

INTRODUCTION/BACKGROUND:GSID and EOE are recognized gastrointestinal (GI) disorders, but their co-morbidity is not fully understood. GSID is a deficiency of sucrase-isomaltase (SI) at the brush border of the small intestine and results in diarrhea, gas, abdominal pain, and distension. EOE is a chronic allergic inflammatory disease of the esophagus with a buildup of eosinophils (white blood cells) in the esophagus.OBJECTIVE:Describe presenting history and steps to diagnosesReview treatment plansProvide insight for healthcare providers who work with complex GI co-morbiditiesMETHODS: The medical and diet records of two brothers' history were reviewed by the authors. Patients were contacted to report any gaps.HISTORY: Sibling 1 CB●First 5 months of life - explosive diarrhea, failure to thrive, and poor muscle tone–Failed formulas: Nutramigen™ (corn syrup solids), Alimentum™ (sugar, modified tapioca starch), and Elecare™(corn syrup solids)●Diagnosis ruled out–Cystic Fibrosis–Parasites●GSID diagnosed by esophagogastroduodenoscopy (EGD) with biopsy for disaccharidase assay at 8 months●Formula switched to 3232A™ with fructose●Sucraid<sup>®</sup> (sacrosidase) Oral Solution prescribed at 3 years (were previously unaware it was available)●Modified starch intake●Repeat EGD at 21 years of age confirmed diagnosis of EOE–egg and dairy eliminated; follow-up EGDs identified egg as EOE trigger●Currently at 26 years–Avoids most starch–Sucraid<sup>®</sup> with sucrose containing foodsSibling 2 GB●Treated for GSID from birth based on brother's GSID diagnosis (confirmed by biopsy)–Formula - 3232A™ with fructose–Sucraid<sup>®</sup> started when solids initiated–Low sucrose fruits, and some starch (mostly bread)●At 14 presented with fatigue, chest pain, and tingling in throat when eating peanuts–EKG and x-ray normal–6 months later EGD confirmed EOE–Dairy, egg, wheat, soy, beef, corn, shellfish, and nuts eliminated–

Despite diet and PPI, EOE persisted • Elemental diet via Naso-gastric (NG) tube initiated—Elecare™ - developed significant diarrhea (corn syrup solids)—3232A™ with fructose was tolerated—Diet- low sucrose, low starch fruits and vegetables (limited amounts)—Discontinued NG-tube, took formula by mouth—Received a “clean scope” (no eosinophils)—Began systematically adding foods back into diet—Steroids initiated prior to starting college • Currently at 21 years:—Drinks 3232A—Modifies starch—Takes Sucraid<sup>®</sup> with sucrose containing foods—Remains on steroids with plans to wean—Recent EGD remained clear of eosinophils

**DISCUSSION:** It took 6 - 8 months to reach the first GSID and EOE diagnoses, both of which require elimination diets. The top eight foods eliminated for EOE are dairy, egg, wheat, soy, beef, corn, shellfish, and nuts. GSID eliminates sucrose and starch. With a dual diagnosis of GSID and EOE, 10 food groups may need to be eliminated initially. Two decades later, at 21 and 26 years old both men are clear of EOE and manage their GSID diet with starch modification and Sucraid<sup>®</sup> therapy.

**CONCLUSIONS:** • Reaching a GSID and EOE diagnosis can take time • Elimination diets require an experienced dietitian to ensure nutrient requirements are met • Foods must be gradually reintroduced to avoid over-restriction and to promote a healthy diet that incorporates the usual pleasures associated with eating

**186 AN UNUSUAL CASE OF NUTRITIONAL HYPOCALCEMIC SEIZURES IN AN ADOLESCENT WITH AUTISM SPECTRUM DISORDER.** A.S. Kota, L. Palezac, R. Bargman, K. Kumar, Pediatrics, Nassau University Medical Center, East Meadow, New York, UNITED STATES.

**Introduction:** The key features of Autism spectrum disorders (ASD) include repetitive and stereotypical behaviors along with sensory issues. These behaviors contribute to restricted diets resistant to change. We report a case of an autistic adolescent who presented with hypocalcemic seizures due to extremely low calcium levels solely attributed to his diet.

**Case Description:** A 14 year old male with ASD presented to the emergency department for increased aggression and developed generalized tonic-clonic seizures while being evaluated. After he was stabilized, workup was done for a new onset seizure. It included complete blood count with differential, electrolytes (including calcium and magnesium), hepatic panel, prolactin, TSH and neuroimaging. Most of the workup was negative except for undetectable calcium levels (<5 mg/dl; Normal (8.4-10.2 mg/dl), low vitamin D (5.6 ng/ml; Normal: 30-100ng/ml) and elevated alkaline phosphatase levels (430 U/L; Normal: 117-390U/L). Review of the diet revealed that he was a picky eater and for four months preceding this admission, his diet exclusively consisted of chips and juice. He was taken care of by his elderly grandparents because of parental neglect as a child. Due to these social issues, his psychological well-being was paid more attention to than nutrition. Physical examination was significant for a non-verbal, non-cooperative male with weight and height at 10<sup>th</sup> and 5<sup>th</sup> percentile respectively. He had bad dentition with multiple cavities. Further investigations for identifying the cause of hypocalcemia were done. Serum magnesium and albumin levels were normal. Parathyroid hormone levels were increased (638pg/ml Normal 15-65pg/ml) along with elevated phosphorus levels (4.9 mg/dl Normal 2.7-4.5 mg/dl). Wrist radiographs did not show any signs of rickets. Treatment of his hypocalcemia was particularly difficult due to his aggressive behavior. He was given intravenous infusions of calcium gluconate along with vitamin D supplementation. PO intake of calcium was encouraged. Diet was slowly transitioned to calcium fortified snacks, juices, pediasure and more varied diet eventually. Prompt response to calcium supplementation over the course of 6 days as evidenced by normalized calcium and parathyroid hormone levels indicated a nutritional etiology of his hypocalcemia especially given his history of restricted diet. He remained asymptomatic for the rest of his hospital stay. He was followed up after 3 months. He was on a varied diet and his calcium, vitamin D and PTH levels were within normal limits.

**Conclusion:** Several studies have shown that children with ASD consume less calcium and vitamin D compared to their typically developing peers. However there can be extreme cases like our patient with a very restricted diet. It is essential to consider frequent and thorough nutritional screening in children with ASD as they have several risk factors which can exacerbate these deficiencies. Caretakers should be advised to diversify the diet or correct the deficiencies either through supplements or fortified foods.

**187 AN UNUSUAL CASE OF SHORT BOWEL SYNDROME COMPLICATED BY FOOD PROTEIN INDUCED ENTEROCOLITIS AND ANASTOMOTIC ULCERS.** A. Kastl, C. Bales, L. Albenberg, J. Kelsen, Gastroenterology, Hepatology, and Nutrition, Children's Hospital of Philadelphia, Chicago, Illinois, UNITED STATES.

**Background:** Anastomotic ulcers are a poorly recognized complication of intestinal resection in children with short bowel syndrome. They are a rare, unexpected, and potentially life-threatening entity that may occur years after surgery. Patients typically present with blood loss, manifesting on a spectrum of indolent iron deficiency anemia

with hemocult-positive stools, to frank gastrointestinal hemorrhage. Abdominal pain, diarrhea, poor appetite, and growth retardation may also be present. Although various therapeutic approaches have been proposed, including antibiotics, bile-acid sequestrants, 5-ASA derivatives, biologics, and surgical resection, none of these have been consistently successful in healing anastomotic ulcers. As such, therapy is currently empiric with no clear management guidelines. Case description: We report a unique case of a 5 year old girl with short bowel syndrome associated with perforated necrotizing enterocolitis (NEC) and a history of severe food protein induced enterocolitis syndrome (FPIES), who presented with bowel inflammation and bleeding anastomotic ulcers several years after bowel resection, and shortly after dietary liberalization. She was born prematurely at 27 weeks EGA, and developed NEC on DOL 28. She underwent ileocolonic resection with ileostomy creation and subsequent ostomy closure, which left her with 43 cm of small bowel anastomosed to her right colon at 3 months of age. Her postoperative course was complicated by watery diarrhea, which was marginally improved by metronidazole and cholestyramine, and significantly (>50% reduction) improved with a transition from breast milk to elemental formula. She was ultimately discharged on a combination of elemental feeds and parenteral nutrition. By 9 months of age, she had achieved enteral independence on an exclusively elemental diet. As solid foods were introduced, beginning with rice cereal, she developed episodes of profound watery diarrhea and vomiting with severe metabolic acidosis within 1-2 hours of food ingestion. An upper endoscopy revealed only mild villous blunting without increased cellularity in the duodenum. Skin prick testing to rice was negative, and patch testing was positive. A diagnosis of FPIES was confirmed with an inpatient rice food challenge. Over the following year, 9 additional foods were found to precipitate these symptoms, and were excluded from her diet. She thrived on a highly restricted diet until 4 years of age, when the diet was liberalized, starting with rice. These liberalizations were initially well tolerated, but she presented 2 months later with asymptomatic iron deficiency anemia. Within two weeks, she developed worsening anemia, associated with diarrhea, fevers, elevated inflammatory markers, and hemocult-positive stools. Fecal calprotectin was normal, and stool studies for infectious organisms were negative. Endoscopy revealed an ulcerated anastomotic site, 5 cm in length, most significantly on the colonic face. The perianastomotic biopsies demonstrated acute inflammation, with fibrinopurulent exudate and pyloric gland metaplasia. The colon mucosa was unremarkable. Upper GI study did not suggest proximal small bowel ulceration. Her diet was not altered during that admission. Her symptoms, anemia, and elevated inflammatory markers resolved after 2 months of combination therapy with mesalamine, Flagyl, and prednisolone. This improvement was sustained on mesalamine alone for 18 months, at which point she represented with abdominal pain, anorexia, melena, and anemia. A diet history revealed progressive liberalization, specifically with rice products. Repeat endoscopy again demonstrated ulceration at the ileocolonic anastomosis without change in visual or histologic character. Her presentation was refractory to several weeks of steroid reinitiation. She was ultimately started on sirolimus and a more restrictive diet with elimination of rice. Symptoms have improved and anemia is resolving on this regimen for the past month. Discussion: A diagnosis of anastomotic ulceration should be considered for any child with delayed gastrointestinal bleeding after intestinal surgery. It is a clinical conundrum that can be refractory to both medical and surgical management. This case raises the possibility that food antigen exposures may be driving an inflammatory response in select children with surgical anastomosis, and that dietary manipulations may play a role in treating anastomotic ulcers.

**189 FILLING THE FEEDING GAP: INTERVENTIONS IN A NUTRITION NICU GRADUATE CLINIC.** C. Jump, Pediatric Gastroenterology, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. Antonio, X.T. Zhang, L. Sams, Pediatrics, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. N. Taylor, Neonatology, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. Patients in the Neonatal Intensive Care Unit (NICU) are at high risk of growth failure, particularly in the early period following hospital discharge, with long-term consequences on their growth and development. We describe a cohort of patients discharged from our institution's Level III nursery with outpatient follow-up in a designated "Nutrition NICU Graduate Clinic" with a pediatric gastroenterologist and dietitian, to identify nutritional deficits and early signs of growth failure with the goal of early intervention to improve outcomes in short-term growth and prevent hospital admissions for growth failure. Over a period of six months, a total of 85 patients were seen for an initial visit, typically between 3-4 weeks after discharge. Patients were referred to clinic based upon birth weight, discharge feeding regimen, and/or concern by practitioner for growth as outpatient. Data from this visit was retrospectively reviewed for demographics at birth (Table 1), feeding regimens at discharge, and need for nutritional intervention. At initial follow up, 29 (34%) had adequate weight gain indicated by growth velocity  $\geq 30$

grams/day (g/d), 33 (39%) had moderate growth failure with 20-29 g/d, 18 (21%) had severe growth failure with <20 g/d, 5 (6%) had weight loss since hospital discharge. In total, 62 of 85 patients (73%) received some nutritional intervention. Of those with a recommendation to change nutritional regimen, the intervention was aimed to increase caloric intake in 34 (55%) and to decrease caloric intake in 18 (29%). After initial clinic visit, 20 patients (24%) were discharged from clinic for follow up with their pediatrician due to adequate growth with no physician or family concerns. Of note, 11 of 85 (13%) of families received education due to inappropriate feeding preparation, e.g. formula mixing. Six of the 40 (47%) infants receiving maternal breastmilk at the time of discharge were no longer receiving breastmilk leaving 40% of the total population receiving breastmilk at their initial follow up. Infants seen in our Nutrition NICU Graduate Clinic represent a high risk cohort with a diverse racial makeup (49% of patients African American, 46% Caucasian, and 5% Hispanic) and a high percentage (59%) with Medicaid as their primary insurance. Our preliminary results suggest an opportunity for quality improvement prior to discharge with emphasis on improvement in delivery of formula mixing instructions and working to sustain breastfeeding and adequate growth post-discharge. Prospective data collection will be collected to continue to characterize the impact of these interventions as well as identify potential risk factors for outpatient growth failure.

**193 COPPER DEFICIENCY-INDUCED BICYTOPENIA CAUSED BY POOR COMPLIANCE IN A PEDIATRIC PATIENT WITH CHRONIC MALNUTRITION.** D. Yi, M. Kim, Pediatrics, Chung-Ang University Hospital, Seoul, KOREA (THE REPUBLIC OF).

Background: In children with intestinal failure (IF), supportive management for preventing chronic malnutrition that results in hematological complications is highly important. Careful treatment including that for psychogenic problems and medical management is required in adolescent patients. Case presentation: A 12-year-old boy who underwent gastric wedge resection was transferred to our hospital because of chronic loss of appetite, vomiting, growth failure, and weight loss. As he was able to ingest orally, we tried to restore his general condition and increase his weight by maintaining additional nutritional supply through peripheral parenteral nutrition (PN). However, continuous vomiting, weight loss, and superior mesenteric artery syndrome persisted because of low treatment compliance. The findings of hyponatremia and bicytopenia did not improve. Bone marrow biopsy was performed, and it revealed copper deficiency. PN was increased via the central route, and additional micronutrient agents, including copper, were administered. In particular, invasive diagnosis and treatment, and adequate education improved the treatment compliance of the child. Since then, he avoided intake of large amounts of beverages and thus no longer had repeated vomiting and weight loss. His copper deficiency and bicytopenia improved, and his weight and dietary intake also increased. Conclusions: We confirmed that treatment compliance is important in pediatric patients with malnutrition. In chronic malnutrition, attention should also be paid to deficiency of micronutrients such as copper, which can lead to hematologic problems. In patients with pediatric IF, a multidisciplinary approach such as medical, surgical, and psychological management will prevent further complications and provide better therapeutic results.

**194 NEUTROPENIA AS A RESULT OF COPPER DEFICIENCY IN A PEDIATRIC INTESTINAL FAILURE PATIENT ON FULL ENTERAL NUTRITION.** D.A. Stamm, M. McGivney, C. Duggan, Gastroenterology, Hepatology and Nutrition, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES. D.A. Stamm, M. McGivney, C. Duggan, Center for Advanced Intestinal Rehabilitation (CAIR), Boston Children's Hospital, Boston, Massachusetts, UNITED STATES. D.A. Stamm, M. McGivney, Surgery, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES.

Introduction: Copper deficiency has various physiologic sequelae including anemia, neutropenia and osseous abnormalities. Risk factors for development of copper deficiency include altered intestinal anatomy, competitive uptake with zinc and iron, and nutrient malabsorption due to chronic diarrhea. Previous studies have documented copper deficiency in pediatric intestinal failure patients dependent on parenteral nutrition (PN), however no research exists on the frequency or outcomes of copper deficiency in this population following PN cessation. This case report describes a fully enterally fed pediatric intestinal failure patient who had unexplained neutropenia for 5 months, which resolved following diagnosis and treatment of copper deficiency. Case Presentation: Our patient was a 2 year old girl with intestinal failure due to multiple intestinal atresias (type IIIb jejunal and type I ileal). She had a complex surgical history including resection of atresias and jejunoileal anastomosis on day of life 1, and serial transverse enteroplasty (STEP) procedure at age 8 weeks. She was PN dependent until age 11 months at which time she was transitioned to a full enteral diet, on a combination of oral and GTube feeds. Serum copper concentrations were assessed 4 and 6 months after PN cessation and were within normal limits. Subsequently, she

developed persistent neutropenia. One year after PN cessation her absolute neutrophil count (ANC) was critically low at 0.22 K cell/ $\mu$ L. She was found to be profoundly copper deficient, with a serum copper concentration of 27 mcg/dL (range 85-150). She was initiated on an enteral copper supplement at a dose of 5 mg daily and serum copper, ceruloplasmin and ANC concentrations normalized. Table 1 details ANC levels over time before and after copper supplementation. Conclusion: Copper levels are routinely measured in pediatric intestinal failure patients on PN, however may not be assessed after progression to full enteral nutrition. Furthermore, untreated copper deficiency can lead to severe neutropenia, as occurred in this case. Serum copper concentrations should be routinely monitored in pediatric intestinal failure patients who are fully enterally fed, and copper deficiency should be considered as a possible etiology for neutropenia. Provision of an enteral copper supplement may be effective in correcting this deficiency.

**196 ESSENTIAL FATTY ACID DEFICIENCY WITH SMOF LIPID MINIMIZATION IN A NEONATE: A CASE REPORT.** N.

Memon, A. Herdt, I. Griffin, MidAtlantic Neonatology Associates, Morristown, New Jersey, UNITED STATES. Hussein, T. Hegyi, Pediatrics, Rutgers Robert Wood Johnson Medical School, New Brunswick, New Jersey, UNITED STATES.

Background: Long-term use of parenteral nutrition (PN) in neonates is associated with the development of parenteral nutrition-associated liver disease (PNALD). The lipid emulsion component of PN, which has traditionally been derived from soybean oil (SO) is a known contributing factor to the pathogenesis of PNALD. Strategies to minimize lipid exposure, through dose reduction or use of alternative lipid emulsions have been shown to improve biochemical markers of PNALD. Restricting the lipid dose, however, carries the potential risk of essential fatty acid deficiency (EFAD), predisposing neonates to compromised central nervous system development and growth failure. Newer intravenous lipid emulsions, such as SMOF (Fresenius Kabi), containing 30% soybean oil plus 25% olive oil, 15% fish oil, and 30% medium-chain triglycerides have recently been FDA approved for adults in the United States. There is limited data regarding the safety and development of EFAD with SMOF lipid minimization, especially in neonates. Objective: To describe a case of moderate essential fatty acid deficiency with SMOF lipid minimization in a late preterm infant. Case description: The patient is a male born at 35 1/7 weeks' gestation with a birth weight of 2300 g via NSVD to a 19-year-old G3P0020 mother with prenatal history significant for gestational diabetes diet controlled and abdominal wall defect in the fetus. Apgars were 2 and 7, and infant required endotracheal intubation at 2 minutes of life due to apnea during resuscitation. Of note, scaphoid abdomen and bowel loops covered with a thin membrane through 1.5 cm diameter abdominal was noted on exam. Infant was taken to the OR on day of life (DOL) 0 where an enterectomy, resection of necrotic bowel due to in utero volvulus, repair of bowel perforation, appendectomy, and reduction of omphalocele with abdominal wall closure was performed. He was made NPO (nil per os) and started on parenteral nutrition. Traditional 100% SO-based emulsion, Intralipid 20% (Fresenius Kabi) was increased per protocol to 3 g/kg/d by DOL 3. On DOL 12, infant was noted to have direct hyperbilirubinemia (1.8 mg/dL), and lipid minimization with SO at 1 g/kg/d was instituted (Figure). Infant continued to develop worsening direct hyperbilirubinemia and on DOL 29, SMOF was started at a reduced dose of 1 g/kg/d. Prior to starting SMOF, EFA profile were obtained and infant had normal, albeit borderline high triene to tetraene (T/T) ratio of 0.075. (Criterion for essential fatty acid deficiency (EFAD), according to the Mayo Medical Laboratories is  $> 0.08$  if 44 4/7 weeks and less and  $> 0.05$  if older). Direct hyperbilirubinemia continued to worsen while the infant received SMOF and EFA profile obtained on DOL 70 revealed moderate essential fatty acid deficiency with a T/T ratio of 0.306 (moderate EFAD is defined as T/T ratio  $> 0.2$  and severe EFAD as  $> 0.4$ ). SMOF was increased to 3 g/kg/d and EFAD was corrected within 2 weeks. Of note, infant did not have worsening of his direct hyperbilirubinemia while receiving higher doses of SMOF. Once the T/T ratio was corrected, SMOF was decreased to 2 g/kg/d, on which the infant currently remains and he continues to have a normal T/T ratio. Conclusion: We have described a case of significant EFAD and worsening direct hyperbilirubinemia with SMOF lipid minimization in a late preterm infant with parenteral nutrition-associated liver disease. SMOF and newer generation lipid emulsions have significantly less essential fatty acids ( $\alpha$ -linolenic acid and linoleic acid) than traditional soybean oil-based lipids. Given that recent studies have also shown EFAD with SO lipid minimization, extra caution and frequent monitoring of T/T ratio should be used during lipid minimization with newer generation lipids.

**200 A COMPLICATED CASE OF A SINGLE EPISODE OF NECROTIZING PANCREATITIS AND PSEUDOCYST LEADING TO EXOCRINE PANCREATIC INSUFFICIENCY.** A. Glinky, P. Arias, N. Santucci, Pediatrics, Louisiana State University Health

Sciences Center, New Orleans, Louisiana, UNITED STATES. Arias, N. Santucci, Gastroenterology, Hepatology and Nutrition, Children's Hospital, New Orleans, Louisiana, UNITED STATES.

**Introduction:** We present a case of necrotizing pancreatitis in a morbidly obese 15 year old male. His course was complicated by a persistent pseudocyst requiring surgical drainage leading to exocrine pancreatic insufficiency. **Case:** KD is a 15 year old morbidly obese male who presented with three days of bilious vomiting and abdominal pain. On exam, he had abdominal striae and diffuse abdominal tenderness to palpation. His initial white blood cell count was 33,000, BUN 57 mg/dL, creatinine 1.3 mg/dL, total bilirubin 2.6 mg/dL, lipase 6737 U/L and amylase 1042 U/L. Abdominal CT scan showed necrotizing pancreatitis, fatty liver, and bilateral pleural effusions. **Management:** He received supportive treatment with intravenous fluids and analgesics. Early in his stay, he developed tachypnea, concerning for ARDS or pulmonary embolism. He required 24 hour intensive care monitoring and antibiotics, metronidazole and zosyn for infected pancreatitis. The remainder of his two week recovery was unremarkable. He was discharged on a fat free diet. In the next month, he developed a large pseudocyst measuring 18 cm x 22 cm, palpable on abdominal examination. The pseudocyst persisted on serial ultrasounds for 5 months, but he remained otherwise asymptomatic. He required a cyst gastrostomy and in situ drainage by the advanced endoscopy team at an outside hospital. He also required 5 settings of ERCP for manual drainage to ensure complete removal. An endoscopic ultrasound showed multiple small gallstones for which he also required a cholecystectomy. An MRCP post pseudocyst removal showed a large defect in the pancreatic body and hypoplastic tail. He has subsequently developed exocrine pancreatic insufficiency (fecal elastase = 41ug/g – normal: >201 µg/g) and fat soluble vitamin deficiencies (vitamin D = 7 ng/mL, vitamin E = 5 mg/L) and is currently receiving oral pancreatic enzyme replacement therapy. **Discussion:** The complex course of this patient's first episode of pancreatitis is a cause for concern. Pancreatitis is one of the most common causes of GI hospital admission in the US. Pseudocysts and acute fluid collections occur in 10% of cases, and mortality is 17% in patients with necrosis. Exocrine pancreatic insufficiency (EPI) is common in acute recurrent and chronic pancreatitis. Although it has been reported, it is unusual to develop EPI after a single episode of acute pancreatitis.

**207 SAMD9-ASSOCIATED ENTEROPATHY IN MIRAGE SYNDROME.** B. Chen, V. Avinashi, Pediatric Gastroenterology, Hepatology and Nutrition, British Columbia Children's Hospital, Vancouver, British Columbia, CANADA. J. Terry, Pathology and Laboratory Medicine, British Columbia Children's Hospital, Vancouver, British Columbia, CANADA. **Introduction:** Recently in 2016, MIRAGE (myelodysplasia, infection, restriction of growth, adrenal hypoplasia, genital phenotypes, and enteropathy) syndrome was described involving a group of individuals with SAMD9 mutations<sup>>1</sup>. SAMD9 protein is described to have a role in cellular growth. Little is known of the nature and outcome of enteropathy in these individuals. **Objective:** To describe two cases of children presenting in infancy with chronic diarrhea related to SAMD9 genetic mutations. **Case descriptions:** Two ex-premature female infants presented with persistent diarrhea with stool output exceeding 100ml/kg/day. Diarrhea had a secretory and osmotic component, as fasting decreased stool output. Clinical features are outlined in the table. Both had negative work-up for infectious colitis, pancreatic insufficiency, cystic fibrosis, protein losing enteropathy and immunodeficiency. Genetic testing revealed pathogenic mutations in the SAMD9 gene on chromosome 7 for both patients. They had multiple medical comorbidities, some consistent with MIRAGE syndrome. They also had features of dysautonomia, which was not part of the initial description of MIRAGE syndrome. One patient underwent endoscopy with histology that demonstrated features resembling an autoimmune enteropathy. She responded favorably to immunosuppressive therapy and eventually gained independence from parenteral nutrition, receiving full blenderized diet via G-tube. For the second patient, due to poor prognosis, a palliative approach was taken. Although no endoscopic studies were performed, finding of SAMD9 mutation increases likelihood of enteropathy associated with MIRAGE syndrome. This patient remains dependent on parenteral nutrition but a possible empiric trial of prednisone to improve enteral autonomy is being explored. **Description of endoscopic and histologic findings for patient 1:** Her initial upper endoscopy and sigmoidoscopy at 6 months of age was macroscopically and microscopically normal. She had repeat endoscopy at 13 months due to worsening diarrhea. Findings included linear gastric erythema and erosions in the duodenal bulb. Colonoscopy showed multiple scattered erosions from transverse colon to rectum. Duodenal, antral and gastric body biopsies showed focal apoptosis. There were nonspecific reactive changes in the duodenum with normal villi and chronic active gastritis. Interestingly, distal colonic biopsies (sigmoid and rectum) showed new finding of increased epithelial apoptotic bodies of up to 5 per crypt base, which may be seen in autoimmune enteritis. Testing for anti-enterocyte antibodies were negative and electron microscopy of the duodenal mucosa was non-diagnostic. Histopathological</sup>

features of tufting enteropathy, microvillus inclusion disease and enteroendocrine cell dysgenesis were not seen. There were normal activity levels of sucrase, maltase, palatinase and lactase. She was treated initially with steroids with improvement in diarrhea as well as her comorbid thrombocytopenia and respiratory stability. Azathioprine was subsequently added. Her improvement initially seemed to predominantly stem from effects of prednisone as increased stool output was observed when steroids were weaned despite adequate azathioprine metabolite levels. Follow-up endoscopy was performed 5 weeks later. Upper endoscopy showed few non-specific antral pinpoint hemorrhages and sigmoidoscopy was normal macroscopically. Histology revealed reduction in number and distribution of apoptotic figures and reduction of plasma cells in the duodenum. Conclusion: These are two reports of patients with SAMD9-associated enteropathy along with some features of MIRAGE syndrome. SAMD9 mutation is a novel cause of non-short gut related intestinal failure. One patient's enteropathy had favorable outcome to immunosuppression. Further information is needed to elucidate prognosis and outcomes for SAMD9-associated enteropathy. 1. Narumi S et al. Nat Genet 2016.

**208 USE OF AN IN-LINE DIGESTIVE ENZYME CARTRIDGE IN PEDIATRIC CYSTIC FIBROSIS PATIENTS.** C. Jump, Pediatric Gastroenterology, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. Hendrix, S. Michel, Nutrition, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. Flume, Pulmonology, Medical University of South Carolina, Charleston, South Carolina, UNITED STATES. Introduction: Optimal nutrition and growth in cystic fibrosis (CF) is associated with improved clinical outcomes. The CF Foundation recommends enteral nutrition (EN) to improve age-dependent anthropometrics in patients that are not able meet nutrition goals with dietary intake alone<sup>1</sup>. Twelve percent of people with CF and exocrine pancreatic insufficiency (EPI) receive supplemental EN given overnight via gastrostomy tube<sup>2</sup>. There are no recommendations for the use of pancreatic enzyme replacement therapy (PERT) with EN due to the absence of clinical trials. A new in-line digestive enzyme cartridge is FDA approved for use in adults on EN. The lipase containing cartridge has been shown to be safe and well tolerated in adults and children age 5 and older<sup>3</sup>; however, there are no long-term studies demonstrating effectiveness, safety, and tolerability in children less than 5 years of age. Methods: This was a single-center retrospective review of pediatric patients using the cartridge in place of traditional PERT with overnight EN. None of the patients used oral PERT in conjunction with this device during overnight EN, although they did continue oral PERT during oral feeding. Patient baseline demographics, change in growth parameters, gastrointestinal (GI) symptoms, and tolerability were recorded. Results: We have treated 13 pediatric patients (8 males) with a mean age of 8.3 ( $\pm$ 5.2) years with the cartridge. Four patients were started on the cartridge when EN was initiated, while 9 had previously been on EN for a mean of 3.4 years. Indication for initiation of the cartridge in those already on EN included poor growth on traditional PERT (n=6) and/or reported poor tolerance of EN due to GI symptoms (n=7). All caregivers reported tolerance of EN with the cartridge. Resolution of GI symptoms was reported in 6 (46%) patients, with the most common report being improvement in morning fullness and bloating (n=3). One child stopped using the cartridge after 3 months related to issues with the cartridge disconnecting during overnight EN. Growth parameters improved overall (Table). At 3 months 83% of patients (n=10) had improvement in BMI or weight-for-length percentiles, and at 6 months 71% (n=5) had improvement in BMI percentiles compared to baseline. Conclusions: Based on our observation, use of this in-line digestive enzyme cartridge provides a safe and practical way to deliver absorbable fats from overnight EN. Use of this device was well tolerated, and promoted weight gain and reduction in GI symptoms of malabsorption in children age 3 months to 16 years, even in the absence of oral PERT with EN. References: 1. Schwarzenberg S, et al. J Cyst Fibros. 2016 Nov;15(6):724-735. 2. CFFPR: 2015 Annual Data Report. Bethesda: CCF; 2016. 3. Orenstein D, et al. Poster presented at NACFC, Oct 27-29, Orlando, FL

**212 LYMPHANGIECTASIA AS A CAUSE OF REFRACTORY ASCITES.** C. Baker, B. Kaj, S. Hardy, U. Shah, Pediatric Gastroenterology, Hepatology and Nutrition, Mass General Hospital for Children, Boston, Massachusetts, UNITED STATES. Diamond, Plastic Surgery, Shriners' Hospital for Children, Boston, Massachusetts, UNITED STATES. Gee, Pediatric Radiology, Mass General Hospital for Children, Boston, Massachusetts, UNITED STATES. BACKGROUND: Primary intestinal lymphangiectasia (PIL) is a pathologic dilation of lymphatic vessels of unknown etiology. Clinical symptoms include diarrhea, peripheral edema and ascites. Here, we described three unique patients with different management options for the treatment of ascites. OBJECTIVE: To describe variation in the treatment of primary intestinal lymphangiectasia. METHODS: A retrospective review of PIL patients treated at the Massachusetts General Hospital for Children over a 10 year period. RESULTS: All three patients were found to have

hypoalbuminemia and refractory ascites. Whilst albumin diuresis was used in all patients, novel treatments were also applied. Patient 1 has tuberous sclerosis with refractory ascites and severe protein losing enteropathy. Intestinal lymphangiectasia was diagnosed on endoscopy and imaging confirmed that there was no secondary lymphatic obstruction with angiomyolipomas. Sirolimus was used for its effect as an inhibitor of vascular proliferation. Response was followed clinically and via measurements of VEGF C and D (vascular endothelial growth factor). Patient 2 presented with severe refractory ascites. He was subsequently found to have a lymphatic leak at the thoracic duct and had surgical closure. Patient 3 presented with left hemi-hypertrophy and refractory ascites and on MR lymphangiogram was found to have absent cisterna chyli and thoracic duct. Her management included albumin diuresis and liposuction for treatment of limb edema by plastic surgery. With further surgical options under review. **DISCUSSION:** Primary intestinal lymphangiectasia is associated with hypoalbuminemia and clinical symptoms of refractory ascites. We describe our experience with three patients with variation in management options.

**216 SUPERIOR DIAGNOSTIC YIELD OF GADOXETATE DISODIUM CONTRAST AGENT IN MAGNETIC RESONANCE CHOLANGIOPANCREATOGRAPHY (MRCP) FOR PANCREATICOBILIARY MALJUNCTION IN A PEDIATRIC PATIENT.** D. Sierra, A.C. Huang, Pediatrics, Nicklaus Children's Hospital, Miami, Florida, UNITED STATES. J. Reeves-Garcia, E. Hernandez, R. Gomara, R. Arboleda, Gastroenterology, Hepatology and Nutrition, Nicklaus Children's Hospital, Miami, Florida, UNITED STATES.

Background Acute pancreatitis has an incidence of about 1 in 10,000 children per year. Systemic disease, drugs, trauma and anatomic abnormalities are common causes in pediatrics. Recurrent pancreatitis is defined as 2 episodes meeting the INSPPIRE criteria without irreversible structural damages to the pancreas. The workup recommended by the INSPPIRE group includes routine laboratory testing, magnetic resonance cholangiopancreatography (MRCP) and genetic testing for CFTR, SPINK1 and PRSS1 mutations, among others. Choledochal cysts have been associated to the development of pancreatitis. In turn, pancreaticobiliary maljunction (PBM) has been proposed in the pathogenesis of choledochal cysts. This occurs when the biliary duct and pancreatic duct join away from duodenal wall, leading to pancreaticobiliary reflux (PBR), which has been associated to the development of biliary malignancy. The mechanism is likely related to chronic inflammation from activation of pancreatic enzymes. We present the case of a patient with recurrent pancreatitis who was diagnosed with choledochal cyst and PBM with the use of Eovist® (gadoxetate disodium), a hepatospecific contrast agent used for MRCP. Case Our patient is an 11-year-old female presenting with a third episode of pancreatitis in the span of 2 years. Her weight and height z-scores were -2.77 and -1.71, respectively. Her laboratory work-up was remarkable for elevated amylase (2800 IU/L), lipase (23415 IU/L) and transaminases. Her thyroid function, cystic fibrosis screen, and celiac serology had been negative. Her computed tomography (CT) abdomen 2 years prior showed thickening of the pancreatic head and tail; gallbladder was unremarkable. In her second episode of pancreatitis, her ultrasound (US) showed enlarged pancreatic tail with slightly coarse echotexture with a prominent common bile duct (CBD) tapering back to normal at the most distal portion. Her US gallbladder showed normal size and shape; mild fusiform dilatation of the CBD which measured 0.4 cm and tapering to 0.3 cm, suggesting a choledochal cyst. Our patient had a magnetic resonance cholangiopancreatography (MRCP) with Eovist® which showed fusiform dilatation of the common hepatic duct tapering down to the mid CBD and terminating abruptly approximately 1 mm before the confluence of the right and left hepatic ducts, suggestive of a pancreaticobiliary maljunction and a gallstone. She was transferred to another institution for endoscopic retrograde cholangiopancreatography (ERCP). Discussion Eovist® is a gadolinium-based contrast agent that is taken up by hepatocytes within 20 minutes after an intravenous bolus. It utilizes the same transporter that uptakes bilirubin into hepatocytes, OATP1. The use of this contrast media in pediatrics has been shown to have clinical utility in the diagnosis of biliary strictures or communications, as well as for evaluation of hepatic functional status. One study showed that its use provided important diagnostic information that led to change in management for 88.2% of patients. This case highlights the use of a non-invasive approach for diagnosis of anatomic pancreaticobiliary abnormalities. In our case, MRCP with Eovist® provided a better image of the anatomic malformation in this patient improving diagnostic accuracy. Although our patient ultimately required ERCP, the use of Eovist® for MRCP in children with normal anatomy may altogether obviate the need for the more invasive approach.

**217 THE PRESENCE OF ANSA PANCREATICA IN ASSOCIATION WITH ACUTE PANCREATITIS IN PEDIATRICS.** A. Panchoo, Pediatrics, Jackson Memorial Hospital, Miami, Florida, UNITED STATES. D. Rivera Rivera, Pediatrics - Gastroenterology, University of Miami, Miami, Florida, UNITED STATES. J.C. Infante, Radiology - Pediatrics, University of Miami, Miami, Florida, UNITED STATES.

Introduction: Ansa Pancreatica is a rare anatomical abnormality of the pancreatic ducts. In this variant the accessory duct of Santorini takes an atypical looped or curved course before joining the main pancreatic duct of Wirsung. The clinical significance of ansa pancreatica remains debatable, but recent research suggests an association with recurrent acute pancreatitis. However, to our knowledge, no pediatric cases of pancreatitis in the literature have been attributed to the presence of ansa pancreatica. Case: We report the case of a 15 year old female who presented with a 2 week history of worsening abdominal pain and 1 day of nausea, vomiting, diarrhea and decreased oral intake. Physical examination was significant for diffuse lower abdominal tenderness with stable vital signs. Initially consideration was given to acute gastroenteritis by the general pediatrics service, but with persistence of symptoms despite therapy further work up was pursued. Laboratory evaluation revealed elevated amylase at 284 units/L [Ref range 30-110 units/L] and lipase at 1565 units/L [Ref range 23- 300 units/L]. Extensive work up was done to identify the source of the pancreatitis and no culprits were identified. Her triglyceride level, electrolytes, bilirubin and hepatic function panel were all within normal limits. Also, she had a negative autoimmune screen. An MRI of the pancreas, in order to rule out anatomical abnormalities that could potentially explain this episode was significant for the finding of an ansa pancreatica. No other abnormalities were found in the abdomen or pancreatobiliary tract. With continued care the patient's oral tolerance steadily improved with resolution of nausea, vomiting, diarrhea and abdominal pain. Lipase levels decreased through discharge and normalized (68 units/L) on outpatient follow up. Discussion: With this case we aim to highlight the rare entity of ansa pancreatica as a consideration in patients with recurrent attacks of acute pancreatitis. Although there has been literature suggesting an association between ansa pancreatica and acute pancreatitis, all of these reported cases have been in adults. Also there have been very few articles linking the presence of ansa pancreatica with pancreatitis in the setting of alcohol consumption. There is no established incidence in the population, but some studies point that ansa pancreatica is present in around 1% of adult patients with acute pancreatitis. To our knowledge, this is the first report in the literature of ansa pancreatica as the cause of acute pancreatitis in a pediatric patient. Although still possibly a rare cause of pancreatitis in the pediatric population, this abnormal anatomical variation is worth to be considered when no other abnormalities are found in the patient's work up. Further population studies in order to determine prevalence and significance in the pediatric population are needed and long term follow up must be considered since this can lead to other episodes of pancreatitis during the patient's life.

**225 RECOVERY OF TOTAL VILLOUS ATROPHY IN A PEDIATRIC PATIENT AS IDENTIFIED BY VIDEO CAPSULE ENDOSCOPY.** J.M. Colombo, D. El Tawil, Pediatric Gastroenterology, Children Mercy Kansas City, Kansas City, Missouri, UNITED STATES. Radhi, Pediatric Bone Marrow Transplant, Children's Mercy Kansas City, Kansas City, Missouri, UNITED STATES.

Capsule endoscopy (CE) provides a safe, painless, full color view of the small intestine for the detection of gastrointestinal pathology beyond the reach of a traditional endoscopy.<sup>1-7</sup> Additionally, it allows examination of intestinal villi when biopsy cannot be performed. Follow-up CE assessment of the small bowel aides in the management of children with chronic diseases such as Celiac Disease and Crohn's Disease.<sup>8-11</sup> This is the first case report of a child presenting with total villous atrophy as detected by CE and follow-up CE demonstrating regeneration of intestinal villi. A 7 year old girl presented for inpatient consultation for a one month history of diarrhea at six months post-transplant for beta Thalassemia Major. Stools were entirely liquid with oil droplets. Frequency ranged from 5-8 times/day and occurred consistently after meals. She required total parenteral nutrition (TPN) to maintain adequate fluid and electrolyte balance. There were no associated fevers, weight loss, vomiting, abdominal pain or distention. She was afebrile with normal vital signs. Anthropometric parameters plotted at the 44<sup>th</sup> percentile for weight and 40<sup>th</sup> percentile for height. Physical examination revealed a well appearing child. Abdomen was soft, non-tender, non-distended, with normal bowel sounds without hepatosplenomegaly. Laboratory investigation was notable for hypokalemia (2.6 mmol/L) and hypoproteinemia (albumin 2.2 gm/dL and total protein 4.1 gm/dL). IgA tissue transglutaminase antibody was normal. C reactive protein was elevated (3.5 mg/dL). Stool for infection, C. difficile, occult blood, and reducing substances were negative. Fecal fat testing was positive. Pan endoscopy was visually normal. Biopsies from the

antrum and rectum were unremarkable. Testing for human herpes virus 6 on the gastric and rectal mucosa was negative. Biopsies from the duodenum were not obtained. CE demonstrated total villous atrophy throughout the entire small bowel. She was discharged home on a gluten free diet plus TPN and diphenoxylate/atropine twice daily. Approximately 2 months following discharge, the diarrhea resolved and TPN and diphenoxylate/atropine were discontinued. Repeat CE demonstrated regeneration of small intestinal villi throughout the entire small bowel. Our case is unique in its use of CE to demonstrate regeneration and recovery of small intestinal villi within 2 months. Biopsies of the small intestine could not be performed given our patient's history of bone marrow transplant and thus increased risk of duodenal bleeding<sup>12</sup>; however, CE provided vital information regarding this patient's clinical symptoms. Previous studies have demonstrated the utility of monitoring mucosal healing in Celiac disease and Crohn's disease.<sup>13-14</sup> To our knowledge, no other case has been described demonstrating regeneration of small intestinal villi documented by CE at 8 weeks.

**Poster Session II**  
**Friday, November 3, 2017**  
**12:00pm – 2:00pm**

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**243 COLONIC LYMPHANGIOMATOSIS IN A 16 YEAR OLD.** D. Sankepalli, A. Aktay, Pediatric Gastroenterology , University Hospitals Rainbow Babies and Children's Hospital, Beachwood, Ohio, UNITED STATES.  
Introduction: Lymphangioma is considered a benign tumor of lymphatic vessels. The incidence of lymphangiomas in Gastrointestinal tract is low in pediatric population. We report a case of 16 year old boy who presented with intermittent lower abdominal pain and rectal bleeding and was found to have colonic lymphangiomas. Case Presentation: A 16 year old male with history of constipation presented with lower abdominal pain and rectal bleeding for few months and unintentional weight loss. He had normal laboratory evaluation. He underwent colonoscopy which showed cluster of nodular polypoid lesions in the right colon, one of the lesion was biopsied. Endoscopic appearance was suggestive of lymphangiomas. He also had a hemorrhoid. Biopsy of the polypoid lesion showed prominent lymphoid aggregates. He underwent capsule endoscopy and Magnetic resonance enterography which were normal. His rectal bleeding was thought to be due to hemorrhoids. Discussion: Lymphangioma is a benign anomaly of lymphatic vasculature. When lesions are multiple or widespread, the term lymphangiomas is used. The incidence of lymphangiomas in GI tract is low and they are infrequently reported in pediatric population. They are diagnosed by typical endoscopic appearance and histopathology. Endoscopically lesions usually appear round and flat polypoid like with wide base and are of same color as surrounding mucosa. Endoscopic ultrasound further helps in defining the characteristics and qualitative assessment of these lesions. Ultrasonographic features are multilocular cystic strictures, homogenous echo pattern and localization of submucosal layer. There have been no reports of malignant transformation of lymphangiomas. Management is based on size of the lesions. Smaller lesions are usually managed with endoscopic polypectomy. Large size lesions can cause intussusception or obstructive symptoms and may need surgical excision. Polypectomy has risks of bleeding and perforation. In our case we are observing and closely following patient and will consider repeat colonoscopy if symptoms recur.

**244 AN UNCOMMON DIAGNOSIS FOR A COMMON GI PRESENTATION.** D. Moores, Pediatric Surgery, Loma Linda University Children's Hospital, Loma Linda, California, UNITED STATES. Truong, M. Shah, Pediatric Gastroenterology, Loma Linda University Children's Hospital, Loma Linda, California, UNITED STATES. L. Denham, Pathology, Loma Linda University Children's Hospital, Loma Linda, California, UNITED STATES. F. Ali, Pediatrics, Loma Linda University Children's Hospital, Loma Linda, California, UNITED STATES.  
Learning Objectives● Recognize that Birt-Hogg-Dube Syndrome (BHDS) is diagnosed by clinical findings and genetic molecular testing, with FLCN (Folliculin, a.k.a. BHD) as the only known gene associated with BHDS● Recognize that colonic neoplasms can be a feature of affected individuals with BHDS  
Case PresentationA 6 year old boy who underwent a heart transplant for cardiac rhabdomyoma presented with a 2 month history of worsening profuse watery diarrhea, weight loss, and anemia without associated fevers, vomiting, or abdominal pain. Symptoms were unresponsive to changes in diet or immunosuppressive therapy. Extensive initial screening tests for lymphoproliferative, infectious, and malabsorptive causes were negative. Colonoscopy showed rectosigmoid

masses with histology consistent with high grade dysplasia, solid areas of intramucosal carcinoma, and foci of poorly differentiated invasive adenocarcinoma. An EGD and subsequent push-enteroscopy grossly identified multiple polypoid masses throughout the small bowel with histologic evidence low grade and high grade dysplasia. Gene testing revealed negative APC gene, but whole exome sequencing detected “heterozygous pathogenic variant in the FLCN gene” consistent with Birt-Hogg-Dube Syndrome (BHDS). The child subsequently underwent a partial colectomy, with plans for total proctocolectomy and end ileostomy. Discussion BHDS is a rare autosomal dominant condition clinically characterized by skin, pulmonary, and renal findings. Cutaneous manifestations include fibrofolliculomas, angiofibromas, and acrochordons. There may be lung cysts with high risk of spontaneous pneumothorax and various types of renal tumors that are typically bilateral and slow growing. The condition is caused by a germline mutation in the FLCN (Folliculin, a.k.a. BHD) gene, which has been linked to the mTOR pathway in tumor suppression. There is great variation in FLCN expression, and the severity of disease can vary significantly even within the same family. There have been rare reports of other tumor types affecting individuals with BHDS into their adulthood including thyroid cancer, squamous cell carcinoma, Hodgkin disease, rhabdomyoma, and neuroendocrine carcinomas. Additionally, while it was thought BHDS did not cause colon cancer, there have been multiple reports of individuals affected with it, such as our patient. He is currently the youngest patient with colon cancer in BHDS. Diagnosis is based on molecular genetic testing, where the FLCN gene is the only pathogenic variant known to cause BHDS. These reports may very well explain both the cardiac rhabdomyoma and colonic adenocarcinoma of our patient, and help us to consider colon cancer screening as part of the potential cancers associated with this syndrome. BHDS should be suspected in patients with more than one cancerous or pre-cancerous nidus.

**245** *EUS WITH FNA IN THE EVALUATION OF A PEDIATRIC PATIENT WITH A MEDIASTINAL MASS.* F.C. Lee, Pediatrics, Baylor College of Medicine, Houston, Texas, UNITED STATES. P. Shah, V. Ramachandran, Baylor College of Medicine, Houston, Texas, UNITED STATES. Fallon, S. Keswani, A. Vogel, B. Naik-Mathuria, Pediatric Surgery, Baylor College of Medicine, Houston, Texas, UNITED STATES. Keswani, J. Hicks, S. Vasudevan, K. Canadas, A. Vogel, B. Naik-Mathuria, D.S. Fishman, Texas Children's Hospital, Houston, Texas, UNITED STATES. Barton, D.S. Fishman, Pediatric Gastroenterology, Hepatology and Nutrition, Baylor College of Medicine, Houston, Texas, UNITED STATES. Hicks, Pathology and Immunology, Baylor College of Medicine, Houston, Texas, UNITED STATES. Vasudevan, K. Canadas, Otolaryngology, Baylor College of Medicine, Houston, Texas, UNITED STATES.

Introduction: Common mediastinal masses, such as esophageal duplication or bronchogenic cysts, are difficult to differentiate by routine cross sectional imaging. Endoscopic ultrasound (EUS) and endoscopic ultrasound-guided fine needle aspiration (EUS-FNA) have been used in the diagnosis of mediastinal masses in adult patients, but use in pediatric patients has not been well-established. Case: A 9 year old female with a 2-week history of worsening cough is found to have a posterior mediastinal mass thought to be a bronchogenic or esophageal duplication cyst. The mass was partially obstructing the trachea and esophagus, but surgical intervention was postponed for treatment of Influenza B pneumonia. She subsequently developed acute desaturation and was taken for urgent endoscopic evaluation of her airway. Bronchoscopy demonstrated marked obstruction at the carina and right bronchus. EUS showed a heterogenous, hypoechoic echotexture mass with fluid that appeared contiguous with the esophageal wall. EUS-FNA was performed to decompress the lesion and relieve some mass effect, with a Cook Medical 25 and then 22 EchoTip, with a small amount of thick mucoid fluid removed. This likely provided a conduit for drainage as the saturations improved over the next 24 hours. She subsequently underwent an uneventful thoracoscopic excision of the mass. Pathologic evaluation showed respiratory mucosa, seromucinous glands, and cartilage, consistent with a bronchogenic cyst. Conclusion: EUS can be used to help characterize mediastinal masses when diagnosis is uncertain. EUS-FNA may be used as part of a diagnostic and management strategy in situations necessitating urgent drainage. This case illustrates the potential of EUS and EUS-FNA in the diagnosis and evaluation of mediastinal masses in the pediatric population.

**246** *GASTROCOLIC AND COLOCUTANEOUS FISTULAS AS A COMPLICATION OF PERCUTANEOUS ENDOSCOPIC GASTROSTOMY IN AN ADOLESCENT BOY WITH EXCESSIVE WEIGHT LOSS.* G.H. Ustundag, Y.D. Soysal, Pediatric Gastroenterology, Hepatology and Nutrition, Bulent Ecevit University, Zonguldak, TURKEY. E. Piskin, N. Hosgöl, Pediatrics, Bulent Ecevit University, Zonguldak, TURKEY. D. Tatli, Pediatric Surgery, Bulent Ecevit University, Zonguldak, TURKEY.

**Introduction:** Percutaneous endoscopic gastrostomy (PEG) is a method commonly used for providing long-term nutrition for patients unable to swallow but with a functioning gastrointestinal tract. Despite the advanced imaging techniques, and within the hands of an experienced endoscopists serious complications of PEG insertion can still be encountered. Here we present a neurologically impaired adolescent boy with the formation of gastrocolic and colcutaneous fistulas as a result of migration of the feeding tube to the transvers colon. **Case Presentation:** A 15-years-old boy with chronic malnutrition due to feeding difficulties related to cerebral palsy had been inserted a feeding tube by PEG pull through method in another medical center. Two months after the procedure, he presented with recurrent vomiting and diarrhea after each feeding session and with significant weight loss. It was noticed by the parents that a material resembling feces was coming retrogradely from the feeding tube occasionally. The patient had applied several times to the medical center that placed the feeding tube, but since the symptoms were transient and the tube was thought to be functioning properly, he was discharged with gastroesophageal reflux medications. Suspecting of a gastrocolic fistula, an endoscopy was performed in our unit, and the mushroom end of the tube was not present in the stomach. When the tube was pushed from outside a protuberance was seen on the stomach surface, but it was totally covered with gastric mucosa. Water soluble contrast material was given from the feeding tube under fluoroscopy to demonstrate if there was any connection of the tube with the stomach. The contrast material did not flow to the stomach lumen but probably to the colon. Therefore, a computer tomography of the abdomen was conducted to locate the exact placement of the bumper, which turned out to be in the transvers colon. Then, the patient was consulted to the pediatric surgery department and he was operated. The bumper of the feeding tube was removed from the colon, and both of the fistulas tracts were repaired. A new gastrostomy tube was placed surgically. In the follow up, the patient was able to be fed from the feeding tube within a few days without further problems, and he started to gain weight. **Conclusion:** Major complications of PEG tube placement such as gastrocolic fistula should be suspected earlier with prompt endoscopic or radiological examination in a patient with continuous feeding difficulty, diarrhea and significant weight loss.

**247 ACUTE APPENDICITIS ON ENDOSCOPY: A CASE REPORT AND REVIEW OF THE LITERATURE.** G.N. Bahia, W. Elfar, E. Prince, T. Rossi, Pediatric Gastroenterology & Nutrition, University of Rochester Medical Center, East Syracuse, New York, UNITED STATES.

**Background** Acute appendicitis is a clinical diagnosis. Imaging studies, such as ultrasound and CT scan, assist in confirming this diagnosis. Endoscopy is considered a relative contraindication in cases of acute appendicitis due to the theoretical increase risk of perforation. However, features of acute appendicitis can be detected at colonoscopy. We report a case of acute appendicitis that was initially unclear on clinical and imaging diagnostics, which then became apparent on colonoscopy. **Case report** 18 years old female, 9 weeks postpartum, who presented to the emergency room with worsening left flank pain and new onset right lower quadrant pain. Two weeks prior to this presentation, she was admitted to the hospital for the management of left pyelonephritis complicated by bacteremia. CT scan of the abdomen with contrast was performed which showed thickening of the proximal appendiceal wall and a fecalith. The surgery team was consulted but felt that her clinical examination did not correlate with the imaging findings. We were asked to perform a colonoscopy with biopsies to assess suspected appendiceal - cecal mass. Fecal occult blood and fecal lactoferrin prior to performing the procedure were negative. She underwent colonoscopy with successful cecal intubation. To our surprise, we discovered what appeared to be an inflammatory mass bulging from the appendiceal orifice. This inflamed protrusion was visually consistent with images of acute appendicitis seen in the literature. No biopsies were obtained at that time. Subsequently, she underwent exploratory laparotomy with ileocectomy in anticipation of finding an appendiceal mass. The surgical pathology came back consistent with acute appendicitis in the proximal appendix. The tip of the appendix was not involved, and there were no inflammatory changes in the adjacent cecum. **Discussion** Acute appendicitis is the most common abdominal emergency, and diagnosing this condition can be challenging. Literature review reveals only a few cases of appendicitis diagnosed by colonoscopy. Appendiceal masses can mimic appendicitis, and it is difficult to distinguish between the two conditions on imaging studies without obtaining biopsies. This report highlights the role of colonoscopy in challenging cases especially to distinguish appendicitis from appendiceal masses.

**254 A CAMERON ULCER IN A 3-YEAR-OLD BOY PRESENTING WITH PERSISTENT ANEMIA.** J.E. Mathew, Department of Pediatrics, Children's Hospital at Montefiore, Bronx, New York, UNITED STATES. J.E. Mathew, I. Novak, E. Prendaj,

D. Jan, G. Tomer, Albert Einstein College of Medicine, Bronx, New York, UNITED STATES. D. Jan, Division of Pediatric Surgery, Children's Hospital at Montefiore, Bronx, New York, UNITED STATES. I. Novak, E. Prendaj, G. Tomer, Division of Gastroenterology and Hepatology, Children's Hospital at Montefiore, Bronx, New York, UNITED STATES. A 3-year-old male with past medical history of congenital right diaphragmatic hernia status post Nissen fundoplication and repair who is G-tube dependent was admitted due to fever and coffee ground secretions from his G-tube. His workup revealed decreased hemoglobin from 11.3 to 9.4 g/dL, and iron studies were as follows: ferritin 9 ng/mL, iron 21 ug/dL, TIBC 449 ug/dL, transferrin 359 mg/dL, and 5% saturation. A respiratory viral panel was negative, and electrolytes and liver function tests were normal. He was started on Lansoprazole and iron supplements, and he was discharged home. He was followed in the gastroenterology and surgery clinics for several months, and due to persistent anemia despite iron supplementation, an esophagogastroduodenoscopy (EGD) was performed that showed a hiatal hernia. Because of worsening anemia (Hb 6.9 g/dL) and dark stools, he was hospitalized for further evaluation. An upper GI series revealed a partially unwrapped fundoplication with herniation through the diaphragmatic hiatus (Figure 1). He received a blood transfusion and an EGD demonstrated an ulcer in the hiatal hernia. A video capsule demonstrated erosion from the same area, but was otherwise normal. Histopathology was consistent with chronic gastritis, Helicobacter pylori testing was negative, and a colonoscopy was normal. His hemoglobin remained stable, and he was discharged home on Omeprazole and iron supplements. He was re-hospitalized in the setting of decreasing hemoglobin, pallor, tachycardia, and dark stools. Exploratory laparotomy and intraoperative EGD were performed, showing erythematous erosions in the cardia consistent with a Cameron ulcer (Figure 2). He underwent lysis of adhesions, repair of the hiatal hernia, and revision of the Nissen fundoplication with mesh placement. Since the surgical repair, the patient has been doing well with a stable hemoglobin at 11.9g/dL. Cameron ulcers are linear mucosal erosions or ulcers in large hiatal hernias where they are constricted by the thoracic diaphragm. The etiology is unclear but presumed to be due to ischemia, mechanical trauma, and/or gastric acid. Lesions occur most often in older adults. They are often asymptomatic but can present as occult gastrointestinal bleeding and can usually be treated with iron supplementation and gastric acid suppression. Cameron ulcers less often can cause acute bleeding and need surgical repair, as in our case. Cameron ulcers are rare and the literature is limited to case series and case reports. To our knowledge this is the youngest patient reported with a Cameron ulcer.

**255 CONGENITAL ESOPHAGEAL STENOSIS.** J. Barry, P. Conjeevaram Selvakumar, S. Patel, Pediatric Gastroenterology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES. E. Park, Pediatric Radiology Imaging Institute, Cleveland Clinic, Cleveland, Ohio, UNITED STATES.

An 11-month-old female presented with symptoms of progressive dysphagia since 3 months of age. Dysphagia progressed from solids to liquids at 11 months of age, with associated weight loss. An upper GI series found a 3 cm segment in the lower esophagus with severe concentric and irregular narrowing (Fig 1). CT scan documented no paraesophageal mass or external compression. Endoscopy visualized a stricture in the lower esophagus measuring 3 mm in diameter (Fig 2) through which a neoscope was unable to advance. Histology demonstrated non-peptic stricture, without eosinophils, consistent with a diagnosis of congenital esophageal stenosis (CES). Serial Savary and Maloney dilation from 12F to 46F diameter was performed with resolution of dysphagia. CES results from malformation of the esophageal wall architecture in utero resulting in stenosis. It has a reported incidence of 1 in 25,000-50,000 infants. Diagnosis is often apparent on esophagogram. In our patient, worsening dysphagia, along with the abovementioned test findings all support the diagnosis of CES. Unusual in our case is the long segment of esophagus involved. CES is a rare entity that should be suspected in children with early onset dysphagia.

**257 MALAKOPLAKIA: A RARE CAUSE OF RECTAL BLEED IN CHILDREN.** K. Sadiq, Pediatrics and child health, Aga Khan University, Karachi, Sindh, PAKISTAN. K. Sadiq, Aga Khan University, Karachi, Sindh, PAKISTAN.

**INTRODUCTION** Rectal bleeding has frequently been attributed to polyps and anal fissures. Malakoplakia is a chronic inflammatory disorder seen mostly in the genitourinary tract, although other organ systems have also been known to be affected. Herein, we describe a case of rectal bleeding due to malakoplakia, a rare disorder that is not typically described in children.

**CASE REPORT** A 4-year-old boy presented with recurring episodes of bleeding per rectum for the last 2 years. On colonoscopy, multiple small, pale nodules were seen in the rectum on endoscopy while the rest of the large intestine appeared normal. The histological features of the rectum were strongly suggestive of malakoplakia.

**DISCUSSION** Malakoplakia is a rare chronic inflammatory disorder. The commonest site is genitourinary system followed by the gastrointestinal system. The exact pathophysiology of

malakoplakia remains elusive, but is thought to be due to a defect in the bacteriocidal activity of macrophages. Grossly, malakoplakia appears as soft pale plaques or nodules. Histologically, it is identified by the presence of sheets of ovoid histiocytes (von Hansemann cells) with accumulation of granular, basophilic PAS-positive inclusions and calcified Michaelis-Gutmann bodies. TREATMENT Antibiotics that concentrate in macrophages (eg, quinolone, trimethoprim-sulfamethoxazole) are used to treat malakoplakia

**258 AN ESOPHAGEAL MAGNET IMPACTION IS MISSED RADIOGRAPHICALLY WITH POTENTIAL COMPLICATION OF ESOPHAGEAL-GASTRIC FISTULA.** K. Black, R. Abdelhadi, Pediatrics, Children's Mercy Hospital, Kansas City, Missouri, UNITED STATES.

Foreign body ingestion is common in the pediatric population with an annual incidence rate of over 125,000 ingestions/year according to the American Poison Control Centers 2007 report. Multiple magnet ingestions have deleterious potential consequences of the digestive tract including perforations secondary to perfusion compromise and pressure necrosis of the bowel walls caught between magnets. This can result in emergent bowel resection and subsequent short bowel syndrome. A toddler presented with sudden onset abdominal pain and 2 episodes of emesis while in a room unsupervised with small magnetic toys. The father ran one of the excess magnetic rods along her stomach and felt movement of objects inside her stomach. Her physical exam was significant for palpable mobile rigid mass in the right upper quadrant. A chest and abdomen AP and lateral views revealed 10 tubular opaque shadows partially aligned in the upper and mid-abdomen with the radiology report 'likely intragastric'. Approximately 5 and half hours after the alleged ingestion time, and unexpectedly, the endoscopy revealed that one magnet was fixated horizontally in the distal esophagus catching the esophageal wall and gastric fundus between it and the remaining 9 aligned magnets in the stomach. After successful detachment and removal of that esophageal magnet, a well-defined ulcer was visible in the distal esophageal wall. The remaining 9 intragastric magnets were adherent to one another and were removed one by one with Roth net and alligator forceps. Another matching ulcer was seen in the gastric fundus. The patient tolerated the procedure well and was discharged with no subsequent esophageo-gastric fistulas or mediastinitis. This case illustrates the limitation of radiologic studies in accurately diagnosing foreign body location, predicting complications, or assisting in triaging the urgency level and timing of intervention. In this patient, an esophageal magnet impaction was completely missed and a pressure necrosis process was underway towards perforation within less than 6 hours of ingestion. Radiologic studies are limited in discerning with accuracy magnet locations since multiple magnets tend to distort anatomic landmarks, and contrast studies may provide insight however will likely delay the planned endoscopic evaluation and intervention; CT scans will likely show streak artefacts. This reiterates that with higher and additive magnetic force the grave danger with multiple magnet ingestion rises exponentially, highlighting the need for emergent removal.

**259 A CASE OF HEREDITARY GASTRIC CANCER DETECTED INCIDENTALLY IN A 17 YEAR-OLD GIRL.** K.M. Johnson, F.C. Giangiacomo, L. Hattar, Pediatrics, University of Kansas-Wichita, Wichita, Kansas, UNITED STATES. Tofteland, Internal Medicine/Pediatrics, University of Kansas- Wichita, Wichita, Kansas, UNITED STATES.

A 17 year old Caucasian female with past medical history of developmental delay and seizure disorder presented with a one year history of abdominal pain. She was found to have H. pylori infection based on serology and received eradication therapy. However, as abdominal pain persisted, she was referred to pediatric gastroenterology. Relevant family history was significant for a paternal grandmother who successfully underwent treatment of gastric cancer in her 40's, a maternal grandmother with esophageal cancer in her 70's, and other close relatives with various types of cancer. After routine workup for abdominal pain did not identify an etiology, a decision was made to proceed with esophagogastroduodenoscopy (EGD) and colonoscopy. EGD did not reveal any gross abnormalities, but random gastric biopsies revealed signet ring cell adenocarcinoma. Colonoscopy with biopsy was unremarkable. Patient underwent repeat EGD with endoscopic ultrasound (EUS) for further diagnosis and staging. Mapping biopsies of the stomach were obtained and one out of six areas confirmed adenocarcinoma. Endosonographic images of the stomach were unremarkable, with normal gastric wall thickness and no local or regional lymphadenopathy. Staging PET-CT did not reveal any distant metastases. Genetic testing was completed due to concern for hereditary diffuse gastric cancer (HDGC), the results of which returned positive for a mutation in the CDH-1 gene. She subsequently underwent exploratory laparoscopy with total gastrectomy, small bowel resection, extensive lymph node dissection, and Roux-en-Y esophagojejunostomy. Post-operative pathology revealed poorly-differentiated signet ring cell adenocarcinoma with negative margins and no evidence of

metastatic disease. Multiple patchy foci of adenocarcinoma were identified microscopically throughout the gastric mucosa, each measuring 2-3 mm in size. Her mother and half-sister were subsequently found to have the same genetic mutation and underwent prophylactic gastrectomy without evidence of malignancy on histologic evaluation. Gastric cancer is an extremely rare pediatric diagnosis. Gastrointestinal malignancies account for just 5% of all pediatric neoplasms and primary gastric adenocarcinoma (GAC) represents 0.05% of all childhood cancers. Literature on GAC in children and adolescents is limited and management is based on adult studies. Etiology of GAC may be multifactorial, but there are reports of patients with hereditary diffuse gastric cancer who have a germline mutation in E-Cadherin (CDH-1). This mutation is associated with high penetrant susceptibility with autosomal dominant inheritance. Prophylactic gastrectomy is strongly advised in carriers of a proven pathogenic germline CDH1 mutation. Genetic evaluation and counseling is important for affected patient and family members. Treatment of gastric cancer depends on the histologic type and stage. EUS is a valuable method for locoregional staging, as it detects depth of gastric wall invasion and the potential presence of regional lymph node involvement. In addition, it provides an opportunity for fine needle aspiration of deeper histologic specimens than mucosal biopsies allow and sampling of lymph nodes that endosonographically appear suspicious for malignancy. Its use in the staging of adult GI malignancy is well established, and has a limited but promising role among pediatric patients with respect to tumor staging as well as diagnostic and therapeutic intervention for a variety of pancreatico-biliary diseases.

