

Thursday, October 8, 2015

POSTER SESSION I

Exhibit Hall

5:00pm – 7:00pm

Presenters at posters from 5:30pm – 6:30pm

ENDOSCOPY/QI/EDUCATION

1 *ESOPHAGEAL SQUAMOUS PAPILLOMA (WART) IN TWO PEDIATRIC PATIENTS: AN UNUSUAL LESION*. A.R. Shahein, Pediatrics, K. Hovnanian Children's Hospital, Tinton Falls, New Jersey, UNITED STATES|T. Matulewicz, A. Soroush, Pathology, Jersey Shore University Medical Center, Neptune City, New Jersey, UNITED STATES|.

Case Summary: Esophageal squamous papilloma (ESP) is a rare benign lesion observed in pediatric and adult patients. Few case reports showed an association with gastroesophageal reflux disease (GERD) or human papilloma virus infection. The natural course of esophageal papilloma is not well described and according to current evidence, most patients are asymptomatic. We report two cases of ESP in adolescent patients. Case #1 is a 14 year-old female with history of long standing constipation, who was being evaluated for new onset diffuse abdominal pain, nausea, heartburn and loss of appetite. Patient had suboptimal response to proton pump inhibitors and interim weight loss, which prompted endoscopic evaluation. Upper endoscopy showed friable esophageal mucosa, 4 x 2 mm polypoid lesion in mid esophagus, gastric hyperemia and small hiatal hernia. Lesion was successfully removed with cold biopsy forceps. Histopathological examination of the lesion showed squamous cell hyperplasia suggesting a benign papilloma with negative in-situ hybridization testing for low and high-risk human papilloma virus in esophageal tissue. Distal and upper esophageal mucosa demonstrated basal cell hyperplasia suggesting acid reflux. Stomach biopsies visualized mild chronic inflammatory cell infiltration in the antrum with negative staining for H. Pylori. Case #2 is a 14 year-old male with history of multiple food allergy, GERD and indeterminate colitis managed with oral proton pump inhibitors (PPI) and Mesalamine. Patient was being evaluated for persistent acid reflux symptoms and new onset diffuse abdominal pain. Upper endoscopy exposed esophageal nodularity, furrowing and mild trachealization. A small 3 x 4 mm polypoid lesion was visualized in upper esophagus and successfully removed with cold biopsy forceps. Histopathological examination confirmed typical appearance of squamous papilloma with no eosinophil infiltration. Gastric mucosal microscopic appearance was suggestive of erosive gastritis with negative staining for H. Pylori. Conclusion: Squamous cell papilloma is defined as a small benign growth involving squamous cells found in the epidermis, the respiratory and digestive tract mucosa. Our patients who were found to have ESP suffered from GERD with failure of the oral proton pump inhibitor therapy to resolve their reflux related symptoms. Previous clinical reports described similar association, yet further studies including larger number of patients are required to better characterize this possible relationship. In contrast to laryngeal papillomatosis, the contribution of Human papilloma virus in solitary esophageal squamous papilloma is unclear. Endoscopists will do well being aware of this rare entity and should thoroughly assess the entire esophagus.

6 *OCCULT MAGNET INGESTION PRESENTING AS MULTIPLE GASTROINTESTINAL FISTULAE IN AN OTHERWISE HEALTHY CHILD*. A. Grover, V.L. Fox, M. Mobassaleh, Gastroenterology, Hepatology and Nutrition, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES|D. Jarrett, Radiology, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES|T. Jaksic, Surgery, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES|.

A fistula is an abnormal communication between two epithelial lined surfaces; enteric fistulas are often the result of surgical intervention, trauma or other inflammatory injury. We present an unusual case of a 4-year-old girl with multiple enteric fistulae in the absence of prior surgery or clinical evidence of active inflammatory disease. CASE DESCRIPTION: A four year old previously healthy female presented for evaluation of intermittent abdominal pain, emesis and diarrhea of 4 month's duration. At the onset, her symptoms consisted of severe abdominal pain and fever for 2 days. She also had multiple bouts of non-bloody, non-bilious, non-feculent emesis and watery stools for the course of 4 days. Thereafter, her symptoms resolved, but would return every 2 weeks and last for one day. In

the interval periods, she was asymptomatic, tolerated a full diet and passed formed stool daily. Review of systems was unremarkable. The family history was negative for inflammatory bowel disease (IBD). PHYSICAL EXAM was normal. LABORATORY examination included normal thyroid function studies, celiac serology, hematological, and biochemical profiles. Her erythrocyte sedimentation rate and C-reactive protein level were mildly elevated. Plain abdominal radiography revealed a moderate stool burden. Given the chronicity of her symptoms and elevated inflammatory markers, endoscopy was performed to investigate possible IBD. Colonoscopy revealed 2 prominent, separate fistulae at the level of the transverse colon. The surrounding mucosa appeared normal. Traversing one fistula led to normal appearing small bowel. The second, smaller fistula could not be passed with the colonoscope. The remainder of the colon and the terminal ileum appeared normal. Biopsies confirmed normal tissue from the colon, terminal ileum, and small-bowel (obtained via the fistula). Esophagogastroduodenoscopy revealed a fistula from the body of the stomach to small bowel. The surrounding mucosa again appeared normal. Barium contrast studies of the upper GI tract, small bowel and colon revealed multiple enteroenteric fistulae. Upon further questioning, it was determined that the patient's father owned multiple "Bucky Ball" neodymium magnets, and the patient had been found playing with these prior to the onset of her symptoms. Laparotomy confirmed the presence of a gastro-colonic, jejuno-colonic, and gastro-jejunal fistula, requiring limited small bowel resection. No inflammation was found in the resected bowel. The patient's symptoms resolved completely after surgery. DISCUSSION: This case illustrates both the significant morbidity associated with accidental magnet ingestion and the potential for using such devices to create stable enteroenteric anastomoses. The application of high-power magnets to achieve desired gastrointestinal anastomoses is an active area of endoscopic and surgical research.

7 *MASQUERADING MECKEL'S? GASTROINTESTINAL PECOMA CAUSING HEMATOCHYZIA IN 9 YEAR-OLD FEMALE.* A. Srivastava, J. Thorn, Y. Zeng, B. Braunhut, A. Har, University of Arizona, Tucson, Arizona, UNITED STATES|.

Background: Perivascular endothelial cell neoplasms (PEComas) are rare mesenchymal tumors that occur in variety of anatomic locations including abdominopelvic and retroperitoneal sites, visceral organs and soft tissues of extremities, skin and bones. Coined in 1996, PEComas are smooth muscle and melanocytic marker positive (smooth muscle actin, HMB45, Melan-A, MiTF) and are part of a family of tumors which include angiomyolipomas, lymphangioliomyomatosis, clear cell "sugar" tumor of the lung, extrapulmonary clear cell "sugar" tumor, and clear cell myomelanocytic tumor. Most recent reviews estimate pediatric PEComa cases involving the GI tract to be under 20. Due to their relative rarity, no formal system currently exists regarding classification, associated prognostic factors, and treatment. Case: A 9 year old previously healthy female presented with 2 episodes of hematochezia. Her hemoglobin decreased from 12.7 (g/dL) to 9.5 in the first 35 hours then down to 6.9 in the following 24 hours. Presentation of massive painless rectal bleeding with currant jelly stools appeared classic for Meckel's diverticulum and patient was taken to the OR without Meckel's scan. Surgical evaluation did not note the presence of a diverticulum; appendix was removed per parents request. Patient hemoglobin continued downtrending post-operation and tagged red blood cell scan was performed, revealing physiologic distribution of tracer and no abnormal radiotracer pooling consistent with no active gastrointestinal bleeding. Decision was made for an esophagogastroduodenoscopy and colonoscopy; colonoscopy revealed a 2.5 cm multilobulated polyp at the hepatic flexure. Location of the polyp was tattooed endoscopically and later removed surgically. Histopathology was consistent with PEComa with no features of malignancy, confirmed from several independent histopathology labs. Although patient's PEComa showed no features of malignancy the margins were not clear; a repeat colonoscopy was performed 6 months post-resection did not reveal recurrence of the lesion and her hemoglobin has since been stable. Conclusion: Although rare in presentation, diagnosis of gastrointestinal PEComa should be entertained in cases of hematochezia without an obvious source of bleeding. Patients with a negative Meckel's scan or seemingly atypical presentation of a Meckel's diverticulum may warrant additional workup.

10 *SUCCESSFUL ENDOSCOPIC REMOVAL OF 12CM TOOTHBRUSH FROM DUODENUM.* A. Lakhole, C. Lin, T. Grikscheit, Y. ZHENG, Pediatric gastroenterology, Children's Hospital Los Angeles, Los Angeles, California, UNITED STATES|.

16 year old female with medical history of bulimia and illicit drug use presented with abdominal discomfort and intermittent epigastric pain. 5 days prior to the presentation, the patient incidentally swallowed a toothbrush handle without the bristles part in an attempt to force emesis. The patient was asymptomatic initially, then developed some abdominal pain and presented to our ED. On presentation physical exam her abdomen was soft, with mild tenderness at the epigastrium. She was afebrile, non-toxic appearing. Lab work included complete blood count and chemistries which unremarkable. CT scan showed 12.5cm long opaque density which was located vertically in the C-loop portion of duodenum and there was no evidence of bowel perforation. She was taken to OR within 12 hours. With gastroscope, a broken toothbrush handle (without bristles) was found in the bulb of duodenum. The duodenum mucosa was severely stretched and the toothbrush tip against the mucosa site was initially mistakenly thought as the downward direction of the lumen. With technical difficulty, the distal end of the toothbrush could not be reached nor visualized. Given the toothbrush was tightly against the duodenum wall, there was no much room to manipulate around the toothbrush. Once confirmed the proximal end of the toothbrush, a polypectomy snare was inserted through the gastroscope. The snare loop was opened completely and placed right next to the tip of the toothbrush. Then the gastroscope was used to push the toothbrush towards the snare loop with some force. The toothbrush fell into the snare loop after few attempts, and a circular sharp edged pressure ulcer was exposed underneath the toothbrush tip with no active bleeding. The proximal end of the toothbrush (~2-3cm) was able to be pulled into the antrum. However, pulling the whole toothbrush out of the duodenum turned out to be the most difficult part given the odd alignment and steric congestion. With surgeons present as back ups, we inflated the stomach to the maximal distend, meanwhile applied great force to pull the snare along with the gastroscope. With multiple attempts, finally we were able to pull the whole toothbrush into the stomach with only minimal mucosal trauma was caused at the duodenum bulb. The toothbrush was then taken out of the stomach without any difficulty or left pieces. Given the deep appearance of the pressure ulcers caused by the toothbrush tip, 3 resolution clips were applied to the ulcer to prevent possible perforation. Post operatively, abdominal X- ray did not show signs of perforation. Patient appeared well, had minimal post-operative pain. She tolerated the advancement of diet and was discharged 2 days after. Follow up at 2 week, the patient did well and presented with no issues since discharge. Conclusion –It is a common practice that a toothbrush can be successfully removed from stomach endoscopically. However, there is no case has been reported that a toothbrush could also be successfully removed from duodenum endoscopically. We demonstrated a case of successful endoscopic removal of a toothbrush located vertically in the c-loop of duodenum, thus avoided surgical intervention and minimized post-operative complications.

12 *THE SAFETY AND EFFICACY OF ERCP IN THE MANAGEMENT OF BILIARY COMPLICATIONS AFTER PEDIATRIC LIVER TRANSPLANTION*. B. Maksimak, K. Radhakrishnan, V. Hupertz, P. Conjeevaram Selvakumar, N. Alkhouri, Pediatric Gastroenterology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES| M. Parsi, Adult Gastroenterology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES|.

Background: Biliary complications (BC) following liver transplantation (LT) in children are common affecting up to 15%- 30% of patients. In adult LT patients, endoscopic retrograde cholangiopancreatography (ERCP) is often the first line diagnostic and therapeutic intervention for BC. Data on the use of ERCP to manage BC in pediatric LT recipients are limited. Methods: We retrospectively reviewed our pediatric LT recipients who underwent an ERCP for BC. We collected information on the indication for LT, biliary reconstruction type, biliary complications, ERCP treatment measures, and complications from the procedure. Results: Eight patients who underwent treatment with an ERCP for suspected BC were identified. The indication for LT was autoimmune hepatitis (n = 4), primary sclerosing cholangitis (n = 2), Byler's disease (n = 1) and Wilson disease (n = 1) with a mean age 16.1 ± 4.4 years. The youngest patient to undergo ERCP in our study was 10 years old at the time of the procedure. Seven of the 8 patients had duct reconstruction in the form of duct-to-duct (D-D) anastomosis and 1 patient had a choledochojejunostomy reconstruction. In all 7 patients with D-D anastomosis, the ERCP was completed successfully. The ERCP on the patient with the choledochojejunostomy was unsuccessful due to difficulty navigating the Roux-en-Y loop. Of the 7 patients on which the ERCP was completed, 4 patients were noted to have an anastomotic stricture, with 2 of these patients also having common bile duct stones and/or sludge. 1 of the 7

patients was noted to have duct size mismatch and the final 2 patients were found to have normal studies. Stent placement was carried out in the 5 patients with anastomotic stricture and duct size mismatch. 4 of these patients underwent further ERCPs with re-stenting of the narrowed regions. In 4 out of the 5 patients treated, intervention with ERCP treatment alone was successful. One patient required surgical treatment. No serious complications developed in any of the patients who underwent ERCP. Conclusions: ERCP is an effective and safe procedure in pediatric patients post LT for the identification and treatment of BC and should be considered in those with D-D biliary reconstruction.

14 *A 17 YEAR OLD BOY WITH FAMILIAL ADENOMATOUS POLYPOSIS AND UNEXPECTED ENDOSCOPY FINDINGS.*
E.J. Brenner, M. KARJOO, SUNY Upstate Medical University, Syracuse, New York, UNITED STATES | .

Familial Adenomatous Polyposis (FAP), an autosomal dominant disease characterized by the presence of numerous adenomatous polyps in the colon, is associated with various extracolonic conditions, including duodenal malignancy. Likewise, celiac disease, an autoimmune small bowel enteropathy, has been reported in conjunction with conditions including small bowel malignancy and autoimmune diseases. To our knowledge, however, FAP has not previously been reported in association with celiac disease. In this case, we describe a 17-year-old male with FAP who was incidentally found to have celiac disease on duodenal biopsy and serology. This report raises the question of whether a connection exists between these pathologies, or if our patient represents an exceedingly rare case. Currently, no readily apparent mechanism links FAP and celiac disease, although the possibility of a genomic link merits consideration. Alternatively, FAP could theoretically induce duodenal inflammation that would precipitate an autoimmune reaction and prompt a response against dietary gluten. To our knowledge, however, no reports exploring this possibility exist in the current literature. At this time, we recommend that physicians caring for FAP patients retain a reasonable index of suspicion for celiac disease, and observe for indicators of celiac disease, including flattening and scalloping of the duodenal mucosa, during upper endoscopy.

15 *EUS ASSISTED RENDEZVOUS PROCEDURE FOR COMPLETE ESOPHAGEAL STRICTURE IN A PEDIATRIC PATIENT..*
F. Lee, D. Agrawal, B.A. Barth, D.M. Troendle, UT Southwestern Medical Center, Dallas, Texas, UNITED STATES | B.A. Barth, D.M. Troendle, Children's Health, Children's Medical Center, Dallas, Texas, UNITED STATES | .

Introduction: Management of complete esophageal strictures (CES) can be challenging. Successful treatment utilizing a simultaneous anterograde-retrograde (rendezvous) endoscope approach has been reported in adult patients. We present a pediatric patient in which endoscopic ultrasound (EUS) was utilized as part of the rendezvous technique to successfully treat a CES. Presentation: A 7-year-old boy presents with progressive dysphagia and inability to tolerate secretions. His history is significant for esophageal transection and gastrostomy placement 9 months prior for treatment of aorto-esophageal fistula secondary to foreign body ingestion with subsequent esophageal re-anastomosis 5 months prior to presentation. Esophagram suggested the diagnosis of CES. The patient was scheduled for an endoscope rendezvous procedure, which identified a benign appearing CES, 22 cm from the incisors and approximately 6mm in length. Contrast injection and failure to advance a guidewire during endoscopy confirmed the diagnosis. Attempts to pass a needle-knife under endoscopic/fluoroscopic guidance were unsuccessful at reestablishing esophageal patency. The patient was subsequently scheduled for an endoscope rendezvous procedure with EUS assistance. Intervention: A linear array echo-endoscope (GF-UC140P, Olympus, Center Valley, PA) was introduced through the mouth and advanced to the level of the stricture. A second endoscope (GIF-XP 160; Olympus) was advanced through the gastrostomy site in a retrograde fashion. A 15mm biliary extraction balloon catheter (Boston Scientific, Marlborough, MA) was passed through the gastrostomy alongside the per-gastrostomy endoscope and positioned just below the stricture using endoscopic/fluoroscopic guidance and was then inflated. The inflated balloon was located endosonographically from the proximal esophagus and served as a target for a 19G needle, which was passed into the distal esophagus under endosonographic/fluoroscopic guidance. A 0.035" guidewire was then passed through the needle into the distal esophagus where it was secured with a snare through the per-gastrostomy endoscope, pulled through the gastrostomy site and secured externally. With dual endoscopic guidance, a needle knife was passed over the wire

to create a tract which was then dilated with 4F,5F,7F, and 10F Sohendra dilators (Cook Medical, Winston-Salem, NC) and then a 15F Savary dilator (Cook Medical). A 10mm x 80mm fully covered Wallflex biliary stent (Boston Scientific) was placed across the stricture under endoscopic/fluoroscopic guidance. After the procedure, the patient experienced no pain or emesis, and was once again able to tolerate his own secretions. He was discharged after 48 hours of observation. 1 month following his procedure, he continues to undergo serial endoscopic dilations, which he is tolerating well. Discussion: For patients with CES, EUS can be utilized as apart of the rendezvous technique to re-establish patency in the pediatric population even when previous attempts at non-EUS guided endoscopic treatments have failed. Successful recanalization can allow the patient to avoid surgery and greatly improve quality of life.

17 *ENDOSCOPIC DIAGNOSIS OF ANTRAL WEBS*. J. Fritz, A. Martinez, D. Lerner, Pediatric Gastroenterology, Children's Hospital of Wisconsin, Wauwatosa, Wisconsin, UNITED STATES].

Introduction: An antral web is a rare cause of obstructive symptoms in children. Diagnosis may be delayed for months or years due to the nonspecific symptoms and only partial obstruction. Data on children with pre-pyloric obstructive anomalies is limited to small case series and emphasize radiographic evaluation for diagnosis (1-3). With this retrospective review, we add our patient experience to the existing literature and discuss the utility of endoscopy and the endoscopic appearance of these lesions. Methods: We reviewed 5 cases of endoscopically identified antral webs diagnosed between 1/1/2005 and 12/31/2012. We then queried our surgical database for all diagnoses of gastric outlet obstruction and antral web during the time frame of 1/1/2012 – 12/31/2014 and reviewed the presentation, method of diagnosis, treatment, and outcomes of these patients. Results: A total of 9 patients with antral webs were reviewed, table 1. The mean age at the time of diagnosis was 4.21 years (1 month – 11.5 years). Presenting symptoms included vomiting, failure to thrive/weight loss, abdominal pain, and abdominal distension. The duration of symptoms at the time of diagnosis ranged from 2 weeks to 11 years. Of the patients who underwent surgery for antral webs from 2012 – 2014, 3 out of 4 were diagnosed endoscopically (1 immediately prior to planned pyloromyotomy). Radiologic evaluation was nondiagnostic in all of our patients, table 1. All 9 patients reviewed underwent resection of their antral web, with 1 being endoscopically resected at an outside hospital. All had improvement in their symptoms following resection with one child transitioning to full oral feeding within 3 months of surgery and three with drastic improvements in their weight for height percentiles. Discussion: An antral web is an uncommon cause of partial obstruction in children. Our experience highlights the importance of considering antral webs as a potential cause of vomiting. Upper endoscopy with a detailed evaluation of the antropyloric region should be considered early in the work-up given that imaging can be falsely reassuring. References: 1) Bell, M., Ternberg, J., McAlister, W., Keating, J., & Tedesco, F. (1977). Antral Diaphragm - a cause of gastric outlet obstruction in infants and children. *Journal of Pediatrics*, 196-202. 2) Feng, J., Gu, W., Li, M., Yuan, J., Weng, Y., Wei, M., & Zhou, X. (2005). Rare causes of gastric outlet obstruction in children. *Pediatric Surgery International*, 635-640. 3) Lui, K., Wong, H., Wan, Y., Hung, C., Ng, K., Tseng, J. Antral web – a rare cause of vomiting in children. *Pediatric Surgery International* (2000), 424 – 425.

19 *CLINICAL OUTCOMES IN BUTTON BATTERY INGESTION IN CHILDREN: A CASE SERIES*. K. Leinwand, D. Brumbaugh, R. Kramer, Digestive Health Institute, Children's Hospital Colorado, Denver, Colorado, UNITED STATES].

BACKGROUND: Clinical outcomes in button battery ingestion are often unpredictable and present a management challenge, as medical complications can range in severity from mild esophageal erosion to tragic death. United States surveillance data has demonstrated increased morbidity and mortality due to the button battery ingestions in the last two decades, raising public health awareness and advocacy efforts to broadcast the danger of button batteries for small children. METHODS: We describe the clinical outcomes in 13 cases of children with button battery ingestion using retrospective chart review. Several objective parameters were compared, including time from button battery ingestion to diagnosis, time from diagnosis to removal, total button battery exposure time, anatomic location of retained button battery, distance from patient location to removal center, clinical outcome,

hospital stay duration, and timing from removal to significant complication. RESULTS: On chart review, the ages of patients with button battery ingestion ranged from 11 months to 6 years of age. All 13 patients experienced medical complications from ingestion. Outcomes range in severity from mild to devastating, and include esophageal ulceration, strictures, esophageal perforation, tracheoesophageal fistula, or in the most extreme cases, death by exsanguination due to aorto-esophageal fistula, as seen in three previously healthy children. CONCLUSIONS: Clinical outcomes appear to vary and be unpredictable, as a relationship between severity of injury and duration of button battery exposure did not directly correlate to the severity of clinical outcome. Because of this prognostic uncertainty, a high level of clinical suspicion for a spectrum of battery-related complications including unknown esophageal injury must be maintained. Further knowledge of outcomes in button battery ingestions will allow clinicians to develop appropriate clinical care pathways to rapidly assess and manage these dangerous foreign body ingestions.

21 *RETAINED WCE : IMPORTANCE OF SHORT INTERVAL BETWEEN PATENCY CAPSULE AND WCE.* M. Gabel, R. Abell, Pediatric GI, University of Rochester, Mendon, New York, UNITED STATES |.

Wireless capsule endoscopy (WCE) is an important diagnostic tool employed to image and evaluate small bowel not visualized on upper or lower endoscopies. 63% of WCEs performed in children are for diagnosis or monitoring of Crohn's Disease (CD). It is more sensitive than small bowel follow through (SBFT) and appears to be complementary with MRE. The overall risk of WCE retention in pediatrics has been reported to be 2.3% but can be increased with active CD. Passage of patency capsule in under 40 hours has a better odds for successful passage of WCE. Rare cases of perforation have been reported in adults due to retained capsule, but none in children. We present the case of a 14 yo female with a small bowel perforation and retained WCE despite successful passage of patency capsule, normal SBFT and no evidence of stricturing on MRE. Our patient initially presented with a one year history of abdominal pain, weight loss, loose stools and iron deficiency anemia. Labs were significant for a Hct of 37, Albumin of 3.8, CRP of 83, and ESR of 31. EGD and Colonoscopy were performed and visually demonstrated ulcers in the duodenum and cecum, and erythema in the sigmoid and rectum. Perianal exam had an indurated area and scarring consistent with a past perianal abscess. Pathology was significant for peptic duodenitis with focal acute inflammation, but colon biopsies were normal. At the time of the procedure, patient was started on 40 mg of prednisone daily, flagyl, and Prilosec. Pentasa was started shortly thereafter. An UGI with SBFT was normal and MRE demonstrated a segment of thick walled terminal ileum (TI), but no stricturing. WCE was pursued to determine extent of small bowel disease. Patency capsule was ingested and passed within 24 hours. Due to family preferences the WCE was performed 6 weeks later. At that point, the patient was on Pentasa, and prednisone 10 mg daily. The WCE demonstrated patchy erythema in distal small bowel but did not exit the small bowel at the end of the study. The family was instructed to watch for its passage in the stool. Less than a week later the patient had a significant increase in abdominal pain. She was taken to the ED and found to have free air on KUB. Patient was taken for Ex-lap and was found to have a perforation at 2 cm proximal to the TI. Proximal small bowel loops were distended and TI was completely strictured and demonstrated no passage. Proximal to this area, the capsule was identified and was mobile in the lumen. It is unusual that despite normal passage of a patency capsule, an unremarkable SBFT, and MRE without stricture, that the capsule would be retained. It is likely that over the course of the 6 weeks between these studies and ingestion of WCE that her disease progressed, especially as her steroids were tapered. We suggest based on these findings that WCE should be performed within 1-2 weeks of patency capsules and that medical management should remain constant over this time period as well.

22 *SPECTRUM OF UPPER GI ENDOSCOPY AT A TERTIARY CARE CENTER OF A DEVELOPING COUNTRY.* M. KHAN, S. Ali, S. Ahmed, Pediatrics & Child Health, The Aga Khan University Hospital, Karachi, PAKISTAN |.

Introduction: Gastrointestinal disorders are important healthcare problem globally. Although upper GI endoscopy is considered to be gold standard in diagnosis of many pediatric gastrointestinal disorders, this is still an underutilized tool in developing countries. The information regarding its efficacy is scanty in most of the developing countries. This can be due to failure to identify the diagnostic importance of procedure, lack of trained

pediatric gastroenterologists, or lack of facilities for endoscopy. Objectives: We carried out this hospital based case series to report the common indications, endoscopic & histopathological findings and complications of pediatric upper GI endoscopy in our setup. Material and Methods: This retrospective study was done at Aga Khan University Hospital, Karachi, Pakistan from 2009-2013. The electronic medical records of all pediatric patients were reviewed retrospectively using EMR. All of the pediatric patients in which upper GI endoscopy was performed during the study period were included in the study. Results: During the study period, a total of 200 upper GI endoscopic procedures were performed. Mean age of patients was 8.5 years. About 2/3 of the patients (66%) received general anesthesia for the procedure. Failure to thrive with suspected coeliac disease was the most common indication for the procedure, seen in 31% patients, followed by recurrent abdominal pain and upper GI bleeding, seen in 18.5% and 15% respectively. Gastritis was the most common abnormal endoscopic finding, seen in 14.5% patients, followed by esophageal varices seen in 5% of children. Almost half of the children (46%) had normal endoscopic findings. Likewise, gastritis was the most common histopathological finding on biopsy, seen in 31% (n=62). Findings were consistent with coeliac disease in 18% (n=36), duodenitis, 10.5% (n=21) and esophagitis, 4% (n=8) of patients. No immediate post procedure complication was noted in our patient population in the study. Conclusion: Upper GI endoscopy is a safe procedure in children. The awareness about its diagnostic and therapeutic role should be raised amongst pediatricians in developing countries. There is also a need to develop training programs of pediatric gastroenterology and pediatric endoscopic suites in developing countries so that children may benefit from this state of the art diagnostic modality.

26 *WHAT ABOUT SWALLOWED HEAVY OBJECTS?*. W. Elfar, N. Patel, Pediatric gastroenterology, University of Rochester Medical Center, Rochester, New York, UNITED STATES].

Introduction: Over 100,000 cases of foreign body ingestions are reported each year in the United States. Among the foreign bodies (FBs) that come to medical attention, 80% to 90% pass spontaneously, 10% to 20% require endoscopic removal and <1% of cases ultimately require surgical intervention. The management of ingested foreign bodies in children is well described in the literature and it varies based on the object ingested, its location, and the patient's age and size. We review a case of a child who intentionally ingested multiple heavy metallic balls, his subsequent management and examine the current guideline and literature available. No guidelines exist for the management of ingested heavy objects. Case report: The patient is a 13 year old male with Asperger's syndrome who presented to the ED with abdominal pain and vomiting after intentionally swallowing 35 metallic balls from the game Magnetix® while reading a book. Despite repeated trials of aggressive bowel clean out with polyethylene glycol (15 caps/255 grams) he only passed four metallic balls spontaneously. Abdominal tenderness was worsening and localized in the right lower quadrant (RLQ). Abdominal radiograph confirmed the presence of 31 round metallic densities in the RLQ. He was unsure if he swallowed magnetic pieces along with the metallic balls. It was initially unclear if the metallic balls were being held together by concomitant ingestion of magnets versus gravitation pull. Complete blood count (CBC) was normal and stools were hemoccult negative. A decision was made to perform a colonoscopy to retrieve the balls. We were able to successfully retrieve all 31 remaining metallic balls with 23 colonoscopies with cecal intubation using the retrieval net. Total weight of the balls was 436 grams (0.961 lbs/15.39 oz), approximately 14 grams (0.5 oz) per ball. The patient tolerated the procedure well with no complications. (Images to be included in the poster). Discussion: Despite aggressive bowel clean out, our patient was unable to evacuate the impacted 31 heavy metallic balls from the cecum. We conclude this is partially due to the effect of gravity, as well as the heavy load of the metallic balls. Although we were able to retrieve the impacted balls, we believe that pressure created from the impaction might have compromised the blood supply to the area, potentially causing perforation and other major complications. In this report, we describe a case where highly dense objects can spontaneously pass through the ileocecal junction, but get impacted in the cecum due to its inability to travel against gravity. This might warrant some additions to the current guidelines for management of pediatric foreign bodies.

EOE/GERD/AERODIGESTIVE

30 *ESOPHAGEAL GRANULAR CELL TUMORS IN THE SETTING OF EOSINOPHILIC ESOPHAGITIS: A CASE SERIES.* B. Kuhn, Pediatric Gastroenterology and Nutrition, Geisinger Health System, Danville, Pennsylvania, UNITED STATES|G.A. Morris, Pediatrics, Geisinger Health System, Danville, Pennsylvania, UNITED STATES|A. Johal, Gastroenterology, Geisinger Health System, Danville, Pennsylvania, UNITED STATES|.

Intro Granular cell tumors (GCT) are benign neoplasms derived from Schwann cells, with only 6 reported GI cases of GCT in kids. There are two reported cases of GCT in individuals with Eosinophilic Esophagitis (EoE), ages 16 and 41 years. We present two additional pediatric cases of esophageal GCT discovered during EoE follow-up endoscopy. Both EoE cases were in remission at the time of GCS finding. Case 1 A 9-year-old boy with autism, asthma, allergic rhinitis, and EoE underwent esophagogastroduodenoscopy (EGD) for EoE follow-up. He was diagnosed with EoE at age 2 years and was treated with 6-food elimination diet and lansoprazole. At 9 years of age, he was asymptomatic and underwent follow-up surveillance EGD on a stable diet. A 2 mm by 4 mm firm nodule was found in the middle third of the esophagus. Biopsy showed squamous mucosa containing submucosal granular cell tumor, which was positive for S100, CD68, and inhibin-alpha immunostaining. Previous EGD one year earlier was without lesion. He underwent removal of GCT via EGD with endoscopic ultrasound and endoscopic mucosal resection. Endosonographic findings were a single oval intramural (subepithelial) hypoechoic lesion in the middle third of the esophagus within the submucosa (Layer 3) and measured up to 3 mm in thickness with well-defined borders. For removal, 2 mL of saline were injected to raise the lesion. Band ligator and snare mucosal resection with Roth net retrieval was performed. A follow-up EGD 3 months later showed a grossly normal esophagus and biopsies without evidence of GCT. Case 2 A 16-year-old boy with asthma, allergic rhinitis, and EoE underwent follow-up EGD. He was initially diagnosed with EoE at age 14 years after a work-up of dysphagia, GERD-like symptoms, and vomiting refractory to acid suppression. His EoE failed 6-food elimination, swallowed fluticasone, and now relies on an elemental diet with isolated single food reintroductions. At age 16 years, follow-up EGD revealed a 2 mm by 4 mm hard, firm nodule in the middle third of the esophagus. Biopsies of the nodule revealed S-100 positivity, consistent with granular cell tumor. Prior endoscopy 3 months earlier was without lesion. He underwent removal of GCT via EGD with endoscopic ultrasound and endoscopic mucosal resection. Endosonographic findings revealed a single small nodule hypoechoic lesion within the submucosa (Layer 3). The mass measured up to 4 mm in thickness with well-defined borders. Resection was performed using band ligator and snare mucosal resection with Roth net retrieval after 2 mL saline injection to raise the lesion. Follow-up EGD 3 months later showed a grossly normal appearing esophagus and biopsies without evidence of GCT. Discussion The first esophageal GCT in a pediatric patient was reported in 2003. GCTs are very rare in children and only one prior pediatric patient has been reported to have esophageal GCT and EoE. Our cases continue to support saline-assisted endoscopic mucosal resection as a safe procedure. The association of GCT and EoE is unclear and may warrant additional investigation with regards to its development in the setting of chronic eosinophilic inflammation.

31 *TOPICAL STEROIDS FOR EOSINOPHILIC ESOPHAGITIS: IS INHALED FLUTICASONE THE EFFICACIOUS AND SAFER CHOICE?* E. Foglio, A.F. Porto, Yale University, New Haven, Connecticut, UNITED STATES|.

Topical steroids (fluticasone propionate or oral viscous budesonide (OVB)) are highly effective for clinical and histological remission in children with eosinophilic esophagitis. When compared to oral steroids they are perceived to have less systemic side effects. Given a disease that is chronic and relapsing, young patients can expect to remain on these medications for long periods of time. There are no studies comparing efficacy and side effects between the two topical therapies in children. We present a case in which a young boy had a significant side effect on OVB with similar efficacy and without any side effects when switched to inhaled flovent. Case: 12 yo male with known lactose intolerance presented with a 6 month history of crampy, epigastric, abdominal pain, and intermittent vomiting. Initial labs were unremarkable and the patient was started on a trial of PPI for possible gastroesophageal reflux. His symptoms initially improved, however 2 months later he reported an increase in

symptoms despite PPI and an endoscopy was performed. Histology was consistent with EoE (25 eos hff in the proximal esophagus). He was continued on a PPI and referred to an allergist. Skin and blood testing were unremarkable therefore the patient was started on budesonide 2mg BID. Four weeks later, mom and his teachers noted that he became sad, withdrawn and could not focus at school. He remained on budesonide until a repeat endoscopy was done 4 months later which revealed complete resolution of esophageal eosinophils. Due to parental concern and persistence of his behavioral changes, budesonide was stopped and he continued on a PPI only. Three months later he underwent a repeat endoscopy, which was again consistent with EoE (innumerable eosinophils in the proximal esophagus). He was started on inhaled flovent 110mcg BID. Repeat EGD 3 months later showed partial improvement, and his flovent was increased to 220mcg BID. Four months after the increase, EGD revealed complete resolution of his eosinophilia. The patient therefore continues on flovent 220mcg BID and nexium. He has done well on this regimen with no clinical symptoms as well as no changes in mood or other side effects. Conclusion: We report a young male with a history of eosinophilic esophagitis who responded to OVB, however developed a side effect of dramatic changes in mood and behavior. He was switched to inhaled flovent, which also led to remission of his eosinophilic esophagitis however without any noted side effects. Currently the only noted side effect with topical steroids includes candidiasis esophagitis. Our case highlights an apparent systemic side effect with oral administration of viscous budesonide. This was not encountered with inhaled flovent. With the anticipated long term maintenance treatment needed for eosinophilic esophagitis, could flovent serve as an efficacious yet safer drug with less systemic absorption? Further studies are needed in children to compare efficacy and side effect profiles between flovent and OVB. In addition, side effects and growth assessments need to be fully examined with long term use of these topical steroids.

34 *FIBROSTENOTIC DISEASE IN CONCURRENT EOSINOPHILIC ESOPHAGITIS AND ESOPHAGEAL ATRESIA.* A. Gupta, A.J. Benitez, M. Chilutti, A. Muir, GI, The Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES].

Eosinophilic Esophagitis (EoE) is a chronic allergic disease characterized by eosinophilic infiltration of the esophagus. Esophageal atresia (EA) is a congenital defect in which the esophagus ends in a blind pouch without connection to the stomach distally. Both conditions are associated with, feeding difficulties failure to thrive, and progressive dysphagia often with esophageal stricture, sometimes requiring esophageal dilation. While an increased prevalence of EoE in patients with EA has been reported, the prevalence of EA within large EoE cohorts has not been previously described. Furthermore, little is known about the clinical course of patients with comorbid EoE and EA. Using an EPIC database search between 2007-2015, we searched for diagnostic codes 750.3 (EA), 530.13 (EoE), and esophageal stricture (530.3) and we looked for overlap of these diagnosis. We performed chart review of patients with EA and EoE as well as those with EoE and stricture to confirm diagnosis of EoE with >15 eosinophils per high power field while on high dose proton pump inhibitor therapy. Stricture was confirmed in these patients as luminal narrowing by esophagram or endoscopy. We identified 2041 patients diagnosed with EoE at The Children's Hospital of Philadelphia. We identified 12 patients (0.59%) with comorbid EoE and EA (EoE/EA). Of the 12 patients with EoE/EA, 8 (67%) had a history of esophageal stricture compared to 12/2041 (0.59%) EoE patients without comorbid EA. Age at diagnosis of EoE in patients with EoE/EA ranged from 3-10 years, and average age of onset of stricture was 4.7 years, compared with average age of stricture onset at 12 years in our EoE alone population. All 8 patients with EoE/EA with stricture, required at least one dilation, compared to 16.7% of EoE patients with stricture. Seven (87.5%) EoE/EA patients required more than 2 dilations, and 2 (25%) required more than 10 dilations, while no EoE patient without EA required more than 1 dilation. Strictures were found to be at the site of anastomosis in 6 of patients in the EoE/EA cohort, and all 6 patients had additional areas of stricture located in the mid/distal esophagus. Despite therapeutic intervention and histologic remission, 62.5% of EoE/EA patients continued to require subsequent dilation. In conclusion, in our population of patients with EoE/EA, we found a more severe fibrostenotic phenotype compared to patients with EoE alone. These patients were more likely to have strictures at an early age and more likely to require multiple dilations despite therapeutic intervention. While these strictures are likely multifactorial, further understanding of this population may lead to greater understanding of the pathophysiology of fibrotic disease of the esophagus.