**260 ENDOSCOPIC ULTRASOUND IN A PEDIATRIC PATIENT WITH LYNCH SYNDROME.** K.P. Shah, V. Ramachandran, Office of Undergraduate Medical Education, Baylor College of Medicine, Houston, Texas, UNITED STATES. F.C. Lee, Department of Pediatrics, Baylor College of Medicine, Houston, Texas, UNITED STATES. S. Vasudevan, Department of Pediatric Surgery, Texas Children's Hospital, Houston, Texas, UNITED STATES. R. Venkatramani, Department of Pediatrics, Section of Hematology-Oncology, Texas Children's Hospital, Houston, Texas, UNITED STATES. D.S. Fishman, Pediatric Gastroenterology, Hepatology and Nutrition, Texas Children's Hospital, Houston, Texas, UNITED STATES.

**Background:** Colorectal cancer is a rare condition in the pediatric patient population. Less than 0.1% of new cases of colorectal cancer are diagnosed in patients less than 20 years of age. Hereditary nonpolyposis colorectal cancer (HNPCC), otherwise known as Lynch syndrome, is the most common hereditary colorectal cancer syndrome. It is an autosomal dominant condition that increases the risk of many cancers including colorectal, endometrial, gastric, and ovarian cancers. Typically, HNPCC is caused by a germline mutation in one of the following DNA mismatch repair genes: *MLH1*, *MSH2*, *MSH6*, or *PMS2*. Patients with HNPCC have a 52-82% lifetime risk of developing colorectal cancer. The mean age of HNPCC diagnosis is between 44-61 years. It is incredibly rare for de novo HNPCC germline mutations to be discovered before adulthood. **Case Presentation:** A previously healthy 15-year old Hispanic female presents with bloody diarrhea, unexpected 7 lb. weight loss, and stabbing pain in the lower abdomen after bowel movements. Workup revealed anemia with a hemoglobin of 7.8 g/dL treated with oral iron supplements that paradoxically increased bowel movements from 3-4 stools per day to 5-6 times per day. The patient eventually underwent an esophagogastroduodenoscopy with biopsy and a flexible sigmoidoscopy with endoscopic ultrasound (Figure 1), which revealed ulcerated areas with friable mucosa and exudates/edema seen at 15 cm from the anus. Pathology confirmed the presence of invasive adenocarcinoma in the rectosigmoid colon. Further examination showed metastasis to an intrapelvic lymph node. A colorectal hereditary panel revealed a de novo c.307-2A>G (IVS3-2A>G) change in the *MLH1* gene, confirming a diagnosis of HNPCC. Management of this condition began with a complete course of chemoradiation with capecitabine followed by a sub-total colectomy and colostomy for both prophylaxis and primary removal of colorectal adenocarcinoma. Adjuvant chemotherapy with capecitabine and oxaliplatin was then administered for six cycles over four months prior to final ostomy takedown and re-anastomosis. At the last follow-up, 3 months post-ostomy takedown and 13 months post-diagnosis, the patient was in remission with no complaints. She is able to eat a regular diet with painless bowel movements. **Conclusion:** This vignette illustrates the use of endoscopic ultrasound in parallel with imaging for the diagnosis of an adolescent with a de novo germline mutation of *MLH1* resulting in HNPCC. Future research should be directed at cost-effective methods to screen pediatric patients for genetic conditions such as HNPCC.

**261 FECAL IMPACTION: UNUSUAL PRESENTATION OF COLONIC ADENOCARCINOMA.** L. Diaz Calderon, L. Conklin, Pediatric Gastroenterology, Hepatology and Nutrition, Children's National Medical Center, Washington, District of

Columbia, UNITED STATES. J.L. Lyles, Medical Education, Children's National Medical Center, Washington, District of Columbia, UNITED STATES.

Colorectal carcinoma is a rare disease in pediatric population. We present a 15-year-old Hispanic male who was initially diagnosed with fecal impaction that later progressed as partial bowel obstruction and ultimate diagnosis of colonic adenocarcinoma. Patient initially presented with abdominal pain and difficulty stooling. Abdominal Xray showed large amount of stool throughout the colon and a barium enema was remarkable for narrowing of the lumen of the descending colon which was attributed to fecal impaction that was managed with bowel clean out. Six-months later, he had similar symptoms. No history of vomiting, weight loss, recurrent fevers or feeding intolerance. On exam patient seemed well nourished with abdominal tenderness to palpation without acute peritoneal signs. There was evidence of bowel obstruction on abdominal Xray. CT abdomen showed a 5cm long segment of severe bowel wall thickening and stricturing within the mid descending colon with surrounding inflammation. Attempted colonoscopy was able to advance up to 40cm in the descending colon due to luminal obstruction by a circumferential, firm, erythematous friable mass with pin point lumen. Patient underwent left hemicolectomy and was diagnosed with stage 3 adenocarcinoma of the distal colon. He completed chemotherapy 8 months later and is currently doing well. This case elucidates the importance of having high index of suspicion of mechanical causes of chronic constipation and fecal impaction refractory to treatment.

**264 THE PYLORUS...A GALAXY FAR, FAR AWAY. GASTRIC OUTLET OBSTRUCTION CAUSED BY AN R2-D2 CHILDREN'S TOY.** M.S. Rosario, C. Maier, Internal Medicine-Pediatrics, USF Health, Lutz, Florida, UNITED STATES. Wilsey, E.K. Swan, Florida State University, Tallahassee, Florida, UNITED STATES. V. Falconer, S. Son, University of Florida, Gainesville, Florida, UNITED STATES. Wilsey, Pediatrics, University of South Florida, Tampa, Florida, UNITED STATES. Wilsey, Gastroenterology, Johns Hopkins All Children's Hospital, St. Petersburg, Florida, UNITED STATES.

Background Foreign body ingestion (FBI) is a common pediatric issue. At least 100,000 cases are reported in the U.S. yearly, 75-80% of which are by children. Ingested items are typically readily available within the home. Furthermore, pediatric FBIs are witnessed in only half of reported cases. FBIs can occasionally be asymptomatic, depending on where the foreign body (FB) is located. However, typical symptoms vary and include drooling, abdominal pain, chest pain, emesis, wheezing, stridor, and respiratory distress. Gastric outlet obstruction (GOO) caused by a FB is an extremely rare complication. The incidence of pediatric GOO is about 2-5/1000 per year, with the most common cause being pyloric stenosis. We present the case of a child with acute onset emesis and epigastric abdominal pain secondary to GOO. To our knowledge, this is the first case of GOO caused by FBI of a children's toy treated successfully with endoscopic removal.

Case Summary A 4-year-old male with no past medical history was directly admitted to the procedural suite for intractable, non-bloody, non-bilious emesis. The night prior to admission, he was asymptomatic upon telling his parents about ingesting an Angry Birds R2-D2 Egg™. He went to bed without any untoward events. Early in the morning, however, he developed uncontrollable, non-bloody, non-bilious emesis with epigastric abdominal pain. He was taken to his pediatrician's office where an X-ray was unremarkable. There was no clear explanation for his symptoms. Yet, the physician's Jedi instincts prompted him to have the patient directly admitted for endoscopic search and retrieval of said missing R2-D2 toy. On arrival, vitals and physical exam were unremarkable aside from abdominal tenderness. The patient was taken to the special procedures unit and placed under anesthesia. A GIF-160 endoscope was passed into the stomach, where the plastic toy was discovered, impacted within the pyloric channel. The toy was grasped with forceps and retrieved through the mouth without complications. A second look showed resolution of the pyloric outlet obstruction without evidence of ulcers or erosions. The patient tolerated the procedure well and was back to baseline upon waking from anesthesia. His epigastric pain and intractable emesis resolved.

Discussion We report the first case of GOO caused by FBI of a children's toy, treated successfully with endoscopic removal. There are guidelines for the management of FBI, specifically involving button batteries, pointed/sharp objects, magnets, coins, blunt objects, and superabsorbent objects. However, ingested items are not always radio-opaque. Endoscopic retrieval is considered earlier on if the patient is symptomatic. In our case, the cause of the patient's symptoms would have been unclear, given his unrevealing radiographs, had he not confessed to FBI. Newer techniques, such as sonography, should be considered to detect radiolucent objects. Many case reports have cited ultrasound as a useful, non-invasive, non-radiating tool for the diagnosis of FBI. A recent case study proposed the use of ultrasound to evaluate radiolucent FBs in the clinical setting of GOO. Overall, it is important to consider FBI in the differential of a toddler/child with intractable emesis and abdominal pain.

**265 GASTRIC BEZOARS IN CHILDREN: OFTEN OVERLOOKED, NEVER UNIMPORTANT.** J.M. Rhoads, Department of Gastroenterology and Nutrition, UT Health Science Center at Houston, Houston, Texas, UNITED STATES. Shah, J.M. Rhoads, Department of Pediatrics, UT Health Science Center at Houston, Houston, Texas, UNITED STATES.

**Introduction:** A bezoar (derived from Persian bazahr) is a mass found trapped in the gastrointestinal system (usually the stomach, occasionally the rectum). A bezoar in the esophagus or upper gastrointestinal tract is sometimes reported in developmentally delayed or institutionalized children. We have identified 19 pediatric patients diagnosed with gastric bezoar during a span of 9 years (2008-2017) at the University of Texas Health Sciences Center/Children's Memorial Hermann Hospital, Houston. We performed a retrospective case analysis to see how our findings compare to the pediatric literature relating to the occurrence of bezoars which are thought to be relatively rare causes of vomiting and abdominal pain in children. **Study method:** The study method used is a retrospective case series by chart review in patients identified by ICD-9 codes 938 and 935. We used the following Medical Subject Headings: 1. MeSH terms bezoar, 2. Keywords gastric bezoar\* or gastric foreign body\* and searched Pubmed, MEDLINE OVID, and TMC library resources data. **Results:** Of the 19 pediatric patients recognized with bezoars, only one had a trichobezoar; all the rest were phytobezoars. The patients ranged from age 2 to 18; all but one were older than 6. In contrast to prior published cases, we found developmental delay in only 5 of the 19 children. 5 patients had dysautonomia, evidenced by tilt-table testing, and 3 of these had pain-associated disability syndrome. Abdominal pain (70%), nausea and vomiting (70%), a decrease in appetite (40%), unintentional weight loss (30%) were the most common presenting symptoms. 6 children had a normal BMI for age, while 9 had a BMI < 5%tile, and 4 had a BMI > 95%tile. Nuclear medicine gastric emptying scan was performed in 7; surprisingly, 4 of the 7 had normal gastric emptying. 17 of the 19 patients were treated with endoscopic removal of the bezoar, but the child with trichobezoar required surgical removal. One with known liver disease had esophageal variceal band ligation without bezoar removal; on repeat endoscopy, the bezoar was gone. Endoscopic removal of most bezoars by Roth net generally required multiple passes (6-20 passes). At follow-up, most of the children had improvement of symptoms (n=13) or complete relief of symptoms (n=4). **Discussion:** This is one of the largest studies till date on gastric bezoars in pediatrics after the extensive study by DeBakey et al in 1938 enlisting 303 existing cases and 8 cases of their own, both in the adult and pediatric population, 40.4% of which were noted to be phytobezoars. Associated developmental delay and impaired gastric motility have been commonly described in older literature, although no description about association with dysautonomia has been noted in prior studies. Bezoar removal usually resulted in resolution of symptoms while reviewing literature, but no particular mention about long term outcome on follow up visits was noted. **Conclusion:** Phytobezoars may be underdiagnosed in pediatrics because of a low clinical suspicion. Bezoars should be considered in patients presenting with chronic abdominal pain, nausea and vomiting--even in developmentally normal children and even in those with normal gastric emptying. Symptoms will often improve with endoscopic removal. **References:** 1. DeBakey M, Ochsner A. Bezoars and concretions. *Surgery* 1939;4:132:60. 2. C. Roche, E. Guye, E. Coinde et al., "Five cases of trichobezoars in children," *Archives de Pediatrie*, vol. 12, no. 11, pp. 1608-1612, 2005. 3. V. N. Sehgal and G. Srivastava, "Trichotillomania ± trichobezoar: revisited," *Journal of the European Academy of Dermatology and Venereology*, vol. 20, no. 8, pp. 911-915, 2006. 4. R. R. Gorter, C. M. F. Kneepkens, E. C. J. L. Mattens, D. C. Aronson, and H. A. Heij, "Management of trichobezoar: case report and literature review," *Pediatric Surgery International*, vol. 26, no. 5, pp. 457-463, 2010. 5. W. B. Wadlington, M. Rose, and G. W. Holcomb Jr., "Complications of trichobezoars: a 30-year experience," *Southern Medical Journal*, vol. 85, no. 10, pp. 1020-1022, 1992.

**266 NON-H. PYLORI GASTRIC INTESTINAL METAPLASIA IN CHILDREN.** S.M. Camacho-Gomez, N.A. Tipnis, Pediatrics, University of Mississippi Medical Center, Jackson, Mississippi, UNITED STATES. Bernieh, A. Saad, Pathology, University of Mississippi Medical Center, Jackson, Mississippi, UNITED STATES.

**Introduction:** In children, Gastric Intestinal Metaplasia (GIM) is a finding with unknown frequency and clinical implications. We present two children with GIM and a review the literature. **Case Series** Case 1: A 16 yo female presented with progressive worsening of dysphagia to solid foods, a sensation of fullness in the chest and sour taste in her mouth but no heartburn or chest pain. She had no weight loss and no family history of gastric cancer. The physical exam was unremarkable and labs were normal (including H. pylori antibody). Her current medication include omeprazole. Esophagram was normal. She underwent esophagogastroduodenoscopy (EGD), which revealed a 4 mm pre-pyloric nodule. NBI demonstrated a villus pattern. H. pylori rapid urease was negative. Hematoxylin and eosin-stained sections on the pre-pyloric nodule biopsy show a antral-type gastric mucosa. The lamina propria is distended by a chronic inflammatory cell infiltrate consisting of lymphocytes and plasma cells.

Numerous signet ring cells, characteristic of intestinal epithelium, are identified. The features are those of chronic gastritis with intestinal metaplasia of the complete type. No dysplasia is present. H. pylori immunostain showed no organisms. Patient reports improvement in her symptoms with PPI therapy. Repeated EGD 1 year after showed the same findings on EGD and biopsies. Case 2: An 8 year old female presents with generalized cramping and sharp abdominal pain for one year associated with nausea and intermittent emesis. No family history of gastric cancer. Labs were normal, including H. pylori antibodies. EGD showed a pre-pyloric nodule and bile lake. Histology of the nodule revealed antral mucosa, chronic inflammation and incomplete GIM. Rapid urease testing for H. pylori as well as immunostains were negative. She was treated with double dose PPI therapy. Repeat EGD showed continued presence of the pre-pyloric nodule and incomplete gastric intestinal metaplasia without progression to dysplasia. Discussion: Gastric Intestinal metaplasia (GIM) is defined as replacement of gastric columnar cells by cells of intestinal morphology characterized by mucin-containing goblet, Paneth, and absorptive cells. These cells are easily distinguished in the gastric mucosa, because they are not present in healthy gastric mucosa. The prevalence of intestinal metaplasia in children is unknown. Furthermore, endoscopic features of gastric intestinal metaplasia in pediatric patients have been poorly defined. White opaque substance visualized by magnifying endoscopy with narrow-band imaging (M-NBI) appears to be a useful indicator of the histological diagnosis of GIM. GIM is a common finding on routine endoscopy in adults and is more frequently associated with H. pylori than in children. The frequency of GIM in children related to H. pylori-positive gastritis versus H. pylori-negative gastritis is variable. Shabib *et al.*, reported a frequency of 42% in children with pylori-positive gastritis versus 6% in children with H. pylori-negative gastritis. However, Kato *et al.*, documented no difference in the presence of intestinal metaplasia between the study groups of children with and without H. pylori-infection and Carvalho *et al.*, documented an study done in pediatric patient where Intestinal metaplasia was not found in the H. pylori-infected group. Reactive gastropathy represented the second most common cause for the occurrence of age-dependent mucosal alterations. Primary duodenogastric reflux could cause gastric mucosal lesions manifested as intestinal metaplasia histologically in children and probably an independent etiological factor. It might play a synergistic role in the pathogenesis of gastric mucosal lesions along with gastric acid and H. Pylori infection. Other causes associated with GIM in adults include: high pH and total bile acid concentration, smoking, and denervation after surgery for benign disease. The association of GIM and adenoma/dysplasia and carcinoma is rarely seen in children, primarily because the time required for these to develop takes the individual to adulthood. The malignant potential of gastrointestinal metaplasia has been shown to vary based on histologic subtype, location, and extent of mucosal involvement. Family history on initial evaluation and incomplete-type intestinal metaplasia was associated with increased risk of subsequent gastric cancer in adult patients. In adults, yearly endoscopy is justified in GIM patients with at least one of these conditions: (1) extension > 20% of the gastric surface area; (2) the presence of incomplete type IM; (3) first-degree relative of gastric cancer patients; and (4) smokers. Controversy exists regarding whether routine surveillance should be performed in individuals with GIM in low-prevalence regions such as the United States. In less intensive GIM, 2-3 years could be proposed. Additional natural history studies are required in children.

**281 A PEDIATRIC PATIENT WITH SEVERE REFRACTORY EOSINOPHILIC ESOPHAGITIS AND ATOPIC DERMATITIS FOUND TO HAVE HETEROZYGOTE BARRIER MUTATIONS IN FILAGGRIN AND KALLIKREIN-RELATED PEPTIDASE 7.** J.B.

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Rationale: Eosinophilic Esophagitis (EoE) is an inflammatory esophageal disease predominately mediated by food allergens. It is frequently associated with other atopic conditions, including atopic dermatitis (AD). Here we present a patient with severe refractory EoE and AD found to have novel mutations in proteins related to squamous epithelial barrier function. Methods: Whole exome sequence analysis was performed by GeneDx. Results: A 4-year-old African American male with severe EoE, AD, and multiple food allergies remained poorly controlled despite standard therapies plus trials of elemental diet, systemic steroids, and immunomodulators. Initial work-up revealed an IgE of 5746 IU/mL, but was otherwise normal including the CBC/differential, other immunoglobulins, lymphocyte enumeration and vaccine titers. Subsequent testing found a further elevated IgE of 46,478 IU/mL. Additional laboratory work-up was unremarkable, including T-cell mitogen proliferation; FOXP3 protein expression; bone marrow biopsy including a myeloproliferative disorder sequencing

panel (PDGFRA, PDGFRB, FGFR1, BCR/ABL1/ASS1); skin biopsy; and, sequencing of DOCK8, FOXP3, SPINK5, STAT1, STAT3 and TYK2. Whole exome sequence showed genetic mutations in two proteins associated with mucosal barrier function, filaggrin (p.R826X mutation, c.2476 C>T) and kallikrein-related peptidase 7 (KLK7)(IVS5+5, c.606+5 G>A).Conclusions: To our knowledge, this is the first report that associates these two genetic mutations. Filaggrin, part of the epidermal differentiation complex (EDC) gene, is a keratin filament aggregating protein, playing a key role in terminal differentiation of the epidermis and epithelial barrier function. Filaggrin has been associated with AD and EoE. The filaggrin mutation found in our patient, R826X has been identified in an African American patient with AD, however no reports to our knowledge of this specific mutation have been reported in EoE. KLK7 has been shown to have a role in skin disease as a desquamation related protease and has been identified in psoriasis, Netherton syndrome and AD patients. We postulate this genotype may depict a phenotype of severe and refractory EoE with AD.

**282 BURIED BUMPER SYNDROME (BBS): RARE COMPLICATION AFTER ACCIDENTAL TRACTION OF PEG TUBE.** K.

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Introduction: Percutaneous endoscopic gastrostomy (PEG) is a well established procedure in toddler who cannot be orally fed due to several medical problem such as oral aversion, primary aspiration. Buried Bumper Syndrome (BBS) is uncommon and late complication of PEG tube surgery. Incidence of BBS in pediatric patient population is between 2.4%-6.7%. It typically presents with resistance to feeds/flush, leakage, redness, and abdominal pain. Case Description: A two and half year old toddler with moderate malnutrition, oral aversion, and eosinophilic esophagitis underwent PEG tube surgery. She was tolerating 4 ounces of bolus feeds every 4 hours infused over few minutes. G tubing accidentally snagged on a toy and pulled the PEG tube. She had some abdominal pain but was able to tolerated the feeds well. Five days later, mother noted a bulge next the PEG tube site, pain with G tube feeds, and gradually increase resistance to tube feeds. She underwent fluoroscopy study at a local hospital with showed intact ostomy track with contrast entering to the stomach. Given her continued symptoms, she underwent upper endoscopy following day. Upper endoscopy showed no bolster or patent ostomy track confirming BBS. As described in the literature simultaneous pull through and extraction technique was used to remove the BBS but it was unsuccessful. New PEG tube was placed using pull through technique in the same tract. BBS was removed by extraction technique and few mini incisions on anterior abdominal wall. Discussion: BBS was first described in 1980s in adult. BBS symptoms triad consists of inability to insert, leakage around PEG, and loss of patency. It is generally thought be from tight apposition of the external bolster and internal bumper. Here we present a case of BBS after accident traction of PEG tube. Several endoscopy techniques have been described in the literature to remove BBS. In this case we report successful removal of BBS using skin incision technique after failed simultaneous pull through and extraction technique.

**289 EXOCRINE PANCREATIC INSUFFICIENCY IN CHILDREN WITH EOSINOPHILIC ESOPHAGITIS: A CASE SERIES.** P.

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Background and Aims: Eosinophilic esophagitis (EoE) is an emerging cause of dysphagia, nausea, emesis, feeding aversion, abdominal discomfort, and subsequent failure to thrive (FTT) in children. Though we continue to learn more about EoE and Exocrine pancreatic insufficiency (EPI), there are no case reports of the two occurring simultaneously. This study evaluated the association between EoE and EPI occurring in a single pediatric EoE center. Methods: Patients with EoE and EPI from our center were identified from the electronic medical record. Demographic, clinical, pathological and growth velocity histories were collected. Patients with EoE were diagnosed based on the published consensus guidelines. EPI was diagnosed based on Secretin pancreatic stimulation test. Results: Six patients with EoE and EPI were identified (2.4% of all EoE patients) over a 10-year time period (Table-1). The average age at the time of EoE diagnosis was 4.8 years (range 1-8 years) while the average age at the time of EPI diagnosis was 5.8 (range 3-10 years). Four patients are treated with diet elimination and 2 are treated with topical corticosteroids for EoE. All 6 patients suffered from FTT which resolved after pancreatic enzymes supplementation. Conclusion: We are following six patients in our practice with both EoE and EPI. Our suspicion is that these patients are at risk for being under diagnosed because poor weight gain in EoE is often times multifactorial ie: feeding aversion, poor variety in diet and vomiting. Given these overlapping symptoms, EPI may be missed. We present 6 patients with EoE who were diagnosed with EPI after management of EoE failed to

provide improvement in weight gain. After initiation of therapy with pancreatic enzymes, all of our patients had improvement in the velocity of weight gain. We suggest evaluation for EPI in EoE patients who are persistently failure to thrive despite appropriate EoE management.

**296 ESOPHAGOGASTRIC JUNCTION OUTFLOW OBSTRUCTION IN A PATIENT WITH EHLERS-DANLOS SYNDROME AFTER NISSEN FUNDOPLICATION.** V. Ramachandran, K.P. Shah, Baylor College of Medicine, Houston, Texas, UNITED STATES. F.C. Lee, D.S. Fishman, G.S. Gopalakrishna, E.H. Chiou, Pediatrics, Baylor College of Medicine, Bellaire, Texas, UNITED STATES. D.S. Fishman, G.S. Gopalakrishna, E.H. Chiou, Section of Pediatric Gastroenterology, Hepatology and Nutrition, Texas Children's Hospital, Houston, Texas, UNITED STATES. V. Mazziotti, Surgery, Baylor College of Medicine, Houston, Texas, UNITED STATES.

Background: Ehlers-Danlos syndrome (EDS) is a connective tissue disorder diagnosed in childhood that has associated gastrointestinal and neurologic dysfunction, including gastroesophageal reflux disease (GERD) in up to 58% of patients. The treatment of GERD includes lifestyle modifications, medical management and, if unimproved, surgery. The possible complications of fundoplication include gas bloat syndrome, dumping syndrome, diarrhea, dysphagia, and rarely, functional esophagogastric junction (EGJ) obstruction. Case Presentation: A 10-year old male with a past medical history of developmental delay, hypotonia and EDS type 3 was referred to the gastroenterology clinic for evaluation of persistent GERD symptoms. He presented with frequent episodes of postprandial regurgitation and vomiting. The patient also reported halitosis, but denied chest pain, dysphagia, loss of appetite, weight loss, or change in bowel movements. Esophagogastroduodenoscopy (EGD) was performed which revealed mild-to-moderate esophagitis. Histology was suggestive of GERD but without significant eosinophilic inflammation. Proton pump inhibitor therapy was initiated and reflux symptoms decreased in severity. A year later despite continued acid suppression, the patient developed new-onset episodes of cough, difficulty swallowing, vomiting, and retrosternal pain. A repeat EGD showed progression of reflux esophagitis and histology showed up to 4 eosinophils per HPF in the esophagus. 48-hour Bravo wireless esophageal pH monitoring study also confirmed abnormally elevated acid exposure in the distal esophagus. An upper GI contrast study was normal, including normal esophageal caliber, contour and peristalsis. The patient underwent laparoscopic Nissen fundoplication shortly thereafter. Three weeks status-post surgery, the patient reported increasing difficulty swallowing solids and liquids, gagging, and retching. A contrast esophagram showed an intact fundoplication wrap with a markedly narrow channel. A flexible esophagoscopy and fluoroscopically-guided balloon dilatation of esophagogastric junction up to 15 mm was subsequently performed, but dysphagia and regurgitation of food and mucous secretions persisted. Esophageal manometry revealed evidence of esophagogastric junction outflow obstruction (Figure 1). EGJ relaxation was abnormal, reflected by an elevated mean integrated relaxation pressure (IRP) at 30.6 mmHg. Mean intrabolus pressure (IBP) with swallows was also elevated at 15.4 mmHg. Esophageal body peristalsis and upper esophageal sphincter motility was normal. A repeat endoscopic balloon dilatation to 20mm was performed and the patient has had interval improvement in dysphagia. Conclusion: Gastrointestinal comorbidities, including GERD, are common findings in patients with Ehlers-Danlos syndrome. Due to the connective tissue abnormalities in these patients, they may be at increased risk of complications from fundoplication. This case highlights the use of esophageal manometry in the clinical assessment of EDS patients status-post Nissen fundoplication.

**297 HYPERTENSIVE ESOPHAGEAL PERISTALSIS - NUTCRACKER, JACKHAMMER, OR DOESN'T MATTER?.** J. Rosen, J. Cocjin, Gastroenterology, Children's Mercy Hospital, Kansas City, Missouri, UNITED STATES.

Conventional manometric criteria for hypercontractility includes distal esophageal peristaltic amplitude greater than 220mmHg (nutcracker esophagus). Hypercontractile esophagus is now defined with high resolution manometry and esophageal pressure topography using the Chicago Classification (v3.0) when the distal contractile integral (DCI) is greater than 8000 mmHg\*s\*cm for 2 swallows (jackhammer esophagus). Both definitions generally are associated with normally propagating peristaltic contractions. Although these definitions evolved over time to improve clinical relevance, there are still few cases identified and treatment options are limited. It remains unclear whether targeting treatment to symptom resolution alone, or symptom resolution plus decreased esophageal contractile force is indicated. A 17 year old male of relative adult height and weight (175cm, 61kg) presented for evaluation with chronic recurrent severe chest pain sometimes followed by forceful emesis and that did not have specific exacerbating or alleviating factors. He did not have weight loss, hematemesis, dysphagia or odynophagia. Prior diagnostic testing included normal chest x-ray, EKG, echocardiogram, esophagram, upper endoscopy with

biopsy (mid and distal esophagus), and pH/impedance study. The pH/impedance study was performed on proton-pump inhibitor therapy and one episode of chest pain occurred during the study which was not associated with reflux. Initial esophageal manometry was performed using a solid state catheter (Unisensor K103659-E-1157-D, Medical Measurement Systems software) with 5mL water swallows. Esophageal contractions had normal peristalsis, and both upper esophageal and lower esophageal sphincter resting pressure and relaxation with swallows were normal. Distal esophageal contractile amplitude of >220mmHg and DCI of 4212 mmHg\*s\*cm (max 4730 mmHg\*s\*cm) were noted (Figure 1). Gastroesophageal outflow obstruction was not identified (Figure 2). These findings are consistent with conventional manometric criteria for hypercontractile esophagus, but are normal in the Chicago Classification v3.0. Treatment was initiated with imipramine and increased to 50mg daily. Mild to moderate relief of chest pain resulted, and repeat esophageal manometry was performed using a modified protocol. Baseline esophageal contractile amplitude and DCI (4182 mmHg\*s\*cm) were unchanged from the prior study. Midazolam 0.5mg IV was given and swallows were repeated with no change in qualitative or quantitative characteristics (DCI 4016 mmHg\*s\*cm). After a recovery period, nifedipine immediate release 5mg was given by mouth and water swallows were again repeated. Distal esophageal contractile amplitude (>220mmHg) and DCI (3024 mmHg\*s\*cm) decreased as a result of nifedipine (Image 2). No chest pain occurred during the manometry testing. Daily calcium channel blocker therapy was added to imipramine with no change to baseline symptoms over 3 months. Most recently, imipramine was weaned and mirtazapine started with excellent symptom control and cessation of most chest pain and vomiting. Utilizing esophageal manometry in the evaluation of noncardiac chest pain identified a putative cause/diagnosis. However, distal esophageal body pressures were abnormal only by conventional criteria, not according to the newer, widely adopted Chicago Classification v3.0. Additionally, medications targeted at visceral hypersensitivity were moderately effective in control of symptoms with no effect on manometry, and calcium channel blockers provided no symptom relief despite decreasing esophageal contractile forces. The clinical course of this patient highlights the complexities in disorders of the esophageal body both in utilization of evolving diagnostic criteria, and identification of physiologic abnormalities that may be irrelevant to clinical care.

**300 SMALL GUT VOLVULUS IN AN ADOLESCENT.** K.L. Ahuja, E. Mok, G. Nambiar, Pediatrics, The Brooklyn Hospital Center, Brooklyn, New York, UNITED STATES. K.L. Ahuja, Pediatrics, NYU Langone Medical Center, New York, New York, UNITED STATES.

A 15 year old adolescent male with past medical history of chronic recurrent abdominal pain presented to emergency department with sudden onset of periumbilical abdominal pain and non-bilious and non-bloody vomiting. His vitals were significant for tachycardia. On physical examination he had periumbilical tenderness and hypoactive bowel sounds. Labs were unremarkable. Abdominal CT was obtained which was suggestive of small bowel volvulus. The patient underwent surgery and had Ladd's procedure with appendectomy.

**DISCUSSION** Malrotation of the midgut has usually been estimated to occur in approximately one in 500 newborns and presents within the first month of life in 64–80% of patients. However, some patients will present later, even in adulthood. Atypical symptoms such as sudden abdominal pain with (bilious) vomiting over a period of months or years are typical and may eventually lead to further diagnostics. Although there are case reports describe the presentation of malrotation in late childhood, only few cases are reported occurring this late in life. Intestinal malrotation is defined as any deviation from the normal 270° counterclockwise rotation of the midgut during embryologic development, also known as nonrotation. The failure of the normal physiological rotation of the midgut leads to various anomalies. Most commonly, the entire small bowel remains on the right side of the abdomen, while the caecum remains on the left, as a result of the absence of a ligament of Treitz. The caecum however, remains attached to the right site of the abdominal wall through persisting peritoneal fibrous bands (known as Ladd's bands). These bands often entrap the descending duodenum causing intermittent gastrointestinal obstruction. Alternatively, the intestines can be situated correctly except for a small vertical attachment of the small bowel mesentery resulting in limited fixation to the retro peritoneum. This makes the small bowel highly mobile and therefore prone to midgut volvulus. This anomaly is known as a malfixation. During the operating procedure, no anomalies indicating a complete malrotation were seen in our patient. Therefore in retrospect, a malfixation was suspected to be the cause of this specific case of small bowel volvulus. Diagnosing a malrotation or malfixation is challenging to physician. Upper gastrointestinal contrast series are the diagnostic method of choice in pediatrics. In cases of malrotation, a contrast enhanced CT scan shows an inversion of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV). **CONCLUSION** Acute abdominal pain in any

age should raise concerns of the possibility of a midgut malrotation with or without intermittent volvulus. This case highlights the demand of high index of suspicion to diagnose midgut malrotation in adolescents.

**301 TREATMENT RESISTANT FUNCTIONAL CONSTIPATION MAY BENEFIT FROM ADDITION OF SSRI.** K. Lamparyk, Pediatric Behavioral Health, Cleveland Clinic Children's, Cleveland, Ohio, UNITED STATES. L. Mahajan, L. Feinberg, Pediatric Gastroenterology, Cleveland Clinic Children's, Cleveland, Ohio, UNITED STATES.

A case series of six pediatric patients treated with addition of SSRI medication for treatment resistant functional constipation is described. All patients were previously treated by both a board certified pediatric gastroenterologist and pediatric psychologist with minimal progress for initial months and subsequently trialed on oral SSRI medications. All patients presented with a strong family history of anxiety, many of whom had also been prescribed SSRI medication as treatment of the underlying anxiety condition, and additional personal symptoms of anxious temperament. However, none of the patients had been previously diagnosed with an anxiety or any other psychiatric condition. All patients were developmentally typical. Prior medical and behavioral treatments are described, including treatment length, medications, stool consistency, and relevant patient medical history. Course of treatment of each patient is described, including bowel movement status at point of initiation and conclusion, SSRI medication and dose, treatment progression, duration of treatment, and treatment outcome. Patients generally improved in constipation and independent toileting status following the initiation of SSRI medication. No adverse outcomes were reported and all parents reported satisfaction with treatment. Results suggest that the addition of SSRI medication for this population may provide a unique benefit to treatment resistant functional constipation after standard medical and behavioral therapies have been exhausted.

**302 A CASE OF SUPERIOR MESENTERIC ARTERY SYNDROME RESULTING IN MASSIVE GASTRIC DILATION, PNEUMATOSIS, AND NECROSIS FOLLOWING CONGENITAL SCOLIOSIS SURGERY.** K.A. Moyer, J. Splawski, Pediatric Gastroenterology, Hepatology, and Nutrition, UH Rainbow Babies & Children's Hospital, Cleveland, Ohio, UNITED STATES.

Introduction: Superior mesenteric artery syndrome is caused by compression of the third portion of the duodenum as it crosses between the aorta and the superior mesenteric artery (SMA). The normal angulation between the SMA and the aorta can be disrupted by several means, including weight loss and scoliosis surgery, thus leading to a narrowed angle, and subsequent compression of the duodenum. The incidence of SMA syndrome following scoliosis surgery is approximately 1%. Symptoms include, post-prandial epigastric pain, anorexia, abdominal distension, and vomiting.

Case Presentation: An 11 year old female with history of congenital scoliosis s/p prior cervical spinal fusion developed abdominal pain and vomiting one week after T3-L3 posterior spinal fusion. She was able to tolerate a regular diet post-operatively and was discharge on POD #3; however, on POD #6, she developed vomiting, abdominal pain and distention. KUB showed a markedly distended stomach. A CT scan demonstrated a massively distended stomach, distention of the first and second portions of the duodenum, extensive gastric and duodenal pneumatosis, and extensive portal venous gas. CT findings were thought to relate to compression of the duodenum as it crossed anterior to the spine. Endoscopic evaluation revealed partial thickness necrosis of the stomach. She was made NPO, a nasogastric tube was placed for decompression, and she was started on hyperalimentation. UGI was consistent with SMA syndrome. Repeat endoscopy 1 week later showed resolution of necrosis. Following this, nasoduodenal feeds were initiated. As she continued to improve clinically, she was gradually advanced to full oral nutrition. Gastric emptying study was completed and was normal. One year later, she continues to tolerate a regular diet well with appropriate weight gain. Discussion: SMA syndrome is a well-known, though rare, complication following scoliosis surgery. Low pre-operative BMI, use of halo traction, and greater correction of spinal deformity increase the risk. The majority of patients with SMA syndrome following scoliosis surgery, can be successfully managed conservatively with nasogastric decompression and IV hydration and nutrition; though approximately 25% require surgical intervention. Regardless of whether the underlying etiology of the SMA syndrome is related to scoliosis surgery or another cause, it is incredibly rare to have gastroduodenal pneumatosis, portal venous air, or gastric necrosis, with only isolated case reports in the literature. The radiologic evidence in our case points to SMA syndrome as the cause of our patient's presentation. However, given the severity, it is possible that other factors contributed, such as vascular or neurologic compromise. Also, despite the severe nature of our case, it is important to note that she had complete resolution with conservative management alone.

**304 PELVIC FLOOR BIOFEEDBACK THERAPY IN THE TREATMENT OF FUNCTIONAL CONSTIPATION AND FECAL INCONTINENCE IN CHILDREN WITH AND WITHOUT PELVIC FLOOR DYSSYNERGIA: A CASE SERIES.** K. Osborne, D. Yacob, P. Lu, R. Chaney, M. Saps, C. Di Lorenzo, K. Vaz, Pediatric Gastroenterology, Nationwide Children's Hospital, NEW ALBANY, Ohio, UNITED STATES.

Background: Pelvic floor biofeedback therapy is an established treatment in the management of adult patients with anal dyssynergia. In children, biofeedback therapy is not readily available in most centers and the current NASPGHAN/ESPGHAN guidelines for functional constipation state that there is no evidence to support the use of biofeedback therapy in the treatment of childhood functional constipation. The objective of this case series is to illustrate that pelvic floor biofeedback therapy can be effective in the treatment of children with functional constipation. Case Presentations: We describe 3 cases of children who underwent pelvic floor biofeedback therapy for chronic constipation and retentive fecal incontinence. The first case is that of a 13 year old male with a long history of severe constipation and fecal incontinence refractory to conventional treatment. Contrast enema showed a redundant and dilated recto-sigmoid colon. Anorectal manometry showed significant pelvic floor dyssynergia and colonic manometry showed colonic dysmotility in the distal 20-30 cm. It was recommended that he start pelvic floor biofeedback therapy while plans were made for anal sphincter Botox injection and possible Malone appendicostomy. He underwent 4 sessions of biofeedback therapy while remaining on his oral stimulant medication. After completion of biofeedback therapy, he was able to sustain an adequate push and was having daily, soft bowel movements without fecal incontinence. Anal sphincter Botox injection and appendicostomy were avoided. The second case is that of a 10 year old male diagnosed with constipation and occasional fecal incontinence. Anorectal manometry was normal but he continued to have incomplete evacuation of the colon despite taking osmotic and stimulant laxatives. Pelvic floor biofeedback was offered to improve his defecation mechanism. He participated in 4 sessions of pelvic floor biofeedback therapy. Upon completion, he was able to sustain an adequate push and was passing daily, soft bowel movements without fecal incontinence. At his most recent follow up appointment, he was continuing to have daily bowel movements without the use of laxatives. The last case is that of a 9 year old female with a long history of severe constipation and fecal incontinence refractory to laxative treatment. Anorectal manometry showed pelvic floor dyssynergia. She participated in 4 sessions of pelvic floor biofeedback therapy. Upon completion, she was able to maintain an adequate and effective push. She is now having daily bowel movements without fecal incontinence with continued use of stimulant and osmotic laxative medications. Conclusion: Biofeedback therapy combined with concurrent assessment of progress and medication adjustment is a valuable tool for the treatment of functional constipation in children with functional constipation and fecal incontinence with or without pelvic floor dyssynergia. Large, randomized, controlled trials are needed to evaluate this further.

**305 DE NOVO ACTG2 GENE MUTATION IN A PATIENT WITH MEGACYSTIS MICROCOLON INTESTINAL HYPOPERISTALSIS.** K.A. Ito, T. Sugiura, S. Ito, T. Endo, T. Togawa, S. Saitoh, Pediatrics, Nagaoya City University, Nagoya, Aichi, JAPAN. Sugiura, Gamagori municipal hospital, Gamagori, JAPAN.

Introduction Megacystis microcolon intestinal hypoperistalsis (MMIHS) is a rare disorder characterized by distended nonobstructed bladder, microcolon, and decreased intestinal peristalsis. Most of the patients have to be maintained by parenteral nutrition, which often leads to life threatening complications, such as parenteral nutrition-associated liver disorder and catheter-related blood stream infections. Recently, familial and de novo *ACTG2* mutations, encoding  $\gamma$ -2 smooth muscle actin have been implicated in the etiology of familial visceral myopathy as well as MMIHS. Case presentation The patient was the first child of non-consanguineous Japanese parents. A fetal abdominal cyst was detected by ultrasound at 26 weeks of gestation. The baby was born vaginally at 38 weeks, and the birth weight was 3640 g. She was hospitalized in our NICU for the purpose of a close inspection. Megacystic bladder was confirmed by ultrasound and Computed Tomography. On day 1 of life, abdominal distention became apparent. A plain abdominal X-ray was consistent with an enlarged stomach. Although she required intermittent urinary catheterization, her abdominal symptoms were relatively mild, and she was able to take milk orally enough. She was discharged from NICU at 19 days of life. During infancy, she had several episodes of abdominal distention and vomiting which needs intravenous infusion. Although a plain abdominal X-ray always showed multiple dilated loops of small and large bowel, she was able to take foods orally. She was not dependent on TPN for two years. At a year of life, she underwent a rectal biopsy, which showed grossly normal bowel mucosa and no increased acetylcholinesterase expression. Contrast enema showed a grossly dilated large colon. At the age of 2 years, failure to thrive was apparent, so she admitted to our hospital to start

TPN. During hospital stay, she suddenly died. Autopsy revealed a massive necrotizing enterocolitis with perforation. Genetic analysis Through whole-exome sequencing, we identified a de novo mutation, c.640C>T (p.214R>C), in the gene encoding the smooth muscle gamma-2 actin, *ACTG2*. Discussion and conclusions We recommend considering genetic testing for *ACTG2* mutations in cases of fetal megacystis with sonographic findings not indicative of an obstructive etiology. Establishing the correct diagnosis of MMIHS is important to avoid unnecessary invasive procedures such as the biopsy.

**308 GASTROPARESIS IN ADOLESCENTS WITH TREATED CELIAC DISEASE IMPROVES WITH STW-5 (IBEROGAST).** L. Feinstein, A. Defelice, N. Reilly, J. Khlevner, Pediatric Gastroenterology, Columbia University, New York, New York, UNITED STATES. N. Kadakia, Pediatrics, Columbia University, New York, New York, UNITED STATES.

Celiac disease is associated with several clinical and manometric abnormalities related to altered gastrointestinal motility. Delayed gastric emptying and slow oro-cecal transit have been observed in untreated celiac patients and thought to be related to altered neuroimmunomodulation, hormonal deregulation and unabsorbed starches and fats in the intestine. Similarly, several studies have demonstrated an association between celiac disease and functional dyspepsia, and some authors have suggested routine screening for celiac disease in patients with refractory dyspeptic symptoms. Data suggests that gastroparesis and functional dyspepsia improve following treatment of celiac disease with a gluten-free diet (GFD). A previous study examining gastric emptying rates in children with celiac disease demonstrated an association between untreated celiac disease and delayed gastric emptying which completely normalized with introduction of a GFD. Several studies have shown STW-5 (Iberogast), an herbal preparation which consists of nine plant extracts, to be an effective treatment option for functional dyspepsia but evidence of an effect on gastric emptying is limited. In this case series we present four pediatric patients with confirmed celiac disease that developed gastroparesis despite adherence to a strict GFD and normalized celiac antibodies. These patients were adolescent females aged 12-16 years old who developed symptoms of nausea, vomiting, bloating and early satiety. One patient had these symptoms at the time of celiac disease diagnosis while the others developed symptoms 1-3 years after diagnosis. Tissue transglutaminase Ig A which was followed as a marker of adherence to GFD normalized in all patients despite worsening dyspeptic symptoms. One patient underwent a repeat duodenal biopsy which was normal. All patients had a 4hrs solid gastric emptying scintigraphy (GES) and were diagnosed with gastroparesis based on established criteria. Erythromycin and cyproheptadine were trialed without relief. Two patients underwent endoscopic intrapyloric botulinum toxin injection without clinical improvement. All patients were eventually trialed on STW-5 with improvement in dyspepsia. Two patients had a repeat GES on treatment with STW-5; one with normalized gastric emptying and the other showed rapid gastric emptying time. Current data suggesting that gastroparesis and functional dyspepsia in individuals with celiac disease resolve on a GFD may not apply to all patients. In this case series we present four adolescents with celiac disease and gastrointestinal dysmotility whose symptoms persisted despite GFD and normal celiac antibodies. All patients failed therapy with pro-kinetic agents but did show clinical response with the addition of STW-5. This case series highlights that dyspepsia and gastroparesis can develop in patients with celiac disease despite adherence to dietary treatment which to our knowledge has not been reported before. In addition STW-5 should be considered for the treatment of gastroparesis and functional dyspepsia in patients with celiac disease.

**313 ACQUIRED MYOPATHIC INTESTINAL PSEUDO-OBSTRUCTION DUE TO AUTOIMUNE ENTERIC LEIOMYOSITIS IN A PREVIOUSLY HEALTHY 10-YEAR-OLD MALE.** Y.A. Mattos, M. Bettiol Nogueira, Gastroenterology, Hospital da Criança de Brasília, Brasília, BRAZIL. H. Bender Kohnert Seidler, Pathology, Hospital de Base do Distrito Federal, Brasília, BRAZIL.

A 10-year-old boy with symptoms of chronic intestinal pseudo-obstruction (CIPO) was hospitalized five times during a period of 6 months because of abdominal distension, vomiting and weight loss. Surgery was performed in the second hospitalization, but no intestinal resection was necessary, as there was not any sign of mechanical obstruction; a meso torsion was found and solved. The other admissions were treated conservatively, with nasogastric tube (for feeding or gastric drainage) and parenteral nutrition. Abdominal enterotomography as well as all X-ray performed over the time showed signs of intestinal obstruction as distension of intestinal loops, hydro-aero levels and lack of gas in the rectum. The child progressed with pleural and pericardial effusion, ascites and persistent fever. Therefore, it was decided to start methylprednisolone besides antibiotic therapy, with dramatic improvement of patient's clinical condition. Surgical full thickness intestinal biopsies were performed from

jejunum, ileum, transverse colon, cecum and sigmoid which demonstrated visceral myopathy due to severe fibrosis present on all muscular layers. Based on the results of laboratory tests (homogenous nuclear FAN 1:640, nuclear dotted 1:320, anti-DNA double positive helix, hypocomplementemia, nephrotic proteinuria) and clinical criteria (pericardial and pleural effusions, ascites, alopecia, skin rash, photosensitivity) the patient was given the diagnosis of Juvenile Systemic Erythematous Lupus (JSEL). CIPO secondary to JSEL is a rare syndrome described recently. In this case, it was a more difficult diagnosis because autoimmune enteric leiomyositis was found in the intestinal biopsies rather than any signs of vasculitis.

**317 MODIFIED SPECIFIC CARBOHYDRATE DIET IN COMBINATION WITH UZTEKINUMAB TO TREAT A CASE OF SEVERE CROHN'S DISEASE AFTER FAILURE OF CONVENTIONAL THERAPY.** C. Jaramishian, K. Coleman, C.S. Huang, Pediatric Gastroenterology, Valley Children's Hospital, Madera, California, UNITED STATES.

Severe pediatric Crohn's disease remains a challenge to the clinician when all conventional treatment seems to fail. Exclusive enteral nutrition (EEN) has been shown to induce and maintain remission in patients with Crohn's disease. However, strict adherence to a formula based diet is difficult to sustain due to interruption of normal eating habits. Recent efforts to develop an alternative non-pharmacological treatment for Crohn's disease show that the specific carbohydrate diet (SCD) has comparable effects to that of EEN. Here we report the case of a 21-year-old female with a 12-year history of severe, early onset Crohn's disease with a significant stricture at the terminal ileum associated to fistulizing, perianal disease with nutritional failure and severe erythema nodosum. After extensive combination therapy trials including systemic and localized steroids, Azathioprine, Infliximab, Adalimumab, Methotrexate and exclusive enteral therapy, she was placed on a modified SCD three months post-induction of Ustekinumab. Prior enteral nutrition in combination with anti-TNF therapy failed to induce remission in the patient. Following the introduction of this modified diet, she experienced clinical remission for the first time in 7 years with improvement in laboratory markers, including Hemoglobin, erythrocyte sedimentation rate, C-reactive protein and fecal calprotectin. The patients follow up colonoscopy demonstrated significant mucosal and terminal ileum stricture improvement. Our findings warrant further investigation of the SCD in combination with a biologic in the treatment of IBD.

**318 TAKAYASU'S ARTERITIS IN PEDIATRIC PATIENTS WITH INFLAMMATORY BOWEL DISEASE.** C. Scherer, A.F. Porto, D. Pashankar, Pediatric Gastroenterology and Hepatology, Yale, New Haven, Connecticut, UNITED STATES.

Introduction: Takayasu's arteritis (TA) is a rare nonspecific chronic inflammatory condition affecting the aorta, main aortic branches, pulmonary and coronary arteries. The inflammation is characterized as a granulomatous vasculitis. TA and IBD are two chronic inflammatory diseases, that have been previously reported to co-exist, both respond to anti-TNF-alpha treatment, and underlying pathophysiology may be related. We present two cases of pediatric patients diagnosed with TA in the context of known IBD. Case Reports: A 13-year-old boy with history of Crohn disease, presented with a 10-day history of abdominal pain. His past medical history is significant for type-1 diabetes mellitus, autism, asthma, and recent course of antibiotics for *Clostridium difficile* infection. Laboratory testing on admission was significant for WBC  $7 \times 10^3/\mu\text{L}$ , Hgb 11.9 g/dL, ESR 120 mm/hr, CRP 142.1 mg/dL, and albumin 4.6 g/dL. A CT scan of the abdomen showed an unusual wall thickening of the abdominal aorta. Magnetic resonance angiogram of the chest and abdomen revealed wall thickening of the abdominal aorta and distal descending aorta. He was diagnosed with TA, and treatment with infliximab 5 mg/kg and methotrexate 25 mg weekly was initiated. Abdominal pain improved. Inflammatory markers significantly improved, ESR 33 mm/hr and CRP 1.7 mg/dL, within 7 days of treatment. He has since remained on infliximab 5 mg/kg every 4 weeks and methotrexate 25 mg weekly, and has done well. A 12-year-old girl with history of Von Willebrand disease and ulcerative colitis, presented to the ED with 4 days of left upper extremity numbness and tingling, and 1 day of altered mental status. MR and CT imaging of the head, neck, and chest showed findings suggestive of TA with large vessel vasculitis of the proximal left subclavian artery, in addition to a left vertebral artery thrombus. She was treated for TA with methylprednisolone 1 g for 3 days, followed by a steroid taper. Infliximab 5 mg/kg and methotrexate 18 mg weekly were started. She was treated with anti-coagulants for the thrombus. Her numbness improved. The tingling and thrombus resolved. Due to concern of potential persistent vasculitis while receiving infliximab, medications were changed to azathioprine and tocilizumab, methotrexate was continued. Tocilizumab was later discontinued due to neutropenia. Four years after diagnosis of TA she remains on azathioprine and methotrexate and has been well controlled. Discussion: IBD and TA co-occur more frequently than expected. Review of previous case reports and series suggest a frequency of IBD in 6-9% of patients with TA. A diagnosis of

IBD typically precedes the development of arteritis and both diseases tend to respond well to anti-TNF-alpha therapy. It is unknown if the pathophysiology of IBD may predispose patients to developing TA. Gastroenterologists should be aware of this relationship and have a low threshold for pursuing evaluation of TA when patients present with abnormal blood pressures, inflammatory markers elevated more than would be explained by current IBD symptoms, unexplained pain or weight loss, and neurologic symptoms including changes in mental status.

**319 CROHN'S DISEASE AND NEPHROTIC SYNDROME: AN ATYPICAL CAUSE FOR HYPOALBUMINEMIA.** D. Rosen, Pediatric Gastroenterology, Yale University, New Haven, Connecticut, UNITED STATES. Antala, Pediatrics, Yale University, New Haven, Connecticut, UNITED STATES.

Introduction: While cases of medication-induced nephrotic syndrome have been described in IBD patients, the presence of Crohn's disease and primary nephrotic syndrome has not been previously reported. Here we report on an adolescent patient with an atypical presentation of hypoalbuminemia. Case report: A 13-year-old male with a history of steroid-sensitive nephrotic syndrome diagnosed at age 3 presented with weight loss, abdominal pain and guaiac positive stool. EGD and colonoscopy confirmed a diagnosis of Crohn's disease. The family was resistant to medication and he was lost to follow up until 2 years later. At that time he was reportedly asymptomatic except for poor growth. Calprotectin was elevated at 753.7 mcg/g, erythrocyte sedimentation rate (ESR) was mildly elevated at 34 mm/hr, and albumin was low at 2.6 g/dL. Repeat EGD showed mild duodenitis. Colonoscopy biopsies were significant for moderate chronic active colitis of the cecum and mild chronic inflammation of the terminal ileum. MRE showed thickening of the distal and terminal ileum. Due to concern for growth failure the family agreed to infliximab treatment. After receiving the standard 3 induction doses, albumin decreased to 1.8 g/dL. Bowel movements were formed, 1-2 times per day and he was not exhibiting any signs of peripheral edema. Budesonide was added and repeat labs 2 weeks later showed a further decrease in albumin to 1.6 g/dL. Infliximab level at 14 weeks showed a serum infliximab concentration of 0.0 ug/mL with no antibodies detected. At that time a urinalysis showed 4+ protein, and he was started on a 4-week prednisone course for suspected relapse of his nephrotic syndrome. At his 8 week infusion, albumin improved to 3.5 g/dL and ESR decreased from 53 mm/hr to 2 mm/hr. Discussion: Renal involvement in inflammatory bowel disease can be categorized as drug-induced or secondary to underlying inflammation. Granulomatous interstitial nephritis is a rare extra-intestinal manifestation of Crohn's disease that may present with impaired renal function. Renal amyloidosis can also be seen in Crohn's disease and may result in nephrotic syndrome. Mesalamine is a known nephrotoxic agent that more commonly leads to interstitial nephritis, although cases of mesalamine-induced nephrotic syndrome have been reported. Additionally, several case reports in both adult and pediatric patients have described nephrotic syndrome associated with adalimumab therapy. This case is unique in that the underlying nephrotic syndrome was seemingly not related to his Crohn's disease or to drug toxicity. Hypoalbuminemia is a common finding in IBD patients, typically in the setting of small bowel disease or colitis with excessive stool losses. This patient did not have extensive small bowel disease or significant colonic disease, which is why other routes of protein loss were investigated. While the steroid course for nephrotic syndrome likely affected his Crohn's disease, it is encouraging that he is off steroids and continues to improve. Conclusion: IBD and nephrotic syndrome can co-exist. In the absence of significant small bowel or colonic disease, other causes for hypoalbuminemia such as renal losses should be considered.

**323 TAILORING ACCELERATED INFlixIMAB INDUCTION TO REDUCE THE NEED FOR EARLY COLECTOMY IN A PATIENT WITH ACUTE SEVERE ULCERATIVE COLITIS.** J. Larson, C. Forges-Voigt, A. Fifi, E. Rivera-Rivera, A. Langshaw, Pediatric gastroenterology, hepatology, and nutrition, Jackson Memorial Hospital, Miami, Florida, UNITED STATES. Fifi, E. Rivera-Rivera, A. Langshaw, Pediatric gastroenterology, hepatology, and nutrition, University of Miami, Miami, Florida, UNITED STATES.

Acute severe ulcerative colitis (ASUC) is a medical emergency and severe disease is defined by a score of 65 or more based on the Pediatric Ulcerative Colitis Activity Index (PUCAI). This index assists clinicians in determining the need for second-line medical therapy or early colectomy. Intravenous (IV) corticosteroids at 1.5mg/kg (maximum 60mg daily) have been the mainstay of management for ASUC for the past 40 years,<sup>1</sup> but approximately one-third of patients fail to respond.<sup>2</sup> For non-responders rescue medical therapy includes infliximab or a calcineurin inhibitor. The standard infliximab induction regimen of 5mg/kg/dose at 0, 2, and 6 weeks, is based on the original ACT studies in adults which excluded hospitalized patients with steroid-

refractory ASUC.<sup>3</sup> However, ASUC is associated with higher circulating levels of tumor necrosis factor and patient's with hypoalbuminemia, increased CRP, male, and high BMI are known to have higher clearance rates of infliximab, which poses a significant clinical challenge.<sup>4</sup> Case Description: We describe the case of a 15 year-old female diagnosed with UC at outside facility two months prior to admission. Pathology confirmed moderate active chronic colitis in the rectum and sigmoid colon, and mild activity in the cecum. She was started on budesonide and mesalamine and followed up as outpatient. However, two months later she was admitted to an outside hospital with fever, hemoglobin of 8.3dl/L, and >8 loose, bloody stools per day. She was started on hydrocortisone 40mg IV every 6 hours with minimal response. Nine days after her admission the first dose of infliximab at 5mg/kg was given. She continued with multiple episodes of loose, bloody stools and a second dose of infliximab at 10mg/kg was given on day 11. The patient was transferred to our institution on day 14, when she developed fever, increased CRP, ESR, and PUCAI score of 70. Sigmoidoscopy revealed erosive, ulcerating, acute and chronic moderate to severe colitis in the sigmoid colon and rectum. Colonic biopsy negative for CMV, EBV, HSV, and adenovirus and stool negative for C. diff toxin. Surgery was consulted for possible colectomy and infliximab 10mg/kg was given on days 21 and 28. Prior to the fourth dose, no antibodies were present to infliximab and level was >34ug/ml. The PUCAI score decreased by 30 points after the fourth dose. The fifth dose was given on day 44 and sixth dose on day 72 at which time patient was in clinical remission with a PUCAI score of 5. Discussion: Algorithms for suggested management of pediatric ASUC exist and guide clinicians. This acts as a guide only and does not replace clinical assessment of individual patients. The ACT studies established the current infliximab regimen in UC which consists of both the induction dose of 5mg/kg at 0, 2, and 6 weeks, and maintenance dose of 5mg/kg every 8 weeks and did not find significant difference in the efficacy when infliximab doses were increased to 10mg/kg.<sup>5</sup> However, hospitalized patients with steroid-refractory ASUC were excluded from these studies. Recent literature in hospitalized adult ASUC patients found that the use of accelerated infliximab induction reduced the need for early colectomy.<sup>6</sup> In our patient's case she had hypoalbuminemia and increased CRP. The third induction dose was given on day 21 due to lack of clinical improvement and overall severity of her clinical condition. The use of salvage therapy with infliximab in pediatric ASUC, including dosing and timing of these infusions are underreported. To our knowledge, this is the first case report in the literature of such an accelerated induction regimen, 3 doses in 21 days, and 6 doses within 72 days. Rescue therapy in this patient led to a positive outcome without colectomy. By 3 months her PUCAI score was zero and to date, a year later, she remains in clinical remission.

**326 PERSISTENT LARGE VESSEL VASCULITIS IN A CHILD WITH INFLAMMATORY BOWEL DISEASE.** J. Tung, Pediatrics, Mayo Clinic, Rochester, Minnesota, UNITED STATES. Ulrich, Medical School, University of Minnesota, Minneapolis, Minnesota, UNITED STATES.

A 15 yo female was diagnosed 2 years prior with indeterminate colitis (gastric erosions and pancolitis). She had partial improvement with mesalamine. Twenty-three months after diagnosis, she was started on 6-mercaptopurine (6-MP) after experiencing bloody diarrhea. Repeat esophagogastroduodenoscopy was normal, while a colonoscopy again demonstrated pancolitis. Several weeks after initiation of 6-MP, she developed bilateral suppurative tonsillitis which was treated with antibiotics and Decadron. Testing for streptococcus, influenza and Epstein Barr viruses was negative. Budesonide was started and 6-MP was held, her gastrointestinal symptoms worsened. At presentation, she had mild abdominal pain and arthralgias (sPCDAI 65). Heart rate was 96 bpm, and blood pressure 127/68 mmHg. She was pale but in no acute distress. No murmurs were present. Abdomen was soft, and without masses or organomegaly. Peripheral pulses were intact. Bloodwork revealed a normocytic anemia (Hemoglobin 10.2 gm/dL), normal white blood count and platelet count, c-reactive protein 65 mg/L (normal <8), and erythrocyte sedimentation rate 52 mm/hr. Stool for Clostridium difficile was negative. MR enterography was negative for small bowel involvement, but revealed diffuse circumferential thickening of the abdominal aorta wall, extending from the bifurcation to the thoracic aorta. MR angiogram (MRA) of the chest demonstrated additional involvement of the aortic arch, right brachiocephalic artery, and left subclavian artery consistent with large vessel vasculitis (Takayasu's arteritis, TAK). Review of gastrointestinal biopsies did not show evidence of small vessel vasculitis. Additional labwork included a positive neutrophil specific antibody, equivocal ASCA IgA, mildly elevated ANA (1.5), negative myeloperoxidase antibody and negative proteinase. Clinical remission was induced with infliximab and 3 months of corticosteroids. Repeat MRA of the chest and abdomen were stable. Echocardiogram was normal. Methotrexate was added, and infliximab levels were optimized. At 10 month follow-up, she remained in clinical remission, and repeat MRA showed improvement. At 17 months, she

was switched to adalimumab due to an infusion reaction. At the time, she was in clinical remission but had mild endoscopic pancolitis. MRA did not show further improvement, nor was there worsening at the time of an IBD flare 23 months after infliximab initiation. Discussion The literature on vasculitis in patients with inflammatory bowel disease (IBD) is limited, particularly in the pediatric population. Takayasu arteritis (TAK) is most frequently associated with IBD, but granulomatosis with polyangiitis, isolated cutaneous vasculitis, and central nervous system vasculitis have also been reported. Drugs used to treat patients with IBD and vasculitis include 5-aminosalicylates, corticosteroids, methotrexate, azathioprine and anti-tumor necrosis factor alpha agents (anti-TNF). Most patients with TAK treated with anti-TNF agents reportedly experience remission of both their vasculitis and IBD. However, while this patient showed radiologic improvement along with clinical remission on anti-TNF therapy, resolution was not achieved even with subsequent combination therapy. In addition, there was no radiologic worsening during a flare of Crohn's disease. It is unknown to what extent the disease activity of TAK parallels gastrointestinal activity.

**335 ISCHEMIC COLITIS IN A TEENAGE GIRL.** K. Whaley, K.I. El-Chammas, L. Denson, K.E. Bove, S. Pentiuk, R. Sheridan, C. Socec, Gastroenterology, Hematology and Nutrition, Cincinnati Children's Hospital and Medical Center, Cincinnati, Ohio, UNITED STATES.

Background: Thrombotic microangiopathy (TMA) is a pathological finding that can be seen in both atypical hemolytic uremic syndrome (aHUS), as well as catastrophic antiphospholipid syndrome (CAPS). The alternative complement gene mutation predisposes patients to aHUS, but a trigger is still required; triggers for aHUS are currently not well understood, but lead to significant complications. Treatment with Eculizumab blocks terminal C5 and the activation of the alternative complement pathway. For patients with both aHUS and CAPS, intestinal involvement is a rare but potentially devastating complication. Our case highlights the significant morbidity that may occur and discusses the need for further knowledge and research to prevent these sequelae. Case Presentation: A 16 year old previously healthy female presented to the emergency room with acute pharyngitis, painful lymphadenopathy, fevers and bloody diarrhea. Her symptoms began three days prior to presentation with pharyngitis and fevers. She then developed abdominal pain and bloody diarrhea the day of presentation. She denied recent travel, food or animal exposures, or sick contacts. Family history is pertinent for Ulcerative Colitis and Anti-Phospholipid Syndrome in the mother. At presentation, her infectious work-up, including stool studies, was negative. Her labs were notable for thrombocytopenia, microcytic anemia and a coagulopathy with an elevated PT and aPTT, and IV antibiotics were initiated. One day after presentation, her pain and diarrhea worsened and she developed vulvar edema and necrosis. She was started on IV methylprednisolone. However, after a few days, she developed acute kidney injury and required continuous renal replacement therapy for 48 hours in the pediatric intensive care unit. She underwent an upper endoscopy and flexible sigmoidoscopy with duodenal and rectosigmoid biopsies notable for acute on chronic intestinal ischemia with transmural thrombi. She was treated with Eculizumab and began to improve. She received Eculizumab every 5 days with levels greater than 100 and CH50 levels less than 50. After three weeks of treatment, she developed acute onset of abdominal pain and vomiting. Abdominal X-ray was notable for pneumoperitoneum, so she was taken to the operative room and found to have necrotic colon with a perforation in the transverse colon. She underwent a subtotal colectomy with ileostomy formation and preservation of her sigmoid colon. Biopsies from her colon were notable for transmural coagulation necrosis consistent with ischemic injury. She was also found to have extensive vasculopathy with arterial and venous thromboses in both her large and small vessels. Labs were notable for an abnormal anti-phospholipid panel with a moderately positive StaClot LA test of 25.4 seconds and a negative DRVVT lupus anticoagulant test. Paroxysmal nocturnal hemoglobinuria testing was negative. She was redosed with Eculizumab and was started on a heparin drip and Plaquenel. She is currently being maintained on Lovenox and Eculizumab with slow advancements in feeding given her recent significant ischemic injury. Discussion: Our patient is a rare case of ischemic colitis with biopsies consistent with both TMA, as well as large vessel thrombosis. While treated with Eculizumab, she developed significant morbidity related to her vasculopathy. Our case highlights the need for further knowledge and research in pediatric patients with intestinal involvement in TMA given the high risk of poor outcomes.

**337 COMBINATION BROAD-SPECTRUM ANTIBIOTICS IMPROVES DISEASE ACTIVITY IN CHILDREN WITH STEROID DEPENDENT OR RESISTANT ULCERATIVE COLITIS.** K. Kordy, Division of Pediatric Gastroenterology, Children's Hospital of Los Angeles, Los Angeles, California, UNITED STATES. Romeo, Department of Paediatrics, University of

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**Background:** Intestinal microbiota influences mucosal inflammation in Inflammatory Bowel Disease (IBD) (Flavell, 2014). Antibiotics have been effectively utilized in adult patients during disease flares (Sato, 2009 and 2010; Kato, 2011; Ledder, 2014). In a small cohort, half of children with moderate-severe refractory Ulcerative Colitis (UC) achieved remission with a regimen of 3-4 antibiotics (Turner, 2014). We report our experience with oral antibiotics in symptomatic children with UC. **Methods:** A retrospective review was conducted on children with histology-confirmed, steroid dependent or resistant UC treated with oral combination of antibiotics based on the Turner study of: amoxicillin (50 mg/kg divided TID, max 500 mg TID), metronidazole (5 mg/kg TID, max 250 mg TID), doxycycline (2 mg/kg BID, max 100 mg BID) for ages over 7, and vancomycin (250 mg QID) if hospitalized. In some, fluconazole (200 mg qd) was added per clinician discretion. Allergenic drug was replaced with gentamicin (2.5 mg/kg TID). Intended duration was 2-3 weeks pending PUCAI response at end of week 2. Concomitant medications continued per standard of care. Disease activity (PUCAI) and available labs were evaluated at baseline, during and post antibiotics up to 6 months. Primary endpoint is a change of >20 PUCAI points at end of therapy. Secondary endpoints include end of therapy inflammatory markers (CRP, ESR, calprotectin), hemoglobin, albumin, and remission (PUCAI  $\leq$ 10). Mixed effects regression was applied to outcome parameters as a function of time compared to pre-treatment baseline while controlling for the individual as a random effect. All p-values were adjusted for multiple comparisons using the Benjamini-Hochberg method. **Results:** Eight steroid dependent or refractory UC patients ages 8-20 years (mean 14.6), 38% female, most with extensive colitis (n=7), and history of poor response to multiple immunosuppressants including biologics, had a mean pretreatment PUCAI of 60 (25-80). Duration of antibiotics ranged from 6-84 days (mean 25) per provider's judgement. PUCAI significantly declined at all timepoints (p<0.001) with half achieving a PUCAI drop >20 at end of antibiotics (mean 37.5), while 3 (37.5%) achieved remission (Figure 1). One patient with severe refractory disease did not respond after 7 days (PUCAI 75 from 80), and subsequently underwent colectomy the following week. One steroid-dependent, infliximab primary non-responder relapsed within 48 hours post-antibiotics after initially responding to 21 days of antibiotics (PUCAI 35 from 65) with subsequent lowering of maintenance steroids. A second patient who achieved remission (PUCAI 75 to 10) with 6 days of therapy developed moderate disease a week after antibiotic completion followed by fecal transplantation. Two patients maintained remission (PUCAI 0) at 2 and 6 months post-therapy, one of whom was able to wean off steroids and de-escalate therapy. Two patients had PUCAI of 20-25 (from 60) at 2 months. Though decreased, there were no significant changes in CRP (N=5, p=0.70, p=0.28), ESR (p=0.91, p=0.29), and calprotectin (N=3, p=0.15, p=0.22) at 2 and 6 months. Albumin was significantly increased at 6 but not 2 months post-therapy (p=0.02, p=0.09) (Figure 2). There were no adverse events related to the antibiotics. **Conclusion:** Overall, 7 out of 8 steroid dependent or resistant UC patients with active disease treated with antibiotics had a clinically significant reduction of PUCAI scores of more than 20 points (p<0.001), inducing remission in 38% by end of therapy. Albumin increased post-therapy (p=0.02) possibly due to improved nutrition and decreased gut leakiness. No other parameters had significant change likely limited by sparse sampling and small size. Two patients relapsed within a week post-antibiotics, though were confounded by timing of steroid wean and recent fecal transplantation. Brief duration of broad-spectrum antibiotic coverage may be a safe and beneficial therapeutic option for short-term control of active disease in refractory pediatric UC patients, but larger sample sizes and long term follow up are needed. Future studies will longitudinally catalogue intestinal microbial succession pre- to post-therapy and during disease recurrence to elucidate microbial responses to non-targeted antibiotic therapy.

**341 HIGH DOSE INFLIXIMAB FOR PEDIATRIC CROHN'S DISEASE: A CASE SERIES FROM A SINGLE TERTIARY HOSPITAL.** L. Hart, S. Forget, E. Seidman, Dept Pediatrics, Division of Gastroenterology, McGill University, Montreal, Quebec, CANADAM. Chavannes, Dept Pediatrics, Division of Gastroenterology, University of British Columbia, Vancouver, British Columbia, CANADAS. Restellini, E. Seidman, Dept Medicine, Division of Gastroenterology, McGill University, Montreal, Quebec, CANADA.

**INTRODUCTION:** Infliximab (IFX) is indicated for induction and maintenance of remission in pediatric patients with moderate to severe active inflammatory bowel disease. Escalation of IFX dose has been shown to be a safe and effective to recapture response in the presence of persistently active disease or loss of response. However, there is no pediatric data on effectiveness and safety of high doses of this medication. **AIMS:** We aimed to assess the safety and effectiveness of high dose (HD) IFX (15mg/kg every 4 weeks) for pediatric Crohn's disease (CD). **METHODS:** We present a case series. Data was collected from the Montreal Children's Hospital from 2015-2017. Inclusion criteria

were pediatric patients with CD who received IFX at a dose of 15mg/kg every 4 weeks. Retrospective chart reviews were performed to obtain data on duration of high dose IFX, IFX drug levels, changes in clinical status (using the abbreviated pediatric Crohn's disease activity index) and biochemical markers (hemoglobin, albumin, C reactive protein (CRP) and fecal calprotectin (FC)). We also collected data on adverse events that occurred during HD IFX therapy. RESULTS: Nine patients met the inclusion criteria: 8 male, 1 female. All patients had luminal non-fistulizing CD. 33% were steroid dependent when starting IFX. All patients were on concomitant therapy at the start of HD IFX (5 with methotrexate, 1 with Imuran, 2 with oral steroids; 3 additionally with rectal therapy). Reasons for HD IFX included persistent symptoms and/or low IFX level with high FC. FC at start of HD IFX ranged from 275 to greater than 1800 ug/g. All non-responders continued to have elevated FC (>700 ug/g). Median time (IQR) on HD IFX was 7mo (8.55). Median (IQR) IFX trough level pre-HD was 4.65 (8.1) ug/mL and at last dose was 17.87 (18.4) ug/mL, with all responders showing an increase in their IFX levels. All anti-IFX antibodies were negative. Median (IQR) CRP pre-HD was 5.5 (7.6) mg/L and at last dose was 5.1 (9.3) mg/L, with all responders who initially had elevated CRP showed a normalization of this value. 3-month durability of response of HD IFX is 75% and 6-month durability is 60%. 55% remained on HD IFX, while 45% switched to another biologic treatment (ustekinumab n=3, adalimumab n= 1). 10 infectious events were reported in 6 patients. Gastrointestinal infections were most common. 1 patient required hospitalization for salmonella bacteremia (after travelling overseas). CONCLUSION: In this case series, HD IFX (15mg/kg every 4 weeks) allowed recapture of response in 55% of the patients. HD IFX could therefore be considered as a feasible option for treatment in patients with Crohn's disease who have failed standard and regular optimization dosing. However several mild adverse events, notably gastrointestinal infections were reported. Further large studies are therefore warranted to better identify which patients would benefit most from this therapeutic strategy.

**342** *SHORT BOWEL SYNDROME ASSOCIATED WITH INFLAMMATORY BOWEL DISEASE.* L. Bauman, M. Farrell, S. Kocoshis, Gastroenterology, Hepatology, and Nutrition, Cincinnati Children's Medical Center, Cincinnati, Ohio, UNITED STATES.

Short Bowel Syndrome (SBS) causes intestinal failure in children and has heterogeneous etiologies. There is a small body of literature describing an association between SBS and subsequent inflammatory Bowel Disease (IBD) in pediatric patients. IBD is more commonly diagnosed in older children without underlying SBS. Here we describe 4 cases of SBS requiring surgical management who later developed intestinal inflammation consistent with IBD. Three eventually required biologic therapy to control their IBD. In conclusion, although the biologic mechanism of SBS with later IBD is not well-delineated, describing this association in more detail is important to heighten awareness in providers and to elucidate the pathophysiologic connection. Case 1DL had extensive small intestinal resection secondary to multiple intestinal atresias, with 100 cm of bowel distal to the ligament of Treitz after resection. He developed a perianal abscess at age 9 after intestinal rehabilitation, and went on to be diagnosed with Crohn's Disease (CD) after colonoscopy with patchy ileal inflammation and granulomas noted on histopathology. He initially was placed on colazol and budesonide. His disease is currently under good control with adalimumab. Case 2MP had moderate resection of the small bowel and extensive resection of his colon secondary to necrotizing enterocolitis, with jejunosigmoid anastomosis. At the age of 11 he developed deep linear ulcerations in the terminal ileum and remaining colon, and was diagnosed with Crohn's Disease. His disease was well controlled on sulfasalazine at follow up at age 19. Case 3NK was diagnosed with Hirschsprung disease as an infant. He underwent colectomy and ceco-anal pull through. He also was diagnosed with hypothyroidism and eosinophilic esophagitis. At the age of 13 he developed frequent soiling and growth failure, and was noted to have persistent ileal and cecal inflammation. He was started on sulfasalazine, but due to persistent anemia was changed to infliximab with monthly iron infusions. Case 4KH was a preterm infant who developed necrotizing enterocolitis requiring ileocolonic resection. Initially she required parenteral nutrition for several years but was able to transition to complete enteral feeds. At age 20 she developed diarrhea, weight loss, and had endoscopy diagnosing CD. She was noncompliant with oral medications and required infliximab to control her disease. She transitioned to adult care, and again required parenteral nutrition for her poor nutrition and weight loss. She passed away at age 27 from complications of her malnutrition.

**344** *MESENTERIC VASCULITIS: AN UNUSUAL MIMICKER OF INFLAMMATORY BOWEL DISEASE.* L. Thorp, J. Fridge, I. Kalampokis, J. Hanson, Pediatrics, UNM, Albuquerque, New Mexico, UNITED STATES.

**INTRODUCTION**Inflammatory bowel disease (IBD) may present with a myriad of nonspecific symptoms. There are many mimickers of IBD and it is important to consider a broad differential diagnosis.**CASE DESCRIPTION**A 15-year-old Chinese female with recently diagnosed autoimmune hemolytic anemia (AIHA) presented to our Emergency Department with 2 months of intermittent abdominal pain, weight loss, emesis and joint pain. On physical exam, she had symmetric polyarthritis of her hand/finger joints, hyperactive bowel sounds and diffuse abdominal tenderness without guarding or rebound. Fecal occult blood testing was positive. Abdominal radiographs and sonography showed hepatosplenomegaly but no obstruction. During her inpatient stay, she required multiple blood transfusions for her anemia and tested positive for cold agglutinins.Colonoscopy revealed ulceration in the sigmoid colon. Pathology identified chronic colitis with cytomegalovirus (CMV) cytopathic effect confirmed by immunohistochemistry. Computed tomography (CT) showed focal areas of inflammation and strictures involving the distal descending colon and ileum, suggestive of Crohn's disease (CD). Abdominal CT angiography was normal. After 7 days of ganciclovir treatment, repeat colonoscopy showed CMV resolution with ongoing ulceration. The IBD panel was positive for CD. Based on the presumptive diagnosis of CD, she started infliximab therapy. 10 days later she developed acute abdomen. Abdominal radiographs showed small bowel obstruction. She had small bowel resection and end ileostomy following exploratory laparotomy. Histopathological examination of the resected ileum revealed eosinophilic granulomatous vasculitis of small and medium sized mesenteric vessels (Fig. 1). She had elevated fibrinogen, von Willebrand factor and D-dimers; ANCA was negative. She had no peripheral eosinophilia or elevated IgE.Ophthalmology and neurology consults ruled out involvement of other organ systems typically involved in eosinophilic granulomatosis with polyangiitis. CT imaging of her chest was normal. IV methylprednisolone and daily oral prednisolone were initiated and her clinical status improved rapidly; she required no more blood transfusions and her inflammatory markers started trending down. Intravenous immune globulin (IVIG) was used as a steroid-sparing agent. She developed intra-abdominal abscesses post-operatively; episodes of septic shock were handled in the intensive care unit with pressors and broad-spectrum antibiotics. Following a 70 day hospitalization, she was discharged on IV ceftriaxone and metronidazole, weekly methylprednisolone pulses, and daily oral prednisolone. She will start IV cyclophosphamide following CT documentation of intra-abdominal abscess resolution.**DISCUSSION**~10-15% of patients with IBD have atypical features. In our case, histopathology of a superficial biopsy sample obtained by endoscopy showed chronic active colitis, a feature of IBD; however, other typical features of IBD (i.e. dense lymphoplasmacytic infiltrate, crypt abscesses, granulomas) were not present. The finding of CMV colitis, which can mimic or complicate IBD, is of unclear significance since the rest of the clinical presentation was not typical of CMV colitis. The diagnosis of vasculitis was made only after examining the mesenteric vasculature of a full thickness biopsy sample from the resected ileum.The patient does not satisfy the diagnostic criteria of any systemic vasculitis syndrome. She is diagnosed with isolated eosinophilic mesenteric vasculitis. Mesenteric vasculitis is rare, especially in pediatrics, and can clinically mimic IBD. When the diagnosis of IBD is not certain or the response to therapy is not as expected, alternative diagnoses must be considered.

**345 HYDROURETERONEPHROSIS CAUSED BY CROHN'S DISEASE IN TWO PEDIATRIC PATIENTS.** L. Feinstein, S.E. Fleet, S. Shrager Lusman, E.S. Lamou  -Smith, Pediatric Gastroenterology, Columbia University, New York, New York, UNITED STATES.

Urologic complications of Crohn's disease include urinary tract infections, urolithiasis, enterovesical fistulization and ureteral obstruction leading to hydronephrosis. Urinary symptoms are often minimally reported and overshadowed by the foremost intestinal manifestations of the disease. Ureteral dilation has been reported in Crohn's disease due to renal calculi, pelvic abscesses and transmural inflammatory changes. Ureteral obstruction has been reported to occur in 6% of adults with Crohn's disease. In one small retrospective series of pediatric patients with Crohn's disease, 1% had ureteral obstruction. In this case series we present two adolescent males with hydroureteronephrosis at the time of diagnosis of Crohn's disease.The first patient is a 14 year old male who initially presented with recurrent fevers, weight loss, intermittent abdominal pain, diarrhea and perianal skin tags. Inflammatory markers and fecal calprotectin were significantly elevated. Endoscopy with biopsy confirmed the diagnosis of ileocolonic Crohn's disease. Infliximab monotherapy was initiated. He continued to have intermittent fever, abdominal pain and diarrhea with elevated C-reactive protein. After treatment with oral antibiotics, he defervesced and inflammatory markers began to normalize. Magnetic resonance enterography (MRE) showed bowel wall thickening in the terminal ileum and moderate right sided hydronephrosis and hydroureter. There was no evidence of obstructing lesion, fistulizing disease or pelvic abscess. Urologic evaluation including voiding

cystourethrogram and renal scan was otherwise normal. The second patient is a 15 year old male who presented with intermittent hematochezia, rectal pain and large perianal skin tags. Inflammatory markers and fecal calprotectin were also significantly elevated. MRE showed a normal appearing intestine and severe left sided hydronephrosis. Similar to the first patient there was no evidence of obstructing lesion, pelvic abscess or fistulizing disease. Endoscopic findings included scattered aphthous ulcerations in the colon with most significant ulcerations and inflammation in the descending and transverse colon. In this case series we present 2 pediatric patients with hydronephrosis at the time of diagnosis of Crohn's disease. The laterality of hydronephrosis was correlated with location of disease which to our knowledge has not been previously reported. Urologic complications of Crohn's disease are not widely reported in pediatrics but represent a significant extra-intestinal manifestation of the disease. These cases illustrate the importance of recognizing urologic complications which may be more prevalent than previously reported in pediatrics. In addition, hydronephrosis has the potential to lead to long term kidney damage, therefore selecting therapeutic options for these patients may require consideration of the nephrotoxic potential of medications used to treat Crohn's disease.

**346 AN UNEXPECTED CAUSE OF ABDOMINAL PAIN IN CROHN'S DISEASE.** L. Alrabadi, D. Pashankar, Pediatric Gastroenterology & Hepatology, Yale University School of Medicine, New Haven, Connecticut, UNITED STATES. Cleary, D. Ozgediz, Pediatric Surgery, Yale University School of Medicine, New Haven, Connecticut, UNITED STATES. Morotti, Pathology, Yale University School of Medicine, New Haven, Connecticut, UNITED STATES. Escalera, Pediatric Gastroenterology Specialists, North Haven, Connecticut, UNITED STATES.

**Introduction:** Intussusception is an acquired invagination of the distal small bowel into the proximal bowel. In most cases, this occurs in the 3 months–3 years old age range and is due to lymphadenopathy. It is uncommon in persons > 3 years of age and, pathologic lead points comprise <5% of cases overall but occur at increased frequency in older patients. Focal causes for lead points can include Meckel's diverticula, inflammatory processes, as well as benign and malignant tumors. Here, we describe a 15-year old female with Crohn's disease who presented with small bowel intussusception secondary to an inflammatory myofibroblastic tumor. **Case Report:** A 15-year old female patient with a past medical history of Crohn's disease diagnosed 2 years prior to presentation. She was initially managed with 6-MP therapy, however, for worsening and development of fibrostenosing disease, she had been started on Infliximab therapy. For the 9 months prior to presentation, she had been maintained at 10 mg/kg every 8 weeks with clinical improvement. She acutely presented with a 2-week history of intermittent, generalized, crampy, post-prandial abdominal pain. Associated symptoms included nausea, non-bloody bilious emesis and non-bloody diarrhea. There was no history of fevers. Patient reported a 4-pound weight loss in the past 1 month. Labs were generally unremarkable with normal inflammatory markers. Stool studies were negative. Abdominal x-ray showed an overall paucity of bowel gas with no evidence of obstruction. MR enterography showed a short segment of small bowel-small bowel intussusception within the mid-distal ileum and no evidence of active Crohn's disease. A uniformly-enhancing mass-like lesion thought to serve as the lead point for intussusception was also found, measuring approximately 2.7 x 2.9cm. In retrospect, the mass was thought to be present on previous MRE imaging 9 months prior, however, was significantly smaller, measuring approximately 1.0 x 1.3cm. Subsequently, the patient underwent diagnostic laparoscopy and laparoscopic-assisted reduction of the intussusception, and a moderate size small bowel tumor was identified as the lead point. A small bowel resection was performed and an enlarged adjacent lymph node was excised. Pathology was consistent with an inflammatory myofibroblastic tumor with clear margins, invading the bowel wall but not with transmural involvement. Immunostain and FISH for anaplastic lymphoma kinase (ALK) was negative. The lymph node was normal. She had an uneventful recovery. Her Crohn's disease is well controlled on maintenance 8-weekly Infliximab infusions. **Discussion:** Intussusception in an adolescent is a rare event. Generally, pathologic lead points as cause of intussusception can be secondary to etiologies including Meckel diverticulum, intestinal polyps, duplications, lymphoid hyperplasia, benign and malignant tumors. We describe a case of an intestinal inflammatory myofibroblastic tumor causing ileal intussusception. These are rare neoplastic tumors classified in the family of spindle cell lesions. The optimal treatment is complete surgical excision. Radiotherapy and chemotherapy have been tried with minimal benefit. Residual disease that is ALK positive may respond to chemotherapy. Recurrence rates range from 15-37% and commonly occur locally in the first year after removal, and distant metastasis is rare. This case highlights the rarity of this tumor and clinical approach utilized for the diagnosis and management.

**350 INFLIXIMAB RELATED INFUSION REACTIONS IN VERY EARLY ONSET INFLAMMATORY BOWEL DISEASE.** M.A. Conrad, J. Kelsen, Pediatric Gastroenterology, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES. N. Dawany, Department of Biomedical and Health Informatics, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES. A. Conrad, K.E. Sullivan, M. Devoto, J. Kelsen, Pediatrics, University of Pennsylvania School of Medicine, Philadelphia, Pennsylvania, UNITED STATES. K.E. Sullivan, Immunology, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES. M. Devoto, Epidemiology and Biostatistics, University of Pennsylvania School of Medicine, Philadelphia, Pennsylvania, UNITED STATES.

Children with very early onset inflammatory bowel disease (VEO-IBD), diagnosed before the age of 6, frequently present with a different phenotype and more severe disease course that may be secondary to an underlying immune dysregulation. Thus, these children often have limited effective treatment options. Infliximab has been shown to be safe and effective therapy in pediatric IBD, but may require special consideration when being used in children with VEO-IBD. A 3 year old girl with poor appetite, growth failure, bloody diarrhea, anemia, and elevated inflammatory markers was subsequently diagnosed with colonic VEO-IBD. Despite treatment with sulfasalazine, rectal 5-ASA, and metronidazole, she continued to have loose stools, growth failure, weight loss, and an elevated calprotectin. She was then treated with infliximab and had a good response with rapid improvement in stool quality, blood in stool, appetite, and energy level. Within minutes of starting her 3<sup>rd</sup> infusion of infliximab, she complained of difficulty breathing, abdominal pain, followed by vomiting, pallor, diffuse erythema, and subsequent vital sign changes including tachycardia with heart rate in 150s, oxygen desaturation to 88%, and tachypnea with respiratory rate 40. The infusion was discontinued, and she received epinephrine 0.15mg intramuscularly and diphenhydramine 1mg/kg intravenously. In addition to protocol, due to the severity of the reaction, methylprednisolone 2mg/kg IV was also given with gradual improvement in her presentation over several minutes. Subsequently, she was admitted to the pediatric intensive care unit for further monitoring. She did not have any further events. She had a normal tryptase level and notably, she had no detectable infliximab neutralizing antibodies at the time of this event. A retrospective chart review of randomly selected IBD patients cared for at The Children's Hospital of Philadelphia was performed to identify infusion reactions to infliximab. 381 subjects (81 VEO-IBD, 300 diagnosed with IBD at age 10 years or older) who were treated with infliximab were reviewed. Of these 81 with VEO-IBD, 19 (23%) had an infusion reaction, while 41/300 (14%) older onset IBD had reactions ( $p=0.04$ ). These reactions included oral itching, skin erythema, chills, shortness of breath, tachycardia, and anaphylactic-like reactions requiring treatment with epinephrine. Of the 41 older onset IBD subjects with infusion reactions, these reactions occurred within a median of 28.7 weeks (range 2 weeks to 4 years, 10 weeks) from initiating the first dose of infliximab. In the 19 VEO-IBD subjects, reactions occurred within a median of 6 weeks (range 2 weeks to 38 weeks). Here we demonstrate an increased rate of infusion reactions among the VEO-IBD population as compared to the older subjects. Additionally, the VEO subjects had a shorter duration of IFX therapy at the time of their reaction, often occurring during induction whereas the patients diagnosed after age 10 had a longer therapy course at the time of infliximab reaction. The normal tryptase level in our index case supports that this reaction was not true anaphylaxis. The prominent role of immune dysregulation in the VEO-IBD population likely contributes to the infusion reactions in this group, but this requires further investigation. The risk and severity of infusion reactions and the importance of prompt attention at the time of these unpredictable reactions warrants close monitoring by medical staff during infliximab infusions, particularly in the pediatric patient population.

**354 KAWASAKI DISEASE IN A PATIENT WITH INFLAMMATORY BOWEL DISEASE.** M. Huckaby, J. Huang, Pediatrics, University of California, San Diego, La Jolla, California, UNITED STATES. M. Huckaby, J. Huang, Pediatric Gastroenterology, Hepatology and Nutrition, Rady Children's Hospital, San Diego, California, UNITED STATES. Background: Several inflammatory conditions have been reported concomitantly in patients with inflammatory bowel disease. Kawasaki disease is a systemic inflammatory disease that targets the vascular system that has only been previously reported once in the literature. Case: A 10 year old male with a history of ulcerative proctitis previously controlled on rectal mesalamine and hydrocortisone enemas, had repeat endoscopy for persistent abdominal pain a year after diagnosis. Endoscopy and pathology demonstrated moderate chronic active inflammation with crypt abscesses in the descending colon (new from prior endoscopy). Sulfasalazine therapy was initiated for extension of disease. Gastrointestinal symptoms resolved. Approximately 6 weeks after starting sulfasalazine, the patient developed fevers, sore throat, rash, periorbital edema, neck pain, abdominal pain,

bilateral hand swelling, and arthralgias. Labs were remarkable for a mildly elevated ESR (21mm), hypoalbuminemia (3.0 gm/dl) elevated liver chemistries (AST 227 U/L, ALT 268 U/L, GGT 169 U/L), microcytic anemia (Hgb 11.9 g/dL, MCV 72.5 fL), mild coagulopathy (PT 15.8 sec, INR 1.3) and normal urinalysis. Abdominal ultrasound demonstrated gallbladder hydrops. Echocardiogram revealed mildly dilated left main coronary artery (z score 2.26) and left anterior descending artery (z score 3.59) without evidence of aneurysm and a trivial inferior pericardial effusion. His electrocardiogram was notable for a QTc of 458, which on repeat demonstrated RSR prime pattern suggestive of myocarditis. He was treated with IVIG, infliximab (10 mg/kg x 1) and high dose aspirin. Sulfasalazine was discontinued because of concern of drug reaction. His fever resolved within 48 hours of treatment and his rash became purpuric on both upper and lower extremities, thought to be hemorrhage from capillary fragility. Because of persistence and worsening of rash along with concern for drug reaction, aspirin therapy was also discontinued. The patient was discharged home on mesalamine suppositories and received no further therapy. Within 5 months, coronary artery dilations had fully resolved and patient continued asymptomatic of his IBD. Discussion: To our knowledge, this is the second report in the literature of a pediatric patient with inflammatory bowel disease developing Kawasaki disease. Kawasaki disease is a systemic inflammatory disease that targets the vascular system. While it is generally a self-limited disease, it may result in severe complications, with up to 25% of cases resulting in coronary aneurysms or ectasia. Interestingly, the prior reported case was also associated with sulfasalazine administration in a patient with IBD. It is possible that some of the atypical symptoms in our patient may have represented a sulfasalazine or mesalamine hypersensitivity reaction, which has been associated with pericardial effusions and myocarditis in patients with inflammatory bowel disease. Furthermore, pericardial effusions, endothelial dysfunction, and valve insufficiency have been reported as extraintestinal manifestations of inflammatory bowel disease. Coronary artery changes however, have not been reported in the literature in association with mesalamine or sulfasalazine hypersensitivity. In addition, our patient did not discontinue his mesalamine exposure while his coronary issues resolved. Gastrointestinal inflammation and colitis have been reported as a manifestation of Kawasaki's disease in the literature. One case in the literature reports a case of Kawasaki's disease presenting with a focal left sided colitis diagnosed on imaging, but without any knowledge of biopsy proven diagnosis of inflammatory bowel disease. We report a case of Kawasaki's disease developing in a patient with IBD. Potential mechanisms include drug sensitivity and/or common inflammatory pathways. Physicians should have a high-index of suspicion for other inflammatory conditions including Kawasaki's in patients with IBD.

**356 GRANULOCYTE-MACROPHAGE COLONY-STIMULATING FACTOR (SARGRAMOSTIM) INDUCES REMISSION AFTER LICHENOID DRUG ERUPTION DUE TO INFlixIMAB IN CROHN'S DISEASE.** A. Lyon, L. Cooke, M.B. Heyman, S.G. Verstraete, Pediatrics, University of California, San Francisco, San Francisco, California, UNITED STATES. Small, K. Cordora, Dermatology, University of California, San Francisco, San Francisco, California, UNITED STATES. Development of lichenoid eruptions is a known complication of TNF $\alpha$  inhibitors. We report a case of lichenoid drug eruption in a teenager on infliximab for Crohn's disease. A 15-year-old male was diagnosed with ileocolonic Crohn's disease in 2012. He maintained clinical remission on infliximab 5mg/kg monotherapy since 2014, but after one year of treatment, he developed a waxing and waning rash that coincided with infliximab infusions. The rash appeared on his trunk and extremities and was described as numerous erythematous, slightly indurated plaques, many with a dusky center, that were 1-4cm in diameter; these were accompanied by several hyperpigmented round patches. Dermatology initially diagnosed pityriasis rosea, thought to be unrelated to his underlying Crohn's disease or medication. Rash persisted despite topical steroids and worsened with each infusion. A punch biopsy in early 2016 revealed a lichenoid interface dermatitis versus recurrent erythema multiforme, likely drug-induced. Infliximab was stopped, and methotrexate treatment was initiated. However, he developed symptoms of abdominal pain and diarrhea, so he was admitted to start adalimumab given concern for a severe acute reaction should he be sensitive to anti-TNF $\alpha$  medications. He tolerated adalimumab well, but within a few weeks the rash worsened, so adalimumab was discontinued. A second skin biopsy confirmed a lichenoid drug eruption and was less suggestive of drug-related erythema multiforme. The patient declined treatment with another anti-TNF $\alpha$  agent due to the severity of the rash. All immunosuppression was stopped, and he was started on an initial 8-week course of sargramostim. The rash resolved, with minimal residual hyperpigmentation, and he achieved complete clinical remission of his IBD symptoms. Lichenoid reactions are a rare yet well described adverse effect to TNF $\alpha$  inhibitors<sup>1,2</sup> and are hypothesized to be caused by a paradoxical inflammatory response due to upregulation of other cytokines such as interferon alpha and CD8+ T-cells. They often resolve upon discontinuation

of the anti-TNF $\alpha$  agent. Granulocyte-macrophage colony-stimulating factor (GM-CSF) is a myeloid growth factor that is also expressed by CD4+ T cells and Paneth cells of the intestinal epithelium. Exogenous GM-CSF may augment host defenses and ameliorate inflammatory responses associated with Crohn's disease and has been shown to induce remission and improve clinical symptoms in these patients<sup>3-6</sup>. It is unknown whether the improvement in our patient's lichenoid reaction was facilitated by the same mechanism. References:1.

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**357 A RARE CASE OF GALLBLADDER AND COMMON BILE DUCT CAPILLARY HEMANGIOMA IN A TODDLER.** A.R. Shahein, D.A. Moya, A. Buckley, R.D. Baker, S.S. Baker, Digestive Diseases & Nutrition Center, Women & Children's Hospital of Buffalo, Buffalo, New York, UNITED STATES. John, Department of Radiology, Women & Children's Hospital of Buffalo, Buffalo, New York, UNITED STATES. Kozielski, Department of Pathology, Women & Children's Hospital of Buffalo, Buffalo, New York, UNITED STATES.

Jaundice is a common symptom in pediatrics with a broad differential diagnosis that in some circumstances requires an extensive work-up. Obstruction of the biliary tract is an important cause of cholestatic jaundice. We present a case of a 2-year-old male previously healthy who presented with progressive obstructive jaundice and elevated transaminases. Imaging studies revealed dilatation of the hepatic ducts, and stenosis of the common bile duct (CBD). A liver biopsy showed biliary changes consistent with acute cholangitis. He required transient percutaneous cholecystostomy tube and hepatic duct drain placement. Eventually he underwent a laparoscopic cholecystectomy with partial resection of proximal CBD and hepatico-duodenostomy reconstruction. His post-operative course was complicated by left hepatic stricture that required dilatation. Histological examination of the gallbladder and extrahepatic bile duct stricture revealed a vascular proliferation consistent with capillary hemangioma. We present a unique case of a pediatric patient with a hemangioma located in a very uncommon site. None of the previously reported patients with hemangioma have presented with cholestasis or involvement of the common bile duct. Similar to our patient, in these reports, the pre-operative diagnosis was not suggestive or conclusive for hemangioma. Our case highlights the importance to keep a high index of suspicion for uncommon causes of obstructive jaundice when evaluating children with progressive cholestasis.

**358 VERY EARLY ONSET INFLAMMATORY BOWEL DISEASE WITH SCLEROSING CHOLANGITIS.** A. Chan, R. Abell, N. Patel, N. Kerkar, Pediatric Gastroenterology, University of Rochester Medical Center, North Chili, New York, UNITED STATES. Lambert, Pediatric Gastroenterology, Buttermilk Falls Pediatrics, Ithaca, New York, UNITED STATES.

Introduction:Uridine diphosphoglucuronate-glucuronosyltransferase 1A1 (UGT1A1) is an enzyme that is responsible for conjugation of bilirubin to glucuronic acid. A defect in UGT1A1 gene can lead to syndromes of elevated unconjugated bilirubin, such as Gilbert syndrome or Crigler-Najjar (CN) syndrome (Types I or II). CN Type I is the most severe form with complete absence of hepatic bilirubin-uridinediphosphoglucuronate glucuronosyltransferase activity. CN Type II presents with intermediate levels of hyperbilirubinemia, resulting from an incomplete deficiency of hepatic UGT1A1 activity. We describe a case of a novel variant mutation of UGT1A1 that does not meet criteria for diagnosing either CN Type I or II. Case presentation:A 2 year old Sudanese immigrant male presented from Egypt for evaluation of liver transplantation. He was diagnosed early in life with presumed CN Type I due to severity of hyperbilirubinemia and response to phenobarbital. There was a history of consanguinity and maternal hyperthyroidism for which his mother received thiouracil. He had been receiving weekly phototherapy to decrease his hyperbilirubinemia and reduce his risk of kernicterus. One year prior to his

arrival to the United States, his parents stopped his phototherapy treatment. Despite lack of medical interventions, he remained asymptomatic with normal development. Upon arrival to the hospital, his total bilirubin was 27mg/dL and 26.8mg/dL of this was indirect. He was treated with intensive phototherapy, phenobarbital and cholestyramine with good response (down to 9.4). Neurologic workup was unremarkable except for abnormal otoacoustic testing on the right ear. Abdominal ultrasound demonstrated patent hepatic and portal vasculature. To confirm the diagnosis, UGT1A1 gene sequencing was done, showing that the patient has a homozygous mutation in the UGT1A1 gene of uncertain significance. The sequence variant is designated c.824\_826delTTG, which is predicted to result in the in-frame deletion of an amino acid (p.Val275del). This mutation has not previously been reported in literature or public databases. Due to other patients with in-frame deletions in exon 1 of UGT1A1 having significant disease, there is suggestion that this patient's variant is possibly pathogenic, but clinical significance is not entirely certain. The patient is also homozygous in the UGT1A1 gene for c.-41\_-40dupTA, a dinucleotide repeat variation in the A(TA)<sub>n</sub>TAA motif in the promoter region, specifically the A(TA)<sub>7</sub>TAA allele, which has been associated with Gilbert syndrome. However, the frequency of this allele is common, and has been classified as a benign variant. Discussion: Currently, our patient just received a deceased donor liver transplant and we are awaiting further genetic testing and histopathology of the native liver. To our knowledge, this is the first reported case of this particular sequence mutation. Given that it does not fit with a classic presentation of Type I or II, this case report may expand the spectrum of Crigler-Najjar syndrome.

**359 TRANSIENT FACTOR VII DEFICIENCY POST PEDIATRIC LIVER TRANSPLANTATION.** A. MacLellan, Medicine, Dalhousie University, Halifax, Nova Scotia, CANADAA. Noble, Division of Gastroenterology & Nutrition, IWK Health Centre, Halifax, Nova Scotia, CANADAK. Kulkarni, Division of Hematology-Oncology, IWK Health Centre, Halifax, Nova Scotia, CANADAA. Noble, K. Kulkarni, Pediatrics, IWK Health Centre, Halifax, Nova Scotia, CANADA. Introduction: Factor VII deficiency (FVII deficiency) can be inherited or acquired. Causes of acquired FVII-D include liver disease, Vitamin K deficiency, and oral anticoagulant therapy. We present a case of de novo transient FVII deficiency post liver transplant in a 4-year-old girl with alpha-1-antitrypsin deficiency (A1AT). Case Description: A 4-year-old girl diagnosed with A1AT at age 1 year required a liver transplant secondary to decompensated cirrhosis. Past medical history was negative for significant bleeding episodes, and INR at diagnosis was normal (0.99-1.0). She received a living-donor related liver transplant from her mother at age 4 years. Her mother had a normal INR at the time of transplantation. Two months post-transplant, our patient developed non-bloody diarrhea positive for *C. difficile*. INR at time of infection was 1.4 (normal range 0.88-1.12). Abdominal ultrasound demonstrated normal liver morphology. Elevated liver enzymes normalized after treatment of the *C. difficile* infection, but the INR did not normalize (range 1.36-1.42) even following Vitamin K administration (5 mg I.V., then 3x5 mg PO once daily). Additional blood work revealed she had a low FVII (0.38 U/mL, normal range 0.51-1.54 U/mL). She was asymptomatic (no increased bruising or bleeding). The donor's FVII (0.54) and INR (1.1) levels were normal post-donation. Ten months post-transplant, our patient's repeat INR was 1.3 (normal 0.8 – 1.2) and FVII was 0.54. She is clinically well and liver graft healthy. Discussion: Our patient had a normal coagulation profile until she developed decompensated cirrhosis. Post liver transplant, she had no bleeding diathesis. INR was measured secondary to elevated liver enzymes and the potential need for a liver biopsy. FVII deficiency was not inherited because the liver donor does not have FVII deficiency. Our patient's INR did not normalize following Vitamin K supplementation, ruling out Vitamin K deficiency. She was not on oral anticoagulants. She has remained clinically well with otherwise normal labs, so other causes of FVII deficiency are not applicable. Our patient most likely acquired FVII deficiency following liver transplant. Other coagulation factor deficiencies (Factors XI, Factor XII, Protein S, Protein C) following liver transplantation have been reported. We hypothesize that the FVII deficiency was secondary to relative graft/recipient weight ratio or alternatively secondary to the liver donor's low normal FVII levels. The FVII deficiency is resolving and INR improving. We expect that the INR will completely normalize with liver growth over time. Conclusion: Our patient represents the first pediatric patient reported with a transient FVII deficiency post-liver transplantation. Coagulation factor deficiencies should be investigated in patients with unexplained elevation of INR post liver transplantation.

**360 CHOLESTATIC LIVER DISEASE ASSOCIATED WITH STEVENS-JOHNSON SYNDROME: A CASE REPORT.** A.J. Peasley, C. Potter, Gastroenterology, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES. Introduction: Several case reports have described cholestasis and vanishing bile duct syndrome (VBDS) associated with Stevens-Johnson syndrome (SJS). We describe a case of an adolescent who developed persistent cholestatic

liver disease from SJS presumably secondary to vanishing bile duct syndrome who was treated with tacrolimus. Case Description: A previously healthy 16 year old female was admitted with high fever, progressive oral and genitourinary mucosal ulceration, bullous skin lesions, eye pain, and blurry vision, concerning for SJS, after ibuprofen use for neck pain. She received 3 g/kg intravenous immunoglobulin. Extensive infectious work up was unremarkable. Non-steroidal anti-inflammatory drugs were discontinued as the presumed etiology for SJS. She required TPN for 12 days due to inability to eat secondary to oral mucosal involvement. At admission, liver function tests were remarkable for elevated total and direct bilirubin (2.1 mg/dL and 1.8 mg/dL, respectively), elevated ALT (388 U/L), elevated AST (206 U/L), elevated alkaline phosphatase (158 U/L), and elevated GGT (369 U/L). During the course of her hospitalization, these abnormalities progressively worsened. Abdominal ultrasound with doppler and MRCP were unremarkable. Ceruloplasmin, alpha-1 antitrypsin pi-typing, and serologies for autoimmune hepatitis were unremarkable. She was started on ursodiol when able to tolerate oral intake. She underwent liver biopsy about 1 month after presentation which demonstrated marked centrilobular cholestasis, in a pattern most consistent with drug-induced liver injury and cholestasis. No significant portal tract inflammation and minimal lobular inflammation. By the time of the liver biopsy, patient had been clinically improving from her SJS and was eating entirely by mouth, and was on day 11 of a systemic steroid taper for SJS. She was discharged from the hospital the day after the liver biopsy on hospital day 28. LFTs were monitored closely as an outpatient while on ursodiol over the next 2 months. She had a transient worsening of LFTs 1 week later, but then a gradual downward trend without normalization. Nutrition was closely monitored while cholestatic. She was maintained on fat-soluble vitamin supplementation. Vitamin levels were checked frequently. Vitamin D required additional supplementation. Otherwise, vitamin levels remained normal. In January 2017, GGT and alkaline phosphatase significantly increased (776 U/L and 754 U/L, respectively), despite improving total bilirubin, direct bilirubin, and transaminases. Repeat abdominal ultrasound was unremarkable. Serum bile acids were significantly elevated at 330 umol/L, raising concern that her cholestasis was worsening despite the slow improvement in her bilirubin. We worried that her progressive cholestasis was related to VBDS and decision was made to start tacrolimus in an attempt to halt the progression of the liver disease. Patient was started on tacrolimus 0.15 mg/kg/day initially, with goal drug levels of 6-8 ng/mL. Within 6 weeks of starting and optimizing the tacrolimus, her LFT abnormalities and serum bile acids had normalized. The synthetic function of her liver has remained intact since the initial insult. She remains on tacrolimus at this time, with plans to discontinue the medication after about 6 months if she continues to do well. Additionally, she remains on ursodiol. Her course has been complicated by bronchiolitis obliterans (FEV1 <40%). The progression of her severe obstructive pulmonary disease has not been affected by tacrolimus. Conclusion: SJS and VBDS are likely both immune-mediated processes, with similar causative drugs/medications. Because of the rarity of both of these diseases, and lack of great evidence available in the literature, a standardized approach to treatment is not available. Treatment with steroids, choleric agents like ursodiol, and immunosuppressants including tacrolimus and biologic agents, have been described with varying results. In our patient, cholestatic liver disease presumed to be secondary to VBDS resolved with tacrolimus. If progression of cholestasis cannot be slowed, these patients ultimately require liver transplant.

**363 OUTCOME OF CHILDREN WITH ABERNETHY MALFORMATION TYPE II AFTER REMOVAL OF CONGENITAL SHUNT.** I. Batsis, W. Karnsakul, Pediatric Gastroenterology, Johns Hopkins, Baltimore, Maryland, UNITED STATES. Mitchell, Interventional Radiology, Johns Hopkins, Baltimore, Maryland, UNITED STATES. Stewart, Pediatric Surgery, Johns Hopkins, Baltimore, Maryland, UNITED STATES. Dunn, Pediatric Radiology, Johns Hopkins, Baltimore, Maryland, UNITED STATES.

Background: Congenital portosystemic shunt is a rare congenital condition first reported by John Abernethy in 1793. Abernethy malformation type II (AM II) is defined with portal blood being partially drained to systemic venous system resulting into hypoplastic intrahepatic portal vein and atrophy of the liver. The diagnosis is often incidental based on imaging studies or occasionally with presentations of hyperammonemia/neurological abnormalities, liver tumors, pulmonary hypertension, hepatopulmonary syndrome and lower gastrointestinal bleeding (LGIB). LGIB is an uncommon presentation. LGIB occurs as a result of the shunt draining portal blood commonly from inferior mesenteric vein into the internal iliac veins (IIV) via the inferior mesenteric vein (IMV). Surgical intervention with shunt ligation or removal (from portal vein or IMV to IIV) is one approach in most patients. We describe clinical outcomes of two cases after the ligation of congenital shunt, one with LGIB and second one with hepatopulmonary syndrome and liver mass. Case 1: A 13 year old female presented with intermittent rectal bleeding since age 3 years. Upon investigation, a CT scan of the abdomen demonstrated small

liver size (without focal lesions) and hypoplastic intrahepatic portal vein with diversion of portal flow from superior mesenteric and splenic veins into dilated IIV consistent with Abernethy malformation type II and a shunt from PV to IIV, and variceal formations in perirectal veins. Shunt ligation was performed at age 10 years resulting in increasing in the volume of the right lobe and caudate lobe of the liver. Mesenteric and perirectal veins, however remained dilated without visible drainage to portal vein (PV) due to possible thrombosis. Pelvic arteriogram, celiac axis arteriogram, superior and inferior mesenteric arteriogram in multiple projections revealed multiple complete long segment occlusions in distal branches of the superior mesenteric vein. Collateral flow was seen to the left PV (evidently not sufficient to reduce portal hypertension) with a backflow into large descending colonic varices, and numerous rectal varices with a small connection draining to the right IIV. She is currently under evaluation for a mesocaval shunt to bypass blood flow proximal to SMV occlusions and to reduce venous flow to rectal vein. Case 2: A 12 year old presented with a 3 year history of dyspnea and exercise intolerance. Her chest X-RAY showed diffuse increased interstitial pattern. Transesophageal echocardiography indicated multiple pulmonary microscopic arteriovenous malformations with of LVEF at 60% and bubble echocardiogram demonstrating large interatrial shunting. Hereditary hemorrhagic telangiectasia was initially suspected. Chest and abdominal CT scans were performed - 1 possible small AVM in lower lobe of the left lung and 9 cm mass in the right hepatic lobe with cavernous transformation of the PV and portovenous collateral extending to the left common iliac vein. Portal venogram/arteriography showed direct communication between the portosystemic shunt and the native portal vein and confirmed Abernethy malformation type II. Liver biopsy confirmed that the liver mass was a hyperplastic nodule or adenoma and no evidence of malignancy. Surgical ligation performed after portal pressures were repeated. Dyspnea significantly improved 6 months after removal of the shunt (improved exercise tolerance with 6 minute walk test). At a 3 year follow up, her liver mass reduced by 1 cm with a normal AFP level. No evidence of LGIB has been observed. Conclusion: Although the mechanisms of the development of the shunt from IMV to IIV or IVC collaterals remains unknown in Abernethy malformation type II, portal hypertension seems to be the driving factor and the cause of LGIB as a complication. Also, it has been postulated that vasoactive mediators bypass the hepatic metabolism and cause intrapulmonary dilation contributing to the pulmonary symptoms. Surgical correction of the anomalous hepatic vasculature seems to be imperative, but even after removal of congenital shunt symptoms such as LGIB and liver mass may persist and require close follow-ups.

**365 THE ROLE OF THE PEDIATRIC GASTROENTEROLOGIST IN DIAGNOSIS AND MANAGEMENT OF HLH PRESENTING WITH ACUTE LIVER FAILURE: A CASE SERIES.** J.A. Bitong, S. McDiarmid, Y. Bulut, I.K. Weiss, E.A. Marcus, Department of Pediatrics, DGSOM at UCLA, Los Angeles, California, UNITED STATES.

Hemophagocytic Lymphohistiocytosis (HLH) is a rare, potentially fatal disorder characterized by hyperactivation of the immune system initiated by a cytokine storm. Primary HLH is a well-studied genetic disorder typically presenting in infancy, whereas secondary, or acquired, HLH, is a complication of another disease process. Both primary and secondary HLH can pose significant diagnostic challenges. This sepsis-like disease involves multiple organ systems and can be triggered by different conditions including infection, malignancy, immunodeficiency, and autoimmune disease. Missed or late diagnosis is the major cause of mortality. The disorder often presents with fever, cytopenia of at least 2 cell lines, organomegaly, and some degree of liver dysfunction, manifest by elevated transaminases, a slightly elevated bilirubin, or the subtler finding of a decreasing albumin level. HLH is often confused with sepsis or disseminated intravascular coagulation (DIC) but does not respond to antibacterial treatment. Input from multiple subspecialists including pediatric gastroenterology is needed in distinguishing HLH from other entities, as early diagnosis and treatment is crucial. Here, we compare and contrast two patients, one with acquired HLH and one with primary HLH, both referred to our center in the setting of acute liver failure (ALF) for liver transplant evaluation. The first patient is a 19-year-old male, previously healthy, who initially presented with the chief complaint of bilateral leg weakness and fever. Over the course of 3 months, he was admitted 6 times, with 3 prolonged hospitalizations at different centers. Despite extensive multi-specialist evaluation, no diagnosis was made prior to transfer to our center for liver transplant evaluation. In the 3 months prior to his presentation to our center, his transaminases and bilirubin increased, and albumin decreased – all were normal on initial presentation. He also developed worsening pancytopenia, splenomegaly, hypofibrinogenemia, hypoalbuminemia, hyperferritinemia, and hypertriglyceridemia, all in the setting of persistent fever despite multiple courses of antibiotics. Upon transfer to our center, his AST was 3575 u/L, ALT 1554 u/L, and INR 2 following multiple units of plasma prior to transfer. He met diagnostic criteria for HLH with ferritin >20,000 mg/L, fibrinogen 73 mg/dL, triglycerides 732 mg/dL, pancytopenia, splenomegaly, and fever. He was started on HLH

treatment with etoposide and high dose steroids. Shortly after treatment was initiated, we received confirmation of hemophagocytosis on bone marrow biopsy performed at a prior institution. The patient died 5 days after admission in multi-organ failure (MOF). Following his death, soluble IL-2 receptor returned high at 42,941 u/L and natural killer (NK) cell activity was decreased. Genetic analysis was negative for HLH mutations. The second patient is a 7-week-old male, previously healthy and with normal birth history, who presented with a 3-day history of fever, fussiness, and abdominal distention, stooling and feeding normally. In a local emergency department, his platelet count was 16 and he was admitted, treated for severe sepsis and DIC. 2 days later he was transferred to our center for liver transplant evaluation in ALF, with AST 2207 u/L, ALT 643 u/L, and INR 2.6 after multiple units of plasma prior to transfer. He presented febrile, already intubated, and on pressor support. He had a ferritin >20,000 mg/L, pancytopenia, fibrinogen 51mg/dL, and hepatomegaly. The patient was too unstable to undergo bone marrow biopsy, and empiric treatment for HLH with etoposide and high dose steroids was initiated. However, the patient died within 72 hours in MOF. Soluble IL-2 receptor later came back high at 27,300 pg/mL. The results of clinical exome sequencing were consistent with autosomal recessive familial HLH. As evidenced by these 2 patients, the initial presentation of HLH can be extremely variable and potentially ambiguous. Up to 30% of patients with HLH initially present with vague neurological symptoms (Sen et al. 2017). They can also present with fever and constitutional symptoms. ALF can be seen in HLH in the setting of MOF, bringing these patients to the attention of the pediatric gastroenterologist. Although liver transplantation is not indicated for HLH due to immune hyperactivation, the pediatric gastroenterologist can evaluate for causes of liver failure, including the complex HLH diagnosis. A liver biopsy showing hemophagocytosis, prior to onset of MOF, may be helpful. A high ferritin can be paramount in diagnosis. Ferritin >10,000 mg/L in children is 90% sensitive and 96% specific for HLH (Sen et al. 2017). Complexity of HLH diagnosis requires high level of suspicion, education, and multiple subspecialist involvement.

**367 IDENTIFICATION OF OBSCURE GASTROINTESTINAL BLEEDING POST-LIVER TRANSPLANTATION: THE ROUX OF THE CAUSE.** T. Beduschi, D. Goldner, A. Tekin, J. Garcia, Miami Transplant Institute, Miami, Florida, UNITED STATES. J.T. Hochberg, D. Goldner, J. Garcia, Pediatric Gastroenterology, Hepatology and Nutrition, University of Miami, Miami, Florida, UNITED STATES. J.T. Hochberg, Department of Pediatrics, Jackson Memorial Hospital, Miami, Florida, UNITED STATES. Beduschi, A. Tekin, Department of Surgery, Jackson Memorial Hospital, Miami, Florida, UNITED STATES.

Roux-en-Y hepaticojejunostomy is a common technique for biliary reconstruction in pediatric liver transplantation; however, rarely is this anastomotic site the etiology of obscure gastrointestinal (GI) bleeding. The etiology of GI bleed, its incidence and its timing post-transplantation has only been described in a small number of publications, with the majority of these retrospective reviews focused on an adult population. Though the overall incidence of GI bleeds post-liver transplant has been reported to range between 4-16%, these bleeds are most often noted to be an early complication occurring in the immediate post-operative period, with the majority occurring within the first three months. The most common etiologies reported include ulcers, enteritis, esophageal varices, and anastomotic bleeds. Here we describe the evaluation and treatment of an occult GIB in a young male, 8 years status post orthotopic liver transplantation (OLT) with hepaticojejunostomy, who was found to have an unusual etiology for bleed: varices at the hepaticojejunostomy anastomosis. This 9 year old boy with history of biliary atresia s/p failed Kasai portoenterostomy received a left lateral segment OLT with hepaticojejunostomy at the age of 11 months. He subsequently did well, maintained on sirolimus monotherapy; however 8 year post-transplantation he presented to the ER with complaints of melena and weakness, and was found to have a hemoglobin of 5.8. The patient underwent an extensive work up including esophagogastroduodenoscopy (EGD), colonoscopy, push enteroscopy and tagged red blood cell study, all with normal results and no identifiable source of the bleed despite persistent heme positive stools and acute drops in hemoglobin requiring multiple transfusions. Of note, push enteroscopy was performed and was unable to reach the Roux-en-Y anastomosis site, which led to suspicion of a possible ulcer in that area. Repeat EGD and colonoscopy were again negative, with the enteroscopy scope unable to reach the Roux, and the decision was made to undergo exploratory laparotomy (ex lap) with hopes of identifying the site of the bleed. Ex lap was notable for findings of varices at the site of the hepaticojejunostomy with signs of recent bleeding, in addition to areas of diverticula with weak mucosa noted in the Roux. Both areas of bowel were resected followed by formation of a new Roux without complication or further bleeding. The majority of bleeds post-OLT occur early in the post-operative period and are related to gastric/duodenal ulcers or surgical issues surrounding a fresh anastomosis. Varices in the roux limb secondary to the development of portal

hypertension post-liver transplant is an unusual etiology of occult GI bleed and has only been described in rare case reports. This case highlights the importance of evaluating the Roux-en-Y anastomosis as a source of GI bleeding well beyond the post-transplant period, even in the absence of outward signs of portal hypertension. The diagnosis of Roux-en-Y variceal bleeding requires a high degree of clinical suspicion given the need for a targeted and at times more invasive workup.

**369** *ESCITALOPRAM OXALATE INDUCED ASYMPTOMATIC HYPERBILIRUBINEMIA IN A PEDIATRIC PATIENT.* J.M. Dailey, Pediatrics, Lehigh Valley Hospital, Allentown, Pennsylvania, UNITED STATES. Paul,, Pediatric Gastroenterology, Lehigh Valley Hospital, Allentown, Pennsylvania, UNITED STATES.

Introduction: Escitalopram oxalate, commonly referred to as Lexapro, is a selective serotonin reuptake inhibitor (SSRI) and one of the most widely used antidepressants in adolescents and adults. This medication has known gastrointestinal and metabolic side effects including dyspepsia, nausea, vomiting, abdominal pain or discomfort, increased or decreased appetite, diarrhea, and constipation that is reported in 1-10% of users. Escitalopram is metabolized by the liver mainly through the cytochrome P450 system, but hepatotoxicity and other hepatobiliary side effects remain rare. In adults there have been few case reports citing acute cholestatic liver injury, hepatitis, and elevated liver enzymes associated with use of Escitalopram, but in the pediatric population adverse reactions involving the hepatobiliary system are unreported. Case Report: A 14 year old female with history of general anxiety and major depressive disorder was referred to pediatric gastroenterology for concerns of jaundice and a total bilirubin of 3.9 mg/dL obtained three days after she was started on Lexapro 10 mg. At time of evaluation by GI she had been on Lexapro for 1 week. She was taking no other medications and had no known history of liver or gallbladder disease. The patient was asymptomatic with reported weight loss of 18 kilograms over the past 3-4 months related to a decreased appetite, but no other particular symptoms. Her exam findings were only pertinent for a thin appearing female with scleral icterus and jaundice. Laboratory testing 8 days after starting medication revealed an elevated total bilirubin of 4.2 mg/dL, with normal direct bilirubin 0.3 mg/dL, AST 12 U/L, ALT 24 U/L, and alkaline phosphatase 67 U/L. The remainder of her work up included complete blood count, reticulocyte count, comprehensive metabolic panel, thyroid function studies, celiac serologies (TTg IgA and IgG autoantibody, IgA, and Gliadin autoantibody profile), autoimmune work up (ANA screen, alpha-1-antitrypsin, LKM type 1 autoantibody, smooth muscle autoantibody, mitochondrial autoantibody), PT with INR, gamma-glutamyl transferase, haptoglobin, ceruloplasmin, and lactate dehydrogenase were all within normal limits. A complete abdominal ultrasound was obtained which was normal and showed no evidence of biliary obstruction. After 3 weeks of Lexapro use, her painless jaundice and indirect hyperbilirubinemia had not improved. In discussion with gastroenterology, her psychiatrist discontinued this medication and switched her to Bupropion. Her indirect hyperbilirubinemia was monitored with comprehensive metabolic panels and direct bilirubin level measurements for 2 months after discontinuation of Lexapro, and showed return to near normal bilirubin levels. Her jaundice resolved over this time period as well. Throughout her monitoring while on Lexapro and during the 2 months after discontinuation, her AST and ALT remained within normal limits and her alkaline phosphatase remained mildly low, which was thought to be related to her nutrition status in the setting of all other lab work being unremarkable and her recent significant weight loss. Conclusion: Escitalopram oxalate induced hyperbilirubinemia is rare with only a few case reports in the adult population. Unlike the child in this case, who only had elevation in total bilirubin levels, the adult case reports show other laboratory findings of liver injury or hepatotoxicity including elevated AST, ALT, or alkaline phosphatase levels. Our patient's onset of symptoms and elevation in bilirubin level does seem to be consistent with the timing of hepatobiliary complications seen in adults, which usually start within days and almost always within the first 8 weeks from starting Escitalopram. Though complications of hyperbilirubinemia or cholestasis remain rare in both the adult and pediatric population, it is important to consider medication induced hyperbilirubinemia in patient's recently started on Escitalopram.

**372** *POST-HEMORRHAGIC HYDROCEPHALUS SECONDARY TO ALPHA-1 ANTITRYPSIN DEFICIENCY IN A NEONATE.* J. Mani, S. Madani, Pediatrics, Children's Hospital of Michigan, Detroit, Michigan, UNITED STATES.

Background: Alpha-1 antitrypsin deficiency (A1ATD) is the most common genetic condition causing liver disease in neonates and children. Severe A1ATD with PiZZ phenotype occurs in approximately 1:3500 live births. Clinically significant liver disease usually presents as neonatal cholestasis and is seen in approximately 10% of patients. Here we report a case of A1ATD presenting as post-hemorrhagic hydrocephalus in a neonate. Case report: A 4 week old girl presented to the emergency room with a three week history of jaundice, vomiting with feeds and a one week

history of increasing head circumference. She was born full term without any complications and was exclusively breast fed since birth. There was no ABO incompatibility. She did receive standard vitamin K prophylaxis at birth. Jaundice was attributed to breast feeding and mother was encouraged to breast feed more frequently. She was reported to have vomiting with feeds since the sixth day of life. US-abdomen ruled out pyloric stenosis and she was started on oral ranitidine for possible gastro esophageal reflux disease. During the last two days of presentation, she had several episodes of uprolling of her eyes lasting a few seconds. She had developed scattered bruises and had a self-limited episode of nose bleed. Physical examination showed a fussy, irritable neonate with bulging anterior fontanellae, macrocephaly, diffuse jaundice and several ecchymotic patches. Laboratory analysis was significant for transaminitis, conjugated hyperbilirubinemia, prolonged PT/ INR and PTT. MRI-head demonstrated diffuse intraventricular hemorrhage with hydrocephalus. She was treated symptomatically for acute liver failure with Vitamin K, Fresh frozen plasma and oral ursodiol. A ventriculoperitoneal shunt was placed and she was monitored closely in the intensive care unit. She underwent further evaluation for neonatal cholestasis. Infectious etiologies were ruled out with negative blood culture, urine culture, cerebrospinal fluid culture and TORCH panel. She had unremarkable alpha fetoprotein, ferritin, urine organic acids, serum amino acids, urine reducing substances, morning cortisol level, free T4 and TSH levels. Hepatobiliary iminodiacetic acid scan (HIDA) was negative for biliary atresia. Serum alpha 1 antitrypsin level was noted to be low (44mg/ dl) and alpha 1 antitrypsin phenotype came back positive for PIZZ phenotype. Liver biopsy revealed cholestasis and hepatocytes with periodic acid Schiff (PAS) positive, diastase resistant globules consistent with the diagnosis of A1ATD. Patient improved symptomatically and was discharged home with close gastroenterology follow-up. Discussion: A1ATD usually presents with neonatal cholestasis. Jaundice clears in most patients by 4 months of age. 2-3 % of these patients develop progressive liver disease with cirrhosis and hepatic fibrosis. Liver damage occurs due to accumulation of the mutated protein within hepatocytes. Patients with neonatal cholestasis can develop Vitamin K deficiency secondary to fat malabsorption and poor oral intake. A1ATD can rarely present with coagulopathy and bleeding diathesis secondary to late onset hemorrhagic disease of newborn (HDN). Exclusively breast fed infants are at higher risk for this presentation as breast milk is a poor source of Vitamin K. Conclusion: Intraventricular hemorrhage and hydrocephalus secondary to late onset HDN can be a presenting symptom for infants with A1ATD. Clinicians should maintain a high index of suspicion and consider A1ATD in the differential diagnoses for patients presenting with neonatal cholestasis and clinical manifestations of late onset HDN.

**373 GRANULOMATOSIS WITH POLYANGIITIS (GPA) PRESENTING AS INFLAMMATORY BOWEL DISEASE (IBD) IN A CHILD WITH AUTOIMMUNE LIVER DISEASE (AILD).** M. Gizaw, University of Pennsylvania, Philadelphia, Pennsylvania, UNITED STATESK. Graham, H.C. Lin, Gastroenterology, Hepatology and Nutrition, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES.

Introduction: Auto-immune liver disease (AILD) and arthritis are well described extraintestinal manifestations of inflammatory bowel disease (IBD). IBD and Granulomatosis with Polyangiitis (GPA) share common symptoms of such as arthralgia, fever, skin abnormalities, and weight loss which can present a clinical challenge when trying to make the correct diagnosis. We describe a case of GPA presenting as IBD in a patient with AILD. Case Report: A 13 year old girl with a history of autoimmune hepatitis (AIH) on azathioprine, primary sclerosing cholangitis (PSC) on ursodiol and juvenile idiopathic arthritis (JIA) presents with a 10 lb weight loss, diarrhea, abdominal pain, and poor appetite. She had a WBC count of 20, CRP 4, ESR 58 and a fecal calprotectin of 858. Results from her colonoscopy showed severe colitis with large ulcers and pathology consistent with chronic active colitis without granulomas. She was started on infliximab, and clinically improved. Two years later she presents with dyspnea, pleuritic chest pain and a non-productive cough. Labs were notable for an ESR 85 and CRP of 1.4. A chest x-ray revealed a right pneumothorax and subsequently chest CT showed mild bronchiectasis, interstitial and peripheral opacities with scattered groundglass opacities and small parenchymal and peripheral cysts. The infectious work-up was negative. Repeat AIH panel remained positive, she was p-ANCA positive and her MP3 proteinase antibody (PR-3) was markedly elevated. Given her clinical symptoms and lab findings, it was determined the unifying diagnosis of her pulmonary, intestinal and joint symptoms was GPA. She was started on induction therapy with high dose methylprednisolone and rituximab with good response. Discussion: IBD and GPA are both associated with ANCA. ANCA in small vessel vasculitis are directed against cytoplasmic antigens such myeloperoxidase (MPO) and PR-3. Approximately 90% GPA patients are seropositive for c-ANCA. There are no known liver complications of GPA and there are very few cases of concurrent AILD and GPA reported in the literature. Recent data suggest cytotoxic T-

lymphocytes antigen 4 (CTLA-4) polymorphisms may have a common pathogenic pathway in the development of autoimmune liver disease and small vessel vasculitis.