38 *THE PERFECT STORM: A CASE OF EOSINOPHILIC GASTROENTERITIS IN A PATIENT WITH GLYCOGEN STORAGE DISEASE.* C. Thornhill, N. Tatevian, J. Rhoads, F. Navarro, The University of Texas Health Science Center at Houston, Houston, Texas, UNITED STATES].

Introduction: Eosinophilic gastroenteritis (EGE) is a rare disorder characterized by eosinophilic infiltration of the bowel wall. Multiple nonspecific gastrointestinal symptoms can be observed. Patients may present with evidence of malabsorption such as iron-deficiency anemia, hypoalbuminemia, hypogammaglobulinemia, and protein-losing enteropathy. Here, we report a unique case of a patient with glycogen storage disease type presenting with ketotic hypoglycemia and lactic acidosis subsequently diagnosed with EGE. Case Presentation: The patient is an 8-year-old male with glycogen storage disease type Ia (GSDIa, von Gierke disease) with no previous history of atopy. Prior to presentation, he was maintained on a sucrose-, fructose-, and galactose-restricted diet along with a daily cornstarch regimen. Over 6 weeks, he was admitted multiple times for episodes of vomiting, hypoglycemia, and lactic acidosis. After receiving continuous dextrose-containing crystalloid solution, the patient would improve only to return days later with comparable metabolic derangements. His cornstarch regimen was increased after each hospitalization. Physical exam and labs were otherwise unremarkable, except for a gradually increasing eosinophil count from 800 at his initial presentation to a peak of 5,800. Pediatric gastroenterology was consulted when the patient developed abdominal pain. Endoscopy was performed and was grossly normal. Histology revealed eosinophilic gastroenteritis (eosinophils: esophagus >40 per HPF; stomach >30 per HPF; and duodenum >100 per HPF). Infectious work-up, including evaluation for parasites, was negative. Radioallergosorbent testing revealed allergen-specific IgE to corn, egg white, peanut, soybean, and wheat, while skin allergy testing showed a strong reaction to corn and weak reactions to soybean and rice. The patient was placed on a 2 month prednisone taper with dietary modification consisting of elimination of corn, soy, fish, wheat, and egg. Cornstarch was replaced with tapioca starch. Repeat endoscopy off steroids showed resolution of eosinophilia, and most recent peripheral eosinophilic count was 200. Discussion: EGE in the setting of GSDIa has not been described in the literature. Cornstarch is used as a slowly absorbed carbohydrate to prevent glycogenolysis and gluconeogenesis which is defective in GSDIa. Primary to our case was corn acting as a contributing allergen. Commercially available cornstarch has a protein content of 0.3 g per 128 g cornstarch. Large quantities of cornstarch were consumed on a daily basis (283 g cornstarch with 0.7 g corn protein). Dietary interventions were effective in this case with cornstarch included in the corn elimination. We propose that mucosal involvement and subsequent malabsorption in EGE led to poor utilization of cornstarch, resulting in the classic hypoglycemia and lactic acidosis of GSDIa.

41 *MANAGING EOSINOPHILIC ESOPHAGITIS, LACTOSE INTOLERANCE, & SEVERE IRON DEFICIENCY ANEMIA IN A CHILD WITH AUTISM WHO EXCLUSIVELY DRINKS COW'S MILK.* B. McElhanon, Division of Pediatric Gastroenterology, Hepatology and Nutrition, Emory University School of Medicine, Atlanta, Georgia, UNITED STATES|B. McElhanon, Division of Autism and Related Disorders, Emory University School of Medicine, Atlanta, Georgia, UNITED STATES|R. Berry, L. Nguyen, R. Lesack, Pediatric Feeding Disorders Program, Marcus Autism Center, Atlanta, Georgia, UNITED STATES].

Introduction: Feeding disorders are five times more prevalent in children with autism spectrum disorders (ASD) as compared to their typically developing peers. Children with ASD are also more likely to have gastrointestinal (GI) complaints including diarrhea, constipation, and abdominal pain for unclear reasons. It is widely accepted that they are, at a minimum, just as likely to be diagnosed with GI diseases as children without ASD like celiac disease and eosinophilic esophagitis (EoE). Behavioral treatment of feeding disorders is the only empirically supported treatment. In cases with extensive GI and nutrition involvement, a multidisciplinary approach to treatment must be utilized. We describe a case in which medical, nutrition, and behavioral concerns were addressed simultaneously to produce the quickest and most effective outcome for the patient. Case Report: A 7-year-old male with a diagnosis of ASD presented to a pediatric GI clinic for feeding difficulties. The patient only drank 1 gallon of whole cow's milk via bottle plus occasional banana infant food daily for years. His mother and speech therapists at school had tried getting him to eat other foods for years making no progress. He had chronic and frequent non-bloody loose stools, diffuse severe desquamating rash with excoriations, sleeping while "sitting up",

food allergies, stunted linear growth, and iron-deficiency anemia. Family history was notable for an asthmatic sister and mother with EoE with strictures requiring dilations. A coordinated and detailed medical evaluation revealed EoE, lactase deficiency, deficiencies in iron, vitamin C, and vitamin E, and multiple previously unknown food allergies by skin testing (milk, egg, shellfish, peanuts, tree nuts, mustard, squash, and peas). He initially received intravenous iron replacement because he refused all supplements by mouth. The patient was urgently referred to the intensive 8-week Day Treatment program for severe feeding disorders to increase food variety while providing treatment for his EoE, growth delay, and vitamin deficiencies. With heavy medical and nutritional oversight, the behavioral intervention was targeted to avoid all known allergenic foods as well as foods restricted on a 6-food elimination diet. Therapy was also targeted to wean from cow's milk and replace with an elemental formula. Following the day treatment admission, he was eating a balanced diet of 12 foods. The follow-up evaluation indicated complete resolution of nutrient deficiencies, microcytosis, and anemia with hemoglobin increasing from 8.4 to 12.9 g/dL and reduced numbers of eosinophils per high power field on esophageal biopsies.

43 *EOSINOPHILIC ESOPHAGITIS PRESENTING AS SEVERE NECROTIZING ESOPHAGITIS AND VASCULITIS*. C. Biaggi, A. Langshaw, Pediatric Gastroenterology, University of Miami, Miami, Florida, UNITED STATES|A. Hirzel, Pediatric Pathology, University of Miami, Miami, Florida, UNITED STATES|.

Eosinophilic esophagitis (EoE) is a chronic immune mediated clinicopathological entity characterized by an isolated, inflammatory eosinophilic infiltration of the esophagus. Endoscopy (EGD) findings may reveal esophageal rings, a thickened pale mucosa with linear furrows and white exudates. Diagnosis is confirmed by the histological presence of ≥ 15 eosinophils (eos) per high power field (hpf), in at least one esophageal mucosal biopsy, and/or the presence of other microscopic features of eosinophilic inflammation, such as eosinophilic microabscesses, superficial layering or extracellular eosinophil granules. The diagnosis of EoE includes demonstration of persistent esophageal eosinophilia after a 2 month course of proton pump inhibitor (PPI). Mucosal breaks with erosions and ulcerations are not indicative findings, instead more suggestive of other diagnosis such as GERD, Crohn's disease or infectious esophagitis. We present a case with an unusual presentation of EoE. This is a 17 y/o male with history of bronchial asthma and environmental allergies who presented to the emergency room with a 3 day history of worsening dysphagia and odynophagia. EGD findings revealed a diffusely ulcerated, friable esophageal mucosa. Microscopic findings revealed mild chronic duodenitis and gastritis, mid and distal esophagus with severe acute and chronic necrotizing esophagitis, organizing thrombosis, severely necrotic smooth muscle and associated vasculitis. Immunohistochemistry was negative for HSV, CMV, adenovirus and fungi. He had no anemia, peripheral eosinophilia, inflammatory marker elevation, or hypoalbuminemia. Given histological findings, autoimmune disease, vasculitides and IBD were considered. Laboratory workup was unremarkable. He was treated with PPI and carafate. EGD was repeated after 3 month course of PPIs on our asymptomatic patient and histologic findings revealed normal gastric mucosa, chronic duodenitis, and >25 eos/hpf in the mid and proximal esophagus. Blood allergy testing revealed class 3 (high level) IgE response to milk. He was started on strict dairy free regimen and repeat EGD after 2 months on this diet revealed no esophageal pathology. Our patient had a classic clinical presentation of EoE, but macro/microscopic findings that were not suggestive and diagnostic of it initially. Conservative treatment with PPIs and carafate were implemented which led to symptom resolution. Repeat EGD after 3 months of treatment revealed diagnosis of EoE. To our knowledge, this is the first reported case of EoE with initial presentation of mucosal tears, severe ulceration and histology consistent with acute vasculitis. If EGD had not been repeated in our asymptomatic patient, the diagnosis would have been missed. We were able to achieve histologic remission on target elimination diet. Whether initial findings were due to long standing reflux esophagitis as a late complication of EoE, do both entities coexist, or is this the initial presentation of future IBD or systemic vasculitis remains to be determined. In this case, it was essential to follow up histologic findings to discover diagnosis of EoE in an asymptomatic patient that presented with an abnormal pathology.

50 *CASE SERIES: PEANUT ALLERGY ORAL IMMUNOTHERAPY INDUCING EOSINOPHILIC ESOPHAGITIS*. W.N. Sayej, Digestive Diseases, Hepatology and Nutrition Center, Connecticut Children's Medical Center, Hartford,

Connecticut, UNITED STATES|E. Semancik, W.N. Sayej, University of Connecticut School of Medicine, Farmington, Connecticut, UNITED STATES|.

Background: We describe three pediatric patients with confirmed eosinophilic esophagitis (EoE) following oral immunotherapy (OIT) for treatment of peanut allergies. The EoE symptoms, endoscopic and histologic findings resolved with cessation of the OIT. **Case Reports:** The first patient, 6 years of age, presented with a two-month history of non-bloody, non-bilious emesis and an associated sore throat that began two weeks after starting a peanut desensitization program using OIT, which was discontinued one week prior to our evaluation. Upper esophagogastroduodenoscopy (EGD) demonstrated furrows and white specks throughout the esophagus, which biopsies revealed severe esophagitis (30-40 eos/HPF) and marked basal layer hyperplasia. After discontinuation of OIT and elimination of peanuts from the diet he had complete resolution of symptoms. Repeat endoscopy four months later showed a normal appearing esophagus and histology revealed resolution of the eosinophilia (0 eos/HPF) and inflammation. The second patient, 11 years of age, presented with a two-week history of non-bloody, non-bilious emesis that started two weeks after the patient progressed in his OIT for peanut allergy desensitization to ingesting whole peanuts. EGD demonstrated furrows and white specks and histology showed severe esophagitis (25-30 eos/HPF) with marked basal layer hyperplasia. The symptoms resolved after discontinuation of OIT and elimination of peanuts. Repeat EGD three months later demonstrated complete resolution of endoscopic and histologic findings (0 eos/HPF). The third patient, 7 years of age, presented with a two-month history of dysphagia, which began two weeks after he began maintenance dosing for his OIT for peanut desensitization. EGD revealed severe esophagitis (50-60 eos/HPF) with marked basal layer hyperplasia. Symptoms resolved soon after discontinuation of OIT. Repeat EGD three months later, showed significant improvement with mild to moderate esophagitis (15-18 eos/HPF) with moderate basal layer hyperplasia. **Conclusion:** This is one of a limited number of case reports of EoE induced in pediatric patients after OIT for peanut allergy desensitization. We also documented the reversal of clinical symptoms and histologic findings with cessation of OIT.

52 *CONCURRENT HERPES SIMPLEX ESOPHAGITIS AND EOSINOPHILIC ESOPHAGITIS IN TWO*

IMMUNOCOMPETENT PATIENTS. J. Yeh, Department of Gastroenterology, UCSF Benioff Children's Hospital Oakland, Oakland, California, UNITED STATES|S. Kerbawy, Graduate Medical Education, UCSF Benioff Children's Hospital Oakland, Oakland, California, UNITED STATES|E. Cham, Department of Pathology, UCSF Benioff Children's Hospital Oakland, Oakland, California, UNITED STATES|.

Background: Herpes simplex virus (HSV) esophagitis is a rare finding in immunocompetent patients. The diagnosis of eosinophilic esophagitis (EoE) in association with HSV esophagitis is a rarely reported but possibly related pathologic process. **Case Presentations:** Two patients presented with acute on chronic dysphagia. The first was a 7 year old boy with history of perioral dermatitis who reported 1 week of substernal chest pain and odynophagia even with liquids, however without emesis. On history, mom reports cutting his food into smaller pieces for years but no previous complaints of pain. Upper GI was negative for esophageal narrowing. EGD revealed pan-esophageal ulcerations but no other stigmata of EoE. Omeprazole and sucralfate were initiated. Pathology was positive for esophageal eosinophilia of 80 per high power field (HPF) and EoE was diagnosed. A separate biopsy sent for viral culture was positive for HSV but he improved on PPI without acyclovir. His oral dermatitis resolved after beginning a food elimination diet. Repeat endoscopy three months after presentation showed abnormal esophageal mucosa with linear furrowing and 50 eosinophils/HPF. Swallowed viscous budesonide was initiated. The second case was a 16 year old female with history of asthma, remote failure to thrive, and 3 prior episodes of pneumonia, who reported 5 days of fever, odynophagia, vomiting, and 8 pound weight loss. EGD revealed severe and diffuse esophageal ulcerations. However, biopsy only showed 5 eosinophils/HPF, therefore a diagnosis of EoE could not be made. No viral organisms were identified and HSV stain was negative, but tissue HSV PCR was positive. She was discharged on omeprazole and sucralfate. Patient was presumed immunocompetent but was started on acyclovir given symptom severity and need for hospitalization. Infectious disease work-up showed normal immune function but elevated IgE. Repeat EGD performed two months after presentation revealed esophageal sloughing, linear furrowing, and 45 eosinophils/HPF on biopsy. Swallowed viscous budesonide was

initiated. Discussion: The relationship between EoE and HSV esophagitis is reported sporadically with an unclear mechanism of relationship. These two cases demonstrate concurrent presentations of EoE and HSV esophagitis. In the first case, EoE was diagnosed on initial endoscopy whereas in the second, only follow up endoscopy revealed EoE, which in retrospect was likely present but diagnosis was not possible given near complete loss of intact mucosa from severe ulcerations. Previous literature posits theories including loss of barrier function secondary to HSV infection and resulting immune system hyper-responsiveness (HSV leading to EoE), versus susceptibility of tissue to HSV due to atopy and T-helper cell dysregulation (EoE leading to HSV). Histopathology may not always diagnose HSV esophagitis, however additional biopsies for viral culture and/or HSV PCR can help. Follow up endoscopy to evaluate for EoE should be considered in immunocompetent patients who present with HSV esophagitis but do not meet diagnostic criteria for EoE on initial histology.

INFLAMMATORY BOWEL DISEASE

56 *RARE GENITAL MANIFESTATIONS OF CROHN'S DISEASE; A CASE SERIES.* A. Russell, M.R. Nicholson, Pediatrics, Vanderbilt University, Nashville, Tennessee, UNITED STATES | H. Correa, Pathology, Vanderbilt University, Nashville, Tennessee, UNITED STATES | S. Tanaka, Urology, Vanderbilt University, Nashville, Tennessee, UNITED STATES | U. Rani, Vanderbilt University, Nashville, Tennessee, UNITED STATES |.

Although cutaneous manifestations are the most common extra-intestinal manifestation of inflammatory bowel disease, metastatic Crohn's disease (MCD) is rare. MCD is defined as the presence of non-caseating granulomatous inflammation and perivascular infiltrate in the cutaneous tissue that is non-contiguous to the gastrointestinal tract. MCD very rarely involves the genitourinary tract in children. When it does, it can present as external genitalia swelling, erythema, plaques or ulcerations. Here we present two pediatric cases of MCD involving the genitourinary tract. The first case was a 7-year-old female, previously diagnosed with Crohn's disease (CD) of the large bowel at age 3, who presented with a 2 cm, non-tender, non-pruritic mass at the clitoral hood. Her magnetic resonance imaging (MRI) showed asymmetric enhancement of the labia minora without involvement of clitoral structure. Pathology of the mass showed diffuse granulomatous inflammation and chronic perivascular inflammation with lymphocytic infiltrate confirming the diagnosis of MCD. The second patient was an 11-year-old male, previously diagnosed with CD and psoriatic arthritis at the age of 7, who presented with penile swelling. MRI showed mild scrotal and penile edema and no associated mass or adenopathy. Pathology of penile skin revealed many non-caseating granulomas with multinucleated giant cells throughout the dermis, consistent with MCD. These cases highlight the fact that despite the rarity of genitourinary Crohn's disease in pediatric patients, it should be considered in the differential diagnosis of genitourinary lesions.

58 *COINCIDENTAL EOSINOPHILIC ESOPHAGITIS AND CROHN'S DISEASE IN 3 PEDIATRIC PATIENTS.* A. Fifi, A. LANGSHAW, T. MILLER, Pediatric Gastroenterology, University of Miami, Miami, Florida, UNITED STATES |.

Introduction: Eosinophilic esophagitis (EoE) is an inflammatory condition of the esophagus with a poorly understood etiology that may be primarily associated with T-helper lymphocyte (Th) type 2 response. A number of basic studies support a role for atopy in the pathogenesis of EoE. The incidence of EoE is increasing in recent times due, in part, to increased awareness of this condition. Crohn's disease (CD) is an autoimmune disease characterized by chronic inflammation of the gastrointestinal (GI) tract that can also affect the esophagus. It is associated with Th 1 and Th 17 cytokine profile. EoE is reported with increased incidence in patients with other autoimmune diseases, however there is limited literature on the coexistence of EoE and CD in both children and adults. Case presentation: We present a case series of 3 patients who developed both EoE and CD. All patients were male from ages of 9-15 years at initial diagnosis. Two of the three developed EoE first, then CD two years later. The other developed CD first, then EoE three years later. Two patients had positive family history of autoimmune disease and two had a personal history of atopy. All three patients had positive radioallergen sorbent test (RAST) to multiple food allergens and positive anti-saccharomyces cerevisiae antibodies (ASCA). Baseline histology on all patients showed inflammation consistent with their primary diagnosis only. However, follow-up histology 2-3 years later revealed further changes that prompted evaluations for the secondary diagnosis (eg.

patient with EoE developed focal, active gastritis, duodenitis, and lymphoplasmocytic infiltrate of colon; patient with CD developed esophagitis with >50 eos per HPF). Treatments followed conventional recommendations with elimination diet and swallowed steroids for EoE and immunosuppressants (methotrexate and infliximab) for CD. Discussion: The coexistence of EoE and CD may provide new insight into the complex interactions of the immune system in the pathogenesis of both conditions. Interestingly these patients tested positive for RAST and ASCA and had a strong history of both atopy and family history of autoimmune disease, suggesting a possible overlapping interaction between allergy and autoimmunity in these conditions. It is important to note that when there is involvement of sites other than just the esophagus in patients with EoE, that the possibility of a coexisting condition like CD should be explored. Finally these patients had refractory disease to therapies directed against their primary disease. Additional therapies with swallowed steroids for EoE and immune-modulators and/or biologics for treatment of CD resulted in improvement in symptomatology, including growth failure, GERD and abdominal pain; only when both conditions were treated adequately.

62 *PERIANAL FISTULIZING CROHN'S DISEASE PRESENTING AFTER SURGERY ON A PERIANAL LESION.* A.A. Singer, J. Adler, Pediatric Gastroenterology, University of Michigan, Ann Arbor, Michigan, UNITED STATES | S. Gadepalli, S. Eder, J. Adler, Child Health Evaluation and Research (CHEAR) Unit, University of Michigan, Ann Arbor, Michigan, UNITED STATES | S. Gadepalli, Department of Surgery, Pediatric Surgery Section, University of Michigan, Ann Arbor, Michigan, UNITED STATES |.

Background: Perianal skin lesions can be an early presenting sign of Crohn's disease (CD). Surgical intervention on these lesions may increase the risk of fistula development and lead to worse outcomes in those who have a perianal fistula present. Some patients have undergone procedures on what was thought to be a simple perianal lesion, only to reveal a fistula leading to the diagnosis of CD. However, these cases are rare within the literature and no reports exist within pediatrics. This case series examines patients with perianal fistulas in whom CD was identified after surgical intervention for perianal skin lesions. This study attempts to determine how frequently patients with perianal CD undergo procedures prior to CD diagnosis. Methods: We conducted an IRB-approved retrospective review of the electronic medical record of all patients with CD followed in the Pediatric Gastroenterology division at the University of Michigan between 1/2005-2/2014. Inclusion required perianal fistulizing CD and a surgical procedure on a perianal lesion prior to diagnosis. Patients with prior history of inflammatory bowel disease, development of fistula later in the disease course, or perianal lesion without fistula were excluded. Results: Of the 318 patients with CD cared for during the study timeframe, 41 (13%) had perianal fistula at time of CD diagnosis. Eight of these patients (19.5%) underwent surgical intervention on perianal lesions prior to diagnosis with CD. The time from discovery of perianal lesion to diagnosis of CD ranged from 3-58 months. Patients presented with perianal abscess (4/8), skin tags alone (3/8), or external hemorrhoids (1/8). All patients had other symptoms that, in retrospect, may have been attributed to CD. 3 patients had constipation, to which their perianal lesion was attributed. Of the patients with documentation available, 43% (3/7) had weight loss or growth failure present at the time of either identification of or surgical procedure on their perianal lesion. Two patients were not adherent with recommendations upon perianal lesion identification, possibly delaying the time to CD diagnosis. The patients underwent a range of surgical procedures, with some undergoing multiple procedures before CD diagnosis. Once diagnosed, 88% (7/8) of patients were treated with infliximab. 7 out of 8 patients had confirmed healing of their fistula with medical therapy at the time of analysis. The time required for fistula healing, defined as resolution on MR enterography (MRE) or physical exam if a repeat MRE was not obtained, was 8 to 34 months. Conclusions: We identified patients with CD and perianal fistula at diagnosis who had surgical procedures on perianal lesions prior to CD diagnosis. The finding of perianal lesions should raise suspicion for CD, especially if other symptoms such as growth failure or weight loss are present. Earlier identification of CD may facilitate prompt medical therapy and avoid unnecessary surgical procedures, which may increase the risk of fistula formation. Future studies are needed to understand the risk factors for perianal fistulas and the role surgery may play in both their development and healing.

64 *INFLAMMATORY BOWEL DISEASE IN A 14-YEAR-OLD MALE WITH HISTORY OF CELIAC DISEASE.* B. Alabd Alrazzak, P. Patel, F. Navarro, Gastroenterology, University of Texas- Houston, Houston, Texas, UNITED STATES].

Celiac disease is a permanent gluten sensitivity disorder presents as an autoimmune mediated enteropathy. Although a strong relationship between celiac disease and other autoimmune disorders such as diabetes mellitus type 1, autoimmune thyroiditis is well established; however, studies showed different results regarding association between celiac disease and inflammatory bowel diseases in adults. In this case, we present a case of ulcerative colitis in a 14 year old male with a history of celiac disease. To our knowledge only few similar cases were reported in children. Case: A 14 year-old male presented to the clinic with hematochezia, the patient has history of asthma and celiac disease diagnosed at age of 12; at that time upper endoscopy showed blunting of the duodenal villi with increased intraepithelial lymphocytes. Tissue transglutaminase as well as anti-endomysial antibody were significantly elevated at time of diagnoses (40 and more than 100 respectively). The patient did well after 6 months on gluten free diet and a follow up labs as well as endoscopy were normal. Past medical, surgical and family history were not contributory. Physical exam showed pale conjunctiva and mild tachycardia otherwise it was unremarkable. CBC revealed hemoglobin of 7, albumin of 3.1, stool calprotectin of 2000, sed rate of 32 and CRP was 16. Colonoscopy showed pancolitis. Biopsies revealed chronic active colitis without granuloma but with terminal ileitis. The patient was treated with methylprednisolone to induce remission and is currently doing well. Conclusion: Although the relationship between celiac disease and Inflammatory bowel disease is not well understood yet; however physician should have a high index of suspicion for inflammatory bowel disease in patients with Celiac disease when they develop new symptoms or do not improve on gluten free diet.

66 *INDUCTION WITH HIGH DOSE INFlixIMAB IN PEDIATRIC SEVERE ULCERATIVE COLITIS; A CASE SERIES.* A. Gjokopulli, Z. Molle Rios, Gastroenterology, Nemours Al duPont Hospital for Children, Wilmington, Delaware, UNITED STATES].

INTRODUCTION Infliximab (IFX) is a monoclonal antibody against tumor necrosis factor alpha (TNF α) used to induce and maintain remission in pediatric ulcerative colitis (UC). Although higher than 5mg/kg dosing has not been studied in children, in clinical practice higher doses are used. Severe UC is associated with high circulating TNF levels and higher fecal concentrations of IFX likely leading to more rapid drug clearance. Such patients may benefit from higher or more frequent IFX dosing. This case series reviews 4 patients with severe UC induced with IFX at 10 mg/kg/dose. Case 1: 14-year-old female diagnosed with severe UC (pancolitis) and induced 12 days later with IFX (10 mg/kg) after losing initial response to steroids. At week 54 of maintenance therapy with 10 mg/kg/dose Q8w, she continued in remission with appropriate IFX levels and no antibodies, so IFX was weaned to 5 mg/kg/dose Q8w and she continues to do well. Case 2: 17-year-old male diagnosed with UC (proctitis) maintained on mesalamine. He relapsed 7 months later with severe pancolitis. He received IFX (10 mg/kg) and was weaned off steroids. Currently, after completion of induction, he is clinically under remission and awaiting his first maintenance dose of IFX after 8 weeks. Case 3: 15-year-old male with HLA-B27 spondyloarthritis and UC (proctitis) maintained on mesalamine. 1 year later, his disease relapsed and extended to pancolitis. He ultimately failed steroids and adalimumab (40mg Qweek). He was hospitalized for IV steroids, TPN, blood transfusion and pain control. High dose IFX induction was elected as rescue but failed. The severity of his disease warranted a subtotal colectomy with end ileostomy. He has recovered well and is planned to have primary anastomosis in the near future. Case 4: 15-year-old female diagnosed with UC (pancolitis) requiring blood transfusion. She initially responded to IV but later failed oral steroids. High dose IFX was given for remission induction. She slowly responded to an accelerated induction dosing. An intercurrent *C. difficile* colitis after completion of induction complicated her course but is currently doing well. RESULTS 2 patients were de novo presentations and 2 were relapses. 2 cases responded well to standard 0-2-6 week induction, 1 required accelerated induction (0-1-5 weeks) and 1 failed. None were on immunomodulator and none developed drug antibodies (ATI). CONCLUSION 3 of the 4 patients with severe UC responded to induction with high dose IFX at 10 mg/kg/dose. High and accelerated dosing of IFX was well tolerated and should be considered in severe cases of UC. Drug monitoring may aid in determining therapy response.

67 *EFFICACY OF COMBINATION ANTIBIOTIC THERAPY FOR REFRACTORY IBD AT CHOP: A SINGLE CENTER CASE SERIES.* B. Godwin, S. Demuynck, E. Maxwell, R. Baldassano, A. Grossman, J. Kelsen, L. Albenberg, Pediatric Gastroenterology, Hepatology and Nutrition, The Children's Hospital of Philadelphia, Haverford, Pennsylvania, UNITED STATES].

Background: The pathogenesis of Inflammatory Bowel Disease (IBD) is thought to involve an inappropriate immune response to gut microbes in genetically susceptible individuals. Current therapies for IBD are focused on immunosuppression, but early data suggests that combination oral antibiotics may be an effective strategy in refractory Ulcerative Colitis (UC), including a recent paper by Turner and colleagues, which demonstrated that therapy with 3 or 4 oral antibiotics improved rates of remission in refractory UC. We sought to determine the efficacy of a similar regimen in our patients with refractory IBD. Here we report the first North American case series describing the use of combination antibiotic therapy in pediatric Crohn's disease (CD) and UC. Methods: We performed a retrospective review of our experience treating refractory IBD with combination antibiotics. Information collected included demographics, disease characteristics, immunotherapy history, indication for antibiotic therapy, and type, dosage, and duration of antibiotics prescribed. Eligible patients were ages 3-21 and prescribed three or more antibiotics concomitantly for the treatment of IBD. Clinical outcomes were evaluated based on changes in the Pediatric Crohn's Disease Activity Index (PCDAI) or the Pediatric Ulcerative Colitis Activity Index (PUCAI) at the time of initiation of antibiotic therapy and at multiple subsequent time points. Disease morbidity outcomes were also measured, including plan for surgery and escalation to experimental therapy. The incidence of adverse events during and following antibiotic therapy was also assessed. Results: Of the enrolled patients (n=14), four had CD (29%) and the remainder had UC or indeterminate colitis (IBDU). In most subjects, the indication for antibiotic therapy was disease refractory to conventional therapy. Our data was most complete for PUCAI scores at baseline and two weeks. The mean decrease in PUCAI was 30 points, which was significant ($p=0.014$). There was also a decrease in PCDAI scores, though this was not powered to show significance. The most common antibiotic regimen was metronidazole, amoxicillin, and ciprofloxacin, and the average length of treatment was 29.5 days. This regimen prevented four patients from having surgery and five from being escalated to experimental therapy. There were four documented adverse reactions that occurred during the course of, or shortly following, therapy. All were mild and only two were likely related to the regimen; all resolved without sequelae. Conclusions: Combination antibiotic therapy for refractory IBD may improve disease activity and may circumvent the need for surgery or escalation to experimental therapy in some patients. Adverse events were rare.

69 *HENOCH-SCHONLEIN WITHOUT PURPURA: A CASE REPORT AND REVIEW LITERATURE.* C. Jarasvaraparn, Pediatrics, University of South Alabama, Mobile, Alabama, UNITED STATES | C. Lertudomphonwanit, K. Pirojsakul, S. Treepongkaruna, Pediatrics, Ramathibodi Hospital, Mahidol University, Bangkok, THAILAND | S. Worawichawong, N. Angkathunyakul, Pathology, Ramathibodi Hospital, Mahidol University, Bangkok, THAILAND].

Henoch-Schönlein purpura (HSP) is a multi-organ vasculitis involving skin, joints, gastrointestinal tract, and kidneys. We report a 5-year-old boy who presented with colicky abdominal pain, vomiting, bloody diarrhea and protein-losing enteropathy for 15 days. Esophagogastroduodenoscopy showed swelling and erythematous mucosa with hemorrhagic spots at duodenal bulb to the third part of duodenum. Investigations for infectious enteritis and other vasculitis disorders were negative. His abdominal pain and bloody diarrhea improved after corticosteroid therapy. However, 2 weeks later, he developed nephrotic-range proteinuria, thus kidney biopsy was performed. Renal histology was consistent with IgA nephropathy, supporting the diagnosis of HSP. HSP should be considered in children with abdominal pain and bloody diarrhea despite of absence of purpura.

71 *DON'T BE FOOLED: PRIMARY ADDISON'S DISEASE PRESENTING AS CROHN'S DISEASE IN AN ADOLESCENT PATIENT.* A. Pierog, M. Glassman, Pediatric Gastroenterology, New York Medical College, Valhalla, New York, UNITED STATES].

A 16-year-old male of Native American descent presented with 1 month of abdominal pain and a weight loss of 30 pounds. He reported no fevers, no vomiting, no diarrhea and a decreased energy level. Initial laboratory

evaluation including comprehensive metabolic panel, thyroid testing, C-reactive protein, sedimentation rate, serum cortisol, aldosterone and celiac testing was negative. He then developed a fever to 102, vomiting and diarrhea and was admitted for IV hydration. An upper endoscopy and colonoscopy were performed and were grossly normal. The pathology report revealed some chronic inflammation in the esophagus consistent with reflux. The small and large intestinal biopsies were normal. As the clinical suspicion for Crohn's disease was still quite high, an MRE was performed and revealed a degree of mucosal thickening in the distal terminal ileum. Because of this finding and his persistent anorexia, diarrhea and abdominal pains, the patient was started on a 5-aminosalicylate. His fever resolved, his abdominal pains and appetite improved and he was discharged home. Once home, he was unable to regain any weight despite increasing calories in his diet. He was then placed on a trial of oral prednisone. On the medication, his diarrhea resolved and his appetite and energy level improved greatly. A slow wean of the steroids was initiated while the patient remained on the 5-aminosalicylate. The patient developed lethargy with this wean, so steroids were resumed. MRE was repeated and this time it was normal without any ileal mucosal thickening. Again steroids were weaned, this time much more slowly, and he seemed to do well until they were completed off, when he developed poor circulation in his hands and feet, depression, and complete lack of energy. He was seen by the PMD at this time and started on sertraline, which seemed to help him to feel better and get back to school. 1 month later, he presented to the ER with vomiting, severe abdominal pain and diarrhea. He was found to be tachycardic, hypotensive and hyponatremic. The patient required multiple normal saline boluses to maintain his blood pressure and control his heart rate and he was admitted for suspected Addison's disease. Emergent CT scan of the abdomen and pelvis was unremarkable. After evaluation by endocrinology and an adrenocorticotrophic hormone stimulation test, Addison's disease was confirmed and he was started on fludrocortisone and hydrocortisone and his gastrointestinal complaints completely resolved. This case highlights the importance of thinking of Addison's disease in pediatrics when tissue pathology is not diagnostic of Crohn's disease. Addison's disease can present with anorexia, weight loss, fatigue, vomiting, diarrhea, depression and respond to steroids, much like Crohn's disease does. This patient did have a fever, which does not typically occur with Addison's disease, which could have been related to a viral infection or another acute inflammatory process in the intestine, which would explain the thickening seen in the ileum on the initial MRE that subsequently resolved.

72 *TAKAYASU'S ARTERITIS IN A PATIENT WITH CROHN'S DISEASE TREATED WITH INFLIXIMAB.* A. Singh, R. Mittal, C.M. Marshall, Pediatric Gastroenterology, University of Oklahoma Health Sciences Center, Oklahoma City, Oklahoma, UNITED STATES].

Introduction: Takayasu's arteritis (TA) and Crohn's Disease are both chronic granulomatous inflammatory diseases. An association has been reported in several patients, but optimal treatment of these patients is unclear. We present a case of TA in a patient with Crohn's ileocolitis, who had been treated with infliximab. Case Presentation: 14 year old female with a two year history of Crohn's ileocolitis, treated with infliximab 5mg/kg every 8 weeks, was admitted with right lower quadrant abdominal pain, poor appetite, bloody diarrhea, vomiting, and weight loss. She received infliximab without improvement. Physical exam was pertinent for abdominal tenderness, an audible bruit in the periumbilical region and diminished femoral and lower extremity pulses. Hemoglobin was low (10.5), CMP normal, ESR (>100) and CRP (136.8) were elevated. A CT scan of the abdomen revealed thickening of terminal ileum, sigmoid colon, rectum and a soft tissue density around the superior mesenteric artery (SMA) and aorta causing narrowing. She was started on IV solumedrol. Diarrhea and pain improved, CRP decreased but ESR continued to be >100. She was transitioned to oral steroids and azathioprine was added. Due to concerns of vascular wall inflammation on CT, a duplex ultrasound was obtained that revealed stenosis of the distal aorta and SMA. She was diagnosed with Takayasu's arteritis. After 2 months, when unable to wean steroids due to recurrent diarrhea and abdominal pain, infliximab was increased to 10 mg/kg every 4 weeks. Azathioprine was continued and nine months after the diagnosis of TA, she was finally weaned off steroids. At that point, MRI/MRA showed normal caliber of aorta with patent branches and patent SMA with only proximal segment of mild to moderate stenosis, subsequently regaining normal caliber. On physical exam, abdominal bruit disappeared and femoral and lower extremity pulses were normally palpable. On continued treatment with infliximab and azathioprine, TA and

CD both have been in remission for 2 years. Discussion: The anti-TNF α monoclonal antibody infliximab, is an effective therapy for both TA and Crohn's disease. However, vasculitis can develop secondary to infliximab; most commonly presenting as cutaneous lesions, or involving small to medium sized vessels. Our patient was successfully treated for both Crohn's disease and TA with the combination of prednisone, infliximab, and azathioprine; with maintenance of remission for 2 years without steroids. Takayasu's arteritis may be an extra intestinal manifestation of inflammatory bowel disease. Early diagnosis and treatment is important for patients with TA to prevent renal hypertension, aortic regurgitation, congestive heart failure, and cerebrovascular accident.