**374** *DYSKERATOSIS CONGENITA AND GASTROINTESTINAL BLEEDING: CLINICAL AND ENDOSCOPIC FINDINGS.* K. Queliza, R.W. Himes, T. Miloh, Pediatric Gastroenterology, Hepatology and Nutrition, Baylor College of Medicine, Houston, Texas, UNITED STATES. A. Bertuch, Pediatric Hematology and Oncology, Baylor College of Medicine, Houston, Texas, UNITED STATES.

Background: Dyskeratosis congenita (DC) is a rare telomere disorder characterized by classic mucocutaneous features, bone marrow failure, and cancer predisposition. Gastrointestinal (GI) manifestations include esophageal stenosis, enteropathy, enterocolitis, and portal hypertension (PHTN). GI hemorrhage has been described in isolated case reports though the etiology is still not well understood. In this case series, we describe our institutional experience with four unrelated individuals with DC who experienced GI hemorrhage. Methods/Results: We identified four patients with DC (ages 2-23 years) who presented to Texas Children's Hospital between 2009 and 2016 for GI bleeding (Table 1). All patients had DC confirmed with *TINF2* mutations and underwent bone marrow transplantation for severe bone marrow failure. Hepatic synthetic function was preserved in all cases, and transaminases were mild to moderately elevated (ALT 60-200s). Patients B, C, and D demonstrated features compatible with PHTN (splenomegaly, ascites, thrombocytopenia with platelets 10-140K) in the absence of cirrhosis on liver biopsy. Patients C and D had hepatocyte vacuolar degeneration and portal fibrosis on liver pathology, while patient B had nodular regenerative hyperplasia. Patient C was later found to have PHTN secondary to Budd-Chiari syndrome (progressed to hepatopulmonary syndrome). Despite evidence of varices on angiography in these three patients, none were observed endoscopically. Endoscopic findings and management of their first GI bleed are found in Table 1. All four patients had at least two significant GI bleeding events. Patient D later experienced bleeding despite trialing a beta-blocker and having argon plasma coagulation performed during endoscopy. Both patients C and D were noted to have vascular ectasias in the stomach, duodenum, and/or colon on subsequent endoscopies. Patients A, B, and C are deceased from causes not directly related to GI bleeding. Conclusion: In this case series, the majority of patients had evidence of non-cirrhotic PHTN without overt varices, as well as poor survival not directly associated with GI bleeding. Portal gastropathy may have contributed to bleeding in one case but was not thought to be the definitive source. DC may be associated with disruption of GI mucosal integrity and vascular ectasias, which further predispose to GI bleeding. Heightened suspicion for combined liver/GI pathology in DC patients is essential for targeted management of GI hemorrhage.

**375** *X-LINKED HYPER IGM SYNDROME PRESENTING WITH END STAGE LIVER DISEASE IN INFANCY.* K. Bigaj, University of Miami, Miami, Florida, UNITED STATES. D. Cheung, J. Garcia, D. Goldner, Pediatric Gastroenterology, University of Miami, Miami, Florida, UNITED STATES.

Introduction: X-linked Hyper IgM Syndrome (HIGM Type I) is a rare primary immunodeficiency characterized by defective immunoglobulin class switching leading to antibody deficiency of IgG, IgA and IgE with normal/elevated IgM, as well as a defect in cell-mediated immunity. Liver disease in these patients has been described to occur later in childhood, often in context of chronic infections. However, we describe a patient who, in addition to developing liver disease in infancy, was diagnosed with HIGM after presenting with liver failure. Case: Our patient is an ex 32 week premature infant who had a 3 month NICU course complicated by a left leg bulla requiring incision, drainage and skin grafting, failure to thrive, >2 months total parental nutrition and neonatal cholestasis with coagulopathy. He had a liver biopsy at 3 months which showed neonatal giant cell hepatitis, proliferative bile ducts and intracellular and canalicular cholestasis. He was lost to follow up and presented at 5 months of age with persistent epistaxis. He was in liver failure with an INR of 2.24 unresponsive to vitamin K, portal hypertension with splenomegaly, pancytopenia, direct hyperbilirubinemia of 19.9mg/dL and elevated transaminases. Newborn screen was concerning for tyrosinemia which was ruled out with formal testing. An extensive evaluation ruled out TORCH infections, viral hepatitis, HLH, Hyper IgE, CGD, GALD, hypothyroidism, alpha 1 antitrypsin deficiency, biliary atresia and metabolic disorders. He had an elevated alpha fetoprotein of 665,400 ng/mL (at 5 months), but PET/CT and Oncology evaluation revealed no evidence of malignancy. He developed PCP pneumonia and an immune workup was initiated. He was diagnosed with HIGM based on absent expression of CD40 Ligand, with normal lymphocyte mitogen stimulation. He had negative testing for *Cryptosporidium*, a known cause of liver disease in HIGM patients. Discussion: It has been reported that four-fifths of patients with HIGM develop liver disease by the age of 20 in the form of primary cirrhosis, carcinomas, sclerosing cholangitis and hepatitis. Though rare, HIGM is

the most common cause of immunodeficiency associated sclerosing cholangitis. In the majority of patients, it initially presents as mild cholangiopathy which eventually progresses to cirrhosis, often in the second decade of life. Given our case, HIGM should be considered in the differential of neonatal cholestasis in the setting of abnormal immunoglobulins despite absence of recurrent infections. Bone marrow transplantation (BMT) may be curative, however, it is recommended prior to onset of significant liver disease due to hepatotoxic conditioning regimens. There has been a single report of successful combined liver and bone marrow transplant; our patient is being evaluated for liver transplant followed by subsequent BMT.

**377 THE SUCCESSFUL TREATMENT OF CHRONIC HEPATITIS C GENOTYPE 2 INFECTION WITH SOFOSBUVIR/VELPATASVIR IN A 14 YEAR OLD ADOLESCENT.** K. Almeida, A. Vigni, K. Christensen, K. Hinojosa, M. Castillo, E. Lawitz, N. Alkhouri, Texas Liver Institute, San Antonio, Texas, UNITED STATESK. Hinojosa, CommuniCare, San Antonio, Texas, UNITED STATES.

**Introduction:** The recent FDA approval of the sofosbuvir/ribavirin regimen for the treatment of adolescents with chronic hepatitis C virus (HCV) infection genotype 2 and 3 represents a significant advancement in the field of pediatrics. Although this direct acting antiviral therapy is superior to the old interferon based regimens, it is not the best option currently available for adult patients. Data from the large randomized ASTRAL-2 study indicates that treatment with sofosbuvir/velpatasvir has greater cure rates when compared to sofosbuvir/ribavirin. The sustained virologic response (SVR) for patients treated with sofosbuvir/velpatasvir was 99% while those treated with sofosbuvir/ribavirin achieved only a 94% SVR rate. Also, the use of ribavirin is associated with a more burdensome side effect profile, which can be avoided by using sofosbuvir/velpatasvir.**Case:** A 14 year old male patient was seen at a referral center with a diagnosis of chronic HCV genotype 2b that was acquired vertically. His baseline viral load was 4,521,123 IU/mL, he was treatment naïve, and non-cirrhotic based on HCV Fibrosure test. The child was overall healthy with blood tests resulting in a normal CBC and CMP. He was prescribed a 12 week regimen of sofosbuvir/velpatasvir 400/100mg. The patient tolerated the regimen well all throughout treatment with no side effects or concerns. At each follow up visit he reported feeling well and his accompanying family member confirmed this. At treatment week 4, testing for the HCV RNA resulted in "Target Not Detected" and the viral load remained undetectable throughout treatment. The patient was brought back in to evaluate SVR after the post treatment week 12 time point. The HCV RNA level again resulted in "Target Not Detected" and the patient was declared cured of the virus.**Conclusion:** This pediatric patient was successfully and safely cured with the use of sofosbuvir/velpatasvir. Although the current FDA approved regimen of sofosbuvir/ribavirin provides a formidable cure rate of 94% SVR per the ASTRAL 2 trial, sofosbuvir/velpatasvir is superior at 99% SVR. It also is associated with less side effects due to the absence of ribavirin. Given these benefits, sofosbuvir/velpatasvir should be considered a reasonable alternative to sofosbuvir/ribavirin in the pediatric population, especially for patients that have anemia or psychiatric related co-morbidities that can be exacerbated by a ribavirin containing regimen.

**379 UNUSUAL CFTR MUTATIONS AND LIVER FAILURE IN INFANCY.** H.H. Khan, S. Kaufman, K.M. Khan, Transplant Institute, MedStar Georgetown University Hospital, Washington, District of Columbia, UNITED STATES.

Mutations in the Cystic fibrosis trans-membrane conductance regulator (CFTR) gene especially the delta F 508 account for the majority of disease related to CF. We report presence of CF gene mutations not known to cause disease in infants that developed end-stage liver disease in infancy.**Case 1**. A premature male infant with negative CF screening at birth developed small bowel obstruction in the neonatal period requiring an ileostomy and subsequently cholestatic liver disease with portal hypertension and histology showing neonatal hepatitis. In addition he was noted to have frequent respiratory infections prompting a sweat test which was positive. Genetic testing revealed that he was heterozygous for P.1177F. He underwent a successful liver transplant.**Case 2**. An African-American female infant developed progressive cholestasis with poor weight gain. Investigations included a liver biopsy that showed neonatal hepatitis, negative sweat test though genetic screening showed her to be heterozygous for CFTR and PEX26 genes mutation. She subsequently developed pneumatosis involving the cecum that was treated conservatively and underwent a successful liver transplant.**Case 3**. A male infant developed progressive liver disease with a liver biopsy showing neonatal hepatitis. He was extensively investigated showing a negative sweat test on repeated studies. Genetic testing revealed that the patient was heterozygous P.K186N variant in AKRID1 gene and homozygous for P.R75Q variant in CFTR gene. He remained on the wait list for some time with a portal vein thrombosis that precluded a partial graft. Unfortunately he succumbed to an acute upper GI hemorrhage.**Conclusion:** Rare and unusual CFTR mutations even in the heterozygous form may be a

feature in otherwise undiagnosed endstage liver disease of infancy. More work is required to understand their precise role leading to liver failure in these patients.

**381** *GGT DISCORDANCE IN NEONATAL CHOLESTASIS: A CASE SERIES OF THREE PATIENTS.* K.M. Critelli, P. McKiernan, L. Siebold, D. Squires, J.E. Squires, Pediatric Gastroenterology, Hepatology, and Nutrition, UPMC - Children's Hospital of Pittsburgh, Pittsburgh, Pennsylvania, UNITED STATES. Ranganathan, Pathology, UPMC - Children's Hospital of Pittsburgh, Pittsburgh, Pennsylvania, UNITED STATES.

Background:  $\gamma$ -Glutamyltransferase (GGT) is a microsomal enzyme distributed in tissues involved in secretory and absorptive processes, particularly bile canaliculi. The magnitude of plasma GGT elevations in infants with liver disease can vary with 'low-GGT cholestasis' associated with panhypopituitarism, genetic disorders of bile acid synthesis, hepatic canalicular transport proteins, and tight junction protein 2 (TJP2); the latter being a key component of cell-cell junctional complexes. In contrast, 'high-GGT' cholestasis is typically associated with conditions resulting in intra- and/or extrahepatic biliary injury such as biliary atresia and Alagille syndrome. We describe 3 cases of inharmonious plasma GGT in neonatal cholestasis. Methods: Retrospective chart review was performed on 3 children with neonatal cholestasis and GGT discordance presenting to the Children's Hospital of Pittsburgh. Data was extracted from the medical record. Histology was reviewed by a single hepatopathologist. Results/Cases: *Patient 1*: Term male born to consanguineous parents, presented at 2 weeks of life (WOL) with a direct bilirubin (DB) of 3.2 mg/dL and GGT of 238 IU/L. An abdominal ultrasound (AUS) was normal. Liver biopsy showed cholestasis with ductular reactivity, mild portal and pericentral inflammation, and mild portal and focal sinusoidal fibrosis. Genetic testing revealed a homozygous c.782delA pathogenic variant in the *TJP2* gene. GGT elevation remains at 6 months of life (mean 158 IU/L). *Patient 2*: Term male born in El Salvador, with jaundice that resolved with phototherapy, presented at 16 WOL with recurrent jaundice, direct hyperbilirubinemia, mild elevation of serum aminotransferase levels, leukopenia, anemia, thrombocytopenia, coagulopathy, and a normal GGT (mean 62 IU/L). Echocardiogram revealed peripheral pulmonary artery stenosis (PAS). Liver biopsy showed bile duct paucity with cirrhotic changes. Syndromic features not present included dysmorphic facies, butterfly vertebrae, posterior embryotoxon, and renal anomalies. Genetic testing revealed a novel *JAG1* mutation (c.1395+3A>C). While classified as a variant of unknown significance, a c.1395+3A>G variant has been reported in Alagille syndrome. The patient underwent liver transplant at 2 years of life. *Patient 3*: Term male presented at 7 WOL with DB 8.8 mg/dL, elevated serum aminotransferase levels, and normal GGT (90 IU/L). Echocardiogram showed mild peripheral branch PAS. An AUS showed a small gallbladder and mild splenomegaly. HIDA scan confirmed biliary excretion into the intestine. Chest and thoracolumbar X-rays and ophthalmology examination were normal. Facies suggestive of Alagille Syndrome were present. Genetic testing demonstrated pathogenic *JAG1* mutation (c.2096\_2100delGAAAAG) confirming Alagille. Conclusion: We report on 3 cases of GGT discordance in neonatal cholestasis, 1 with a novel *JAG* pathological variant. While GGT is useful for differentiating neonatal cholestatic syndromes, these cases emphasize the limitations of exclusively relying on GGT to categorize certain liver disorders. Rather, laboratory assessment of the patient with suspected or clinically obvious liver disease is context dependent, and our differential diagnosis of neonatal cholestasis should remain broad.

**382** *CASE SERIES- TREATING CHILDREN WITH HEPATITIS C 1A WHO ARE TREATMENT-EXPERIENCED AND CIRRHOTIC WITH LEDIPASVIR-SOFOSBUVIR UNDER COMPASSIONATE USE.* K.M. Reed, J. Gamblin, J. O'Connor, S. Palle, Pediatrics, OU Childrens, Oklahoma City, Oklahoma, UNITED STATES.

Purpose of Study: Despite significant advances in landscape of treatment for adults with Hepatitis C virus (HCV) infections, there is limited data concerning those same therapies in children. For pediatric patients, PEG-IFN- $\alpha$  and Ribavirin (PEG-IFN/RBV) still used as first-line. Multicenter studies have shown that in children with genotype 1 HCV infections, only a 53% sustained virological response (SVR) using this therapy. We present three cases of vertically acquired HCV-genotype 1a infections and their response to therapy with ledipasvir-sofosbuvir (LDV/SOF), after previously failed therapy with PEG-IFN/RBV, within a single center under compassionate use. The aim is to increase awareness of outcomes of children treated with LDV/SOF as we await results from pediatric HCV drug trials. Methods Used: Three patients with HCV-Type 1a genotype who had failed previous PEG-IFN/RBV treatment, continued to have progressive liver disease and received therapy with LDV/SOF were studied. Demographic, Clinical, laboratory, and treatment characteristics were analyzed. Summary of Results: All patients completed their treatment course without significant side effects. All three had a rapid virological response. One patient has had a

sustained virological response (SVR12) and the other two will be tested at 12 weeks post-treatment (as shown in the table below) Conclusions: Current first-line FDA approved therapy for children with PEG-IFN/RBV has a long course, significant side effects, and poor response rates for those especially with HCV genotype 1a infection. In this case series we show successful re-treatment of pediatric HCV 1a infected children using LDV/SOF. Each patient tolerated their treatment course and had viral clearance by treatment week 4 and all subsequent testing has remained negative

**383** *HEPATOPULMONARY SYNDROME ASSOCIATED WITH ABERNETHY MALFORMATION.* L. Ng, J. Mait-Kaufman, Pediatric Gastroenterology, Cohen Children's Medical Center of NY, Lake Success, New York, UNITED STATES. H. Kholwadwala, Pediatric Cardiology, Cohen Children's Medical Center of NY, New Hyde Park, New York, UNITED STATES.

Hepatopulmonary syndrome is an uncommon clinical syndrome affecting patients of all ages with liver disease. We present a case of hepatopulmonary syndrome associated with Abernethy malformation in a medically complex patient. 16 year old female with a complex medical history inclusive of anoxic brain injury, abdominal heterotaxy with intestinal malrotation status post repair as infant, repaired transitional AV septal defect, hypothyroidism and peptic ulcer disease, was found to have persistent hypoxia and digital clubbing on routine follow up. Bloodwork was notable for a hemoglobin of 14.8mg/dL, concerning for developing polycythemia from presence of chronic hypoxemia. She then was admitted for cardiac catheterization, which revealed multiple micro-arteriovenous malformations in bilateral lung fields with a right to left shunt that was consistent with pulmonary arterio-venous malformations. Abdominal MRI and CT subsequently revealed the presence of a congenital extrahepatic portosystemic shunt, associated with heterotaxy, polysplenia and non-visualization of the intrahepatic portal vein. These findings were consistent with a diagnosis of Hepatopulmonary syndrome (HPS) with Abernethy malformation. Hepatopulmonary Syndrome is characterized by 1) defect in arterial oxygenation, 2) intrapulmonary vascular dilatations, and 3) advanced liver disease, portal hypertension or congenital portosystemic shunts, such as cavopulmonary shunts and Abernethy malformation. Abernethy malformation is rare congenital vascular malformation defined by diversion of portal blood away from liver, into the systemic circulation. Non-invasive cross-sectional imaging techniques, such as CT or MRI, may demonstrate abnormal portal circulation, hence being the diagnostic mode of choice. No effective medical therapies for HPS currently exist, and liver transplantation is the only effective treatment proven to improve survival rates. In patients with HPS and Abernethy malformation, treatment often depends on the type of shunt, associated congenital anomalies and the extent of liver disease. Our patient was treated at a liver transplantation center where an interventional radiological study revealed a hypoplastic portal vein and a portorenal shunt; the portorenal shunt was successfully balloon occluded with an Amplatzer vascular plug, after which time adequate flow was demonstrated into the portal vein, thereby restoring portal blood flow to the liver. Follow-up sonogram with doppler now demonstrates a patent portal vein with normal hepatopedal flow.

**384** *TPNOMA" AS A RARE COMPLICATION OF LIPID EXTRAVASATION WITH UVC MISPLACEMENT: A CASE SERIES."* L.M. Alfaro Cruz, K. Barton, P.M. Hertel, B.A. Carter, D.S. Fishman, Pediatric Gastroenterology, Baylor college of Medicine/Texas Children's Hospital, Houston, Texas, UNITED STATES. E.M. Bergner, J.A. Garcia-Prats, Neonatology, Baylor College of Medicine/ Texas Childrens Hospital, Houston, Texas, UNITED STATES.

Introduction Umbilical venous catheterization (UVC) is commonly performed in premature infants to obtain central intravenous access. Placement requires proficiency and experience to avoid complications that can lead to morbidity and mortality (e.g. infection). We describe 2 cases where UVC misplacement lead to a cystic infiltrative lesion in liver followed by cholestasis. Case 1A full term female infant born at 38 weeks to a diabetic mother presented with neonatal hypoglycemia and respiratory distress. The patient was intubated and UAC/UVC lines were placed, and was then started on empiric antimicrobials. She was made NPO, and TPN was initiated. At 24 hours of life her direct bilirubin was 0.3 mg/dl, but increased to 3.7 mg/dl on DOL 3. Aminotransferases were also elevated: ALT 622 U/L and AST 277 U/L. Abdominal ultrasound showed hepatomegaly and a cystic lesion in the right hepatic lobe containing a complex fluid collection (Figure 1a). On DOL 4 patient was transferred to our center. Repeat imaging showed an enlarging complex fluid collection in the liver involving both hepatic lobes. The UVC catheter was removed and TPN discontinued. The conjugated bilirubin decreased to 1.2 mg/dl, as did the AST/ALT to 97/65 U/L respectively. Ultrasound showed decreased size of the heterogeneous intrahepatic lesion, and the patient was discharged on DOL 40. Case 2A 34 week twin born to a mother with premature rupture of membranes

presented with respiratory distress requiring continuous positive airway pressure. UVC catheter was inserted. TPN and intralipids were started with enteral feeds on DOL 2. Intralipids were discontinued on DOL 4. At 24 hours of life, the total bilirubin was 3.9. On DOL 5, the patient developed abdominal distension, apnea, bradycardia and lethargy. Labs revealed anemia, thrombocytopenia, lactic acidosis, hyponatremia, and conjugated bilirubin of 0.5 mg/dL and AST/ALT of 995/205 respectively. Abdominal and chest radiograph revealed a low-lying UVC at T10-T11 level. An abdominal US revealed a 5cm round multi-septated cystic structure in the posterior right hepatic lobe, UVC in the liver and diffuse ascites. A CT abdomen (Figure 1b) confirmed the lesions and had corresponding fat signal. On DOL 7, the patient continued to deteriorate requiring intubation and the patient was transferred to our center for coagulopathy and increasing size of liver lesions. Repeat imaging with Doppler revealed reversal of flow within the left portal vein. Conjugated bilirubin peaked at 21.3mg/dL, AST 4373 at U/L, ALT 1158 at U/L. Hospitalization was complicated by prolonged intubation secondary to competitive abdomen from significant ascites, feeding intolerance requiring prolonged TPN use, adrenal insufficiency, and catheter associated thrombi. After weeks of supportive care, abdominal distension improved and the infant was able to tolerate enteral feeds. Patient was discharged on DOL 88. Recent labs and imaging showed improvement in AST 181 U/L, ALT 137 U/L, conjugated bilirubin 4 mg/dL, and decreasing size of the hepatic collection (2.8cm). Discussion UVC placement is a common neonatal procedure that carries a risk of significant morbidity and mortality in up to 25% of cases. Infection, hepatic necrosis, portal vein thrombosis, arrhythmia, and extravasation of fluids are the most common complications. TPN hypertonicity with elevated pH causes chemical irritation, which can lead to extravasation of fluid from the vasculature with external or internal compression of other structures. Factors affecting risk of catheter malposition include physician experience, technique of insertion, and type of parenteral infusion. In these cases, extravasation of TPN led to formation of a cystic lesion (likely containing fat) in the liver, with secondary cholestasis and hepatic inflammation. Although liver injuries post-UVC have been described with frequency approaching around 100 cases worldwide, TPN fat accumulations are rarely described. Distinct from hematomas and related cysts, only one prior case of TPN fat accumulation has been reported, which the presence of blood and TPN fat was confirmed by US guided fine needle aspiration. Conclusion UVC placement can lead to serious liver injuries that are not only related to the procedure itself but also to the content infused. Although rare, TPN fat accumulation can have a varied clinical presentation, it can lead to significant liver disease if not promptly recognized and corrected.

**385 HEPATIC ADENOMA IN A PATIENT WITH KABUKI SYNDROME.** L.H. Gutierrez Sanchez, K. Furuya, Pediatric Gastroenterology, Mayo Clinic, Rochester, Minnesota, UNITED STATES. Kolbe, Radiology, Mayo Clinic, Rochester, Minnesota, UNITED STATES. Babovic-Vuksanovic, Clinical Genomics, Mayo Clinic, Rochester, Minnesota, UNITED STATES. Zhang, Pathology, Mayo Clinic, Rochester, Minnesota, UNITED STATES.

Introduction: Kabuki syndrome (KS) is a rare disorder with an estimated incidence of 1 in 32,000 newborns. Mutations in the KMT2D or KDM6A genes have been associated with this disorder. It has an autosomal dominant pattern of inheritance for the KMT2D gene and x-linked dominant for the KDM6A gene. Most patients have anomalies that include distinctive faces, skeletal abnormalities, intellectual disability and growth deficiency. Feeding difficulties, visceral malformations, cleft palate, cardiac defects and seizures are also common. No clear predisposition to cancer is known. There have been case reports of patients with KS and acute lymphoblastic leukemia, neuroblastoma, hepatoblastoma, Burkitt's lymphoma and fibromyxoid sarcoma. We present a case of a child with KS and multiple hepatic adenomas. Case report: 14 year old female with KS, with a heterozygous mutation in exon 31 of the MLL2 gene, who was found to have a liver mass on a follow up cardiac echocardiogram. Upon presentation she was asymptomatic from a liver stand point. She had no jaundice, pruritus, easy bleeding, changes in mental status, acholic stools, hematochezia or melena. Initial blood work included Hgb 13.1 g/dL, WBC  $5.9 \times 10^9$ /L, platelets  $272 \times 10^9$ /L, Na 139 mmol/L, K 4.3 mmol/L, Alk Phos 176 U/L, AST 65 U/L, ALT 50U/L, total bilirubin 1.2 mg/dL, direct bilirubin 0.6 mg/dL, GGT 85U/L, Cr 0.4mg/dL, alpha fetoprotein 0.7 ng/mL, CA19-9 8 U/mL, CEA 0.5ng/mL. Screening tests for Wilson disease, alpha-1-antitrypsin deficiency, viral hepatitis and autoimmune hepatitis were negative. As part of her evaluation she had an abdominal MRI with and without contrast which showed a large heterogenous mass (15 x 11 x 14.5 cm) within the left hepatic lobe with retention of hepatobiliary contrast agent. Abnormal background liver parenchyma with multiple other similar-appearing smaller masses in both hepatic lobes was also noted. She underwent an US guided percutaneous liver biopsy; histology demonstrated an atypical hepatic adenoma with suspicion of early malignant transformation in her left lobe. The parenchyma of her right lobe demonstrated fibrosis without a definite etiology. The patient has

been listed for transplant. Discussion: KS is a rare disorder which can involve multiple organs; there is no clear relationship between this disorder and malignancies. However, mutations in the MLL2 gene (also known as KMT2D gene) have been associated with in vitro genome instability which can lead to tumorigenesis. In this case, we describe a new malignancy that has never been reported in KS patients. The number of cases in the literature is very low but one can consider screening strategies for these patients due to their likely increased risk of malignancy.

**386 LIVER CIRRHOSIS IN A 7-YEAR OLD MALE LEADING TO A RARE DIAGNOSIS.** M. Cohen, Pediatrics, Cleveland Clinic, Beachwood, Ohio, UNITED STATES. Radhakrishnan, P. Conjeevaram Selvakumar, Pediatric Gastroenterology, Cleveland Clinic Childrens, Cleveland, Ohio, UNITED STATES. Patil, Pathology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES. Gupta, Radiology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES. Lakshmanan, Cleveland Clinic, Cleveland, Ohio, UNITED STATES.

Background: Primary malignant liver tumors in childhood are rare and represent only 1-2% of all pediatric tumors. Hepatocellular carcinoma (HCC) is the second most common primary hepatic malignancy in children after hepatoblastoma. Here we describe a case of a 7 year old male child who was diagnosed with HCC with no identifiable etiology. Case Presentation: A 7 year old male, with no significant medical history, was admitted for 10 days of periumbilical abdominal pain following acute gastroenteritis and fevers. He had significant abdominal distension and tenderness on physical exam. Initial labs were significant for platelet count of 73 k/uL, INR of 1.6, alanine transaminase (ALT) of 65 U/L and aspartate transaminase (AST) of 88 U/L. Abdominal X-ray showed mildly dilated loop of bowel suggestive of ileus. CT abdomen was significant for liver cirrhosis a small hypodense lesion in right lobe of liver and splenomegaly. Liver ultrasound showed a heterogenous liver with irregular contour consistent with cirrhosis. Evaluation for infectious causes including viral hepatitis (A, B and C), Cytomegalovirus and Epstein-Barr virus were negative. Evaluation for autoimmune hepatitis, alpha-1 antitrypsin deficiency and Wilson disease were also negative. Alpha fetoprotein was obtained due to small lesion seen in CT abdomen and was significantly elevated at 5493.6 ng/ml. Due to significantly elevated AFP, MRI abdomen with HCC protocol was performed which showed cirrhotic liver with innumerable regenerative nodules and 2 distinct lesions of the right hepatic lobe demonstrated arterial enhancement and subsequent washout of contrast on delayed imaging with T2 hyperintensity, concerning for dysplastic nodules. Transjugular liver biopsy revealed regenerating nodules surrounded by thick fibrous bands indicative of cirrhosis. Due to HCC and cirrhosis, patient received orthotopic liver transplant with excellent post-operative course. The histology of explanted liver showed moderately differentiated HCC with no lymphovascular invasion. Discussion: HCC accounts for about one-third of primary hepatic malignancies in children and about 0.5% of all pediatric malignancies. The majority of HCC cases arise de novo in children without preexisting liver diseases. Risk factors include chronic viral hepatitis, progressive familial intrahepatic cholestasis type 2, hereditary tyrosinemia, type I glycogen storage disorder and uncorrected congenital porto-systemic shunts. Children often present with advanced disease and larger tumor size compared to adults, likely due to delayed detection due to vague nonspecific symptoms. Diagnosis of pediatric HCC is mostly made with CT or MRI of abdomen with contrast which shows arterial enhancement of the lesion with washout during the venous phase with T2 enhancement on MRI with significantly elevated AFP. Since HCC is relatively chemoresistant, surgical resection is preferred in children with no metastasis. Treatment strategies for metastatic disease and unresectable tumors include neoadjuvant chemotherapy, ablation and transcatheter arterial chemoembolization but prognosis remains poor.

**390 SUCCESSFUL TREATMENT OF ABERNATHY MALFORMATION WITH LIVER TRANSPLANTATION: A CASE REPORT.** M. Ayers, Pediatrics, Cleveland Clinic, Cleveland, Ohio, UNITED STATES. Mamoun, Radiology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES. Radhakrishnan, P. Conjeevaram Selvakumar, Pediatric Gastroenterology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES.

BACKGROUND: Abernathy malformation (AM) is an extremely rare congenital extrahepatic portosystemic shunt (CEPS) where portomesenteric blood drains into systemic circulation bypassing liver through a complete or partial shunt. Owing to the rarity, literature on AM is limited with most of our current knowledge coming from case reports/series. Here in we report a child who was found to have AM and was successfully treated with liver transplantation (LT). CASE: An 18-month-old male presented to our center for evaluation and management of known AM diagnosed at birth. He was born full term and at birth, he had generalized edema with no dysmorphic features. Neonatal echocardiogram and abdominal ultrasound obtained for evaluation showed evidence of CEPS with no

other congenital anomalies. He was being managed with lactulose and low protein diet. Despite this, he was admitted multiple times for dehydration and hyperammonemia in the setting of illness approximately every 3-4 months. In our center, his portal circulation anatomy was evaluated with MR angiogram which revealed absence of the extrahepatic and intrahepatic portal vein (PV). The splenic vein (SV) and superior mesenteric vein (SMV) formed a common trunk which drained directly into inferior vena cava (IVC). He was listed for liver transplant due to his repeated hyperammonemia episodes and the future risk of hepatic nodules and dysplasia. Consequently, he had a successful orthotopic LT at the age of 3 years. Subsequent imaging studies indicated appropriate blood flow in portal circulation with excellent clinical outcome.

**DISCUSSION** First reported in 1793, only about 80 cases of AM have been described. There are 2 types of AM: In type I, there is congenital absence of PV with SV and SMV draining into IVC either separately or after forming a common trunk. In type II, PV is intact with partial shunting to IVC by a side-side shunt. Accordingly, our patient had type 1 AM. Clinical presentations are variable which include neonatal hypoglycemia, cholestasis, hepatic encephalopathy and hepatopulmonary syndrome. Type 1 AM is also associated with other congenital anomalies which are not seen in our patient. Diagnosis of AM is initially made with US Doppler and confirmed with MR/CT angiography. Type 1 AM is associated with hepatocellular carcinoma and hepatoblastoma if left uncorrected. Treatment is limited to LT in type 1 AM as in case of our patient whereas shunts could be coiled surgically or percutaneous transcatheter approach in type 2 AM.

**397 A CASE OF SPONTANEOUS COMMON BILE DUCT (CBD) PERFORATION.** M. Halabi, D. Ashok, N. Merritt, Paediatrics, Children's Hospital - London Health Sciences Centre, London, Ontario, CANADA.

**Background** Spontaneous common bile duct (CBD) perforation is a rare cause of abdominal pain. It is mainly seen in children. The etiology is not completely understood, however theories included: congenital weakness of the CBD, tumors of the bile duct, choledochal cyst, and pancreatic reflux. We report the presentation of a child with spontaneous CBD perforation and subsequent management.

**Case Report** A 22-month-old (ex 25+2 week's gestation) female presented with a one-month history of abdominal pain, vomiting, abdominal distension and jaundice to her local paediatrician. Investigations were performed which showed a conjugated hyperbilirubinemia and large ascites on ultrasound. On examination the abdomen was visibly distended, tense and tender to palpation. Initial metabolic, and hepatic blood work was normal. An MRI of the chest and abdomen was normal. A paracentesis with insertion of peritoneal drainage catheter, and liver biopsy was performed. Paracentesis demonstrated biliary ascites. Hence a biliary scintigraphy was performed which showed a bile leak. She developed compartment syndrome and intra-abdominal sepsis. An urgent exploratory laparotomy was performed due to clinical deterioration despite treatment with broad-spectrum antibiotics and percutaneous drainage of bile. On laparotomy, a large amount of bile-stained ascitic fluid was drained with large amount of inflammatory reaction and fibrinous deposits at the porta hepatis. She was managed conservatively with 3 weeks of external intraabdominal drains. A transhepatic cholangiogram was performed two weeks post operatively, which, demonstrated no extravasation or biliary leak. Follow up at 6 months showed complete recovery.

**Discussion** Very few cases of spontaneous CBD perforation have been described in the literature. It is most commonly seen in children within the age range of 25 weeks to 7 years of age, with a peak incidence age of 6 months old. Patients may present with sub-acute (80%) or acute (20%) symptoms of abdominal pain, jaundice, and abdominal distension. Management can vary from simple drainage, suture closure of defect and/or cholecystectomy. However conservative management, as we have used in our case has been shown to have successful outcome.

**Conclusion** In conclusion, due to the rarity of spontaneous CBD perforation, delayed diagnosis and intervention is common. Physicians must consider the diagnosis in infants and children, if biochemical analysis of ascitic fluid confirms bilious ascites. This will prompt timely diagnosis by non-invasive biliary scintigraphy and management to prevent further complications.

**398 SUCCESSFUL USE OF SILIBININ IN A CHILD WITH MUSHROOM-INDUCED ACUTE LIVER FAILURE.** M. Kabbany, P. Conjeevaram Selvakumar, C. Pasquarella, K. Radhakrishnan, Department of Pediatric Gastroenterology and Hepatology, Cleveland Clinic Foundation, Cleveland, Ohio, UNITED STATES.

Mushroom poisoning is an uncommon cause of acute liver failure in children. *Amanita phalloides* is the most common cause of fatal mushroom poisoning with high mortality and morbidity up to 50%. It exerts its effect through Amatoxin; a toxin that binds to DNA-dependent RNA polymerase II causing inhibition of protein synthesis. Tissues with high rates of protein synthesis are most affected leading to the initial gastrointestinal symptoms and later hepatic and renal failure. Amatoxin can also induce a TNF- $\alpha$  mediated hepatocyte apoptosis. About 60% of

absorbed Amatoxin is excreted into bile and undergoes enterohepatic circulation increasing the hepatocytes exposure. We report a case of a 9 year old Nepali immigrant male who presented to the emergency department along with other family members with abdominal pain, vomiting and diarrhea. Symptoms started 2 days after mushroom ingestion. Initial laboratory findings showed AST >7000 (Normal 5-50 U/L), ALT >7000 (Normal 5-50 U/L), total bilirubin 2 (Normal 0-1.5 mg/dL), direct bilirubin 1.1 (Normal 0-0.4 mg/dL), ammonia 30 (Normal 17-47  $\mu$ mol/L) and INR 2.3 (Normal 0.8-1.2) which increased to 3.8 within 24 hours of presentation. Patient continued to be awake and alert with no signs of hepatic encephalopathy or active bleeding. Patient received aggressive IV hydration along with Octreotide drip, Penicillin G, N-Acetylcysteine and vitamin K. Emergent permission to use Silibinin from our institution's institutional review board was obtained and the patient received the medication 12 hours after presentation as an IV bolus of 5 mg/kg followed by continuous infusion of 20 mg/kg/24 hour for 72 hours. Within 12 hours of starting Silibinin, his labs were downtrending with INR 3.2, AST 4554 and ALT 5927. The INR normalized by day 3 of admission. Patient was discharged after 10 days with almost normal liver enzymes. Silibinin is an antidote that is undergoing FDA approval process. It blocks the enterohepatic circulation of the Amatoxin as well as ameliorates the TNF- $\alpha$  mediated hepatocyte apoptosis. Only few case reports on using this medication in children exist, most of which show efficacy. In conclusion, early recognition of mushroom poisoning as well as early administration of the antidote is crucial for the outcome in these patients. We would like to acknowledge Dr. Todd Mitchell and Madaus Inc. for their help obtaining the medication.

**400 BENIGN INFLAMMATORY BILIARY STRICTURES IN INFANTS: NOVEL APPROACHES WITH NON-SURGICAL INTERVENTIONS.** P. Reddy, Y. Rivas, D. Kogan, N. Ovchinsky, Pediatrics, Montefiore Medical Center, New York, New York, UNITED STATES. Golowa, Radiology, Montefiore Medical Center, Bronx, New York, UNITED STATES. Ho, Medicine, Montefiore Medical Center, Bronx, New York, UNITED STATES.

Background: Benign idiopathic biliary strictures are exceedingly rare in infants. Historically, these cases are managed surgically, however less invasive percutaneous and endoscopic treatments may be promising. Here, we present a report of two infants with benign inflammatory biliary strictures that were managed with a minimally invasive approach. Case 1: A 5 month old boy with history of microcephaly and hypoxic ischemic encephalopathy presented with jaundice and acholic stools. Initial work-up revealed: total bilirubin (TB) 12.4 mg/dL, direct bilirubin (DB) 9.7 mg/dL, Aspartate Aminotransferase (AST) 184 U/L, Alanine Aminotransferase (ALT) 148 U/L, and Gamma Glutamyl Transferase (GGT) 732 U/L. Ultrasound (US) and MR Cholangiopancreatography showed diffuse intrahepatic bile duct dilatation suggestive of biliary stricture. Endoscopic retrograde pancreatography (ERCP) was attempted, however cannulation of the common bile duct (CBD) was unsuccessful. A percutaneous transhepatic cholangiogram (PTC) demonstrated a stricture of the common hepatic duct (CHD), and a catheter was left in place for external drainage to allow the decompression of the biliary system. Subsequently, a combined PTC and ERCP procedure was performed, which had been described as a "Rendezvous technique." The CBD was easily cannulated and a stent was placed via endoscopy with removal of biliary drain. Patient's laboratory values normalized 6 weeks post-procedure. Resolution of ductal dilatation was confirmed on US. Case 2: A 3 month old girl presented with acholic stools and scleral icterus. Initial work-up revealed: TB 6.7 mg/dL, DB 4.3 mg/dL, AST 121 U/L, ALT 92 U/L, and GGT 1120 U/L. After a Hepatobiliary Iminodiacetic Acid (HIDA) scan showed no excretion into the small bowel, an intraoperative cholangiogram was performed for concern of biliary atresia. Dilatation of the right and left hepatic bile ducts with a stricture of the CHD and CBD was seen. CT scan was significant for an ill-defined mass within the porta hepatis surrounding a strictured duct. Subsequently, a PTC was performed at our liver center confirming a long segment stricture involving the confluence of the right and left main hepatic ducts. An internal/external biliary drain was placed, followed by serial balloon dilations of the stricture. Biopsy of the surrounding mass revealed fibrosis and collagen deposition with inflammatory cells. Staining for CD-31 to suggest endothelioma and for CD1A and S-100 to suggest histiocytosis were negative. The drain was removed after a cholangiogram demonstrated patent biliary system and appropriate intrabiliary ductal pressures by biliary stress manometry test. Laboratory values normalized 3 months post-procedure and remain normal at 9 months follow-up. US demonstrated complete resolution of the intrahepatic ductal dilatation and of the hypoechoic mass in the porta hepatis. Discussion: We present these cases of benign inflammatory biliary strictures in infants and highlight similarities in presentation to other causes of obstructive cholestasis including biliary atresia and extrinsic compression. Importantly, less invasive percutaneous and endoscopic interventions, including a novel rendezvous technique, may be utilized to treat these patients with excellent outcomes.

**401 EBV ASSOCIATED MALIGNANCIES IN CARDIAC PATIENTS WITH PROTEIN LOSING ENTEROPATHY.** S. Rudra, H.C. Lin, Pediatrics, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES.

Introduction: Protein losing enteropathy (PLE) is a complication in single ventricle patients after Fontan surgery due to altered circulation physiology. We discuss two cases of adolescents with cardiac disease and PLE who developed EBV-associated malignancies. Case Presentations: A 14-year-old with transposition of great arteries, VSD, coarctation of aorta, and chronic lung disease was admitted for abdominal pain, food refusal, and exacerbation of bronchiectasis. Work-up was significant for hypoalbuminemia (2.3 g/dl), elevated stool alpha-1 antitrypsin, T-cell immunodeficiency, and worsening pulmonary function tests. Chest CT was obtained and showed pulmonary and liver nodules confirmed by MRI. Liver biopsy of four liver nodules were positive for EBV-associated diffuse large B-cell lymphoma. Treatment included chemotherapy for lymphoma, Entocort and furosemide for PLE, and IVIG for T-cell immunodeficiency. An 18-year-old with double outlet right ventricle status post Fontan, mitral and pulmonary atresia, and PLE was admitted for daily fevers, diarrhea, pedal edema and fatigue. She had hypoalbuminemia (2.9g/dL), hypoproteinemia (4.8g/dL), low IgG level, and EBV PCR with 2000 copies. CT imaging showed hepatosplenomegaly and lymphadenopathy (mesenteric, retroperitoneal, supraclavicular). Bone marrow and lymph node biopsies were positive for EBV-associated Hodgkin's lymphoma. Treatment included chemotherapy for lymphoma; Entocort, furosemide and albumin for PLE; and IVIG for immunodeficiency. Conclusion: These two cases highlight concern that PLE gives rise to lymphocyte dysregulation increasing malignancy potential. Clinicians should assess for complications of PLE including, but not limited to, immunodeficiency disorders and EBV-associated malignancies in patients with cardiac etiologies.

**403 ESOPHAGEAL INTRAMURAL PSEUDODIVERTICULOSIS WITH TRACKING IN A CHILD WITH AUTOSOMAL DOMINANT HYPER-IGE SYNDROME.** V. Ramachandran, K.P. Shah, Baylor College of Medicine, Houston, Texas, UNITED STATES. S. Fishman, Pediatric Gastroenterology, Hepatology and Nutrition, Baylor College of Medicine, Houston, Texas, UNITED STATES. F.C. Lee, Pediatrics, Baylor College of Medicine, Houston, Texas, UNITED STATES. F.O. Seeborg, D.S. Fishman, Texas Children's Hospital, Houston, Texas, UNITED STATES. F.O. Seeborg, Immunology, Allergy, and Rheumatology, Baylor College of Medicine, Houston, Texas, UNITED STATES.

Introduction: Esophageal intramural pseudodiverticulosis (EIP) is a condition in which the submucosal excretory glands of the esophagus form outpouchings, or, pseudodiverticuli. It is thought to be due to obstruction or dilation of the submucosal excretory glands, and has been associated with chronic esophageal inflammation and motility disorders. Patients most commonly present with dysphagia, and EIP carries a small risk of perforation that could result in mediastinitis. Case: An 11 year old boy with history of chronic lung infections leading to severe bronchiectasis, eczema, and skin abscesses presents from Saudi Arabia for workup of a suspected primary immunodeficiency disorder. He was diagnosed with autosomal dominant Hyper-IgE syndrome (AD-HIES) and incidentally noted to have pneumomediastinum with esophageal pneumatosis on CT scans that worsened over a 3 month period. Barium esophagrams were significant for air in the esophageal wall and bilateral intramural tracking of contrast (Figure 1). Endoscopic evaluation showed a pseudodiverticulum at 24 cm, as well as esophagitis, gastritis, and duodenitis with eosinophilia and budding yeast seen on pathology evaluation. Cultures of biopsies grew *Candida glabrata* and *Saccharomyces cerevisiae* and a plan was made for long-term anti-fungal therapy. The patient returned to Saudi Arabia before appropriate anti-fungal treatment could be initiated, and eventually underwent a lung transplant for severe bronchiectasis. Conclusion: While AD-HIES is known to be associated with fungal infections of the GI tract, few other gastrointestinal complications have been documented in adult or pediatric literature. EIP with tracking has rarely been documented in the pediatric population. Recognizing EIP in children is important as it carries a risk of perforation.

**407 HELICOBACTER PYLORI AND HEPATITIS A COINFECTION COMPLICATED BY ACALCULOUS CHOLECYSTITIS IN AN ADOLESCENT REFUGEE.** J. Tumba, C. Kim, Medical Student, University of Illinois College of Medicine in Rockford, Rockford, Illinois, UNITED STATES. K. Nguyen, Division of Pediatric Gastroenterology, Mercy Health, Rockford, Illinois, UNITED STATES.

*Helicobacter pylori* (*H. pylori*) is the primary cause of peptic ulcers and is linked to gastric cancer. *H. pylori* prevalence in southern and eastern Europe, South America, and Asia is greater than 50%. The isolation of *H. pylori* from feces, dental plaque, and saliva suggest fecal-oral transmission, but the mode of transmission has not been proven and continues to be a controversial topic. Several recent studies have used Hepatitis A virus (HAV) as a sensitive marker of fecal-oral transmission and propose that parallel HAV and *H. pylori* seropositivity rates may

support fecal-oral mode of transmission for *H. pylori*. Unfortunately, these studies fail to acknowledge the differences in immunological responses between *H. pylori* and HAV infections, and therefore impact the interpretation of seropositivity. We present a case of an Iraqi refugee with hepatitis A and *H. pylori* coinfection, presenting with acute abdominal pain, jaundice, vomiting and elevated liver enzymes. Further investigation with right upper quadrant ultrasound and magnetic resonance cholangiopancreatography revealed acalculous cholecystitis. This case may lend support to the fecal-oral mode of transmission for *H. pylori*. To our knowledge, this is the first case report of Hepatitis A complicated by acalculous cholecystitis and symptomatic *H. pylori* infection.

**412 A UNIQUE CASE OF A 7 YEAR OLD WITH HEMATEMESIS AND GASTRITIS ASSOCIATED WITH YERSINIA ENTEROCOLITICA INFECTION.** K. Mudambi, R. Bensen, Pediatric GI, Stanford, Sunnyvale, California, UNITED STATES. *Yersinia enterocolitica* is a gram-negative bacillus shaped bacterium, primarily animal-borne and acquired by ingestion of contaminated water, milk or meat. In humans, infection predominantly leads to self-limited symptoms of enterocolitis or terminal ileitis. This typically presents as a diarrheal illness and can include hematochezia. It can occasionally cause symptoms of appendicitis, known as "pseudo-appendicitis" through invasion of Peyer's patches in the ileum, causing focal inflammation and abdominal pain in the right lower quadrant. There is a single case report in the literature of severe upper gastrointestinal (GI) symptoms associated with *Yersinia* infection in a 53-year-old woman who presented with abdominal pain and nausea, found on endoscopy to have multiple erosions, ulcers, and pus-like lesions in the gastric antrum, which was cultured positive for *Yersinia enterocolitica*. We are not aware of any case reports illustrating upper GI manifestations of *Yersinia* infection in children. Here we present a unique case of a child who presented with hematemesis without diarrhea, and on endoscopy was found to have histological and gross evidence of diffuse upper GI inflammation, with evidence of elevated stool inflammatory markers (calprotectin) and protein loss in stool (elevated Alpha-1 Anti Trypsin.) He was started on PPI therapy, and upon discharge was found to have *Yersinia enterocolitica* in his stool. After follow up in clinic several weeks later he was completely asymptomatic and had improved stool markers, along with interval weight gain. Although this clinical scenario is rare, *Yersinia* can be considered in the differential diagnosis of nausea, vomiting and hematemesis as a possible cause of gastritis in an otherwise healthy child, even without diarrhea or typical right lower quadrant abdominal pain.

**415 CASE REPORT: ATYPICAL PRESENTATION OF HENOCHE SCHONLEIN PURPURA.** L.R. Rhodes, Pediatric Gastroenterology, Louisiana State University Health Sciences Center New Orleans, New Orleans, Louisiana, UNITED STATES.

Introduction: This is the case of a patient who initially presented to an outside Emergency department (ED) with acute onset of peri-umbilical pain with radiation to the right lower quadrant. Further presentation and workup lead to the diagnosis of Henoch Schonlein Purpura (HSP). Case description: 12 y/o Caucasian male presented twice to an outside ED with peri-umbilical pain with radiation to right lower quadrant initially suspected to be an acute appendicitis. At the second ER visit, he had a low grade fever of 100.4F, white blood cell count at 16.1, ESR at 7, and CRP elevated at 2.98. A CT scan of the abdomen/pelvis with contrast revealed mildly distended small bowel loops, abnormal free fluid within the lower pelvis, and poor visualization of the appendix that was normal in caliber. Surgery evaluated patient and performed aspiration drainage of the free pelvic fluid. Surgery further gave impression that clinical picture was not consistent with an abscess or appendicitis. Patient was admitted and started on antibiotics. Following admission patient developed dark, maroonish colored stools. Adult Gastroenterology was consulted due to concern for gastrointestinal (GI) bleed. Esophagogastroduodenoscopy (EGD) was done revealing duodenal ulcers. Patient was transferred to higher level of care facility with pediatric gastroenterology. He was transferred to our facility for further evaluation and management. After the transfer, he had persistent leukocytosis and dark, maroonish stools with new onset purpura and petechiae, arthralgia, hypertension, and worsening coagulation studies. Extensive workup was done involving multiple consultants to rule out infectious causes and possible HSP. EGD and colonoscopy revealed inflammation of the esophagus, stomach, small intestine, and colon with severe focal chronic active duodenitis. Skin biopsies revealed purpura with mild perivascular dermatitis, leukocytoclastic vasculitis, and negative IgA immunoperoxidase. Cultures, viral studies, and other infectious workup for purpura were all negative. The patient was diagnosed with HSP. Management and Outcome: Initially treated with broad spectrum antibiotics which were discontinued after the diagnosis of HSP. Then, started on IV steroids which were switched to oral Prednisone with tapering dose

outpatient and Enalapril for high blood pressure. Patient improved clinically and was followed up outpatient by gastroenterology, rheumatology, and nephrology. Patient's symptoms resolved and he is doing well. Discussion: This case illustrates that a patient can present atypically for HSP with presentation of acute abdomen and GI bleeding with the development of typical symptoms later.

**417 SUCCESSFUL TRANSITION OFF TUBE FEEDING IN A CHILD WITH HYDROPS FETALIS: A MULTIDISCIPLINARY TEAM APPROACH.** D.M. Dunn, G.R. Markowitz, S. Galbally, K. Pucci, L. Brochi, S.S. Cohen, Feeding and Swallowing Center, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES.

Background: Many infants and young children with complex medical conditions rely on supplemental nutrition from tube feeding. For some, the transition to oral feeding occurs slowly and progresses in line with a child's developmental skills; however, in some cases, tube weaning can be a complex process. Tube dependency is considered an, "unintended result of long-term tube feeding in infants and young children" (Dunitz-Scheer et al., 2009), and can impact a child's development, health, and nutrition, in addition to the multitude of social and emotional sequelae for children and families alike (Edwards et al., 2016). The transition from tube dependence to oral feeding is multi-systemic involving the child's oral motor skills, ability to physiologically tolerate food, developmental and behavioral factors, and caregiver readiness. A multidisciplinary team approach to weaning is indicated to address each of these complex factors. The following case details a multidisciplinary approach to treating significant feeding difficulties in a child with a complex medical history. Case Vignette: Diagnosed prenatally with fetal hydrops, "Hattie" was delivered by cesarean section at 31 weeks gestation and spent the first 9 months of her life in the Neonatal Intensive Care Unit. She was discharged dependent on nasogastric tube feeding. Hattie presented to the Pediatric Feeding and Swallowing Center at the Children's Hospital of Philadelphia at 9 months of age. Her medical presentation included ascites, pulmonary hypoplasia, and chronic lung disease with ongoing requirements for supplemental oxygen and diuretics. She had significant gastroesophageal reflux and was receiving 100% of her nutrition via nasogastric tube by a complex mixture of breastmilk, diluted formula, fat soluble vitamins, and added oils to maintain her essential fatty acid profile. The Feeding Team comprised of a physician, nurse practitioner, dietitian, speech-language pathologist, occupational therapist, and psychologist closely followed Hattie at regular outpatient visits. Initial feeding goals included Hattie accepting a spoon near her mouth, initiating developmental therapies to support her oral motor skill development, and managing her nutrition. At 23-months of age, she was enrolled in an intensive behavioral feeding program where Hattie learned to consume 25 bites of 15 foods of pureed texture at each of 3 meals daily, but was still 100% dependent on a feeding tube for nutrition. The dietitian continued to increase the formula concentration to support age-appropriate weight gain. Unexpectedly, five months after completing the program, Hattie's spirited personality led to her refusing all caregiver spoon feeding attempts. Recommending a child-directed approach, the Feeding Team suggested decreasing the demand on Hattie to increase her willing acceptance of more food and formula by mouth, and Hattie presented at 34-months receiving 80% of total calories from a tube. Only two months after that, her tube dependence decreased to 66% of her calories from the tube. With close Team followed-up, increased oral motor skill development, Hattie's willing acceptance, and medically-adherent parents, Hattie overcame the odds and is now a full oral feeder at age three. Conclusion: Many children present to CHOP's Feeding Team for help weaning off of supplemental tube feeds, but few families are as memorable as Hattie's. Hattie's complex medical history and severe oral aversion in early infancy were signs that Hattie may never be a full oral feeder. Her extremely slow progress weaning from the tube and accepting a spoon, in combination with her family's investment in her feeding progress and infallible follow-through with team recommendations made her case an example of the importance of patience and persistence both for the Feeding Team as well as patients and families. Hattie's story exemplifies the vital collaboration between families and a multidisciplinary medical team in establishing oral acceptance for children in early childhood.

**427 ESSENTIAL FATTY ACID DEFICIENCY AFTER PARTIAL BILIARY DIVERSION FOR PFIC.** I. Batsis, G. Felix, A. Scheimann, Pediatric Gastroenterology, Johns Hopkins, Baltimore, Maryland, UNITED STATES. Stewart, Pediatric Surgery, Johns Hopkins, Baltimore, Maryland, UNITED STATES.

Background: Progressive familial intrahepatic cholestasis (PFIC) is a heterogeneous group of disorders, caused by a defect in bile acid transport, resulting in high serum bile acid concentrations and severe intrahepatic cholestasis. The resulting cholestasis causes depletion of intraluminal bile acid concentration and contributes to fat malabsorption. Partial biliary diversion (PBD) is a surgical alternative to liver transplantation that has been

demonstrated to resolve cholestasis and pruritus, improved growth, and dampen disease progression. Patients with PFIC almost always have fat-soluble vitamin deficiencies prior to surgery. Recent studies have shown that after PBD, fat-soluble vitamin deficiencies often persist – especially Vitamin D and E and patients require supplementation. This case demonstrates a patient who developed essential fatty acid (EFA) deficiency after undergoing PBD; to the best of our knowledge, no other reports exist in the current literature. Case study: This is a 7 year old boy who was diagnosed with PFIC 3 (ABCB4 mutation, by Jaundice Chip, Cincinnati) at age 3. By age six, this patient was having significant pruritus not controlled by rifampin or hydroxyzine. He was referred to surgery for partial biliary diversion and liver biopsy. Liver biopsy was significant for cholestasis, marked bile duct proliferation, and rare bridging fibrosis. Six months after surgery, it was found that his Vitamin D level was very low at 7 despite adequate dietary intake. Vitamin A and E were also found to be low and the INR was mildly prolonged at 1.2. It was also noted that he had an eczematous-type, weepy rash on bilateral upper extremities. Additionally, the patient's mother reported that he was having significant behavioral and cognitive issues in school. Essential fatty acid profile was sent and revealed that the percentage of linoleic acid, C18:2w6, was low. The patient was supplemented with fat-soluble vitamins and put on a "healthy fats" diet. Vitamin levels normalized on supplementation and essential fatty acid profile also normalized with dietary changes. At three month follow-up, the rash had improved and his cognitive/behavioral issues had resolved. Summary: Persistent deficiencies of fat-soluble vitamins are a known risk after partial biliary diversion. However, it should be noted that essential fatty acid deficiencies can also occur. Jankowska et al. demonstrated that lipid profiles (specifically total cholesterol, triglycerides and phospholipids) are often abnormal/elevated in patients with PFIC and that these profiles normalize after PEBD. The mechanism of changes in lipid metabolism is unclear and there is limited data available regarding post-surgical monitoring of fat-soluble vitamins, lipid profiles and essential fatty acid (EFA) profiles. Close follow-up and dietary counseling are necessary for these patients.

**429 METHEMOGLOBINEMIA AND FAILURE TO THRIVE AS A RESULT OF FPIES TO SOY FORMULA WITH RESOLUTION ON AMINO ACID BASED FORMULA.** J. Lee, Pediatrics, Brooke Army Medical Center, Ft. Sam Houston, Texas, UNITED STATES. S. Jones, Pediatric Gastroenterology, Brooke Army Medical Center, Ft. Sam Houston, Texas, UNITED STATES.

**Introduction:** Food protein-induced enterocolitis syndrome (FPIES) is a severe, non-IgE mediated gastrointestinal food allergy syndrome. Despite being a recognized entity, its rarity and presentation variability makes diagnosis difficult. Soy protein is a recognized trigger for FPIES, but case reports of severe FPIES associated with methemoglobinemia (methHgb) from soy alone are scant. We describe an infant who presented with lethargy and methHgb caused by FPIES in the hopes of improving recognition and treatment. **Case Presentation:** A 1 month old male infant who was born at term to a GBS-negative mother presented to our emergency department (ED) with one day of acute vomiting, decreased oral intake, and listlessness. Birth weight was 3.7kg and newborn screens were normal. He was on soy formula after switching from cow's milk protein formula at 1 week old due to excessive spitting up. Parents were mixing formula according to the instruction labels with bottled spring water. The parents denied sick contacts, rash, blood or mucus in stools, delayed passage of meconium, constipation, or use of teething gels. He did have more frequent, looser stools in the week leading up to his presentation, but was brought to ED by mother because that day he was consuming only 1 ounce every few hours, had several episodes of non-bilious, non-bloody emesis, and a rectal temperature at home was 97F. In the ED he was grey in color and lethargic. Initial vital signs were: weight 2.9kg, rectal temp 96.5F, pulse 132 bpm, BP 88/56 mmHg, respirations 42 bpm, and oxygen saturation of 93% on room air. A sepsis evaluation was initiated prior to PICU admission. Pertinent laboratory findings included euglycemia, hyperchloremia, hypernatremia, hypokalemia, and non-anion gap metabolic acidosis. Arterial blood gas revealed methemoglobin of 15.1%. Further evaluation of the metabolic acidosis returned normal urine electrolytes, stool pH of 6 and positive (3+) reducing substances. His cultures (stool, blood, CSF, urine) remained negative. Given the possibility of FPIES in the setting of feeding intolerance and negative cultures, an amino acid-based formula with pre- and probiotics was started and tolerated without emesis. Within days, his lethargy and diarrhea disappeared and his acidosis and other electrolyte abnormalities resolved. Patient has continued to demonstrate appropriate weight gain, without feeding intolerance or recurrent methHgb. **Discussion** The rarity and variation in clinical presentation of FPIES often make early diagnosis difficult. This case reflects the severity of FPIES and reinforces soy protein as a potential allergen that can induce FPIES and the association of FPIES with methHgb. Additionally, the case emphasizes several of the unique laboratory abnormalities

of FPIES. When patients present with a sepsis picture and metHgb in the newborn period, providers must consider non-IgE mediated food protein allergies, specifically FPIES along with other etiologies such as ingestions.

**434 LACTIC ACIDOSIS SECONDARY TO THIAMINE DEFICIENCY.** J. Chang, G. McLaughlin, Pediatric Intensive Care, Jackson Memorial Hospital, Miami, Florida, UNITED STATES. J.T. Hochberg, Pediatrics, Jackson Memorial Hospital/University of Miami, Miami, Florida, UNITED STATES. D. Goldner, Gastroenterology, University of Miami, Miami, Florida, UNITED STATES.

Introduction: In 1985 Velez et al., reported a previously unknown complication of total parenteral nutrition (TPN) without thiamine: acute onset of life threatening metabolic acidosis. The transformation of pyruvate (from glucose) into acetylcoenzyme A is performed by pyruvate dehydrogenase, an enzyme dependent on thiamine as a co-factor. Without thiamine, pyruvate is converted to lactate through anaerobic metabolism. Thiamine deficiency is more likely in the critically ill, secondary to underlying malnutrition and increased catabolic state. As the body's thiamine storage are exhausted clinical symptoms usually become evident, resulting in a significant degree of morbidity and mortality. Case report: A 17-year-old male with phenylketonuria was on specialized total parenteral nutrition due to bowel obstruction. He developed auditory and visual hallucinations followed by a complaint of chest pain with the associated physical findings of tachypnea but not hypoxia. He was transferred to the intensive care unit with a vasodilatory pattern of shock, strong pulses, warm extremities and hypotension. Initial laboratory investigations revealed a metabolic acidosis with a serum lactate level up to 13.8mmol/L which failed to improve with fluid administration and antibiotics. The patient received 100 mg of thiamine and, within hours, hypotension, tachycardia and lactic acidosis resolved. On further investigation it was noted that vitamins were omitted from the specialized total parenteral nutrition for approximately three weeks. In a patient population at increased risk of sepsis, clinical differentiation between sepsis and thiamine deficiency is difficult. Thiamine deficiency manifests clinically as wet or dry beriberi. The signs and symptoms of dry beriberi reflect peripheral nervous system involvement and vasodilation with high cardiac output. Overtime the high output state can progress to wet beriberi with decreased cardiac function. Both sepsis and thiamine deficiency can result in severe lactic acidosis. However, the differential diagnosis for refractory lactic acidosis in patients with malnutrition or on TPN should always include thiamine deficiency.

**435 WERNICKE ENCEPHALOPATHY IN A PEDIATRIC PATIENT WITH DYSPHAGIA.** L.E. Mullinax, L. Siebold, Pediatric Gastroenterology, Children's Hospital of Pittsburgh at UPMC, Pittsburgh, Pennsylvania, UNITED STATES.

Introduction: Wernicke encephalopathy (WE) is a critical neuropsychiatric condition due to thiamine deficiency that is commonly associated with alcoholism. It is characterized by a triad of symptoms: ophthalmoplegia, ataxia and encephalopathy. We report a case in which WE was diagnosed in a pediatric patient who severely limited his intake for 3 months. Case Description: A 17-year-old male patient with a past medical history of generalized anxiety disorder and obesity was hospitalized for the evaluation of dysphagia complicated by restriction of oral intake and 30kg weight loss over a 3-month period. Leading up to this admission, the patient self-restricted his diet to Kool-Aid and stopped taking his psychiatric medication due to fear of choking. Modified barium swallow study showed normal swallowing with thin liquids but inability to swallow puree material without the help of thin liquids. Esophagram was significant for mild mucosal irregularity at the junction of the hypopharynx and cervical esophagus, thought to be related to mild inflammation. Esophagogastroduodenoscopy revealed esophageal candidiasis and treatment for esophageal candidiasis with fluconazole was initiated. His clinical history was non-concerning for any iatrogenic causes or underlying immunodeficiency that would predispose him to esophageal candidiasis. Over the course of the admission, he complained of worsening confusion, memory loss and visual difficulties and was found to be with nystagmus and ataxia on physical exam. Brain MRI confirmed the diagnosis of WE and he responded well to treatment with thiamine. As the patient slowly advanced his oral feeds, his electrolytes were closely monitored to prevent refeeding syndrome. His psychiatric medication was also restarted. At discharge, he was transferred to a pediatric specialty rehabilitation hospital with a functional feeding program to continue psychiatric treatment and oral therapy. At follow-up, his anxiety is well controlled, he is no longer symptomatic from WE after thiamine repletion, he is eating normally, and is without complaints of dysphagia. Discussion: WE can be fatal if left untreated. The gravity of WE if left undiagnosed and thus untreated only emphasizes the importance of careful history taking, a proper and comprehensive physical exam and most importantly, disease awareness, even in the pediatric population.

**436** *MANAGING MEDNIK SYNDROME ASSOCIATED INTESTINAL FAILURE: A BALANCE BETWEEN ZINC AND COPPER.* L. Diaz Calderon, C. Torres, Pediatric Gastroenterology, Hepatology and Nutrition, Children's National Medical Center, Washington, District of Columbia, UNITED STATES.

MEDNIK syndrome stands for mental retardation, enteropathy, deafness, neuropathy, ichthyosis and keratoderma. It is caused by mutations in AP1S1 gene, which is involved in intracellular copper trafficking with secondary deficiency of copper-dependent enzymes. Abnormal copper metabolism results in its accumulation in liver, enterocytes and CNS. We present a 22-month-old male born at 36 weeks of gestation with intestinal failure secondary to congenital diarrhea associated with homozygous changes in the AP1S1 gene. He has intestinal failure associated liver disease, sensorineural hearing loss, metabolic bone disease and developmental delay. Since birth, patient had feeding intolerance and was 100% parenteral nutrition dependent. He had high fluid, electrolyte and micronutrient requirements due to excessive stools losses. Intestinal electron microscopy was normal. Metabolic and endocrine workups were negative. He had extremely low serum copper and ceruloplasmin levels. Initial liver biopsy showed stage 3-4 fibrosis, marked cholestasis and low copper quantitation in liver. After enteral zinc supplementation was started, patient had multiple episodes of sepsis, enterocolitis and central line infections. We found an association between extremely low serum copper levels and impaired immunity therefore the dose of supplemental zinc was adjusted to allow normal serum copper levels and avoid the complications of its deficiency. His recurrent episodes of sepsis and chronic diarrhea resolved. He is currently tolerating jejunal feeds, which provide 70% of his daily caloric needs and receives parenteral nutrition that gives the remainder of his calories. This case elucidates the importance of maintaining a strict balance between serum zinc and copper levels in the treatment of patients with MEDNIK syndrome.