76 *TRISOMY 21 AND PERIANAL CROHN'S DISEASE: A CASE SERIES.* E. Bishop, T. Jester, Pediatrics, University of Alabama at Birmingham, Homewood, Alabama, UNITED STATES|.

Introduction: Perianal disease can be a cause of significant morbidity in patients with Crohn's disease. It has not been previously shown that there is an association with Trisomy 21 and Crohn's disease; more specifically cases describing perianal disease in this patient population. Case descriptions: Patient 1 was a 13 year old female at presentation with a 2 year history of abdominal pain, weight loss and intermittent episodes of emesis and diarrhea. On exam she was found to have a perianal abscess as well as a large anal tag. Her upper endoscopy and colonoscopy showed gross pancolitis and histology confirmed these findings in addition to granulomas. Remission was induced with prednisone and Infliximab and she continues on Infliximab. Patient 2 was 9 years of age when she presented with hematochezia and abdominal pain. Her first upper and lower endoscopies demonstrated pancolitis. She was lost to follow-up with parental non-adherence to therapy. Two years later she again presented with worsening symptoms and repeat endoscopy revealed an anal stricture in addition to distal colitis. Her colitis is now improved on every 4 week Infliximab infusions, but her anal stricture still requires regular dilations by pediatric general surgery. Discussion: There are no case reports in the medical literature of patients with Trisomy 21 and perianal Crohn's disease. These two cases may be isolated events or could represent a genetic association. Further sharing of patient data in pediatric inflammatory bowel disease networks with collaboration of other centers may help determine if there is an increased risk of perianal Crohn's disease in patients with Trisomy 21.

78 *INCIDENTAL CARCINOID DURING BOWEL RESECTION FOR CROHN DISEASE: COMPLICATION OF THERAPY OR FORTUNATE DISCOVERY?.* D.M. Dykes, S.A. Saeed, Pediatric Gastroenterology, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, UNITED STATES|A. Munaco, J. Frischer, Pediatric Surgery, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, UNITED STATES|J. Hata, Pathology, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, UNITED STATES|.

Introduction: Carcinoid tumors are slow-growing neuroendocrine tumors (NETs) which are rare in adults and less common in pediatrics with estimates of about 2.8 cases per million under age 30. We are reporting the experience of a previously healthy 15 year old male with newly diagnosed Crohn disease who was found to have an incidental appendiceal carcinoid tumor at the time of surgical resection of his stricturing disease. Case. The patient was diagnosed with Crohn disease (CD) after a 6 month history of nausea and weight loss. The extent of bowel involvement was ileocolonic as well as mid small bowel (mid-jejunal stricture) based on radiographic and endoscopic evaluation. He responded briefly to prednisone and initiation of an anti-tumor necrosis factor alpha agent (anti-TNF) but symptoms relapsed as steroids weaned. Due to persistent symptoms concerning for partial obstruction and ongoing inflammation in the setting of appropriate anti-TNF level, he underwent a limited ileocecal and jejunal resection. Pathology was notable for inflammation consistent with Crohn disease in both resected areas of intestine (total of 13 cm) but was noted to have an incidental carcinoid tumor in the distal appendix, 2 mm in size with no serosal compromise and negative margins. Clinically, he has done well with quiescent disease following surgery and has had normalization of blood and stool markers of inflammation with documented endoscopic mucosal healing six months after surgery. He has not had any further symptoms of carcinoid tumor and has had negative urine HIAA screening at diagnosis and 6 months following surgery. Discussion: Carcinoid tumors have been described in up to 0.2% of children undergoing appendectomy without IBD, but the incidence of incidental carcinoid tumors in IBD is not described. Cases of carcinoid tumors and other

NETs have been described in an older adolescent and in adult patients with IBD, although the time to identification of these tumors may be up to 10 years or more from diagnosis. Most carcinoids are found incidentally at the time of an IBD related surgery but do tend to occur in areas with active disease. One case of carcinoid-associated death has been reported in which combination anti-TNF and immunomodulator therapy was felt to play a possible role, but in general this association has been difficult to prove due to the low number of cases reported. Conclusion: Most carcinoid tumors in adult IBD are incidental findings at the time of surgery, and it is unclear whether these tumors are related to primary inflammation or as a consequence of a particular medical therapy. With growing concern over safety of treatment regimens for IBD, particularly in younger patients, this case represents an opportunity to review potential risks of medications compared with the risk of active disease. While collaboration through learning health networks and multi-center patient registries may provide better ability to characterize the incidence and natural history of these tumors over time, cases such as this warrant close scrutiny to ensure optimal safety of our patients.

79 *SARCOIDOSIS OF THE BOWEL IN A CHILD*. H.W. Hamandi, S. Ogholikhan, C. Cuffari, Gastroenterology & Nutrition, Johns Hopkins University, Baltimore, Maryland, UNITED STATES|L. Voltaggio, Pathology, Johns Hopkins University, Baltimore, Maryland, UNITED STATES|.

9 year old African American male who presented with 2 weeks of worsening abdominal pain accompanied by loose stools and incontinence. Initial laboratory evaluation did not show evidence of anemia or hypoalbuminemia, but inflammatory markers were elevated: CRP 3.3 mg/DL, ESR 36 mm/h. Infectious stool studies were negative. Imaging with CT scan abdomen and pelvis pursued first and notable for bowel wall thickening of the ascending colon. The pediatric gastroenterology team postulated that inflammatory bowel disease was the most probable differential diagnosis. The patient ultimately underwent a colonoscopy, which revealed increased nodularity in the terminal ileum and severe inflammation in the ascending colon as evidenced by bowel wall edema, loss of vascular markings, erythema, and ulcerations. The pathology was remarkable for terminal ileum with well-formed compact granulomas in superficial lamina propria, ascending to transverse colon with well-formed compact granulomas in lamina propria with associated ulceration, descending colon to rectum with non-diagnostic findings. There was no significant architectural distortion and no activity to suggest inflammatory bowel disease. The well-formed granulomas in lamina propria are consistent with the diagnosis of sarcoidosis. Subsequent MRE also showed bowel wall thickening and contrast enhancement extending from the distal ileum to the transverse colon. Upon further investigation, the patient revealed a remote history of vision problems that were treated with an ophthalmic corticosteroid ointment; outside records from an ophthalmologist described uveitis and granulomatous keratic precipitates consistent with sarcoidosis. Of note, chest X-ray was negative for hilar adenopathy and angiotensin converting enzyme activity level was within normal range. Induction therapy consisted of high dose steroids with dramatic improvement in abdominal pain and stool quality. After reviewing the adult literature on treating sarcoidosis of the bowel, the patient was started on 6-mercaptopurine for maintenance therapy. Repeat MRE obtained 4 months after diagnosis showed complete resolution of bowel wall thickening. Repeat colonoscopy performed 9 months after diagnosis revealed mucosal healing and no granulomas on surgical pathology. Discussion: The incidence and prevalence of sarcoidosis of the bowel is extremely rare, particularly among children. The presentation can mimic ulcerative colitis and Crohn's disease with abdominal pain and loose stools, elevation in inflammatory markers, bowel wall thickening on various imaging modalities, and abnormal mucosa on colonoscopy. The surgical pathology is essential to making the diagnosis. Interestingly, this clinical vignette highlights that sarcoidosis can initially manifest in the gastrointestinal tract without evidence of pulmonary involvement. Additionally, this patient was successfully treated with steroids for induction and 6-mercaptopurine for maintenance with evidence of mucosal healing. Anti-TNF monoclonal antibody agents such as infliximab were considered, but the literature reported mixed results in the treatment of sarcoidosis.

83 *DIVERTING ILEOSTOMY FOR THE TREATMENT OF SEVERE, REFRACTORY, PEDIATRIC INFLAMMATORY BOWEL DISEASE*. E.C. Maxwell, R. Baldassano, M.A. Conrad, N. Dawany, P. Mamula, P. Mattei, L. Albenberg, J. Kelsen, GI, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES|.

Background: Diverting ileostomy is a potentially temporizing intervention in patients with inflammatory bowel disease (IBD) refractory to medical therapy. Though traditionally performed for perianal disease, at the Children's Hospital of Philadelphia (CHOP), ileal diversion has also been utilized in the management of severe colonic IBD. In addition to potentially inducing clinical remission, the procedure provides an opportunity for the appropriate diagnosis to be made in patients with indeterminate colitis (IC), thus avoiding the complications associated with ileal pouch-anal anastomosis (IPAA) in patients who in fact have Crohn disease (CD). There is a paucity of literature in the pediatric population investigating the efficacy or risks of this procedure. Aims: To evaluate whether diverting ileostomy improves disease outcomes, as defined by decrease in corticosteroid use and number of blood transfusions, and improvement in hemoglobin and growth velocity. Secondary aim was to determine whether diversion led to a change or clarification in IBD diagnosis, potentially altering the ultimate therapeutic course. Methods: We performed a retrospective study of pediatric patients who underwent diverting ileostomy at CHOP from January 1, 2000 to November 1, 2014 for management of severe, refractory, colonic IBD. Patients were excluded if they had diverting ileostomy performed for any other indication. Demographic and clinical data were collected at 1 year prior to ileostomy, and 6 months, 1 year, and 3 years post surgery. Results: Twenty four patients underwent diverting ileostomy, 15 (63%) of whom were male. Twelve patients (50%) were diagnosed with IBD at age 5 years or younger. Eleven patients (46%) had an initial diagnosis of CD, and 13 (54%) had IC. All patients failed therapy with biologic agents prior to diversion. Eighteen of 24 patients (75%) had chronic steroid exposure in the 1 year prior to diversion, versus 6 of 18 (33%) patients in the 1 year post diversion. Average height velocity prior to diversion was 2.9 cm/year, compared to 5.6 post diversion ($p=0.034$). Average weight velocity prior to diversion was -2.1 kg/year, versus 6.2 post diversion ($p=0.008$). The number of blood transfusions required in the 1 year prior to diversion was 2.1, compared to 1.1 in the 1 year post diversion ($p=0.047$). There was no statistical improvement in the average hemoglobin at 1 year post diversion. There were 11 patients with complete follow up data available at 3 years. Of those, 3 remained diverted, 4 had undergone colectomy, and 4 were successfully reanastomosed. At the end of the study, 7 patients (29%) had a change in diagnosis. There were no significant complications or deaths. Conclusions: In a subset of pediatric patients with refractory IBD, diverting ileostomy can be a successful intervention to induce clinical stability as measured by improvement in height and weight velocity, and decreased blood transfusion requirement. Importantly, diversion is steroid-sparing, and allows additional time for the correct diagnosis to be made. Prospective data is being collected to further define these outcomes.

PANCREAS/CELIAC/MALABSORPTION

88 *PRIMARY INTESTINAL LYMPHANGIECTASIA DIAGNOSED BY CAPSULE ENDOSCOPY*. H. Aaron, K. Chen, Pediatric Gastroenterology, Drexel University College of Medicine, Philadelphia, Pennsylvania, UNITED STATES |.

Background: Primary Intestinal Lymphangiectasia (PIL) is characterized by dilated intestinal lacteals resulting in lymph leakage into the bowel lumen and leading to protein-losing enteropathy. It generally presents in children under 3 years of age with bilateral pitting edema and anasarca. Diagnosis is typically made by endoscopic observation of intestinal lymphangiectasia with visualization of dilated lacteals on corresponding histology. We present a case of PIL diagnosed by capsule endoscopy in a 3-year-old boy with two prior normal upper endoscopies with unremarkable histology. Case: A 3-year-old boy presented with 5 days of progressive bilateral lower extremity swelling and abdominal distention. He had been ill one week prior with viral gastroenteritis. He did not have history of constitutional symptoms, chronic diarrhea or constipation. He had been hospitalized twice for similar symptoms. Previous work-up included upper endoscopy with normal biopsies, and he had spontaneous resolution of symptoms. He had no other significant medical or surgical history. Labs on this presentation were significant for serum total protein 3.8g/dL (6.0-8.0), albumin 2.3g/dL (3.7-5.5), normal serum transaminases and coagulation factors, urinalysis was normal with no evidence of proteinuria, total 25-OH vitamin D <4ng/mL (30-100), IgG 141mg/dL (507-1305), and fecal alpha-1-antitrypsin >1.13mg/g (0.0-0.5). Abdominal ultrasound demonstrated ascites and normal liver anatomy. Due to his anasarca, albumin infusion and IV furosemide were given, with resolution of edema within 2 days. Repeat upper endoscopy with biopsy was normal. Capsule endoscopy then revealed patchy areas of lymphangiectasia throughout the small bowel, and he was diagnosed with PIL. In

consultation with the pediatric dietitian, we placed him on a low-fat, high-protein diet using age-appropriate table foods and supplementation with 16 ounces daily of Portagen formula (87% medium chain triglyceride [MCT], Mead Johnson Nutrition). We also recommended water-soluble formulation of fat-soluble vitamins. Conclusion: PIL can cause protein-losing enteropathy that is a challenge to diagnose. In this case, our patient's clinical picture was complicated by the presence of waxing and waning symptoms that improved without intervention, as well as previous workup that included two normal endoscopic studies. Upper endoscopy often does not provide sufficient visualization. As was the case in our patient, capsule endoscopy can be the best modality to make a diagnosis. Lifelong dietary management with a low-fat, high-protein diet with MCT supplementation is the mainstay of treatment. By using MCT oils, therapy aims to reduce engorgement of intestinal lymphatics and prevent rupture. In cases when this diet is not tolerated, enteral or parenteral nutrition may be considered. Octreotide therapy and a few case reports of antiplasmin therapy are other options. However, pharmacotherapy should be reserved for cases in which dietary management is ineffective.

90 *A CASE OF AUTOIMMUNE PANCREATITIS IN AN ADOLESCENT MALE.* H. Al-Atrash, A. Maheshwari, Z. Molle Rios, Gastroenterology, Nemours A I Du Pont Hospital, Wilmington, Delaware, UNITED STATES].

Background: Autoimmune pancreatitis (AIP) a more commonly recognized disease in adults; remains an extremely uncommon presentation in children. We hereby present a case of an adolescent male who presented with obstructive jaundice and later diagnosed with probable AIP type 1. Case presentation: A previously healthy 14 year old male presented with acute onset obstructive jaundice. Laboratory results were significant for total bilirubin of 8.7 mg/dl, Aspartate aminotransferase of 441 IU/L, Alanine aminotransferase of 646 IU/L, gamma-glutamyl transferase 505 IU/l, serum lipase of 446 U/L, amylase of 65 U/L and ESR of 14 mm/hr. His IgG 4 level was normal. Autoantibodies and infectious hepatitis workup negative apart from a positive ANA of 1:80. Imaging was significant for an ultrasound showing an enlarged gallbladder with a dilated common bile duct, which was later confirmed with a magnetic resonance cholangio-pancreatography that also revealed marked distension and dilation of the biliary tree with concerns for long segment stricture in the pancreatic head versus pancreatic mass. Based on those findings, he underwent an endoscopic ultrasound which revealed echogenic changes to the pancreas suggestive of inflammation or fibrosis. An Endoscopic retrograde choangiopancreatography was also done at the time with a placement of a stent and an ampullary biopsy obtained. Biopsy was significant for benign villous mucosa with acute and chronic inflammation and an immunohistochemical stains positive for an average of 12 IgG4 expressing plasma cells/HPF. These cells represented 18% of total tissue plasma cell populations. Based on these finding he was diagnosed with type 1AIP and started on steroids. His repeat labs and imaging studies confirmed resolution and he remains asymptomatic to date. Discussion: AIP is a rare disease with an incidence of 1.9% of adults with chronic pancreatitis and a mean age of 36 to 60 years at diagnosis. It is a rare presentation in our pediatric population and we are aware of only 16 cases reported in children. Patients with AIP have variable presentations with more than half experiencing obstructive jaundice caused by stenosis of the bile duct, similarly to our patient. Diagnosis of AIP pancreatitis is challenging and typically involves radiological imaging, serological markers and histology. AIP is further classified into two subtypes: Type 1 which is more commonly seen in adults and characterized by serum elevation of IgG4 (above twice the normal range), a histology showing massive infiltration with IgG4 plasma cells and association with other autoimmune diseases. Type 2 is the more commonly reported in children in whom serological markers are absent with no systemic manifestation and histology showing neutrophilic infiltration with granulocytic epithelial cells (GEL). While both subtypes respond to steroids type 1 tends to have a high relapse rate. While all most of the documented case reports of children with AIP have been of type 2 AIP, our patient's histology is highly suggestive of type1 AIP making him a unique presentation.

91 *UNUSUAL PRESENTATION OF PANCREATIC NEUROENDOCRINE TUMOR.* A. Baig, R. Nathan, K. Viswanathan, Pediatrics , Brookdale University Hospital and Medical Center, Brooklyn, New York, UNITED STATES].

Background: Neuroendocrine tumors (NETs) are distinctive tumors that can arise in almost any organ of the body. A significant proportion of these tumors are not associated with syndromes related to hormonal

hypersecretion. These patients often present with non-specific GI symptoms rather than the endocrine symptoms which often makes diagnosis a challenge. We present a case of non-functional NET who presented with abdominal pain. Case Report: A 14 year old male was admitted with one year history of self-remitting intermittent episodes of vague abdominal pain approximately every 3-4 months, lasting a day or two followed by occasional vomiting. The parents reported occasional nausea, anorexia & fatigue. They denied any history of jaundice, weight loss, diarrhea or night sweats. Complete physical examination & workup done for suspected viral syndrome by the PMD was reported normal. Abdominal sonogram was done which showed an isoechoic mass-like lesion adjacent to the body of the pancreas which was confirmed on CT as a 4.6 x 3.3 x 4.8 cm mass. Tumor markers CA19-9, Gastrin, VMA, CEA, AFP and 5HIAA were normal. CT-guided biopsy was done and immunohistochemistry was positive for Chromogranin A, Synaptophysin, CAM 5.2 clone 5D3, CD 56 N-CAM, NK marker cells, Ki-67 (10-15%); and negative for TTF1. Octreotide scan was reported negative. Subtotal pancreatectomy with splenectomy was done a month later. The pancreas and associated mass were densely adhered to the portal vein/SMV junction. The tumor was classified as an invasive low grade, well differentiated pancreatic endocrine neoplasm which had metastasized to 1 of the 17 lymph nodes. Follow up scans have been negative. Discussion: Clinical manifestations of nonfunctional NET mimic a variety of disorders and therefore do not lend themselves to identify the specific underlying tumor. Patients are often first alerted to disease because of symptoms caused by the growth of these tumors. Approximately 37-78% of the patients present with abdominal pain followed by weight loss, anorexia and nausea (1). Our patient was found to have one positive lymph node because of the initial delay in workup. These tumors should be considered as a differential after an acute process or infectious cause is ruled out. Noninvasive workup such as an ultrasound or biological markers may aid in the detection of these tumors. Because of the significant incidence of local and liver metastasis, complete excision should be done at the time of diagnosis. Conclusion: Detecting nonfunctional NET is often a challenge because of its vague presentation as compared to its functional counterpart. These tumors should be kept in the differential diagnosis once the more common differentials have been ruled out. Early workup may prevent the local and distant spread of tumor. References: 1. Well-Differentiated Pancreatic Nonfunctioning Tumors/Carcinoma Massimo Falconi a Ursula Plöckinger c Dik J. Kwekkeboom d Riccardo Manfredi b Meike Körner e Larry Kvols f Ulrich F. Pape g Jens Ricke h Peter E. Goretzki i Stefan Wildi j Thomas Steinmüller k Kjell Öberg l Jean-Yves Scoazec m and all other Frascati Consensus Conference participants. *Neuroendocrinology* 2006;84:196–211 DOI: 10.1159/000098012

92 *EXOCRINE PANCREATIC INSUFFICIENCY PRECEDING THE ONSET OF INFLAMMATORY BOWEL DISEASE*. T. Bhatia, R.D. Baker, R.H. Alkhoury, D. Moya, A. Chapa- Rodriguez, R. Abdou, S.S. Baker, University of Buffalo, Buffalo, New York, UNITED STATES |.

Inflammatory bowel diseases (IBD) including Crohn's disease (CD) and Ulcerative colitis (UC) are systemic diseases characterized by chronic intestinal inflammation. Pancreatic involvement with IBD is rare compared to other extra intestinal manifestations, such as skin, eye, liver or joints. Exocrine pancreatic insufficiency (EPI) is the most frequent manifestation of pancreatic involvement with IBD; however, it usually does not precede the symptoms of IBD. We report two cases where exocrine pancreatic insufficiency preceded the onset of inflammatory bowel disease. First case is of a 13 year old female who presented with steatorrhea for 6 months. She denied weight loss, blood in stools or abdominal pain. Pancreatic elastase was low at < 15 mcg/g (Normal >200 mcg/g). Pancreatic stimulation test was consistent with pancreatic insufficiency. CBC, IgG, IgG4 and Cystic fibrosis mutation analysis were unremarkable. MRI abdomen and pelvis with contrast revealed a small pancreas without calcifications or fatty infiltration. She was started on 500 units lipase/kg/meal of Pancrelipase and had resolution of her symptoms. A year later, she developed bright red rectal bleeding with increased frequency of bowel movements alternating between Bristol 2 and Bristol 6. Her laboratory evaluation showed leukocytosis with left shift, anemia and elevated ESR. Stool studies were negative for infection. Upper endoscopy and colonoscopy showed pancolitis with crypt abscesses with normal terminal ileum, consistent with Ulcerative colitis with phenotype E4 S1 (Paris classification). Our second patient who is a 13 year old male, also presented similarly with steatorrhea and weight loss of 30 lbs. for 9 months. He denied abdominal pain or blood in stools. Laboratory evaluations including CBC, CRP, ESR, Cystic fibrosis mutation analysis were normal. Stool samples for occult blood, lactoferrin and infection were negative.

Pancreatic elastase was low at <15 mcg/g (Normal >200 mcg/g). Diagnosis of pancreatic insufficiency was confirmed by pancreatic stimulation test. CT abdomen and pelvis with contrast revealed small pancreas but no other abnormalities. He was started on Pancrelipase 500 units lipase/kg/meal. 10 months after his initial presentation, he developed abdominal pain and rectal bleeding. EGD and colonoscopy showed active colitis in cecum, ascending colon and transverse colon with skip lesions and normal terminal ileum, consistent with the diagnosis of Crohn's disease with phenotype A1b L3 B1 G0 (Paris Classification). In conclusion, exocrine pancreatic insufficiency can precede the diagnosis of inflammatory bowel disease and should be thought of as one of the differentials in patients with EPI of unknown etiology.

93 *AN UNREPORTED SINGLE CYSTIC FIBROSIS MUTATION RESULTING IN PANCREATIC INSUFFICIENCY.* K. Bittar, N.H. Patel, D. Mehta, K. Horvath, Pediatric Gastroenterology, Orlando Health, Orlando, Florida, UNITED STATES|.

Introduction: Pancreatic insufficiency (PI) is the most common gastrointestinal manifestation of cystic fibrosis (CF), affecting about 85% of patients. In most individuals, a normal sweat chloride (SC) result is sufficient to rule out CF. However, a normal SC concentration is seen in approximately 1% of patients with CF, who have unusual genotypes. We report a child who presented with poor weight gain and PI whom SC testing was normal, yet genetic testing revealed CF variant c.2620-26 A>G. Case: The Caucasian male initially was referred to our gastroenterology clinic at 1 month of age for evaluation of fussiness and constipation. He was born at full term and passed meconium on the first day of life. He was initially switched to an amino acid based formula for suspicion for milk protein intolerance. At 12 months of age, failure to thrive was noted despite appropriate caloric intake of 90 kcal/kg/day. He also had foul smelling stools. His stool studies revealed extremely low pancreatic elastase (<15 mcg/g) and elevated qualitative fat, suggestive of malabsorption due to severe pancreatic insufficiency. He underwent an upper endoscopy, flexible sigmoidoscopy, and pancreatic stimulation test with secretin. He was challenged with whole milk prior to his scope. His histologies were normal. However, generalized decrease in his pancreatic enzymes was found. He was started on pancreatic enzyme (PE) supplementation and responded well with appropriate weight gain. A SC test was normal (7-8 mmol/l). A repeat pancreatic function test assessing both his acinar and ductal function was performed. Both his acinar cell (enzyme production) and ductal cell (bicarbonate secretion) functions were abnormal suggesting a chronic pancreatitis (CP). His genetic testing for CF mutation indicated that he is heterozygous for CF variant c2620-26 A>G. Conclusion: There is no report available in the literature on this single mutation and its clinical significance was commented as unknown. The only case reported in the past was a 2 year-old child with CF whom SC was 43-56 mmol/l, but had also mutation of IVS8-5T. Our case has CP without imaging evidence of pancreatic damage that responded well to PE replacement making it likely that this is the etiology of his pancreatic dysfunction. The possibility of additional mutations in SPINK, PRSS1 genes should also be considered.

94 *USE OF PROPRANOLOL IN A SEVEN-YEAR-OLD MALE WITH INTESTINAL LYMPHANGIECTASIA AND LYMPHEDEMA.* B. Cunningham, M. Goldman, P. Rogers, Pediatrics, Walter Reed National Military Medical Center, Bethesda, Maryland, UNITED STATES|J. Weeks, Pediatrics, Madigan Army Medical Center, Tacoma, Washington, UNITED STATES|B. Cunningham, M. Goldman, Pediatrics, Uniform Services University of the Health Sciences, Bethesda, Maryland, UNITED STATES|.

In primary intestinal lymphangiectasia small intestinal lymph drainage is impaired due to dilated enteric lymphatics. Lymphatic abnormalities may occur elsewhere in the body. The hallmark of this condition is protein-losing enteropathy (PLE) which is associated with diarrhea, nausea, vomiting, hypoalbuminemia, and peripheral edema. Serum immunoglobulins and clotting factors are also frequently decreased. Treatment options are limited. We present a patient with intestinal lymphangiectasia and congenital lymphedema successfully treated with propranolol. A 7-year-old male with a history of congenital lymphedema, intestinal lymphangiectasia, PLE, adrenal insufficiency, asthma, eczema, and sickle cell trait presented to this institution for care after relocation. He was born at term after an unremarkable prenatal course. At delivery the patient was noted to have right lower extremity and left upper extremity edema. The clinical evaluation suggested Milroy's disease but genetic markers

were negative for this condition. At age 2 years he was seen following a 5-6 month history of abdominal distention and multiple loose, nonbloody stools daily. He was found to have hypoalbuminemia, hypoproteinemia, low immunoglobulins, and a high fecal alpha-1 antitrypsin level. The patient was diagnosed with PLE and started on a high protein, low fat formula with medium chain triglyceride (MCT) oil. Histology from an upper endoscopy confirmed a diagnosis of intestinal lymphangiectasia. The patient's diarrhea seemed to improve markedly as an incidental finding when high dose corticosteroids (prednisone 45mg) was started for the treatment of eczema at age 4 years. Attempts at weaning steroids resulted in return of diarrhea, so this therapy (1.4mg/kg/day) was continued for 3 years until presentation. In spite of these interventions the patient had ongoing edema in his arms, legs, scrotum, and penis that was treated with furosemide and intermittently applied compression dressings. On steroids, height velocity significantly slowed. After a multidisciplinary evaluation, the patient's low fat with high MCT oil content/high protein diet was optimized. He was started on propranolol at 0.6mg/kg/dose twice daily and a steroid wean was instituted. After a week of treatment, the patient had lost 2kg, had dramatic improvement in his edema, and was having one formed stool daily. He continued to improve as his propranolol increased to a target of 1.7mg/kg/dose twice daily, and his steroid dose was adjusted to physiologic levels. Optimal management of patients with congenital lymphangiectasia syndrome is unclear. The current mainstay of treatment is dietary with the focus on a low-fat, high MCT, and high-protein diet. Octreotide has been suggested by case report to improve symptoms, possibly due to a reduction in splanchnic blood flow and reduced lymphatic pressure. There has been recent case report evidence that propranolol may be a treatment option for children with localized cystic lymphatic malformations and in a neonate with generalized lymphangiectasia. This is the first report of successful use of propranolol in an older child with congenital lymphedema and intestinal lymphangiectasia.

95 *ACUTE ONSET OF EPIGASTRIC ABDOMINAL PAIN IN A PREVIOUSLY HEALTHY 14 YEAR OLD FEMALE SECONDARY TO SOLID PSEUDOPAPILLARY TUMOR OF THE PANCREAS.* J.E. Dranove, Division of Pediatric Gastroenterology, Levine Children's Hospital, Charlotte, North Carolina, UNITED STATES|N. Fleishman, Department of General Pediatrics, Levine Children's Hospital, Charlotte, North Carolina, UNITED STATES|.

Introduction: Solid pseudopapillary tumors of the pancreas are exceedingly rare, representing less than 2% of all pancreatic neoplasms, and seldom seen in children. A high index of suspicion is necessary to entertain this diagnosis when patients with severe epigastric pain out of proportion to modestly elevated amylase/lipase are encountered. The symptoms can easily be mistaken for pancreatitis. Case Report: A previously healthy 14 year old African American female presented with acute onset of left upper quadrant and epigastric abdominal pain with accompanying nausea, vomiting, and dizziness. She presented 2 days after initial onset of symptoms, where labs revealed mildly elevated lipase (79), normal CBC and CMP. CT scan of abdomen and pelvis with IV and oral contrast showed no evidence consistent with stranding or inflammation typical of pancreatitis, however noted was a 2.8 x 3.2 cm mass in the tail of the pancreas read as concerning for pancreatic cyst vs. pseudocyst. Followup ultrasound showed that the mass appeared solid in nature, and not cystic. Due to lack of historical symptoms and imaging findings consistent with pancreatitis, and the severe pain out of proportion to lab findings, adult gastroenterology colleagues were consulted and an EUS with FNA biopsy was performed. Grossly the mass felt solid to the endoscopist. Pathology results from FNA were consistent with solid pseudopapillary tumor of the distal pancreas including strong nuclear and cytoplasmic staining for beta catenin, moderate nuclear staining for progesterone receptors, variable keratin staining, and negative chromogranin stain. Hepatobiliary surgery performed distal, subtotal pancreatectomy with splenectomy. The resected specimen was well demarcated and partially encapsulated with no gross invasion of the spleen or surrounding vasculature. Surgical pathology confirmed diagnosis of solid pseudopapillary tumor with staining consistent with FNA specimen. Postoperatively the patient had improvement in nausea, vomiting, and decreased abdominal pain. Patient was discharged home on POD 6 in stable condition. Discussion: Presenting symptoms of solid pseudopapillary tumors of the pancreas in pediatric patients include epigastric and upper abdominal pain that can be very similar to a variety of other conditions, including most notably pancreatitis or peptic ulcer disease. . Rarely a mass will be discovered on physical exam, and initial diagnosis is typically suspected via imaging (CT, US or MRI) when a solitary solid pancreatic mass without associated pancreatitis is encountered. Caution must be taken not to attribute this condition to pancreatitis with a

psudocyst, as a pseudocyst takes several weeks to form and should be associated with radiographic signs of interstitial pancreatitis. Once the diagnosis is suspected via imaging, confirmation via tissue sampling, most commonly via EUS, is necessary. This tumor rarely presents with constitutional symptoms or metastases. Outcomes in patients with disease isolated to pancreas and complete resection are excellent with cure rates of approximately 95%.

98 *WIRSUNGORRHAGIA FROM A PEUDOANEURYSM RUPTURE OF THE PANCREATODUODENAL ARTERY IN A 6 YEAR OLD GIRL WITH PANCREATIC PSEUDOCYST FROM RECURRENT PANCREATITIS.* R.A. Gomez Suarez, . Franciosi, J.M. Denham, R. Sandler, Pediatric Gastroenterology, Nemours Children's Hospital, Orlando, Florida, UNITED STATES|R.C. Burns, T. Westmoreland, Surgery, University of Central Florida, Orlando, Florida, UNITED STATES|R.C. Burns, T. Westmoreland, Pediatric Surgery, Nemours Children's Hospital, Orlando, Florida, UNITED STATES|R.A. Gomez Suarez, . Franciosi, J.M. Denham, R. Sandler, Pediatrics, University of Central Florida, Orlando, Florida, UNITED STATES|F. Weber, C. Johnson, Interventional radiology , Nemours Children's Hospital, Orlando, Florida, UNITED STATES|F. Weber, C. Johnson, Radiology, University of Central Florida, Orlando, Florida, UNITED STATES|.

Gastrointestinal bleeding is an infrequent complication of pancreatitis in the pediatric patient. Wirsungorrhagia (pancreatic duct hemorrhage) is a rare and potentially fatal complication from recurrent pancreatitis that generally occurs from ruptured pseudoaneurysms at either the pancreatic or splenic arteries. We present a 6 year old girl with a two year history recurrent episodes of pancreatitis and occult GI bleeding who was seen at multiple hospitals without an established etiology. No obvious source of bleeding was identified despite an extensive evaluation that included: Meckel Tc 99 scan, upper and lower endoscopy, and small bowel capsule endoscopy. Abdominal MRI and ERCP showed a 1.5 x 1.1 cm cystic lesion in the head of the pancreas. Celiac artery arteriography was performed which demonstrated a pseudoaneurysm arising from a gastroduodenal artery branch along the lining of the pseudocyst and consistent with an etiology for hemorrhage. Next, multiple Concerto™ microcoils were deployed into the gastroduodenal artery until assurance of complete occlusion of the feeding branch vessels was identified. Pancreatoduodenal or splenic aneurysm should be suspected if there is presence of melena or hematochezia in a patient with recurrent pancreatitis or pancreatic pseudocyst.

99 *A RARE CAUSE OF DUODENAL ULCER IN AN ADOLESCENT MALE.* V. Grossi, W. Sayej, Pediatric Gastroenterology, Connecticut Children's Medical Center, Hartford, Connecticut, UNITED STATES|C. Finck, Pediatric Surgery, Connecticut Children's Medical Center, Hartford , Connecticut, UNITED STATES|N. Bezler, Hematology Oncology, Connecticut Children's Medical Center, Hartford , Connecticut, UNITED STATES|.

Introduction: Symptomatic peptic ulcer disease (PUD) in pediatric patients is rare, with an incidence of 0.5 to 4.4/100,000 individuals. It is commonly associated with *Helicobacter pylori* infections or use of non-steroidal anti-inflammatory drugs (NSAID). When either etiology is not identified by history or diagnostic workup, evaluation for other causes must be pursued. We present a case of a duodenal ulcer initially thought to be from NSAID use, which later presented as a metastatic pancreatic neuroendocrine tumor (PNET). Case: The patient is a 15-year-old male with a history of asthma and migraine headaches presenting to the emergency room with dizziness. He was found to be severely anemic, with a hemoglobin of 3.9 g/dL; remainder of his complete blood count (CBC) was normal. He was mildly tachycardic but otherwise hemodynamically stable. History revealed that he was taking high dose ibuprofen for headaches every 6 hours for the past eight days. He denied abdominal pain, nausea, vomiting, hematochezia, or melena. Further laboratory evaluation revealed normal liver function tests, electrolytes, C-reactive protein and uric acid. Erythrocyte sedimentation rate was mildly elevated at 18 mm/hr. Stool hemoccult was weakly positive. After slow transfusions over 2 days, esophagogastroduodenoscopy (EGD) and colonoscopy revealed small gastric ulcerations with a large ulcer in the duodenal bulb; colonoscopy was normal. Duodenal, gastric, and esophageal biopsies were normal and negative for *H. pylori*. He was treated with pantoprazole 40 mg BID and sucralfate 1 gm TID. Six weeks later, the patient remained asymptomatic, with a hemoglobin of 9.7 g/dL. Repeat EGD revealed multiple small ulcerations in the duodenal bulb and mild bulging in the gastric antrum with a narrowed pylorus. Biopsies of the duodenum and esophagus were normal with mild eosinophilic gastritis. He was

scheduled for a 3-month follow-up EGD, but did not return until 6 months. At that time, he remained asymptomatic. Hemoglobin was stable at 9.5 g/dL; however, he endorsed a 20-pound weight loss. EGD revealed duodenal inflammation and ulceration creating a “second lumen” in the duodenal bulb. Again noted was a larger bulge in the antrum with evidence of gastric outlet obstruction. Computed tomography (CT) of the abdomen revealed a 4.5 x 4.4 x 3.6 cm mass in the head of the pancreas with mass effect on the duodenum and innumerable lesions in the liver, consistent with metastatic disease. Biopsy of the pancreatic mass was attempted via endoscopic ultrasound, but the specimen was nondiagnostic. The patient then underwent an exploratory laparoscopy. Wedge biopsy of a liver lesion confirmed metastatic PNET, well-differentiated and staining diffuse-positive for chromogranin and synaptophysin; gastrin, CD10, and vimentin were negative. The patient soon initiated treatment with lanreotide, capecitabine, and temozolomide. Discussion: This case highlights a rare, but significant, cause of a duodenal ulceration. While PNETs only compromise 1-2% of pancreatic tumors, it is important to investigate other etiologies of PUD when a history of NSAID use or *H. pylori* is not identified, or recurrent ulcers are refractory to medical treatment.

101 *THREE INTERESTING CASES WITH PROBLEMS OF LYMPHATIC SYSTEM OF GASTRO INTESTINAL TRACT.* B. Kerur, J. Shapiro, M. Herzlinger, N.S. LeLeiko, Pediatric Gastroenterology, Hasbro Childrens Hospital, Providence, Rhode Island, UNITED STATES|.

The diseases of lymphatic system of the gastro intestinal tract are rare. It is important to understand physiology of lymphatics, before medical management. We describe 3 cases with problem in lymphatics of intestines. The first case is 2 year old with infantile systemic hyalinosis. A rare, progressive, fatal condition, characterized by widespread deposition of hyaline material in many tissues including gastrointestinal tract. Protein-losing enteropathy (PLE) has been described in many of the cases with infantile systemic hyalinosis. Intestinal lymphangiectasia is the underlying cause of PLE in infantile systemic hyalinosis. She was lost for follow up before starting treatment. The 2 case was PLE due to increased right heart pressure, secondary to single ventricular physiology following palliative heart surgery. He was successfully treated with SC octreotide with normal fecal alpha1 antitrypsin and stable growth. The 3 case was 4 week old baby, with history of malrotation and post Ladd procedure in neonatal period. He was admitted with progressive ascites, tap was suggestive of chylous ascites, upper endoscopy was normal. Ultrasound revealed ascites with thickened intestinal loops on right side compared to loops on left side of abdomen, suggestive of lymphangiectasia. Ascites did not improve with holding enteral feeding and octreotide infusion. Upper GI series showed suspicion of volvulus. Operative findings showed multiple adhesions with lymphatic obstruction due intermittent volvulus. Following surgical adhesion lysis, he was switched to pregestamil with MCT oil, as he had developed reaction to high MCT formulas (Portagen, Infaport, monogen). Octreotide could worsen intestinal perfusion, caution should be exercised when starting it and correctible surgical complications should be rule out before starting octreotide. Conclusion: We report a case of chylous ascites as complication of post-surgery adhesions. In patients with chylous ascites, octreotide should be used with caution, surgical problems and conditions that may lead to ischemia should be ruled out. Octreotide may risk perfusion of intestine.

102 *PRIMARY INTESTINAL LYMPHANGIECTASIA, A MISSED DIAGNOSIS.* E. Khan, Pediatric, Riley Hospital for Children, Indiana University School of Medicine, Indianapolis, Indiana, UNITED STATES|A. Watts, G.C. Subbarao, Pediatric Gastroenterology, Riley Hospital for Children, Indiana University, Indianapolis, Indiana, UNITED STATES|.

Primary intestinal lymphangiectasia (PIL) is a rare protein-losing enteropathy caused by dilated lymphatic channels in the intestine. Lymphatic fluid leaks into the gut, resulting in hypoproteinemia, hypogammaglobulinemia, hypoalbuminemia, and lymphopenia. It is a rare disease most commonly affecting children under the age of three. Patients may present with failure to thrive, edema, lymphedema, ascites, or diarrhea. PIL is diagnosed by characteristic findings on endoscopic evaluation with confirmatory histopathological evaluation of small intestine biopsies. PIL can be either severe, affecting the entire small bowel, leading to lifetime disease, or a transient disorder only affecting part of the small bowel. Treatment focuses on a low fat, protein rich diet with

supplementation of medium chain triglyceride (MCT), though in some cases, total parenteral nutrition (TPN) is needed. Our case is of a 2 year old Hispanic female who presented to the Pediatric Gastroenterology team with failure to thrive and diarrhea. She had initially presented to the General Pediatric Hospitalist team three months prior with presumed infectious gastroenteritis. Her initial hospital course was complicated by poor tolerance of enteral fluids requiring a short course of TPN, as well as peripheral edema due to hypoalbuminemia requiring albumin infusion. Stool alpha 1 antitrypsin was normal. While her vomiting and edema resolved with supportive treatment, her diarrhea improved but persisted. Subsequently at the clinic visit, weight loss was noted with ongoing diarrhea. Laboratory studies revealed iron, zinc, and Vitamin D deficiencies as well as hypocarbia, hypocalcemia, hyperchloremia, and hypoalbuminemia. Fecal fat studies suggested malabsorption. Endoscopic examination revealed abnormal duodenal and jejunal mucosa containing diffuse raised white lesions of variable size consistent with lymphangiectasia. Subsequent biopsies confirmed PIL. She was placed on a high protein, low fat diet with supplementation of MCT. She was also given iron, zinc, and vitamin D supplements. This case demonstrates the importance of considering lymphangiectasia in pediatric patients presenting with diarrhea and hypoalbuminemia. Since fecal alpha 1 antitrypsin (A1AT) has sensitivity of only 58%, relying on these levels can lead to delay in diagnosis. Dietary modification remains the cornerstone in the management of PIL, though octreotide has been used for refractory cases.

103 *COLLAGENOUS GASTRITIS/DUODENITIS PRESENTING AS CLINICALLY SIGNIFICANT ANEMIA*. A. Kriegermeier, Gastroenterology, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES].

A 12 year old boy presented to his pediatrician with 2-3 months of progressive fatigue and was found to have a hemoglobin of 4.3 g/dL (13-16 g/dL), MCV-60 fl (78-98 fl), RDW-18.6% (12.4-14.5%), Reticulocyte count -1.8%. Other cell lines were normal. One year prior he had a normal MCV and hemoglobin of 12.7g/dL. Parents and patient denied any obvious source of bleeding such as hematemesis, hematochezia, melena or epistaxis. He also denied any abdominal pain, vomiting or diarrhea but did have decreased intake over the past several months and had lost 2.7kg in the past year. He was admitted to the hospital and transfused. Laboratory evaluation was consistent with iron deficiency anemia and he had no evidence of hemolysis. He had a normal abdominal ultrasound and negative celiac panel, PPD and infectious stool studies. Fecal calprotectin was <15 ug/g. He had 1 out of 3 fecal occult blood tests that were positive. He was discharged home on oral iron supplements to follow up with GI for further evaluation. He underwent EGD and colonoscopy. Colonoscopy was visually and histologically normal. The EGD was notable for mucosal oozing in the stomach prior to any manipulation or biopsies but no obvious source of bleeding was seen. There were multiple stellate and superficial ulcers in the body of the stomach and the duodenal bulb appeared edematous but the rest of the duodenum was visually normal. The pathology from the EGD initially reported moderately active chronic gastritis and mildly active duodenitis with mild focal villous blunting. However, upon further review of the biopsies with the pathology department there was noted to be a thickened subepithelial collagen layer within the stomach and duodenum with detachment of the surface epithelium. This finding was confirmed by Masson's trichrome stains. This finding was consistent with a diagnosis of collagenous gastritis and duodenitis. The patient was continued on oral iron supplements and started on omeprazole. He has subsequently maintained an appropriate hemoglobin, remains asymptomatic and gained 4kg in the subsequent 3 months. Repeat endoscopy has not been performed due to clinical stability. Collagenous gastritis/duodenitis is a rare disorder, but one that should be considered in the differential of patients that present with anemia even without abdominal pain or diarrhea. Special attention should be paid to the size of the subepithelial collagenous band deposition when considering collagenous gastritis/duodenitis as a diagnosis.

104 *PANCREAS DIVISUM IN CHILDREN WITH ACUTE RECURRENT AND CHRONIC PANCREATITIS*. T.K. Lin, J. Palermo, L. Hornung, L. Fei, M. Abu-El-Haija, Gastroenterology, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, UNITED STATES].

Introduction: Pancreas divisum (PD) is the most common congenital anomaly of the pancreas. The prevalence of this anomaly is ~7% based on autopsy studies. It has been implicated as a contributing factor to the development

of acute recurrent pancreatitis (ARP) and chronic pancreatitis (CP). The recognition of ARP/CP in children is increasing. The association of PD in children has been poorly defined. Through an internal hospital registry of children with ARP or CP, we sought to identify the occurrence rate of PD and to report characteristics such as genetic mutations that may better define this patient population. Methods: Children and adolescents ≤ 21 years of age defined as having ARP or CP were enrolled prospectively within REDCap database. A retrospective data review of patient information included demographics, pancreatitis symptom characteristics, gene testing results for mutations associated with ARP/CP, radiographic findings, and endoscopic retrograde cholangiopancreatography (ERCP) findings. Results: Over a twenty-month period, forty-three ARP/CP patients were enrolled into the database. A diagnosis of PD was made in seven children (5 female) based on findings by magnetic resonance cholangiopancreatography (MRCP) and/or ERCP. Mean age at the time of PD diagnosis was 10 years (range 2 to 14 years). Genetic testing for cationic trypsinogen (PRSS1), cystic fibrosis transmembrane conductance regulator (CFTR), and serine protease inhibitor, Kazal type 1 (SPINK1) was performed in all PD patients, with three patients having had additional gene testing for chymotrypsinogen C (CTRC). Mutations were found in 4 patients, including CFTR mutation (n=3) and SPINK1 mutation (n=2). One child was compound heterozygous for CFTR mutation and another had a combined CFTR and SPINK1 mutation. The remaining two patients had single mutations in CFTR and SPINK1, respectively. No differences in demographics or symptom characteristics were identified when comparing ARP/CP patients with PD to those without PD. The frequency of finding any genetic mutation in patients with PD was similar to patients without PD. MRCP was the initial radiologic modality that identified PD in all patients. Five patients subsequently had an ERCP with confirmation of complete PD in four and incomplete PD in one. Fluoroscopic changes suggestive of CP were found in all patients including a dilated dorsal pancreatic duct with/without ductal irregularity, ectatic side branches and/or a pancreatic duct stricture. Conclusion: Pancreas divisum was found in 16% (7/43) of our cohort of children with ARP/CP. This prevalence is greater than that of the general population; however, it remains unclear whether PD is a factor and to what degree in the development of ARP or CP in children. Patients with PD confirmed by ERCP are likely to have pancreatic ductal changes suggestive of CP.

105 *RED HERRING SIGNS OF DUODENAL BLUNTING AND HEPATIC GLYCOGENOSIS IN A PATIENT WITH SHWACHMAN-DIAMOND SYNDROME.* M. Lowry, L. Gillis, K.F. Thomsen, Pediatric Gastroenterology, Vanderbilt University, Nashville, Tennessee, UNITED STATES].

Case Description: A previously healthy female was referred at 10 months of age for elevated transaminase enzymes and failure to thrive. Laboratory testing revealed AST 133, ALT 231 and elevated anti-gliadin immunoglobulin (Ig) A antibodies. Due to low total IgA, tissue transglutaminase (TTG) antibodies were not reported. An esophagogastroduodenoscopy (EGD) revealed duodenal villous blunting and increased intraepithelial lymphocytes (IEL). Her growth and celiac serologies improved after initiation of a gluten-free diet, but transaminase enzymes did not improve. A liver biopsy revealed hepatocellular glycogenosis, confirmed by periodic acid-Schiff (PAS) staining. A Glycogen Storage Disease (GSD) Liver Panel by Massively Parallel Sequencing returned negative for 10 known genetic mutations. Urine organic acids, plasma amino acids, biotinidase profile and glycogen storage enzyme testing were essentially normal. At 4 years of age she developed rectal prolapse and constipation. CFTR genetics were negative. She was treated with Miralax without subsequent prolapse. Further screening labs revealed slightly low absolute neutrophil count (ANC), slightly elevated LDH, low fecal elastase and low total IgA and IgM. An abdominal US revealed increased echogenicity of the pancreas. She was started on pancreatic enzyme replacement. Genetic testing for Shwachman-Diamond Syndrome (SDS) resulted positive for one heterozygous mutation and one homozygous mutation consistent with SDS. A repeat EGD on a gluten-free diet continued to show mild villous blunting without increased IEL. A bone marrow biopsy revealed mild hypocellularity without signs of myelodysplasia. The transaminase enzymes improved, however pancreatic elastase remained low. SDS testing of her immediate family revealed a maternal carrier of two mutations, a paternal carrier of one mutation and a sibling carrier of one mutation. Discussion: This patient was diagnosed with SDS based on clinical findings and genetic confirmation. Two red herring signs, duodenal blunting and hepatic glycogenosis, delayed the diagnosis. The diagnosis of celiac disease remains in question as duodenal changes suggestive of celiac disease

have persisted after dietary removal of gluten, however, her growth and positive celiac serologies improved on a gluten-free diet. Our literature review found one case series reporting 11 out of 15 patients with SDS had duodenal changes of varying degrees. Although transaminase enzyme elevation is commonly seen in SDS, our search has not found any documented cases of hepatocellular glycogenosis. This patient's transaminase enzymes improved and she has not had any associated findings of glycogen storage disease suggesting that SDS caused the hepatic glycogenosis.

107 *IDENTIFICATION AND MANAGEMENT OF PANCREATIC ASCITES IN AN ADOLESCENT PATIENT*. B. Maksimak, P. Conjeevaram Selvakumar, V. Hupertz, Pediatric Gastroenterology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES|.

Background: Pancreatitis is a relatively common condition seen by pediatric gastroenterology providers. Most pediatric patients improve with supportive care measures with little to no significant long-term complications, but in a select number of patients the complications of pancreatitis can be encountered. Herein we report a case of pancreatic ascites in an adolescent male and its subsequent diagnosis and management. Case: The patient is a 14 year old male with a PMH significant for acute on chronic pancreatitis (positive for homozygous SPINK-1 mutation), chronic abdominal pain and a pancreatic pseudocyst who presented to the hospital with worsening epigastric abdominal pain and elevated amylase and lipase. He was subsequently made NPO, started on IV fluids and given pain medications. An MRCP was obtained which showed a decreased size of his pseudocyst but now with a fluid collection around the right kidney and paracolic gutter with only a small amount of peritoneal ascites. During the course of admission he continued to have worsening abdominal pain despite supportive care and improving laboratory values. Follow-up imaging was again obtained and now showed an increase in his ascites and increased dilation of the pancreatic duct. An ERCP was obtained and showed pancreatic duct disruption with a leak in the head of the gland with mild dilation and irregularity of the duct. A sphincterotomy along with pancreatic stenting was completed. US guided aspiration of the ascites was completed and showed an amylase of 120,392 U/L and lipase of 260,580 U/L. After the diagnosis, he had a peritoneal drain placed. Over time the ascites improved and he was eventually able to have the drain removed and we advanced his diet. Following discharge from the hospital he underwent work-up for pancreatectomy with islet cell transplantation and subsequently had the operation several months later. He now is doing well with complete resolution of his chronic abdominal pain. Discussion: Pancreatic ascites is defined as the accumulation of pancreatic fluid in the peritoneal cavity with peritoneal lipase being at least three times that of the plasma value. The most common cause in the adult population for pancreatic ascites is chronic pancreatitis due to alcohol abuse. History of a pancreatic pseudocyst is also fairly common in pancreatic ascites but in many cases the cause can be idiopathic. In pediatric patients, pancreatic ascites is quite rare and commonly being associated with traumatic pancreatic injury. The first step in managing these patients is supportive and starting on TPN to minimize pancreatic secretions. If no improvement, current literature suggests that ERCP is the modality of choice for evaluation and identification of a pancreatic leak. This also allows for therapeutic intervention with possible transpapillary pancreatic stenting, which reduces pressure in the ductal system and possibly occluding the leak. The hope is to allow for healing of the leak to avoid the need for more invasive surgical measures. Although rare in the pediatrics, pancreatic ascites needs to be considered with a history of a pancreatic pseudocyst or when they are not responding to supportive care alone.

108 *INTRACTABLE DIARRHEA, HYPERNATREMIA AND METABOLIC ACIDOSIS IN A NEONATE - A DIAGNOSTIC CHALLENGE*. C. Mandelia, S. Krishna, Pediatrics, Cleveland Clinic Children's, Cleveland, Ohio, UNITED STATES|B. Maksimak, P. Conjeevaram Selvakumar, V. Hupertz, N. Alkhouri, Pediatric Gastroenterology, Cleveland Clinic Children's, Cleveland, Ohio, UNITED STATES|.

Introduction: Glucose-galactose malabsorption (GGM) is a rare autosomal recessive disorder of intestinal glucose and galactose absorption. The infrequency with which it is encountered and the nonspecific presentation makes it a diagnostic challenge. Case Report: An 8 day old Amish female neonate was admitted for intractable diarrhea and failure to thrive with 19% weight loss since birth (2.72 kg from 3.35 kg). She was born at 39 weeks after an

uneventful pregnancy and was taking 16-20 Oz/day of Similac advance with up to 20 watery stools daily. Labs on admission revealed normal CBC, Na-177, K-6.5, Cl-147, HCO₃- 10.6, BUN-104, and creatinine-1.3. Hypernatremic dehydration, metabolic acidosis and pre-renal azotemia were treated with appropriate intravenous fluids and bicarbonate. Stool studies revealed low pH, high osmolar gap, positive stool reducing substances and a 24 hour NPO challenge resulted in dramatic improvement in hypernatremia and stool output pointing towards osmotic diarrhea. EGD and flexible sigmoidoscopy with biopsy were normal and later revealed normal disaccharidase activity and normal electron microscopy exam. In view of her Amish descent and clinical picture, a presumptive diagnosis of Glucose Galactose Malabsorption (GGM) was made and she was started on a carbohydrate free formula with added fructose which led to rapid resolution of diarrhea, correction of metabolic abnormalities and good weight gain. Genetic testing confirmed the diagnosis of GGM. Conclusion: GGM is a rare autosomal recessive genetic disease caused by deficiency in the intestinal sodium/glucose transporter, encoded by the solute carrier family 5, member 1 gene (SLC5A1). It presents with severe osmotic diarrhea in the neonatal period that can lead to significant metabolic derangement. Early recognition and institution of the correct formula can prevent further metabolic deterioration.

109 *NEONATAL DIARRHEA DUE TO SPINT2 MUTATION IS ASSOCIATED WITH ABNORMAL EPCAM LOCALIZATION.* A. Martinez, A. Muise, Y. Avitzur, Gastroenterology, Hepatology and Nutrition, Hospital for Sick Children, Toronto, Ontario, CANADA|C.E. Thoeni, E. Cutz, Pathology, Hospital for Sick Children, Toronto, Ontario, CANADA|P. Wales, Group for the Improvement of Intestinal Failure and Treatment, Hospital for Sick Children, Toronto, Ontario, CANADA|H. Huynh, Gastroenterology, Hepatology and Nutrition, Stollery Children's Hospital, Edmonton, Alberta, CANADA|A. Lacson, Pathology, Stollery Children's Hospital, Edmonton, Alberta, CANADA|.

Introduction: Neonatal diarrheas are associated with high morbidity and mortality and often result from genetic defects. SPINT2 mutation was recently reported in two different phenotypes of neonatal diarrhea – syndromic tufting enteropathy (TE) and congenital sodium diarrhea (CSD). The mechanism leading to the diarrhea in SPINT2 mutation and the association of SPINT2 mutation with these phenotypes are unclear. Methods: We present 2 children with SPINT2 mutation and TE / CSD phenotypes. Clinical characteristics and histopathological findings including Electron Microscopy (EM) and immunostaining for epithelial cell adhesion molecule (EpCAM), the abnormal protein in non-syndromic TE, and Ubiquitin C are described. Results: Patient 1 was born at 36 weeks of gestational age after an uneventful pregnancy with choanal atresia, choroid plexus cyst and congenital glaucoma. The parents are consanguineous with an Amish ethnic background. The patient developed secretory diarrhea soon after birth and requires total parenteral nutrition (TPN). Patient 2 was born at 36 weeks of gestational age from consanguineous parents. At birth he developed respiratory distress leading to the diagnosis of choanal atresia and ophthalmologic exam revealed punctate keratitis. At 2 weeks of age he presented with secretory diarrhea and severe hyponatremia and is TPN dependent. Genetic testing revealed the same homozygous mutation, c488A-G (Y163C), in both patients. Both patients underwent upper and lower gastrointestinal endoscopy which appeared macroscopically normal. Duodenal biopsies in patient 1 showed a relatively normal looking mucosa with mild focal partial villous blunting. PAS stain and EM analysis demonstrated normal brush border microvilli and prominent secretory-like granules in the cytoplasm of crypt enterocytes. Patient 2 showed epithelial tufts, most prominent in villus enterocytes, but no evidence of secretory-like granules on neither PAS stain nor EM. Immunohistochemistry for EpCAM in both patients showed mislocalized EpCAM in the cytoplasm of villus and crypt enterocytes. EpCAM was not detected at the basolateral membrane, where it is normally located. In double immunostaining studies, Ubiquitin C co-localised with EpCAM. In control samples (healthy patients and disease control), Ubiquitin C was mainly found apically and rarely localised with EpCAM in villus enterocytes. Network analysis suggested a potential interaction between EpCAM and ubiquitin C, a molecule involved in regulation of protein degradation. Conclusions: EpCAM is mislocalized in the cytoplasm of enterocytes in patients with neonatal diarrhea due to SPINT2 mutation. Immunostaining for EpCAM can be used as a diagnostic marker in these cases. Although SPINT2 mutation was reported with two different clinical phenotypes (syndromic TE and CSD), our findings suggest that these phenotypes could be part of a disease spectrum with common mechanism, potentially via EpCAM

mislocalization. Further studies are needed to elucidate the role of EpCAM and ubiquitin C in SPINT2 neonatal diarrhea.

110 *DIAGNOSIS OF CHRONIC PANCREATITIS WITHOUT PRECEDING EPISODES OF ACUTE DISEASE*. R. Paspulati, University Hospitals Case Medical Center, Cleveland, Ohio, UNITED STATES|M.S. Mehta, S. Sankararaman, T.J. Sferra, J. Moses, Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES|.

Background: Chronic pancreatitis (CP) is uncommon in children. The majority of children with CP have a history of acute recurrent pancreatitis (ARP); however, the diagnosis of CP can be challenging in the rare cases with no prior history of acute pancreatitis (reported to be $\leq 10\%$). Here, we highlight the early utilization of magnetic resonance cholangiopancreatography (MRCP) based on a high index of suspicion in diagnosing CP in three children who presented with no prior episodes of acute pancreatitis or abdominal pain. Case 1: 16-year-old male with a history of dopamine-sensitive dystonia and tethered cord presented with abdominal pain for a week and was found to have a serum lipase of 13,499 U/L. He had no previous illnesses suggestive of pancreatitis. An initial abdominal ultrasound (US) demonstrated mild hepatomegaly and a mildly dilated pancreatic duct. MRCP showed patchy areas of abnormal pancreatic signal enhancement with a stricture of the main pancreatic duct and upstream dilatation consistent with chronic pancreatitis. Case 2: 17-year-old female with a history of HLA-B27 positive uveitis presented with acute abdominal pain, normal amylase and lipase, and an abdominal US notable for hepatomegaly with mild intra-hepatic biliary dilation. She had no previous illnesses suggestive of pancreatitis. MRCP revealed changes consistent with chronic pancreatitis including diffuse mild pancreatic enlargement with loss of normal lobulated contour, peripheral rind-like enhancement, and mild prominence of the pancreatic duct with mass effect on the distal common bile duct and second portion of duodenum. Case 3: 14-year-old male with a history of end-stage renal disease secondary to Eagle Barrett syndrome dependent on peritoneal dialysis who presented with acute abdominal pain. Lipase was elevated with a peak value of 654 U/L. He had no previous illnesses suggestive of pancreatitis. Abdominal computed tomography and US identified a pancreatic pseudocyst that required percutaneous drainage. MRCP demonstrated a long segment pancreatic duct stricture within the body and head of the pancreas consistent with CP. Conclusions: Chronic pancreatitis should be considered in patients with hepatopancreatobiliary abnormalities on initial imaging despite lack of classic preceding symptoms or recurrent ARP. MRCP can be a useful diagnostic tool to diagnose chronic pancreatitis in these individuals. Early detection of chronic pancreatitis may lead to earlier interventions that can prevent long-term complications.

111 *12-YEAR-OLD MALE WITH SPLENIC ARTERY PSEUDOANEURYSM IN THE SETTING OF ACUTE PANCREATITIS*. G.A. Morris, C. Walsh, Pediatrics, Geisinger Health System, Danville, Pennsylvania, UNITED STATES|W.J. Cochran, Pediatric Gastroenterology, Geisinger Health System, Danville, Pennsylvania, UNITED STATES|.

Introduction Splenic artery pseudoaneurysm (SAP) is a rare complication of abdominal trauma or pancreatitis. In pediatrics, SAP usually follows abdominal trauma. We present a 12-year-old on valproic acid with pancreatitis that caused SAP. Case A 12-year-old African American male with complex past medical history including seizure disorder presented with two days of abdominal pain and irritability following a week of cough, rhinorrhea, and fatigue. He had decreased appetite. No emesis, diarrhea, fever, trauma, rash, or recent seizure. He had a history of constipation treated successfully with polyethylene glycol. Labs revealed a lipase of 206 U/L, amylase of 217 U/L, elevated valproic acid level, and platelet count of 45,000. Additional labs included normal liver and kidney panels, white blood cell count, and hemoglobin. Parvovirus B19, Epstein-Barr virus, and cytomegalovirus titers were negative. Hereditary pancreatitis panel (CFTR, PRSS1, SPINK1) was negative. Two months prior he had symptomatic acute pancreatitis. Amylase was 219 U/L and lipase was 257 U/L. At that time, abdominal ultrasound was normal with a poorly visualized pancreas. Outpatient management included low fat diet. Lipase normalized within 10 days. Past medical history included: hyperthermia-induced cerebral infarct and hemorrhage at age 8 months, global developmental delay, cerebral palsy, complex partial seizures, and constipation. Medications were valproic acid, rufinamide, clonazepam, clonidine, omeprazole, folic acid, and polyethylene glycol. Family history unknown, as patient was adopted. Physical exam at second admission revealed a developmentally delayed non-verbal male in

no distress. Vital signs and general physical exam were normal. Abdomen was soft, non-tender, non-distended with no organomegaly. Normal bowel sounds. On admission, a clear liquid diet, IV fluids and pantoprazole were started. Pain was controlled with ketorolac. Valproic acid was discontinued. Due to his second episode of pancreatitis, non-contrast abdominal magnetic resonance imaging and magnetic resonance cholangiopancreatography were obtained, revealing 3.1 centimeter heterogeneous mass with evidence of blood products arising from pancreatic tail. Subsequent pancreatic computed axial tomography scan with IV contrast showed nearly completely thrombosed pseudoaneurysm of the splenic artery at splenic hilum with 4.2 centimeter maximum axial dimension as well as edematous pancreatic tail. He underwent successful coil and Gel-Foam embolization of actively bleeding branch of the splenic artery. There were no prior signs of bleeding. Lipase normalized two days after coil embolization. Three weeks later, follow up abdominal CT angiogram showed resolution of splenic hilar artery hematoma and pseudoaneurysm without complicating infarct. He has not had any further episodes of pancreatitis. Conclusion Patient was diagnosed with pancreatitis while on valproic acid, which he had been on for two years. He also had SAP, which is very rare in the pediatric population. SAPs can be complications of pancreatitis as pancreatic enzymes likely weaken blood vessel walls. Endovascular coil embolization can successfully treat SAP.

112 *HEREDITARY PANCREATITIS PRESENTS WITH PANCREATIC DUCT STRICTURE*. M. Prero, M. Love, O. Almadhoun, University of Kansas, Kansas City, Kansas, UNITED STATES |.

A six year old African American male presented with severe recurrent abdominal pain and vomiting for the past four years. Initial work-up failed to identify the cause of pain. Due to pain severity, a CAT scan of the abdomen was done and showed mild enlargement of the pancreatic head with pancreatic duct dilatation, which was concerning for acute focal pancreatitis. Due to normal amylase and lipase enzyme levels that did not correlate with pancreatitis, MRCP was done and revealed a high grade proximal pancreatic duct obstruction with dilation of the main pancreatic duct. ERCP confirmed the pancreatic duct stricture in the pancreatic head. A sphincterotomy with balloon dilation and stent placement were done. Family history revealed a paternal great aunt died of pancreatic cancer and had been diagnosed with pancreatitis, and a paternal aunt also had pancreatitis. Given the family history of pancreatitis, genetic testing for hereditary pancreatitis was completed and revealed a heterozygous mutation of p.R122H in the PRSS1 gene, in addition to a heterozygous pathogenic mutation for the p.R1070Q in the CFTR gene. This case shows that hereditary pancreatitis can present with recurrent abdominal pain due to pancreatic duct stricture in the setting of normal pancreatic enzymes. Recognition of this disease is essential due to an increased risk of developing pancreatic cancer in the fifth decade of life.

114 *SOLID PSEUDOPAPILLARY TUMOR MASQUERADING AS GALLSTONE PANCREATITIS*. A.A. Singer, C.J. Dickinson, Pediatric Gastroenterology, University of Michigan, Ann Arbor, Michigan, UNITED STATES |.

Introduction: SPT (solid pseudopapillary tumor) is an epithelioid tumor that makes up 1-3% of pancreatic cancers. It may represent a higher proportion in pediatrics due to the relative rarity of other pediatric pancreatic cancers. Long-term single institution case series suggest SPT makes up 8-16% of pancreatic cancers in children. Approximately 2800 cases have been reported in the English literature since first described in 1959. In the last 15 years, the incidence has increased sevenfold as more advanced imaging techniques have become widely available. SPT occurs most commonly in adulthood and primarily in women, with a median age in the third decade of life. SPT may be asymptomatic. When symptoms are present, abdominal pain and mass are most common. Unlike pancreatic adenocarcinoma jaundice and weight loss are rare. Also unlike adenocarcinoma, serum levels of the CA 19-9 tumor marker are normal. Grossly, SPT can be solid, cystic, or mixed. It often contains hemorrhagic portions, is usually well circumscribed, and occurs most frequently in the pancreatic tail. Histologically, SPT consists of uniform appearing cells that form a solid mass that may degenerate into cystic hemorrhage and a pseudopapillary appearance. There is no clear relationship with pancreatic cell lines and the pathogenesis is unclear. SPT is usually indolent but has malignant potential, with 9-15% of cases causing local invasion or metastases. Accordingly, surgical resection is the standard of care, with distal pancreatectomy or pancreaticoduodenectomy being selected

based on tumor location. The prognosis is significantly more favorable than other pancreatic tumors, with a 5 year survival of 94-97%. The potential role of chemotherapy and radiation therapy requires further investigation. Case Report: A 13 year old Hispanic female presented with 2 days of epigastric pain and emesis. Her history included inherited hemolytic anemia with resultant cholelithiasis and splenomegaly. She had previously been hospitalized multiple times for cholelithiasis and family had deferred cholecystectomy. She had received three abdominal ultrasounds in the past, though the distal pancreas was obscured by bowel gas in all three. She had never received cross-sectional imaging. At the referring hospital, abdominal ultrasound demonstrated cholelithiasis and possible CBD dilation. The pancreas was not visualized. Her lipase was 726 IU/L and amylase was 128 IU/L. Due to concern for possible gallstone pancreatitis, she was transferred to our institution for more conclusive imaging and possible need for an ERCP. Repeat US on admission demonstrated a 2.4cm heterogeneous pancreatic tail lesion without central Doppler flow. MRI showed a 2.5 x 2.6 x 3.4 cm heterogeneous lesion with solid and cystic components, small foci of hemorrhage, a thickened peripheral enhancing rim, and minimal surrounding edema. The remainder of the pancreas was unremarkable. She was diagnosed with SPT. The patient is scheduled to receive distal pancreatectomy at the time of submission. Conclusion: We described an atypical presentation of an uncommon pancreatic tumor in children in order to increase awareness of an unusual lesion with rapidly increasing incidence.

116 *A CASE OF MULTICYSTIC LESIONS OF THE PANCREAS SECONDARY TO PANCREAS DIVISUM*. V. Sood, Y. Rekhtman, Pediatric Gastroenterology and Nutrition, Medstar Georgetown University Hospital, Washington , District of Columbia, UNITED STATES|K.M. Khan, Pediatric Transplant , Medstar Georgetown University Hospital, Washington, District of Columbia, UNITED STATES|.

Recurrent abdominal pain is a common indication for older children and teenagers to seek medical attention. While organic disease is not often found, atypical features warrants investigation. Here we report a case that might have otherwise presented in adult life. A 15 year old male was admitted with sharp, left upper quadrant abdominal pain of two months duration. Laboratory work up was normal including CBC, CMP, amylase and lipase. CT scan and MRCP of the abdomen revealed enlarged pancreatic tail with numerous cysts in the pancreas suspicious of pseudocyst, no cystic lesions in kidney and liver were found. The CEA, Ca 19-9, IgG levels, and pancreatic elastase were normal, C-Peptides, HbA1c was mildly elevated. Genetic testing for hereditary pancreatic cancer, Von Hippel Landau's syndrome, hereditary pancreatitis (PRSS1, SPINK1, CFTR and CTRC genes) and cystic fibrosis was normal. Endoscopic ultrasound demonstrated multiple cystic lesions, some connected to the pancreatic duct with largest cyst located near the tail of pancreas. Aspiration of cystic fluid was normal for cytology, amylase and low CEA level. Subsequent ERCP revealed pancreas divisum, an endoscopic sphincterotomy of minor papilla was performed with stent placement. He continues to experience abdominal pain, is undergoing evaluation for total pancreatectomy with pancreatic auto islet cell transplantation. Conclusion: Cystic pancreatic lesions in children are rare. Our patient has obstruction of the major pancreatic duct secondary to pancreas divisum resulting in variant of Wirsungocele with increased risk of recurrent pancreatitis or he may have congenital cystic lesions of the pancreas.

118 *GLUTEN-RELATED METHEMOGLOBINEMIA*. G. Tumgor, M. Agin, Pediatric Gastroenterology, Hepatology and Nutrition, Cukurova University Medical Faculty , Adana, TURKEY|G. Leblebisatan, Pediatric Hematology, Cukurova University Medical Faculty, Adana, TURKEY|.

Background: Acquired methemoglobinemia may develop due to drugs, chemicals, and some foodstuffs. Dapsone is the most commonly implicated drug, and even topical forms have been shown to lead to methemoglobinemia. Methemoglobinemia can also develop due to consumption of some nitrate-rich foods (such as spinach). Case: A 4.5-year-old girl presented to our clinic with failure to thrive, abdominal distension, and cyanosis. The patient's history revealed that cyanosis had begun at the age of 2 years, echocardiography performed at another center was normal, and hemoglobin electrophoresis 1 year previously had yielded levels of CO Hb 11%, HHb 11.8%, O Hb 40.6%, and MetHb 46.8% (0.4-1.5), at which time she had been diagnosed with methemoglobinemia. The patient had been started on methylene blue 1 mg/kg per day at another hospital. Cyanosis had decreased during drug therapy, but recurred when drug therapy ended. The patient had no history of use of any other drug or chemical

substance. On physical examination at presentation to our hospital, her body temperature was 36.7 °C, blood pressure 110/70 mmHg, heart rate 98/min, weight 12 kg (<3p; Z score -2.5), and height 84 cm (<3p; Z score; -5.2). The patient was suffering from cyanosis, malnourishment, abdominal distension, and genu valgum deformity in her right leg. There were no findings of dehydration in the case. Other systemic parameters were normal. Oxygen saturation at pulse oxymetry was 83% while breathing ambient air. Hemoglobin level was 12g/dL, Hct: 38%, MCV: 78 fL, serum iron level: 10 mg/dl, total iron binding capacity: 401 and ferritin: 4.7. Serum electrolyte values were normal. The genu valgum deformity in the right leg was identified as spontaneous healing of a right femur supracondylar femur fracture following a fall 2 months previously. The bones were osteoporotic. Celiac disease was considered on the basis of clinical and laboratory findings (tTG IgA > 160 U/ml and EMA IgA > 300 U/ml). Upper gastrointestinal endoscopy and histopathology were compatible with celiac disease (Marsh 3C). The patient was started on a gluten-free diet, and the diarrhea, abdominal distension, and cyanosis improved after 15 days. Diet compliance at 12-month follow-up was good. Control methemoglobin value was 11.2%, and there was no recurrence of cyanosis and there was no recurrence of cyanosis (weight 20 kg, Z score +0.33; height 96 cm, Z score -3.0). tTG (-) and EMA (-) were determined. Conclusion: We do not know the mechanism by which methemoglobinemia develops in celiac disease. We think it may be due to the aniline ring, insufficient response to oxidative stress, or endogenous intoxication seen in celiac disease. We know that celiac disease presents with very different, atypical clinical findings. We think that an atypical form of celiac disease may lead to methemoglobinemia in individuals with a genetic disposition. This is the first report of methemoglobinemia in celiac disease and that clinicians should consider this entity in patients with celiac disease and cyanosis/low saturation that fails to correct with supplemental.

119 *A 39 MONTH OLD WITH DIARRHEA AND HYPOKALEMIA*. J. Tung, C.H. Cramer, A.N. Lteif, C.A. Arndt, C. Moir, Mayo Clinic, Rochester, Minnesota, UNITED STATES|B. Anderson, Metropolitan Pediatric Specialists, Minneapolis, Minnesota, UNITED STATES|.