**438** *NORMAL SOMATIC GROWTH IN THE CONTEXT OF MULTIPLE MICRONUTRIENT DEFICIENCIES IN AN ADOLESCENT FEMALE WITH SHORT BOWEL SYNDROME.* M. McGivney, D.A. Stamm, B. Depaula, C. Duggan, Center for Advanced Intestinal Rehabilitation, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES. Depaula, C. Duggan, Division of Gastroenterology, Hepatology and Nutrition, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES.

Background: Malabsorption of micronutrients is a well described phenomenon in pediatric patients with intestinal failure, and those who have undergone ileal resection are particularly at risk. This is related to the loss of the primary absorption site for vitamin B12 as well as bile salt malabsorption resulting in steatorrhea and impaired uptake of fat soluble vitamins A, E, D and K. Furthermore, patients who have undergone ileocecal valve resection experience loss of the ileal brake, resulting in increased stool losses. Deficiencies in both zinc and copper have been described in the context of high volume diarrhea. Micronutrient deficiencies can cause macrocytic anemia, xerophthalmia and retinal abnormalities, hemolytic anemia and peripheral neuropathy, metabolic bone disease, coagulopathies, growth failure, and neutropenia. Given this wide range of deleterious impacts on normal development, adequate micronutrient absorption is often assumed a prerequisite for normal somatic growth in the pediatric patient. Methods: Retrospective chart review was performed on an adolescent female with intestinal failure who presented with multiple micronutrient deficiencies despite normal somatic growth and bone mineral density. Results: Our patient was born at 31 weeks gestation with development of necrotizing enterocolitis (NEC) on day of life 9. She underwent exploratory laparotomy resulting in significant ileal resection including her terminal ileum and ileocecal valve, with restoration of intestinal continuity at 4 months of age via jejunocolonic anastomoses. She weaned from parenteral nutrition and progressed to full oral feedings before one year of age. She continued as a patient in our intestinal rehabilitation program with increasingly sporadic follow up as her age progressed. She was diagnosed with vitamin B12 and vitamin D deficiencies at age 15 years, and was started on 1000 mcg of cyanocobalamin intramuscularly once monthly and 8000 IU of ergocalciferol once daily. Serum values of B12, 25OHD, homocysteine and methylmalonic acid subsequently normalized. Zinc deficiency was noted at age 16 years and she was started on 44mg elemental zinc per day with normalization of serum levels. After being lost to follow up for 2 years, our patient presented to our intestinal rehabilitation program at age 18 years for routine evaluation. She reported 5 liquid stools per day and intermittent mild post-prandial right lower quadrant pain which was alleviated by defecation. She was eating an unrestricted diet of oral solids without high calorie supplements. She reports a regular pattern of menstruation. She reported non-adherence to prescribed cyanocobalamin, ergocalciferol and zinc sulfate supplements for greater than 1 year. On physical exam our patient's weight was 54.7kg (40th%ile wt/age z-score -0.24) with an interval weight gain of 3kg, height 157.5cm (19th%ile ht/age z-score -0.88) without significant interval change in ht/age z-score, BMI 22.1kg/m<sup>2</sup> (58th%ile

bmi/age z-score 0.2) with interval increase in bmi/age from -0.2. She was alert, interactive and in no acute distress, skin warm and free of pallor or rash, conjunctiva pink, normal contour and expansion of the chest with tanner 5 breast development. Normal S1 and S2, lungs clear to auscultation. Her abdomen was soft and non-tender without hepatosplenomegaly. She had tanner 5 external genitalia with a clean, dry and intact perineum. Neurologic exam was non-focal with 2+ patellar deep tendon reflexes. No evidence of extremity or gluteal wasting. As part of routine screening, a DEXA scan was obtained showing z-scores of 0.2 at the lumbar spine and -1.05 for total body less head (TBLH). Biochemical assessment with evidence of vitamin D (8.9 ng/ml), zinc (52 mcg/dl), and copper (58 mcg/dl) deficiency with vitamin B12 insufficiency supported by borderline low serum concentration (191 pg/ml) along with macrocytosis (MCV 89.7 fl) and hyperhomocysteinemia (13.8 umol/L). Our patient was treated with 1000 mcg of intramuscular cyanocobalamin monthly, 12,000 IU ergocalciferol daily, 44mg elemental zinc daily and a complete chewable multivitamin providing 2 mg per day of copper. Conclusion: Significant micronutrient deficiencies can exist in pediatric patients with intestinal failure in the context of normal somatic growth and reassuring physical exam. Monitoring of micronutrient levels should be consistently included as part of routine care. More frequent monitoring may be required in adolescent patients who may have challenges with adherence to prescribed therapies.

**440 TWO CASES OF PERNICIOUS ANEMIA IN TEENAGE MALES WITH LOWER EXTREMITY WEAKNESS.** A. Glinky, P. Arias, N. Santucci, Pediatrics, Louisiana State University Health Sciences Center, New Orleans, Louisiana, UNITED STATES. Arias, N. Santucci, Gastroenterology, Hepatology and Nutrition, Children's Hospital, New Orleans, Louisiana, UNITED STATES.

Introduction: This case series presents two cases of pernicious anemia diagnosed in teenage males with other autoimmune conditions between March and October of 2016. Case presentation: RJ is 16 year old male with past medical history of Evans syndrome and schizophrenia who presented with two months of progressive bilateral lower extremity weakness and pain leading to inability to walk. On exam, he had a wide-based gait, positive Romberg sign, 4/5 strength, and decreased sensation to pinprick and vibration in his lower extremities. Laboratory studies showed normocytic anemia (Hb 8 g/dL) and vitamin B12 level of 53 pg/mL (normal >200 pg/mL). HW is a 19 year old male with type I diabetes since age two who presented with progressive bilateral lower extremity weakness and numbness. On exam, he had bilateral lower extremity weakness and 4/5 strength as well as decreased sensation of vibration and proprioception. He had pancytopenia with macrocytic anemia (Hb 3.9 g/dL and MCV of 113.3 fL) and B12 level of <50 pg/mL. Serum methylmalonic acid was elevated in both patients. Both patients had positive intrinsic factor blocking antibodies. HW did not have anti-parietal cell antibodies and RJ was untested. Both were deficient in vitamin D and HW was deficient in folate. Both patients had upper and lower endoscopy with evidence of gastritis but no malabsorption. Management and outcome: RJ and HW both received daily intramuscular cyanocobalamin injections for 5-7 days followed by weekly injections for a month with marked symptomatic neurologic improvement as well as supplementation for their respective vitamin deficiencies. RJ completed two weeks of inpatient neurorehabilitation while HW only required outpatient physical therapy, which he did not complete. Both patients currently receive monthly home B12 injections. Discussion: Pernicious anemia is commonly seen in patients with other autoimmune conditions. The cases in these vignettes had intrinsic factor blocking antibodies and elevations in serum methylmalonic acid which were important for making this diagnosis. Schillings tests are much less commonly used but still useful in the setting of negative antibodies and high clinical suspicion. Intrinsic factor antibodies are highly specific for pernicious anemia but only 50-70% sensitive while anti-parietal cell antibodies are much less specific, thus of decreased diagnostic utility. It is notable that both cases had other vitamin deficiencies without evidence of malabsorption. A literature review revealed vitamin D deficiency to be common in auto-immune disorders. Interestingly, neither of these patient had evidence of atrophic gastritis, which is a common finding in patients with pernicious anemia.

**441 NOT SO TUFT: DIAGNOSING CONGENITAL TUFTING ENTEROPATHY BEFORE VISIBLE TUFTING.** L. Klein, Pediatrics, Duke University Medical Center, Durham, North Carolina, UNITED STATES. Purohit, Internal Medicine and Pediatrics, Duke University Medical Center, Durham, North Carolina, UNITED STATES. M. Mavis, R. Noel, N. Venkatasubramani, Pediatric Gastroenterology, Hepatology, and Nutrition, Duke University Medical Center, Durham, North Carolina, UNITED STATES.

Background: Congenital tufting enteropathy (CTE) is an autosomal recessive disorder of intestinal mucosal development characterized by intractable diarrhea resulting in failure to thrive, severe caloric-protein

malnutrition, total parenteral nutrition (TPN) dependence, and in most cases, need for small bowel transplant. *EpCAM*, the gene most commonly implicated in CTE, encodes for epithelial cell adhesion molecules. We present a rare case of CTE with a homozygous frameshift mutation in *EpCAM*. Case: A 4-week old boy was hospitalized for evaluation of diarrhea, emesis, and failure to thrive. Prenatal and birth history was positive for consanguineous parents but otherwise unremarkable. Multiple formula changes for weight loss and diarrhea made no improvement in symptoms. Physical examination showed a cachectic infant with temporal wasting. His laboratory workup was remarkable for a non-anion gap metabolic acidosis. Inflammatory and infectious studies were negative. Initial genetics and renal evaluations were normal. Endoscopy at 3-months of age revealed villous atrophy but no evidence of tufting or other abnormalities on electron microscopy. A gastrostomy tube was placed for growth failure and persistent diarrhea. The patient developed hypovolemic shock and hypoglycemia and was unable to be weaned from intravenous fluids. Although original electron microscopy of small intestine was non-diagnostic, immunohistochemistry (IHC) staining for congenital diarrheas was performed on the intestinal biopsy. CD10 and chromogranin highlighted an intact brush border and evenly distributed neuroendocrine cells, respectively. However, MOC31 staining for *EpCAM* expression was negative within the epithelium, concerning for CTE. Genetic testing discovered homozygosity for *EpCAM* variant c.38\_66dup, which is predicted to result in a frame shift and premature protein termination. The patient was started on TPN, but developed multiple line infections and liver fibrosis necessitating small bowel transplant. The patient is clinically doing well after isolated intestinal transplant with appropriate growth on full enteral nutrition. Discussion: Diagnosis of CTE is often delayed or missed because the characteristic findings on small bowel biopsies can be subtle, focal, or even absent, since tufts are usually not identifiable until after two years of age. This case demonstrates the importance of immunohistochemical staining and genetic analysis despite normal ultrastructure on electron microscopy. Additionally, there are reports of favorable outcomes based on genetic mutations, further emphasizing the importance of complete genetic testing.

#### **442 GASTROINTESTINAL INVOLVED LANGERHANS CELL HISTIOCYTOSIS IN AN INFANT WITH FAILURE TO THRIVE.**

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Abstract/Background: Langerhans cell histiocytosis (LCH) is a rare disease with a heterogeneous presentation. Of children diagnosed with LCH, gastrointestinal (GI) tract involvement is exceedingly rare. Most children diagnosed with gastrointestinal tract involved LCH have significant chronic upper or lower GI symptoms such as hematochezia, diarrhea, or vomiting that initiates an evaluation. [1] Here, we present a case of a four month old infant who was diagnosed with multisystem LCH involving the GI tract after presenting to a general pediatrics clinic with a rash and failure to thrive. Case Report: Our patient was born at term after an unremarkable pregnancy and delivery. At 6 weeks of age, he developed non-blanching salmon colored papules in the scalp and eyebrows. His rash spread to involve the proximal extremities and the trunk. He also developed erythematous atrophic plaques without scale in the bilateral axilla and inguinal creases. Weight gain and linear growth almost completely ceased beginning at two months of age. He had one self-limited episode of vomiting and diarrhea, but otherwise parents described a normal stool pattern. He never developed melena or hematochezia. At four months of age, a failure to thrive evaluation was initiated. He had a complete blood count and complete metabolic panel performed as part of this evaluation, which were remarkable for low albumin and normocytic anemia. He was referred to dermatology where a skin biopsy was performed due to suspicion for LCH. Cells on his biopsy showed immunoreactivity to CD1a, confirming the suspected diagnosis. After confirmation of the diagnosis of LCH, an upper endoscopy was performed due to hypoalbuminemia and failure to thrive. Superficial erosions in the duodenum were visible on endoscopy. A biopsy of the lesions demonstrated cells that also stained positive for CD1a, confirming the diagnosis of multifocal LCH with gastrointestinal involvement. Discussion: Multifocal Langerhans cell histiocytosis is a rare disease and its wide variety of clinical presentations often delays diagnosis and treatment. Involvement of the gastrointestinal mucosa in LCH is infrequent, estimated to occur in less than two percent of LCH patients in published case series. [2] [3] Gastrointestinal involved LCH is associated with high morbidity and mortality despite its rarity, underscoring the importance of early diagnosis and treatment. Previously published case reports of this clinical entity almost always involve infants and young children with chronic vomiting, hematochezia, or bloody diarrhea. To our knowledge, this is one of the first published cases of GI involved LCH where the diagnosis was not preceded by chronic diarrhea or GI bleeding, thus expanding the presentation of this disease. This case underscores importance of maintaining a broad differential when evaluating failure to thrive in the

infant. References: 1. Hait E, Liang M, Degar B, Glickman J, Fox VL. Gastrointestinal tract involvement in Langerhans cell histiocytosis: case report and literature review. *Pediatrics.* 2006; 118(5):2. Geissmann F, Thomas C, Emile JF, et al. Digestive tract involvement in Langerhans cell histiocytosis: the French Langerhans Cell Histiocytosis Study Group. *J Pediatrics.* 1996; 129(6):836–8453. Shima, H et al. Protein-Losing Enteropathy Caused by Gastrointestinal Tract–Involved Langerhans Cell Histiocytosis. *Pediatrics.* 2010; 125; e426.

**443** *MIRAGE SYNDROME: A RARE DIAGNOSIS PRESENTING AS REFRACTORY INFANTILE CHRONIC DIARRHEA.* L. Alrabadi, A. Alper, A.F. Porto, Department of Pediatric Gastroenterology & Hepatology, Yale University, New Haven, Connecticut, UNITED STATES. Okafor, Pediatrics, Yale University School of Medicine, New Haven, Connecticut, UNITED STATES. Jeffries, J. McGrath, Genetics, Yale University School of Medicine, New Haven, Connecticut, UNITED STATES.

Introduction: Chronic diarrhea is a common gastrointestinal symptom in infants, mostly due to infections, drug adverse reactions and food sensitivities. Here we describe an infant who was evaluated for failure to thrive and refractory chronic diarrhea and was subsequently diagnosed with an extremely rare genetic disorder. Case Report: A six-month old boy with an extensive past medical history including prematurity (ex-26 weeker, birth weight: 600g), necrotizing enterocolitis, s/p gastrotomy and small bowel resection, bronchopulmonary dysplasia, ambiguous genitalia and adrenal insufficiency, presented to clinic with failure to thrive, reflux symptoms and feeding intolerance. The patient presented with fussiness and back arching, associated with feeds, with no emesis. Also, he had frequent loose and non-bloody stools, notably after every feed, up to 6-7 times a day. Patient was growing at <sup>st</sup> percentile for both weight and height. Physical exam was significant for dysmorphic cranial facies, microphallus with fused labiosacral folds, and hypospadias, otherwise unremarkable. Labs were notable for hyperkalemia (K= 5.5 mmol/L) and low bicarbonate (HCO<sub>3</sub>= 18 mmol/L). Initial management included increased caloric density of amino-acid based formula and initiation of a histamine 2 receptor antagonist. Feeding tolerance gradually improved; however, he continued to have poor weight gain and diarrhea. Feeds were later augmented to increase calories by 31%. This modification improved weight gain but worsened his chronic diarrhea. As the patient continued to work with the gastroenterology team, he was simultaneously being evaluated by both genetics and endocrinology to determine an etiology linking the ambiguous genitalia and adrenal insufficiency. Evaluation included whole exome sequencing, with negative initial screening for common genetic causes of ambiguous genitalia and adrenal insufficiency. Further extended genetic testing identified a mutation of the SAMD9 gene leading to the diagnosis of MIRAGE syndrome. Discussion: A multi-disciplinary approach in the evaluation of this patient led to the diagnosis of MIRAGE syndrome; which is a constellation of diagnoses that develop secondary to a gain-of-function mutation to the SAMD9 gene. The diagnoses include: Myelodysplastic syndrome; Infections; Restriction of growth; Adrenal hypoplasia; Genital hypoplasia; Enteropathy (chronic diarrhea and colonic distension). Though MIRAGE syndrome is rare, with only 19 patients described in the literature, the syndrome is described as often fatal with death common in the first year of life secondary to severe invasive infections. This case report indicates the importance of considering this diagnosis in the differential of a patient with unexplained chronic diarrhea, especially when the above-mentioned co-morbidities are present.

**448** *SCHWACHMAN DIAMOND IN THE ROUGH: A STORY OF NEONATAL FAILURE TO THRIVE AND NEUTROPENIA.* M.L. Gallant, J. SODEN, Gastroenterology, Hepatology, and Nutrition, Children's Hospital of Colorado, Denver, Colorado, UNITED STATES.

Introduction: This is the case of a patient who presented with fever, neutropenia, abdominal distension, poor weight gain, and loose stools and was diagnosed with Schwachman Diamond Syndrome. The case is unique based on the neonatal presentation, as well as the surgical abdominal manifestation. Case Description: This is a 46 day old male with a history of IUGR, skeletal rib dysplasia, a prior NICU admission for abdominal distension and feeding intolerance who presented with fever, fussiness, and neutropenia. He was born at term, SGA, and had a prior NICU admission with concern for abdominal distension, however he was able to achieve full enteral feeds without significant intervention at that time. Three weeks later, he began to have NBNB emesis and fussiness with feeds, and he then developed fever and diarrhea. He was evaluated in an outside ER, with evaluation notable for severe neutropenia with an ANC of 50. He was transferred to our NICU once again for fever and neutropenia. He was also noted to have poor weight gain. Infectious studies were normal. On day four of his admission, he began to develop abdominal distension. Surgical suction rectal biopsy did not show evidence of aganglionosis. However, after the procedure, he became febrile with a tense abdomen and had labs consistent with DIC. He was taken to the OR for

an exploratory laparotomy, which was significant for tip colitis of the cecum and a gangrenous appendix similar in appearance to neutropenic typhlitis, requiring ileocecal resection and diverting ileostomy. Given his skeletal dysplasia, diarrhea (and later confirmed pancreatic insufficiency), neutropenia, and failure to thrive, genetic testing for Schwachman Diamond was sent and confirmed the diagnosis. Management of his Schwachman Diamond as an outpatient has involved G-CSF twice weekly for chronic neutropenia, pancreatic enzyme replacement, and further monitoring for associated nutritional complications. He has malabsorptive symptoms despite pancreatic enzyme replacement suggesting at least some component of short bowel syndrome as well. Discussion: Schwachman Diamond is an autosomal recessive disorder characterized by exocrine pancreatic insufficiency and bone marrow dysfunction along with other associated anomalies. It should be considered in the differential for neonates with severe bacterial infections, neutropenia or pancytopenia, and failure to thrive. While commonly considered for children assessed for failure to thrive and pancreatic insufficiency later in life, it is also well described in the neonatal period and early diagnosis has important implications for progressing to myelodysplasia and potentially requiring more aggressive therapies like bone marrow transplantation.

**453 NINE-YEAR-OLD WITH AUTOIMMUNE PANCREATITIS AND EXOCRINE PANCREATIC INSUFFICIENCY.** M.S. Mehta, I. Rojas, Pediatric Gastroenterology, Hepatology and Nutrition, University of Texas Southwestern Medical Center, Dallas, Texas, UNITED STATES. S. Mehta, I. Rojas, Pediatric Gastroenterology, Children's Health, Dallas, Texas, UNITED STATES.

Background Autoimmune pancreatitis (AIP) is a rare inflammatory form of chronic pancreatitis most commonly seen in adults. There are few published pediatric cases and type 1 AIP with IgG4 elevation is rarely seen. We report a case of AIP with elevated IgG4 who presented with transaminitis and jaundice, and later developed exocrine pancreatic insufficiency (EPI). Case This 9-year-old male presented with transaminitis and jaundice, following 1.5 weeks of left upper quadrant pain, weight loss and fatigue. An abdominal ultrasound revealed a mass at the head of the pancreas with dilation of the proximal biliary duct, which was confirmed on abdominal MRI. Lipase, amylase, alpha-fetoprotein and carcinoembryonic antigen were normal; serum IgG4 was elevated to 141 mg/dL (<95mg/dL). He underwent endoscopic ultrasound guided fine-needle aspiration of the head of the pancreas. Histology revealed rare clusters of benign epithelial cells; IgG4 staining could not be done due to pauci-cellularity. After starting prednisone, IgG4 normalized and pancreatic mass resolved; however, follow-up ultrasound revealed an atrophic pancreas. He has been followed for 2.5 years off therapy without recurrence of disease or other organ involvement. Transaminitis and jaundice resolved after one month, but he then developed greasy stools and persistent weight loss. He had low fecal elastase 43ug/g (>201ug/g) and elevated fecal qualitative neutral and split fat, suggesting EPI. Steatorrhea improved one week after starting pancreatic enzyme replacement therapy (PERT). Four months later, at time of steroid completion, patient self-discontinued PERT. He continued to have persistently low fecal elastase levels, but has remained asymptomatic with weight gain, normal fat soluble vitamin levels and no steatorrhea. Discussion This patient was diagnosed with probable type 1 AIP in setting of clinical symptoms, imaging findings, IgG4 elevation and improvement on steroids; however, diagnosis could not be confirmed due to inadequate tissue sample on pancreatic biopsy. Improved biopsy techniques are needed to assist in making diagnosis and ruling-out other causes of pancreatic mass, such as malignancy. Despite resolution of pancreatic mass on imaging, patient developed atrophy of pancreas, as well as persistent laboratory evidence of EPI, but no clinical symptoms. In absence of steatorrhea or malnutrition, PERT may not be necessary. Available literature suggests 61% of pediatric patients develop atrophic pancreas, but only 16% develop EPI. Conclusion This patient had a rare case of probable type 1 AIP with IgG4 elevation without evidence of systemic IgG4 disease. Better biopsy techniques for pediatric patients need to be developed to confirm diagnosis of AIP. Pancreatic atrophy and EPI can develop, despite therapy for AIP. Fecal elastase may remain low, but in absence of clinical symptoms, PERT may not be necessary.

**454 CONGENITAL TUFTING ENTEROPATHY AND PROTEIN ALLERGY: AN UNUSUAL PRESENTATION.** M.S. Mehta, S. Barlow, Pediatric Gastroenterology, Hepatology and Nutrition, University of Texas Southwestern Medical Center, Dallas, Texas, UNITED STATES. S. Mehta, S. Barlow, Pediatric Gastroenterology, Children's Health, Dallas, Texas, UNITED STATES.

Background Congenital tufting enteropathy (CTE), is a rare congenital condition with abnormal enterocyte development or differentiation leading to intractable diarrhea and intestinal failure. Histological features include villous atrophy and epithelial dysplasia with a "tufting" appearance. Typically, CTE presents with early-onset

chronic diarrhea. We report an unusual case of tufting enteropathy presenting with blood in stool. Case This full-term male infant developed blood in stool at 4 days of life while on breastmilk, which resolved after switching to a hydrolyzed formula. He was briefly admitted at two weeks of life with a urinary tract infection, and noted to have failure to thrive (weight 0.23%ile,  $z=-2.83$ ), for which feeds were switched to amino-acid based formula. At 4.5 weeks of life bloody diarrhea recurred, which worsened after mother resumed hydrolyzed formula. He was subsequently admitted for the diarrhea and persistent failure to thrive (weight <0.01%ile,  $z=-4.59$ , 91g below birth weight) despite adequate caloric intake. Diarrhea and blood partially improved when enteral intake was held. After unrevealing infectious work-up, patient underwent an esophagogastroduodenoscopy and flexible sigmoidoscopy, which showed an edematous duodenum. The histology revealed patchy duodenal villous blunting with rare focal epithelial tufting. Immunohistochemical staining showed absence of intestinal epithelial cell membrane adhesion molecule (EpCAM) throughout the GI tract, confirming diagnosis of CTE. He also had mild eosinophilic colitis suggestive of allergic colitis from protein allergy. He was discharged on total parental nutrition and amino-acid based trophic feeds with resolution of bloody diarrhea and stable weight gain. Discussion While there is heterogeneity in presentation, CTE usually presents in early infancy with chronic watery diarrhea and failure to thrive; bloody diarrhea has not been described. CTE is associated with EpCAM deficiency, which results in enterocyte disorganization with increased permeability of intestinal barrier resulting in intractable watery diarrhea. Epithelial disorganization and villous blunting can also lead to accompanying malabsorptive diarrhea. These features are unlikely to be related to the non-IgE mediated pathway causing this patient's protein allergy. In this patient, bloody stools from protein allergy proved to be a red herring masking his CTE. Epithelial tufting is the histopathological hallmark of this disease, but can be rare or absent and often misconstrued for artifact. Therefore, EpCAM immunostaining is important if there is suspicion for CTE. Conclusion CTE is a rare condition with variable presentation. EpCAM immunohistochemical staining should be done because other histological features may not be present. Protein allergy has not previously been reported with CTE and may be unrelated.

**455 ACUTE PANCREATITIS IN A PATIENT WITH DIABETIC KETOACIDOSIS AND SICKLE CELL VASO-OCCLUSIVE CRISIS.** C. Ossai, M. Hamza, M. Peichev, R. Nathan, K. Viswanathan, Pediatrics, Brookdale University Hospital and Medical Center, Brooklyn, New York, UNITED STATES.

Background Acute pancreatitis is an increasingly recognized condition in the pediatric population. Few case reports have described acute pancreatitis in patients with diabetic ketoacidosis (DKA)<sup>1</sup> or acute sickle cell vaso-occlusive crisis (VOC).<sup>2</sup> Case Report A 13-year-old girl with history of sickle cell anemia (HbSS) and type 1 diabetes mellitus presented to the ED with vomiting and severe pain in the chest, back and epigastrium of one day duration. Initial laboratory studies were WBC  $33 \times 10^9$ /L, Hb 8 g/dl, glucose 658 mg/dl, HCO<sub>3</sub> 6.5 mmol/l, pH 7.11, LDH 3560 U/L, bilirubin 3.2 mg/dl, ALP 215 U/L, ALT 21 U/L, AST 34 U/l. She was admitted for management of DKA and sickle cell VOC. She received IV fluids, insulin, ceftriaxone and morphine. On day 2, she continued to have epigastric pain with radiation to the back. Labs showed WBC  $37 \times 10^9$ /L, Hb 6.8 g/dl, glucose 228 mg/dl, HCO<sub>3</sub> 16 mmol/l, pH 7.30, LDH 11000 U/L, ALP 191 U/L, ALT 25 U/L, AST 328 U/L, Amylase 132 U/L, Lipase 450 U/L, Triglyceride 53 mg/dl and GGT 25 U/L. Abdominal sonogram showed cholelithiasis with normal common bile duct (CBD). Patient was continued NPO, started on omeprazole and received one unit of packed red blood cell transfusion (PRBC). On day 3, amylase was <30 U/L, lipase 20 U/L, WBC  $22 \times 10^9$ /L, Hb 7.8 g/dl, LDH 8690 U/L. Clinical symptoms improved; she was started on clear liquids and advanced to regular diet on day 4. Discussion We had a broad differential diagnosis for our patient's presentation including gall stones obstructing CBD, DKA, hypertriglyceridemia. Lab studies and imaging ruled out these possibilities and the presence of elevated amylase and lipase after resolution of acidosis ruled out DKA. The persistent nausea, abdominal pain and drop in Hb increased our clinical suspicion of acute pancreatitis secondary to acute sickle cell VOC. Sickle cell VOC has been proposed to cause acute pancreatitis through the process of microvascular occlusion and ischemic injury.<sup>2</sup> After receiving PRBC, there was a significant drop in amylase and lipase which supported our suspicion. Blood transfusion has a beneficial role in patients with sickle cell disease and vaso-occlusive crisis by improving the sickling process and increasing oxygen delivery to the tissues. We believe this was the case in our patient. It is likely that she initially presented with DKA and dehydration which triggered an acute vaso-occlusive process leading to pancreatitis. Conclusion Acute pancreatitis should be considered in the differential diagnosis of patients with acute sickle cell VOC or DKA who present with persistent abdominal pain. References 1. Singla AA, Ting F, Singla A. Acute pancreatitis secondary to diabetic ketoacidosis induced hypertriglyceridemia in a young adult with undiagnosed type 2 diabetes. JOP. 2015 Mar 20;16(2):201-4. 2. Ahmed S, Siddiqui AK, Siddiqui RK, Kimpo

M, Russo L, Mattana J. Acute pancreatitis during sickle cell vaso-occlusive painful crisis. *Am J Hematol.* 2003 Jul;73(3):190-3.

**456 CHYLOMICRON RETENTION DISEASE: A CASE OF INFANT PRESENTING WITH VOMITING AND FAILURE TO THRIVE WITHOUT DIARRHEA.** M. Woods, S. Parkash, S. Chowdhury, M. Rashid, Department of Paediatrics, Dalhousie University, Halifax, Nova Scotia, CANADA.

**BACKGROUND:**Chylomicron retention disease (Anderson's disease) is a very rare autosomal recessive disorder caused by mutation of the SAR1B gene that leads to hypocholesterolemia and accumulation of lipoproteins in the enterocytes. This leads to malabsorption and deficiencies of fat soluble vitamins with serious clinical sequelae. Patients most commonly present in infancy with nonspecific symptoms such as vomiting, diarrhea and failure to thrive. Diarrhea is reported to be universally present in all cases.**OBJECTIVE:**We report the first case of an infant with chylomicron retention disease presenting without diarrhea.**CASE:**A 5-month-old term baby boy of French Canadian descent was referred from a community hospital with vomiting and severe failure to thrive. By 2 months of age, he had only grown to 4.8 kg. He was exclusively breastfed for the first 3 months of life but had been transitioned to formula due to poor growth. By 4 months of age, his length and head circumference plateaued. He was spitting up small amounts in the first few weeks of life and then progressed to have frequent non-bilious vomiting. There was no history of diarrhea. An upper GI series showed minimal reflux. A barium enema done for Hirschsprung's disease was negative. Sweat chloride was normal. Treatment with PEG3350 for presumed constipation and lansoprazole had been initiated along with NG tube feeds with a partially hydrolyzed formula. On examination at transfer to the Paediatric unit, the infant appeared emaciated with poor muscle mass. The abdomen was distended with no masses or organomegaly. Muscle strength was significantly decreased. Investigations revealed normal urine, blood gas, electrolytes, glucose, renal function, bilirubin, ammonia, and organic acids. The liver transaminases were mildly elevated and albumin was low. Stool microscopy showed fat globules. Upper GI endoscopy showed milky white appearance of the duodenal mucosa. Biopsies showed normal villous height and architecture with significant steatosis of the enterocytes. Electron microscopy confirmed large amount of lipid droplets in the cytoplasm of enterocytes. Further workup revealed markedly reduced LDL-cholesterol, borderline low HDL-cholesterol and normal triglycerides. Vitamin A, D and E levels were decreased. Genetic testing revealed two different heterozygous variants in the SAR1B gene; c.537T>A and c.409G>A. Parents were both carriers of the variants in SAR1B, confirming that each variant was on a separate copy of the SAR1B gene. The infant was started on a partially hydrolyzed formula with a higher medium chain triglyceride content given orally and by NG tube. Supplementation with high dose vitamin A, D, E and K was initiated. In follow-up at 10 months of age, he was doing very well, gaining weight with normal fat soluble vitamin levels.**CONCLUSIONS:**Chylomicron retention disease can present without diarrhea. A high index of suspicion for a disorder of hypocholesterolemia and early endoscopy is recommended in infants with persistent vomiting and failure to thrive. A timely diagnosis and treatment is essential to avoid serious clinical sequelae, especially neurological impairment.

**458 A 2 YEAR OLD MALE WITH A MYSTERIOUS PANCREATIC MASS LEADING TO A RARE DIAGNOSIS: A CASE REPORT.** N. Bhesania, K. Radhakrishnan, P. Conjeevaram Selvakumar, Pediatric Gastroenterology, Cleveland Clinic Children's Hospital, Cleveland, Ohio, UNITED STATES. D. Patil, Department of Anatomic Pathology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES. Walsh, Department of Surgery, Cleveland Clinic, Cleveland, Ohio, UNITED STATES. E. Rodriguez, Department of Cardiovascular Pathology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES. **Background:**Segmental arterial mediolysis (SAM) is an arteriopathy of unknown etiology that commonly involves the abdominal, retroperitoneal, cardiac and cerebral arteries. This abnormality leads to increased susceptibility to ischemia, hemorrhage and dissection. Few case reports have been described among adult population, but there is only one case of pediatric SAM described in a preterm infant. Herein, we present a case of arterial abnormality that is most consistent with SAM in a 2 year old male. **Case Presentation:**A 2 year old boy with no significant past medical history presented with a 2 day history of fever, anorexia, abdominal pain and vomiting. Physical exam revealed mild abdominal tenderness. Initial labs were significant for a lipase of 2754 U/L and was admitted for acute pancreatitis. A CT scan of abdomen revealed a cystic structure measuring 28 mm localized to the region of the pancreatic head. MRCP revealed a hyperintense structure in the pancreatic head (2.8 x 2.1 x 2.6 cm) with connection to the main pancreatic duct and a mildly dilated pancreatic duct. The patient had another episode of acute pancreatitis after 6 months. Repeat MRCP was consistent with enlargement of the pancreatic lesion now

found to be compressing the portal vein, with signs of portal hypertension including multiple varices in the stomach and splenomegaly. He underwent explorative laparotomy with open pancreatoduodenectomy, cholecystectomy with pancreaticojejunostomy, open en-bloc resection of portal vein and appendectomy. Significant intraoperative findings were: an 8 cm poorly circumscribed pancreatic head mass with invasion of the portal vein resulting in near complete occlusion of the lumen. The histological findings included evidence of foci of organizing hemorrhage surrounded by fibrosis and bland myofibroblastic proliferation admixed with dense collagen fibers. Adjacent to this large area of hemorrhage, there was evidence of an abnormal artery with intimal hyperplasia. A Movat stain revealed segmental loss of the internal elastic lamina and tunica media with features indicative of aneurysm formation. Additional vascular imaging studies did not reveal any other vascular involvement. While organizing hemorrhage secondary to prior episode of pancreatitis is in the differential, the overall findings are most indicative of a ruptured aneurysm secondary to a non-inflammatory vasculopathy, most consistent with SAM. Discussion: SAM is a rare vasculopathy of unknown etiology that is non-inflammatory and nonatherosclerotic. The incidence of SAM is unknown with most of our knowledge coming from existing case reports. The etiology of SAM is not clearly understood but it is believed to result from vasospasm due to high levels of catecholamines and other vasoactive substances in the pediatric population. The clinical presentation of SAM is reported to be highly variable ranging from asymptomatic or abdominal pain to dramatic life-threatening hemorrhage in abdomen, brain or retroperitoneum. Histologic findings in SAM include lytic degeneration of the tunica media which can lead to tearing or separation from the adventitia and fibrosis. Visceral angiography helps in identifying the vasculature involved and patterns of involvement such as stenosis, dissections or aneurysms. Diagnosis is based on a combination of clinical features, histopathological findings and visceral angiography. It is very important to distinguish SAM from other mimics causing vasculopathy such as vasculitis, connective tissue diseases or fibromuscular dysplasia. Treatment strategies include surgical management or arterial embolization.

**460 SYSTEMIC LUPUS ERYTHEMATOSUS PRESENTING WITH PROTEIN LOSING ENTEROPATHY.** P. Guha, S. Hourigan, C. Port, Pediatrics, Inova Fairfax Children's Hospital, Fairfax, Virginia, UNITED STATES. Honigbaum, S. Hourigan, Gastroenterology, Pediatric Specialists of Virginia, Fairfax, Virginia, UNITED STATES. Way, Rheumatology, Pediatric Specialists of Virginia, Fairfax, Virginia, UNITED STATES.

Case: A 17 year old female presented with 4 months of progressively worsening edema and arthralgia and 2 months of daily nausea, emesis and diarrhea with cramping. She had over 5 stools a day, including nocturnal stooling, that were watery and non-bloody. She also reported epigastric pain that was worse after eating. Initial laboratory results showed: normal blood counts and liver enzymes; low albumin (1.9g/dL), positive stool *Clostridium difficile* toxin B PCR; positive CMV IgM, negative *Helicobacter pylori* stool antigen and elevated stool alpha 1 antitrypsin (135mg/dL). She was diagnosed with Protein Losing Enteropathy (PLE) and received multiple doses of albumin and furosemide for her edema secondary to hypoalbuminemia and was started on Vancomycin for *Clostridium difficile* infection. She was also started on a low-fat, high-protein diet. Upper GI series showed prominent area gastricae thought to be secondary to gastritis, and she was started on omeprazole. Further into her hospitalization, her ANA came back positive at 1:640 as well as positive dsDNA, SSa, Sm, RNP, Lupus Anticoagulant and histone antibodies. Her C3 was very low at 29 and her C4 was undetectable. She was diagnosed with Systemic Lupus Erythematosus (SLE) and was started on treatment with IV methylprednisolone 1000mg daily for three days followed by initiation of hydroxychloroquine therapy with significant resolution of her diarrhea and anasarca. Discussion: While there is no pathognomonic characteristic to Systemic Erythematosus Lupus (SLE), some presentations are more common than others, namely arthritis/arthralgia and skin rashes. However, rarely, patients present with other symptoms, such as protein losing enteropathy, as was the case with our patient. PLE is an uncommon initial manifestation of SLE, particularly in children, with patients presenting with non-specific symptoms such as edema and low albumin. Patients with protein-losing enteropathy usually have a positive ANA, anti-double stranded DNA, and low serum complement levels. The combination of low serum albumin and low 24 hour urine protein has a high positive predictive value and specificity for SLE associated PLE. Pulse steroids followed by immunosuppressants are the mainstay of treatment for patients with SLE regardless of presenting symptoms. Given the high frequency of misdiagnosis in patients presenting with PLE, SLE should always be considered in the differential diagnosis.

**461 ACUTE PANCREATITIS SECONDARY TO PAPILLARY STENOSIS IN A PATIENT WITH CELIAC DISEASE IN REMISSION.** P. Lin, J. Wolfe, Gastroenterology, Childrens National Medical Center, Washington, District of Columbia, UNITED STATES.

Background: There are multiple reports of patients with celiac disease developing pancreatitis, although the mechanism behind this increased risk is not yet known. One proposed theory is that inflammation within the second portion of the duodenum also affects the Ampulla of Vater, leading to papillary stenosis and an obstructive pancreatitis. However, this has been seen typically in uncontrolled celiac disease or at diagnosis. We present here a case of acute pancreatitis secondary to papillary stenosis in a patient with celiac disease in remission. Case Presentation: 14 year-old male, with long-standing celiac disease in remission, developed an acute episode of epigastric abdominal pain. He had an elevated lipase with mildly elevated bilirubin and normal transaminases. Abdominal CT showed edema of the pancreatic head, with dilated common bile duct and intrahepatic ducts. This was his first instance of acute pancreatitis. ERCP demonstrated a stenotic biliary papilla with a dilated CBD to 10 mm. Sphincterotomy was performed and a common bile duct stent placed. Duodenal biopsies taken during ERCP showed no inflammation on histology. The stent was removed 6 weeks later. However, the patient then developed a recurrence of pancreatitis 7 weeks after stent removal. Repeat ERCP showed a pancreatic duct stricture that required placement of a pancreatic duct stent. Given the degree of stenosis, he required progressive stent upsizing with a repeat ERCP 3 weeks later, and followed by final removal of the stent 3 weeks later. Testing for genetic causes of pancreatitis was negative. Conclusion: This patient with celiac disease in remission for over 10 years developed acute pancreatitis with papillary stenosis. While this has been seen in uncontrolled celiac disease, this patient had no evidence of histologic inflammation on his biopsies. He later developed a reoccurrence of pancreatitis with a pancreatic duct stricture. His presentation raises a potential association between celiac disease and increased risk of ductal strictures despite adequate gluten-free diet control.

**Poster Session III**  
**Saturday, November 4, 2017**  
**12:00pm – 2:00pm**

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**506 USE OF LUMEN APPOSING METAL STENTS FOR ENDOSCOPIC DRAINAGE OF INTRA-ABDOMINAL FLUID COLLECTIONS IN PEDIATRIC PATIENTS.** P. Costa, Y. Rivas, I. Novak, Children's Hospital at Montefiore, Bronx, New York, UNITED STATES. Ho, K. karia, Montefiore Medical Center, Bronx, New York, UNITED STATES.

Background: Technological advances in endoscopic devices have improved our ability to treat intra-abdominal fluid collections that had previously required surgical intervention. Pancreatic pseudocysts and perirectal abscesses can now be drained with less invasive techniques, including the use of lumen-apposing metal stents (LAMS). LAMS have a configuration characterized by two large flanges which help to avoid stent migration as well as a large diameter to allow passage of the endoscope for endoscopic debridement of the cavity [5]. Pancreatic pseudocysts are a known complication of pancreatitis in both adult and pediatric patients. The traditional treatment was surgical drainage and debridement of necrosis. It has been shown in adults that advanced endoscopic therapy is an acceptable and safe alternative option [3]. There have been multiple studies showing the efficacy of endoscopic drainage with resolution of pancreatic fluid collections in adults [2]. Following the success of this technique in adults, pediatric gastroenterologists are now beginning to use advanced endoscopic therapies as well. One case report was published describing the use of LAMS for drainage of a pancreatic fluid collection in a single pediatric patient [1]. Endoscopic drainage of perirectal abscesses has also been recently performed using these stents. One study showed both immediate and long-term success in perirectal and perisigmoid abscess endoscopic drainage in adults [4]. This retrospective chart review evaluates the indications, outcomes, and complications of LAMS for drainage of pancreatic fluid collections and perirectal abscesses in pediatric patients at our institution. Methods: A retrospective chart review was performed to investigate the pediatric patients who underwent endoscopic drainage of fluid collections using LAMS at Children's Hospital at Montefiore between 2015 and 2017. Patients up to 21 years of age when diagnosed and treated were included. Six patients were identified and included in the study. Results: Between 2015 and 2017, six patients were identified that required endoscopic ultrasound-assisted drainage of intra-abdominal collections using LAMS. There were 2 female and 4 male patients, age ranges 9 to 20 years. Three patients had perirectal abscess post perforated appendicitis and 3 patients had pancreatic pseudocysts. Stents were in place for 2-100 days (average for perirectal abscess 16.3 days, average for pseudocyst

72 days). All patients had complete resolution of the collections. One patient had a small amount of bleeding on day 2 after stent placement and flexible sigmoidoscopy was performed with EUS showing resolution of the collection and the stent was removed. One patient required debridement of the pseudocyst twice while the stent was in place. None of the patients had recurrence of the collection. Discussion Intra-abdominal fluid collections had previously been managed surgically but recently it has been shown that endoscopic techniques can be therapeutic as well. Following success in adults, we have begun using these techniques in pediatric patients. Although the number of patients in our study is limited, we demonstrate that the use of LAMS to drain perirectal abscesses and pancreatic pseudocysts is both effective and safe in pediatric patients. Our case series shows the importance of less-invasive therapeutic options when treating these conditions. Post-procedure bleeding noted in our youngest patient (9yr) may have been avoided with the use of a smaller device. Development of smaller LAMS may be beneficial in a pediatric population. References 1. Pediatric application of the lumen-apposing metal stent for pancreatic fluid collections. MJ Giefer, BL Balmadrid - Gastrointestinal endoscopy, 2016. Safety and Efficacy of Endoscopic Ultrasound-Guided Drainage of Pancreatic Fluid Collections With Lumen-Apposing Covered Self-Expanding Metal Stents. RJ Shah, JN Shah, Irving Waxman, TE. Kowalski, A Sanchez-Yague, Jose Nieto, BC Brauer, Monica Gaidhane, Michel Kahaleh. Clin Gastroenterol Hepatol 2015; 13: 747-752. Equal efficacy of endoscopic and surgical cystogastrostomy for pancreatic pseudocyst drainage in a randomized trial. Varadarajulu S1, Bang JY, Sutton BS, Trevino JM, Christein JD, Wilcox CM. Gastroenterology. 2013 Sep;145(3)4. Long-term outcome of endoscopic ultrasound-guided pelvic abscess drainage: a two-center series. Laurent Poincloux, Fabrice Caillol, Christophe Allimant, Erwan Bories, Christian Pesenti, Aurelien Mulliez, Frederic Faure, Olivier Rouquette, Michel Dapoigny, Armando Abergel, Marc Giovannini. Endoscopy 2017; 49(05): 484-490. A new lumen-apposing metal stent for endoscopic transluminal drainage of peripancreatic fluid collections. Piotr S Wrobel, Jeremy Kaplan, and Ali A Siddiqui. Endosc Ultrasound. 2014 Oct-Dec; 3(4): 203-204

**512 COMBINED ENDOSCOPIC ULTRASOUND AND SECRETIN PANCREATIC FUNCTION TEST IN PATIENTS EVALUATED FOR CHRONIC PANCREATITIS** ROBERTO GUGIG M.D. R. Gugig, UCSF, Visalia, California, UNITED STATES.

Endoscopic ultrasonography (EUS) has increasingly been used as single modality to diagnose chronic pancreatitis (CP). While the morphologic changes seen on EUS have been well characterized, there is disagreement about whether morphologic EUS examination alone should be used as the gold standard for diagnosis, especially in patients with early or "minimal change" disease. Poor inter-rater reliability, disagreement about which features are physiologic vs. pathologic, and lack of consensus as to which morphologic features should receive more relative weight in diagnosis have led to concerns over the accuracy of EUS to diagnosis CP. Endoscopic Ultrasound (EUS) evaluation of pancreatic duct compliance following secretin-stimulation (sEUS) along with EUS morphologic examination (EUS) and duodenal fluid [HCO<sub>3</sub><sup>-</sup>] measurement (ePFT) in one endoscopic session has been reported as a means of evaluating for chronic pancreatitis (CP) in adults. We evaluated the feasibility of the combined examination and compare EUS measurements of pancreatic ductal compliance with duodenal [HCO<sub>3</sub><sup>-</sup>] for diagnosing CP in children. METHODS—Prospective case series of patients with suspected CP who underwent combined EUS, sEUS and ePFT examination in one endoscopic session. The main outcome measures were the feasibility of performing the combination examination and the correlation between ductal compliance and duodenal fluid [HCO<sub>3</sub><sup>-</sup>] measurement. RESULTS—All examinations were completed in one endoscopic session and there were no complications in 14 patients. There was a trend toward less change from baseline in head and body ductal diameter in patients with CP, but only the percent change from baseline in the tail was significant (CP 144.3% vs. normal 240.9%, p<0.01). Regression analysis demonstrated fair correlation between maximum change in ductal diameter and duodenal [HCO<sub>3</sub><sup>-</sup>]. CONCLUSIONS—Combined EUS, sEUS, and ePFTs is feasible and safe in children with suspected CP, preliminary results demonstrate positive correlation between pancreatic ductal compliance and duodenal fluid [HCO<sub>3</sub><sup>-</sup>]. Further studies are needed to determine the best protocol to follow, and if findings are reproducible at a larger scale

**515 LAPAROENDOSCOPIC RENDEZVOUS FOR CHOLECYSTOCHOLEDOCHOLITHIASIS IN AN INFANT.** S. Bitton, Pediatric Gastroenterology, Steven and Alexandra Cohen Children's Medical Center of New York, Hofstra Northwell School of Medicine, Lake Success, New York, UNITED STATES. G. Nagendra, Pediatrics, Steven and Alexandra Cohen Children's Medical Center of New York, Hofstra Northwell School of Medicine, New Hyde Park, New York, UNITED STATES. N.P. Heinzerling, J.M. Prince, Pediatric Surgery, Steven and Alexandra Cohen Children's Medical Center of New York, Hofstra Northwell School of Medicine, New Hyde Park, New York, UNITED STATES. R. Gal, M.J. McKinley,

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Choledocholithiasis is uncommon in the healthy pediatric population and rare in the infant population. Endoscopic retrograde cholangiopancreatography (ERCP) is the gold standard for treatment. However, the procedure does carry risks, most commonly post-ERCP pancreatitis. In cases with increased pre-procedure risk, laparoendoscopic rendezvous (LERV) to clear the common bile duct (CBD) and remove the gallbladder (GB) has been described as a safe option in adults. We describe a difficult case where both pediatric gastroenterology and pediatric surgery worked together to treat cholecystocholedocholithiasis in an infant. A 4 month old full term male, with an uncomplicated birth history, presented to the emergency department with concern for weight gain and recurrent irritability with feeds. On exam, vitals were stable, weight was 5.83 kg (2nd percentile), and abdominal exam was normal. Labs revealed AST/ALT 223/95 u/L, total/direct bilirubin 0.8/0.5 mg/dL. An abdominal ultrasound (US) was negative for intussusception, but revealed sludge and stones within an otherwise normal GB, a normal liver, and a 3mm echogenic focus within a 2 mm CBD. The patient was admitted and started on ursodeoxycholic acid. Serial abdominal US showed CBD dilation to 4.8mm with a persistent 3mm echogenic focus in the CBD, with increasing hyperbilirubinemia (max total/direct bilirubin 1.8/1.5 mg/dL), consistent with extrahepatic biliary obstruction. The patient was brought to the OR by the pediatric surgery and ERCP team for LERV. During laparoscopy, the liver appeared cholestatic and the GB wall thickened. The GB was dissected, mobilized, and sutured to the abdominal wall creating a cholecystostomy through which a contrast cholangiogram revealed multiple CBD stones. Several antegrade saline flushes with intravenous glucagon were unsuccessful in clearing the CBD. A guidewire was advanced through a torturous cystic duct, the CBD, and into the duodenal lumen. With surgical fields remaining intact, the ERCP team then introduced a neonatal duodenoscope to grasp the guidewire in a rendezvous fashion to access the CBD and perform a 5 mm sphincterotomy. An 8 mm balloon was advanced over the guidewire to sweep the CBD of black stones and sludge. The surgeons then flushed saline antegrade from the cystic duct until clear fluid flowed into the duodenal lumen. An occlusion cholangiogram demonstrated no filling defects. The endoscope was withdrawn, and the surgeons completed the laparoscopic cholecystectomy. The patient recovered from the procedure without complication, tolerated full feeds, and was discharged with improved labs on post-op day 2. At 1 week follow up, patient continued to have improved feeding and weight gain, and continued to thrive at the 12 month follow up visit. In conclusion, our case demonstrates that LERV is a safe and effective treatment option for infantile cholecystocholedocholithiasis. Within a single sedation, the CBD was endoscopically cleared of calculus and the GB was removed laparoscopically without the risks associated with open cholecystectomy and bile duct exploration. Furthermore, the patient was discharged within 2 days of the procedure, thriving. We believe that this is the youngest LERV reported in literature, and suggest that LERV be considered as a treatment option when the required resources are available to treat high risk pediatric cases safely.

**516** *MESENTERIC SIDED MECKEL'S DIVERTICULUM PRESENTING AS INTERMITTENT ABDOMINAL PAIN IN A PEDIATRIC PATIENT.* S. Hassan, D. Montoya Melo, B. Barth, D.M. Troendle, Pediatric Gastroenterology, University of Texas Southwestern, Dallas, Texas, UNITED STATES. Hassan, D. Montoya Melo, F. Qureshi, B. Barth, D.M. Troendle, Children's Health, Dallas, Texas, UNITED STATES. F. Qureshi, Department of Surgery, University of Texas Southwestern, Dallas, Texas, UNITED STATES.

Background Meckel's Diverticulum (MD) is one of the most common congenital anomalies of the gastrointestinal tract (GI). Clinical manifestation is usually painless rectal bleeding; however, presentation is variable, and a high level of suspicion is needed to make the correct diagnosis. <sup>99m</sup>Tc pertechnetate scintigraphy (Meckel's scan) is the primary diagnostic method for evaluation of MD but other modalities may be necessary to make the diagnosis, especially in cases of atypical clinical presentation. Case 11 year old female presented with intermittent abdominal pain, prompting multiple emergency room visits. Her pain was epigastric and occasionally radiated to the right lower quadrant. Intensity was 10/10 at its worst, and unrelated with defecation. She denied nausea, emesis, diarrhea, hematochezia or melena. Laboratory workup was unremarkable. Abdominal X-Ray and CT were obtained, showing findings of mild mesenteric lymphadenopathy in the ileocolic region with normal appendix. Her pain improved with stool softeners and anti-spasmodic medications. Eight months later, patient presented with worsening of her abdominal pain now associated with microcytic anemia. She subsequently underwent endoscopic evaluation, which showed small erosions in the terminal ileum and fresh blood in the ileum and colon. No definitive source of bleeding was identified. Capsule endoscopy was performed with findings suggestive of multiple polypoid lesions in the proximal and mid small bowel. MR enterography was obtained for further characterization,

however no intraluminal mass was identified within the duodenum, jejunum, or ileum. With no definitive diagnosis, surgery was consulted to assist with single balloon enteroscopy. A small mesenteric sided diverticulum was identified on the serosal aspect of the mid jejunum and it was resected. Histopathology revealed gastric oxyntic-type mucosa with an adjacent deep ulcer which extended to the muscularis propria consistent with mesenteric sided MD. Discussion MD is a congenital GI abnormality that results from the incomplete obliteration of the omphalomesenteric duct. It is considered a true diverticulum because it contains all three layers of the intestinal wall. MD is commonly described using the rule of 2s: 2 percent of the general population, children younger than 2 years of age, 2 feet from the ileocecal valve, 2 inches in length and contains 2 types of ectopic tissue. While typically located on the anti-mesenteric side, they have rarely been described to be mesenteric sided. The typical presentation is painless rectal bleeding, however patients can present with a variety of symptoms including diffuse abdominal pain and anemia. Meckel's scan is the diagnostic modality of choice; however MD may be diagnosed during advanced endoscopic procedures, laparoscopy or laparotomy. Conclusion Even in the absence of typical symptoms, MD should be considered in the differential diagnosis of abdominal pain.

**517 RECURRENT HSV ESOPHAGITIS IN AN IMMUNOCOMPETENT 8-YEAR-OLD.** S. Faasse, C. Sauer, Pediatric Gastroenterology, Hepatology, and Nutrition, Emory University, Atlanta, Georgia, UNITED STATES.

Introduction HSV is an uncommon cause of esophagitis in immunocompetent hosts. There are several case reports in the literature that describe HSV esophagitis (HSVE) in immunocompetent adolescents and adults, but few reports describe HSVE in school aged children, and none describe recurrent episodes. Here, we present an immunocompetent child with 3 episodes of HSV esophagitis. Case Presentation Patient initially presented at age 6 with odynophagia, fever, and inability to tolerate PO. She was admitted for IV hydration and endoscopy was performed. Grossly, multiple aphthous ulcers were visualized throughout her esophagus (Image 1). Histopathology revealed epithelial hyperplasia consistent with moderate reflux esophagitis, immunostaining for HSV I and II was positive. After starting omeprazole and valacyclovir, she improved significantly and was discharged home. 19 months later, she again presented with odynophagia. Valacyclovir was started outpatient, but she was unable to tolerate PO. She had one similar but less severe episode of odynophagia in the interim and was treated by her PCP with valacyclovir. On EGD, an aphthous ulcer in the proximal esophagus, as well as a large ulcer in the distal esophagus were visualized (Image 2). Histopathology showed active esophagitis with ulceration, granulation tissue formation, and necrotic debris. Stains for HSV I and II, CMV, and fungus were negative. However, given her history and presenting symptoms, she was treated with a 7 day course of valacyclovir. Due to recurrent HSVE episodes, she was evaluated by immunology. HIV testing was negative, TBNK profile was normal, and she was not noted to have other recurrent infections, thus it was determined that an underlying immunodeficiency was unlikely. Discussion HSVE is a rare cause of odynophagia in school age children. Because exposure to the virus is less common in this age group, underlying susceptibility to disease, whether congenital or acquired, should be considered. Additional factors, such as uncontrolled gastroesophageal reflux, may contribute to symptoms and diminish mucosal integrity. Negative immunostaining should be taken in the context of concurrent antiviral treatment, and treatment threshold should remain low in cases of high clinical suspicion. Conclusion Although rare, HSVE should be considered in children with odynophagia. In children with recurrent HSVE, it would be prudent to evaluate for underlying immunodeficiency. References Al-Hussaini AA, Fagih MA. Herpes simplex ulcerative esophagitis in healthy children. Saudi J Gastroenterol. 2011 Sep-Oct;17(5):353-6 Canalejo Castrillero E, García Durán F, et al. Herpes esophagitis in healthy adults and adolescents: report of 3 cases and review of the literature. Medicine (Baltimore), 2010, Vol.89(4), p.204-210 Ramanathan J, Rammouni M, Baran Khatib R. Herpes simplex virus esophagitis in the immunocompetent host: an overview. Am J Gastroenterol, 2000, Vol.95(9), p.2171-2176 Rodrigues F, Brandão N, et al. Herpes simplex virus esophagitis in immunocompetent children. J Pediatr Gastroenterol Nutr. 2004 Nov;39(5):560-3

**518 WHAT COMES FIRST? HEPATOBLASTOMA OR FAMILIAL ADENOMATOUS POLYPOSIS.** S. Walji-Virani, M. Semrin, Pediatric Gastroenterology, Childrens Medical Center, Dallas, Plano, Texas, UNITED STATES. Semrin, Pediatric Gastroenterology, University of Texas Southwestern, Dallas, Texas, UNITED STATES.

Familial adenomatous polyposis (FAP) is a rare genetic condition that is inherited as an autosomal dominant trait. The gene responsible for FAP is adenomatous polyposis coli (APC). The most common extracolonic manifestation of this gene is hepatoblastoma (HB) and desmoid tumors in children therefore sharing the same germline as in AFC. Given this knowledge, children with FAP are routinely screened for HB. However, children who are successively

treated for HB are routinely followed and screened for recurrence of HB or other cancers but not for FAP. A 17 years old African American male is referred by the After Cancer Care clinic for evaluation of anemia and guaiac positive stools. He had history of HB at age 4 years and was successfully treated with partial hepatectomy and chemotherapy. He was routinely followed by the above clinic and this was the first presentation of these symptoms. Initial history from patient was not significant, he denied any constipation, abdominal pain or hematochezia. Physical exam was benign except for gross blood on rectal exam. Consequently, upper and lower endoscopies were obtained that showed diffuse sessile polyps in the gastric fundus, antrum and duodenum and from the terminal ileum to the rectum. The pathology showed adenomatous dysplasia on upper endoscopy and tubulovillous adenoma on the lower endoscopy. Genetic testing performed confirmed presence of the APC gene consistent with FAP. He had laparoscopic total proctocolectomy with ileal J pouch, anal anastomosis and diverting loop ileostomy. The ileostomy was closed 3 months later. Granted occurrence of FAP is rare in children, but knowing that it shares the same gene as HB, all children with history of HB should be genetically tested for the APC gene and monitored for FAP routinely

**519 AN ELUSIVE CARCINOID TUMOR: THE IMPORTANCE OF CAPSULE ENDOSCOPY.** S. Rao, Child Health, University of Missouri - Columbia, Columbia, Missouri, UNITED STATES. J. Kantor, Child Health Gastroenterology, University of Missouri - Columbia, Columbia, Missouri, UNITED STATES.

**INTRODUCTION:** Gastrointestinal carcinoid tumors are rare in the pediatric population with most diagnosed during the 6th decade of life. Gastrointestinal carcinoid can be very difficult to suspect due to nonspecific symptoms. Small intestinal tumors present a difficult challenge, as they are not easily visualized on esophagogastroduodenoscopy (EGD) and colonoscopy and may require capsule endoscopy to avoid radiation and anesthesia risks in the pediatric population. **CASE REPORT:** A 15 year old white male with a past medical history of iron deficiency anemia, mild chronic gastritis, and ileitis presented with a 3 week history of fatigue and abdominal pain. The patient began evaluation for anemia approximately 2 years ago, which included a CBC and iron studies consistent with iron deficiency. Further workup for inflammatory bowel disease was unremarkable demonstrating a normal ESR <7 mm/h, normal CRP 0.1 mg/dL, and a negative celiac panel. An abdominal CT demonstrated distal small bowel thickening with multiple enlarged mesenteric lymph nodes, and a short segment of ileo-ileal intussusception, however the patient denied any abdominal pain at that time. Approximately 2 years later, the patient presented with chronic abdominal pain and failure to thrive. EGD and colonoscopy were unremarkable, but biopsy pathology reports noted esophagitis, chronic gastritis, and chronic duodenitis and omeprazole was initiated. A capsule endoscopy demonstrated large abscesses and ulcers with broadened villi over the distal jejunum and proximal duodenum without erythema or hyperemia. Diagnostic laparoscopy revealed a large matted section of the distal jejunum with short and thick mesentery, multiple enlarged lymph nodes, and a hard mass with multiple areas of scarring. During an exploratory laparotomy, the section of bowel with mass was resected. Histopathology revealed a multifocal well-differentiated neuroendocrine carcinoma with staining consistent with carcinoid tumor. The patient had an unremarkable postoperative course and noted increased energy, weight gain, and complete resolution of abdominal pain. **CONCLUSION:** Carcinoid tumors are rare in pediatric patients with most diagnosed in the 6th decade of life. Although the incidence of these tumors in pediatric patients is very low, the small bowel is a common location for carcinoid tumors in this population. It is imperative that we recognize and treat these cases since studies indicate a poor overall survival of those with small bowel neuroendocrine tumors. Pediatric carcinoid tumors of the small bowel are often not well visualized with EGD or colonoscopy. Diagnosis can be difficult given the nonspecific symptoms of gastrointestinal carcinoid tumors, which are often not suspected until resection. Capsule endoscopy may be advantageous in the pediatric population and also avoids the risks of radiation and anesthesia.

**520 NOT JUST FOR ADULTS: THE USE OF PUSH ENTEROSCOPY TO DIAGNOSE MECKEL'S DIVERTICULUM IN A 16-YEAR-OLD PATIENT WITH IRON DEFICIENCY ANEMIA AND OCCULT GASTROINTESTINAL BLEEDING.** S. Kolli, A. Frade Garcia, A.C. Huang, E. Hernandez, Nicklaus Children's Hospital, Miami, Florida, UNITED STATES.

Meckel's diverticulum (MD) affects 2-3% of the population. Located approximately 100 cm from the ileocecal valve, MD is a remnant of the omphalomesenteric duct. The Technetium-99m pertechnetate scintigraphy (Tc-99m) detects the MD by reacting with its gastric mucosa. It is the first-line imaging utilized for pediatric patients with gastrointestinal bleeding with a sensitivity of 85%. The Tc-99m scan is less sensitive (63%) in the adult population. When multiple imaging and scoping techniques cannot detect the site of GI bleeding, push enteroscopy has been

rarely used as an alternative technique in pediatrics. We present a case of a 16-year-old male with persistent lower gastrointestinal bleeding that was diagnosed with MD via the push enteroscopy. Our patient is a 16-year-old previously healthy male who presented with fatigue, dyspnea, and decreased exercise tolerance. He had an unremarkable birth and family history. He had generalized pallor. His weight was in the 88 percentile; his height was in the 40 percentile. His complete blood count was remarkable for low hemoglobin at 7.2 g/dL with an elevated reticulocyte count; iron studies were consistent with iron deficiency anemia. Complete metabolic panel was remarkable for transaminase elevation. His heme-occult stool was positive. His fecal calprotectin was mildly elevated at 326. His inflammatory markers and infectious work-up was negative. His Prometheus inflammatory bowel disease panel, celiac serology, hemolytic anemia genetic panel, Helicobacter pylori stool antigen, gastrointestinal panel, and bone marrow biopsy were negative. Our patient underwent an extensive but unremarkable imaging: abdominal ultrasound, computed tomography abdomen, and magnetic resonance imaging enterography. His Tc-99m scan was negative. His upper endoscopy (EGD) and colonoscopy showed mild colitis. Our patient's capsule endoscopy showed white lesions in the small intestine and a non-actively bleeding ulceration in the ileum. Our patient required blood transfusions and intravenous iron supplementation. Patient finally underwent push enteroscopy and a Meckel's diverticulum was identified. Patient underwent surgery and had clinical resolution of symptoms. Although, push enteroscopy may cause discomfort or intestinal perforation, it is a safe technique often used to evaluate proximal small bowel disease in adults. The site of the MD may be challenging to access with upper endoscopy. The push enteroscopy is able to insert almost 160 cm beyond the ligament of Treitz. Push enteroscopy have been used in children to evaluate a variety of conditions, like gastrointestinal bleeding, chronic diarrhea and failure to thrive, but have not been routinely utilized. Push enteroscopy have been shown to have a higher diagnostic yield with the same procedure time compared to the EGD in children. Push enteroscopy was able to safely and effectively diagnose MD in our patient after extensive, unremarkable imaging and scoping techniques. We propose to include push enteroscopy as a safe, viable option in evaluating gastrointestinal bleeding in children.