A 39 month old male was referred with a 20 month history of diarrhea (4-5/daily). Birth history was unremarkable. Initially breastfed through 18 months of life, his parents reported persistent loose stools. The stools were never watery, but did occur while fasting. There was no history of frequent antibiotic use or foreign travel. Trials of dairy, fructose and gluten elimination were unsuccessful. He began to have growth deceleration, though normal development. Prior workup included a normal complete blood count, electrolytes, albumin, ESR, c-reactive protein, celiac serology, and IgE to foods. Stool studies including culture, ova and parasites, Giardia antigen, Clostridium difficile toxin, occult blood, and fecal elastase were negative. EGD and flexible sigmoidoscopy biopsies were normal. At 37 months of age, hypokalemia was detected (2.1 mmol/L, normal 3.8- 5.1), with normal to mild hyponatremia and non-anion gap metabolic acidosis. A small-bowel follow-through was negative, and an abdominal ultrasound showed normal kidneys and dilated colon with significant liquid stool. Urine studies did not suggest a renal etiology, but rather normal renal compensation. Potassium citrate supplementation was initiated at 4mEq/kg BID. On examination, he was well-appearing and interactive. Cardiopulmonary exam was notable for pectus carinatum, with normal heart sounds. Maximal blood pressure was 91/55. Abdominal exam revealed no hepatosplenomegaly, mass, or tenderness. Echocardiogram was normal. Thyroid, zinc, and gastrin levels were normal. Stool analysis was notable for absence of leukocytes, normal sodium and chloride levels, but osmolar gap of -32 mOsm/kg. Hormone testing revealed the following abnormalities: serum vasoactive intestinal peptide (VIP) 500 pmol/L (normal <75), free normetanephrine 1.9 nmol/L (normal < 0.9), urine metanephrines 440 mcg/24hr (normal 34-169), norepinephrine 91 mcg/24 hr (normal 4-29), and dopamine 4401 mcg/24 hr (normal 40-260), urine vanillylmandelic acid (VMA) 43.6 m/gm Cr (normal <16), and homovanillic acid (HVA) 150.7 mg/gm Cr (normal <25). Imaging (CT chest/abdomen and MIBG scan) revealed a heterogenous tumor in the left retroperitoneum without metastases. Resection of the 5.7 x 4.3 x 4.0 cm mass was consistent with ganglioneuroblastoma. Bone marrow biopsy was normal, without features of malignancy. Within one week VIP, VMA, and HVA levels normalized. The oral potassium citrate was discontinued and one week later serum potassium and bicarbonate were also normal. He began to have formed stools. VIP-producing tumors cause a syndrome of watery diarrhea, hypokalemia, and achlorhydria (WDHA). Although ganglioneuroblastomas generally

present in young children, they rarely produce VIP. Furthermore, this patient did not present with typical profuse watery diarrhea and dehydration. Despite elevated serum and urine catecholamines, he did not present with pallor, sweating or hypertension. Tumors of the sympathetic nervous system can secrete different patterns of catecholamines and methylated metabolites. It is crucial they be measured in puzzling pediatric GI cases. Elevated levels will help identify occult tumors and lead to safe perioperative planning and followup for recurrence.

FUNCTIONAL/MOTILITY

123 *UTILITY OF MOTILITY STUDIES IN SELECTED CASES OF INTESTINAL FAILURE.* A. Algotar, M. Dienhart, D. Yacob, J. Balint, Division of Gastroenterology, Hepatology and Nutrition, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES|D. Bates, Division of Pediatric Radiology, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES|P. Minneci, Division of Pediatric Surgery, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES|.

While it is known that intestinal dysmotility can be an inherent problem or develop following injury to the bowel, formal motility testing is not routinely indicated in the care of patients with intestinal failure. However, determining the cause of failure to advance enteral feeds can be challenging and in select circumstances, motility testing can provide key data that aids management. Case 1: 15 month old with multiple intestinal atresias, the first at 43cm from the Ligament of Treitz with 3 additional short segments of small bowel including 4 cm of patent terminal ileum, an ileocecal valve and colon. Tapering enteroplasty of the proximal bowel was performed with 2 primary anastomoses leaving 54 cm of small bowel and entire colon in continuity. Post operatively, she required ongoing gastric decompression. At 1 month, a barium enema demonstrated a microcolon with free flow of contrast into a dilated distal small bowel. Upper GI with small bowel follow through showed a dilated small bowel with a colon of small caliber but no obstruction. Suction rectal biopsy demonstrated ganglion cells. With pro-motility medications on board, drip feeds were started with tolerance of only low volumes due to recurrent distension and vomiting. Repeat imaging studies documented progressive dilation of the small bowel with persistence of a non-obstructed very small caliber colon. As medical therapy failed, surgery was deemed necessary. The question was whether the abnormal appearing colon was also functionally obstructive leading to proximal dilation in which case an ostomy would be indicated. Colonic manometry demonstrated normal motility leading to a serial transverse enteroplasty with preservation of the entire colon. A small section of the distal small bowel was also resected with end-end ileal anastomosis. She has since been able to advance steadily on enteral feedings. Case 2: 6 yr old with Trisomy 21 who developed necrotizing enterocolitis requiring surgery, initially left with 45 cm of small bowel to a stoma, distal end of colon brought up as mucus fistula although this subsequently strictured, and a Hartmann's pouch. At 7 months ostomy was taken down with anastomosis to rectosigmoid colon. She was unable to remain free of parenteral support for any length of time. Imaging studies demonstrated a dilated residual colon. Anorectal and colonic manometry at age 4 years demonstrated normal anorectal manometry, but poor motility in the distal 16 cm of the colon. End ileostomy was created with the rectosigmoid colon left to a mucus fistula. Persistent high ostomy output prevented advancement to full feeds. Anterograde motility study from the mucus fistula showed good motility in the proximal 10 cm of the residual colon. She underwent ileostomy takedown and anastomosis to the colon with creation of colostomy 10-12 cm distally, providing improved absorptive capacity. She achieved enteral autonomy. While it is recognized that there is often dysmotility in short bowel syndrome, further manometric characterization when medical therapy has failed can allow for a more precise and objective surgical approach that results in clinical improvement while salvaging any piece of bowel that is functional.

139 *PERSISTENCE OF BILIARY PAIN AFTER CHOLECYSTECTOMY IN AN ADOLESCENT GIRL.* I. Naimi, Pediatrics , SUNY Upstate Medical University , Syracuse, New York, UNITED STATES|M. Beg, Division of Pediatric Gastroenterology, SUNY Upstate Medical University, Syracuse, New York, UNITED STATES|.

Introduction: Sphincter of Oddi Dysfunction (SOD) may be due to either stenosis or dyskinesia of the Sphincter of Oddi, a circular muscle at the distal aspect of biliary tree that controls flow of bile and pancreatic juices into the

duodenum. SOD can lead to a clinical syndrome that may include abdominal pain, elevation of pancreatic or liver enzymes, common bile duct dilation or episodes of recurrent pancreatitis. Case Report: A 16 year old female with chronic abdominal pain presented to the pediatric gastroenterology clinic in 2014 for second opinion. In 2011 she had undergone laparoscopic cholecystectomy for cholelithiasis, yet continued to experience recurrent abdominal pain (RAP) with nausea and vomiting. Laboratory tests had revealed persistent transaminitis with no visible abdominal pathology on repeat imaging. In 2014 we performed endoscopic gastroduodenoscopy that showed mild reflux esophagitis and mild chronic gastritis; Omeprazole was started with no improvement in symptom frequency or severity. Magnetic resonance cholangiopancreatography (MRCP) did not reveal any pathology. Subsequent endoscopic retrograde cholangiopancreatography (ERCP) with manometry revealed elevated biliary sphincter pressure of 86 mmHg leading to diagnosis of SOD. Endoscopic biliary sphincterotomy was performed without complications and the patient reported resolution of symptoms. Conclusion: SOD is an uncommon but emerging cause of RAP in children. Gastroenterologists should include it on their list of differential diagnoses, specially in patients who have undergone cholecystectomy. Data on safety and longterm efficacy of endoscopic sphincterotomy in children with SOD is scarce and requires prospective randomized trials for further evaluation.

140 *CHEMICAL DISSOLUTION OF A PEDIATRIC GASTRIC BEZOAR USING ORAL COCA-COLA.* A. Nelson, N. Romo, I. Batsis, A.S. Kota, J. Gershel, D. Levanon, Pediatrics, Jacobi Medical Center, Albert Einstein College of Medicine, New York, New York, UNITED STATES|E. Blumfield, Radiology, Jacobi Medical Center, Albert Einstein College of Medicine, New York, New York, UNITED STATES|.

Introduction: Bezoars are gastric or intestinal foreign bodies typically comprised of ingested food, hair, medications, and/or mucus. The patient may be asymptomatic or present with various gastrointestinal symptoms, or the bezoar may be diagnosed incidentally on abdominal imaging. We report the case of a 5 year old girl with an incidental finding of a gastric bezoar and its treatment with oral Coca-Cola. The use of the Coca-Cola is reported in the adult literature in a variety of regimens. The proposed mechanism of action involves the high phosphoric acid content and the carbonation of the Coca-Cola combining to degrade the biological materials making up the bezoar. Case Description: A 5 year old girl with a history of chronic constipation presented to the emergency department with abdominal pain. She was afebrile and denied vomiting or diarrhea. She was initially diagnosed with a urinary tract infection with positive urinalysis and culture, but did not complete her prescribed antibiotic course. When she returned to the emergency department with recurrent abdominal pain an abdominal radiograph revealed the presence of a gastric bezoar. The patient had no history of pica, trichotillomania, trichophagia, or extended-release medication use. She did have history of chronic constipation, swallowing chewing gum and being a picky eater. Her physical examination showed a well-appearing child with a normal abdominal examination, and no tenderness to palpation or palpable masses. The abdominal radiograph was repeated and showed persistence of the bezoar. Pediatric Gastroenterology and Pediatric Surgery were consulted, and the patient was admitted for evaluation and treatment. She was asymptomatic and clinically stable, so - based on cases reported in the adult literature - a trial of oral Coca-Cola was recommended, with endoscopy or surgery to follow if needed. The patient was treated with a regimen of oral Coca-Cola, small sips of water, and no other foods or liquids. During the first 24 hours of admission, she drank approximately 2 liters of Coca-Cola. The abdominal radiograph was then repeated, and it showed complete resolution of the gastric bezoar. She was discharged on a regular diet with polyethylene glycol for her constipation, and at a subsequent clinic visit she was asymptomatic and had a normal physical examination. Conclusion: The use of Coca-Cola to dissolve gastric bezoars is well-documented in the adult literature, but it has never previously been described in a pediatric patient. This case suggests that dissolution with oral Coca-Cola may be a safe, inexpensive, and well-tolerated approach to the treatment of gastric bezoar in clinically-stable, well-appearing children. Further research is required to validate this method and to clarify in what cases this may or may not be an appropriate and useful technique.

142 *USE OF REGULAR SUCROSE LOLLIPOPS AS A METHOD OF DISTRACTION DURING PEDIATRIC ANORECTAL MANOMETRY TO AVOID THE USE OF SEDATION.* J.K. Peacock, R. Medina-Centeno, R. Sanghavi, Outpatient

Gastroenterology, Children's Medical Center Dallas, Dallas, Texas, UNITED STATES | M. Craft, D. Ali Boucher, GI Lab, Children's Medical Center Dallas, Dallas, Texas, UNITED STATES |.

Introduction: For patients undergoing anorectal manometry (ARM), sedation is sometimes needed for anxious patients. However, sedation may affect the results of the ARM and limits the ability to test for sensation, defecation dynamics and other voluntary maneuvers. Sedative medications are also costly and require post-procedure monitoring, further increasing healthcare costs. Alternatives to the use of sedation during minor gastroenterology procedures have been examined, and the analgesic effect of oral sucrose has been especially observed in infants and young children. We report a series of ARMs during which a regular sucrose lollipop was used as a method of distraction to achieve patient cooperation. **Materials and methods:** Charts of all patients who received ARM at our institution over the past 29 months were retrospectively reviewed. Charts of patients who had documented non sedative distraction methods were selected for review. Methods of distraction and tolerance of the procedure were documented by the nurse attending the procedure. **Results:** One hundred thirty-five patients, ranging from seven months to 18 years, received anorectal manometries between January 1, 2013 and May 12, 2015. Of these, 12 patient charts were eligible for our review. Regular sucrose lollipops were used to distract all 12 of these patients during their anorectal manometries. Ages of these patients ranged from three years to 16 years. All 12 patients tolerated the procedure without adverse effects. The study was able to be completed in all these patients; sensation and defecation dynamics were successfully completed in these patients. **Conclusions:** Use of regular sucrose lollipops may be an effective method of distracting the anxious child during pediatric anorectal manometry. It may be used as a less expensive alternative to sedation, decreasing patient anxiety, increasing patient cooperation, promoting studies allowing testing for voluntary maneuvers, and decreasing healthcare costs.

LIVER

159 *LONG TERM REMISSION OF IMMUNE THROMBOCYTOPENIC PURPURA POST LIVER TRANSPLANTATION WITH RITUXIMAB: A CASE REPORT.* D.N. AlBogami, L.P. Szonyl, D.C. Broering, M. SHAGRANI, Department of Liver & Small Bowel Transplantation & Hepatobiliary/Pancreatic Surgery (Dpt)-R, King Faisal Specialist Hospital & Research Center, Riyadh, SAUDI ARABIA |.

ABSTRACT: **Introduction:** Thrombocytopenia is common among children following liver transplantation. Most cases are mild and transient. Late onset, severe, symptomatic immune thrombocytopenia is less common. **Method:** We report the case of a 4-year-old boy who underwent living related liver transplantation (LDRLTx) for biliary atresia. The patient had no episode of ITP before. At 23 months post-LTx, he presented with widespread purpurae and bruises. His platelet count was 1000 thr/ml. The common causes of thrombocytopenia (drug, infection, malignancy, hypersplenism) were excluded. Bone marrow biopsy showed adequate megakaryocytes. Platelet associated antibodies were detectable (Anti Platelet AB). The patient diagnosed as immune thrombocytopenic purpura and treated initially with intravenous immunoglobulin (1g/kg) for two consecutive days and methylprednisolone (2mg/kg/day for one week), however he did not respond to the conventional therapy or modification of immunosuppression (Tacrolimus was changed to cyclosporin). **Result:** Child was successfully respond to RITUXIMAB (Anti CD 20). He planned to receive a total of five doses weekly based on platelet count and immunoglobulin level. Now the patient 10 month after ITP. He seen regularly in the clinic and his platelet count normalized. **Conclusion:** The use of the rituximab (Anti CD 20) in immune thrombocytopenic purpura showed satisfactory long term outcome for patient post liver transplant.

161 *MECKEL-GRUBER SYNDROME: A RARE CAUSE OF ELEVATED TRANSAMINASES.* A. Andrews, J. O'Connor, Pediatric Gastroenterology, Hepatology and Nutrition, University of Oklahoma Health Sciences Center, Oklahoma City, Oklahoma, UNITED STATES |.

A 5-year-old male presented in acute renal failure (BUN 102/Cr 4.5). Renal ultrasound showed bilateral renal parenchymal disease. Laboratory findings included anemia, metabolic acidosis, hypocalcemia, hyperphosphatemia,

elevated GGT (397) and transaminases (AST 122, ALT 207). Alkaline phosphatase, total protein and bilirubin were within normal limits. Liver ultrasound revealed mild hepatomegaly (12.7 cm), increased echogenicity and a normal portal venous system with normal doppler flow. Genetics was consulted secondary to the combination of multisystem findings and a history of developmental delay and hypotonia as an infant. Genetic testing and head MRI were ordered, the later revealing a molar tooth sign. Patient was subsequently started on dialysis secondary to renal failure due to nephronophthisis and six months later he underwent a living donor kidney transplant. Hepatology was consulted four months post-transplant secondary to persistently elevated transaminases despite discontinuation of hepatotoxic medications. It was noted that for one year prior to transplant he had been treated for pruritus, thought to be allergic or psychosomatic. His evaluation included a normal alpha 1 antitrypsin level and phenotype MM, negative acute and chronic infectious hepatitis panels, negative autoimmune hepatitis panel and normal serum copper level. He had a positive EBV titer (peak 326) without clinical signs of acute EBV infection. Bile acid level was elevated (135.9). Liver biopsy revealed portal expansion by fibrosis and irregular ductal and ductular proliferation (biliary dysgenesis), bridging fibrosis, and mild focal chronic cholangitis. Pruritus was refractory to hydroxyzine, rifampin, and ursodiol and was causing the majority of his discomfort and pain. He has since had significant relief with naltrexone. Past medical history is significant for 37 week gestation complicated by oligohydramnios. He has global developmental delay and mild cerebral palsy. Other diagnoses include: esotropia, astigmatism, poor depth perception, chronic hypertension, oculomotor apraxia and bilateral hand tremor. Family history is remarkable for a sister born at 34 weeks who lived only a few minutes. Records from partial autopsy report that she had multicystic dysplastic kidneys and bilateral double ureters. Genome wide testing confirmed mutations in the TMEM67 gene leading to the diagnosis of Meckel-Gruber syndrome. Meckel-Gruber syndrome with associated nephronophthisis is an autosomal recessive lethal disorder characterized by central nervous system malformation (occipital encephalocele), bilateral renal cystic dysplasia, cleft palate, polydactyly, ductal proliferation in the portal area of the liver, pulmonary hypoplasia and situs inversus. Mutations are found in a number of genes that encode proteins involved in the function of primary cilia, basal bodies and centrosomes. The molar tooth sign is diagnostic on brain MRI for these ciliopathies. This case highlights a rare cause of elevated transaminases. Other more common causes should always be considered in the initial investigation as comorbidities often exist.

162 *A CASE OF EBV HEPATITIS REVEALING UNDERLYING PRIMARY SCLEROSING CHOLANGITIS IN A FIVE YEAR-OLD GIRL.* A.G. Carlson, Internal Medicine-Pediatrics, University of Kansas-Wichita, Wichita, Kansas, UNITED STATES|N. Farha, University of Oklahoma, Normal, Oklahoma, UNITED STATES|F.C. Giangiacomo, Pathology, Via Christi Hospitals Wichita, Inc, Wichita, Kansas, UNITED STATES|L. Hattar, Pediatric Gastroenterology, University of Kansas Medical School- Wichita, Wichita, Kansas, UNITED STATES|.

Primary sclerosing cholangitis (PSC) is a rare autoimmune condition that results in inflammation, fibrosis and stricturing of the bile ducts leading to chronic cholestasis and often end-stage liver disease. The incidence of PSC in children less than 18 years of age is 0.23 cases per 100,000 person years. Infectious mononucleosis is commonly caused by the Epstein-Barr virus (EBV) and is associated with hepatic inflammation. In most cases of EBV hepatitis, liver function studies may not necessarily be followed to resolution if the diagnosis of infectious mononucleosis is made. However, the following case suggests that a close follow up of hepatitis, even if presumed to be due to a viral illness, may be necessary. A 5 year-old female presented to her primary care physician with a 10-day history of intermittent fevers and was diagnosed with a viral syndrome. Patient represented a week later with persistent fevers, polyuria, and abdominal pain. Laboratory evaluation revealed leukocytosis, normocytic anemia, and transaminitis [AST 254mg/dL, and ALT 212 mg/dL] with alkaline phosphatase of 1,192mg/dL. Total bilirubin, INR, and albumin were all within normal limits. In the following days, patient developed hepatomegaly and lymphadenopathy, and was diagnosed clinically and by EBV titers with acute infectious mononucleosis. One month later, her laboratory evaluation showed a rising transaminitis [AST 532mg/dL, ALT 282 mg/dL] with alkaline phosphatase of 1258 mg/dL, and GGT of 662mg/dL. Again, albumin, INR, and total bilirubin were all within normal limits. Due to continued abdominal pain, fatigue and fevers, patient underwent an EGD and colonoscopy with biopsy that showed microscopic ileocolitis. Serological evaluation was done that resulted in mildly positive ANA

and total IgG. The remainder of the hepatitis evaluation was negative. Four months after initial presentation patient underwent liver biopsy due to persistently elevated liver enzymes that revealed stage 3 chronic biliary disease, most consistent with PSC. A MRCP was obtained that showed stricturing disease at the bifurcation of the right posterior hepatic duct consistent with PSC. Patient fevers resolved and abdominal pain improved with mesalamine and budesonide treatment. Fatigue and poor appetite continue to be her most significant complaints, reflecting the status of a chronic disease. In this particular case, the acute EBV infectious mononucleosis revealed her chronic undiagnosed liver disease. This case underscores the importance of following every patient with abnormal liver enzymes, even if the diagnosis is presumed to be an acute viral illness. Close follow-up allows for identification of other possible underlying liver diseases and monitoring for disease progression. This case also uncovered the rare diagnosis of PSC in a 5 year-old female, a disease with unidentifiable incidence in children less than 10 years of age.

164 *IDIOPATHIC NEONATAL HEPATITIS ASSOCIATED WITH HYPERINSULINISM AND HYPERAMMONEMIA*. A. Chugh, K. Shah, R. Azzam, Pediatrics, University of Chicago, Chicago, Illinois, UNITED STATES|E. Whitcomb, J. Hart, Pathology, University of Chicago, Chicago, Illinois, UNITED STATES|.

Introduction: Liver dysfunction and cholestasis presenting as idiopathic neonatal hepatitis (INH) is well associated with hypopituitarism. No cases of INH have been described in neonatal hyperinsulinism hyperammonemia (HIHA). This report describes a case of a neonate with HIHA and cholestasis with histological changes consistent with idiopathic neonatal hepatitis. Case Description: A full term AA male was born via c/s to a 29-year-old G3P3 mother with pregnancy complicated by gestational diabetes A1, hypertension, and asthma. Maternal serologies were all negative with the exception of GBS positive status. APGARs were 9 and 9. Birth weight was 3.65 kg. Physical exam was unremarkable. Postnatally, the patient was found to have hypoglycemia (20 mg/dL) that persisted and required titration of fluids to a GIR of 9.7 mg/kg/min even with adequate oral feeding. The infant was found to be hyperinsulinemic with a peak insulin level of 37.2 uIU/mL (nl < 28.5 uIU/mL) in conjunction with non-ketotic hypoglycemia. Evaluation for pan-hypopituitarism showed no abnormalities of the hypothalamic-pituitary axis including normal ACTH, cortisol, growth hormone, LH, FSH and thyroid studies. Studies for infectious etiologies were also done and were notably negative. At around 3 weeks of age, the ammonia level was found elevated on three separate measurements (up to 178 mcg/dL, nl 20-70 mcg/dL). A genetic test was sent for certain mutations associated with familial hyperinsulinism hyperammonemia but was negative. The patient developed a gradual rise in direct bilirubin and transaminases levels during the first two weeks of life. On DOL 19, he developed acholic stools. Direct bilirubin level peaked at 3.0 mg/dL, AST and ALT at 209 and 103 U/L, respectively, with albumin and INR within normal limits. Liver ultrasound was normal. Phenobarbital was given x 5 days before a HIDA scan was performed on DOL 26 showing normal uptake and excretion. A liver biopsy was performed at 4 weeks of age. It revealed mild lymphocytic infiltrates in the portal tracts, diffuse hepatocellular ballooning, and hepatocellular and canalicular cholestasis consistent with INH. Post-prandial hypoglycemia continued intermittently and thus Diazoxide was begun at 1 month of age, and was titrated from 5 mg/kg/day to 15 mg/kg/day to achieve blood glucose > 70 mg/dL. Bilirubin and liver tests significantly improved within the first 3 weeks into Diazoxide therapy. Treatment continued until 10 months of age, at which time a fasting glucose test and LFTs were normal. Discussion: To our knowledge, this is the first case of neonatal hepatitis associated with hyperinsulinism and hyperammonemia in the absence of hypopituitarism. The relationship between endocrinopathies and cholestasis has been well described. Generally, hormones are thought to play a role in bile secretion and bile salt independent flow. The specific mechanisms of action of individual hormones, however, are less clear. Normalization of hormonal dysregulation leads to resolution of INH and prevents permanent liver disease.

166 *REVERSIBLE NEUROLOGIC SEQUELAE OF REFRACTORY VITAMIN E DEFICIENCY SECONDARY TO PROGRESSIVE FAMILIAL INTRAHEPATIC CHOLESTASIS TYPE 2 IN A PEDIATRIC PATIENT FOLLOWING LIVER TRANSPLANT*. E. Collyer, V. Hupertz, K. Radhakrishnan, Department of Pediatric Gastroenterology, Hepatology, and Nutrition, Cleveland Clinic Children's, Richmond Heights, Ohio, UNITED STATES|B. Eghtesad, Transplant Center, Cleveland Clinic

Foundation, Cleveland, Ohio, UNITED STATES| R. Garcia, Pediatric Gastroenterology and Nutrition, Akron Children's Hospital, Akron, Ohio, UNITED STATES|.

Case: 15 yo Caucasian M who presented at age 13 with presumed diagnosis of PFIC Type 1 after presenting to OSH at age 1 with cholestasis and severe pruritus unresponsive to ursodiol and rifampin. Underwent Roux-en-Y partial biliary diversion at age 3, with resolution of pruritus à Normal growth and development until age 12 at which point he had acute regression of gross and fine motor & speech milestones. Brain MRI & 24 hr video EEG WNL. Neurologic findings all attributed to severe vitamin E deficiency: level <0.1 •He was started on high oral doses of vitamins A, D, E, and K with progressive worsening of neurologic function and admitted to OSH for GJ feedings and TPN for optimal provision of micronutrients. Discharged with total of 241 IU/kg/day vitamin E (RDA 25-50 IU/kg/d in cholestasis). Transdermal vitamin E attempted and 1 year high dose supplementation, tocopherol levels remained undetectable. He had no other past medical history, had no Amish or Mennonite roots, and no history of consanguinity. Physical Exam: weight and height were <3 percentile. He was anicteric, had bilateral clubbing, and no HSM. Neurologic exam was remarkable for absent DTRs in LE BL, +ataxic gait, +intention tremor, +dysynergia, +dysmetria, +dysarthria, +slowing of rapid alternating movements. He also had decreased sensation to vibration bilaterally in toes, decreased muscle bulk in LE. He had intact speech comprehension, but had significant dysarthria and scanning speech. Genetic testing sent to investigate underlying etiology: Returned with compound heterozygote for two missense mutations (c.3457C>T & c.499G>A) in the ABCB11 gene, clinching diagnosis of PFIC 2, or BSEP deficiency. •Given significant neurologic morbidity attributed to PFIC2, he was listed for liver transplant with MELD of 6, with appeal for MELD Exception. He received an orthotopic left lateral segment LDLT from an anonymous donor. After extensive PT and OT, he was ambulating with walker by hospital discharge. Walking unassisted with improved gait 2 months post-transplant. Discussion: Vitamin E deficiency can present as myopathic or neuropathic forms. Neuropathy is typically a spinocerebellar syndrome, with ataxia, hyporeflexia, and loss of proprioceptive and vibratory sensation, skeletal myopathy and a pigmented retinopathy Generally, vitamin E deficiency can lead to severe, irreversible neurologic effects if left untreated. There are case series of tocopherol repletion and improvement in neurologic symptoms though most cases are irreversible. Conclusion: In PFIC2 patients with severe refractory vitamin E deficiency and neurologic sequelae, liver transplant may reverse neurologic pathology when long term supplementation with high dose vitamin E does not improve levels or symptoms.

168 *PATHOLOGIC FRACTURES IN PEDIATRIC PATIENTS WITH HISTORY OF CHRONIC CHOLESTASIS*. D. Goldner, J. Vittorio, S. Lobritto, M. Martinez, Pediatrics, Columbia University Medical Center, New York, New York, UNITED STATES|.

Background: Bone disorders are common in patients with cholestatic liver diseases, and include hepatic osteodystrophy, vitamin D deficiency rickets, osteopenia, and fractures. As in other chronic conditions, the majority of the available information on skeletal complications of cholestasis is derived from published experience in adults. Aim: The aim of this study was to describe the clinical features, biochemical data, and radiologic bone changes of children with fractures related to a history of chronic cholestasis. Methods/Results: We surveyed all pediatric patients with a history of chronic cholestasis at our institution from 2010-2014 for incidence of fracture. Details characterizing each patient's medical history were obtained through retrospective review of the electronic medical record. Fourteen patients were found to have a total of 42 fractures. Fractures occurred primarily in the upper and lower extremity long bones and ribs (79%) with 7 of the 14 patients sustaining multiple fractures. There was little to no trauma documented in 11 of the 14 cases (79%). Eight of the 14 patients (57%) with fractures had biliary atresia. In 9 of the 14 patients (64%), fracture occurred post- orthotopic liver transplant with 6 of those 9 cases (67%) occurring within 1 year post-transplant. Age at time of fracture ranged from 6 months to 17 years (median 2.3 years). Features of metabolic bone disease were present in 86% of the x-rays obtained at the time of fracture. Only 6 of the 14 (43%) patients had biochemical evidence of active cholestasis with elevated GGT and/or bilirubin at time of fracture. 25-OH vitamin D was abnormally low in 7/14 patients (50%) and normal serum levels of calcium and phosphorus for age were noted in all but 1 patient at time of fracture. Six of the 14 children (43%) had been

exposed to high dose systemic steroids in the 6 months prior to fracture occurrence. Conclusion: Children with a history of cholestatic liver disease appear to be at risk for pathologic fractures that present in early childhood, despite normalization of laboratory parameters including 25-OH vitamin D and biochemical markers of cholestasis. These findings emphasize the need for increased clinical awareness of the fracture risk even after resolution of cholestasis, particularly during the peri-transplant period when this risk may be greatest. Further studies are necessary in order to quantify the risk of fracture in this population, to explore factors contributing to bone fragility in these children and to design clinical interventions to improve overall bone health.

170 *A NEW ASSOCIATION BETWEEN LATE ONSET ACUTE LIVER FAILURE AND TRMU GENE MUTATION REQUIRING LIVER TRANSPLANTATION.* A.S. Huang, R.E. Quiros-Tejeira, Pediatric Gastroenterology, UNMC, Omaha, Nebraska, UNITED STATES|W.B. Rizzo, Pediatrics Metabolism, UNMC, Omaha, Nebraska, UNITED STATES|W. Grant, L. Vargas, D. Mercer, A. Langnas, Surgery-Transplant, UNMC, Omaha, Nebraska, UNITED STATES|.

INTRODUCTION: Mitochondrial DNA (mtDNA) depletion syndromes remain a rare cause of liver failure. Usually these disorders are manifested during early infancy with poor feeding, vomiting, jaundice, hypo activity and impaired hepatic function. The majority of these patients resolve the initial episode after several months of supportive care. Among neonatal mitochondrial diseases, mitochondrial hepatopathies have become increasingly more common, demonstrating a combination of mtDNA-dependant complex deficiencies and a decrease in the number of mtDNA copies. Few genes have been identified to be responsible for the synthesis of stable mtDNA. There have been 18 cases reported of TRMU gene mutation. Five of them have progressed to overwhelming multiorgan failure and have died. The human TRMU gene plays an important role in mitochondrial translation; therefore, mutations on the TRMU gene have been associated to chronic liver disease with some cases presenting progressing to death. CASE PRESENTATION: Our patient a 12 month old female, presented with jaundice, intractable vomiting and lethargy that progressed to acute liver failure (ALF) and need for ventilatory support. She had history of poor feeding, vomiting and hypoactivity before developing ALF. Because of progression of her liver failure, it was decided to operate on her and do a liver transplantation (LTx). Before LTx, she had abdominal US showing findings consistent with fatty infiltrates of her liver. Her explanted liver had "microvesicular steatosis" consistent with a possible mitochondrial disorder. A dual genome panel by massively parallel sequencing was performed to study her mtDNA with findings of a homozygous missense variant of the TRMU gene. After her LTx, She is doing well. She is slowly regaining lost milestones before her LTx. CONCLUSION: Few cases of TRMU gene mutation have been reported causing significant liver disease yet there is no report of TRMU gene mutation leading to late onset ALF and need for liver transplantation. We are reporting the first case of late onset ALF secondary to TRMU gene mutations that successfully underwent a liver transplantation. TRMU gene mutation can lead to severe and acute liver disease and should be suspected in infants and toddlers presenting with evidence of a metabolic disorder leading to acute liver failure."

172 *REVERSING HEPATIC FIBROSIS WITH HIGH DOSE VITAMIN E IN A CASE OF ABETALIPOPROTEINEMIA.* C. Jackson, M.T. Geraghty, C. Jimenez-Rivera, Pediatrics, University of Ottawa, Ottawa, Ontario, CANADA|M.T. Geraghty, D. El Demellawy, C. Jimenez-Rivera, Children's Hospital of Easter Ontario, Ottawa, Ontario, CANADA|D. El Demellawy, Pathology, University of Ottawa, Ottawa, Ontario, CANADA|R.A. Hegele, Robarts Research Institute, London, Ontario, CANADA|.

Abetalipoproteinemia (ABL) is a rare disorder due to recessive mutations in the genes encoding microsomal triglyceride transfer protein (MTP) or apolipoprotein B (APOB). This results in hypocholesterolemia and lipid malabsorption. Symptoms usually present in childhood and include steatorrhea, failure to thrive and profound fat soluble vitamin deficiency with neurological sequelae as well as fatty liver. A 15 year old asymptomatic and healthy male was seen in clinic with elevated liver enzymes on routine blood tests. BMI was 19.5. On exam he had no hepatosplenomegaly or signs of liver disease. His AST was 50 IU/L, ALT 85 IU/L, ALP, GGT, bilirubin, albumin and coagulation tests were normal. Investigations for underlying liver disease were all negative. His ANA was positive (1:160). Abdominal US revealed a slightly coarse and echogenic liver suggesting fatty infiltration. Abdominal MRI

was normal except for fatty infiltration. Fasting lipid profile revealed total cholesterol 1.4 mmol/L (3.3-5.8 mmol/L), triglycerides 0.76 mmol/L (0.85-1.7 mmol/L), HDL 0.84 mmol/L (0.9-1.7 mmol/L), LDL <0.78 mmol/L (1.6-3.4 mmol/L). Lipid profile was repeated and results were similar. Vitamin A and D were normal and Vitamin E was low at 8 umol/L (12-46 umol/L). Liver biopsy showed steatohepatitis with severe extensive bridging fibrosis and early cirrhosis (stage 3/4) Fig 1. ABL was suspected, eye exam was normal and there were no acanthocytes. Molecular analysis showed compound heterozygous mutations in the MTP gene (c.1618>T:p.Arg540Cys; c.1901>G:p.Asp634Gly), which confirmed the diagnosis. He was started on high dose vitamin E, 800 IU BID. Liver biopsy was repeated one year later and revealed similar degree of steatohepatitis with improved fibrosis (stage 1c/4). This case exemplifies a case of NASH secondary to ABL in a non-obese patient on whom progression of hepatic fibrosis was halted and reversed by the use of high dose vitamin E.

178 *3 YEAR OLD FEMALE WITH ACUTE LIVER FAILURE SECONDARY TO TREATMENT WITH DEFERESIROX FOR BETA THALASSEMIA.* A. RAMASWAMI, D.J. Rosen, R. Arnon, Pediatrics & Recanati-Miller Transplant Institute, Mount Sinai School of Medicine, New York, New York, UNITED STATES].

Deferesirox (DFX) is an oral chelating agent used for a variety of diseases to treat chronic iron overload including beta thalassemia, sickle cell disease, and myelodysplastic anemia. DFX is generally well tolerated with the exception of GI disturbances and rash, although cases of renal toxicity, as well as acute and chronic liver failure, have been reported in adults. Here we describe a 3 year old female with beta thalassemia undergoing treatment with DFX and chronic transfusions who presented with RSV infection, Fanconi's syndrome, and acute liver failure. On presentation INR was 2.9 with ALT 199 IU/L, AST 263 IU/L, GGT 40 IU/L, total bilirubin 2.2 mg/dL, direct bilirubin 2.1 mg/dL, ammonia 447 mcg/dL, and ferritin of 1013 µg/L. Recent routine labs had shown normal liver enzymes and normal renal function, although most recent ferritin two weeks prior to presentation was low at 600 µg/L. Other causes of acute liver failure such as Wilson's disease, organic acidemias, fatty acid oxidation defects, hemophagocytic lympho-histiocytosis (HLH), autoimmune hepatitis, and viral etiologies including EBV, CMV, Hepatitis A and Hepatitis B were ruled out. RSV infection has been shown to cause acute hepatitis but not liver failure and so was ruled out as a potential etiology. Unfortunately the patient rapidly went into respiratory arrest with cerebellar tonsillar herniation, and was deemed too unstable for a liver transplant. Autopsy was declined. While up to 50% of cases of acute pediatric liver failure are indeterminate, given the acute presentation with Fanconi's syndrome and reported cases of acute liver failure from DFX in adults, it is likely that the acute onset of liver failure was due to DFX. The low ferritin prior to presentation suggests that over chelation with DFX may have been a factor, and it is possible that the concomitant viral infection also contributed to the acute hepatic failure. This case was reported to the FDA. It is important for pediatric gastroenterologists, hepatologists, and hematologists to be aware that the commonly used drug DFX may lead to acute liver failure in children.

MICROBIOLOGY/INFECTIONS/PROBIOTICS

180 *THE SPECTRUM OF IMMUNODEFICIENCY IN GASTROINTESTINAL ATRESIA AND TTC7A MUTATION.* A. Singh, J. Picoraro, M. Martinez, J. Khlevner, Pediatric Gastroenterology, Hepatology & Nutrition, Columbia University Medical Center, New York, New York, UNITED STATES | P. Jain, Department of Pathology and Cell Biology, Columbia University Medical Center, New York, New York, UNITED STATES | Y. Demirdag, Pediatric Allergy & Immunology, Columbia University Medical Center, New York, New York, UNITED STATES].

Gastrointestinal tract is the largest lymphoid organ in the body and it is not uncommon for intestinal manifestations to be associated with immunodeficiency. Recent studies implicate biallelic mutations in tetratricopeptide repeat domain 7a (TTC7A) gene responsible for combined immunodeficiency with multiple intestinal atresias (CID-MIA). Histologically, intestinal tissue exhibits enterocolitis with disruption of gut mucosal architecture and cell apoptosis. CID-MIA is a life-threatening condition evolving in utero with intestinal obstructions that require immediate postnatal surgical intervention. Prognosis is extremely poor; mortality often in infancy attributed to surgical complications, intestinal failure or opportunistic infection. We present a 5-year-old girl born at 35 weeks gestation to consanguineous parents with a complicated medical course in the neonatal

period for gastric outlet obstruction, congenital annular pancreas and duodenal atresia with malrotation leading to multiple small bowel resections. She later developed recurrent intestinal obstruction, enterocutaneous fistulas and remains dependent on parenteral nutrition due to severe intestinal failure. Recurrent polymicrobial sepsis since infancy and life-threatening gastrointestinal bleeding at age 4 attributed to CMV gastritis strongly suggested an underlying immunodeficiency. She has persistent severe lymphopenia associated with severe CD4+ lymphocytopenia. Transient neutropenia has also been observed during infections. Interestingly, she has normal quantitative immunoglobulins (IgG, IgM, IgA), yet there was no antibody response to protein or polysaccharide vaccinations. Whole exome sequencing revealed novel homozygous missense variant c.206T >C (p.L69P) detected in TTC7A, inherited from heterozygous parents. This variant was predicted to be deleterious and damaging by Sift and Provean (in silico protein deleteriousness prediction programs). The mutation is absent in 1000G and ESP population databases with the nucleotide change evolutionarily conserved. Expanding the phenotype of previously described cases of TCC7A mutations, this patient demonstrates an atypical form of combined immunodeficiency resulting in recurrent CMV and abdominal infections. In cases of intestinal atresia, immunodeficiency must be part of diagnostic considerations. Whether her immunodeficiency is intrinsic to lymphoid cells or extrahematopoietic defects remains unclear, although the patient is currently stable on antibiotic prophylaxis and IVIG infusions to circumvent infection. Genotyping has opened discussion for future therapeutic plans, which may include a combination of bone marrow, thymus and small bowel transplantation. Discovery of new variants make it essential to investigate their causality in human disease, establishing clinically relevant mutations. As genotyping becomes increasingly prevalent, clinical implications and management may be tailored for these monogenic diseases. The unique immunologic and clinical presentation in this case suggests that TTC7A gene has a clinical-pathologic spectrum more variable than previously recognized and should inform future management of this disease.

Friday, October 9, 2015

POSTER SESSION II

Exhibit Hall

1200pm – 2:00pm

Presenters at posters from 12:30 – 1:30pm

ENDOSCOPY/QI/EDUCATION

188 *TODDLER WITH SHORT BOWEL SYNDROME PRESENTING WITH OBSCURE RECURRENT GASTROINTESTINAL BLEEDING: A DIAGNOSTIC AND THERAPEUTIC DILEMMA.* C. Biaggi, L.J. Febo, A. Langshaw, Pediatric Gastroenterology, University of Miami, Miami, Florida, UNITED STATES | J. Garcia, H. Neville, E. Perez, Pediatric Surgery, University of Miami, Miami, Florida, UNITED STATES |.

Serial transverse enteroplasty (STEP) may be performed in patients with short bowel syndrome (SBS) who have significant bowel dilation leading to malabsorption and dependence on total parenteral nutrition (TPN) or have small bowel bacterial overgrowth (SBBO) refractory to medical management. Although published data is scarce, gastrointestinal bleeding (GIB) has been reported at staple line sites. Treatment modalities for such pathology is limited and individualized. We describe a case of a 16-month-old boy with history of SBS secondary to multiple intestinal atresias presenting with obscure recurrent GIB 8 months post STEP procedure. Patient underwent two resections with primary anastomosis shortly after birth. At 4 months of age he was referred to our institution where he underwent repair of previously unrecognized malrotation and STEP procedure. Post STEP small bowel length was 61cm, which allowed for enteral autonomy by 9 months of age. At 12 months of age, 8 months after his STEP procedure, he presented to the emergency room (ER) with profuse melena and pallor. He was noted to have hemoglobin of 6 and an albumin of 2.4. Endoscopy (EGD)/colonoscopy was performed and no ulcerations, polyps or inflammation were noted. Histological findings were unremarkable. Over the course of 4 months, he presented 7 times to the ER with similar symptoms and symptomatic anemia requiring 7 blood transfusions. Workup included

multiple nuclear medicine bleeding scans (1 with heparin induction), Meckel's scan, CT angiography, repeat EGD/colonoscopy, upper GI series and barium enema- all of which failed to identify source of bleeding. Capsule endoscopy could not be performed due patient's age and size. Therapeutic modalities included amino acid based formula for potential allergy, sulfasalazine for possible anastomotic ulcer and cyclic antibiotics for SBBO. After 4 months of failed medical management, diagnostic procedures, and dependence on blood transfusions, he underwent exploratory laparotomy. Lysis of adhesions was performed followed by EGD and push enteroscopy with an open abdomen. Two ulcers were found at STEP staple line in the jejunum with scattered small ulcers proximal to the site. Remeasurement of the small bowel revealed 126cm, double the length documented at 4 months of age. Hence, given sufficient bowel length and enteral autonomy, 6cm of affected bowel was resected. Tissue obtained had no ischemic changes, vasculitis, eosinophils or changes of inflammatory bowel disease. Since surgery, melena resolved and hemoglobin has remained stable. He continues on an elemental diet, thriving, with no need of parenteral support. This case provides insight into the diagnostic and therapeutic options in a toddler SBS patient with enteral autonomy who presents with GIB after STEP procedure. Exploratory laparotomy and/or push enteroscopy should be considered in cases where medical interventions have failed, capsule endoscopy is not an option due to patient's age and size, STEP portion cannot be reached by conventional EGD, and frequent bleeding requiring transfusions pose high morbidity to the patient.

190 *ZINC TOXICITY WITH FOREIGN BODY INGESTION*. O. Choudhry, F. Zapata, UNMC, Omaha, Nebraska, UNITED STATES|O. Choudhry, F. Zapata, Children's Hospital and Medical Center, Omaha, Nebraska, UNITED STATES|.

A 3 y/o male, ex-33 week premie with known history of abnormality in CHD7 gene (possible CHARGE association) and seizure disorder, presented to primary physician with persistent vomiting and found to have a foreign body in stomach. Mom was instructed to watch for passage in stools but did not see it pass and did not follow up. Two months later admitted for increased seizure activity and on repeat x-ray found to have the foreign body still in stomach. Was taken for endoscopy where seven pennies were found in the stomach, one of which was corroded and breaking apart. Each was individually removed and mucosa was observed to be normal appearing. Zinc levels were checked the same day and found to be severely elevated at 494 ug/dl (Normal: 60-120 ug/dl). Repeat levels checked in one week and two weeks from the procedure showed consistent decline in zinc levels to normal showing that the pennies were the source of elevated zinc in blood. Copper levels remained normal, seizure activity improved in the meantime. Zinc toxicity can present with seizures and can affect levels of other elements such as copper. In our case it is difficult to predict whether concurrent increase in seizure activity was due zinc toxicity or underlying seizure disorder, no other neurological changes from baseline were noted.

191 *INTESTINAL TRANSPLANT FOR MANAGEMENT OF INFANTILE JUVENILE POLYPOSIS SYNDROME (IJPS)*. O. Choudhry, P. Palomo, R. Quiros, E. Reyes, A. Langnas, W. Grant, D. Mercer, L. Vargas, UNMC, Omaha, Nebraska, UNITED STATES|O. Choudhry, P. Palomo, R. Quiros, E. Reyes, Children's Hospital and Medical Center, Omaha, Nebraska, UNITED STATES|.

A 3 y/o female with known diagnosis of Infantile Juvenile Polyposis syndrome was transferred to our center for intestinal transplant evaluation. She was found to have hundreds of polyps through out her gastrointestinal tract. Due to ongoing anemia and hypoalbuminemia, small bowel resection and subsequent small bowel transplant was performed at our center. She had an ileostomy and developed polyps in the stoma and subsequent graft failure. She underwent explantation of the graft, was stabilized on TPN and brought back for Liver, Pancreas, Small bowel and our first Pediatric Colonic transplant. Surveillance biopsies were done from both the ileum and colon, showed no signs of rejection. Patient showed good recovery and is being maintained on immunosuppression.

192 *IMPLEMENTATION OF SPECIFIC ROUNDING PRACTICES YIELDS IMPROVED COMMUNICATION AND ENHANCES PARENTS' PERCEPTION OF HEALTHCARE DELIVERY*. D. Galloway, J. Gossett, S. Miller, L. Cope, E. LaTulippe, J. Higby, S.A. Saeed, Gastroenterology, Cincinnati Childrens Hospital Medical Center, Cincinnati, Ohio, UNITED STATES|.

Background Effective communication and collaboration with patients and their families is essential to the care of the hospitalized patient. Multidisciplinary, family-centered rounds are an ideal method utilized to accomplish this objective and have been implemented at Cincinnati Children's Hospital Medical Center since 2003. However, as medical team size and medical complexity has increased, the ability to consistently meet the needs of the patients' families in addition to sustain effective communication among both parties has been challenged. Methods A 7-question survey derived from the Inpatient Child Hospital Consumer Assessment of Healthcare Providers and Systems (HCAHPS) questionnaire was utilized to solicit objective data regarding families' perceptions of the core aspects of the rounding process. Each question solicited four answers and was assigned a numerical value (Never-1, Sometimes-2, Usually-3, Always-4). The percentage of questions answered "Always" served as the outcome measure for improvement and tracked using a run chart. This measure was assessed at baseline across a 12 week period and throughout the study period of 3 months where five different Plan-Do-Study-Act (PDSA) ramps were tested. Results The baseline median percentage of the family survey questions answered "Always" was 78%. Interventions targeted more effective communication, team work, accountability, defined roles and family involvement on rounds. The median percentage increased to 93% by the end of the study period. All interventions were adopted into practice and incorporated into the rounding guidelines for the inpatient gastroenterology service. Conclusions The implementation of specific rounding practices targeting more effective communication, team work, accountability, defined roles and family involvement on rounds leads to a more consistent and reliable rounding process. Furthermore, it improves Parent/Guardian perceptions of delivery of care and communication with the healthcare team during family-centered rounds, and may have impact on satisfaction with care and overall experience.

193 *CHOLEDOCHAL ABNORMALITY WITH CHOLEDOCHOLITHIASIS IN 13 MONTH OLD WITH SUDDEN ONSET JAUNDICE*. R.P. Dillard, C.A. Hieronymus, University of Kentucky, Lexington, Kentucky, UNITED STATES].

A 13 month old previously healthy male presented with a two-day history of jaundice and light colored stools. His mother had noticed generalized fussiness in days prior, but no other symptoms. He was found to have transaminitis, elevated INR, elevated lipase, and conjugated hyperbilirubinemia. Exam was also significant for hepatomegaly and right upper quadrant tenderness. Abdominal ultrasound demonstrated intrahepatic and extrahepatic biliary ductal dilation. MRCP showed a stone in the common bile duct. ERCP with cholangiogram performed a few days later identified an anomalous pancreaticobiliary junction and a 10 mm long common channel as well as dilation of main bile duct and central intrahepatic biliary tree. Successful sphincterotomy with stone and sludge extraction was performed. The patient was discharged the following day. At his one month follow-up appointment, transaminases, bilirubin, lipase, alkaline phosphatase and GGT had normalized and jaundice had completely resolved. This case illustrates the importance of early cholangiopancreatography in young patients presenting with cholestatic jaundice. Biliary stones in conjunction with choledochal abnormalities should be considered in the differential diagnoses of obstructive jaundice in otherwise healthy children.

194 *FECAL CALPROTECTIN: A POSSIBLE ROLE IN SCREENING FOR JUVENILE POLYPS?*. S.K. Hourigan, Johns Hopkins School of Medicine, Greatfalls, Virginia, UNITED STATES|F. Khan, H. Mani, Inova, Fairfax, Virginia, UNITED STATES|C. Chao, S.K. Hourigan, Pediatric Specialists of Virginia, Fairfax, Virginia, UNITED STATES].

Fecal calprotectin is commonly used as a quantitative marker for intestinal inflammation, predominantly in inflammatory bowel disease; however the role of this laboratory value has not been well established for patients with juvenile polyps. We present two cases which highlight the association of raised fecal calprotectin with large juvenile polyps. Case 1 A 10-year-old male presented with intermittent bloody stools and iron deficiency anemia. A calprotectin level was obtained which was abnormally elevated at 575 mcg/g (normal < 50 mcg/g). Subsequently, colonoscopy was performed which showed a large pedunculated single polyp at the hepatic flexure (4cm x 2.5cm x 2.5cm) which was removed. The histology was consistent with a juvenile polyp. Six months after the polypectomy, a repeat calprotectin level normalized to <50 mcg/g. In addition, due to the large size of the polyp, he had a repeat colonoscopy at 6 months which was normal without any further polyps being found. Case 2 A 10-year-old girl with

several years of intermittent of abdominal pain, poor growth and a family history of Crohn's disease, presented with a recent history of bloody stools. The patient had a calprotectin level of 1386 mcg/g. An endoscopy and colonoscopy performed showed a large complex polyp at the hepatic flexure (5cm x 4cm x 4cm) and two smaller polyps in the sigmoid colon and rectum which were removed; pathology was consistent with juvenile polyps. A repeat calprotectin level one month after polypectomy had normalized to <50mcg/g and the patient had no further gastrointestinal complaints. Both of these cases illustrate the association of raised fecal calprotectin with juvenile polyps, with return to normal levels after polypectomy. It is not surprising that calprotectin, a protein present predominantly in neutrophils, is increased with juvenile polyps given the high presence of inflammatory cells, including many neutrophils within the polyps. This then raises the possibility of using fecal calprotectin, along with the current standard stool hemoccult as a screening tool for juvenile polyps, to assess complete removal of all polyps and also possibly for polyp recurrence. This lab marker may also serve as a noninvasive measure instead of frequently repeating colonoscopy procedures for detecting polyps.

197 *COLON ADENOCARCINOMA IN PEDIATRICS: A REVIEW OF 4 CASES FROM A COMMUNITY HOSPITAL*. L.S. Lawrence, Pediatric Residency, Advocate Children's Hospital, Oak Lawn, Illinois, UNITED STATES|C. Smith, R. Nagpal, Pediatric Gastroenterology, Advocate Children's Hospital, Oak Lawn, Illinois, UNITED STATES|.

Colon adenocarcinoma is a common malignancy in the adult population, but is believed to be a rare occurrence in the pediatric population. The objective of this review is to highlight that pediatric colon adenocarcinoma is not an uncommon malignancy in the pediatric population and that it can present in a variety of ways that the physician needs to be aware of in order to make a timely diagnosis. We present our experience in a community hospital with four patients, two to be discussed now and two further in the poster [as pending IRB approval], of patients diagnosed with colon adenocarcinoma in the pediatric population [age <20 years old] over the past ten years. A 15 year-old female [Patient A] presented with three months of epigastric abdominal pain and one month of anorexia, fatigue, diarrhea, emesis and weight loss; and an 18 year-old male [Patient B] with cystic fibrosis presented with progressive periumbilical abdominal pain and diarrhea. Physical exam notable for mild pallor in both patients and was otherwise unremarkable for Patient A, while for Patient B a mass was palpated under the liver. Laboratory testing notable for microcytic anemia in both. In Patient A and Patient B, on colonoscopy an ulceroinflammatory mass was seen in the ascending colon along with two small polyps. Histopathology for both was consistent with poorly differentiated adenocarcinoma with mucin-producing and signet ring features. Initial management with right hemicolectomy, where pathology confirmed stage IV adenocarcinoma with peritoneal seeding. Patient A is currently undergoing palliative chemotherapy while Patient B has since deceased. As there are no screening methods in place to catch colon adenocarcinoma in its early stages for the pediatric population, children often present in later stages and have worse outcomes compared to adults. As such, colon adenocarcinoma in the pediatric patient should be on the physician's radar and considered as the potential cause of epigastric or periumbilical abdominal pain, pallor, anorexia, weight loss or diarrhea even with a normal physical exam.

199 *COLLAGENOUS GASTRITIS: A MEDICAL MYSTERY*. M. Malik, Jinnah Sindh Medical College, Karachi, Sindh, PAKISTAN|M. Beg, Division of Pediatric Gastroenterology, SUNY Upstate Medical University, Syracuse, New York, UNITED STATES|.

Mariam Z. Malik, M.B.B.S, Mirza B. Beg, MD, Jinnah Sindh Medical College, Karachi, Pakistan, Division of Pediatric Gastroenterology, SUNY Upstate Medical University, Syracuse, New York Abstract: Collagenous gastritis is a rare histo-pathological disorder characterized by thick sub-epithelial collagen bands (>10 µm) associated with inflammatory infiltrate of the gastric mucosa. It affects two age groups: children presenting with anemia with gastric mucosal diseases and adults presenting with chronic diarrhea with collagenous colitis. We discuss the case of a 7-year old with collagenous gastritis and associated colitis. Patient presented with hematemesis and spatial disorientation. Patient had a history of iron deficiency anemia, abdominal pain, failure to thrive and constipation. Esophagogastroduodenoscopy and colonoscopy showed no source of active bleeding. Stoolcalprotectin test suggested inflammatory bowel disease but blood serological markers were normal. Gastric body and antral

biopsies revealed dense chronic inflammation and epithelial fibrosis suggestive of collagenous gastritis. Duodenal and ileal biopsies were normal. Colon biopsies showed non-specific colitis. There was no significant architectural distortion and no observable fibrosis. Upper GI and small bowel series were normal. Capsule endoscopy revealed abnormal mucosa with edema, flat areas in proximal bowel and scalloped mucosa in ileum. Celiac disease antibody test was negative. Empirically, the patient was treated with a gluten-free diet and therapy with mesalamine, ferrous sulphate and lansoprazole with improving symptoms. Due to collagenous gastritis's unknown etiology, a definite treatment regimen does not exist. Symptoms are relieved through therapy of iron supplement, proton pump inhibitor, anti-inflammatory agent(s) and systemic corticosteroid. Physicians should consider collagenous gastritis when evaluating a child with refractory iron deficiency anemia and abdominal pain along with findings of sub-epithelial collagen deposition within the gastric mucosa. Randomized double blind placebo control studies are needed for establishing a treatment regimen.

203 *SEVERE ESOPHAGITIS WITH STRICTURE DUE TO LICHEN PLANUS IN A FEMALE TEEN*. D. Molla Hosseini, Pediatrics Gastroenterology, Hepatology, and Nutrition Fellowship, University of Colorado School of Medicine, Aurora, Colorado, UNITED STATES|G. Kobak, Digestive Health Institute, Children's Hospital Colorado, Aurora, Colorado, UNITED STATES|C. Torres-Zegarra, L. Prok, Dermatology, Children's Hospital Colorado, Aurora, Colorado, UNITED STATES|.

This is the case of a Caucasian patient with a history of significant dermatologic problems including poorly controlled seborrheic dermatitis with frontal alopecia, for which she has been closely followed in the Dermatology clinic since her initial presentation at 6 years of age. Given the lack of improvement with treatment over the course of a year, a scalp biopsy was performed revealing lichenoid changes. Two years later, she presented to the Gastroenterology clinic for abdominal pain, dysphagia, and poor weight gain. Empiric treatment with a proton pump inhibitor failed to improve her symptoms and an endoscopy with biopsies was performed detecting a severe lymphocytic esophagitis. At the time, there were concerns about an atypical autoimmune process and corticosteroids were empirically started. The patient had resolution of her pain and dysphagia but was then lost to follow up. Meanwhile, she continued to be seen by Dermatology for her seborrheic dermatitis and frontal alopecia, which remained poorly controlled despite multiple combinations of topical steroids, keratolytic agents, and antifungal shampoos. Four years later, she again presented to the Gastroenterology clinic with abdominal pain, dysphagia and odynophagia, vomiting, and poor weight gain. A repeat endoscopic evaluation revealed chronic esophagitis as well as a stricture requiring dilation. Her ongoing dermatologic issues and persistent gastrointestinal problems lead to the concurrent re-evaluation of the skin and esophageal biopsies. The two biopsies were found to show a similar lymphocytic inflammatory process, leading to the diagnosis of cutaneous and enteral lichen planus. Lichen planus is a chronic inflammatory papulosquamous disorder of unknown etiology that is rarely seen in children. It is a well-recognized disorder involving the skin, nails, and mucosal surfaces but is predominantly a disease of middle aged women, affecting less than 1% of the general population. Lichen planus esophagitis is otherwise extremely rare and has only been described within a handful of adult patients in their fourth decade of life or later. Given the rarity of esophageal involvement, there is often a delay in diagnosis and subsequent initiation of the appropriate treatment. We present the first reported case of lichen planus esophagitis and subsequent stricture in the pediatric population. A unifying diagnosis of lichen planus involving both scalp and esophagus was critical in our patient due to its prognostic and therapeutic distinction from other causes of alopecia, dermatitis, esophagitis, and stricture formation.

204 *THE RADIOLOGIC JOURNEY OF AN ABDOMINAL MASS: A CASE OF CASTLEMAN'S DISEASE*. K. Nelson, A.S. Kota, J. Gershel, D. Levanon, Pediatrics, Jacobi medical Center, Albert Einstein College of Medicine, New York, New York, UNITED STATES|E. Blumfield, Radiology, Jacobi Medical Center, Albert Einstein College of Medicine, New York, New York, UNITED STATES|.

Introduction: Castleman's disease is a rare lymphoproliferative disorder in childhood. It may involve any extra-thoracic site where lymphoid tissue is present, and therefore can be very difficult to diagnose due to its variable

presentations. We report a case of a 16 year-old female with an incidental finding of an abdominal mass, which ultimately proved to be Castleman's disease on histopathology. Case Description: A 16 year-old morbidly obese female presented with a history of three months of nausea, vomiting and weight loss, which began during a trip to Africa. The initial work-up was negative, including chemistries, liver function tests, pancreatic enzymes, and stool cultures. An abdominal ultrasound was subsequently obtained. It showed a mildly enlarged, but otherwise normal liver, as well as a normal gallbladder and pancreatic body and head. However, a rounded, hypoechoic lesion was identified in the region of the pancreatic tail. As the study was limited by the patient's body habitus it was not possible to determine whether the lesion was cystic or solid. Initial differential diagnosis included a pancreatic pseudocyst, a mesenteric cyst and other gastrointestinal and mesenteric neoplasms. On a computed tomography scan the lesion was solid, 6 cm in length, with heterogeneous contrast enhancement. At this point, the differential diagnosis included a gastrointestinal stromal tumor (GIST), a solid pseudopapillary tumor of the pancreas (SPEN), a neuroendocrine tumor, as well as mesenteric fibromatosis and an inflammatory myofibroblastic tumor. In addition, the enhancement pattern of the tumor was very similar to what was seen in the spleen, so the possibility of an accessory spleen was raised. However a sulphur colloid liver/spleen scan did not demonstrate uptake in the mass. An attempt to perform a EUS was unsuccessful and an MRI/MRCP did not contribute additional information for better characterization of the lesion. There were no additional masses or lymph nodes. Further workup, such as markers of malignancy, HIV and TB were also negative. During the last stages of the evaluation, the patient's symptoms resolved. The mass was excised and intra-operative frozen section was consistent with a large lymph node. Further histopathological evaluation revealed reactive lymphadenopathy, consistent with hyaline vascular Castleman's disease. Conclusion: The radiologic evaluation of epigastric masses may be challenging as it is often difficult to determine the origin of the lesion. The differential diagnosis is wide and includes a multitude of inflammatory and neoplastic lesions. Although Castleman's disease is very rare, it should be considered in the case of a solid, enhancing mass. This would be typically an incidental finding with an excellent prognosis. Nevertheless, the final diagnosis is confirmed on pathology

206 *COLONIC VASCULAR MALFORMATION IN PATIENT WITH OCULAR ALBINISM*. M. Wyneski, S. Kassabian, R. Garcia, Pediatric Gastroenterology, Akron Children's Hospital, Akron, Ohio, UNITED STATES|C. Seif-Pasquarella, Pediatric, Akron Children's Hospital, Akron, Ohio, UNITED STATES|O. Soldes, Pediatric Surgery, Akron Children's Hospital, Akron, Ohio, UNITED STATES|R. Novak, Pathology, Akron Children's Hospital, Akron, Ohio, UNITED STATES|.

10 year old Caucasian male with history of ocular albinism presents with a 6 year history of recurrent rectal bleeding and anemia requiring blood transfusion for a minimum hemoglobin of 6.2 g/dL. The history includes episodes of painless hematochezia every 6 to 8 weeks. There was no history of fever, weight loss, constipation, abdominal pain, oral lesions, rash, or perianal disease. On colonoscopy, a prominent non-bleeding intramural vein was identified in the region of the splenic flexure that extended to the cecum and terminal ileum. An extensive workup including multiple endoscopies, capsule endoscopy, tagged red blood cell scans, Meckel's scan, ultrasound of abdomen and liver with Doppler, CT angiogram and magnetic resonance angiography were performed which did not definitively define the source of the bleeding. A mesenteric angiogram was performed and was notable for an extensive mesenteric venous malformation of the right, transverse, and proximal descending colon. He subsequently underwent laparoscopic assisted right, transverse and proximal descending colectomy with ileocolonic anastomosis. Pathological examination at all levels of the specimen showed vascular abnormalities which were most prominent in the serosa and submucosa but were also seen within the muscular wall and mucosa. There were areas of thin walled vessels that had the characteristics of a cavernous hemangioma," as well as groups of thin walled vessels suggestive of "lymphangioma". All the vascular channels were lined by flat bland endothelial cells consistent with a vascular malformation rather than a hemangioma. Vascular lesions of the colon are relatively uncommon and are generally grouped into lymphangioma-hemangioma and vascular malformations. We describe such a case associated with ocular albinism presenting with persistent, recurrent lower gastrointestinal hemorrhage and histopathologic findings of an admixture of capillary, cavernous, and venous structures. In such cases mesenteric angiogram is useful for identifying the cause of hemorrhage. These

fascinating cases in addition to the previous described case create the incognita of a potential association between colonic venous malformation and ocular albinism."

207 *TWO GENETIC VARIANTS IN APC IN A SINGLE FAMILY: A RARE CASE SERIES AND LITERATURE REVIEW*. F. Tubito, L. Small, F. Otaki, B. McClure, S. Lipkin, New York Presbyterian Hospital/Weill Cornell Medical Center, New York, New York, UNITED STATES].

Familial adenomatous polyposis (FAP) is a hereditary colorectal cancer (CRC) syndrome characterized by the development of hundreds to thousands of colorectal adenomas. FAP is usually caused by one autosomal dominantly inherited deleterious variant in APC. To our knowledge, no individual or family has been reported with more than one predicted deleterious genetic variant in APC. We report a case of FAP through two generations of a family with two genetic variants in APC: p.S457* and p.I1307K, in which only one family member carries both in trans. This is the first report to describe the associated compound heterozygous phenotype, which is similar to that of classic FAP. This report also adds to the knowledge of the pathogenesis of classic FAP through an 8 year-old carrier of APC p.S457*'s phenotype and of APC p.I1307K through a 19 year-old carrier's phenotype. The 8 year-old underwent cancer surveillance and was found to have multiple colorectal polyps, which supports surveillance for FAP-associated gastrointestinal malignancies beginning before the age of 8 years. The first to have genetic testing in this family was the individual that carries both APC variants; all of his first-degree relatives were at-risk of carrying each variant. Current genetic testing protocols call for pre-symptomatic testing of at-risk family members of individuals with FAP to undergo single-site analysis for the deleterious familial mutation. It is unlikely that it would have been discovered that family members were at risk of and could be tested for APC p.I1307K if the 8 year-old without this variant was tested first. Therefore, current testing protocols may not always be ideal as a family with two APC variants is reported and the incidence of this is unknown. Overall, this case study reports a novel compound heterozygous APC genotype and its segregation through a family.

208 *GASTROINTESTINAL HEMORRHAGE AS A COMPLICATION OF SHORT BOWEL SYNDROME*. K. Suryawala, M. Steele, Pediatric Gastroenterology, Oklahoma University Health Science Center, Oklahoma City, Oklahoma, UNITED STATES].

Introduction Short bowel syndrome (SBS) resulting from small bowel resection, is the most common cause of intestinal failure in children. Perianastomotic ulceration (PAU) is a rare complication after small bowel resection with only few reported cases in the literature. Complications of PAU include gastrointestinal hemorrhage and iron deficiency anemia. We describe 3 patients with iron deficiency anemia (IDA) secondary to GI bleeding who initially underwent small bowel resection with ileocolonic anastomosis in infancy. Case Presentations A male infant born with gastroschisis underwent small bowel resection with ileocolonic anastomosis at 2 months of age. At age 3, he had his first serial transverse enteroplasty (STEP) procedure for bowel lengthening. He has had recurrent episodes of GI hemorrhage requiring multiple blood transfusions since age 5. During an enteroscopy performed at the time of second STEP procedure, he was diagnosed with 7 angiodysplastic lesions which were oversown. Despite this, he continued to have recurrent GI bleeding. One year later, a colonoscopy and capsule endoscopy revealed non bleeding linear ulcer at ileocolonic anastomosis site. He underwent surgical resection of ileocolonic anastomotic ulcer and is currently stable. 11 year old female born with gastroschisis underwent small bowel resection and ileocolonic anastomosis at birth. At age 5, she developed significant IDA related to occult GI hemorrhage. Initially, multiple radiological and endoscopic modalities failed to reveal cause of GI bleeding. At 9 years, a capsule endoscopy revealed non-bleeding lineal ulceration at the site of ileocolonic anastomosis. She is currently stable on iron supplements. 7 year old female born with gastroschisis underwent small bowel resection and ileocolonic anastomosis at birth with 3 subsequent STEP procedures. Since age 5, she has experienced IDA associated with GI bleeding. Recent colonoscopy has revealed ulceration at the anastomosis site. She is currently stable on iron supplements. Discussion The perianastomotic ulceration seen in our patients after an ileocolonic anastomosis in infancy manifested with severe iron deficiency anemia and gastrointestinal hemorrhage. The cause of anastomotic ulcer in short bowel syndrome is not entirely clear. Ulcers may develop at the site of previous bowel resection due

to hypersecretion of gastric acid or poor tissue perfusion. Because of the delayed presentation, clinicians should be aware of the possibility of perianastomotic ulceration in patients with short bowel syndrome with features of iron deficiency anemia associated with gastrointestinal hemorrhage. Endoscopic examination may be necessary to identify the source of bleeding and resection of anastomotic ulcer may be needed.

EOE/GERD/AERODIGESTIVE

211 *EOSINOPHILIC ASCITES AND EOSINOPHILIC ENTERITIS – A RARE PRESENTATION OF AN UNCOMMON DISEASE.* T.R. Adamiak, M.J. Jensen, Sanford Children's, Sioux Falls, South Dakota, UNITED STATES].

Introduction: Eosinophilic ascites is rarely encountered in children. When present, the differential includes parasitic infections, tuberculosis, vasculitis, hypereosinophilic syndrome, and malignancy, among others. Eosinophilic gastroenteritis is also a consideration, particularly if the eosinophilic infiltration involves the subserosal layer. We report a 10-year-old girl who presented with ascites, elevated WBC with significant peripheral eosinophilia, and diffuse small bowel wall thickening on Abdominal CT, whose evaluation proved consistent with eosinophilic enteritis. Case History: A 10-year-old girl with history of eczema was admitted to the hospital with a 1-month history of intermittent abdominal pain, vomiting, and diarrhea. She had increasing abdominal distension in the week prior to presentation. Labs were notable for elevated WBC 18.2 with 50% eosinophils. She had normal electrolytes, hepatic panel, ESR, CRP, celiac panel, IgE level, and urinalysis. PT/INR were mildly elevated. Abdominal Ultrasound + Doppler showed ascites, but was otherwise normal. Abdominal CT showed marked ascites and diffuse small bowel wall thickening. Paracentesis was consistent with eosinophilic ascites, including 7291 nucleated cells with 84% eosinophils, culture was negative. Upper endoscopy and colonoscopy were notable for marked erythema in the terminal ileum. Biopsies showed 1-9 eos/HPF in the mid-esophagus, 27 eos/HPF in the distal esophagus, mild chronic gastritis, terminal ileum with up to 70 eos/HPF, normal duodenum and colon biopsies. Stool studies were negative for infections. Toxocara Ab, Strongyloides Ab, and serum quantiferon were all negative. The patient was diagnosed with eosinophilic enteritis and started on Prednisolone 1mg/kg/day, Singulair, and Prevacid. She continued eating a regular diet. Her CBC was normal 4 days after starting Prednisolone, with WBC 10 and 3% eosinophils. Follow up MR Enterography 3 weeks after starting Prednisolone showed much improvement in the small bowel wall thickening and enhancement. She was weaned off Prednisolone over 8 weeks, and clinically did very well, with normal CBC and PT/INR. Repeat upper endoscopy and colonoscopy were visually normal. There was chronic gastritis present, but otherwise the biopsies were all normal, without any eosinophilia. The patient has continued to feel well, with normal labs. Conclusion: Eosinophilic enteritis can present with eosinophilic ascites, elevated WBC with peripheral eosinophilia, and small bowel wall thickening. In patients with eosinophilic ascites, parasitic infection should be ruled out prior to starting corticosteroid treatment for eosinophilic enteritis. Treatment with corticosteroids can improve clinical symptoms, labs, and histology. Possibly Singulair may also have added benefit. A high index of suspicion is necessary to diagnose this rare condition in pediatric patients.

213 *EOSINOPHILIC GASTROENTERITIS AND COLITIS: POSSIBLE PRECURSORS OF INFLAMMATORY BOWEL DISEASE IN PEDIATRIC PATIENTS.* J. Barry, Pediatrics, Cleveland Clinic Children's, Univeristy Heights, Ohio, UNITED STATES][L. Mahajan, Gastroenterology, Cleveland Clinic Children's, Cleveland, Ohio, UNITED STATES].

Eosinophilic gastrointestinal (GI) disorders (e.g. esophagitis, gastroenteritis and colitis) are characterized by eosinophil-rich inflammation in the absence of other known causes for eosinophilia (e.g. drug reactions, parasitic infections, and malignancy). GI mucosal eosinophilic activation has been identified in patients with inflammatory bowel disease (IBD). Until now, these diagnostic entities have been defined as separate conditions. We report 3 patients who were initially diagnosed and treated for primary eosinophilic GI disorders that eventually exhibited clear cut endoscopic and pathologic evidence of IBD. Patient 1, a Caucasian female, presented at 8 years of age with recurrent abdominal pain and emesis. Endoscopic evaluation confirmed eosinophilic esophagitis and gastritis. Symptoms were controlled for years on PPI therapy, oral cromolyn and pulsed corticosteroids. Endoscopy showed gradual clearance of eosinophils from the upper tract by age 12. At 20 years of age her abdominal pain progressed

and she developed hematochezia. Left sided colonic biopsies showed expansion of the lamina propria by chronic inflammatory cells, crypt architectural distortion and crypt abscesses, consistent with IBD. She is currently symptomatic on immunomodulator therapy and will start biologic therapy. Patient 2, a Caucasian male, presented at 10 years of age with dysphagia, recurrent abdominal pain and diarrhea. Proximal and distal esophageal biopsies showed >20 eosinophils per high power field (hpf); eosinophils were present in the lamina propria of gastric and small bowel biopsies. He was maintained for years on PPI therapy, montelukast, dietary elimination and a 5-ASA with a diagnosis of eosinophilic esophagitis and colitis. At the age of 16, he developed increased abdominal pain and hematochezia. Repeat endoscopy showed clearance of eosinophils with findings of chronic active colitis. Therapy was gradually escalated and he is currently maintained on biologic therapy. Patient 3, a Caucasian male, presented at age 11 with reflux symptoms, diarrhea and weight loss. Pathology on upper endoscopy showed rare intraepithelial eosinophils within the squamous esophageal mucosa. Colonic biopsies showed distorted crypt architecture with eosinophil maximum density within the cecum and rectum of 100/hpf and 95/hpf, respectively. Rare foci of eosinophilic crypt abscesses/ cryptitis were noted within the transverse and descending colon biopsies. Subsequent stool studies for parasites were negative. He was treated empirically with antihelminthic therapy, placed on a steroid taper, and did well for a year on 5-ASA therapy when symptoms recurred. Repeat biopsies showed clearance of eosinophils and were consistent with IBD. He is currently maintained on biologic therapy after initial immunomodulator failure. These cases may suggest a common etiology or inflammatory mediator in pediatric eosinophilic gastrointestinal disorders and IBD. Long term studies are needed in pediatric patients with eosinophilic gastrointestinal disorders to better determine the risk for development of IBD in this patient population and help guide endoscopic surveillance and therapy.

215 *PNEUMOPERICARDIUM, CARDIAC TAMPONAD AND ESOPHAGOPERICARDIAL FISTULA AS A COMPLICATION OF CAUSTIC INGESTION.* T. Ciecieręga, Pediatrics , NYP-Weill Cornell Medical College, New York, New York, UNITED STATES|M. Sultan, S. Shweiki, S.A. Rumeleh, N. Hijeh, Pediatrics , Makassed Hospital, Jerusalem, PALESTINE, STATE OF|R. Zarnegar, Surgery, NYP - Weill Cornell Medical College, New York, New York, UNITED STATES|.