**522 SMALL BOWEL INTUSSUSCEPTION IN A 16-MONTH-OLD CHILD WITH PEUTZ-JEGHERS SYNDROME.** R. Prasad, S. Ciullo, Section of Pediatric General, Thoracic and Minimally Invasive Surgery, St. Christopher's Hospital for Children, Philadelphia, Pennsylvania, UNITED STATES. R. Prasad, S. Ciullo, Department of Surgery and Pediatrics, Drexel University College of Medicine, Philadelphia, Pennsylvania, UNITED STATES. A. Zacharias, Department of Pediatrics, St. Christopher's Hospital for Children, Philadelphia, Pennsylvania, UNITED STATES. G. Mallon, Division of Pediatrics and Radiologic Sciences, St. Christopher's Hospital for Children, Philadelphia, Pennsylvania, UNITED STATES. G. Mallon, H. Pall, Department of Pediatrics, Drexel University College of Medicine, Philadelphia, Pennsylvania, UNITED STATES. Marinovich, Department of Pathology and Laboratory Medicine, St. Christopher's Hospital for Children, Philadelphia, Pennsylvania, UNITED STATES. H. Pall, Section of Gastroenterology, Hepatology, and Nutrition, St. Christopher's Hospital for Children, Philadelphia, Pennsylvania, UNITED STATES.

Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant inherited condition characterized by hamartomatous gastrointestinal polyps, mucocutaneous pigmentation, and a predisposition for malignancy. Most patients with PJS are diagnosed in the second or third decade of life, commonly complicated by intussusception at the median age of onset of 15 to 16 years. This report describes a rare case of a 16-month-old male with PJS who presented with a small bowel intussusception caused by a polyp that served as a lead point within the intestine. He was diagnosed with PJS at the young age of 6 months with a history of rectal and antral polyps. A mutation in the *STK11* gene was detected in this patient and his father, who also has PJS. The patient presented with abdominal distention with subsequent ultrasound revealing a small bowel intussusception. He underwent a diagnostic laparoscopy, where two areas of intussusception adjacent to each other were noted. There was a 4.2 cm polyp serving as a lead point, internally involving most of the circumference of the intestine (Figure 1.A and 1.B). Subsequently, a very short segmental small bowel resection and primary anastomosis were performed. To our knowledge, this is the youngest patient with PJS to have developed a small bowel intussusception due to a polyp. It is important for the medical community to be aware that serious complications such as intussusception can occur early in infants and young children with PJS. Further research is needed to provide evidence-based guidelines on the screening strategy for these young patients.

**527 CHOLANGIOSCOPY FOR RETRIEVAL OF MIGRATED BILIARY STENT IN A PEDIATRIC PATIENT.** T.L. Piester, Pediatric Gastroenterology, Stanford University, Palo Alto, California, UNITED STATES. J. Carroll, D.M. Troendle, B. Barth, Pediatric Gastroenterology, Children's Medical Center, Dallas, Texas, UNITED STATES.

Endoscopically placed biliary stents are frequently used for the treatment of biliary tract obstruction (1). Stents differ by design (pig tail vs straight), size, and material (plastic vs. metal). Plastic stents are commonly used and are usually designed as a straight stent with flaps at each end to avoid migration into or out of the duct (2-3). Despite the flap design, migration can occur with data showing proximal migration in up to 5% of cases (4-5). Methods to retrieve migrated stents include using a snare, Soehendra retrieval catheter, forceps, baskets, and balloon dilators (5-8). In patients with biliary stricture or a tight duct, guidewire cannulation of the stent lumen is thought to be preferred. Cholangioscopy utilizes a through the scope device that allows direct high resolution visualization of the bile duct or pancreatic duct lumen. Here, we report the use of cholangioscopy to retrieve a migrated biliary stent in a pediatric patient following failure of traditional retrieval techniques. **METHODS/CASE** 12 year old (25 kg) male with a history of ornithine-transcarbamylase deficiency underwent orthotopic liver transplantation in October 2005 which was complicated by postoperative hepatic artery thrombosis. He developed intrahepatic biliary strictures requiring multiple ERCPs involving biliary dilation and stenting to establish and maintain drainage. Ultimately, a 7 Fr x 9 cm biliary stent was noted to migrate proximally into the bile duct. The initial attempt at stent retrieval failed using a biliary dilation balloon, stone extraction balloon, stone extraction basket, snare, and Roth net. A guidewire could not be passed through the stent due to difficult position against the wall of the distal bile duct (Image 1). The patient was monitored over months with stable AST, ALT, bilirubin, and GGT and no adverse events related to the stent's position were appreciated. Stent retrieval using cholangioscopy was attempted 6 months after placement. A 10FR cholangioscope did not initially fit in the duct. A 4 to 6 mm Soehendra dilator catheter was used to dilate the strictured areas of the duct in hopes of altering the position of the stent, and loosening any resistance the stricture might cause that would affect removal. Next, a 6 mm biliary dilator balloon was used to dilate the distal aspect of the duct to permit passage of the cholangioscope. The Spyglass cholangioscope (Boston Scientific) was inserted into the duct and a guidewire was passed through the stent under direct visualization (Image 2) and appreciated under fluoroscopy (Image 3). At this point, a snare was passed over the wire, through the duodenoscope channel and ultimately into the bile duct surrounding the retained stent. It was closed around the stent, and the stent was successfully extracted (Image 4). **DISCUSSION** To our knowledge, this is the first reported case of cholangioscopy-assisted retrieval of a proximally migrated biliary stent in a pediatric patient. In cases where conventional, less expensive retrieval methods fail, direct visualization afforded by cholangioscopy may safely facilitate retrieval of migrated biliary stents in pediatric patients.

**532 REFLUX ASPIRATION IN LUNG INJURY AMONG CHILDREN WITH DYSPHAGIA AND ORAL ASPIRATION.** J.B. Osborn, Graduate Medical Education, Phoenix Children's Hospital, Phoenix, Arizona, UNITED STATES. S. Schroeder, D. Williams, Gastroenterology, Phoenix Children's Hospital, Phoenix, Arizona, UNITED STATES. L. Mirea, Clinical Research, Phoenix Children's Hospital, Phoenix, Arizona, UNITED STATES. Rao, J. Woodward, J. Smith, Pulmonology, Phoenix Children's Hospital, Phoenix, Arizona, UNITED STATES.

**Introduction and Background:** Swallowing is a complex mechanism, and dysphagia, defined as difficult or abnormal swallowing, can occur at any time during swallow (oral, pharyngeal, esophageal). Prevalence of dysphagia in typically developing children is estimated at 25-45% and reaches 80% in development disorders. Treatment approaches vary depending on underlying reflux aspiration and presence of chronic respiratory symptoms. Therefore, it is important to identify and characterize patients with significant respiratory symptoms and disease resulting from reflux aspiration, to ensure appropriate treatment of reflux, including the use of laparoscopic anti-reflux surgery. **Methods/Experimental Approach:** This is a descriptive, retrospective chart review study was approved by Phoenix Children's Hospital (PCH) IRB. Subjects included pediatric patients with dysphagia and proven or at risk for aspiration on MBS, seen at the Aerodigestive Clinic at PCH between March 1<sup>st</sup> 2015 to December 31<sup>st</sup> 2016. All study subjects received standard of care testing for dysphagia and oral aspiration. Reflux was measured with a multi luminal channel pH impedance probe and reflux esophagitis was assessed by esophageal biopsies. Bronchoalveolar lavage (BAL) was collected via a standard method at bronchoscopy and pepsin was measured using a pepsin A ELISA assay. Symptom review questionnaires were obtained per ADC clinic protocol at the time of first evaluation or by chart review. **Results:** A total of 12 (out of 100 screened) patients were included, with age 24-127 months old and gestational age 24-39 weeks. Subjects were predominantly female (75%) and Caucasian (75%). MBS results ranged from mild to severe dysphagia with 58%

(7/12) having severe dysphagia. Only 25% (3/12) of patients had an abnormal DeMeester score > 15 and 50% (6/12) had esophagitis on esophageal biopsy. The extent of respiratory symptoms varied, with patients experiencing anywhere from zero symptoms to 17/18 symptoms on a daily basis. No patient had significant pepsin A levels found on BAL, defined as > 3.0. Additionally, there were no trends relating the degree of dysphagia and acid reflux parameters on pH impedance or esophageal biopsy results, demonstrating dysphagia and acid reflux may be independent of each other in this population. No correlation was observed between respiratory symptoms and pathologic reflux or weakly acidic and non-acidic reflux episodes. Conclusions: While it is common to see dysphagia and chronic respiratory symptoms in the young Aerodigestive population, the prevalence of reflux aspiration appears to be much lower. In our small cohort, no patient had a significant level of pepsin found on BAL despite a range of acid reflux burden. This indicates that acid reflux aspiration may not play a significant role in the Aerodigestive population. Moreover, this indicates that pepsin is likely not an accurate biomarker for reflux aspiration in patients with dysphagia. A prospective study is planned to further characterize the mechanisms of persistent symptomatic dysphagia and gastroesophageal reflux disease. Funding: Funding is provided from the University of Arizona College of Medicine Valley Research Partnership grant and the Connell Family Aerodigestive and Research Fund.

**533 FOOD IMPACTION SECONDARY TO PROGRESSIVE HISTOPLASMOSIS.** M.I. Abbas, Pediatrics, Minnesota Gastroenterology, PA, Minneapolis, Minnesota, UNITED STATES. J.J. Short, Pediatric Surgical Associates, Minneapolis, Minnesota, UNITED STATES. W.F. Pomputius, Infectious Disease, Children's of Minnesota, Minneapolis, Minnesota, UNITED STATES.

A 14 year old previously healthy Caucasian male presented with a one month history of chest pain and progressively worsening dysphagia. He was admitted after having reported that he felt "meat stuck" in his chest. He had eaten a hot dog the day prior to admission when his chest pain worsened significantly. Symptoms included foaming regurgitation, chest tightness, dyspnea, chest pain, difficulty swallowing and a 2 kilogram weight loss over the past month. He had been evaluated at an outside emergency department one month prior for the chest pain with workup noting a chest computed tomography (CT) scan that had reported evidence of prior granulomatous infection with calcified and mildly enlarged subcarinal nodes, calcified left hilar nodes and calcified granuloma in left lower lobe (Image). Patient was given anti-acids and discharged with follow-up with family being told that the symptoms were unrelated to the CT findings. Patient had not reported a previous history of cough, extensive travel although lives in Minnesota along the Mississippi river valley. Patient underwent upper endoscopy which noted a 6 cm piece of hot dog lodged in the mid to upper esophagus. This was removed successfully and on visual inspection there was an appearance of external mass effect in the mid-esophagus. Biopsies did not demonstrate esophagitis. Extensive infectious disease workup revealed positive *Histoplasma capsulatum* titer with negative urine antigen. Patient had repeat CT of the chest which showed enlargement of the conglomeration of lymph nodes seen within the subcarinal region which measured approximately 4.8 x 4.4 cm in maximal axial dimension (previously measuring approximately 3.8 x 3.0 cm. Patient underwent mediastinoscopy with excisional biopsy of his posterior mediastinal mass and the pathology report showed necrotizing granulomatous lymphadenitis along with small yeast organisms consistent with Histoplasma. Patient was treated with itraconazole and noted resolution of his dysphagia and chest pain after one month. Esophageal dysphagia is commonly encountered in a number of intrinsic mucosa conditions such as gastroesophageal reflux disease or eosinophilic esophagitis. Patients typically present with difficulty swallowing, chest pain, regurgitation or vomiting. Food impaction can be a also a common presentation of dysphagia and is more commonly associated with esophagitis and esophageal stricturing. Rarely, extrinsic causes are identified stemming from external compression of the esophagus. Mediastinal masses have long been identified as cause of dysphagia in children. Serious conditions of progressive dysphagia with an enlarging mass in the mediastinum can be concerning for malignancy. Benign conditions are the leading cause of mediastinal masses and, in the United States Ohio and Mississippi river valleys, mediastinal lymphadenopathy from Histoplasmosis is a frequent cause of an enlarging mediastinal mass. The mass is typically associated with granulomatous mediastinitis with a conglomeration of lymph nodes that are matted together and under caseation necrosis. We presented an interesting case of food impaction occurring with a rare progressive enlargement of granulomatous mediastinitis associated with pulmonary Histoplasmosis.

**534 THE UTILITY OF CYP2C19 TESTING IN THE MANAGEMENT OF A PATIENT WITH REFRACTORY GASTROINTESTINAL ULCERATION.** M. Gonzalez, I. Absah, Pediatric Gastroenterology, Mayo Clinic, Rochester, Minnesota, UNITED STATES.

**Introduction:** The field of pharmacogenomics has been evolving as an individualized means to achieve better clinical outcomes in patient care. Proton pump inhibitors (PPIs) are one of the most commonly utilized drugs in the pediatric gastroenterology practice. PPIs are known to be metabolized by the cytochrome P450 system, particularly by CYP2C19. Each individual's ability to metabolize PPIs may predict therapeutic response and potential side effects. **Case report:** 16 year old male who presented to the clinic with a 5 year history of recurrent vomiting and diarrhea. He had been previously found to have gastric and duodenal ulcerations by esophagogastroduodenoscopy (EGD) but had failed to respond symptomatically to standard doses of several proton pump inhibitors, with some inconsistent improvement on 40 mg BID of esomeprazole. His work-up was otherwise negative, including complete blood counts, inflammatory markers, vitamin B12, celiac serologies and permissive genes, ANCA, ASCA, gastrin level, plasma and urine metanephrines, parathyroid hormone and phosphorus, prolactin, thyroid function, and a brain MRI. Due to a strong family history of multiple endocrine neoplasia type 1, a PET-CT scan was done, which was negative for an octreotide-avid neoplasm, however showed increased tracer activity throughout the 2nd and 3rd portion of duodenum. Genetic testing revealed no pathogenic mutation or variant of the MEN1 gene. A repeat EGD confirmed the presence of multiple gastric and duodenal ulcers, some with raised margin and deep mucosal involvement. Due to an essentially negative work-up with persistent gastric and small bowel ulcerations, pharmacogenetics testing for CYP2C19 genotype was done. The patient was found to be a CYP2C19 ultrarapid metabolizer. Based on this, esomeprazole dose was increased by 100% to 80 mg BID, which resulted in resolution of his vomiting and diarrhea episodes. **Discussion:** This case demonstrates the value of pharmacogenomics in the clinical management of select patients. As a known cytochrome enzyme involved in the metabolism of PPIs, CYP2C19 genotyping may predict treatment response. As in the above case, ultrarapid metabolizers will often require up to a 100-200% increase of the conventional dosing or a change to another medication class. Testing should be considered in patients who require PPI therapy for specific diseases but who fail to respond despite adequate dosing. **Conclusion:** Cytochromes pharmacogenomics testing is useful in the management of patients with refractory gastroesophageal reflux disease.

**540 PRE-DUODENAL PORTAL VEIN AND IT'S RARE ASSOCIATION WITH A MECKEL'S DIVERTICULUM IN A PEDIATRIC PATIENT.** N. Israel, R. Walia, Pediatric Gastroenterology, Mercer University School of Medicine, Macon, Georgia, UNITED STATES. ISSA, J. Fernandez, A. Pavuluri, Y. Head, Department of pediatrics, Medical center of Georgia, MACON, Georgia, UNITED STATES. J. Glenn, Medical center of Georgia, Department of Pediatric Surgery, Atlanta, Georgia, UNITED STATES.

A pre-duodenal portal vein (PDPV) is a congenital anomaly, that is rare in both children and adults, which results from the persistence of a primitive vitelline vein. Rather than passing inferiorly and behind the pancreas, the portal vein crosses in front of both the duodenum and pancreas. PDPV is usually found as an incidental finding during surgeries of the gastrointestinal tract. Complications resulting in intestinal obstruction, due to an extrinsic compression of the duodenum, are rare. In fact, in a 25-year retrospective study in a single center, PDPV was found only in five neonates. In all of them, the PDPV was asymptomatic, and the duodenal obstruction was due to associated malformations, such as malrotation, duodenal atresia, duodenal web, or annular pancreas. Most of the cases of PDPV reported have been described in association with other congenital anomalies including heterotaxia, polysplenia syndrome, situs inversus, cardiac defects, malrotation, biliary or duodenal atresia, and annular pancreas. To our knowledge, out of the 100 pediatric and adult cases reported, an association with a Meckel's diverticulum (MD) has never been described. We hereby, report the case of a patient with PDPV associated with a MD with emphasis on surgical removal of such Portal vein anomalies, which is controversial. A 14-month old male with a history of heterotaxy syndrome, left atrial isomerism, severe gastroesophageal reflux disease, and failure to thrive with malrotation that was noted on an upper GI series. Surgical exploration revealed affixation of the midgut without dilatation or volvulus of the duodenum. A vascular structure crossing the duodenum anteriorly was identified as a PDPV in association with a MD. Since no signs of duodenal obstruction or inflammation were noted, the PDPV was left in place. A Ladd's procedure to correct the malrotation was performed along with removal of the MD. Since surgery, patient has remained asymptomatic with weight gain and improvement of emesis. Although, an incidental finding, this anomaly is of great surgical importance as it can cause unexpected surgical complications

from accidental injury to the portal vein especially during removal of associated gastrointestinal anomalies. Surgical removal depends on findings of extrinsic compression of the duodenum, leading to obstruction.

**542** *A NOVEL APPROACH TO THE SIX FOOD ELIMINATION DIET FOR EOSINOPHILIC ESPGHAGITIS: BLENDERIZED SFED TUBE FEEDING.* S. Schwartz, R. Shaykin, M. Manual-Rubio, J. Wechsler, A. Kagalwalla, B. Wershil, PEDGI, Lurie Children's, Chicago, Illinois, UNITED STATES. Kagalwalla, Pediatrics, Cook County Health and Hospitals System, Chicago, Illinois, UNITED STATES.

Introduction: Dietary treatment of eosinophilic esophagitis (EoE) that includes exclusive elemental diet therapy with an amino acid based formula has demonstrated 83-97% efficacy in children with EoE. Six Food Elimination Diet (SFED), which excludes foods containing cow's milk, soy, wheat, egg, peanut/tree nut and fish/shellfish is the alternate dietary approach to treating EoE in children and adults and has an efficacy of 74%. Blenderized tube feedings (BTF) which are a combination of foods and liquids that are pureed and administered through a feeding tube are increasingly requested as patients express significant desire to pursue holistic and organic diets. SFED administered as BTF has not been previously reported. Case study: A 22 month old girl with eczema, poor weight gain, solid food aversion, eosinophilia and EoE was started on elemental diet administered by NG tube feeding. Follow up endoscopy on exclusive elemental formulation demonstrated complete histological remission of EoE. Subsequently, the patient was instructed to reduce NG elemental formula by 25% and start toddler on SFED. Repeat endoscopy on this regimen demonstrated continuing histological remission. Patient oral intake however continued to be inadequate requiring G-tube placement and feeding therapy. Concerns about the composition of the elemental formula by the family and the ongoing cost of the elemental formula led to interest in BTF. The family's ability to be able to prepare the blenderized SFED was assessed as well as the feeding tube size as sufficient for the diet. Education on food sanitation and preparation of blenderized feedings in the home as well as recipes were developed within the constraints of the SFED to meet nutritional needs of the toddler. Recipes provided to family started with using elemental formula as a base, and transitioned to allowed alternative milk beverage as tolerated. At the time of the next endoscopy 4 months later, CT had been getting 50% of allowed foods by mouth, and 50% via g-tube feedings; the SFED blenderized diet with allowed alternative milk beverage as a base and elemental diet every other day. EGD with patient on this regimen again demonstrated histological remission of EoE. CT's weight and height, were monitored as well as analysis of recipes for nutritional adequacy and compliance with SFED. CT tolerated BTF with no symptoms of intolerance to feedings was able to maintain growth velocity as had done with elemental formula. Patient went on to successfully add foods back, as evidenced by follow up endoscopies showing continued histological remission using BTF during the reintroduction stage of dietary treatment. Conclusion: This case illustrates the potential to safely provide SFED as a therapy for EoE administered via BTF in the medically stable, motivated patient. This should be studied in a larger series to validate our observations.

**546** *EOSINOPHILIC GASTROENTERITIS PRESENTING AS ACUTE PERFORATED DUODENAL ULCER.* C. Scherer, U.P. Phatak, Pediatrics, Yale University, New Haven, Connecticut, UNITED STATES. Cleary, D. Ozgediz, Surgery, Yale University, New Haven, Connecticut, UNITED STATES.

Introduction: Eosinophilic gastroenteritis is a rare condition of the gastrointestinal tract. The clinical presentation is most often chronic, and depends on the location and degree of eosinophilia present. Patients frequently have a history of atopy, peripheral eosinophilia, and most commonly present with non-specific symptoms such as abdominal pain, vomiting, and diarrhea. We present a case of a previously healthy 13-year-old male who presented with an acute abdomen secondary to duodenal perforation, and was later found to have eosinophilic gastroenteritis. Case Report: A 13-year-old male presented to the emergency department with acute onset of severe abdominal pain, nausea, and emesis. He was previously healthy, with no history of trauma, NSAID use, or abdominal complaints. There was no family history of ulcer disease or GI bleeding. On arrival, he was tachycardic and ill-appearing and physical exam was consistent with peritonitis. Abdominal CT showed marked thickening of the first portion of the duodenum and surrounding inflammatory changes with a large amount of intraperitoneal free fluid and air. During open laparotomy, a 5-mm hole in the 1<sup>st</sup> portion of the duodenum draining bilious fluid was identified, consistent with duodenal perforation. The surrounding duodenum and distal stomach appeared inflamed. An omental graham patch was sutured in place. He had an uneventful post-operative recovery, yet the cause of inflammation and perforation remained unknown. Labs showed a normal hemoglobin 14.1 g/dL, WBC 7.5 x 1000/uL, gastrin level was 35 pg/mL, and no peripheral eosinophilia was present. Despite normal serum

*Helicobacter pylori* IgG, he was treated empirically for *Helicobacter pylori* with Amoxicillin, Clarithromycin and Protonix given the unclear etiology of his duodenal perforation. Few months later, an upper endoscopy with biopsies was performed. Upper endoscopy was visually normal; however, chronic inflammation and marked eosinophilia were present in gastric and duodenal biopsies. He was asymptomatic at time of the endoscopy and was not treated. Stool studies were negative for ova and parasites. Allergy testing was negative. He did implement a dairy restrictive diet empirically. Repeat upper endoscopy and colonoscopy, after dietary changes, were grossly and histologically normal, with no eosinophils present on biopsies. Discussion: We report eosinophilic gastroenteritis as a rare condition that can affect the mucosal, muscular, and serosal layers individually or may occur with transmural inflammation. In the absence of other identifiable etiologies, our patient's acute duodenal perforation was most likely secondary to transmural eosinophilic inflammation. Perforation due to duodenal ulcer is rare in children, with most cases attributed to H Pylori infection, NSAID use, or Zollinger-Ellison syndrome. This case is an atypical presentation of a rare condition in children, and only few other similar cases have been reported. Conclusion: As the prevalence of eosinophilic gastrointestinal disease is increasing, we must be familiar with the wide range of presentation, including life-threatening surgical emergency with peritonitis due to intestinal perforation. Reporting such cases is critical to improve our understanding of disease prevention, pathogenesis, and improving treatment.

**547 AERODIGESTIVE PICK UP : NON PULSATILE COMPLETE VASCULAR RING ON TRIPLE ENDOSCOPY IN A CHILD WITH STRIDOR.** V. Bhardwaj, Pediatric Gastroenterology, Children's Hospital Los Angeles, Los Angeles, California, UNITED STATES.

Background: Vascular rings are known to cause noisy breathing, difficulty in feeding, persistent cough and gastroesophageal reflux; which is a frequently encountered symptom constellation in children seen in an Aerodigestive center. Triple endoscopy is a procedure performed in Aerodigestive centers wherein laryngoscopy, bronchoscopy and esophagogastroduodenoscopy is performed in a single sedation as an evaluation for children with breathing and eating problems. Case Description: 10-month-old male with congenital blindness and developmental delay presented with chronic cough, stridor, feeding difficulties and suspected gastroesophageal reflux since birth. He was born full-term with minimal oxygen requirement at birth and 48-hour neonatal intensive care unit stay. Around 2 months of age he was started on high dose ranitidine for suspected gastro-esophageal reflux disease with which he was less fussy however his loud breathing, cough and congestion persisted. Over the next several months he had repeated bouts of upper respiratory infections that required nebulized albuterol, nebulized steroids and oral antibiotics to which his symptoms responded although only temporarily. He was described to have textural sensitivities to different consistency foods and had intermittent choking/gagging with solids. He was a slow eater however maintained his weight for length at 50th percentile. He was sent to pediatric gastroenterology for an evaluation from where he was referred to the Aerodigestive center. He underwent triple endoscopy which revealed mid esophageal luminal narrowing and external right anterior tracheal compression near carina by a non-pulsatile mass. Type 1 laryngeal cleft and mild bronchomalacia were also noted. A CT angiogram with 3D-reconstruction of airway and an echo were obtained which showed right aortic arch with mirror image branching. Diagnosis of complete vascular ring anomaly with a dominant right-sided aortic arch was confirmed intra-operatively and patient underwent division of double aortic arch, aortopexy and repair of complete vascular ring. At 1-month follow up clinic visits he had no audible stridor or cough. Modified barium swallow study performed post procedure demonstrated laryngeal penetrations with thin liquids and patient did well on nectar thick consistency foods. His genetic work up thus far has been negative for an underlying genetic syndrome. Conclusion: This case demonstrates an atypical presentation of complete vascular ring as a non-pulsatile mass. It was an unusual diagnostic pick up of an uncommon congenital cardiac anomaly. The case highlights the benefits of team approach and collaborative efforts in Pediatric Aerodigestive Medicine.

**551 UNCOMMON CASE OF VOMITING.** C. Chen, C.S. Vijay, Pediatrics, West Virginia University, Morgantown, West Virginia, UNITED STATES.

A 14 year-old female presented with a several year history of vomiting with dysphagia to solids and liquids associated with a 10 lb. weight loss over the past several weeks. She presented to our facility with worsening symptoms. She complained of vomiting without any notice, which had limited her social activities due to the embarrassment of vomiting. Her vomitus had the appearance of undigested food without signs of blood. On admission, she appeared acutely ill, but did not have any other abnormalities on physical exam. Her weight and

height were at the 50th and 83rd percentiles, respectively. The rest of her review of systems was negative. Her vitals were appropriate, and her complete blood count, electrolytes, liver function tests, and thyroid studies were all within normal limits. Pediatric Gastroenterology was consulted, as there was concern for achalasia, eosinophilic esophagitis, and pseudoachalasia. A barium swallow was performed which showed esophageal dilatation with a distal short, smooth tapering segment with complete lower esophageal sphincter relaxation failure (even two hours after observation). Both primary and secondary peristalsis were absent throughout the esophagus, and only tertiary waves were visualized. Manometry was subsequently performed and showed absent peristalsis and a tight lower esophageal sphincter. An endoscopy with biopsy was subsequently performed. The endoscopy showed a significant amount of retained food in the middle and distal esophagus, but showed no other abnormalities. The biopsy showed no histological abnormalities. These results were consistent with a diagnosis of primary achalasia. During her hospital stay, she underwent pneumatic balloon dilatation. Her diet was slowly advanced following the procedure and she was started on a proton pump inhibitor. She experienced improvement in her symptoms during her hospital stay and was able to tolerate regular foods by the time of discharge. Discussion Achalasia is a rare motor disorder of the esophagus that frequently presents with dysphagia, regurgitation, and weight loss. This condition usually occurs in adults between the ages of 25 and 60, and is very rare in pediatric patients. The incidence is about 1 in 100,000 in the general population, and only about 5% of those cases occur in pediatric patients. In many patients, it can be difficult to diagnose and is frequently misdiagnosed as gastroesophageal reflux disease. Many teenagers may be mistakenly diagnosed with an eating disorder. The rarity of this condition means that diagnosis may often be delayed for years, as was seen in this patient. Therefore, diagnosis of this condition requires strong clinical suspicion and referral to a pediatric gastroenterologist.

**553 THE ROLE OF PYRIDOSTIGMINE IN PEDIATRIC MOTILITY DISORDERS.** M. Manini, R. Grothe, M. Hager, Division of Pediatric Gastroenterology, Mayo Clinic, Rochester, Minnesota, UNITED STATES. Manini, M. Camilleri, Clinical Enteric Neuroscience Translational and Epidemiological Research (CENTER), Mayo Clinic, Rochester, Minnesota, UNITED STATES. Lu, C. Di Lorenzo, Division of Pediatric Gastroenterology, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES.

Background: Gastrointestinal (GI) motility disorders are common in children. Treatment is challenging with limited medical and surgical options. Pyridostigmine, acetyl cholinesterase inhibitor, increases acetylcholine at the neuromuscular junction promoting intestinal contractions. Little is known about pyridostigmine role and dosing in pediatric GI motility disorders. Methods: Case series of children with GI dysmotility managed with oral pyridostigmine. Patients' diagnoses include chronic intestinal pseudo-obstruction, gastroparesis with delayed small bowel transit, chronic constipation with failure to thrive, and prolonged ileus after pelvic surgery with chronic opioid use. Results: Pyridostigmine was effective and safe in all cases. Pyridostigmine decreased abdominal distention, increased bowel movements frequency, and improved enteral feeding tolerance. Effective dosing ranged between 0.25-2.0 mg/kg/day. Conclusion: We found oral pyridostigmine to be helpful in children with different GI motility problems. Pyridostigmine should be considered in such patients when other treatment interventions have not been beneficial.

**555 UNDIAGNOSED HYPOTHYROIDISM: THE ETIOLOGY OF MYSTERIOUS HALITOSIS AND CONSTIPATION.** M.B. BEG, Pediatrics, SUNY Upstate Medical University, Golisano Children's Hospital, SYRACUSE, New York, UNITED STATES. Baig, Biological Sciences, University of Illinois at Chicago, Chicago, Illinois, UNITED STATES. Ahmad, Biological Sciences, Le Moyne College, Syracuse, New York, UNITED STATES. SF. anwer, Pediatrics, Auburn Hospital, Auburn, New York, UNITED STATES.

6-year-old was seen by pediatrician for chronic constipation. Over the course of the year, the parents noted halitosis paralleled with bouts of constipation. Work up revealed thyroid stimulating hormone level (1626 IU/mL) exceeding the upper limit of normal value (0.34-4.82 IU/mL) by 33,700%. T4 was 0.21 ng/dL (0.77-1.60). Diagnosis of hypothyroidism was made and treated with resolution of halitosis and chronic constipation. Halitosis or bad breath can be indicative of underlying diseases. In general, intraoral conditions, like insufficient dental hygiene, periodontitis or tongue coating, are considered to be the most important cause (85%) of halitosis. Therefore, dentists and periodontologists are the first-line professionals to be confronted with this problem. Ear, nose, throat-associated (10%) or gastrointestinal and endocrinological (5%) disorders may contribute to the problem as well. The production of volatile sulfur compounds produced by microbial metabolic process is the main cause of halitosis. Hypothyroidism results in intestinal hypomotility, slower stool movement and increased water extraction.

In these cases of dysmotility, a fecal mouth odor may be detectable. We present this case due to its rarity, complexity and unusual nature and look to explore the underresearched relationship between halitosis and hypothyroidism.

**558 HEMORRHOIDS AND RECTAL PROLAPSE": AN UNUSUAL PRESENTATION OF ABERNETHY MALFORMATION".** N.M. Blondet, L. Ambartsumyan, Gastroenterology and Hepatology, Seattle Children's Hospital, Seattle, Washington, UNITED STATES.

**Case Description:**A 6 year old female presented to our clinic for evaluation of rectal prolapse and hemorrhoids, which had been noted before the age of one. She had history of bloody stools, which were not associated to painful defecation. A prior flexible sigmoidoscopy revealed no polyps or mucosal abnormalities and exam under anesthesia had revealed findings consistent with rectal prolapse. She was diagnosed with chronic constipation and encopresis, and started in an aggressive regimen of stool softeners and laxatives. Upon evaluation at our institution, pelvic imaging revealed a large prolapsing rectal varix, which had developed secondary to collateral circulation from an absent portal vein (Abernethy malformation type I). She subsequently underwent surgical correction via mesocaval shunt, and had bowel continence for the first time ever shortly after surgery.**Discussion:**External hemorrhoids and prolapsed internal hemorrhoids are found primarily in older patients and rectal prolapse seldom occurs in children who do not have an underlying predisposing condition. Rectal varices should be included in the differential of children with protruding anorectal lesions that resemble hemorrhoids, especially when associated with bloody stools despite appropriate bowel management.

**567 THE VALUE OF HIGH RESOLUTION ESOPHAGEAL MANOMETRY(HREM) ON ACHALASIA TREATMENT OUTCOMES IN PEDIATRIC PATIENTS.** R.A. Gomez Suarez, Pediatric Gastroenterology, Nemours Childrens Hospital, Orlando, Florida, UNITED STATES. J. Locklear, J.P. Franciosi, Pediatric Gastroenterology, Nemours Children's Hospital, Orlando, Florida, UNITED STATES. R.A. Gomez Suarez, J. Locklear, J.P. Franciosi, Pediatrics, University Of Central Florida, Orlando, Florida, UNITED STATES. Casas Melley, T. Westmoreland, C. Reyes ferral, Pediatric Surgery, Nemours Children's Hospital, Orlando, Florida, UNITED STATES. Casas Melley, T. Westmoreland, C. Reyes ferral, Surgery , University of Central Florida, Orlando, Florida, UNITED STATES.

**Introduction** The role of High Resolution Esophageal Manometry (HREM) among children with achalasia for diagnosis and intervention remains poorly characterized. **Objective** The aim of the study is to explore the value of HREM on achalasia treatment outcomes in pediatrics at Nemours Children's Hospital. **Methods** We reviewed medical records and HREM trace in patients diagnosed with achalasia at Nemours Children's Hospital during the period of 2013-2017. Positive outcomes were defined as having absence weight loss, dysphagia, regurgitation, and chest pain according to the Eckardt score. **Results:** 9 patients underwent HREM for diagnosis of achalasia. eight boys and one girl age  $15 \pm 1.9$  years old underwent HREM. One of the patients had type I, 4 patients had type II, and 4 had type III achalasia. One of the patients had Down syndrome and two patients had Triple A Syndrome. Of the two patients with triple A syndrome one type I and the other had type II. Two patients received dilation therapy, six patients received the Heller Myotomy, one received Botox therapy, and one received perioral endoscopic Myotomy, (POEM) therapy. The patient with type I achalasia underwent all four of the therapies listed above. 8/9 patients showed positive outcomes, one patient showed no change and one patient had worse symptoms. The patient with type I achalasia improved after The POEM. 3/4patients with type II received the Heller Myotomy and all of those patients showed improved symptoms. All patients with type III, three had Heller Myotomy and all improved. The patients who received non-surgical treatment showed no change or worsening in symptoms. **Conclusion** HREM in children may be useful to characterize achalasia subtypes and to help guide intervention.

**569 NOVEL USE OF PROKINETIC MEDICATIONS IN PEDIATRIC OGILVIE'S SYNDROME IN THE SETTING OF ACUTE SICKLE CELL CRISIS.** S. Mansoor, E. Kutsch, Pediatric Gastroenterology, Hepatology and Nutrition , Nemours/Alfred I DuPont Hospital for Children , Wilmington, Delaware, UNITED STATES.

**Introduction:** A rare complication of sickle cell disease (SCD) is acute colonic pseudo-obstruction, also known as Ogilvie's syndrome. Vasoocclusive crisis can involve any organ including gastrointestinal (GI) tract where ischemia of the mesentery and abdominal viscera can lead to multiple complications. Ogilvie's syndrome is not only a rare entity but there are very limited treatment options available. Apart from supportive treatment for the underlying sick cell crisis, there is only one published pediatric case of Ogilvie's syndrome treated successfully with

Neostigmine. Certain prokinetic agents like Erythromycin have been used in adults with good outcomes. Here we report a case of Ogilvie's syndrome where multiple prokinetic medications were utilized with excellent results. To our knowledge this is the first pediatric case reported using these novel therapeutic options successfully. Case: 5 year old female with SCD (SS type) and history of splenic sequestration who presented to our ED with 1 day history of abdominal pain, massive abdominal distention and concerns of acute sickle cell crisis. Otherwise no fever, nausea, vomiting and normal bowel movements. Laboratory tests were notable for anemia requiring transfusion. Obstruction series and CT abdomen/pelvis was remarkable for significantly dilated bowel loops. She was started on a bowel regimen, but had acute worsening of her symptoms. Subsequently we started bowel decompression using rectal irrigations with red rubber tube along with addition of Erythromycin for small bowel prokinesis. Her symptoms resolved completely within a few days and she was discharged home on Erythromycin. 6 months later she was readmitted with similar massive abdominal distention and pain. Her imaging re-demonstrated findings of severe abdominal adynamic ileus. She developed concurrent acute chest syndrome, which was treated with Augmentin and Azithromycin in addition to supportive therapy. Bowel decompression with rectal irrigations were implemented simultaneously. As a result, her clinical course improved rapidly and GI symptoms were completely resolved prior to discharge. Discussion: This clinical vignette highlight a rare but important complication of SCD that requires prompt identification and treatment. There have been reported cases of necrotizing colitis, pseudomembranous colitis, ischemic colitis and necrotic perforation in adult patients with SCD. More importantly, this is the first reported case of Ogilvie's syndrome in pediatrics successfully treated with use of prokinetic medications like Erythromycin, Azithromycin and Augmentin. Even though all these agent work exclusively at the level of stomach and small bowel, we do believe patients with colonic pseudo-obstruction from Ogilvie's syndrome may benefit from these antibiotics as an adjunct to supportive management of underlying sickle crisis. This can be due to the fact that this entity encompasses a generalized dysmotility of the GI tract. Most prokinetic agents have excellent safety profile and in our experience are required only for short-term. Their use can significantly reduce the morbidity associated with this Ogilvie's syndrome and should be considered early in the course of disease.

**571 ACTG2 DELETION MUTATION RELATED VISCERAL MYOPATHY WITH ORAL PYRIDOSTIGMINE TREATMENT.** S.

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Background: Chronic intestinal pseudo-obstruction (CIPO) is diagnosed upon clinical symptoms and signs of bowel obstruction without lumen-occluding lesion. The etiology of CIPO varies from primary causes such as visceral myopathy and neuropathy to secondary causes such as toxins, infections, diabetes, connective tissue diseases, and so on. Diagnostic modality to identify the cause of CIPO has evolved, and there are reports about genetic mutations in CIPO patients using whole-exome sequencing (WES). We acknowledged one female patient with small bowel CIPO who was diagnosed with visceral myopathy with *ACTG2* mutation who successfully recovered her symptom by adding oral pyridostigmine. Case: 11-year-old girl was admitted to our clinic for abdominal pain and vomiting for three days. She was diagnosed with CIPO prior to the admission for repetitive episodes of small bowel ileus without mechanical obstruction in wax and wane manner. Her first admission was at the age of 10, when she first complained about abdominal bloating and vomiting. She had no past history or family history of gastrointestinal motility disorder. On the impression of small bowel obstruction, abdominal pelvic computed tomography (APCT) was performed which was suspicious of bezoar obstruction at ileocecal valve area. After surgical removal of the bezoar, which was considered to be a fecal material, she was treated for constipation and was discharged home. However, her ileus persisted at the outpatient clinic, and she was repeatedly readmitted to our department for CIPO. Many evaluations were done to find the cause of her intestinal obstruction such as infectious, endocrinologic, immunologic causes. Imaging studies did not reveal any focal structuring or obstructing lesion or a passage delay, and gastroduodenoscopy and colonoscopy were performed which all showed normal results. Bowel decompression and low residue diet was tried, and several medications such as probiotics, antibiotics, and prokinetics were applied. Steroid therapy was tried since there were borderline eosinophilia (26/HPF) noted in duodenal biopsy. None of the medications seemed to have effect to relieve her symptoms, and exploratory laparotomy and ileostomy were done for decompression which also did not seem to alleviate her symptom. Intravenous home parenteral nutritional (PN) treatment was started due to severe malnutrition. Whole exome sequencing was done at this time which showed positive result for de novo *ACTG2* mutation with deletion on exon 8~9, confirming visceral myopathy. Although she had ileostomy, her intestinal obstruction worsened gradually and she was readmitted to our clinic. Intravenous neostigmine infusion

was done to maximum dosage of 2.5 mg per day at the rate of 0.5 mg per hour, and the ileus has been improved. We have change the intravenous neostigmine into oral pyridostigmine at total of 180 mg per day. She is off from all other medications and successfully maintained to be free from home PN or admission. Conclusion: WES can be considered for diagnosis of CIPO, and oral anticholinesterase inhibitors may be effective treatment options in refractory small bowel CIPO.

**572 ATYPICAL HIRSCHSPRUNG'S DISEASE: RARE CASE OF SINGLE ZONAL AGANGLIONOSIS WITH POSITIVE RAIR AND GANGLION CELLS PRESENT ON RECTAL BIOSPY.** S. Nanton, Pediatric Gastroenterology, Avera McKennan Hospital and University Health Center, Sioux Falls, South Dakota, UNITED STATES. Bufo, C. Stoos, J. Roskens, S. Feit, B. Davis, Avera McKennan Hospital and University Health Center, Sioux Falls, South Dakota, UNITED STATES. Hirschsprung's disease is usually characterized by congenital aganglionosis beginning in the distal rectum and extending proximally. In the majority of patients with congenital intestinal aganglionosis, the aganglionic segment is restricted to the rectosigmoid colon. Rectal biopsy and anorectal manometry are considered useful diagnostic tools with the diagnosis typically being confirmed by absence of ganglion cells in the distal rectum. Contrast enema typically demonstrates the presence of a transition zone between a dilated proximal bowel and constricted distal colon. We describe an unusual case of zonal aganglionosis in which the aganglionic zone was limited to the sigmoid colon with normally ganglionic rectum and descending colon. A full term male infant presented with bilious emesis, abdominal distension and delayed passage of meconium at 48 hours old. Contrast enema demonstrated transition zone at the junction of the distal descending and sigmoid colon (Figure 1B). Abdominal CT scan confirmed the presence of a narrowed sigmoid colon with no evidence of mass, extrinsic compression or vascular compromise (Figure 1C). Anorectal manometry confirmed the presence of the recto-anal inhibitory reflex (RAIR) (Figure 1A). Suction rectal biopsy returned positive for rare ganglion cells (Figure 2A). Calretinin stain was positive (Figure 2B). The patient continued to be symptomatic and full thickness rectal biopsy again confirmed the presence of ganglion cells (Figure 2C & 2D). The patient subsequently underwent resection of the transition zone at the level of the distal descending colon and colostomy placement. This confirmed the presence of normal ganglionic descending colon and aganglionic sigmoid transition zone. A diagnosis of zonal aganglionosis was made based on these findings. The patient's abdominal distension resolved and he demonstrated normal growth with good stool output from the ostomy site. The infant is scheduled for pull through surgery. Zonal aganglionosis is characterized by a segment of aganglionic bowel preceded and followed by bowel with normal innervation. In our patient, the aganglionic sigmoid colon was preceded by ganglionic descending colon and followed by ganglionic rectum with RAIR positive. Single zonal aganglionosis differs from skip segment Hirschsprung's disease. In skip segment Hirschsprung's disease, the rectum is always aganglionic, however our patient showed ganglion cells in the rectum. Our case highlights the fact that in rare cases, the presence of ganglion cells in the rectum does not necessarily preclude a diagnosis of Hirschsprung's disease.

**576 A RARE ASSOCIATION OF TURCOT SYNDROME AND FISTULIZING CROHN'S DISEASE IN ONE PATIENT: A NOVEL CASE.** M.A. Corbera-Hincapie, G. Beasley, Pediatric Gastroenterology, University of Florida, Gainesville, Florida, UNITED STATES.

**Introduction** This case report illustrates the rare association of Turcot Syndrome and fistulizing Crohn's Disease in the same patient. There are challenges surrounding diagnosis and management of patients, who have this rare combination of disease processes. Turcot Syndrome Type II is autosomal dominant and has an association with Familial adenomatous polyposis (FAP), which carries a strong predisposition of polyps that develop into colorectal adenocarcinoma, with the most common central nervous system tumor being medulloblastoma. Crohn's Disease (CD) is a chronic inflammatory disease of the gastrointestinal tract that is of unknown etiology and may affect any region from the mouth to the anus. Case A 13 year old female with a family history of FAP presented with a history of diarrhea, intermittent rectal bleeding, fevers, rectal pain, poor appetite and weight loss. Colonoscopy was consistent with CD. She carries a mutation on the APC gene, giving her a diagnosis of FAP. Almost a year after initial presentation, patient presented with facial asymmetry and headaches and was diagnosed with medulloblastoma. Genetic mutation consistent with FAP, along with development of medulloblastoma, was diagnostic of Turcot Syndrome. Patient was treated with Adalimumab for her CD and subsequently with radiation and chemotherapy for the medulloblastoma. Yearly surveillance scopes have been performed with the most recent one remarkable for low-grade dysplasia in colon. **Discussion** In a patient with Turcot Syndrome and fistulizing Crohn's Disease, the main points of discussion revolve around the role of immunosuppressive therapy in

acceleration of dysplasia along with surgical management. There has been growing evidence to implicate factors, such as immunosuppressive therapy, previous radiation exposure, chemotherapy and genetic predisposition, in rapid progression of advanced adenocarcinomas. Our patient carries the genetic mutation predisposing her to cancer and underwent radiation treatment with chemotherapy, along with years of immunosuppression with biologic therapy, placing her at high risk for accelerated progression of secondary malignancies. This raises the question of surveillance frequency in these patients. From a surgical management standpoint, restorative proctocolectomy and ileal pouch-anal anastomosis have been the procedures of choice for patients with FAP. However, in patients with fistulizing CD, an ileo-anal J-pouch has been considered to be contraindicated by some due to its high complication and failure rates. Patients with Crohn's Disease have an increased rate of anastomotic strictures, pouch failure and pouch inflammation. Patients should be aware that creation of a diverting ileostomy versus restorative procedure may be a better option for controlling symptoms and improving lifestyle in patients, who are high risk for pouch complications or failure. Conclusion There is only one other reported case in the literature of a patient carrying diagnosis of both Crohn's disease and Turcot Syndrome. This case highlights the need to balance the risks and benefits of treating multiple complex diseases and the need to further identify optimal management.

**587 PENILE AND SCROTAL METASTATIC CROHN'S DISEASE IN PEDIATRIC MALE PATIENT.** S. Hassan, B. Gurram, Pediatric Gastroenterology, University of Texas Southwestern, Dallas, Texas, UNITED STATES. Hassan, B. Cartwright, B. Schlomer, B. Gurram, Children's Health, Dallas, Texas, UNITED STATES. Wu, B. Schlomer, Pediatric Urology, University of Texas Southwestern, Dallas, Texas, UNITED STATES.

Background Metastatic Crohn's disease (MCD) is a rare and under-recognized cutaneous manifestation of Crohn's disease in pediatric population, and it is described as noncaseating cutaneous granulomatous lesions not contiguous with intestinal Crohn's disease. MCD has been reported to involve any part of the body including scalp, face, trunk, extremities, lung and genital areas. Of these, genital MCD is the most frequently reported. A majority of these patients had MCD as their presenting symptom before the diagnosis of Crohn's disease. From the past literature in children, only 16 boys presented with penile or scrotal swelling as their initial manifestation of Crohn's disease. We describe a teenage boy who presented with penile and scrotal swelling as the initial manifestation of Crohn's disease. Case A 15-year-old African American boy presented to his pediatrician with non-tender, non-erythematous swelling of the penile shaft skin and upper scrotum with associated dryness and pruritus for 1 month. No associated dysuria or fever. He did not report any gastrointestinal symptoms at the time of presentation. He was treated as contact dermatitis by urology with diphenhydramine and 1% hydrocortisone topical. In view of non-response to above medications, a penile and scrotal skin biopsies were obtained, which revealed granulomatous inflammation involving scrotum and penis. PAS and AFB stains were negative for microorganisms. This prompted a referral to gastroenterology. Further questioning in the GI clinic revealed, poor weight gain for over a year, intermittent abdominal pain, poor energy levels for 2-3 weeks and recurrent oral ulcers for 6 months. Upon exam he was malnourished (BMI Z score -3.89), had circumferential edema involving penile shaft and edema of scrotum without erythema or induration. He also had 2 skin tags and an anal fissure in the 12'0 clock position. Blood work was significant for iron deficiency anemia, elevated CRP and hypoalbuminemia. Esophagogastroduodenoscopy and colonoscopy showed scattered ulcerations in the distal esophagus, stomach, duodenal bulb and terminal ileum. He had normal appearing colon and 3<sup>rd</sup> portion of the duodenum. Biopsies revealed chronic granulomatous inflammation involving the stomach, duodenum, terminal ileum and colon. He was diagnosed with Crohn's disease and was started on Infliximab. Conclusion Metastatic Crohn's disease can precede the gastrointestinal manifestations of Crohn's disease. A high degree of suspicion is needed for presence of Crohn's disease in patients presenting with cutaneous manifestations especially involving genitalia.

**589 CONTRAST ENHANCED ULTRASOUND: A NEW, SAFE, BEDSIDE METHOD OF CHARACTERIZING ILEAL STRICTURES IN CROHN'S DISEASE.** S.D. Sidhu, C. Cuffari, Pediatric Gastroenterology, Hepatology and Nutrition, Johns Hopkins University Hospital, Baltimore, Maryland, UNITED STATES. Hwang, Pediatric Radiology, Johns Hopkins University Hospital, Baltimore, Maryland, UNITED STATES.

Background: In patients with Crohn's disease (CD) complicated by ileal stricture, both acute inflammation and chronic fibrosis contribute to luminal narrowing and obstruction. New contrast enhanced ultrasound (CEUS) technology can be used to characterize the degree of inflammation and fibrosis involved in ileal strictures. The ability to differentiate between reversible inflammation that responds to medical therapy and a predominantly

fibrotic stricture that requires surgical resection holds important clinical implications. CEUS is a non-invasive, inexpensive, radiation-free and fast modality that provides a functional assessment of ileal strictures. Methods: CEUS was performed on two pediatric patients with CD complicated by ileal stricture. Contrast enhancement kinetics of the distal ileum were assessed, including wash-in slope, peak intensity, time to peak intensity and area under the curve. These quantifiable kinetics reflect the dynamic pattern of blood perfusion in the examined tissue. The same technique was also applied to healthy jejunal bowel, thus allowing each patient to act as their own internal control. Comparative Case Series: Case 1: 17-year-old female with CD with ileal stricture requiring ileocectomy, re-presenting with abdominal pain and distension consistent with obstruction. CEUS of the distal ileum revealed thickened submucosa, decreased peristalsis as well as lower wash-in slope, time to peak, peak intensity and area under the curve as compared to jejunal kinetics. These findings favor fibrotic rather than inflammatory obstruction and correlate to the colonoscopic finding of a tight ileocolonic anastomosis prohibiting further intubation. Case 2: 16-year-old female with CD presenting with abdominal pain and distension consistent with obstruction. CEUS of the distal ileum revealed narrowed lumen, thickened submucosa, and decreased peristalsis as well as increased wash-in slope, time to peak, peak intensity and area under the curve as compared to jejunal kinetics. In comparison to the patient in Case 1, she exhibited higher wash-in slope, time to peak, peak intensity and area under the curve. These findings are consistent with active inflammation rather than fibrosis resulting in stricturing of the terminal ileum and correlate to MRE with significantly inflamed distal ileum, surrounding mesenteric inflammation and ascites. Summary: CEUS is a non-invasive, inexpensive, radiation-free, and fast mode of imaging that provides a functional assessment of ileal strictures. Further prospective exploration of this new technology is needed to better define the role of CEUS in CD and other gastrointestinal diseases.

**590 PERFORATED MECKEL'S DIVERTICULITIS MIMICKING IBD.** S. Talathi, J. Mestre, Pediatric Gastroenterology, UAB, Birmingham, Alabama, UNITED STATES. D. Kelly, Pathology, UAB, Birmingham, Alabama, UNITED STATES. S. Anderson, Surgery, UAB, Birmingham, Alabama, UNITED STATES. Y. Vaid, Radiology, UAB, Birmingham, Alabama, UNITED STATES.

Meckel's diverticulum (MD) is a common congenital anomaly of the gastrointestinal tract. Symptoms of MD can be non-specific leading to diagnostic dilemma. We present a case of complicated MD with findings suggestive of IBD. PRESENTATION: A 13 year old male was admitted with acute hypogastric pain, and fever. He had guarding on physical exam but no signs of peritonitis. Laboratory work showed leukocytosis and CRP (27). A computed tomography of abdomen reported terminal ileal (TI) thickening. His pain, fever and leukocytosis worsened, blood cultures were drawn and intravenous fluids and antibiotics were given. Appendix ultrasound was normal and magnetic resonance enterography (MRE) reported cecitis. Colonoscopy with intubation of TI was normal. Post colonoscopy, patient developed perforation. Exploratory laparotomy revealed a perforated MD (Fig). He underwent small bowel resection with primary anastomosis. DISCUSSION: MD is a common congenital anomaly of the GI tract and the most common cause of GI bleeding in pediatrics. It generally said to follow the rule of 2's, however, can present with non-specific symptoms making diagnosis of MD challenging. Only few cases of MD develop complications, like intussusception, obstruction, diverticulitis and perforation. MD can be diagnosed by a Meckel's scan or a tagged RBC scan. Diagnosing a complicated MD as in our patient, is more challenging as it can mimic several other conditions. In our patient, acute appendicitis was ruled out by imaging, however, he had TI thickening. Meckel's diverticulitis has been misdiagnosed IBD in previous cases. TI is commonly affected in Crohn's disease (CD). Misdiagnosing CD can prove harmful and if suspected, intubation of the TI during colonoscopy is the standard of care. Our patient's upper endoscopy and colonoscopy were normal. TI thickening has been found in many other conditions too. Our patient did not have evidence of rectal bleeding, hypoalbuminemia, weight loss. Looking at the images retrospectively, CT scan did not show any dilation of the bowel loops, or any air fluid levels whereas the MRE did. This suggests that MD had perforated by the time an MRE was performed. CONCLUSION: A high index of suspicion towards a possibility of a complicated MD should be entertained in patients presenting with an acute abdomen, once other causes of acute abdomen are ruled out.

**591 EPIDURAL ABSCESS AND SPINAL OSTEOMYELITIS IN A PATIENT WITH CROHN'S DISEASE.** S. Zavoian, R.D. Baker, S. Choudhury, Pediatric Gastroenterology, University at Buffalo, Buffalo, New York, UNITED STATES. R.H. Alkhouri, Pediatric Gastroenterology, Presbyterian Healthcare Services, Albuquerque, New Mexico, UNITED STATES.

Crohn's disease (CD) is a chronic inflammatory condition of the gastrointestinal tract. It involves the full thickness of the gut and it is known to be complicated with fistulizing disease and intra-abdominal abscess (IAA) in 10-30% of cases. Initial management of IAA includes antibiotics and percutaneous drainage. We present a patient who has failed medical therapy and developed the rare complications of epidural abscess and osteomyelitis. 17 yo male recently diagnosed with ileal Crohn's disease, complained of numbness in the right foot while running, after receiving the first infliximab induction dose. He had mild abdominal tenderness and he was afebrile. Labs showed normal CRP and correcting anemia after iron infusion. An MR enterography (MRE) showed thickening of the terminal ileum with phlegmon formation and possible fistula. He was treated with ciprofloxacin and metronidazole and infliximab has been continued. He showed clinical improvement. Follow up MRE showed persistent but stable phlegmon at 1 month and resolution at 6 months with possible ileal stricture. Three months later he presented with increasing back pain in the right sacroiliac area radiating down the thigh. He also reported hematuria and low grade fever and had significant weight loss. There was right sacroiliac tenderness to palpation while he was afebrile. Urinalysis showed yellow clear urine and was within normal limits. CRP was high at 116, ESR 90, platelets were normal 445. Leukocytosis at 11.8 has been noted with increased band formation to 18% and 66% neutrophils. Hemoglobin was 11.5 and hematocrit 34.2. His PCDAI was calculated at 22.5. An abdomen and pelvis contrast enhanced CT scan showed no evidence of abscess or phlegmon recurrence, however it revealed an ileo-sigmoid fistula and sigmoiditis. Due to persistent symptoms and high clinical suspicion of osteomyelitis, an MRI was obtained and showed presacral phlegmon extending into the epidural space of the sacral spinal canal, and osteomyelitis of the S2 vertebral body. He responded clinically to antibiotic therapy with vancomycin and ampicillin followed by ciprofloxacin and metronidazole. Infliximab was held. Percutaneous drainage of the pre-vertebral collection and subsequent laparoscopic partial resection of the ileum were done. Eventually symptoms resolved, a follow-up MRI showed resolved epidural abscess. The infliximab therapy has been resumed. Surgical intervention will be required for 60-70% of CD patients with IAA, however the timing and indication for surgery are not clearly defined. We present a rare complication suggesting that earlier surgical intervention should be considered. Despite abscess resolution with medical therapy, recurrence and other complications can still occur. A high clinical suspicion has to be maintained for rare complications including neurological complications. Clinical and laboratory findings can precede diagnostic imaging findings.

**593 SEVERE COLITIS FOLLOWING RITUXIMAB THERAPY IN A PEDIATRIC MULTIVISCERAL TRANSPLANT RECIPIENT.**

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Introduction: Cases of severe ulcerative colitis have been reported following rituximab therapy in non-transplant patients. We herein reported the first case of severe colitis with recto-labial fistula in a pediatric transplant recipient after rituximab therapy. Case report: Patient is a 9-year-old female who underwent a liver, small bowel, pancreas transplant at 1 year of age due to short bowel syndrome secondary to gastroschisis. Five years after transplant, she developed post-transplant lymphoproliferative disease (PTLD) and received multiple courses of rituximab and chemotherapy. She then developed abdominal pain, diarrhea and painful defecation. Endoscopy was performed which showed deep linear ulcers in the rectum, deep punctate broad based ulcers in her native colon and ileo-colonic anastomotic ulcers. Pathology revealed severe colitis and ulcerations with negative special stains and cultures. Further rituximab was withheld and she was started on prednisone. Repeat endoscopy one month later showed partial resolution of colonic ulcers. She subsequently developed recto-labial fistula with persistent fecal drainage. She was started on infliximab, but then underwent diverting colostomy due to poor quality of life resulting from multiple admissions for labial cellulitis and/or abscesses. Discussion: Since rituximab is a B-cell depletion therapy, it might lead to immune dysregulation with T-regulatory cell dysfunction and subsequent development or exacerbation of inflammatory bowel disease. We suggest that any patients receiving rituximab therapy who develop lower gastrointestinal symptoms should be investigated for colitis, especially in immunosuppressed patients.

**595 LYMPHADENOPATHY SCARE IN A 10 YEAR OLD FEMALE ON ADALIMUMAB.** S.L. Page, Gastroenterology, Children's Mercy Kansas City, Kansas City, Missouri, UNITED STATES.

A 10 year old girl initially believed to have Eosinophilic Esophagitis in 2012 presented in 2014 with colitis. She was diagnosed with Crohn's disease, with granulomas throughout stomach, terminal ileum and colon, and treated with prednisone, balsalazide and azathioprine. She quickly transitioned to infliximab but developed antibodies and was switched to biweekly adalimumab September 2015. Ultimately, she received weekly adalimumab beginning April 2016 after suffering a flare and having a negative trough level. Methotrexate caused worsening abdominal pain. Repeat EGD/colonoscopy June 2016 revealed severe eosinophilic gastritis and duodenitis with eosinophilia of her terminal ileum. Epithelioid granulomas remained in her colon. She responded well to cromolyn and budesonide. In January 2017, she developed bilateral cervical lymphadenopathy, fevers, sore throat, and ear infections. She responded to Augmentin with decreased lymphadenopathy and fever curve. However, symptoms recurred with discontinuation of antibiotics. She received a course of amoxicillin with similar results. Workup revealed normal hemoglobin, ESR, albumin, uric acid, LDH, and calprotectin. Testing was negative for influenza, strep pharyngitis, CMV, EBV, adenovirus, Histoplasma, and coccidioides. Chest xray was negative. A CT scan of her neck revealed bilateral cervical lymphadenopathy without abscess. She was admitted to the hospital with impressive tender, painful, and large cervical lymph nodes. ENT, infectious disease, and rheumatology were involved. A biopsy of her posterior cervical lymph node showed reactive lymphoid hyperplasia, without evidence for malignancy. EGD revealed esophageal eosinophilia. Further workup revealed negative HIV, RPR, Bartonella, Mycoplasma, and Blastomyces. She had a normal C3, C4, ANA, ttg, ACE, and lysozyme. Lymph node was negative for AFB, anaerobic, aerobic, and fungal organisms. She was discharged home without an etiology found for her lymphadenopathy. Fevers and lymphadenopathy persisted. She developed difficulty swallowing. She was readmitted to the hospital and immunology and oncology were consulted in addition to reconsultation by ENT. A bone marrow biopsy was obtained which revealed normal cellular marrow with trilineage hematopoiesis and no evidence of malignancy. ENT removed her tonsils and adenoids. Tonsils were 4+ in size, clearly purulent and filled with debris. Her adenoids were 3+ and clearly infected as well. Microbiology on the tonsils grew nontypeable Haemophilus influenzae. She was treated with ampicillin-sulbactam and then Augmentin at discharge. She subsequently experienced resolution of her tender cervical lymphadenopathy and fevers. Unfortunately, because we held her adalimumab, her Crohn's disease flared.

**598 BILATERAL SUBMASSIVE PULMONARY EMBOLISM IN A PATIENT WITH ULCERATIVE COLITIS.** A.M. Gupta, B.P. Tullius, D. Say, S. Kaur, Pediatric Gastroenterology, UC Davis Medical Center, Sacramento, California, UNITED STATES. Pawar, Pediatric Hematology, UC Davis Medical Center, Sacramento, California, UNITED STATES.

**Introduction:**Inflammatory bowel disease such as ulcerative colitis (UC) is known to be a prothrombotic condition. Treatment options, such as steroids, can also increase the risk for thrombus. Currently there are no consensus guidelines on when IBD patients should be started on anti-coagulation therapy. **Case Description:**A 17 year old male with UC presented to an OSH ED with worsening abdominal pain and bloody diarrhea, while being treated for a UC flare with corticosteroids started one week prior to presentation. He was also taking his 5ASA for maintenance therapy and 6-MP was started 4 weeks prior to presentation. Pediatric Ulcerative Colitis Index (PUCAI) was 50 on presentation. Further history revealed that he also had headaches and right-sided thoracic pain for approximately 2 weeks prior to presentation. On presentation to OSH ED, the patient was noted to be tachycardic and mildly dyspneic. Blood work was concerning for leukocytosis to WBC of 16K with neutrophilic predominance and a lipase of 4114 U/L. A CT abdomen/pelvis showed evidence of UC flare and pancreatitis, with an incidental right lower lobe pulmonary (RLL) opacity noted in the limited lung field. This opacity was initially read as consolidation concerning for pneumonia and he was started on levaquin and transferred to our facility. On admission to UC Davis the patient's abdominal exam was consistent with UC flare but he was also noted to be tachycardic and a third heart sound was noted on auscultation. In the setting of recent development of respiratory symptoms and a RLL opacity noted on CT, suspicion was raised for pulmonary embolus. Stat CT chest revealed bilateral submassive pulmonary emboli, and the patient was started on a heparin drip. He remained hemodynamically stable throughout his admission and eventually was transitioned to Dabigatran for PO anticoagulation therapy. His UC symptoms improved on steroids, and 6-MP was discontinued given his pancreatitis. He was discharged home on a steroid taper and later started on biologic therapy.

**Discussion:**Adolescent patients with inflammatory bowel disease are a unique population who have multiple prothrombotic risks. The reason for PE in this patient is likely multifactorial: steroids, pancreatitis from 6-MP and active IBD. Pediatric gastroenterologists must have high index of suspicion for PE in patients with IBD on various treatment options, especially with any complaints of respiratory or cardiac symptoms. Our case highlights that

clinical trials and consensus guidelines on thrombotic prophylaxis in IBD patients will be helpful in prevention of life threatening complications from thrombotic events such as PE.

**605** *INFANTILE-ONSET INFLAMMATORY BOWEL DISEASE AND LYMPHOMA AS A PRESENTATION OF IL-10R DEFICIENCY.* T. Patel, M.A. Conrad, J.E. McDermott, J. Kelsen, Pediatric Gastroenterology, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATESK. Sullivan, Immunology, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATESN. Dawany, Bioinformatics, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATESL. Devoto, Pediatrics, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATESL. Duffy, Pediatric Gastroenterology, Inova Pediatric Specialty Center, Fairfax, Virginia, UNITED STATES.

**BACKGROUND:** Patients who present with inflammatory bowel disease (IBD) in the first 2 years of life (and often in the few few months of life), known as neonatal-onset IBD, often have a severe disease course that is unresponsive to conventional therapy. The role of genomics is more pronounced in this age group and identification of monogenic defects can radically change the therapeutic approach. We present a case of a 13-year-old girl with neonatal-onset, fistulizing, severe IBD and diffuse large B cell lymphoma (DLBCL) who was found to have a homozygous IL-10RA mutation.**CASE PRESENTATION:** A newborn presented with bloody diarrhea and fever on day of life 1. Over the next 2 years, her disease course was complicated by severe bloody diarrhea and development of rectovaginal fistulae. She was diagnosed with duodenal and ileocolonic Crohn disease at that time and was treated with multiple medical therapies including steroids, 6-MP, Humira, Remicade and Methotrexate. She ultimately underwent a diverting ileostomy at 9 years of age and a mass was detected intraoperatively. She was subsequently diagnosed with diffuse large B cell lymphoma and underwent chemotherapy. At the time of presentation to our institution at age 13, her DLBCL was in remission, but her IBD remained active, including recurrent oral ulcers, intermittent abdominal pain and watery ostomy output without blood. In addition, she had severe growth failure. Infectious history was notable for 20-25 episodes of sinusitis, 1 pneumonia, 14 episodes of otitis media and 1 pelvic abscess. The combination of DLBCL and fistulizing IBD, known to be consistent with IL-10R defects, prompted genomic evaluation in our Very-Early-Onset IBD clinic. Indeed, we detected a homozygous mutation in IL10RA (Arg101Trp, which has previously been described by Mao et al). Subsequent IL-10R functional testing was abnormal. Immunophenotyping demonstrated absent iNKT cells and overall low NK cells. She will undergo hematopoietic stem cell transplantation as the definitive therapy.**DISCUSSION:** This case highlights the critical importance of identifying unique and defining clinical characteristics and co-morbid conditions in very young children with IBD. Absent iNKT cells have not previously been reported in IL10R defects, but have been reported in B cell lymphoma. This finding may serve as a marker for malignancy risk in IL-10R defects. However, further investigation is needed. The integration of the phenotypic, immunologic and genomic data can allow for identification of monogenic defects in these children and the potential for a targeted, life-saving therapeutic approach.

**607** *ACUTE HEART FAILURE AFTER INFLIXIMAB INFUSION IN A 4-YEAR-OLD WITH CROHN'S DISEASE.* V. Cardenas, J. Adler, G. Lee, Pediatrics and Communicable Diseases, University of Michigan, Ann Arbor, Michigan, UNITED STATES.

**Introduction:** Tumor necrosis factor  $\alpha$  antagonist (anti-TNF $\alpha$ ) therapy has revolutionized the care of pediatric patients with inflammatory bowel disease (IBD). Anti-TNF $\alpha$  therapy is generally well tolerated with infrequent severe adverse drug reactions. Anti-TNF $\alpha$  therapy studies for the treatment of adult heart failure identified increased mortality. Thus, it is contraindicated in patients with pre-existing heart failure. There are limited data on new-onset heart failure in pediatric IBD patients treated with anti-TNF $\alpha$  without pre-existing cardiovascular disease. We present a case of a pediatric patient with Crohn's disease who received infliximab in the setting of an acute disease exacerbation and developed acute severe heart failure.**Case Report:** A 4-year-old female diagnosed with Crohn's colitis in the past month and a half had completed 2 induction doses of infliximab 5 mg/kg without incident. She then developed worsening disease and was admitted to the hospital for management of severe dehydration, acute kidney injury, and electrolyte abnormalities. The initial 48 hours of hospitalization were focused on hydration and electrolyte management. She received a 3<sup>rd</sup> dose of infliximab 10 mg/kg due to severity of illness and low albumin (1.6 g/dl). She had some improvement in inflammatory markers (C-reactive protein of 11.6 to 3.5 mg/dl) but had minimal clinical improvement. She received a 4<sup>th</sup> dose of infliximab 10 mg/kg 72 hours after the 3<sup>rd</sup> dose. She responded well with improvement in stool

frequency, consistency, and hematochezia. She remained hospitalized due to complications including pre-renal acute kidney injury, parenteral nutrition supplementation, central line-associated thrombosis, and intermittent fevers, suspicious for infected thrombus. On hospital day 12, 5 days after the 4<sup>th</sup> infliximab dose, she became tachypneic, tachycardic, and developed a new S3 gallop. Echocardiogram revealed a severely depressed systolic function of both left and right ventricles (ejection fraction 30%). She required intensive care unit admission for medical management. She had a negative evaluation for infections, including herpes simplex virus, cytomegalovirus, Epstein-Barr virus, Influenza A and B, parvovirus, adenovirus, coxsackie virus, and a respiratory viral PCR panel. Her immune evaluation revealed appropriate response to prior vaccinations, normal lymphocyte distribution, negative anti-neutrophil antibody and rheumatoid factor, normal neutrophil oxidative burst, and immunoglobulin quantitation. Given the timing of infliximab administration, her new-onset heart failure was suspected to be secondary to infliximab toxicity. She had a prolonged hospitalization and was ultimately discharged on multiple cardiac medications for ongoing depressed cardiac function. Infliximab was discontinued. She continued azathioprine and oral budesonide for Crohn's disease management and has remained in clinical remission. Given the concern with long-term exposure to steroids and elevated fecal calprotectin, the addition of a non-anti-TNF $\alpha$  medication such as vedolizumab is being considered. Conclusion: Anti-TNF $\alpha$  therapy is a contraindication in patients with pre-existing heart failure. Very little is known about new-onset acute heart failure in patients on infliximab, particularly among pediatric patients without cardiovascular risk factors. This has rarely been reported as a drug reaction in adults, but there are no such reports of pediatric IBD patients. It is a severe adverse reaction that must be recognized. It also has a significant implication on the care of a young patient with Crohn's disease who may no longer be able to receive this class of medications. Clinicians should be aware that heart failure can be a rare but serious adverse reaction to anti-TNF $\alpha$  therapy, requiring prompt evaluation and treatment.

**608 INFLAMMATORY BOWEL DISEASE - A CASE SERIES FROM SOUTHERN BRAZIL.** V.A. Scheeffeffer, C.T. Ferreira, M. Epifanio, A.L. Estevam, A.F. Freitas, L. Nader, C.S. Silva, C. Steinhaus, M.U. Melere, M.R. Ceza, Gastroenterology, Hospital da Criança Santo Antônio, Canoas, Rio Grande do Sul, BRAZIL.

Objective: We present a case series of children with inflammatory bowel disease (IBD). Since there is few Brazilian studies on pediatric IBD, we aimed to describe the characteristic of this disease in our population. Methods: Review in electronic medical records of 41 children with IBD attended between January 2012 and May 2017 in a Brazilian tertiary referral hospital. Results: Twenty one (51%) patients were diagnosed as having Crohn's disease (CD), eighteen (43%) as Ulcerative Colitis (UC) and two (4%) as Undetermined Colitis. 21 patients (51%) were male. Mean age at diagnosis was 11.39 yo (6 mo to 17 yo). Six patients were diagnosed before being referred to our institution, four of them before 2012. The number of diagnosis per year is shown in figure 1. Regarding symptoms at onset, most patients presented with abdominal pain and diarrhea. The frequency of symptoms is shown in table 1. Among those who presented fistula, two had associated perianal pain. Among CD patients, 13 (61%) had disease in mouth or upper gastrointestinal (GI) tract, 11 (52%) in small bowel, 8 (38%) in right colon, 8 in transverse colon, 7 (33%) in left colon, 9 (42%) in sigmoid and 13 had anorectal disease. Among UC patients, 1 (5%) had disease in mouth or upper GI tract, 2 (11%) in small bowel, 12 (6%) in right colon, 13 (72%) in transverse colon, 14 (77%) in left colon, 15 (83%) in sigmoid and 15 had anorectal disease. Regarding treatment, 12 (29%) patients were put into monotherapy at diagnosis. All but two were UC and were in monotherapy with Mesalazine. One patient with CD was in monotherapy with Infliximab due to fistula as only symptom and the other was put into corticosteroids as monotherapy due to low age at onset. Patients with combined therapy followed the institutional protocol and Mesalazine plus corticosteroid was used in moderate to severe UC, Azathioprine plus corticosteroid was used in CD and Infliximab plus Azathioprine was prescribed in CD with fistula. Conclusion Our results show a slight increase in frequency of IBD through the years. This may reflect an increase in prevalence, as seen worldwide. The need of combined therapy in most patients may reflect a higher level of severity in this population. Larger studies are needed to confirm our hypothesis.

**609 BIOLOGICAL THERAPY IN CHILDREN WITH INFLAMMATORY BOWEL DISEASE - EXPERIENCE OF A TERTIARY CARE HOSPITAL IN SOUTHERN BRAZIL.** V.A. Scheeffeffer, C.T. Ferreira, M. Epifanio, A.L. Estevam, A.F. Freitas, L. Nader, C.S. Silva, C. Steinhaus, M.U. Melere, M.R. Ceza, Gastroenterology, Hospital da Criança Santo Antônio, Canoas, Rio Grande do Sul, BRAZIL.

**Introduction**The use of biological therapy (BT) in inflammatory bowel disease (IBD) has changed the outcomes of this entity. In Brazil, there is still some limitation regarding the use of this therapy. The prime reason is that this type of medication is still expensive and access is still limited. Besides that, there is still doubt regarding safety of use of BT among both physicians and patients. We aimed to describe the experience of a tertiary care hospital with BT in children attended between January 2012 and May 2017. The data was obtained after review of electronic medical profile. **Results**At the refereed period, 41 patients with IBD were attended at the institution. Sixteen (39%) patients were in use of BT, three of them (19%) had ulcerative colitis and thirteen (81%) had Crohn's disease. BT was prescribed due the following: fistula (25%), steroid-dependent disease (68%), refractory disease (7%). Infliximab was BT of choice for all patients and two (12%) patients had to change therapy to Adalimumab due to loss of response. Combined therapy with Azathioprine was initially prescribed for 15 (93%) patients and one patient took Infliximab as monotherapy from the beginning. Five patients (31%) are current on monotherapy with Infliximab and one is on monotherapy with Adalimumab. All patients are current asymptomatic. There have been no record of complication during infusion in our center. **Conclusion**In a country where access to BT is limited, the choice of therapy in IBD should still follow a step up approach. The use of BT in selected patients has demonstrated to be safe and has shown good outcomes, therefore there should be no reason to postpone the indication of BT in severe presentations of IBD.

**612 VULVAR CROHN'S DISEASE IN VERY-EARLY ONSET IBD (VEO-IBD).** Z. Khan, St. George's University, St. George's, GRENADAQ. Warsi, Epidemiology and Biostatistics, University of California, San Francisco, California, UNITED STATES. Karjoo, M.B. BEG, Pediatrics, SUNY Upstate Medical University, Golisano Children's Hospital, Syracuse, New York, UNITED STATES.

The incidence of pediatric Inflammatory Bowel Disease (IBD) is on the rise, particularly in children younger than 10-years-old. More recently, very-early-onset IBD (VEOIBD) has been defined as IBD that occurs in children less than 6 years of age. Croon's disease (CD) is a chronic inflammatory bowel disease that can affect any portion of the gastrointestinal tract. Extra-intestinal manifestations of CD may involve skin, eyes, joints and bones. Crohn's disease-related genital involvement is very rare in all age groups. We present a girl diagnosed with perianal and ileo-colonic VEOIBD at age 6 years, treated with infliximab with resolution of gastrointestinal symptoms. At age 10, she developed intermittent and refractory genital swelling. The patient was seen by a gynecologist for labial swelling and itching and was treated with maintenance therapy of good local hygiene, warm sitz baths and topical hydrocortisone without improvement. Magnetic resonance enterography demonstrated normal rectosigmoid colon, bowel wall circumferential thickening with no edema or rectovaginal fistula. The vulvar biopsy demonstrated non-necrotizing granulomas with multinucleated giant cells, confirming the diagnosis of vulvar CD. CD must be considered in the differential diagnosis of recurrent, non-tender, erythematous and edematous lesions of the genital area. We want to bring attention to gynecologists and primary care physicians that patients with refractory genital lesions, with known Crohn's disease, could have an extraintestinal clinical manifestation of cutaneous CD. Its early recognition, diagnosis and appropriate treatment can decrease morbidity. We present this case due to its rarity, complexity and unusual nature.

**614 DEEP CAVITARY SKIN LESIONS LEADING TO THE DIAGNOSIS OF IBD.** A. Kilgore, C. Menard-Katcher, E.F. de Zoeten, Digestive Health Institute, Section of Pediatric Gastroenterology, Hepatology and Nutrition, Children's Hospital Colorado, Aurora, Colorado, UNITED STATES. Kilgore, A. Bruckner, Department of Pediatrics, University of Colorado School of Medicine, Aurora, Colorado, UNITED STATES. C. Menard-Katcher, Gastrointestinal Eosinophilic Diseases Program, Department of Pediatrics, University of Colorado School of Medicine, Aurora, Colorado, UNITED STATES. K.E. Capocelli, Department of Pathology, Children's Hospital Colorado, Aurora, Colorado, UNITED STATES. K.E. Capocelli, L. Prok, Department of Pathology, University of Colorado School of Medicine, Aurora, Colorado, UNITED STATES. L. Prok, A. Bruckner, Department of Dermatology, University of Colorado School of Medicine, Aurora, Colorado, UNITED STATES. E.F. de Zoeten, Mucosal Inflammation Program, University of Colorado School of Medicine, Aurora, Colorado, UNITED STATES.

**BACKGROUND:** Dermatologic extraintestinal manifestations can lead to the initial diagnosis of inflammatory bowel disease (IBD). Below is a unique case of deep cavitory skin lesions as the presenting symptom of IBD and its response to treatment with enteral therapy. **CASE:** A 15 year old female presented with a 3 week history of progressively worsening skin lesions. The skin lesions began as papules on her forehead, which progressed to self-draining pustules and subsequent deep ulcerations on her head, neck and torso. She also complained of 3 weeks of

diarrhea, a 10 lb weight loss, and right hip pain for which she had been seeing a chiropractor. Her abdominal exam was significant for right lower quadrant tenderness. Lab studies revealed microcytic anemia, thrombocytosis, hypoalbuminemia, and an elevated CRP. MRI revealed cutaneous abscesses, subcutaneous soft tissue edema, and diffuse colitis. Drainage from the abscesses showed numerous neutrophils; however, her broad infectious work up was negative except for skin flora. A skin biopsy taken from the edge of a lesion revealed subtle chronic inflammation of the dermis and subcutis only. Immunologic evaluation was unrevealing including a normal neutrophil oxidative burst. GI work up included an EGD and colonoscopy revealing distal esophageal exudate, gastric mucosal atrophy as well as deep ulcerations of the sigmoid colon and cecum, severe pseudopolyps throughout the colon, and a normal terminal ileum. Biopsies identified active esophagitis, chronic active duodenitis, active ileitis, chronic active pancolitis, and apoptotic bodies in the glands of the colon and rectum. Immunohistochemistry of the colonic biopsy was negative for CD1a-staining histiocytes ruling out Langerhans cell histiocytosis. Her c-ANCA and p-ANCA levels were positive. The leading diagnosis was Crohns disease with deep cavitory skin lesions. Family declined traditional therapy and chose to initiate enteral therapy and colonically-released budesonide. Her skin lesions were cleansed and dressed regularly. After 2 months, her family chose to continue enteral monotherapy. On follow up, she denied abdominal pains, diarrhea and blood in her stools. Her skin lesions greatly improved. **DISCUSSION:** This patient's skin findings were believed to be unique extraintestinal manifestations of IBD. Investigation into possible infectious, oncologic and immunologic causes for her dermatologic findings were negative. Her cavitory lesions healed with wound care and enteral therapy, showing its use in the treatment of extraintestinal manifestations of IBD.

**616 THE NECESSITY OF EARLY BONE MARROW TRANSPLANTATION FOR IL-21 RECEPTOR DEFICIENCY PATIENTS TO PREVENT COMPLICATIONS FROM LIVER CIRRHOSIS.** A.C. Huang, E. Gonzalez, Pediatrics, Nicklaus Children's Hospital, Miami, Florida, UNITED STATES. Hernandez, Pediatric Gastroenterology, Nicklaus Children's Hospital, Miami, Florida, UNITED STATES.

IL-21 receptor (IL-21R) deficiency is a rare immunodeficiency disorder. IL-21 is a cytokine that regulates the cell cycle and cytotoxicity. IL-21R deficiency produces abnormalities in B, NK and T cells, manifesting as frequent lung infections, gastrointestinal involvement and sclerosing cholangitis. We present two patients with IL-21R deficiency who died from complications of their worsening ascites and liver cirrhosis. The disease course and outcome of our patients indicates the necessity of early bone marrow transplantation for IL-21R deficiency patients. Patient 1 is a 13-year-old Colombian male. Patient 2 is the 8-year-old brother of Patient 1. There is consanguinity in the family. Both patients presented with recurrent otitis media, recurrent pneumonia, chronic *Cryptosporidium* hepatobiliary infection, and jaundice. Both patients had elevation of transaminases and direct hyperbilirubinemia. Patient 1 had a magnetic resonance cholangiopancreatography (MRCP) that showed intrahepatic and extrahepatic biliary ductal dilatation with mild beading, periportal edema, hepatosplenomegaly, and gallbladder polyps. Patient 1 had poor response to vaccinations, decreased T cell proliferation response and low IGG, requiring IVIG infusions. Patient 2's MRCP and magnetic resonance imaging (MRI) liver showed mild diffuse enlargement of the liver and pancreas, dilatation of the common bile duct and hepatic ducts, and ascites. Patient 2's liver biopsy showed sparse portal chronic inflammatory infiltrates and diffuse macrovesicular steatosis. Patient 2 also had poor response to vaccinations, low CD8, and low IGG, requiring IVIG infusions. His flow cytometry showed the loss of IL-21 mediated signaling and a homozygous deletion of exon 4 of the IL-21R gene. Both patients were diagnosed with IL-21R deficiency. Both siblings had worsening ascites and liver cirrhosis, recurrent cholangitis, increasingly aggressive infections, chronic lung disease and chronic diarrhea associated with malabsorption and malnutrition. Patient 1 had upper esophageal varices bleeding, acute respiratory failure, hypotension, and extended-spectrum beta-lactamases (ESBL) *Escherichia coli* infection. Patient 2 had pancytopenia, malnutrition, and bacteremia. Both patients died from complications of ascites and liver cirrhosis. Only 8 pediatric patients of IL-21R deficiency have been reported in the literature, including our two patients. Three other IL-21R/IL-21 deficiency patients also had gallbladder and liver involvement: idiopathic liver fibrosis, chronic cholangitis, and hepatic steatosis. One of the patients had a liver transplantation but died from post-transplant complications. Three of the patients received hematopoietic stem cell transplantation (HSCT). Two of the three patients died from post-HSCT complications. However, the third patient that had an early HSCT did well, demonstrating early HSCT is necessary. Based on our patients' progressive liver disease and eventual death, early bone marrow transplantation when the condition is identified may be essential in improving the outcome for IL-21R deficiency patients. More IL21-R deficiency

patients need to be identified to further our research on the appropriate interventions for this rare immunodeficiency.

**617 A CASE OF SCARLET FEVER ASSOCIATED WITH HEPATITIS.** A. Panchoo, Pediatrics, Jackson Memorial Hospital, Miami, Florida, UNITED STATES. Fifi, E.D. Rivera Rivera, Pediatrics - Gastroenterology, University of Miami, Miami, Florida, UNITED STATES.

Introduction:Scarlet Fever is a common pediatric illness caused due to a bacterial infection that happens in some patients with strep throat. This is a condition with an overall excellent prognosis. Hepatitis secondary to scarlet fever is a rare complication described in adults and even less frequently in children. Though the association between scarlet fever and hepatitis was first reported in 1931, the pathogenesis still remains largely unknown and there are very few pediatric cases in the literature highlighting the course, prognosis and eventual outcomes of these patients.Case Report:We report the case of a 12 year old male with scarlet fever presenting with a 4 day history of jaundice and dark urine. His illness began 9 days prior to presenting with fever, and the complaint was mostly sore throat and decreased appetite. Then, 3 days later, he developed an erythematous fine papular rash. He was taken to an urgent care center where a rapid Strep test was found to be positive for group A Streptococci. Subsequently the patient was diagnosed with Scarlet Fever and started on Amoxicillin. At presentation to our institution's Emergency Room, his vital signs were stable and normal for age, except for fever up to 39.2. Physical examination was significant for bilateral scleral icterus, an erythematous fine papular rash of the face and upper extremities but no hepatomegaly. Laboratory evaluation was significant for elevated antistreptolysin O titers, elevated liver enzymes and negative antibody tests for hepatitis viruses A, B and C.At that point the Pediatric Gastroenterology Service was consulted and further testing were sent. Some of these included DNA PCR for Epstein Barr Virus, adenovirus, cytomegalovirus, human herpes virus 6, herpes simplex virus 1 and herpes simplex virus 2. All of these were negative. Duplex ultrasound of the liver was unremarkable. The patient maintained a stable course through discharge and on outpatient follow up his liver enzymes continued to decrease with further resolution of icterus.Discussion:Scarlet fever is a common pediatric illness with generally very good prognosis and seldom rarely we see complications. However, hepatitis in association with scarlet fever has been reported before, but many healthcare providers, including general pediatricians are not always aware of this, therefore potentially leading to a more prolonged hospital stay and battery of test that increase the cost of healthcare. With this case we aim to raise awareness among pediatricians regarding possible hepatic involvement with scarlet fever and underline that most of these patients have a benign disease course with complete recovery after several weeks and months.

**619 DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS (DRESS) ASSOCIATED WITH ACALCULOUS CHOLECYSTITIS.** L.M. Alfaro Cruz, B. Chumpitazi, Department of Pediatrics, Baylor College of Medicine, Houston, Texas, UNITED STATES.L.M. Alfaro Cruz, B. Chumpitazi, Division of Pediatric Gastroenterology, Hepatology, and Nutrition, Texas Children's Hospital, Houston, Texas, UNITED STATES.

Clinical Vignette:A 7 y.o F with history of epilepsia partialis continua and epileptic encephalopathy developed high fevers (up to 104.3<sup>o</sup>F), a diffuse macular-papular rash, and had altered mental status. She was subsequently diagnosed with Drug reaction with Eosinophilia and Systemic Symptoms (DRESS) attributed to phenytoin. The gastroenterology service was consulted due to abnormal aminotransferases (AST/ALT: 398 U/L /382 U/L), elevated GGT (609 U/L) and conjugated hyperbilirubinemia (1.3 MG/DL). She had mild abdominal RUQ tenderness. An abdominal ultrasound identified an enlarged liver with normal echogenicity and contour, no focal mass, with periportal edema. It also identified diffuse gallbladder wall thickening (3.2 cms wall thickness) without stones, trace pericholecystic fluid, and normal common bile duct diameter.The patient was started on both steroids (for DRESS) and ciprofloxacin/flagyl (for acalculous cholecystitis). After six days of therapy her RUQ pain resolved and her conjugated bilirubin normalized to 0.0. She also had improvement on repeat ultrasound with decreased gallbladder wall thickening. However her aminotransferases (AST/ALT:1411 U/L, 527 U/L) and GGT (2543 U/L) continued to rise. Following a negative autoimmune and infectious workup a percutaneous liver biopsy was obtained on day 13 of admission; this showed diffuse monocytic inflammation without significant eosinophils or plasma cells. The patient completed a 14-day course of steroids and antibiotics and was discharged with improving aminotransferase and GGT levels.Discussion:DRESS is a drug immune response often usually associated with anticonvulsants (such as phenytoin) that can present between 2-6 weeks after the initiation of the medication. Clinical symptoms and signs may include fever, skin eruptions that affect more than 50% of body

surface area, leukocytosis with eosinophilia, elevation of aminotransferases, and in 80-90%, involvement of at least one organ. Organs commonly affected are the liver, kidneys and lungs; less commonly affected are the pancreas, brain, muscle, and thyroid. Liver involvement in DRESS may be accompanied by elevated aminotransferases, hepatomegaly, and jaundice. Liver injury in DRESS can vary being of a cholestatic type, hepatocellular injury type, or a mixed type (observed in our patient). DRESS liver involvement can be severe, at times progressing to acute liver failure and increased mortality. Steroids are of unproven benefit and there is no consensus on their usage though they are often used in severe cases. New in this vignette is the previously unreported association of DRESS and acalculous cholecystitis. Acalculous cholecystitis, particularly if not treated adequately, can lead to severe gallbladder complications such as gallbladder perforation or gangrene. As high mortality rates of at least 30%, in most cases secondary to its rapid progression to complications such as In the setting of a syndrome that triggers an immune response affecting different organs, it is unclear why the gallbladder is less often affected. Given the more rapid improvement in the gallbladder wall thickening and conjugated hyperbilirubinemia we hypothesize that the therapies (including antibiotics) helped to address the acalculous cholecystitis prior to resolution of the DRESS manifestations. Conclusions: DRESS can affect multiple organs and may be associated with acalculous cholecystitis. Clinical vigilance for this potential association is recommended.

**625 PRESENCE OF COLITIS IN PEDIATRIC YOUNG ADULT PATIENTS WITH PROGRESSIVE FAMILIAL INTRAHEPATIC CHOLESTASIS BEFORE AND AFTER LIVER TRANSPLANTATION.** N. Bhesania, C.S. Pasquarella, P. Conjeevaram Selvakumar, V.F. Hupertz, Pediatric Gastroenterology, Cleveland Clinic Children's Hospital, Cleveland, Ohio, UNITED STATES. plesec, Department of Pathology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES.

Progressive familial intrahepatic cholestasis (PFIC) type 1 is an autosomal recessive cholestatic disorder caused by mutation in FIC1 protein leading to impaired bile secretion that typically present in the neonatal period or first year of life subsequently resulting in intractable pruritus and progressive liver damage, requiring biliary diversion or even liver transplantation. Extrahepatic manifestations include short stature, failure to thrive, diarrhea from underlying enteropathy, and fat malabsorption. Although partial biliary diversion has been shown to alleviate pruritus and progression of liver fibrosis liver transplantation is required in some patients. The effect of liver transplantation (LT) on extrahepatic symptoms of PFIC 1 is variable. In particular, a subset of patients develop worsening diarrhea post-LT for unknown reason. It is believed that improvement in bile secretion post-LT results in a relative increase in bile acid secretion leading to diarrhea. Here in, we present a case series of 5 patients with PFIC 1 who developed intractable diarrhea post-LT who underwent colonoscopy. These patients were 10-23 years of age and mean age at LT was 9.5 years. All the patients were on Tacrolimus for immunosuppression and had negative infectious work up. The histological findings were variable and were not consistent with a particular diagnosis. Four subjects had evidence of focal active colitis with eosinophil predominant expansion of the lamina propria, either localized to one region or involving the entire colon. Multiple therapies were utilized including clonidine, pancreolipas, cholestyramine, loperamide, metronidazole, rifaximin, ciprofloxacin, lactobacillus, budesonide and mesalamine. Two patients improved with cholestyramine, 2 improved with clonidine, and 2 improved with mesalamine. One patient demonstrated no improvement but has only been trialed on pancreolipase at this time. Studies looking at the pathophysiology of diarrhea in patients with PFIC 1 post-LT are limited. A case series by Egawa et al revealed 6 children with intractable diarrhea following liver transplant for Byler's disease who improved with bile acid adsorptive. To our knowledge, this is the first case series to report inflammatory pathology in these patients with PFIC 1 who developed intractable diarrhea post-LT. We could hypothesize that a potential inflammatory component could contribute to intractable diarrhea post-LT in PFIC 1 patients. The underlying pathophysiology for this colitis is not clearly understood. Future studies are necessary to evaluate this association and to consider colonoscopy prior to transplant to determine if these findings are present prior to LT.

**626 SUPPRESSION OF BILE ACID SYNTHESIS WITH PROLONGED PARENTERAL NUTRITION IN A PRETERM INFANT WITH NECROTIZING ENTEROCOLITIS: A CASE REPORT.** N. Memon, N. Ovchinsky, Pediatrics, Goryeb Children's Hospital, Morristown, New Jersey, UNITED STATES. N. Memon, C. Lee, A. Herdt, MidAtlantic Neonatology Associates, Morristown, New Jersey, UNITED STATES. Weinberger, Cohen Children's Medical Center of NY, Hyde Park, New York, UNITED STATES. Hegyi, Rutgers Robert Wood Johnson Medical School, New Brunswick, New Jersey, UNITED STATES. Guo, Rutgers University, Piscataway, New Jersey, UNITED STATES.

Introduction: Maintaining bile acid (BA) and cholesterol homeostasis is important in protecting the liver from injury and parenteral nutrition (PN)-associated liver disease (PNALD). BA synthesis is mainly regulated by cholesterol 7 $\alpha$ -hydroxylase (CYP7A1), the rate-limiting enzyme in the classic pathway. Alterations in CYP7A1 expression in animal models of liver disease have been associated with hepatic inflammation, cholestasis, steatosis, oxidative stress, apoptosis, and fibrosis. Preterm birth is often complicated by infection, inflammation, and prolonged PN exposure (all of which are associated with suppression of BA synthesis). To date there are no published studies investigating CYP7A1 activity in high-risk preterm infants, a population particularly vulnerable to PNALD. Objective: To measure CYP7A1 activity in a preterm infant requiring prolonged parenteral nutrition (PN) and compare it to two healthy control infants. Methods: Plasma levels of 7 $\alpha$ -hydroxy-4-cholesten-3-one (C4, a validated biomarker for CYP7A1 activity) were quantified weekly in three preterm infants. The case study infant developed necrotizing enterocolitis (NEC) on DOL 10 and required prolonged PN (> 2 months). He developed biochemical evidence of cholestasis on DOL 28. The control infants had unremarkable NICU courses, requiring brief PN (< 10 days) and were easily transitioned to full enteral feeds. Phytosterol (PS) and FGF19 concentrations (known regulators of CYP7A1 activity) were also measured in these infants. C4 and PS concentrations were measured using HPLC-MS/MS. FGF19 concentrations were measured using ELISA. Results: CYP7A1 activity was upregulated in both control infants by 2 weeks of age; the case study infant had undetectable CYP7A1 activity out to 2 months of age. FGF19 concentrations remained low in all infants and ps concentrations in the case study were > 100x that of the control. Conclusion: This study describes a preterm infant on prolonged parenteral nutrition with undetectable levels of C4 and very high PS levels. We expect the suppression of CYP7A1 activity leads to a reduction in BA synthesis and bile flow; however, it is unclear whether these changes would decrease or increase cholestasis. Additionally, CYP7A1 participates in the degradation of cholesterol and PS, therefore high PS levels may be associated with the decrease in CYP7A1 activity. FGF19 remains low in all 3 infants, suggesting that it may not regulate CYP7A1 activity in the first few weeks of life. Additional studies are needed to characterize the relationship between prolonged PN and CYP7A1 activity, as well as their role in liver disease in the premature infant.

**632 UNDIAGNOSED BILE ACID SYNTHESIS DISORDER (BASD): IDENTICAL 7-MONTH-OLD TWINS PRESENTING WITH HYPOCALCEMIC RICKETS AND FAILURE TO THRIVE.** O. AHMAD, J. Nogueira, Pediatric Gastroenterology, University of Alabama at Birmingham/ Childrens of Alabama, Birmingham, Alabama, UNITED STATES.

Bile acid synthesis disorders (BASD) are a group of uncommon metabolic disorders caused by inborn genetic defects in the synthesis of bile acids. Bile acids are a chemical compound that are produced in the liver to promote the flow and excretion of bile in the liver, a key element in absorption of fats and fat-soluble vitamins, and also work in the elimination of cholesterol from the body. Defects in the bile acid synthesis process lead to a failure in creating normal bile acids, resulting in malabsorption of vital nutrients and the overproduction of hepatotoxic bile acids. BASD can present with features of cholestasis, poor weight gain, steatorrhea, hepatosplenomegaly, malabsorption of fats, and fat-soluble vitamins. If unrecognized and untreated this can eventually progress to cause life-threatening complications such as liver cirrhosis and eventually liver failure. We analyze a case study of 7-month-old female Hispanic twins, who initially presented to Endocrinology with hypocalcemic rickets and failure to thrive. They had serum and radiographically abnormalities consistent with severe rickets and were started on treatment. At their follow-up appointments with Endocrinology, they were not gaining weight and had mildly elevated alanine transaminase of 50 U/Liter and 81 U/Liter (normal 5-45 U/liter), normal aspartate aminotransferase and bilirubin, so were referred to Gastroenterology. A liver work-up was continued that led to analysis of their urine by fast atom bombardment mass spectrometry showing sulfate and glycol-sulfate conjugates that are characteristic of a diagnosis of 3 $\beta$ -hydroxy- $\Delta^5$ -C<sub>27</sub>-steroid dehydrogenase (3 $\beta$ -HSDH) deficiency. Its deficiency is the most common inborn error of bile acid synthesis. The genetic analysis of the twins showed this is the first reported case of a defect caused by a Nucleotide change at c.890delT. Treatment with oral bile acid replacement called cholic acid was started at a dose of 10mg/kg/day. Within three months of treatment each child had considerable clinical and biochemical improvement. Bile acid synthesis disorders are such a rare disorder patients initially do not always present with liver disease. It is important to understand that clinical and lab values that assist in the appropriate diagnosis. Bile acid synthesis disorders should be suspected in infantile rickets, particularly in a clinical setting of cholestasis, poor weight gain, liver disease of unknown cause, or abnormalities of fat or fat-soluble vitamin absorption. Early diagnosis and treatment is imperative because bile acid replacement will help with overall improvement of liver function, fat-soluble vitamin deficiency, and weight

gain. A thorough work-up and multi-disciplinary approach can improve diagnosis and treatment of a  $\beta$ -HSDH deficiency, preventing the complications of cirrhosis and eventually liver failure in these twins.

**634 BILIARY STRICTURE AS A PRESENTING FEATURE OF ATYPICAL KAWASAKI DISEASE.** P. Reddy, Pediatrics, Montefiore Medical Center, New York, New York, UNITED STATES. D. Kogan, Y. Rivas, Pediatric Gastroenterology and Nutrition, Montefiore Medical Center, Bronx, New York, UNITED STATES. E. Levin, Radiology, Montefiore Medical Center, Bronx, New York, UNITED STATES. S. Ho, Medicine, Montefiore Medical Center, Bronx, New York, UNITED STATES.

Kawasaki disease (KD) is an acute, systemic vasculitis of small and medium size arteries that predominantly affects patients younger than 5 years. While cholestasis and gallbladder hydrops are common biliary manifestations of KD, biliary strictures are less often reported. We present a case in which the diagnosis of KD was made at the time of biliary stenting of a common bile duct (CBD) stricture in a patient who presented with abdominal pain. Case: A 3 year old girl with a history of constipation presented to 3 hospitals with abdominal pain and pruritic rash. Mom reported that these symptoms occurred immediately after eating. She was initially treated for constipation, and then with steroids and diphenhydramine. On the third presentation, patient was febrile to  $38.9^{\circ}\text{C}$  and diagnosed with a viral illness. Two weeks later, she presented to our ED with persistent abdominal pain, and worsening fatigue. Exam was notable for temperature of  $37.4^{\circ}\text{C}$ , scleral icterus, and a liver palpable 4.5cm below the costal margin. While in the ED, patient developed urticaria on her back and abdomen. This rash resolved with diphenhydramine. Later, she was noted to have desquamation of her palms and soles, and flesh colored papules with central umbilication on her abdomen. Laboratory values were significant for WBC 16.5, platelets 533, total bilirubin (TB) 10.3 mg/dL and direct bilirubin (DB) 7.7 mg/dL, aspartate aminotransferase (AST) 341 U/L, alanine aminotransferase (ALT) 280 U/L, alkaline phosphatase (ALP) 1367 IU/L, amylase 27 U/L and lipase 10 U/L and gamma glutamyl transferase (GGT) 456 U/L. Ultrasound showed an echogenic liver with CBD dilation to 1cm and dilated intra and extra hepatic bile ducts. A magnetic resonance cholangiopancreatography (MRCP) revealed stricture and stenosis of the distal CBD. She underwent endoscopic retrograde cholangiopancreatography (ERCP) with sphincterotomy but unsuccessful balloon dilation of the CBD. After 48 hours, a repeat ERCP with successful stenting of CBD was performed. The echocardiogram revealed ectasia of the left coronary artery and an ejection fraction of 45.7%. Mom was able to confirm over 5 days of intermittent tactile fever prior to presentation, though the patient was afebrile on presentation and throughout initial course. The diagnosis of atypical Kawasaki disease was made and she received IVIG and low dose aspirin. Laboratory values were notable for WBC 23.3, Platelets 558, TB 3.8 mg/dL and DB of 2.1 mg/dL; AST 66 U/L, ALT 43 U/L, ALP was 826 IU/L, amylase 15 U/L and lipase 10 U/L. She was discharged home on the 9<sup>th</sup> day of hospitalization and on follow up 6 weeks later, was noted to have resolution of fevers, abdominal pain, and rash. Discussion: Hepatobiliary disease is a rare sequela of KD, and a high index of suspicion is required for prompt diagnosis and intervention. There are several cases in the literature illustrating the development of hepatobiliary disease as a part of KD. However, to our knowledge, there are no reported cases in which the diagnosis of the vasculitis is made after the confirmation of biliary or pancreatic disease. This case highlights the importance of considering KD in an afebrile child who develops a biliary stricture in the absence of trauma or other contributing condition.

**637 FONTAN LIVER DISEASE EVOLUTION AFTER HEART TRANSPLANTATION ASSESSED BY MRI, LIVER BIOPSY AND CLINICAL SCORING.** D.M. Soler Rodriguez, R. Romero, Pediatric Gastroenterology, Emory University, Atlanta, Georgia, UNITED STATES. K. Kanter, Cardiothoracic Surgery, Emory University, Atlanta, Georgia, UNITED STATES. C. Mao, S. Deshpande, W. Mahle, Pediatric Cardiology, Emory University, Atlanta, Georgia, UNITED STATES. J. Magliocca, Liver Transplantation, Emory University, Atlanta, Georgia, UNITED STATES.

Fontan (FON) surgery yields elevated central venous pressures, non-pulsatile lung blood flow and reduced maximal cardiac output, all of which impact the liver. FON liver disease (FLD) blood tests belie changes seen on abdominal magnetic resonance imaging (MRI) or liver biopsy (LB). Assessing hepatic reserve in FON patients (pts) needing heart transplantation (HT) is challenging. We report liver evaluation and follow up in 7 pts referred for HT after FON since October 2015. METHODS: Standard tests, INR, AFP, and MRI and LB were obtained, if not contraindicated, before or with HT evaluation or catheterization (cath). MRI scores were based on both hepatic congestion and fibrosis (1-mild, 2-moderate, 3-severe), extent of varices (1- for splenomegaly/splenic varices, 2- for gastroesophageal varices, 3- for both 1 and 2) and 1 point each for arterially enhancing nodules (NOD) and ascites (max MRI score =11). LB was scored for both sinusoidal dilatation and fibrosis (1= mild, 2 = moderate, 3=

severe; scores = 2 to 6). Blood tests were performed at 6 and 12 months (mo) post HT with heart cath. Repeat MRI and LB were obtained with 12 mo post-HT cath. Pre- and 12 mo post-HT cath data, hepatic venous wedge pressures (HVWP) and hepatic venous wedge pressure gradients (HVWPG), MELD scores, APRI, VAST score (1 point each for varices/ascites/splenomegaly on imaging, or platelet count <150 K; max =4), creatinine clearance (CrCl), MRI score, and LB score were compared. RESULTS: All 7 pts are alive post-HT. 3 pts reached 12 mo post-HT checks. 2/3 improved in post-HT MELD score and CrCl. All 3 lowered VAST and LB scores. 2/3 have had 12 mo post HT MRI's and both had lower MRI scores. NOD in 1 of these 2 persisted post-HT. CONCLUSIONS: Pts with FLD and low MELD or VAST scores can have successful short term HT outcomes even with pre-HT ascites, advanced hepatic congestion and fibrosis on MRI. MRI, LB, MELD and VAST scores along with CrCl can improve in FLD post-HT. NOD may persist post-HT warranting post-HT imaging.

**638 A CASE OF INFANTILE HEPATIC HEMANGIOENDOTHELIOMAS.** S.T. Boston, S. Palle, Pediatric Gastroenterology, OUHSC, OKC, Oklahoma, UNITED STATES.

In the neonatal period, hepatic tumors are rare. The most frequently occurring type of neonatal hepatic tumors are infantile hepatic hemangioendotheliomas (IHHs) which are classified as benign vascular tumors. They originate from the mesenchymal liver tissue and are composed of small vascular channels lined by endothelial cells. The majority of these vascular tumors are diagnosed within the first six months of life as they show rapid growth during this phase. During this proliferative phase, IHHs can cause congestive heart failure, failure to thrive, ascites, anasarca, respiratory conditions, consumptive hypo-thyroidism, consumptive thrombocytopenia, or severe coagulopathy leading to thrombosis. Rapid resolution often occurs during the first year or two of life marked by involution, thrombosis, and scar formation. A 4-month-old female, with a normal birth and past medical history, presented due to symptoms of nonbloody/nonbilious emeses, fever, and nonbloody diarrhea. The parents also noted increased sleepiness, increased fussiness, increasing abdominal distension, and 3 pounds of weight loss in 2 weeks. Her abdominal x-ray showed significant hepatomegaly. An abdominal ultrasound was done which showed trace ascites and multiple large heterogeneous masses in the liver up to 6 centimeters in diameter with concern for a neoplastic process. A CT of the abdomen was then done which showed multiple liver masses consistent with hemangiomas. The best treatment for an individual patient may depend on factors such as the types of lesions present (IHH lesions may be solitary, multifocal, or diffuse), the lesions' effects on other organ systems, and the extent to which they occupy space in the liver. Previously studied treatments have included corticosteroids, interferon alpha, vincristine, hepatic artery embolization or ligation, surgical resection, or even liver transplant for patients who are difficult to stabilize. Traditionally, first-line therapy has been corticosteroids. However, more recent publications indicate that propranolol is more effective and presents fewer side-effects. Our patient was started on 2 mg/kg/day of oral propranolol divided twice daily, and her dose was later adjusted to 3 mg/kg/day divided twice daily. She is currently responding well to therapy. Recent abdominal ultrasounds have shown involution of the hemangiomas.

**639 HOMOZYGOUS MUTATION OF NEK 8 GENE: A NOVEL CAUSE OF PROGRESSIVE NEONATAL CHOLESTASIS AND END STAGE LIVER DISEASE.** S. Hassan, J.M. Andersen, A. Aqul, Pediatric Gastroenterology, Hepatology and Nutrition, University of Texas Southwestern, Dallas, Texas, UNITED STATES. Umana, Pediatrics, Division of Genetics and Metabolism, University of Texas Southwestern, Dallas, Texas, UNITED STATES. Wolf, Pediatric Nephrology, University of Texas Southwestern, Dallas, Texas, UNITED STATES. Hassan, L. Umana, M. Wolf, J.M. Andersen, A. Aqul, Children's Health, Dallas, Texas, UNITED STATES.

Background Neonatal cholestasis occurs in approximately 1 in 2500 term infants. The most common underlying disease includes extrahepatic bile duct abnormalities, hepatocellular disease, genetic disease and inborn errors of metabolism. The advances in molecular genetics helped elucidate molecular mechanisms of a subset of hepatobiliary diseases that begin during childhood and lead to ongoing liver dysfunction in children and adults. Case We report two male and female siblings born to consanguineous parents from Mexico presented with phenotypically similar neonatal cholestatic liver disease progressing to hepatic failure. Their liver biopsies showed paucity of bile ducts (< 0.6 bile ducts/portal area) without facial, ocular or vertebral findings typically seen with Alagille's syndrome. Genetic evaluation for *JAG1* and *NOTCH2* mutations were negative. The older male sibling eventually progressed to liver failure requiring a liver transplant by 21 months of age. Within a year post transplant, he developed chronic kidney failure requiring dialysis and multiple new cardiac anomalies including pulmonary valve stenosis, aortic valve stenosis and dysplastic mitral valve. The younger female sibling is

following a similar clinical trajectory and now has end stage liver disease at 2 years of age with evolving renal and cardiac disease. Whole exome gene sequencing revealed both siblings have identical homozygous mutations in the *NEK8* gene previously implicated in cardiac, liver and kidney development. However, the homozygous missense mutation (c35G>T; p.R12I) reported in the siblings was in the protein kinase region of the *NEK8* gene, unlike the majority of reported cases where the mutations involves one of the RCC1 (Regulation of Chromosome Condensation 1) domains. The affected amino acid is highly evolutionary conserved to *C. elegans* and the mutation is predicted to be damaging. The sequence variation was not identified in 134 Hispanic individuals from the 1000 genome project. Discussion NEK 8 (Never in mitosis A-related Kinase 8) is a highly conserved ciliary protein kinase critical for normal hepatic, renal and cardiac development in many species including humans. Mutations of NEK8 protein in the RCC1 domains have been associated with a similar clinical phenotype leading to hepatic dysfunction, renal disease, and cardiac anomalies. We report the first case of a homozygous missense mutation in the protein kinase region of the *NEK8* gene to present with advanced liver, renal and cardiac diseases. Conclusion Testing for mutation of the *NEK8* gene should be considered in patients with progressive neonatal cholestasis of unknown etiology, particularly patients with paucity of bile ducts. Patients with *NEK8* mutations should be closely observed for development of kidney and cardiac disease.

**641 UNUSUAL CAUSE OF OBSTRUCTIVE JAUNDICE IN A 5 YEAR OLD MALE.** S. Memarian, F. Jose, Pediatrics, Vidant Medical Center, Greenville, North Carolina, UNITED STATES.

Introduction: Obstructive Jaundice in children is most commonly caused by biliary stones or inflammation. (1) Rarely, it can be caused by malignant neoplasms. There are only a few reports of obstructive jaundice in children secondary to lymphoma. (2) We present a case of a 5 year old male with Burkitt Lymphoma (BL) who presented with obstructive jaundice resulting from tumor compression of common bile duct as the initial presentation of Lymphoma. In this case a rapid growing lymphoma caused obstructive jaundice, so early diagnosis and intervention is critical and life-saving. Case report: A previously healthy 5 year old Caucasian male who presented with painless jaundice and acholic stool of 3 days duration prior to admission, not associated with abdominal pain, vomiting, fever, weight loss or fatigue. Physical exam was only remarkable for icteric sclera. Initial blood work showed normal complete blood cell count (CBC) and negative acute viral hepatitis panel. Complete metabolic panel (CMP) was significant for predominantly conjugated hyperbilirubinemia, elevated Gamma Glutamyl Transferase (GGT), Alkaline Phosphatase (ALP), Alanine transaminase (ALT) and Aspartate transaminase (AST). Prothrombin level (PT), INR, and Albumin were within normal range. Initial workup findings were consistent with obstructive jaundice. Abdominal ultrasound showed 2 hypoechoic masses within the posterior segment of the right hepatic lobe and the uncinate process of pancreas. Follow-up CT and MRI of abdomen showed an indeterminate, 3 x 2 cm well-circumscribed low-attenuation extrahepatic mass along the posterosuperior liver edge in a sub diaphragmatic position and a large, 3.6 x 2.5 cm well-circumscribed low-attenuation mass in the pancreatic head causing secondary intra and extra hepatic biliary, gallbladder, and pancreatic ductal dilation. Due to concern for malignancies, ultrasound guided core needle biopsy of the mass adjacent to the posterior segment of right hepatic lobe was obtained and pathology resulted in final diagnosis of High Grade B Cell Burkitt Lymphoma (BL). Patient was started on chemotherapy treatment and responded well with significant improvement in hyperbilirubinemia and remission of abdominal mass size. No surgical intervention for decompression was necessary. He is currently in remission. Discussion: Obstructive jaundice has rarely been reported as a presenting manifestation of malignant neoplasms, especially in children. (1) Tumor compression resulting in obstructive jaundice is uncommon and usually a late presentation. (3) Burkitt lymphoma (BL) is a type of Non-Hodgkin lymphoma (NHL) and is the most common type of lymphoma found below the diaphragm that affects the gastrointestinal system. (5) Obstructive jaundice as the presenting sign of NHL is found in less than 1% of patients. (4) Our patient presented with obstructive jaundice as the initial symptom of BL resulting from tumor compression of common bile duct. BL is a highly aggressive NHL and is considered the fastest growing tumor with doubling speed in 24 hours, so early diagnosis and intervention for treatment is crucial. (2) Ultrasound is the most important initial imaging technique. (6) Confirmatory imaging studies such as CT scan and MRI should be considered if the ultrasound shows a suspected mass. (1) For our patient, abdominal ultrasound, abdominal CT scan and MRI were useful for initial diagnosis while histology finding of ultrasound guided biopsy of the mass confirmed the diagnosis of BL. Patient was treated with chemotherapy with significant improvement of hyperbilirubinemia and remission of abdominal mass size. In conclusion, although NHL manifesting primarily as obstructive jaundice is uncommon, it should be included as a possible differential diagnosis in a child with obstructive jaundice. This case emphasizes

that patient may not present with the stereotypical features of NHL and high index of suspicion may be necessary in diagnostic imaging and guiding towards the appropriate management course. References: 1. John B. Pietsch, Sadhna Shankar, Connie Ford, and Joyce E. Johnson Obstructive Jaundice Secondary to Lymphoma in Childhood *Pediatr Surg* 2001; 36:1792-1795. 2. R. D. Wammanda, F. U. Ali, S. J. Adama, H. W. Idris & U. Abubakar Burkitt's lymphoma presenting as obstructive jaundice *Annals of Tropical Paediatrics* (2004) 24, 103-1063. Hsu C-F, Ko S-F, Hsiao C-C, Shieh C-S, Huang C-C, Huang F-C Obstructive Jaundice as the presenting manifestation of Burkitt's Lymphoma in a 4-year-old boy. *J Formos Med Assoc* 2003;102:105-84. Pietsch JB, Shankar S, Ford C, et al. Obstructive jaundice secondary to lymphoma in childhood. *J Pediatr Surg*. 2001;12:1792-17955. Ravindra KV, Stringer MD, Prasad KR, et al. Non-Hodgkin lymphoma presenting with obstructive jaundice. *Br J Surg*. 2003; 90:845-849. 6. Saini S Imaging of the hepatobiliary tract. *N Engl J Med*. 1997;336(26):1889.

**642 PUZZLING CASE OF PORTAL HYPERTENSION.** S. Nasiri, Z. Molle-Rios, Gastroenterology, Nemours/A.I. DuPont Hospital For Children, Wilmington, Delaware, UNITED STATES. Nasiri, Z. Molle-Rios, Sidney Kimmel Medical College at Thomas Jefferson University, Philadelphia, Pennsylvania, UNITED STATES.

**Introduction:** Portal hypertension (PH) is defined as portal pressure greater than 10mmHg or a hepatic venous pressure gradient greater than 4mmHg. The most common cause of PH is of hepatic origin, with the presence of cirrhosis resulting from either biliary or hepatocellular diseases. We present a case of PH of unclear etiology. **Case:** We present a 9month (mo) old male born at 36 weeks with congenital cytomegalovirus (CMV) infection treated with 7mo of oral valganciclovir. During the neonatal intensive care unit (NICU) stay, an umbilical venous catheter was placed and visualized in the left portal vein. He developed mild direct hyperbilirubinemia which was attributed to CMV infection. An abdominal ultrasound (US) at 5 days of life showed a gallbladder with a normal common bile duct, as well as dilated tubular structures at the porta hepatis. Repeat US at 1mo and 6mo showed possible liver calcifications, enlarged liver and spleen, with a patent portal vein. There were similar findings of dilated tubular structures at the porta hepatis. At 8mo, he presented to an outside hospital with hematemesis, mildly prolonged prothrombin time, anemia, thrombocytopenia, and mild transaminitis. Magnetic resonance venography showed normal liver, gallbladder, and bile ducts. There were findings at the porta hepatis suspicious for cavernous transformation of portal vein with collateral flow along the lesser curvature of the stomach, esophageal varices and recanalized umbilical vein. No ascites was noted. A liver biopsy was performed and revealed normal histology. At this point, he was transferred to our institution for evaluation and management. He received multiple packed red blood cell and platelet transfusions. Further testing showed normal synthetic liver function. A thrombophilia work up was negative. Transjugular CO<sub>2</sub> portography showed an elevated wedge hepatic pressure of 19mmHg, a free hepatic pressure of 3mmHg, with a hepatic wedge pressure gradient (HWPG) of 16mmHg, compatible with severe sinusoidal PH. He had an upper endoscopy showing grade 3-4 esophageal varices. Sclerotherapy was performed successfully with sodium tetradecyl sulfate. Repeat liver biopsy by laparoscopy showed no evidence of cirrhosis, grossly or histologically. It showed minimal nonspecific chronic inflammation with no fibrosis. **Discussion:** Initially, based on imaging studies, the patient's PH was thought to be secondary to extrahepatic portal vein (EHPV) obstruction. However, on evaluation with portography, the finding of a patent EHPV and markedly elevated HWPG made this less likely. In a report by Ghishan, et al., they present a case of "Noncirrhotic Portal Hypertension in Congenital Cytomegalovirus Infection." The patient had an elevated HWPG of 9mmHg, with no gross evidence of cirrhosis on percutaneous liver biopsy, although there was extensive sinusoidal fibrosis on histology. This was not present in our case which makes it an unusual presentation of sinusoidal PH. Our patient continues to undergo upper endoscopies with sclerotherapy. He is scheduled for liver elastography and possible repeat liver biopsy.

**646 ARGINASE DEFICIENCY AND THE HEPATIC COMPLICATIONS.** S. Patel, D.S. Fishman, Pediatric Gastroenterology, Hepatology and Nutrition, Baylor College of Medicine/Texas Children's Hospital, Houston, Texas, UNITED STATES. Burrage, J. Neira Fresneda, Molecular and Human Genetics, Baylor College of Medicine/ Texas Children's Hospital, Houston, Texas, UNITED STATES. de Guzman, Pediatric Rheumatology, Baylor College of Medicine/ Texas Children's Hospital, Houston, Texas, UNITED STATES.

**Background:** Arginase deficiency accounts for 3.5% of all urea cycle defects (UCD) and is inherited in an autosomal recessive manner, with an incidence of ~1 in 950,000. Unlike the majority of other UCDs, arginase deficiency typically presents in later infancy or pre-school age with progressive spastic paraparesis, developmental delay and seizure disorder. Hyperammonemia can occur at any age and hepatic complications can include acute liver failure,

cholestasis, and hepatic fibrosis. Case Presentation: A 2-year-old Hispanic female, with developmental delay and seizure disorder, was referred to hepatology for persistently elevated AST and ALT. Initial work-up included screen for alpha-1-antitrypsin deficiency with Pi Typing, screen for autoimmune hepatitis, and ultrasound imaging. Patient found to be M-type, negative for F-actin antibody, and negative for liver/kidney microsomal antibody. In addition, ultrasound imaging was unremarkable. Yet, subsequent liver biopsy showed granulomatous hepatitis. Considering the differential diagnosis, an infectious and rheumatologic work-up was started. The patient was found to be negative for Brucella, Bartonella, Toxoplasma, Toxocara, HIV, EBV and CMV infection. In addition, she was noted to have only a mildly elevated ACE level of 101 U/L (ref 13-100 U/L), making the concern for sarcoidosis lower. Given her concomitant developmental delay, a genetic evaluation including whole exome sequencing was also done. Whole exome sequencing (WES) showed a homozygous pathogenic variant in *ARG1* (c.466-1G>C), providing evidence for a diagnosis of arginase deficiency. The WES results were confirmed with an elevated plasma arginine level on test of serum amino acids and decreased arginase enzyme activity in RBCs. In this case, the patient has been admitted twice since the time of diagnosis with hyperammonemia. She has had a maximum serum ammonia level of 215  $\mu\text{M}$ /L (ref 22-48  $\mu\text{M}$ /L). Peak AST and ALT values have been 345 U/L (ref 20-39 U/L) and 715 U/L (ref 8-24 U/L), respectively. In addition to protein restriction, she has been started on Ravicti (glycerol phenylbutyrate, a nitrogen-scavenging agent, 7.1 ml/m<sup>2</sup>/day). Her serum AST and ALT have both come down to 32 U/L, about three months following diagnosis and treatment. She continues to have an unremarkable ultrasound image of her liver and shows no additional signs of sarcoidosis. While this patient's hyperammonemia and elevation in AST and ALT can be explained by arginase deficiency, the etiology of her granulomatous hepatitis remains unknown, and not previously reported in arginase deficiency. Discussion: Though rare, it is prudent to keep arginase deficiency on the differential diagnosis in patients who present outside of the neonatal period with progressive neurological signs, such as spastic diplegia, and hepatic involvement.

**651 DIVERSITY OF ABCB11 MUTATIONS IN JAPANESE PATIENTS WITH INTRAHEPATIC CHOLESTASIS ASSOCIATED WITH LOW GAMMA-GLUTAMYL TRANSPEPTIDASE LEVEL.** T. Togawa, T. Sugiura, K. Ohashi, K. Ito, T. Endo, S. Saitoh, Pediatrics and Neonatology, Nagoya City University Graduate School of Medical Sciences, Nagoya, JAPAN. Hayashi, Department of Medical Pharmaceutics, The University of Tokyo, Tokyo, JAPAN.

*ABCB11* (ATP binding cassette family B, member 11) encodes bile salt export pump, which is a transporter protein expressed at the canalicular membrane of hepatocyte. *ABCB11* (NM\_003742.2) is located in 2q24.3-2q31.1 and consists of 27 exons. The results of mutations in *ABCB11* cause several kinds of intrahepatic cholestasis; progressive intrahepatic cholestasis type 2 (PFIC2), benign recurrent intrahepatic cholestasis type 2 (BRIC2), or intrahepatic cholestasis of pregnancy (ICP). To date, approximately 300 *ABCB11* mutations have been reported from Western countries in Human Gene Mutation Database, while *ABCB11* mutations of Japanese patients have not been provided in large series. We aimed to elucidate the diversity of *ABCB11* mutations in Japanese patients with intrahepatic cholestasis associated with low gamma-glutamyl transpeptidase (GGT) level. We carried out molecular genetic analyses with targeted next-generation sequencing by Ion Torrent Personal Genome Machine system (Thermo Fisher Scientific), covering the entire exons of *ABCB11*. We detected 32 mutations in 22 patients from different families. In clinical diagnoses of the 22 patients, PFIC2, BRIC2, ICP, and clinically undiagnosed patients were 10, 4, 1, and 7, respectively. The median value of direct bilirubin and GGT were 5.1 mg/dL (range, 1.5-14.7) and 42 U/L (range, 8-85), respectively. Twelve of the 22 patients had homozygous or compound heterozygous mutations; 7 PFIC2, 4 BRIC2, and 1 ICP. While 10 patients had a mutation on a single allele; 3 PFIC2 and 7 undiagnosed patients. Regarding the mutation type, twenty-eight single nucleotide variants (20 missense and 8 nonsense), 3 small deletions, and 1 duplication were detected. Several mutations occurred in multiple alleles; C129Y in 6 alleles, W330X in 5 alleles, and E636G in 3 alleles. R487H and R575X were also shown in 2 alleles. These 5 mutations were extremely rare variants, referring to 1200 Japanese genetic variation database and Exome Aggregation Consortium dataset. This result was different from the report that E297G and D482G were the most frequent missense mutation in Western country. In our cohort, two patients diagnosed as PFIC2 or BRIC2 were biallelic protein truncating mutation in *ABCB11*. Young patients who have biallelic protein truncating mutations in *ABCB11* are known to be at increased risk of developing hepatocellular carcinoma. Moreover, mutation specific drug therapy of PFIC/BRIC, such as 4-phenylbutyrate therapy, has begun and is showing clinical benefits. Therefore, longitudinal clinical data collection from the patients having mutations in *ABCB11* should be needed. In cases where only a single allelic variant is

identified, we might apply a further genetic investigation, such as an analysis of highly conserved promoter sequences in the non-coding region.

**653** *A SPOTTY SITUATION.* T. Kaenkumchorn, S. Raikar, Pediatrics, Rush Children's Hospital, Chicago, Illinois, UNITED STATES.

**Case Presentation:**Our patient was a 14 day old male born at 31 and 2/7 weeks who was admitted to the Neonatal Intensive Care Unit after birth for prematurity. On day 2 of life, he developed 2 pinpoint cutaneous lesions on his leg and back. The lesions increased in number until day 14 of life when 23 well-demarcated red to violaceous papules were noted on his entire body ranging in size from 1 to 4 mm. Labs were notable for hemolytic anemia (Hct 29.7, retic 4.3%, LDH 418). Liver enzymes were normal. Abdominal ultrasound showed 3 large hepatic hemangiomas, hepatic vessel dilation, and vascular shunting. Echocardiogram showed mild dilation of the left atrium and ventricle and increased systemic cardiac venous return. He developed tachypnea and hepatomegaly concerning for early congestive heart failure. High output heart failure responded well to furosemide 1 mg/kg twice daily. He started propranolol, tolerated dose adjustment well, and was discharged on 3 mg/kg/day divided three times daily. Serial echocardiograms showed improvement of heart function. Furosemide was discontinued prior to discharge.**Discussion:**The differential diagnosis of cutaneous/liver hemangiomas and coagulopathy includes kaposiform hemangioendothelioma (presents with vascular tumors during infancy and early childhood and DIC), multifocal lymphangioendotheliomatosis with thrombocytopenia (presents with mild consumptive coagulopathy and extensive venous and lymphatic malformations), multicentric hepatoblastoma (the most common liver neoplasm in children), and multifocal infantile hemangiomas with extracutaneous liver involvement.**Conclusions:** Our patient was diagnosed with multifocal infantile hemangiomas with hepatic involvement causing high output shunting. Infantile hemangiomas are the most common benign soft tissue tumor of infancy. Most lesions affect the skin and subcutaneous tissues and resolve spontaneously. However, in a small subset of patients there is extracutaneous involvement, most commonly the liver. Multifocal hepatic hemangiomas can lead to arterial to venous vascular shunting and high output cardiac failure. Propranolol has been used to decrease the size of hepatic hemangiomas. Mechanism of action is thought to be vasoconstriction, decreased expression of angiogenic factors, apoptosis, or a direct effect on differentiation of mesenchymal stem cells. Indications for starting propranolol include high output heart failure, hypothyroidism, and rapidly increasing size of hemangiomas. Hypothyroidism may develop due to production of type 3 iodothyronine deiodinase by hemangioma tissue which inactivates thyroid hormone. Side effects of propranolol include hypoglycemia, hypotension, and bradycardia.**Resources:**1. Dickie BH, Fishman SJ, Azizkahn RG, et al. Hepatic Vascular Tumors. *Seminars in Pediatric Surgery* 2014;23(4):168-172.2. Drolet BA, Frommelt PC, Chamlin SL, et al. Initiation and Use of Propranolol for Infantile Hemangioma: Report of a Consensus Conference. *Pediatrics* 2013;131(128): 129-140.3. Mazereeuw-Hautier J, Hoeger PH, Benlahrech S, et al. Efficacy of Propranolol in Hepatic Infantile Hemangiomas with Diffuse Neonatal Hemangiomatosis. *J Pediatr* 2010;157:340-2.4. Mhanna A, Franklin WH, Macini AJ. Hepatic Infantile Hemangiomas Treated with Oral Propranolol - A Case Series. *Pediatric Dermatology* 2011;28(1):39-45.

**656** *ANTIVIRAL THERAPY DURING PREGNANCY SUCCESSFULLY PREVENTED MOTHER-TO-CHILD TRANSMISSION OF HEPATITIS B VIRUS INFECTION.* T. Sugiura, Pediatrics, Gamagori City Hospital, Gamagori, Aichi, JAPAN. Sugiura, T. Endo, K. Ito, S. Saitoh, Pediatrics and Neonatology, Graduate School of Medical Sciences, Nagoya City University, Nagoya, JAPAN. Suzuki, Pediatrics, Juntendo University Graduate School of Medicine, Tokyo, JAPAN. Tajiri, Pediatrics, Osaka General Medical Center, Osaka, JAPAN. Tanaka, Virology and Liver Unit, Graduate School of Medical Sciences, Nagoya City University, Nagoya, JAPAN.

**Introduction** While joint hepatitis B immunoglobulin (HBIG) and hepatitis B vaccine immunoprophylaxis has been shown to reduce mother-to-child transmission of hepatitis B virus (HBV) infection, approximately 10% of chronic HBV infections cannot be prevented. The major risks of chronic HBV infection in the immunization era are maternal hepatitis B surface antigen (HBs) and hepatitis B e antigen (HBe) positivity as well as a high maternal viral load. Antiviral treatment using lamivudine (LMV) or tenofovir disoproxil fumarate (TDF) during pregnancy has been reported to decrease mother-to-child transmission of HBV. Here, we report the results of a multicenter, retrospective study aiming to prevent mother-to-child transmission of HBV in Japan. **Patients** Twenty one infants born to 12 mothers with HBe antigen positive were recruited. The mothers' age were 28 to 37 years old, and 9 were Japanese, 2 were Chinese, and 1 was Vietnamese. Viral markers for hepatitis C and human immunodeficiency virus

were negative in all women. Twelve of 21 infants were the first children and 9 infants were the second children. The upper limit (i.e., 9.0 log copies/mL of HBV DNA) was found for 11 of 12 mothers, while HBV DNA could not be obtained in 1 mother. Methods To prevent mother-to-child transmission of HBV, 3 mothers were treated with LMV (100 mg daily) and 6 mothers were treated with TDF (300 mg daily). Thus, nine infants comprised the treatment group. Antiviral therapy was initiated from 26 to 32 weeks of gestation in 8 of 9 women; however, 1 mother was treated with TDF starting at 22 weeks of gestation because a premature delivery was anticipated. Written informed consents were obtained from all women. This study was approved by the ethics committees of the Nagoya City University Graduate School of Medical Sciences (approval number 694-2) and each hospital. LMV and TDF were discontinued 4 to 8 weeks after delivery, and the viral loads gradually increased to baseline levels. Antiviral therapy was generally well tolerated and there were no significant adverse effects (e.g., gastrointestinal symptoms or elevated alanine aminotransferase levels). Twelve infants were included as the untreated group. All infants were born at term with normal Apgar scores. No congenital anomalies (major or minor) were identified. All 21 infants received immunoprophylaxis with HBIG and HB vaccines. Results HBs antigens were negative at 12 months of age for all 9 children in the treatment group. HBs antigens were positive at 12 months of age for 10 of 12 children in the untreated group. The rate of HBV positivity was significantly lower in the antiviral therapy group than the untreated group ( $p < 0.01$ ). Discussion TDF (pregnancy category B) is categorized as safer than LMV (pregnancy category C) by the United States Food and Drug Administration. In Japan, TDF for chronic hepatitis B was only approved by the Japanese Health Insurance System in 2014. Therefore, TDF was selected in recent patients who were recruited after 2014. Conclusion Antiviral therapy during pregnancy successfully prevented mother-to-child transmission of HBV for high-risk mothers.