Introduction: Caustic ingestions are usually rare but can lead to serious complications. In adults, esophagopericardial fistula is a rare complication of both benign and malignant esophageal diseases. Because it is often associated with extremely high mortality rates, early recognition and proper therapy by pericardial drainage is of paramount importance for the successful management of esophagopericardial fistula. We present a case of a 5 year old male who developed uncommon complication post-causing ingestion. To our knowledge, this is the second case of pediatric esophagopericardial fistula secondary to a caustic ingestion presenting with pneumopericardium and cardiac tamponade. Case Report: A 5 year boy was transferred from an outside facility 4 days after ingesting concentrated drain cleaner powder. He had severe airway edema requiring initially mechanical ventilation. An upper endoscopy was not performed as he was transferred 4 days after the ingestion which would increase his risk of esophageal perforation. He was managed conservatively with subsequent introduction of liquid oral feedings. Feedings were advanced. The patient was discharged in good general condition one week after admission. He tolerated full oral intake at discharge. One month later he presented with increased work of breathing, pallor and low grade fever. His chest Xray showed large pneumopericardium. Physical exam revealed generalized pallor, distended neck veins and distant heart sounds. He had tachycardia with normal oxygen saturation and blood pressure. Urgent cardiac echo confirmed the pneumopericardium with compressed right ventricle suggestive of early cardiac tamponade. Pericardial drain was placed urgently and pneumopericardium has resolved. Broad spectrum antimicrobial therapy was started. Two days later Gastrografin esophagram showed narrowing in the distal esophagus with extravasation of contrast indicative of esophageal perforation. A gastrostomy tube was placed for feeding. He is currently awaiting surgical repair of his esophagus. Conclusion: Ingestion of caustic substances in children became rare occurrence as of late secondary to active prevention and education. Acquired esophageal perforation in pediatrics is rare, but can have a life threatening complication of pneumopericardium and cardiac tamponade. High index of suspicion with early recognition and prompt therapy is life saving.

221 *VASCULAR RING IN AN ADOLESCENT PATIENT WITH NAUSEA AND MILD DYSPHAGIA*. K. Kametz, Pediatric Gastroenterology, Lehigh Valley Hospital, Bethlehem, Pennsylvania, UNITED STATES].

Case: 13 year old male presented to the outpatient pediatric gastroenterology office for approximately eight months of early morning nausea, associated with several episodes of intermittent non-bloody non-bilious emesis. Symptoms were not associated with food or activity. He had mild improvement of nausea with omeprazole 20 mg once daily that was prescribed by his primary care physician. He complained of mild dysphagia with breads, but otherwise denied abdominal pain, dyspepsia, abnormal bowel movements, weight loss or change in appetite. Laboratory testing was unremarkable and included the following: TTG IGA, IGA, TSH, T4, lipase, amylase, chemistry panel, complete blood count and sedimentation rate. Omeprazole 20 mg was increased to twice daily and barium esophagram was ordered that showed a right-sided aortic arch with aberrant left subclavian artery. He was referred to pediatric cardiology who performed a cardiac MRI that confirmed the presence of a vascular ring consisting of the right aortic arch, the left subclavian artery and a left ligamentum compressing on the esophagus and trachea. The patient is scheduled for surgery to ligate and divide the ring. Conclusion: Vascular rings are usually considered in younger children and infants who present with respiratory difficulties or stridor, but it is important to consider them in older pediatric patients with dysphagia. In this case, the presenting symptom was nausea, but in careful history taking it was discovered that the patient also had mild dysphagia which prompted the barium esophagram to be ordered. In the setting of mild dysphagia, even in an older child, barium esophagram should be considered to rule out significant anatomic abnormalities such as vascular rings which was seen in this particular patient.

224 *GASTROESOPHAGEAL ADENOCARCINOMA IN AN 18 YEARS OLD PATIENT WITH CHROMOSOMAL ANOMALY*. G. Gershman, S. Yusing, Pediatric Gastroenterology, Harbor UCLA, Los Angeles, California, UNITED STATES|J. Lasky, E. Panosyan, M. Ruiz, Pediatric Hematology Oncology, Harbor UCLA, Torrance, California, UNITED STATES|M.J. Sun, UCLA David Geffen School of Medicine, Los Angeles, California, UNITED STATES|C. Kurihara, G. Kolchugina, V. Stepensky, Department of Pediatrics, Harbor UCLA, Torrance, California, UNITED STATES].

Gastroesophageal carcinoma is a rare entity in the pediatric population and has been presented in medical literature through only a few case reports. Here, we present the case of an 18 year old male with a chromosomal deletion who was subsequently diagnosed with metastatic gastroesophageal carcinoma. An 18 year old male with chromosome 14q22 deletion presented to our hospital with 3 weeks of non-bloody and non-bilious emesis occurring a few minutes after ingestion of solids and liquids. He had a history of 10 to 15 pound weight loss over one month period and one week of loose stools. There was no prior history of gastroesophageal reflux or corrosive ingestion, and patient denied use of alcohol, drugs, and tobacco. There was no family history of gastrointestinal disease or cancer. His physical exam was notable for several dysmorphic features including significant microcephaly, microtia, and hirsutism but otherwise without significant clinical findings. During his first admission, a CT of the abdomen and pelvis showed intussusception of the small bowel which resolved before any medical or surgical intervention was undertaken. Resolution of the intussusception was confirmed by ultrasound and the patient was discharged home with strict return precautions after being able to tolerate a regular diet. He was readmitted 3 days later with recurrence of non-bilious emesis, epigastric abdominal pain, and underwent diagnostic laparoscopy, as well as an intra-operative EGD. EGD showed an ulcerated circumferential mass at the gastroesophageal junction (Figure 1). Histologic analysis of biopsy specimens from the mass was consistent with poorly differentiated adenocarcinoma with squamous features that was human epidermal growth factor receptor 2 (HER2) positive. MRI and repeat CT imaging showed multiple liver lesions suggestive of metastases. Helicobacter pylori antibody was negative. Lynch syndrome genetic testing and assay for Fanconi anemia were negative, and further diagnostic workup was declined by the family. Due to progressive dysphagia, a gastrostomy tube placed which was mainly used for gastric decompression as well as a central venous catheter for total parenteral nutrition (TPN) administration. Given the metastatic lesions in the liver, and poor prognosis associated with adenocarcinoma, the family elected to forgo further therapies and pursue palliative care for the patient. Discussion: Gastroesophageal carcinoma is a rare disease of childhood. Mean age of diagnosis is 67 years¹.

Environmental factors have been suggested to be a predominant role in gastro-esophageal carcinogenesis, but typically with prolonged exposures and latent periods. In addition chronic reflux, Fanconi anemia, in which the testing in our patient was negative, chromosomal losses of 4q, 5q, 9p, 18q are considered risk factors for developing gastro-esophageal carcinoma^{1,2}. Here, we report for the first time, an incidence of gastroesophageal carcinoma in an individual with a deletion in chromosomal 14. Pathogenesis remains unclear, and further investigation of etiology and management of this disease in a multidisciplinary approach is essential.

226 *THINKING OUTSIDE THE LUMEN: LEIOMYOMATOSIS CAUSING LINEAR ESOPHAGEAL ULCERATION*. N. Mendez, T. Webster, T. Weinstein, J. Markowitz, Pediatric Gastroenterology, NS-LIJ Cohen Children's Medical Center, Lake Success, New York, UNITED STATES |.

Introduction: Leiomyomatosis is a known, albeit seldomly diagnosed, phenomenon in children with a history of Alport's syndrome. Diffuse leiomyomatosis is a progressive condition characterized by benign smooth-muscle cell proliferation. More commonly it affects the esophagus; however, it may also involve the tracheobronchial wall, distal gastrointestinal tract, or uterus. Case Report: A 12 year old female who was diagnosed with Alport's Syndrome at 2 years of age via kidney biopsy, presented with heartburn and progressively worsening epigastric pain for the past 4 years. This burning sensation was worse when lying flat and was associated with coughing while eating. Cough would intermittently wake her from sleep. Sleeping upright would alleviate pain and improve her cough. She would intermittently have emesis, approximately three times per month that was non-bilious and non-bloody. She denied weight loss, mouth ulcers, fever, and diarrhea. Physical examination was unremarkable. Laboratory studies including CBC, CMP, ESR, CRP, TSH, free T4, quantitative IgA and celiac panel were within normal limits. Upper endoscopy showed a thin, linear ulceration in the distal esophagus without any active bleeding. The ulcer was approximately 2 cm in length and ended at the gastro-esophageal junction. Gastric and duodenal mucosa were grossly normal in appearance. Pathology was only remarkable for non-specific and reactive changes of the squamous epithelium and stroma that were consistent with changes immediately adjacent to an ulcer. There was no evidence of fungi or viral cytopathic effect. Following these results, she was started on Omeprazole 40mg by mouth daily. Given the unusual appearance of the ulcer, an esophagram was obtained that demonstrated a smooth posterior and left lateral rounded C-shaped indentation upon the distal esophagus with a mild delay in progression of contrast into the stomach. Subsequent noncontrast MRI of the chest revealed diffuse thickening of the distal 9.5 cm of the esophagus. The esophageal wall measured upwards of 1.8 cm in thickness with an associated narrowed lumen. The wall was noted to be hypointense on T1 and T2 weighted images. Findings were concerning for esophageal leiomyomatosis given her history of Alport's Syndrome, however, a neoplastic or inflammatory process could not be excluded. While awaiting surgical evaluation, the patient experienced increased emesis and developed dysphagia to solids. Shortly thereafter she underwent an Ivor Lewis esophagogastrectomy. Pathology specimens of the distal esophagus and stomach following the resection showed leiomyomatosis involving the wall of the distal esophagus, gastroesophageal junction, and proximal stomach (up to 8.0 x 6.5 x 3.5 cm). Conclusion: This case outlines the importance of recognizing that uncommon conditions can be the cause of common symptoms such as reflux and esophagitis. In particular it highlights that fact that extramural disease can cause mucosal findings.

227 *A PEDIATRIC CASE OF SLOUGHING ESOPHAGITIS*. M. middelhof, L. Saubermann, Pediatric Gastroenterology & Nutrition, University of Rochester Medical Center, Rochester, New York, UNITED STATES |.

A 16-year-old female patient with no significant medical history who presented with symptoms of "throat burning", chest pain, and sniffing associated with a chronic cough. She was initially started on Ranitidine by her PCP, and then Omeprazole following an evaluation by an ENT and concern for laryngeal reflux. There was no improvement in her symptoms on the proton pump inhibitor and she was subsequently referred to the Pediatric GI clinic. An EGD was then scheduled and performed to evaluate her for possible reflux esophagitis or eosinophilic esophagitis. The EGD was normal except for circumferential thickening and ringed appearance of the distal most portion of the esophagus. The remainder of the esophageal mucosa appeared normal. Histopathologic diagnosis demonstrated

that the gastro-esophageal junction had squamous epithelium with abundant parakeratosis with conspicuous intraepithelial bullae filled with inflammatory cells. No fungal organisms were seen with GMS stain. The proximal esophageal biopsy was unremarkable. The histologic findings were unusual but felt to be most consistent with so-called "sloughing esophagitis". EDS is a rare lesion characterized by superficial necrotic squamous epithelium and endoscopic plaques or membranes. EDS is most commonly seen in association with desquamating dermatologic disorders, particularly pemphigus vulgaris, but it has been reported in association with ingestion of oral bisphosphonates, non-steroidal anti-inflammatory drugs, doxycycline, quinidine, celiac disease, as well as complication of rigid endoscopy with esophageal dilation and following the accidental ingestion of fish bone. According to literature review EDS affects older, debilitated patients on polypharmacology. To the best of our knowledge this is the first pediatric case report of esophagitis dissecans superficialis. Our patient responded to the therapy of proton pump inhibitor (omeprazole 40 mg daily for 4 months). It is possible that EDS was an incidental finding and the patient's symptoms were secondary to gastroesophageal reflux disease. Repeat UGI endoscopy did not reflect sloughing mucosa and histopathologic findings were negative as well. The natural course of EGDS appears benign with resolution of symptoms."

228 *A CASE REPORT ON A 17-MONTH-OLD MALE WITH A RARE CONGENITAL MUSCULAR RING.* J. Migliuri, J. Hollon, J. Devito, R. Ricca, Pediatrics, Naval Medical Center Portsmouth, Suffolk, Virginia, UNITED STATES | .

A 17-month-old male with refusal to transition to solid food was evaluated for presumed esophageal dysphagia. He was found to have a rare congenital muscular ring that was successfully treated with thoracoscopic myotomy. His initial evaluation included an esophagram which demonstrated a lower esophageal stricture and subsequent endoscopy revealed food just proximal to the stricture. After ruling-out eosinophilic esophagitis, attempts to dilate the stricture were unsuccessful and resulted in lumen-occluding contractions during peristalsis, distinguishing it from a stricture or mucosal ring. A computed tomography scan confirmed a focal circumferential hypertrophic muscular ring 2 cm proximal to the gastroesophageal junction. Robot-assisted thoracoscopic esophageal myotomy was performed, in combination with intraoperative endoscopic visualization. The patient tolerated the surgical release well and is no longer having symptoms. This case study describes a novel approach for a rarely encountered congenital anomaly.

INFLAMMATORY BOWEL DISEASE

244 *POST OPERATIVE RECURRENCE IN PEDIATRIC CROHN DISEASE.* M.A. Conrad, R. Baldassano, N. Dawany, P. Mamula, P. Mattei, E.C. Maxwell, L. Albenberg, J. Kelsen, GI, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES | .

Background: Despite the use of anti-TNF therapy, immunomodulators, and antibiotics, pediatric patients with Crohn disease have a high risk of requiring intestinal resection. However, surgical resection is not curative and historically, greater than 50% of patients experience post-operative recurrence and require additional surgery. Risk factors for early recurrence of disease post operatively have not been well described in the pediatric population, particularly since the use of biologic therapy has been introduced. The primary objective of this study was to describe a single center cohort of pediatric patients who underwent ileocecal resection for Crohn disease. Methods: This was a retrospective chart review of pediatric patients with Crohn disease who underwent surgical resection at a pediatric tertiary care center, from Jan 1998-2013. Medical records were reviewed for baseline demographic data, clinical variables, laboratory and radiology studies, endoscopy, therapy, surgical interventions, and disease or medication related complications. Data were obtained for 6 months prior to resection, and again 6 months, 1 year, and 3 years post surgery. Exclusion criteria included surgical intervention for the indication of ulcerative colitis, indeterminate colitis, perianal disease, abscess drainage, ostomy revision, or stictureplasty. Results: 150 pediatric patients with Crohn disease who underwent intestinal resection during the study window and followed by pediatric gastroenterology at CHOP were identified. Of the 53 subjects who were analyzed thus far and had complete data available, 42% were female, and the average age at diagnosis was 13+/- 3 years. 65% of patients had ileocolonic Crohn disease of which 23% had ileal and limited cecal disease. Forty patients had

ileocectomy and the remainder of surgical procedures included ileal resections. Indications for surgery included medically refractory disease, growth failure, fibrotic stricture with obstructive symptoms, and penetrating disease. Length of follow up was 3 years. Average PCDAI at the time of surgery was 27. The average time from diagnosis to surgery was 2.27 +/- 2.6 years, with 9 patients whose surgery was performed at diagnosis of Crohn disease. Twenty-five patients were initiated on biologic therapy prior to surgery, although the median time from initial dose of biologic therapy to surgery was 5.4 months. Of these patients, 18 (72%) continued biologic therapy post-operatively. The recurrence rate based on follow up endoscopy among these patients was 67% (12 patients). Nine patients were first started on biologic therapy post-operatively. Of these 56% (5 patients) had recurrence of disease on endoscopy. Three patients underwent a second resection, all were treated with biologic therapy following the initial surgery. Conclusions: Pediatric patients have a high risk of disease recurrence following small bowel resection. Additionally, based on the short interval from initiation of biologic to surgery, surgical intervention may be a potential therapy to induce remission in high risk patients. A larger cohort is necessary to further define predictors of disease recurrence and effect of biologic therapy, which is currently in process.

245 *PULMONARY, HEPATIC, AND SPLENIC LESIONS AS AN INITIAL MANIFESTATION OF CROHN'S DISEASE.* M.

Crespo, V. Baez-Socorro, T.J. Sferra, Pediatric Gastroenterology, Hepatology, and Nutrition, University Hospitals-Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES|.

Introduction: Crohn's Disease (CD) is a systemic disease with up to 25% of patients presenting with extraintestinal manifestations. Nodules and aseptic abscesses within multiple visceral organs is a rarely reported manifestation of inflammatory bowel disease. Case Report: An 11-year-old female was in good health until a month prior to presentation when she developed fevers, abdominal pain, and weight loss. She had hypoalbuminemia, anemia, and elevated inflammatory markers. Infectious workup was negative including multiple blood cultures and evaluation for fungal (*Histoplasma*, *Blastomyces*, *Coccidioides*, *Aspergillus*), viral (HIV, EBV, CMV), and bacterial pathogens (*Bartonella*, *Mycobacteria*, *Treponema Pallidum*, *Brucella*). Rheumatologic evaluation was negative including myeloperoxidase antibody, proteinase-3 antibody, ANA panel, angiotensin converting enzyme, lysozyme, and ferritin. An eye exam and echocardiogram were normal. Computed tomography (CT) of the chest and abdomen showed multiple pulmonary, hepatic, and splenic lesions. She underwent bone marrow and splenic lesion biopsy. The bone marrow was normocellular with mild granulocytic hyperplasia. The splenic lesions were necrotic tissue with chronically inflamed surrounding fibrous capsules. Initial esophagogastroduodenoscopy and colonoscopy with biopsies were normal. Capsule endoscopy demonstrated a small jejunal polypoid lesion. The fevers persisted and, even though an infectious pathogen was not identified, she received a course of intravenous antibiotics without improvement in symptoms. Positron emission tomography (PET-CT) revealed uptake in the lungs, spleen, and liver. Almost 2 months after presentation, she underwent repeat EGD with push enteroscopy and colonoscopy. These studies demonstrated a jejunal polypoid lesion and small colonic aphthous ulcers. The terminal ileum was grossly normal. The intestinal biopsies were significant for lymphoid aggregates within the small bowel mucosa and diffuse focal active colitis. The polypoid lesions was described as lymphoid aggregates. The terminal ileal mucosa was normal. She was placed on solumedrol for presumed inflammatory bowel disease and was transitioned to oral prednisone. Her symptoms improved. The pulmonary, liver, and splenic nodules completely resolved on CT 2 months after initiation of steroid therapy. She was tapered off steroids and a week after completion of therapy developed diarrhea and abdominal pain. Repeat endoscopy revealed grossly normal mucosa. Random biopsies showed gastritis, increased mononuclear cells within the small bowel lamina propria and epithelium and chronic active ileitis. The colonic mucosa was normal. Prednisone was re-started and she was started on immunomodulators. Her symptoms improved and she has been maintained on immunomodulator therapy alone. Subsequent endoscopic evaluation revealed colonic mucosal architectural distortion. Discussion: Systemic involvement in IBD has been well described. Extraintestinal manifestations can occur in up to 25% of pediatric patients with IBD. Similar but scarce reports suggest Crohn's disease as a cause of pulmonary, hepatic, and splenic lesions as first manifestations of the disease.

247 *ORAL SERUM-DERIVED BOVINE IMMUNOGLOBULIN THERAPY TO HELP ACHIEVE CLINICAL REMISSION WITH ASSOCIATED DECREASES IN FECAL CALPROTECTIN IN A PEDIATRIC ULCERATIVE COLITIS PATIENT.* B.P. Burnett, Medical Affairs, Entera Health, Inc., Cary, North Carolina, UNITED STATES| M. Dave¹, Gastroenterology, Texas Digestive Disease Consultants, Plano, Texas, UNITED STATES|.

Current treatment goals in pediatric ulcerative colitis (UC) include: achieving clinical remission with minimal side effects or affecting growth and development while maintaining a normal quality of life. Drugs like steroids can lead to long-term side effects in children with UC. The early use of 5-ASA and immunomodulators are recommended for induction therapy for mild/moderate UC before anti-TNF therapy. Of children with UC ~50% become steroid refractory or dependent by 1 year even with use of immunomodulators and infliximab. Safe, effective therapies are needed for pediatric UC patients. Serum-derived bovine immunoglobulin/protein isolate (SBI) is a prescription medical food product intended for the clinical dietary management of IBD. SBI has a multifaceted mechanism of action (MOA) to help manage gut barrier function by binding to microbial components in the GI tract which may lead to reduced immune activation and better nutrient utilization. A 13-year-old female patient (49.8kg, 159cm, BMI 19.7) diagnosed in 2/2010 with UC by EGD/Colonoscopy experienced hematochezia 5-10 times/day with weight loss. Initial treatment with prednisone, mesalamine and VSL#3 induced remission. The steroid was later discontinued. After one year (2/2011), she relapsed again experiencing hematochezia. A repeat EGD/Colonoscopy confirmed a UC flare. A short course of prednisone with 6-MP was added to VSL#3 and mesalamine suppositories. 6-MP was discontinued by the family to see if she could remain in remission with mesalamine and VSL#3 (7/2014). Within 2 weeks, the patient developed hematochezia 10 times/day. She refused prednisone and was placed on budesonide without effect. Budesonide was discontinued and infliximab treatment was planned. Pending insurance approval of infliximab, SBI was given at 5g BID along with 6-MP/mesalamine suppositories/VSL#3 (10/2014). Fecal calprotectin (FC) was followed before and after addition of SBI. Her symptoms improved within 2 weeks of adding SBI and by 1 month, she attained asymptomatic clinical remission. Before SBI was administered to the patient, even while on 6-MP/mesalamine suppositories/VSL#3, her FC levels were 1717 mcg/g. After starting SBI, the patient's FC decreased to 588.4 mcg/g after 2 weeks, 211 mcg/g in 2 months and <15 mcg/g by 1/2014. The patient is now maintained in clinical remission on SBI at 5g BID/6-MP/mesalamine suppositories/VSL#3. She experiences no hematochezia or abdominal pain and FC remains low. Infliximab was avoided and steroids were not required to achieve and maintain remission. SBI, along with 6-MP/mesalamine/VSL#3, led to clinical remission with associated decreases in FC. This difficult pediatric case demonstrated the possibility of avoiding medication associated with significant adverse events in children by augmenting management of UC with SBI. SBI may provide for distinctive nutritional requirements necessary to manage pediatric UC. Further studies are warranted to assess SBI in pediatric patients with UC, but it may provide a safe option to help manage patients wanting to avoid steroids and anti-TNF therapy.

248 *PERICARDIAL EFFUSION IN AN ADOLESCENT WITH ULCERATIVE COLITIS ON INFLIXIMAB.* K. Dickinson, M. Schaefer, T. Falaiye, Pediatric Gastroenterology, Penn State Hershey, Hershey, Pennsylvania, UNITED STATES|.

Introduction: Extraintestinal manifestations of inflammatory bowel disease are widely recognized, however they rarely involve the pericardium. Inflammatory pericardial effusion is even less acknowledged, especially in pediatrics where few case reports exist. Pericardial effusion can also be an adverse effect of two common therapies for IBD, sulfasalazine and infliximab. This makes differentiating the etiology difficult. Nonetheless, this rare complication is important to recognize in children with IBD who present with chest pain. Case Report: The patient is a 17-year-old female with ulcerative colitis initially treated by an outside provider with mesalamine and prednisone. She discontinued mesalamine due to arthralgias and edema. Sulfasalazine did not improve her arthralgia. Thus, infliximab infusions were started at 5mg/kg in addition to prednisone, budesonide and sulfasalazine. Her hematochezia and stooling frequency improved on infliximab which continued for four months. The patient's maintenance interval was decreased to seven weeks and total infliximab dose was increased due to weight gain. After an infusion of the increased dose, she developed shortness of breath and chest pain. Symptoms improved initially but recurred two weeks later. She presented to an outside ED with sharp substernal chest pain

and exertional shortness of breath. She had no associated fevers, other symptoms, or recent illness. On admission, she was tachycardic and tachypnic with an oxygen requirement. Chest CT revealed pericardial effusion, prompting transfer to our pediatric intensive care unit. Echocardiography confirmed large circumferential pericardial effusion with diastolic right atrial and ventricular free wall collapse. 297 mL of serous fluid was drained, the cultures of which were negative for fungi, viruses, acid fast bacilli, anaerobic and aerobic bacteria. Fluid cytology had 1884 nucleated cells with composition of: PMNs 10%, Lymph 17%, Baso 6%, Eos 40%, Mono 14%, reactive mesothelial cells 13%. Glucose was normal and Light's criteria did not indicate exudate based on LDH and protein. Pediatric rheumatology was consulted for consideration of drug-induced lupus, which was discussed as unlikely due to lack of other symptoms as well as ANA of <1:40 and rheumatoid factor <9. Discussion: There are case reports of adults with pericardial effusions on infliximab or sulfasalazine. Sulfasalazine-associated cardiac reactions typically occur within two weeks of initiation, whereas our patient was on this medication for months. Drug-induced lupus-like illness is associated with elevated ESR, positive ANA and gradual onset of fevers, arthralgias, and arthritis, none of which our patient exhibited. Pericardial effusion is rare in adult IBD populations. Its occurrence has not been described as related to disease activity. Possible etiologies for pericardial effusion in this case may be drug-induced pericardial effusion or a manifestation of extraintestinal IBD. Conclusion: Although described in adults, pericardial effusion should also be considered in pediatric patients with IBD and chest symptoms, as well as pediatric IBD patients on infliximab therapy.

250 *AN INTERESTING CASE OF ORBITAL MYOSITIS IN A PEDIATRIC IBD PATIENT.* K.M. Ellery, S. Kim, Pediatric Gastroenterology, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES | M. Naddaf, Pediatric Gastroenterology, Mercy Children's Hospital, Toledo, Ohio, UNITED STATES | P. Jaggi, Pediatric Infectious Disease, Nationwide Children's Hospital, Columbus, Ohio, UNITED STATES | .

Our patient is a 12 year old Caucasian female diagnosed with ileocolonic Crohn's disease in 2012. On admission to our hospital in April 2015, she presented with left periorbital pain and swelling attributed to infection. Further evaluation revealed orbital myositis (OM) that developed after discontinuing infliximab and initiating vedolizumab therapy. She started steroids and azathioprine (AZA) for Crohn's therapy at initial diagnosis. Due to lack of clinical improvement, therapy was changed to infliximab (IFX) in conjunction with AZA, which induced disease remission. She did relatively well until late 2014; at that time, she developed aggressive alopecia and psoriasis. Severe superinfection at the sites of alopecia and psoriasis required prolonged IV antibiotic therapy for several weeks with minimal resolution. Due to significant dermatologic and infectious issues along with recurrence of Crohn's symptoms, IFX and AZA were discontinued. She subsequently started enteral therapy and restarted AZA in January 2015, which resulted in improvement of her GI symptoms. Her alopecia and psoriasis resolved after cessation of IFX. Vedolizumab was started for maintenance in March 2015. She developed acute periorbital swelling, pain, photophobia, and diplopia in late March 2015. Orbital cellulitis was diagnosed by CT scan. She received 3 weeks of antibiotics (nafcillin, meropenem, vancomycin, and metronidazole) and daily prednisone (60 mg). Despite aggressive treatment, she had waxing and waning of swelling with intermittent fevers (up to 104F). She was referred to our hospital for further evaluation. Repeat CT scan showed medial and lateral rectal muscle enlargement and possible fluid collection in the lateral rectus muscle with normal appearance of all sinuses. Lacrimal gland biopsy was negative and exploration of the lateral rectus muscle showed no abscess; thus, antibiotics were discontinued. She subsequently underwent orbital MRI, which revealed orbital myositis. She received high dose (1 gm/day) IV methylprednisolone for 5 days, followed by weekly methylprednisolone infusions (1 gm/week) for 8 weeks and a prolonged prednisone taper. She had resolution of her ophthalmologic symptoms after high dose steroids were completed. Methotrexate will be started to treat OM in consultation with Rheumatology and Ophthalmology. There are few cases in the literature reporting patients with IBD and orbital myositis. OM most commonly presents with Crohn's disease. It can present prior to the initial gastrointestinal symptoms or during disease remission. There is a report of an adult patient who developed OM 13 weeks after the cessation of IFX. Our patient developed her symptoms 15 weeks after cessation of IFX. Treatment of OM consists of high dose corticosteroids as first line therapy; followed by low dose orbital radiotherapy, methotrexate, cyclosporine, and cyclophosphamide as second line therapies. We speculate that the systemic effects of IFX

suppressed the development of OM and that changing to vedolizumab, a more gastrointestinal-specific therapy, led to emergence of extraintestinal IBD manifestations.

255 *INTRAOPERATIVE INCIDENTAL DIAGNOSIS OF CROHN'S DISEASE FOLLOWING ABDOMINAL TRAUMA*. J. Lynch, A. Rouster, B. Riedel, Pediatrics, WVU, Morgantown, West Virginia, UNITED STATES].

A 16 year old white male presented to the emergency department with RLQ abdominal pain following a crash during an ATV race. His abdomen had struck the handlebar of his vehicle as he was ejected. The pain developed shortly after crashing and continued despite family's treatment with hydration and aspirin. The patient presented to the ED where an US demonstrated free fluid in the pelvis. Abdominal CT showed intraperitoneal air, as well as free fluid. He was taken emergently for exploratory laparotomy and was found to have a 2 cm perforation of the distal ileum, as well as an extremely friable terminal ileum. The surrounding 1.5 feet of bowel showed evidence of chronic inflammation and thickening. These findings raised suspicion for Crohn's disease by the surgeon and specimens were sent for pathology. Diagnosis of Crohn's disease was made based upon the pathologic findings, which included transmural inflammation, crypt abscess formation, and focal granulomas. Upon further questioning he had experienced intermittent bouts of abdominal pain & diarrhea over the past year. Initial labs showed hypoalbuminemia, leukocytosis, markedly elevated C-reactive protein, and anemia with depressed MCV. This presentation is unique in that there is a relative dearth of cases reported in the literature of incidental diagnosis of IBD. This case illustrates the variability of disease manifestation in patients with Crohn's disease.

262 *AN UNUSUAL CASE OF PEDIATRIC INFLAMMATORY BOWEL DISEASE WITH CAVITARY PULMONARY NODULES*. N. Santucci, J. Gallois, B. Keith, LSU, Children's Hospital of New Orleans, New Orleans, Louisiana, UNITED STATES].

Background - Pulmonary manifestations of inflammatory bowel disease are a rare extra intestinal finding in children, with only 17 cases reported previously in the literature. We report a case of a 16 year old female who presented with colitis, asymptomatic pulmonary nodules and SIADH. Case History - In this 16 year old with abdominal pain and bloody diarrhea, colonoscopy showed extensive superficial ulcers in the colon, edema around the terminal ileum and biopsies showed cryptitis and crypt abscesses. Serum IBD SGI suggested indeterminate colitis. Pulmonary nodules were found on chest Xray done for tachycardia. An extensive infectious, rheumatology, and immunology work up failed to reveal the diagnosis for these nodules. Open wedge lung biopsy revealed chronic active bronchiolitis with negative viral stains. Her serum sodium consistently ranged from 128-132 despite fluid restriction. A workup revealed chronic SIADH which was secondary most likely to her pulmonary lesions. Conclusion - Cavitory pulmonary nodules in an adolescent patient would more likely be associated with infectious etiologies including mycobacteria. Bronchiectasis and bronchitis are common findings with IBD, but not bronchiolitis. Fistulizing lung disease and eosinophilic pneumonias are seen in Crohn disease (CD), while pulmonary vasculitis has been associated with ulcerative colitis (UC); however, cavitory pulmonary nodules are not a usual finding. SIADH has not been previously reported as a manifestation of pulmonary nodules related to IBD in children.

266 *SINGLE CENTER EXPERIENCE WITH VEDOLIZUMAB USE IN PEDIATRIC PATIENTS WITH INFLAMMATORY BOWEL DISEASE*. J.H. Weitzner, C. Sauer, S. Kugathasan, Pediatric Gastroenterology, Hepatology, and Nutrition, Emory University, Atlanta, Georgia, UNITED STATES].

Background: Vedolizumab is a monoclonal antibody which binds integrin $\alpha 4\beta 7$ and is approved for adult patients with Crohn's disease (CD) and Ulcerative Colitis (UC). It is limited in its off label use in pediatric patients with CD and UC. We describe the experience using vedolizumab in a pediatric tertiary IBD center. Methods: A retrospective review identified pediatric IBD patients (age <21 years) receiving vedolizumab for CD or UC for at least six weeks. Data on disease activity, demographics, and previous treatments was obtained. Disease activity was evaluated with the pediatric Crohn's disease activity index (PCDAI). Results: Nine pediatric patients, all CD, were identified as having initiated vedolizumab between July 2014 and May 2015. Vedolizumab dose was 6mb/kg/dose with maximum dose 300mg infused at 0, 2, 6 weeks, and then every 8 weeks. Mean age at diagnosis was 10.2 (range 2-

16) years and mean age at vedolizumab initiation is 16.25 (range 7-21) years. Five patients (55.6%) had prior bowel surgery. All patients (100%) had previously failed at least one anti-TNF agent. All patients have received the 6 week dose and 4/9 have received the 14 week dose. Week 6 and 14 remission rates defined by PDAI <10 were 11.1% and 25%, respectively. However, mean PDAI decreased from 38.6 at week 0 to 35 at week 6. There were no infusion reactions, or adverse events reported. All patients remain on vedolizumab. Conclusion: In our experience, vedolizumab is safe in pediatric IBD patients. The efficacy in our population is yet to be understood. These remission rates are lower than those reported thus far in adults. This decreased response rate could be due to the selection bias of studying patients who have previously failed anti-TNF therapy and have the most aggressive disease phenotypes. With increased number of patients receiving vedolizumab, we will have further insight into the efficacy of inducing remission in pediatric IBD patients.

NUTRITION

270 "IRIDA": AN UNUSUAL CAUSE OF IRON DEFICIENCY ANEMIA. E. April, Pediatrics, Laval University, Quebec, Quebec, CANADA | J. Castilloux, Gastroenterologist, Laval University, Quebec, Quebec, CANADA |.

Introduction Iron deficiency anemia is a common pediatric condition. This discovery, sometimes fortuitous, is often caused by inadequate iron intake or excessive milk consumption. When these etiologies are excluded, it is often necessary to pursue with more invasive tests in order to rule out a digestive cause. We report the case of two French Canadian siblings with unexplained iron deficiency anemia not responding to oral iron supplements in whom a rare genetic cause has been found: the IRIDA (iron resistant iron deficiency anemia). Methods and Results The brother was diagnosed at the age of 2, during an episode of febrile seizure. His hemoglobin, mean corpuscular volume, serum iron and ferritin level were low, respectively at 74, 47.1, 2 and 9. Over 4 years, he underwent multiple investigations. A small fundic vascular lesion was found at the first two upper endoscopies and was treated with argon. No other lesion was subsequently found at the 3 following upper endoscopies (normal gastric and duodenal biopsies). Hemoglobin electrophoresis was normal, but a deletion of the alpha-globulin gene revealed a minor alpha thalassemia, not explaining however the severity of microcytic iron deficiency anemia. All other investigations were normal: 2 colonoscopies (with normal biopsies), 2 capsule endoscopies, RBC labeled scan, a small bowel series, a meckel scan, abdominal ultrasound, blood lead levels, nutritional assessment, celiac disease screening, parietal cell antibodies, bone marrow aspiration and anemia screening of both parents. Although asymptomatic, his anemia did not improve with his 6 mg/kg/day of oral iron supplement. His sister was 6 months old when an iron deficiency anemia was also diagnosed. She underwent substantially the same investigations as her brother. Her anemia also remained unexplained so a genetic cause was suspected. A mutation of TMPRSS6, affecting the expression of hepcidin, a key factor of systemic iron homeostasis, allowed the final diagnosis of iron refractory iron deficiency anemia (IRIDA). Conclusion Recognizing IRIDA among the other causes of iron deficiency anemia in children is a challenge for the clinician. IRIDA must be suspected when a child with unexplained anemia does not respond to enteral iron supplements. An early diagnosis might prevent many invasive investigations and allow a faster introduction of intensive parenteral iron therapy that is the only treatment of possible efficacy.

276 TWO CASE STUDIES OF AN INTENSIVE MULTIDISCIPLINARY DAY TREATMENT PROGRAM TO ADDRESS PEDIATRIC FEEDING DISORDERS. C. Ebert, P.W. Powell, K.S. Nersessova, S. Illig, S. Benton, Feeding, Children's Hospital of Richmond at VCU, Richmond, Virginia, UNITED STATES |.

Feeding disorders in infants and children compromise the child's health in the short and long-term, decrease overall quality of life for the child and their family, and increase health care costs for the family and for society as a whole. Using two case studies, we will present the importance of using behavioral interventions in conjunction with medical and sensory interventions to decrease negative feeding behaviors. Both children participated in an intensive eight-week day treatment feeding program where they were fed three meals a day, five days a week, by an occupational or speech therapist using a behavioral protocol in consultation with a clinical psychologist. Parents were also trained in how to implement the behavioral protocol at home. Negative feeding behaviors that were

identified included negative vocalizations, expelling, pushing the presentation away, and turning head away from the presentation. Techniques used to improve feeding behaviors included positive reinforcement, specific behavioral interventions, limiting meal times, proper seating, and oral motor desensitization exercises. By the end of the program both children demonstrated decreases in negative feeding behaviors, increases in variety and volume of foods they ate, and increases in height and weight. The first case study will describe the treatment for a medically complex toddler who was dependent on g-tube feedings upon admission and by discharge was meeting most calorie needs by mouth. The second case study will describe the treatment for a school-age child diagnosed with Autism who was dependent on a complete pediatric formula to meet his nutritional needs at admission and was eating over twenty different table foods at discharge.