**658 IMMUNODEFICIENCY 47 PRESENTING WITH DIRECT HYPERBILIRUBINEMIA AND HEPATIC DYSFUNCTION IN IDENTICAL TWINS.** Y. Gurevich, Y. Lu, Pediatric Gastroenterology, Northwell Health, Cedarhurst, New York, UNITED STATES.

Title: Immunodeficiency 47 presenting with direct hyperbilirubinemia and hepatic dysfunction in identical twins. Introduction: Immunodeficiency 47 is an exceedingly rare X-linked recessive immunodeficiency, with only 11 reported cases in the literature. Patients typically present in infancy with signs of liver disease or recurrent infections. Other manifestations include neurologic findings, such as seizures and intellectual disability. Case Description: Identical male twins (monochorionic, diamniotic) presented to GI clinic at 66 days of life for further evaluation of direct hyperbilirubinemia. They were born at 36 weeks gestation to a 33-year-old, G5P2 mother. On physical exam, each twin had an umbilical and inguinal hernia, an undescended testicle, and hypospadias. Initial blood work was notable for elevated transaminases (AST = 373/365, ALT = 68/66, TB 15.8/17.9, DB of 8.1/8.3 for Twin A and B, respectively). In addition, iron saturation was 100% for both twins. The twins were followed with frequent blood draws to trend their hepatic panel and coagulation markers. The twins were also seen by Hematology secondary to concerns over coagulopathy. Due to progressively worsening coagulation markers (INR 2.39/2.52 in Twin A and B, respectively), the patients were referred to a Liver Transplant Center. A liver biopsy was performed on one of the twins (who had more stable coagulation markers) which was negative for gestational alloimmune liver disease. H&E staining from the liver biopsy revealed non-specific fibrosis, while EM had findings compatible with a lysosomal storage disease. Whole exome sequencing eventually revealed that both twins had immunodeficiency 47. Twin A had persistently elevated transaminases, required TPN, and received a liver transplant 3 months ago. Twin B initially did better with improving transaminases and stable coagulation markers, but eventually did require a liver transplant 1 month ago. Since transplant, both twins have been thriving, though twin B requires tube feedings overnight. Discussion/Conclusion: Immunodeficiency 47 is a rare cause of hepatic dysfunction and immunodeficiency in infancy. Genetic testing should be considered when more common causes have been ruled out, especially in patients who have multiple medical issues.

**664 QUALITY OF LIFE ASSESSMENT AND COST EFFECTIVENESS OF FECAL MICROBIOTA TRANSPLANT FOR RECURRENT C. DIFFICILE INFECTION IN A 3 YEAR OLD CHILD.** L.F. Caicedo Oquendo, A. Medina, Gastroenterology, Nicklaus Children's Hospital, Doral, Florida, UNITED STATES.

Introduction Clostridium difficile is a Gram (+), spore-forming, anaerobic pathogen that causes diarrhea, pseudomembranous colitis, toxic megacolon, septic shock and even death through toxin production. There is a hypervirulent strain called C. Diff NAP1. Recurrence of C. difficile infection (RCDI) can be present in 20-30% of patients after the first infection, increasing to 40-60% after the 3th episode of recurrence. RCDI in the pediatric

population is increasing in prevalence, especially in children ages 1 to 4 years old in both the hospital and community, therefore increasing costs to the healthcare budget. Standard treatment for the first recurrence of CDI is metronidazole. For the second recurrence, oral vancomycin is suggested. For the third recurrence, tapered or pulsed regimen of oral vancomycin is used and consideration of other antimicrobial agents: nitazoxanide, fidaxomicin, and rifaximin. Fecal microbiota transplant (FMT) has been a tolerable, effective treatment in RCDI since the first FMT in 1958. Multiple studies report high success rates and efficacy with FMT for RCDI. FMT have been performed in children as young as 13 months old for RCDI. The delivery of FMT via colonoscopy has shown to have a higher success rate than the nasoduodenal route. The PedsQL is a questionnaire that measures health related quality of life (HRQOL) in children with acute and chronic health conditions that can be used after implementing an intervention. This is a standardized questionnaire that evaluates physical, emotional, social and school functioning within different age ranges and can be disease specific. In the adult population, FMT has shown to be a cost effective intervention for RCDI compared to a vancomycin taper. To our knowledge, there have been no publications that assess cost effectiveness or utilize standardized methods to measure HRQOL before and after FMT for RCDI pediatric patients without IBD. Objective To review the case of a 3-year-old patient with RCDI and to assess the HRQOL through PedsQL and cost effectiveness of FMT for RCDI. Case Presentation A 3-year-old male was referred by pediatric infectious disease due to RCDI with symptoms of abdominal pain, weight loss, and bloody stools (Bristol #5-#7). The patient was found to be NAP1/B1 (+), with negative biomarkers including IBD Sgi Diagnostic (Prometheus). He failed treatments of metronidazole x1, vancomycin x 2, and vancomycin taper x2 and nitazoxanide x1 due to recurrence of symptoms. Following our institution's internal FMT protocol, our patient met the requirement for FMT. Universal FMT donor was used (Openbiome) and delivered via colonoscopy. Gross examination showed mild colitis of transverse colon. There were no intraoperative complications. Within 24 hours post-transplant, patient developed a fever of 102 degrees Fahrenheit that resolved with acetaminophen. Otherwise, FMT was tolerated well. Clinical improvement was noted 48 hours post-FMT. Patient was followed up post-FMT at 2 weeks, 8 weeks, and 6 months. At 8 weeks, C. difficile toxin was negative. His biomarkers remained unremarkable throughout consecutive visits. Patient's HRQOL was assessed pre- and post-FMT with parent reported PedsQL survey. Scores revealed preFMT:66/100 and postFMT:98/100, with higher scores indicating better HRQOL. Discussion We report the case of a 3 year old patient with antibiotic resistant RCDI treated successfully with FMT showing long term engraftment with clinical/biochemical remission and improvement of HRQOL using the PedsQL. His outcome supports prior studies that have shown the success of FMT. Using FMT in a timely manner would have avoided extra costs for our patient and unnecessary usage of antibiotics. Even though using upper FMT is of lower cost, we believe using the colonoscopy route is more cost effective due to higher cure rate and avoidance of diagnostic errors that could present with the same symptoms and potential risk of liability. In addition, to our knowledge, we were the first to utilize a standardized assessment of HRQOL through applying PedsQL to this case.

**667 H PYLORI INFECTION CAUSING HICCUPS IN A PEDIATRIC PATIENT.** N. Yallanki, L. Small-harary, A. Chawla, Peds Gastroenterology, Stony Brook University hospital, New Hyde park, New York, UNITED STATES. J. Lutz, L. Wilks-Gallo, Pediatrics, Stony Brook, Stony Brook, New York, UNITED STATES.

Hiccup is an involuntary, reflex-like activity that begins with contraction of the diaphragm and is terminated by the abrupt closure of the glottis. Multiple systemic disorders can cause intractable hiccup including gastrointestinal pathology. This case focuses on an atypical presentation of *Helicobacter pylori* infection that presented with acute persistent hiccups. Case: An 18-year-old Hispanic male presented to the emergency department (ED) with acute onset of severe persistent hiccups, nausea, and vomiting that occurred up to 12 times a day. Patient was an ill-appearing male with near continuous hiccups and frequent retching. Epigastric tenderness to deep palpation was elicited. Laboratory data were unremarkable. He was admitted to the Pediatric Unit. Over the first 5 days of admission the patient had persistent, progressive symptoms with continuous hiccups and bilious emesis. His hiccups were the most prominent symptom and seemingly triggered the emesis. He tolerated no oral intake and his hydration was maintained with IV fluids and eventually PPN. Multiple therapies to abort the hiccups were attempted including metoclopramide, lorazepam, IV acetaminophen, pantoprazole, magnesium hydroxide, and chlorpromazine without any sustained response. Extensive workup during his admission yielded a negative abdominal ultrasound, abdominal x-ray, abdominal CT scan, neurologic evaluation with brain MRI and EEG, EKG, urine toxicology screen, psychiatric evaluation and LFTs. EGD showed significant nodularity in the gastric antrum. Biopsies revealed severe chronic gastritis with evidence of rod shaped bacteria consistent with *H. pylori*. He

received 3 days of IV triple therapy with erythromycin, metronidazole and pantoprazole as PO medications were not tolerated followed by oral therapy with amoxicillin, clarithromycin, and pantoprazole to complete 10 day course. Symptoms improved within 24hrs with complete resolution by 72hrs. Discussion: Causes of intractable hiccup include gastrointestinal pathology, central nervous system (CNS) lesions, toxic-metabolic disorders, irritation of the diaphragm or vagus nerve, drugs, general anesthesia, postoperative causes, and psychogenic pathology. The hiccup reflex is not fully understood but consists of 3 general components: an afferent limb traveling within the phrenic and vagus nerves, a central processor, and an efferent limb traveling within the phrenic nerve to the diaphragm and accessory nerves to intercostal muscles. Processing is provided by several neurotransmitters, including GABA and dopamine. Triggers for hiccups are conventionally described as central or peripheral. Gastrointestinal pathology, including gastritis, gastric distention and gastroesophageal reflux, is considered a peripheral triggering stimulus for hiccups. A single case series by Rey et al. indicated that 7.9% of male and 10% of female GERD patients had hiccups. Orr and Rowe showed that *H. pylori* infection has been implicated as a cause of hiccup, confirmed via successful bacterial eradication resulting in cessation of hiccups. The putative pathogenesis is that increased acid production following *H. pylori* infection stimulates esophageal mucosa irritating vagal afferents and triggering hiccups. The majority of cases of hiccups are self-limited and resolve spontaneously. Persistent hiccup refers to episodes that last for between 48 hours to 2 months duration. Episodes of hiccups lasting longer than 2 months are considered intractable. Severe and prolonged hiccups may lead to exhaustion, fatigue, malnutrition, weight loss, dehydration and even death in the extreme cases. Patients with intractable hiccups are more likely to have structural or functional irritation involving the reflex arc than patients with self-limited or persistent hiccups. There are case reports in the adult literature suggesting that treatment of *H. pylori* infection can result in resolution of hiccups in cases where it is presumed to be the triggering cause. There are no known case reports in the pediatric literature regarding the association between *H. pylori* and hiccups. This case report is the first description, to our knowledge, of an association between *H. pylori* infection and persistent hiccup in the pediatric age group. Acute persistent hiccup in this case was presumed to be due to moderate to severe gastritis secondary to *H. pylori* infection. *H. pylori* is a gram-negative bacillus that colonizes the mucus layer of the stomach where it can cause gastritis, peptic ulceration and ultimately adenocarcinoma if left untreated. Conclusion: Hiccups are usually self-limited, but with persistent hiccups gastrointestinal pathology should be considered. *H. pylori* is an under recognized etiology of persistent hiccup. Our case clearly elicits an association between *H. pylori* gastritis and hiccups and suggests that *H. pylori* should be considered early in the differential of persistent hiccups.

**668 HEPATITIS E DIAGNOSED AS AUTOIMMUNE HEPATITIS SUCCESSFULLY TREATED WITH CORTICOSTEROIDS.** N.Z. Minkoff, I. Hoffman, Pediatrics, Cohen Children's Medical Center on New York Hofstra Northwell School of Medicine, Northwell Health, New Hyde Park, New York, UNITED STATES. Buzzi, Department of Pediatric Gastroenterology, Cohen Children's Medical Center Hofstra Northwell School of Medicine, Northwell Health, New Hyde Park, New York, UNITED STATES. Haggmann, Division of Pediatric Infectious Diseases, Cohen's Children Medical Center of New York Hofstra Northwell School of Medicine, New Hyde Park, New York, UNITED STATES. K. Williamson, Pediatric Pathology, Cohen Children's Medical Center Hofstra Northwell School of Medicine, Northwell Health, New Hyde Park, New York, UNITED STATES.

Hepatitis E is a challenging diagnosis to make and problematic ailment to treat in pediatric patients who require more than supportive care. It is underdiagnosed in the developed world, often unrecognized or mislabeled due to nonspecific findings which overlap with other causes of hepatitis, including autoimmune hepatitis. While most cases do not require treatment, Hepatitis E can cause rapidly progressing cirrhosis, neurological symptoms and renal injury and there are no high quality randomized control trials for treatment in the pediatric age group. We present the case of an 8 year-old boy who recently emigrated from Bangladesh who presented with liver failure, clinically worsened despite supportive care and had a favorable response to corticosteroid treatment. Initial testing suggested mononucleosis, but confirmatory testing was negative. He was found to have laboratory and histological evidence consistent with autoimmune hepatitis, for which corticosteroid treatment is indicated universally in the pediatric age group. Subsequent to his discharge he was diagnosed with Hepatitis E, for which corticosteroids are not an established treatment, but could be a reasonable choice given the pathophysiology of liver damage due to hepatitis E. This case highlights the diagnostic challenge that hepatitis E poses, the importance of maintaining a high index of suspicion for hepatitis E in those who recently travelled to endemic areas, and illustrates a novel treatment for immunocompetent pediatric patients with liver failure due to hepatitis E.

**670** *PROTEIN-LOSING ENTEROPATHY WITH MUCOSAL EOSINOPHILIA - MENETRIER'S, CMV, EGE, OH MY!*. J. Park, Pathology, University of Texas Southwestern Medical Center, Dallas, Texas, UNITED STATES. P. Kumar, Medical School, University of Texas Southwestern Medical Center, Dallas, Texas, UNITED STATES. S. Mehta, C. Baldwin, E. Cheng, Pediatrics, University of Texas Southwestern Medical Center, Dallas, Texas, UNITED STATES. J. Park, Pathology, Children's Medical Center, Dallas, Texas, UNITED STATES. S. Mehta, C. Baldwin, E. Cheng, Pediatrics, Children's Medical Center, Dallas, Texas, UNITED STATES.

The pathogenesis of protein-losing enteropathies (PLE) can involve inflammatory exudation, increased mucosal permeability, and/or lymphatic dysfunction leading to loss of serum protein via the GI tract. We report a case of PLE associated with mucosal eosinophilia. A previously healthy 2 year old male presented with vomiting, diarrhea, peripheral edema, and hypoalbuminemia. Renal pathologies were initially ruled out. Fecal  $\alpha$ -1-antitrypsin was elevated (1.13 mg/g), indicative of a PLE. Endoscopic findings were notable for large erythematous and edematous gastric folds in the stomach. The gastric mucosa demonstrated cytomegalovirus (CMV)+ immunohistochemical stain and marked foveolar hyperplasia consistent with Menetrier's Disease (MD). However, increased intraepithelial eosinophilia noted in the stomach, terminal ileum, and colon suggested an allergic process such as eosinophilic gastroenteritis (EGE). Quantitative PCR was positive for CMV (3.7 log<sub>10</sub> IU/mL). The patient responded to supportive therapy, including intravenous albumin infusion and diuresis. By 3 weeks, the vomiting and diarrhea ceased without initiating antiviral therapy or steroids. Menetrier's Disease, while commonly seen in adults, is generally a self-limited disease in children. Enlarged gastric rugae and foveolar hyperplasia with glandular atrophy traditionally establish the diagnosis. These findings are more specific if limited to the oxyntic mucosa of the gastric body and fundus with antral sparing. About 1/3 of children with MD have detectable levels of CMV in gastric tissue. Due to the high incidence of childhood CMV infections, a direct causality between CMV and MD has not been shown. An antiviral therapy such as ganciclovir is rarely indicated. Although classic descriptions of MD do not include eosinophilia, ~61% cases have reported peripheral eosinophilia and others have described mucosal eosinophilia. Some speculate the underlying cause of this eosinophilia include an allergic hypersensitivity reaction and increased mucosal permeability due to GI infection allowing for allergen penetration. Eosinophilic gastroenteritis induces PLE via a non-erosive pathology. EGE is a rare condition (8.4/100,000) with highest prevalence in children under the age of 5. Like Menetrier's disease, EGE can present with symptoms including nausea, diarrhea, and ascites. Cases involving the gastric muscular layer can result in gastric wall thickening. EGE is characteristically associated with peripheral and mucosal eosinophilia. However, unlike MD, EGE is a chronic disease that requires either dietary modification or systemic steroid therapy. Fortunately, our patient demonstrated clinical resolution with just supportive care and returned to a healthy state at 1 month follow-up which is consistent with the clinical course of Menetrier's disease.

**671** *HERPES SIMPLEX ESOPHAGITIS IN IMMUNOCOMPETENT CHILDREN: A CASE SERIES*. R. STURM, J. Barreto, M. Clemente, PEDIATRICS, NICKLAUS CHILDRENS HOSPITAL, HOLLYWOOD, Florida, UNITED STATES.

**INTRODUCTION:** Herpes Simplex Virus Esophagitis (HSVE) has been well described in immunocompromised patients, but has rarely been described in the immunocompetent pediatric host. We present a case series of three immunocompetent children diagnosed in our hospital with HSVE and discuss the appropriate workup and management. **METHODS:** Retrospective analysis of the medical records of patients with the combined diagnoses of HSV infection and esophagitis between 1991 and 2016. Three children had confirmed HSVE and had documented immunocompetence. **CASES:** The cases included a 10 year-old male (patient 1), a 16 year-old male (patient 2) and a 16 year-old female (patient 3). All three patients presented to our hospital with acute onset odynophagia/dysphagia, fever and retrosternal pain that failed to improve with supportive management. They were all diagnosed with HSVE based on a combination of tissue immunohistochemical staining, tissue viral culture and tissue HSV PCR. Patient 1 developed a vesicular extremity rash while admitted which was positive for HSV-1. All three children had immunologic testing and were not found to have underlying immunosuppression. Patient 1 had normal lymphocyte subsets and negative HIV serology. Patient 2 had normal lymphocyte subsets and immunoglobulin levels. Work-up for patient 3 included vaccine titers, immunoglobulin levels and lymphocyte subsets, which were remarkable for slight decrease in CD4 lymphocyte count of no clinical significance. All patients achieved complete resolution of symptoms with acyclovir and supportive care. **DISCUSSION:** The most common symptoms of HSVE are acute onset odynophagia, fever and retrosternal chest pain. Interestingly, oropharyngeal lesions are uncommon. Many cases are thought to go unrecognized with symptoms attributed to other self-

resolving causes. There is a male predominance of approximately 3:1. While endoscopic findings of HSVE are non-specific, biopsies are required to differentiate from other infectious and non-infectious etiologies of esophagitis. Immunohistochemistry and viral cultures confirm the diagnosis, although tissue HSV PCR may have higher sensitivity. Immunologic work-up in previously healthy patients is a point of discussion given that infectious esophagitis may be a sign of underlying immunosuppression. While less than twenty pediatric cases have been described, the existing literature does not support the need for immunological testing. Documented immunocompetence in all three of our patients provides further evidence for this. As HSVE is usually a self-limited disease, symptomatic treatment with adequate analgesia, acid suppression and maintenance of hydration remain the mainstay of therapy. Acyclovir has been shown to hasten resolution of symptoms and should be considered. CONCLUSION: HSVE should be suspected in an otherwise healthy patient with acute onset odynophagia, chest pain and fever. A combination of supportive treatment and acyclovir is recommended. Immunological testing is not required in all patients with HSVE, specifically in a previously healthy child with an uneventful recovery.

**674** *HERPES ESOPHAGITIS WITH CONCOMITANT EOSINOPHILIC ESOPHAGITIS IN A CHILD: A CASE REPORT.* T.A. Cristoforo, K. Rietsma, Pediatrics, University of South Florida, Tampa, Florida, UNITED STATES. A. Cristoforo, K. Rietsma, M. Wilsey, Pediatrics, Johns Hopkins All Children's Hospital, St. Petersburg, Florida, UNITED STATES. Wilsey, E.K. Swan, Pre-Medical, Florida State University, Tallahassee, Florida, UNITED STATES.

Introduction: Herpes simplex virus esophagitis (HSVE) is a viral infection of the esophageal mucosa caused by herpes simplex virus (HSV) that usually occurs in immunocompromised patients. However, rare case series and reports have documented occurrences in immunocompetent pediatric patients. Eosinophilic esophagitis (EoE) is an inflammatory condition of the esophagus typically observed in atopic patients. Only a handful of case reports have been published documenting concomitant HSVE and EoE, and the relationship between these two conditions is not well understood. Case: We describe a 13 year-old immunocompetent male who was diagnosed with HSVE via esophagogastroduodenoscopy (EGD) and biopsy. Repeat EGD following treatment of HSVE was consistent with a diagnosis of EoE. He was treated with high dose proton pump inhibitor (PPI) for one month. Repeat EGD findings remained consistent with a diagnosis of EoE. Conclusion: The relationship between HSVE and EoE remains unclear at this time. Practitioners should be mindful of a possible association and continue to publish case reports to further establish the relationship between these conditions. Further studies are needed to determine the best course of treatment for EoE after treatment of HSV.

**676** *AN ORANGE A DAY KEEPS THE DOCTOR AWAY: CHILDHOOD SCURVY AS AN UNUSUAL CASE OF REFUSAL TO WALK.* K. Tribble, A. Broman, S. Garrison, D. Say, Pediatrics, University of California, Davis, Sacramento, California, UNITED STATES.

A 3-year-old male with a history of global developmental delay, repaired myelomeningocele, Chiari type II malformation, and congenital hydrocephalus with ventriculoperitoneal shunt placement presented to the emergency department with increased fussiness and refusal to stand. Initial assessment was notable for right knee swelling as well as gingivitis and stomatitis. Basic laboratory studies were remarkable only for a microcytic anemia. X-rays of the right femur were obtained and revealed a healing femur fracture, prompting a non-accidental trauma work-up. A skeletal survey revealed diffuse osteopenia, and preliminary reads of the MRI right lower extremity were concerning for a subperiosteal abscess of his femur. The patient underwent urgent surgical intervention, where a hematoma in the setting of a healing fracture was noted, rather than an abscess. Nutritional studies were ordered to determine the cause of the hematoma and the osteopenia. Ultimately, he was found to have iron deficiency as well as deficiencies of vitamins A, C, D, and zinc. Based on these lab results and imaging findings, the patient was diagnosed with scurvy. A detailed dietary history uncovered that the patient was extremely selective and only consumed brown and white-colored pureed foods. Further inquiry revealed inadequate calorie consumption by mouth due to oral aversion, prompting nasogastric tube placement and slow titration of his feeds to caloric goals with close monitoring for refeeding syndrome. This case demonstrates that nutritional deficiencies, even those as rare as scurvy, should not be overlooked in the developed world. It also highlights the importance of eliciting a detailed dietary history in all pediatric patients, particularly in those with a history of developmental delay.

**685 COW MILK PROTEIN ALLERGY - THE GREAT MASQUERADER.** N. Laverdure, S. Rached Dastous, K. Grzywacz, V. Marchand, Gastroenterology, hepatology and nutrition unit, Ste Justine Hospital, Montreal, Quebec, CANADA. J. Trebichavsky, Pediatric surgery, Ste Justine Hospital, Montréal, Quebec, CANADA.

Background Cow milk protein allergy is a very common disease, with a prevalence of 2 to 5% in the pediatric population. The diagnosis is often made clinically and appears early, usually during the first few months of life. Common clinical presentations include: reflux, atopic dermatitis, failure to thrive, chronic diarrhea, as well as rectal bleeding or enterocolitis. Severe, life-threatening, presentations are very rare. The 3 reported cases show that cow milk protein allergy may have a very early and severe presentation and can even mimic some surgical diagnoses.

Case n°1 An 8-day-old formula-fed male infant with no significant personal or family history of atopic disease was transferred to our hospital with bilious vomiting and abundant hematochezia. Small intestinal volvulus was suspected, but laparotomy was normal. In the face of persistent rectal bleeding and severe anemia (Hgb 73g/l), rectal biopsy and upper endoscopy were performed and showed mild colitis and severe gastritis with mucosal desquamation. Biopsies revealed an important infiltration of eosinophilic cells (>100/HPF in the stomach and severe eosinophilic proctitis). Peripheral eosinophil count was initially normal, but hypereosinophilia appeared at day 3 of admission (maximum  $2600 \times 10^6/L$  at 16 days of life). After 8 days of bowel rest with total parenteral nutrition, progressive refeeding was initiated, with an amino acid based formula and was well tolerated.

Case n°2 A newborn female infant was transferred to our hospital for suspected intestinal obstruction. She was breastfed and had a brother with cow's milk protein allergy. She presented with bilious emesis and mild rectal bleeding a few hours after birth. Abdominal X-ray showed small bowel dilatation. Ultrasound and small bowel opacification did not demonstrate volvulus or malrotation. Enteral feeding was started with standard formula 48 hours after admission. Three days later, she suddenly deteriorated and developed acute diarrhea with hypernatremic dehydration (Na: 165mmol/l) and metabolic acidosis (pH: 7.145, bicarbonate level: 11mmol/l). Feedings were stopped but 24 hours later she once again developed rectal bleeding. A severe colitis was seen on endoscopy and biopsies showed eosinophilic infiltration of the mucosae with >70 eosinophils/HPF. Rectal bleeding stopped 3 days after she was made NPO. Peripheral eosinophilia (max  $3000 \times 10^6/L$ ) appeared at day 6 of life. She responded well to an amino acid based formula and peripheral hypereosinophilia resolved.

Case n°3 A 10-day-old male infant presented with 24 hours of diarrhea and lethargy. Birth and pregnancy were unremarkable. He was the first child of a mother with severe IgE mediated food allergies and a father with seasonal allergies. He was fed with standard cow's milk based formula. Initial work up revealed severe metabolic acidosis (pH: 6.8, bicarbonate level: 3.7mmol/l) and thrombocytopenia. Diarrhea stopped with bowel rest. Stool sodium was normal and reducing substances were negative. Rectoscopy performed a week after admission showed a pale mucosae and biopsies revealed > 25 eosinophils/ HPF. Peripheral hypereosinophilia was present (maximum of  $1900 \times 10^6/L$ ). This patient is still hospitalized in the ICU and enteral feeding has not yet been initiated.

Discussion The above cases show that cow milk protein allergy can have an acute and severe presentation and can even mimic surgical pathologies in early life. Atypical clinical manifestations of cow milk protein allergy need to be recognized by pediatricians and primary care physicians in order to improve the management of these patients.

**687 AN INNOVATIVE APPROACH TO A CHALLENGING CASE IN INTESTINAL REHABILITATION: DRAMATIC REDUCTION IN PARENTERAL NUTRITION DEPENDENCE WITH NOVEL INTERVENTIONS IN CHILDREN'S HEALTHCARE (NICH).** R. Bensen, J.L. Damman, J. Kerner, D. Naranjo, Pediatrics, Stanford University School of Medicine, Palo Alto, California, UNITED STATES. D. Wagner, M. Harris, Pediatrics, Oregon Health & Science University, Portland, Oregon, UNITED STATES. C. Nesor, Stanford Children's Health, Palo Alto, California, UNITED STATES.

Case: A 13 year old girl with gastroschisis and intestinal atresia was followed at an experienced and multi-disciplinary intestinal rehabilitation center since birth. She had 140cm of small bowel and 40cm colon without an ileocecal valve remaining based on last measurement during a serial transverse enteroplasty procedure for bowel lengthening. Her course was complicated by recurrent Central Line-Associated Bloodstream Infections (CLABSIs), multiple central venous catheter (CVC) replacements, bacterial overgrowth, barriers to adherence, and depression. Despite clinical anticipation of progress toward enteral independence, close follow-up with interdisciplinary care, and the use of multiple community resources to address psychosocial concerns, she still required parenteral nutrition (PN) 5 days per week in order to maintain appropriate weight and hydration. To address her suboptimal intestinal rehabilitation, at the age of 12 years, she was enrolled in an innovative program: Novel Interventions in Children's Healthcare (NICH). The NICH program provides a combination of family- and skills-based interventions to address preventable poor health outcomes in children with medical and psychosocial complexity. NICH

interventionists, intensively trained and heavily supervised lay persons, carry caseloads of 6-10 complex patients and serve as coaches and liaisons with children, caregivers, medical teams, schools and community providers. The interventionists identified and addressed barriers to care. Family supervision of health care management was increased. Family conflict around health care management and feelings of isolation and hopelessness declined. As a result, the patient demonstrated improved adherence to medications prescribed to treat bacterial overgrowth, more consistent tube feed administration, greater oral and gastrostomy tube fluid intake, and engagement with psychiatric care. These changes led to improved weight gain and linear growth, along with a reduction in symptoms associated with bacterial overgrowth. Within 9 months of NICH enrollment, her PN was reduced from 5 to 2 days per week while maintaining hydration and growth. With this reduction, she achieved a longtime goal of changing from a tunneled CVC to an implanted port to allow for participation in desired activities, such as swimming. She experienced no CLABSIs since enrolling in the program. Total hospital days declined from 11 in the year prior to enrollment to 2 during participation. Discussion: The NICH program is a promising interdisciplinary intervention to improve health and quality of life while reducing excess healthcare costs for children with intestinal failure and complex psychosocial challenges. Even without factoring in the reduced costs of hospitalization, using a cost of daily home PN of \$200 (typical estimates range from \$150-\$250), the reduction in PN from 5 to 2 days per week decreases healthcare expenditures by *more than \$31,000* *per year*, substantially greater than the costs associated with the program and increased use of behavioral health services. Despite its high intensity, the NICH program may be cost saving for health insurers for select patients with intestinal failure due to the reduction in high cost PN and associated complications.

**688** *COMMERCIALLY PREPARED BLENDERIZED TUBE FEEDINGS: A SIMPLER ALTERNATIVE.* S. Bibbens, C.A. Collymore, D.S. Jones, San Antonio Military Medical Center, San Antonio, Texas, UNITED STATES.

Introduction Blenderized tube feedings (BTF) have regained popularity after reports of improved feeding tolerance and parental interest in providing whole foods to gastrostomy tube (GT) dependent children. BTF are forsaken for commercial formulas (CF) due to ease of administration, decreased risk of contamination, and standardized nutrient composition. Commercially prepared BTF (CBTF) with standardized nutrient profiles are now available for ease of GT administration. Despite the advent of CBTF and studies showing benefits to BTF, providers are hesitant to recommend BTF. Concerns include adequate nutrient delivery, risk of bacterial contamination, need for frequent follow up, and lack of existing evidence-based outcomes. We sought to examine changes in patient feeding intolerance (FI) and growth parameters in pediatric patients transitioned from CF to CBTF. Methods Four exclusively GT fed pediatric patients with known FI underwent transition from CF to CBTF at two tertiary care teaching hospitals. FI was defined as retching, vomiting, bloating, constipation, and/or diarrhea. With one exception, patients were transitioned from CF to CBTF under the guidance of a physician and dietitian. Parents assessed symptoms before and after the transition. Medical records were reviewed for growth parameters, medication use and co-morbidities. Results Ages ranged from 17 months to 6 years. Duration of CBTF ranged from 4 months to 1.5 years. All patients were tried on more than one CF prior to switching to CBTF. No strict transition protocol was followed. All four patients were on full CBTF by 12 weeks, most within 4 weeks. After the complete transition all parents reported substantial improvements in FI regarding volume tolerated, retching, vomiting, diarrhea and constipation. All patients were able to wean anti-reflux and promotility medications. One patient began taking food by mouth. All patients maintained optimal growth, but patients required 15-20% more calories after the transition. Three patients were managed closely by a dietitian and no infectious or metabolic complications were reported. One patient, not managed by a dietitian, was hospitalized with azotemia and hyponatremia secondary to excessive protein and inadequate sodium provision. Following adequate treatment, this patient was restarted on a CBTF under guidance of a dietitian and follow up at 2 months demonstrated normal electrolytes and renal function. Discussion The results of our small series looking at CBTF were similar to previous studies on BTF. CBTF improved FI, oral intake and use of medications. Growth was slower on CBTF requiring an increased caloric delivery likely from dietary induced thermogenesis. CBTF offer several advantages over customizable diets including ease of preparation, minimal nutrient variability and decreased contamination risks. CBTF appear to be safe when manufacturer directions for preparation and storage are followed and can improve gastrointestinal symptoms. Still, our small series demonstrated that monitoring with a dietitian is beneficial in avoiding insufficient or excessive nutrient provision. Larger cohort studies in future will be important but can be safely undertaken.

**690** *INFANT WITH PINK MILKY BLOOD: A VERY RARE CASE OF CHYLOMICRONEMIA MANAGED WITH FORMULA CONTAINING MCT AND DIETARY MODIFICATION.* S. Nagaraja, R. Nathan, A. Baig, Pediatric , Brookdale University Hospital, Brooklyn, New York, UNITED STATES.

Hypertriglyceridemia with significant elevation in triglycerides are becoming increasingly common in children who are obese and have sedentary lifestyle. But genetic disorders which causes hypercholesterolemia and hypertriglyceridemia in infants are very rarely reported<sup>4</sup>. The latter defect can be induced by an abnormality either in the lipoprotein itself, Lipoprotein lipase deficiency or lipoprotein receptor defect. Here we report a 6 weeks old female infant of middle eastern decent who presented with fever, cough and nasal congestion for 1 week. Physical examination was significant only for eruptive Xanthoma present around the right eyes. During blood draw for sepsis screen, patient noted to be having pink milky blood<sup>3</sup>. Baby was evaluated for sepsis and causes for hyperlipidemia due to the presence of eruptive Xanthomas. Though, the sepsis screen was not significant. The lipid profile showed alarmingly high cholesterol of 975 and triglycerides 1580. Ophthalmology examination was significant for Lipemia Retinalis. US abdomen done in view of pancreatitis due to hypertriglyceridemia was reported normal. EKG and Echo was also normal. Further evaluation of the parents and siblings revealed that the father and the older sister also had pink milky blood with increase in cholesterol and triglycerides, which was undiagnosed. Due to the presence of familial high cholesterol, triglyceride and eruptive xanthoma, further evaluation of genetic causes for hyperlipidemia was done<sup>2</sup>. Genetic analysis showed a T108R mutation in GPIIIBPI gene suggestive of Chylomicronemia due to LPL deficiency<sup>5</sup>. This is an extremely rare type of Hyperlipidemia. Initially mother was advised exclusive breastfeeding with modification in her diet. This induced as sharp increase in triglycerides in the baby. Thus breastfeeding was discontinued. Formula containing 55% MCT was started, which showed good improvement. Hence the after 3 months, formula was changed to those containing 80% MCT. After 8 months, there was a substantial decrease in the cholesterol and triglyceride without administering any lipid lowering medications. The Eruptive Xanthoma spontaneously resolved as the cholesterol and triglycerides showed decreasing trend. The child continues to closely follow up with cardiologist, gastroenterologist and the lipid clinic as chylomicronemia is associated with increased incidence of premature coronary vascular disease, pancreatitis. This case is being presented because of its extreme rarity and unusual presentation in an infant. The use of lipid lowering medications in infants has not been studied because of rarity and hence much data is not available about treating familial hypercholesterolemia and hypertriglyceridemia in infants<sup>1</sup>. Thus it can be a challenge treating these patients. Though the parents consented for the case report on the child. The photos of the patient cannot be published, as parents did not consent for the photo due to religious reasons. Biopsy of the cutaneous xanthoma could not be performed due to cosmetic reasons as lesions were very near to the eyes. The photo of the milky blood is attached.

**693** *DIARRHEA DILEMMA: A NOVEL CONGENITAL DIARRHEAL DISORDER.* J. Sanville, T.J. Menz, C. Cerezo, Pediatric Gastroenterology, Brown University/Hasbro Children's Hospital, North Chelmsford, Massachusetts, UNITED STATES. Gupta, J. Sanchez-Esteban, Neonatology and perinatal medicine, Brown University/Women and Infant's Hospital, Providence, Rhode Island, UNITED STATES.

Congenital Diarrheal Disorders (CDDs) are a rare, complex group of enteropathies that typically present early in infancy and can prove to be a challenging clinical conundrum. Given the wide range of disease severity inherent among this group, the diagnosis is not always readily apparent. Many present with profuse, watery stool after birth resulting in severe dehydration and life-threatening electrolyte abnormalities underscoring the importance of a timely, accurate diagnosis. Currently, there are only 250 cases known worldwide. Here, we report a patient with a likely novel CDD. A term, non-dysmorphic infant was born to consanguineous parents via caesarean section, noted to have marked abdominal distention with dilated, fluid filled bowel loops visualized on abdominal ultrasound. Prenatally, pregnancy was complicated by polyhydramnios with amniotic fluid index (AFI) > 40 cm. Within the first 24 hours of life, the infant had passed 620 cc of fluid per rectum which was initially replaced with intravenous crystalloid and subsequently transitioned to total parenteral nutrition (TPN). Stool output continued to be large volume (> 10 cc/kg/day) despite exclusive TPN the first 7 days of life, raising the question of a possible congenital secretory diarrheal disorder. Upper endoscopy and sigmoidoscopy with biopsies were performed revealing normal gross, histopathologic and electron microscopic findings. Stool electrolytes were difficult to quantify, however stool samples illustrated excessive amounts of sodium (> 100 mmol/L) and a fecal osmotic gap of 4 mOsm/kg consistent with a diagnosis of congenital sodium diarrhea (CSD). Fluid balance and electrolytes were carefully monitored, replaced and supplemented to correct ongoing losses. TPN was discontinued after 7

days as the infant tolerated oral feeds. Serum electrolytes have remained normal on regular diet and supplementation with lactated ringers (LR) via gastrostomy tube in the outpatient setting. Infant is now 7 months old and is growing and developing normally. She continues to have excessive fecal sodium losses and intermittent metabolic acidosis. Chromosomal microarray analysis did not identify mutations responsible for most common causes of CSD including GUCY2C, SLC9A3, or SPINT2. Whole exome sequencing has been sent and results are pending. As the pool of CDDs continues to expand, advances in molecular genetics to further diagnostically elucidate causative relationships among these disorders will hopefully enable advances in therapeutic and prognostic indications for these patients and their families. Here, with our patient we hope to add to that pool of knowledge with a novel disorder as we await final whole exome sequencing results.

**698** *IDIOPATHIC HEMORRHAGIC PANCREATITIS IN A PEDIATRIC PATIENT.* M.L. Harvie, Pediatric, UTHSC, Memphis, Tennessee, UNITED STATES. J.K. Eshun, D.D. Black, Pediatric Gastroenterology, Le Bonheur Children's Hospital, Memphis, Tennessee, UNITED STATES. Temtem, J.K. Eshun, D.D. Black, Pediatric Gastroenterology, UTHSC, Memphis, Tennessee, UNITED STATES.

Background: Hemorrhagic pancreatitis is a rare complication of acute pancreatitis and can occur due to severe inflammation, regional necrosis, or pseudocysts causing major vessel erosion. There are only a few cases in the literature of this condition in the pediatric population, and those reported are associated with an underlying anatomical defect or chronic disease. We report a case of idiopathic hemorrhagic pancreatitis in a pediatric patient. Case presentation: 14-year-old female presented to our emergency department with four days of epigastric pain and non-bloody, non-bilious emesis. She had left upper quadrant and left flank tenderness, voluntary guarding, but no rebound tenderness and no abdominal distention. At presentation, she had no evidence of anemia, lipase 580 U/L, and triglycerides 366 mg/dL. Ultrasonography showed inflammatory changes and free fluid in the right lower quadrant with moderate free fluid in the deep pelvis; negative for biliary obstruction. Computed tomography showed marked inflammatory changes with fluid around the body and tail of the pancreas and prominent interstitial edema consistent with hemorrhagic pancreatitis. Etiology was unclear given absence of risk factors, with only mild elevation in lipase and triglycerides, which did not correlate with severity of her disease. She was not taking any medications known to cause pancreatitis. Autoimmune workup was negative. Magnetic resonance cholangiopancreatography showed normal biliary and pancreatic ducts without choledocholithiasis or abnormal anatomic variant. Eight days into admission, she had an increase in lipase to 2057 U/L. Repeat imaging showed interval decrease in the peripancreatic fluid collection without any evidence of pseudocyst formation. Hemoglobin levels were stable and triglyceride levels decreased after admission. She did not require any blood transfusions during admission. Peak lipase level was 2600 U/L and amylase was 234 U/L. She improved with conservative management. Discussion: We report a pediatric case of hemorrhagic pancreatitis of unclear etiology. Our patient's triglyceride level never exceeded 400 mg/dL, and had normalized to 118 mg/dL prior to discharge. Pancreatitis from hypertriglyceridemia usually occurs with levels >1000 mg/dL, but can be seen with levels >500 mg/dL. She was readmitted two additional times for recurrent pancreatitis with triglyceride levels of 883 mg/dL and >1380 mg/dL on the second and third admission, respectively. While hypertriglyceridemia is the possible etiology of her recurrent pancreatitis, it seems less likely the cause of her hemorrhagic pancreatitis. Regardless of etiology, management including initial vigorous hydration and attention to fluid status and parenteral nutrition may be necessary as these patients have potential to decompensate.

**702** *ENDOTHERAPY FOR POST-PANCREATIC FLUID COLLECTIONS.* P.K. Kapavarapu, J. Cabrera, Pediatric Gastroenterology, Children's Hospital of Wisconsin, Wauwatosa, Wisconsin, UNITED STATES. K.S. Dua, Gastroenterology, Medical College of Wisconsin, Milwaukee, Wisconsin, UNITED STATES. D. Gregg, Radiology, Children's Hospital of Wisconsin, Milwaukee, Wisconsin, UNITED STATES.

We describe two different modalities of treatments to address the pancreatic fluid collections in a 12yr old child with pancreatic duct disruption secondary to her first episode of pancreatitis. A 12yr old, obese (33.14Kg/m<sup>2</sup>) female with no significant past medical history presented with fever, abdomen pain and vomiting. She was diagnosed first episode of acute pancreatitis (abdomen pain, lipase 2445, CT showed inflamed pancreas). At presentation, she had hypotension, decreased urine output, low grade fevers, elevated creatinine (2.0mg/dL), and an elevated CRP (31.7mg/dL). She was managed with low volume fluid resuscitation, analgesia, and empiric antibiotics. She clinically improved with no signs of bacteremia, off antibiotics but showed signs of low grade fevers. Repeat abdominal CT then revealed: a large pancreatic pseudocyst (14cm diameter) near the neck of the

pancreas that was not compressing on any surrounding structures. She was clinically better and discharged home on day 14 of her initial hospitalization. Two weeks after discharge, she returned to GI clinic with poor appetite, intermittent abdominal pain, emesis and weight loss (10kg since the pancreatitis episode). Repeat abdominal imaging showed maturation of pseudocyst (12cm) with a mass effect on the greater curvature of the stomach. Ascites was seen with some fluid interspersed through the mesentery and tracking down inferiorly along the parabolic gutter. She underwent an endoscopic drainage of the pseudocyst with drainage of 2.4L purulent fluid. A 12mm metal stent and a 7Fr by 5cm double pigtail stent were placed into the pseudocyst. Her appetite improved and she was discharged home. Four weeks later she presented with one week of fevers and abdominal pain. Lipase was normal but CRP was elevated (24mg/dL). Imaging showed resolution of the pancreatic pseudocyst with an intact metal and pigtail stent, and normal stomach. A second collection of 18cm long (cranio-caudally) fluid collection along the parabolic gutter was seen to be persistent. This was not accessible to endoscopic or percutaneous drainage. A third complex heterogenous non-drainable phlegmon of 5.8cm (largest diameter) was noted in the anterior mid abdomen. On further detailed review of these images revealed a disruption of the pancreatic duct between the neck and body of pancreas. Repeat ERCP found a disrupted pancreatic duct downstream from the prior cyst drainage. A sphincterotomy was performed and a pancreatic stent was placed splinting the disrupted pancreatic duct. The previously placed metal and pigtail stent were removed. Shortly after this second ERCP she was able to return home and was clinically stable. Repeat MRI showed improvement in the fluid collections. This case illustrates two infrequent treatment modalities for endoscopic treatment of large pancreatic fluid collections related to acute severe pancreatitis in a child: 1) cystogastrostomy stent placement in a large pancreatic pseudocyst and 2) stenting of a disrupted pancreatic duct to both splint the duct and also to divert the pancreatic juices into the duodenum. Endotherapy with direct fistulization of the GI tract and diversion of the pancreatic juices into the duodenum can lead to fluid resolution and should be considered before surgical or radiological approach.

**703** *SUSPENDING DIAGNOSTIC BIAS: A RARE CAUSE OF EMESIS AND ABDOMINAL PAIN.* P.T. Edwards, J.A. Lee, Pediatrics, UTHSC, Memphis, Tennessee, UNITED STATES. J.A. Lee, C. Cavender, Pediatric Gastroenterology, UTHSC, Memphis, Tennessee, UNITED STATES.

A 15 year African American old male with history of cannabinoid use presented to emergency department with one year of emesis and 20% weight loss. He had not been taking proton pump inhibitor (PPI) as previously prescribed. During initial hospitalization, he underwent esophagogastroduodenoscopy (EGD), which showed esophagitis and numerous gastric and duodenal ulcerations. Helicobacter pylori testing by stool antigen and staining was negative. CMP, CBC, and fasting gastrin levels were normal, and he was discharged home with PPI and sucralfate. Symptoms resolved, but he had 9 admissions over the next 4 months for recurring episodes of emesis and abdominal pain. With each admission, patient displayed non-adherence to medications and tested positive for cannabinoids. His symptoms resolved within 24 hours with bowel rest, acid suppression, and intravenous fluids. Cannabinoid hyperemesis was considered the most likely etiology and given rapid resolution of symptoms, repeat endoscopy was not done during hospitalization. After missing 6 outpatient GI appointments, his first hospital follow up, repeat EGD was scheduled and again showed numerous ulcerations. Repeat fasting gastrin level showed significant increase from 18 to 1667 pg/ml. Follow up abdominal CT showed two enhancing pancreatic lesions, which was confirmed by endoscopic ultrasound. He underwent surgery with excision of lymph nodes overlying pancreas and extensive exploration for other sites of malignancy. No other lesions were noted. It is unclear whether these lesions represent metastases from primary sites that have regressed or atypically migrating G cells from fetal pancreas. Genetic testing was negative for any mutations associated with Multiple Endocrine Neoplasia syndromes. Repeat gastrin levels have remained within normal limits six months after the operation. The existence of gastrinomas primary to a lymph node has been questioned though there have been an expanding number of cases reported. Patient noncompliance coupled with substance abuse can lead to delayed diagnosis because of availability bias. Proper counseling to encourage medication compliance is essential, and seeking alternative diagnoses should be pursued if symptoms persist over time.

**708** *CELIAC DISEASE AND NEUROFIBROMATOSIS TYPE 1 IN AND AFRICAN-MERICAN CHILD.* R.L. Mones, L. Topper, N. Jouayed Oundjian, K. Singh, I. Fennoy, Pediatrics, Harlem Hospital, Columbia University, New York, New York, UNITED STATES. Fennoy, Pediatrics, Columbia University, New York, New York, UNITED STATES.

Introduction Neurofibromatosis type 1 (NF1) is an autosomal dominant genetic disorder that affects one in every 3000 people. Celiac disease (CD) is a common disorder in the developed world where the prevalence is approximately one per 100 people. We report the first case in an African-American pediatric patient with both the NF 1 and celiac disease. Case Report Our 7 year old patient was noted to have numerous café-au-lait spots at birth. Genetic testing confirmed that he had NF1. Despite this, he led a normal life and grew along the 10<sup>th</sup> percentile. At age 7 years he was noted to have premature thelarche and his length curve fell to below the 3<sup>rd</sup> percentile. He was referred to the Pediatric Endocrine clinic wherein a Tissue Transglutaminase IgA (TtG-IgA) antibody was noted to be 81 IU. (<20 IU) He was then seen in the Pediatric Gastrointestinal clinic wherein his history and physical were unremarkable for any other signs and symptoms of CD. The elevated TtG IgA was confirmed to be elevated. (65 IU). A duodenal biopsy showed a Marsh IIIA lesion in the duodenum. Commercial genetic testing for ancestry (23 and me) confirmed that his mom was 98% Sub-Saharan. HLA testing was negative for both DQ2 and DQ8. However this testing revealed the presence of DQA1\*01, DQA1\*03/DQB1\*0202, DQB1\*0501. It was recommended that he go on a GFD. A downward trend in his TtG-IgA levels has been noted thus far. Discussion: We report the first case wherein there is an association between CD and NF1 in a pediatric patient. Moreover, our patient is AA. The former association has been rarely reported in adult patients. Patients with NF1 have gastrointestinal problems and their growth length curve is down shifted. It is also of interest that the patient's maternal ancestry is Sub-Saharan. There are groups from that part of the world that have the highest incidence of CD in the world. There have also been large immigrations of Europeans and to Senegal. Moreover, being negative for HLA types DQ2/DQ8 can be seen in up to 5% of people with CD. Finally, this report emphasizes the need to think of CD in AAs if symptoms suggest that CD may exist despite the apparent ethnicity. 1. Listerick R and Charrow J. Neurofibromatosis type 1 in childhood. *Journal of Pediatrics* 1990; 116: ( 845-853) Biagi F, Campanella J et al. Unusual association of neurofibromatosis type 1 and celiac in a single patient. *Functional Neurology* 2005;20(1);33-34 Szudek J, Birch P et al. Growth in North American white children with neurofibromatosis type 1. *2000 Med Genet*;37;973-938 4. Lorete Maria da Silva Kotze, Absence of HLA-dq2 and DQ8 does not exclude celiac disease in Brazilian patients. *Letter to Editor* ;, *Revisyó Espanola de Enfermedades Digestivo* /2014/106/8/561-562

**709 ISOLATED VITAMIN A DEFICIENCY IN AN ADOLESCENT WITH SHWACHMAN-DIAMOND SYNDROME. S.**

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Shwachman-Diamond Syndrome (SDS) is a genetic disorder characterized by exocrine pancreatic insufficiency (EPI), bone marrow dysfunction and skeletal abnormalities. EPI results in malabsorption of nutrients including fat-soluble vitamins - vitamins A, D, E and K. An 18 year old female with SDS presented for routine gastroenterology follow-up. She has a history of weight loss, poor appetite and poor compliance with medications. The patient was prescribed pancreatic enzyme replacement therapy (PERT) at 3600 units/kg/day of lipase and aquADEK (2 chewable tablets daily), without symptoms of malabsorption (denied diarrhea, steatorrhea, flatulence and abdominal distention). On review of systems, she complained of new onset night blindness. Labs showed mildly low serum vitamin A (19.4 mcg/dL, normal 20-80 mcg/dL), low retinol binding protein (RBP) (1.5, normal 3.0-6.0), and normal retinol:RBP ratio (0.95, normal 0.8-1). Vitamin E, 25-hydroxy vitamin D and PT/INR were normal. Celiac screen, C-reactive protein and zinc levels were reassuring. The patient was referred to ophthalmology where she was noted to have a dark-adapted visual threshold and full field electroretinography consistent with night vision deficits and compromised rod mediated retinal function. The patient was started on 50,000 international units (IU) of vitamin A orally for 2 weeks and repeat labs showed improvements in vitamin A and RBP levels as well as symptoms of night blindness. Her retinol:RBP ratio remained normal. Electroretinography was followed over time and showed improvement. Detailed history revealed the patient had not been compliant with the aquADEK and was not taking the aquADEK with PERT. She required another month of high dose vitamin A at 50,000 IU daily orally followed by 1 month of 20,000 IU daily orally and is now taking aquADEK, 2 tablets daily, with compliance. Night blindness has resolved and the electroretinogram has normalized. Isolated vitamin A deficiency is rare and is often diagnosed by low serum vitamin A levels, and/or low serum retinol:RBP ratios. However, a normal vitamin A level, RBP, and/or retinol:RBP ratio does not always accurately reflect true liver and total body stores of vitamin A. Inflammation and protein malnutrition can depress RBP concentrations. Zinc deficiency can impair RBP synthesis causing falsely low levels. RBP is released into the circulation in a 1-to-1 complex with retinol (vitamin A) and can be used as a surrogate tool to evaluate vitamin A deficiency in the absence of inflammation and malnutrition. Our

patient did have significant wasting so her RBP may not have reflected true body stores of vitamin A and thus, may have been difficult to interpret. It is important to screen for vitamin deficiencies in SDS patients with EPI, especially since long-standing vitamin A deficiency related blindness can be irreversible.

**712 A RARE CASE OF LEUKOCYTOCLASTIC VASCULITIS IN AN ADOLESCENT WITH CELIAC DISEASE.** S. Daniel, M.

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Introduction: Celiac disease (CD) is an autoimmune inflammatory disease of the small intestine as a result of reaction to gluten, a protein found in wheat, rye and barley. The classic presentation of the disease comprises of gastrointestinal symptoms. However, extra-intestinal manifestation have also been reported. Dermatitis Herpetiform (DH) is considered as the cutaneous manifestation of gluten-dependent enteropathy. Leukocytoclastic vasculitis (LCV) manifests as palpable purpura as a result of inflammation within small vessels. The incidence rate is reported to be about 30 cases per million people per year. The exact etiology is unknown but multiple factors have been associated with LCV including drugs, infections, collagen vascular diseases, autoimmune disease and malignancy. Literature search has revealed only a few case reports of LCV associated with CD. There was no evidence of any pediatric case reported. We report an interesting case of a 16 year old male who presented with vomiting, abdominal pain and rash diagnosed with CD in the setting of LCV. Case Presentation: 16 year old male with history of recurrent C.diff presents with a 6 month history of chronic abdominal pain. He then developed a nonpruritic rash on his elbows 2 weeks prior to his presentation. His symptoms progressed to nausea, NBNB emesis, and decreased oral intake 4 days prior to presentation. Physical Exam revealed nonblanching erythematous macules and patches on the arms with crusted papules over bilateral elbows, forearms and right wrist. He also had tenderness to epigastrium. His urinalysis revealed microscopic hematuria. CBC, CMP, Thyroid panel, ESR, CPK, amylase and lipase were within normal range. The result of an autoimmune screen, ANA, anti-double-stranded DNA, anti LKM, anti-smooth muscle, HLA-B27 and ANCA were negative. His ASCA IgA was elevated to 29.7 (0-24.9). His CRP was 2.5 (0-0.5). Viral screen including hepatitis panel was also negative. <i>Antistreptolysin</i> O titer was within normal range. His celiac titers were noted to be abnormal, TTG IgA >100 (0-3), TTG IgG 38 (0-5). Nephrology work up for microscopic hematuria was negative. He underwent an upper endoscopy which showed ulcerations in his stomach and villous blunting with increased intraepithelial lymphocytes in duodenum consistent with celiac disease. His colonoscopy was unremarkable with no evidence of inflammatory bowel disease. He was started on Prilosec 40mg daily, Carafate 1gm TID and gluten free diet. Initially, his rash was thought to be DH, which is commonly associated with CD. However, the rash was then noted to spread to his lower legs and dorsal and plantar feet. At that time, he underwent a skin biopsy, which was consistent with LCV. There was no evidence of IgA depositions as seen in DH. Once he started a gluten free diet, his abdominal pain and rash resolved over the course of a month. His celiac titers normalized within 3 months of being gluten free and he has had no recurrence of the rash. Discussion: Cutaneous manifestations of celiac disease (CD) has been recognized in the pediatric population. There is a strong association between CD and dermatitis herpetiformis (DH). DH is an itchy, blistering skin disease with sites of predilection at the elbows, knees and buttocks. Deposition of immunoglobulin A (IgA) in the papillary dermis is a key diagnostic feature of DH. The classic findings of LCV include vascular and perivascular infiltration of superficial and mid-dermal small blood vessels with neutrophils and granulocytic debris (leukocytoclasia), fibrinoid necrosis and disruption of vessel walls, and extravasation of red blood cells. The exact pathogenesis involved in the association between celiac disease and LCV remains unclear. LCV has been reported to be associated with immunocomplex deposition on the vessel wall, where the antigen may be either exogenous or endogenous. There is evidence that increased intestinal permeability has a pathogenic role in celiac disease. This permeability can allow antigens to penetrate and form immunocomplexes that can circulate and eventually be deposited in the skin. Another proposal is that endogenous antigens from damaged small bowel mucosa can also lead to immunocomplexes deposition in skin. Treatment of LCV with corticosteroids have been controversial. The use of corticosteroids and starting a gluten-free diet in patients with CD has proved to be effective in treating LCV. Our patient had responded well to dietary therapy alone with full resolution of his rash. Conclusion: A variety of dermatological changes have been observed in patients with celiac disease. It is important that clinicians recognize this association to optimize patient management and prevent delay of appropriate intervention. Although LCV is rarely associated with CD, clinicians should be mindful that it may be a cutaneous manifestation of celiac disease.

**714** *INTRACTABLE DIARRHEA DUE TO UNDIAGNOSED DGAT1 MUTATION LEADING TO SEVERE ELECTROLYTE DERANGEMENTS, PNEUMATOSIS INTESTINALIS, PROTEIN-LOSING ENTEROPATHY, AND RICKETS IN A FEMALE INFANT.* T.L. Ratchford, A.J. Kirby, H. Pinz, D. Patel, Pediatrics, Saint Louis University, St Louis, Missouri, UNITED STATES.

Congenital diarrhea disorders (CDD) represent a group of uncommon enteropathies that are often diagnostically challenging. Eight patients with congenital diarrhea due to homozygous recessive DGAT1 mutations have been previously described. DGAT1-related disorder is a rare, autosomal recessive condition caused by loss of function mutations in the diacylglycerol o-acyltransferase (DGAT) 1 gene, which affects lipid metabolism. We report on a female with congenital diarrhea due to loss-of-function compound heterozygous mutations in DGAT1, which to our knowledge has not yet been described. The patient initially had a normal life, but in the first few months of age developed diarrhea that gradually worsened and led to persistent acidosis. She was admitted, discharged, and readmitted to multiple tertiary care hospitals. During one hospitalization, she developed pneumatosis intestinalis. At our presentation, she had severe metabolic derangements, including non-anion gap metabolic acidosis, vitamin-D-deficiency rickets with secondary hyperparathyroidism, and protein-losing enteropathy. Extensive laboratory, pathology, and endoscopic evaluation was performed and negative for infectious, autoimmune, and genetic causes, including testing for recognized congenital diarrheal disorders such as microvillus inclusion disease and tufting enteropathy. She was transitioned to a combination of home parenteral nutrition and nasojejunal feeds and discharged. Due to the unknown etiology of her diarrhea, whole exome sequencing was performed and revealed mutations in DGAT1. Though DGAT1-related disorder is rare, with greater availability of whole exome sequencing, we anticipate an increase in the identification of these pathogenic mutations. With increased recognition of this disease, affected patients may receive a prompt diagnosis and appropriate management.

**715** *THE BENEFITS OF ENDOSCOPIC ULTRASOUND IN THE SETTING OF AN ELEVATED PANCREATIC POLYPEPTIDE IN A 17 YEAR OLD MALE WITH UNDERLYING MULTIPLE ENDOCRINE NEOPLASIA TYPE 1 (MEN1).* T.A. Hadley, A. Freeman, Pediatric GI, Hepatology, and Nutrition, Emory University, Avondale Estates, Georgia, UNITED STATES.

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Introduction: Pancreatic masses in children are rare. Routine pediatric imaging modalities may be insufficient in detecting these lesions. Patient is a 17-year-old male with a diagnosis of Multiple Endocrine Neoplasia Type 1 (MEN 1), confirmed by pathogenic heterozygous mutation in the MEN 1 gene (c.1333delc), which encodes for menin- a tumor suppressor gene. MEN1 is an autosomal dominant in its inheritance and is characterized by tumors in the parathyroid gland, pituitary, and pancreas. Individuals with MEN1 are also at an increased risk for the development for carcinoid tumors, adrenocortical tumors, meningiomas, facial angiofibromas, collagenomas, and lipomas. Given the high risk for development of aggressive and multiple tumors, pre-symptomatic screening is critical and performed frequently., including pancreatic polypeptide levels. Pancreatic polypeptide is a 36 amino acid hormone that is primarily located in the head and uncinate process of the pancreas and elevations have been associated with pancreatic neuroendocrine tumors. Its overall function is to self regulate pancreatic secretion activities. Our patients pancreatic polypeptide level was > 1600, with a normal reported level less than 297. Imaging was completed including an MRI Abdomen with and without contrast (x2), which showed no evidence of a pancreatic lesion and or tumor. Due to his continued elevation in his pancreatic polypeptide level he was referred for an endoscopic ultrasound. On evaluation a subtle but clear, well-defined oval mass lesion arising from the body region of the pancreas was displayed. The lesion was 6 mm, oval, hypoechoic with a halo measuring to 9 mm. A fine needle aspiration was performed and pathology showed atypical neuroendocrine cells, positive for chromogranin, synaptophysin, CD56, and ki67 +. The immunoprofile supported neuroendocrine origin and the clonal appearance was more suggestive of an evolving neuroendocrine tumor. These patients with such characteristics tend to develop a spectrum of neuroendocrine proliferations. This lesion in particular, with its very limited cellularity may represent a nodule such as "hyperplasia" or "dysplasia". Additional imaging was also completed and showed no evidence of tumors in the parathyroid or pituitary glands. Our patient was referred to Surgical Oncology, where close surveillance with repeat imaging was recommended since he continues to remain asymptomatic. Conclusion: Endoscopic US should be strongly considered in patients with elevated pancreatic polypeptide levels and normal routine abdominal US.

**716 PANCREATIC PSEUDOCYST FORMATION IN A PEDIATRIC PATIENT WITH LEFT VENTRICULAR ASSIST DEVICE.** V. Ramachandran, K.P. Shah, Office of Undergraduate Medical Education, Baylor College of Medicine, Houston, Texas, UNITED STATES. Adachi, Congenital Heart Surgery, Texas Children's Hospital, Houston, Texas, UNITED STATES. Jeewa, Division of Pediatric Cardiology, The Hospital for Sick Children, Toronto, Ontario, CANADA. L. Whitfield van Buren, D.S. Fishman, Section of Pediatric Gastroenterology, Hepatology and Nutrition, Texas Children's Hospital, Houston, Texas, UNITED STATES. E. Lopez, J.R. Rodriguez, Division of Pediatric Surgery, Texas Children's Hospital, Houston, Texas, UNITED STATES. F.C. Lee, Department of Pediatrics, Baylor College of Medicine, Houston, Texas, UNITED STATES.

**Background:** There is increasing use of implantable left ventricular assist devices (LVADs) for end-stage heart failure in children. The complications of LVADs include bleeding, hemolysis, infection, and inflammation of adjacent structures, potentially including the pancreas. Acute pancreatitis is an infrequent but severe complication in the post-operative patient associated with a 50% mortality and pseudocyst formation. Conditions predisposing to LVAD thrombosis, a major complication occurring in 9% of adult patients within 2 years of implantation, can lead to hemolysis. The association of hemolysis and pancreatitis has been observed in other disease states. However, limited data is available on pancreatic pseudocyst formation in pediatric patients in the setting of LVADs. **Case Presentation:** A previously healthy 7-year old Hispanic female presented at an outside hospital with a 2-week history of diarrhea and abdominal cramping progressing to lethargy, nausea, and dyspnea. She was intubated secondary to cardiogenic shock. She was subsequently transferred to our tertiary care center in multisystem organ failure secondary to probable viral myocarditis. One month after presentation, she was discharged, but was readmitted two days later for exacerbation of her chronic heart failure and was listed for heart transplant. She required an LVAD for mechanical circulatory support. On post-operative day one, she developed elevated amylase (857 U/L) and lipase (3769 U/L), and status epilepticus in the setting of acute subdural hemorrhage. Anti-coagulation therapy was temporarily held. Gradually increasing abdominal pain and persistently elevated amylase and lipase prompted abdominal computed tomography (CT) scan one week post-operatively, which revealed pancreatic inflammatory changes. Interval CT two weeks thereafter showed acute pancreatitis and hemorrhagic pseudocyst. Additional concern for a possible upper gastrointestinal bleed one month later prompted esophagogastroduodenoscopy, which revealed two moderate-sized ulcers of the gastric body/fundus. One ulcer bed included a concerning finding of possible gray foreign body (Figure 1), prompting exploratory laparotomy. Intraoperatively, an area of gastric ischemic necrosis adherent to the LVAD was identified. An omental flap and hepatorrhaphy was used for tissue coverage and the area was imbricated. The pseudocyst was aspirated and sent for culture. Although the patient tolerated the procedure well, she began to decline post-operatively with no evidence of cardiac support from the LVAD. Testing for the LVAD revealed thrombosis. The patient's critical condition did not allow for re-entry into the abdomen for pump exchange and the patient passed shortly thereafter. **Conclusion:** This vignette illustrates pancreatic pseudocyst formation in the setting of LVAD placement with subsequent thrombosis. Since LVADs are far less common in the management of pediatric cardiac patients compared to their adult counterparts, further investigation into the incidence of and association with pseudocyst formation is needed.