280 *IRON SUPPLEMENTATION IMPROVES APPETITE OF AN ANOREXIC CHILD WITH IRON DEFICIENCY ANEMIA AND PROTEIN-LOSING ENTEROPATHY INDUCED BY COW MILK. AND THE LITERATURE REVIEW.*. C. Jarasvaraparn, G.J. Fuchs, Pediatric Gastroenterology, Hepatology and Nutrition, University of Arkansas for Medical Sciences College, Little rock, Arkansas, UNITED STATES|.

Iron deficiency anemia (IDA) is a common cause of morbidity and healthcare cost among children. IDA affects 2.1% of US children 12 to 35 months of age. Protein-losing enteropathy (PLE) is a disorder characterized by abnormal profound enteric protein loss. The diagnosis of PLE is considered in children with hypoproteinemia especially after other causes have been excluded. To date, there was no report of a child with PLE and IDA without anasarca. We report a 3-year and 1 month-old child with PLE and IDA but without anasarca. The history was significant for a reported feeding aversion leading the parents to allow the child to rely on large volumes of whole cow milk for her nutrition. Laboratory evaluation showed IDA with hypoproteinemia, hypoalbuminemia and elevated fecal alpha-1-antitrypsin. Of interest, our case presented with chronic anorexia and loss of appetite. After a few days of iron supplementation, her appetite normalized and the child began eating very well. Iron status indices improved (hemoglobin, hematocrit and ferritin were 11.1 g/dl, 34.1 % and 7.4 ng/ml, respectively) after iron supplementation for 1 month. The literature describing the association of gastrointestinal blood loss and cow milk feeding is also reviewed.

290 *A UNIQUE PRESENTATION OF FANCONI SYNDROME.* E.D. Rivera Rivera, S. Kahn, T. Sentongo, Pediatric Gastroenterology, University of Chicago, Miami, Florida, UNITED STATES|.

Fanconi syndrome is described as a group of defects in the proximal tubule that leads to the inability to reabsorb electrolytes such as phosphorus, glucose, bicarbonate and amino acids among others. Here we report the case of newly diagnosed Fanconi Syndrome in a patient consulted for refeeding syndrome. A 3 year old male with a past medical history of cerebral palsy, seizure disorder, failure to thrive, G tube dependence and status post Nissen fundoplication was brought to his pediatrician's office due to dehydration. After the assessment it was decided that he needed to be hospitalized and was transferred to Comer Children's Hospital at The University of Chicago for further management. Upon arrival he was noticed to be dehydrated hypokalemic and hypophosphatemic (potassium of 2.3 mmol/L [Ref range 3.5-5.0] and phosphorus of 1.4 mg/dL [Ref range 2.5-4.4]). He was started on maintenance IV fluids which were dextrose and potassium containing and feedings were resumed, which were well tolerated. However, four hours later his electrolytes were rechecked and this time the potassium was found to be 2.0 mmol/L and the phosphorus was at 0.6 mg/dL. An electrocardiogram was immediately obtained and "u" waves were seen, which triggered immediate transfer to the Pediatric Intensive Care Unit. After that a Gastroenterology consult was requested due to concerns for refeeding syndrome. Upon evaluation of the patient, it was noticed that despite providing feeds at a very conservative rate (started at 10% of his total estimated required daily calories) and consistently replacing electrolytes potassium and phosphorus, the electrolyte abnormalities persisted. His serum bicarbonate levels were low (ranging from 13-16 mmol/L [Ref range 23-30]) and the urinalysis contained ketones and glucose. Further testing the urine for urine amino acids revealed the presence of large amounts of amino acids in the urine. The diagnosis of Fanconi syndrome was confirmed based on the presence of amino acids, phosphorus, potassium, bicarbonate and glucose in the urine, presence of rickets, growth retardation and failure

to thrive. The patient was subsequently treated with potassium, phosphorus and bicarbonate replacement as is usual in this patient population. Refeeding syndrome can be described as the phenomena that occurs when there are significant shifts in fluids and electrolytes after feeding a patient that has been chronically malnourished. This phenomena can be potentially fatal and its hallmark is hypophosphatemia which is also commonly associated with other electrolytes imbalances such as hypokalemia. The constellation of laboratory results as well as the initial clinical presentation, such as the electrolytes abnormalities, amino aciduria, rickets in the setting of normal vitamin D levels, failure to thrive, and growth retardation lead us to consider Fanconi syndrome as a potential diagnosis since it can also have other extra renal manifestations which were present in this patient. In this case we were able to unmask a diagnosis that presented as refeeding syndrome, but certainly keeping a high index of suspicion in patients that do not fit the typical clinical picture was key in this case.

294 FORMULA SWITCH LEADS TO ENTERAL FEEDING TOLERANCE IMPROVEMENTS IN CHILDREN WITH DEVELOPMENTAL DELAYS. H. Storm, Clinical Sciences, Nestle Health Sciences, Florham Park, New Jersey, UNITED STATES|G. Minor, Children's Center for GI and Nutrition, Hollywood, Florida, UNITED STATES|.

Background and Objectives: Children with developmental delays are often dependent upon enteral nutrition. Common clinical practice challenges encountered include vomiting, constipation, diarrhea, abdominal distention and inability to reach nutritionally adequate feeding volumes. The aim of our study was to evaluate the effects of switching children with developmental delays who were experiencing feeding intolerance on intact protein formulas to a 100% whey peptide-based formula frequently used for GI compromised children. **Methods:** We performed a retrospective chart review of children with developmental delays who suffered from feeding intolerance while receiving intact protein formulas. Charts of children (1-18 years of age at time of formula change) who were receiving gastrostomy tube feedings yielding $\geq 90\%$ caloric requirements and without factors influencing feeding tolerance (recent change of tube location, acute illness or infection, or documented cow's milk protein allergy) who switched from an intact protein formula to a 100% whey, peptide-based formula were reviewed. Data were collected on types of feeding tolerance exhibited pre- and post- switch and use of medications to manage feeding intolerance. Assessments by healthcare providers regarding responses to the switch were categorized as Improved, "No change," or "Worsened." **Results:** From 375 medical records of enterally-fed children with developmental delays screened, records for 13 children (6 males, 7 females) met criteria. Average age at time of formula change was 8.4 years (range 2.4-13.9). Ten subjects (77%) were fed via gastrostomy tube and 3 (23%), were fed by jejunostomy tube. Eleven (85%) had had a Nissen fundoplication. Of the 13 subjects assessed, 12 (92%) had improved feeding tolerance resulting from the switch to 100% whey peptide based formula, and 75% of these reported the time to improvement within 1 week after the switch. Feeding tolerance parameters improved were: vomiting: 6 of 7 cases (86%), gagging and retching: 3 of 4 cases (75%), high residual volumes: 5 of 8 cases (63%), constipation: 3 of 7 cases (43%), diarrhea: 3 of 3 cases (100%) and poor weight gain: 5 of 5 cases (100%). Of the 8 different reported feeding intolerance-related medications used prior to the formula switch, 6 (75%) were reported by a smaller proportion of patients after the switch and none were increased. After switching to 100% whey, peptide based formula 5 of 7 (71%) patients were able to tolerate increased feeding volumes and 4 of 4 (100%) that had concomitant poor weight gain on an intact protein formula achieved an increase in weight after switch. **Conclusion:** Switching to a 100% whey peptide based formula improved symptoms of feeding intolerance in the majority of developmentally delayed children suffering from these symptoms."

PANCREAS/CELIAC/MALABSORPTION

311 FAILURE TO THRIVE DUE TO NON-CELIAC GLUTEN SENSITIVITY IN A 12 MONTH OLD GIRL. M.Y. Phadke, A.F. Porto, Yale University, New Haven, Connecticut, UNITED STATES|.

Gluten related disorders include celiac disease, wheat allergy and non-celiac gluten sensitivity. While wheat allergy and celiac disease both have reliable screening and diagnostic tests, non celiac gluten sensitivity (NCGS) lacks both a clinical definition and screening tests. It can present with gastrointestinal as well as extraintestinal

manifestations and includes patients with negative screening tests for celiac disease or wheat allergy whose symptoms improve on a gluten free diet (GFD). NCGS is considered to be a rare entity in children with prevalence less than that of celiac disease. There have been no reported cases in infancy. We present a 12 month old with NCGS. A 12-month old girl presented with failure to thrive (FTT) and diarrhea. She was born full term and grew well until the age of 6 mo, when she began to gain weight at a slower pace. She dropped from the 37 to the 17 at 10 mo of age, and fell to the 5 at 12 mo. She was a picky eater and fussy. Initial consultation revealed a normal physical exam. We advised caloric supplementation and lab work including a complete blood count (CBC), thyroid function tests, comprehensive metabolic panel, iron studies, celiac panel, and stool tests. These labs were significant for mild eosinophilia on her CBC. She had a normal total IgA and TTG-IgA, with a mildly elevated anti-gliadin IgG and stool positive for 1+ reducing substances. At follow up visit 4 weeks later, weight gain was noted with weight at the 12 but her diarrhea persisted and in the interim, she developed vomiting. Mother started the child on a GFD in light of a positive anti-gliadin antibody and persistent symptoms. Follow up 8 weeks later revealed appropriate weight gain and complete resolution in symptoms. We asked the patient to resume gluten in the diet for 6-8 weeks and then performed an EGD and colonoscopy, which showed no evidence of enteropathy or eosinophilic disease. Celiac genetic markers were negative for DQ2 and DQ8. At this time, we advised a GFD with a likely diagnosis of NCGS. Several months later, a rash after accidental wheat exposure prompted allergy testing. She did have a positive patch test suggesting wheat intolerance, but introduction of other gluten containing grains was unsuccessful. Currently, the patient remains on a GFD and is doing well, with growth along the 34%ile. We present an infant with NCGS who presented with FTT, vomiting, and diarrhea after induction of gluten. Testing for celiac and wheat allergy were negative and her symptoms resolved on a GFD. This case demonstrates that a trial of GFD should be considered in patients with GI symptoms who have been evaluated for celiac disease and/or wheat allergy. Though rare, NCGS is a diagnosis that should be considered not only in children but also in infants who may develop symptoms after the introduction of gluten into their diet. The utility of patch testing to aid in the diagnosis of NCGS had not been fully studied and relevance in this patient is not certain. Further research is needed to understand the pathogenesis and epidemiology of this disease. For now, it is important to consider NCGS as a possible diagnosis in the pediatric population.

312 *CELIAC DISEASE WITH A DOUBLE BURDEN - COMPLICATED WITH INTESTINAL TUBERCULOSIS AND SEIZURES – A CASE REPORT.* A. Azmatullah, S. Ishaque, M. Tetlay, M. Baig, F. Ahmad, K. Sadiq, Pediatrics & Child Health, Aga Khan Univeristy, Karachi, Sindh, PAKISTAN | O. Parkash, Medicine, Aga Khan University, Karachi, Sindh, PAKISTAN |.

Introduction Celiac disease (CD) is combination of an autoimmune disorder and food intolerance that occurs in genetically susceptible individuals following ingestion of gluten. CD is also associated with a number of immune mediated associations. We present a case of a 12 year old child, known case of Celiac disease, presenting with complaints of leg swelling, decreased oral intake and intermittent abdominal pain. Clinical examination revealed bilateral pedal edema and ascites. She was managed as Protein Calorie Malnutrition. During her first week of stay she developed issues of fresh bleeding per rectum followed by melena and hematemesis. CBC showed anemia and thrombocytopenia. An upper GI and lower GI endoscopy was done which showed diffuse oozing from terminal ileum, ulceration and edema of illeocecal valve. Due to suspicion of Tuberculosis (TB), specimens were sent for Expert MTB/RIF assay (geneXpert), Acid Fast Bacilli (AFB) culture, and Histopathology. TB was confirmed based on a positive GeneXpert and granuloma formation on biopsy, and patient was started on Anti-Tubercular therapy (ATT). The child developed seizures during stay, which was diagnosed as partial cerebral venous thrombosis. She was discharged home seizure free on Levetiracetam and ATT. Conclusion Both intestinal tuberculosis and CD can cause malnutrition and failure to thrive, and a high index of suspicion is required to diagnose intestinal tuberculosis in CD. The effect of malnutrition as well as a common genetic association could play an important part in the increased risk of TB. There may be an autoimmune mechanism for celiac disease associated cerebral venous thrombosis as well.

LIVER

317 *ANTI-C MEDIATED HEMOLYTIC ANEMIA IN A CHILD AFTER ONE YEAR OF LIVING-RELATED DONOR LIVER TRANSPLANTATION. SUCCESSFUL TREATMENT WITH STEROIDS AND INTRAVENOUS IMMUNOGLOBULIN* .. M.A. Barr, L. Szonyi, D.C. Broering , M. SHAGRANI, Department of Liver & Small Bowel Transplantation & Hepatobiliary/Pancreatic Surgery, King Faisal Specialist Hospital & Research Center, Riyadh, Riyadh, SAUDI ARABIA | M.A. Barr, Pediatrics, Tanta University Hospital, Tanta, EGYPT |.

INTRODUCTION : Autoimmune hemolytic anemia (AIHA) is a relatively infrequent condition, in which autoantibodies target RBC antigens, resulting in premature destruction with inadequate compensation. The inciting autoantibodies may be warm agglutinins (mostly IgG direct hemolysis in the spleen), cold agglutinins (C3 mediated IgM intravascular hemolysis), or mixed agglutinins. Few case reports described AIHA after solid organ transplantation. Moreover, AIHA is rare after pediatric liver transplantation. Several factors participate in the pathogenesis of post-transplant AIHA, including sensitized donor lymphocytes and post-transplant immunosuppression. CASE STUDY : In this report, we are presenting an A+ boy who underwent liver transplantation at the age of 15 months from his A+ father. Post-transplant immunosuppression was adequately achieved by tacrolimus. Fourteen months later, he presented with acute hemolysis. The boy was A+ K- C+ c+ E+ e+. Direct anti-globulin test (Coomb's test) was positive, and later on, anti-C, anti-K, anti-M, and anti-Fya IgGs were retrieved from the serum. The eluate test showed anti-C as the principal inciting IgG. Treatment included transfusion of PRBCs (five times over 1 month), a short course high dose methyl prednisolone, oral prednisone for 1 month, and two doses of IVIG. Complete recovery was achieved with no recurrent hemolysis till the present time (13 months). CONCLUSION : AIHA may occur more than one year after liver transplantation in children. To our knowledge, this is the first post-liver transplant Anti-C AIHA in children, and it was amenable to steroids and intravenous immunoglobulin IVIG without using rituximab or modification of his immunosuppression therapy.

318 *KERNICTERUS SECONDARY TO A NOVEL MUTATION IN UGT1A1 GENE COMBINED WITH GILBERT TYPE PROMOTER MUTATION*. I. Batsis, A.S. Kota, K. Nelson, J. Gershel, D. Levanon, Pediatrics, Jacobi Medical Center, Albert Einstein College of Medicine, New York, New York, UNITED STATES | E. Pereira, Genetics, Jacobi Medical Center, Albert Einstein College of Medicine, New York, New York, UNITED STATES | E. Blumfield, Radiology, Jacobi Medical Center, Albert Einstein College of Medicine, New York, New York, UNITED STATES |.

Introduction: Gilbert's syndrome/Gilbert-type promoter abnormalities are the most common inherited disorders of bilirubin glucuronidation. Most instances result in a mild, transient unconjugated hyperbilirubinemia, secondary to a two-thirds reduction in hepatic uridinediphosphoglucuronate glucuronosyltransferase (UGT1A1). We recently cared for a two year-old girl with neurological sequelae of kernicterus, who was found to have a homozygous mutation in the promoter area of the UGT1A1 consistent with Gilbert's syndrome, as well as a novel homozygous variant that produced a picture similar to that of Crigler Najjar type II. Case report: A 12 month-old girl presented for a "second opinion" with failure to thrive, generalized hypotonia, global developmental delay, icteric sclera, and diarrhea. The infant was a recent immigrant from Mauritania and was the product of a consanguineous union (parents are first cousins). The infant became jaundiced shortly after birth (Hepatitis B negative) and this persisted at follow-up two months later. At that time the patient was treated with phototherapy thrice a week and placed on phenobarbital "for stiffening of arms and legs". Since then, the patient has had poor weight gain and delayed developmental milestones. The parents presented medical records with a total/direct bilirubin levels of 24.4/1.2 mg/dl and a brain MRI exhibiting hyperintensities in the globus pallidus. While the family believes the patient had normal genetic testing, they could not provide records. At our institution, patient was mildly icteric with marked hypotonia. Initial total/direct bilirubin was 4.2/0.3 mg/dl. Blood count, hemolytic markers, liver enzymes, and thyroid function tests were normal; serologies for infectious hepatitis were all negative. An MRI confirmed the findings on the previous study. At the time of presentation the patient was on a small dose of phenobarbital. The patient was then evaluated by a Pediatric Gastroenterologist, who adjusted the phenobarbital to maintain a stable/low bilirubin level; a Neurologist, who confirmed the diagnosis of kernicterus; and a Geneticist. Genetic

sequencing of the UGT1A1 gene in the child demonstrated 2 distinct abnormalities: homozygosity for a mutation *28 TA7 in the promoter region and a novel homozygous mutation for a variant of uncertain significance. The patient's bilirubin level remained stable on phenobarbital treatment and she is receiving supportive therapies. Conclusion: This case highlights the complexity of phenotypic expression of mutations in the UGT1A1 gene. Consanguinity and country of origin further complicate this case, as the prevalence of Gilbert's syndrome in western Africa is unknown and recessive mutations may become clinically evident due to intermarriages. Since the severity of the functional deficiency is dependent on the particular gene mutation, unknown variants may lead to detrimental effects when combined with a known pathogenic variant. Further studies on bilirubin-UGT enzyme activity would be beneficial to determine if our patient's unknown variant, in combination with the pathogenic variant, causes markedly decreased enzyme activity.

319 SUCCESSFUL ORTHOTOPIC LIVER TRANSPLANTATION AND MISMATCHED UNRELATED STEM CELL TRANSPLANTATION IN A 20 YEAR OLD FEMALE WITH SICKLE CELL DISEASE AND PRIMARY SCLEROSING CHOLANGITIS. C. Anderson, M. Andreansky, Pediatric Hematology-Oncology, University of Miami, Jackson Memorial Hospital, Miami, Florida, UNITED STATES|K. Bigaj, Pediatrics, University of Miami, Jackson Memorial Hospital, Miami, Florida, UNITED STATES|A. Hirzel, Pediatric Pathology, University of Miami, Miami, Florida, UNITED STATES|A. Tekin, Transplant Surgery, University of Miami, Jackson Memorial Hospital, Miami, Florida, UNITED STATES|.

Sickle cell disease (SCD) is a common hemoglobinopathy, the hallmark pathological process being veno-occlusive phenomena which culminates in multiple end organ complications. Hepatic dysfunction in SCD occurs either as sequelae of the hemoglobinopathy or a separate comorbidity, and accounts for 6.5% of overall mortality. There are a few reports in the literature that have described the feasibility of liver transplantation followed by hematopoietic stem cell transplantation (HSCT) as potential treatment modality. This is the first report, to our knowledge, of tandem orthotopic liver transplantation (OLT) followed by a mismatched unrelated HSCT in a 20 year-old female with sickle cell disease hemoglobin SS, with a concurrent primary sclerosing cholangitis (PSC), ulcerative colitis (UC) and liver cirrhosis. Our patient was diagnosed with PSC at 12 years of age, and experienced a rapidly progressive course and resultant stage 4 fibrosis by 16 years of age. One year prior to transplantation, she did not yet exhibit overt signs of end-stage liver disease (ESLD), however she had been sick with cholangitis and liver biopsies were concerning for worsening cirrhosis, PSC and iron overload (figure 1). At the age of 20 years, our patient was admitted for liver transplant followed by HSCT. She had received exchange transfusion to decrease HbSS to <30%. Laboratory work-up revealed mild total and direct hyperbilirubinemia with elevated GGT and ALP, normal coagulation profile and mild hypoalbuminemia. Orthotopic liver transplantation with duct-to-duct biliary reconstruction was performed. Immunosuppression included Methylprednisolone taper and Tacrolimus. Post-operative recovery was uneventful. HSCT was performed one month later with a 9/10 HLA mismatched unrelated donor and reduced-intensity conditioning including Alemtuzumab, Fludarabine, Melphalan and Thiotepa (figure 2). There were no major complications, and she was discharged home on Day +29 post BMT with 100% engraftment. The post-transplant period was complicated with HUS like symptoms contributed to tacrolimus and GVHD of the skin, joints and lung which was managed with immunosuppression. Two years after liver and bone marrow transplantation, our patient is doing well and has not been admitted to the hospital in over one year. She is on Cyclosporine for liver rejection prophylaxis with perfect liver functions, and on Cellcept for chronic lung and joint GVHD which remain quiescent. Her UC has not been active since post-transplant. This case demonstrates that orthotopic liver transplantation and unrelated HSCT with low intensity regimens may offer a successful treatment modality for liver disease with concurrent hemoglobinopathies. Early intervention before development of significant ESLD may result in better prognosis.

320 SUCCESSFUL LIVER TRANSPLANTATION FOR FULMINANT HEPATIC FAILURE WITH DISSEMINATED HSV-2: PRESENTATION OF TWO CASES. M.W. Butler, W.J. Steinbach, Pediatrics, Duke University, Durham, North Carolina, UNITED STATES|D. Sudan, Surgery, Duke University, Durham, North Carolina, UNITED STATES|.

Herpes simplex virus 2 (HSV-2) is an uncommon cause of hepatitis in neonates and may present with fulminant hepatic failure. The standard of care is antiviral treatment; however, there are rare controversial occasions when liver transplantation must be considered. It is unclear whether disseminated HSV-2 should be a contraindication for liver transplantation, as the transplanted graft will be exposed to the virus coupled with the added burden of immunomodulation. Furthermore, the majority of patients may respond to antiviral therapy, and therefore transplantation prior to antiviral optimization may be unnecessary. There are also other concerns regarding persistent injury due to multisystem organ failure. There is a paucity of data regarding the optimal treatment algorithms for transplantation for hepatic failure secondary to neonatal HSV-2 infection. We present two cases of neonatal fulminant hepatic failure caused by disseminated HSV-2 that successfully underwent orthotopic liver transplantation (OLT). Both infants were discharged from the newborn nursery after uneventful perinatal courses and subsequently developed vomiting and feeding intolerance within the first two weeks of life. Neither had a known history of potential HSV-2 exposure or vesicular skin lesions. At initial presentation, both were critically ill with profound coagulopathy, encephalopathy, and associated multisystem organ failure. Empiric antiviral treatment with acyclovir was immediately initiated. However, both infants remained gravely ill despite aggressive medical management. Case 1 underwent living related liver transplant at day of life 14. A diagnosis of HSV-associated liver disease was provided after transplantation by the explanted native liver and serologic studies that were obtained on admission. This child is currently 4 years of age and doing well with no clinical sequela and is maintained on tacrolimus and PRN acyclovir. Case 2 was known to have disseminated HSV-2. Due to progressive liver failure despite aggressive antiviral therapy, orthotopic liver transplantation was performed with a cadaveric segmental graft on day of life 32. Both explanted native livers were remarkable for massive necrosis and immunohistochemistry that was diagnostic of HSV. After transplant, both infants had normalization of liver function. To date, both infants have maintained graft function, and require chronic antiviral therapy with acyclovir based on limited adult literature. While liver failure in the setting of disseminated HSV-2 infection should remain an infrequent indication for transplantation, these two cases demonstrate the potential for successful liver transplantation and subsequent graft survival, as well as present a need for the development of standards of care for this complex neonatal hepatic condition.

321 *HEMORRHAGE AFTER CIRCUMCISION: AN UNUSUAL PRESENTATION OF TYROSINEMIA (TYPE I)*. K.E. Diaz Ayllon, R. Gonzalez-Peralta, R. Zori, Department of Pediatrics, University of Florida and UF Health Shands Children's Hospital, Gainesville, Florida, UNITED STATES|K.E. Diaz Ayllon, Division of Medical Education, Gainesville, Florida, UNITED STATES|R. Gonzalez-Peralta, Division of Gastroenterology, Gainesville, Florida, UNITED STATES|R. Zori, Division of Genetics and Metabolism, Gainesville, Florida, UNITED STATES|.

Introduction: Tyrosinemia (type 1) is a genetic disorder caused by deficiency of fumarylacetoacetate hydrolase. This enzymatic defect leads to elevated serum levels of tyrosine and urine concentrations of succinylacetone. Symptoms usually appear in infancy and include failure to thrive, vomiting, diarrhea, liver dysfunction, cabbage-like odor, and bleeding tendency. Progression of untreated disease leads to liver, kidney and neurological disease and a high propensity for liver cancer in early childhood. Case: AB is a 6-week-old Caucasian-Hispanic male who developed persistent penile bleeding after elective circumcision. The pregnancy and delivery were uneventful and family history was negative for bleeding, liver or metabolic disease. Findings on initial exam were normal, except for tachycardia, the presence of a systolic murmur, and mild penile oozing from the circumcision site. On admission, the hemoglobin was 8.1 g/dL, platelet count was 151,000U/L and INR was 6.5. He continued to bleed from the circumcision site with worsening anemia and persistent coagulopathy requiring multiple transfusions of red blood cells and fresh frozen plasma. Results of initial liver tests were normal (except for alkaline phosphatase of 2,177U/L). On repeat analysis within a few days of admission, the AST and ALT were mildly elevated at 75 U/L and 70 U/L, respectively; the total and direct bilirubin concentrations were normal (1.3 mg/dL and 0.2 mg/dL, respectively). The alfa-fetorprotein (AFP) was markedly elevated (202,852 ng/mL). A bleeding diathesis and other causes of liver dysfunction were excluded. Persistent coagulopathy in the context of mild liver dysfunction raised suspicion for tyrosinemia; results of urine succinylacetone levels were markedly elevated confirming this diagnosis. He was started on Nitisinone (1 mg/kg/day) and low-tyrosine diet. The coagulopathy, liver tests and AFP gradually

normalized and he was discharged home. He later developed attention deficit disorder and oppositional disorder but otherwise has remained clinically stable for the past 14 years with careful clinical monitoring, including periodic ophthalmological, biochemical evaluations, and abdominal sonograms. Conclusion: Persistent bleeding-coagulopathy in the setting of mild liver disease should quickly raise suspicion for tyrosinemia (type 1). Expedient diagnosis is critical to implement appropriate therapy and avert tyrosinemia-related complications including cataracts, kidney and liver disease and hepatocellular carcinoma. Since the incorporation of tyrosinemia testing in the newborn screening program in 2006, affected infants are detected early in the State of Florida.

328 *PROLONGED COAGULOPATHY FOLLOWING STREPTOCOCCUS INTERMEDIUS PYOGENIC LIVER ABSCESS*. L.J. Klein, J.P. Middleton, T. Altes, University of Virginia, Charlottesville, Virginia, UNITED STATES |.

Introduction: Pyogenic liver abscess is an uncommon cause of abdominal pain and fever in children. *Streptococcus intermedius*, usually found in normal oral and gastrointestinal flora, is a rare cause of liver abscess. Although a majority of patients with pyogenic liver abscess have coagulopathy, the longevity of this coagulopathy has not been reported. We describe a patient with coagulopathy lasting three weeks after a *S. intermedius* liver abscess. Case presentation: A 16-year-old previously healthy female presented with two weeks of right-sided abdominal pain and intermittent fever following a dental cleaning. She was evaluated on three separate occasions before imaging with ultrasound and MRI demonstrated a 4.6 x 5.1 cm right lobe liver abscess. She was started on broad spectrum antibiotics. A CT guided percutaneous drain produced 60 mL of purulent fluid but was complicated by 200 mL of bleeding. Aerobic cultures of the abscess aspirate grew *S. intermedius*, while blood cultures remained negative. During admission, despite intravenous vitamin K therapy, her prothrombin time (PT)/international normalized ratio (INR) remained elevated at 16 sec/1.5 with low Factor V and VII activity. After 3 weeks of intravenous antibiotics, a follow-up ultrasound demonstrated improvement of her abscess, but her coagulopathy persisted with prolonged INR and decreased Factor activity. Eight weeks after presentation, her coagulopathy completely resolved.

Discussion: We describe a case of prolonged coagulopathy in a child with a *S. intermedius* liver abscess. Because it is an uncommon cause of prolonged fever and abdominal pain in immunocompetent children, providers often fail to consider pyogenic liver abscess during evaluation. Although gram-negative bacteria, *Staphylococcus aureus*, and anaerobes are the most common etiologic agents, *S. intermedius* should be considered, especially after dental or gastrointestinal manipulation. Coagulopathy in patients with pyogenic liver abscess is not uncommon and is clinically important since treatment can be invasive with percutaneous drainage. Belko, J., Goldmann, D. a, Macone, A., & Zaidi, A. K. (2002). Clinically significant infections with organisms of the *Streptococcus milleri* group. *The Pediatric Infectious Disease Journal*, 21(8), 715–23. 2. Chu, K. M., Fan, S. T., Lai, E. C., Lo, C. M., & Wong, J. (1996). Pyogenic liver abscess. An audit of experience over the past decade. *Archives of Surgery*, 131, 148–152.

329 *EARLY HEPATOCELLULAR CARCINOMA ASSOCIATED WITH FIBROCYSTIC LIVER DISEASE IN A 10 YEARS OLD CHILD --FIRST CASE REPORT.* K. Kumar, M. SHAGRANI, D.C. Broering, Pediatric Transplant Hepatology, Organ Transplant Centre, King Faisal Specialist Hospital and research Centre, Riyadh, SAUDI ARABIA | H. Alhussaini, Pathology And Laboratory Medicine, King Faisal Specialist Hospital and Research Centre, Riyadh, SAUDI ARABIA |.

Background: Fibrocystic liver–kidney disease is caused by a group of rare and genetically diverse disorders that are associated with kidney cysts or dysplasia and ductal plate malformation in the liver. There have been several reports of liver neoplasias arising in hepatobiliary fibrocystic diseases. However, most of them were cholangiocarcinomas and cases involving hepatocellular carcinoma (HCC) are rare and all reported cases are related with adults. Case Report: A 10 years old girl with history of repeated gastrointestinal bleeding underwent multiple times for banding and sclerotherapy and also had history of Porto systemic shunt without any significant benefit referred to us as a case of Fibrocystic liver disease with decompensated liver function for Liver Transplantation. Child underwent Living donor liver transplantation, the explanted liver showed. Gross pathology Explant liver weighed 838 g and measuring 21 x 13 x 8.5 cm with attached gallbladder measuring 7 x 3 x 0.2 cm (in wall thickness). The external surface is covered by multiple white nodules ranging in size from 0.4 to 1 cm. Serial slicing reveals an ill-defined yellow soft lesion (4 x 2.5 x 2.5 cm) localized in the sub capsular area of the left lobe (segment

4). The rest of the cut surface is green and nodular (cirrhotic). Histopathology Microscopy from largest nodule consistent with early hepatocellular carcinoma. The rest of liver is cirrhotic and the morphology consistent with fibrocystic disease of liver. Conclusion: We reported a rare case of HCC associated with fibrocystic liver disease. When diagnosing Fibrocystic liver disease without known risk factors, the presence of HCC must be considered and vice versa. To our knowledge, this is the first reported case of Hepatocellular carcinoma associated with fibrocystic liver disease in a 10 year old child.

330 *SPONTANEOUS BILE DUCT PERFORATION CASE REPORT*. R. Nagpal, C. Smith, Pediatric Gastroenterology, Advocate Children's Hospital, Oak Lawn, Illinois, UNITED STATES|A. Lo, Pediatric Surgery, Advocate Children's Hospital, Oak Lawn, Illinois, UNITED STATES|L.S. Lawrence, Pediatric Residency, Advocate Children's Hospital, Oak Lawn, Illinois, UNITED STATES|.

Spontaneous bile duct perforation is a rare cause of direct hyperbilirubinemia and peritonitis in the pediatric population; to date there are approximately 150 reported cases. Its rarity can lead to morbidity, delay in diagnosis and prolonged hospital course. The objective of this review is to increase awareness among pediatric gastroenterologists of the manifestations of spontaneous bile duct perforation. We present the case of a 4 week-old male with an unremarkable neonatal history presenting with 10 days of fevers to 100-101°F, acholic stools, scleral icterus and progressive abdominal distension. On exam, the patient was irritable, exacerbated by abdominal palpation. Abdomen was firmly distended. Bilateral scrotal edema was noted, as well as scleral icterus and jaundice to the chest. Lab testing was notable for T and D bilirubin of 4.7 and 3.6, respectively; alkaline phosphatase 297; GGTP 1642; albumin 2.1; hepatic transaminases and prothrombin time were normal. Abdominal ultrasound showed mild central biliary ductal dilation, normal size gallbladder, no triangular echogenic cord, and loculated ascites. Paracentesis demonstrated grossly bilious fluid and hepatobiliary iminodiacetic acid scan demonstrated early filling of the liver and gallbladder with free peritoneal spill, and no filling of the duodenum. Spontaneous perforation of the bile duct was confirmed after the patient underwent exploratory laparotomy and intraoperative cholangiogram. Initial management consisted of intraperitoneal drain placement, temporary cholecystostomy and bowel rest. Spontaneous bile duct perforation should be considered in infants presenting with jaundice, acholic stools and ascites. Early surgical intervention is essential to rule out distal obstruction, rule out pathology such as ruptured choledochal cyst, prevent complications, and facilitate spontaneous healing of the perforation.

331 *NEONATAL CHOLESTASIS: PROGRESSIVE FAMILIAL INTRAHEPATIC CHOLESTASIS I AND HYPOTHYROIDISM*. L.K. Lazar, L. Chan, University of Texas at Southwestern, Dallas, Texas, UNITED STATES|.

Case: 7 week old ex-term female presented from PCP for jaundice and direct hyperbilirubinemia. Her jaundice started two weeks prior to admission and had worsened over the previous three days. Further history was negative for poor feeding, dark urine, acholic stools, and family history of neonatal jaundice or liver disease. However, there was parental consanguinity. At presentation, patient had direct hyperbilirubinemia (total bilirubin 7.78 mg/dl, direct 6.42 mg/dl), no cholestasis (alkaline phosphatase was elevated to 426 units/L, GGT 11 units/L), and no hepatitis (ALT 30 units/L, and AST 44 units/L). Urinalysis was unremarkable. Hepatic ultrasound did show a dilated common bile duct but no intrahepatic biliary dilatation. NM hepatobiliary scan was not concerning for biliary atresia. Newborn screen test was normal. Urine organic acids and succinylacetone were negative for inborn errors of metabolism and tyrosinemia respectively. Thyroid studies were notable for TSH 418 microIU/L and T4 of 0.56 ng/dL. Serum alpha-1-antitrypsin level was low, and phenotyping revealed M; however, this did not exclude rare deficiency allele. Due to congenital hypothyroidism, patient began treatment, and his direct hyperbilirubinemia improved. As an outpatient, however, her direct hyperbilirubinemia worsened. The jaundice chip was sent and positive for progressive familial intrahepatic cholestasis I. Discussion: Progressive familial intrahepatic cholestasis (PFIC) describes a class of autosomal recessive genetic mutations that cause neonatal cholestasis. Cholestasis typically presents by 3 months of age and often progresses to cirrhosis within the first decade of life. This patient has PFIC type I, a mutation in the ATP8B1 gene on 18q21-22. The mutation itself causes

dysfunction of the FIC1 protein, a P-type ATPase. The defective protein results in ineffective bile acid secretion in the hepatocytes. Type I is notable for direct hyperbilirubinemia, normal/low gamma-glutamyl peptidase, and elevated bile acids. The interesting thing about this case is that the patient had congenital hypothyroidism, another potential cause of hyperbilirubinemia. Without close follow-up, further work-up may not have been completed.

333 *SUCCESSFUL TREATMENT OF ACUTE FULMINANT HEPATITIS C IN A TEENAGE BOY.* M. Maksimak, Pediatric Gastroenterology, Geisinger Clinic - Janet Weis Children's Hospital, Danville, Pennsylvania, UNITED STATES| R.E. Smith, Gastroenterology and Hepatology, Geisinger Clinic, Danville, Pennsylvania, UNITED STATES|.

Introduction: Hepatitis C is an infection seen occasionally in our adolescent patients. It is most commonly acquired via vertical transmission or from intravenous drug use. Infection usually results in chronic disease with liver manifestations occurring over several decades. We report a 17 year old boy who presented with fulminant acute hepatitis C. Case Report: SS presented as a 17-year-old boy with a chronic history of intravenous heroin abuse. He was interned in a drug treatment program in October 2013. At that time HCV, HIV and HBV testing were normal. However the patient had a positive PPD and was treated with Isoniazid 300 mg daily. In December he developed jaundice and tiredness. Testing for HCV antibody was now positive and he was referred for pediatric gastroenterology consultation in January 2014. Initial testing revealed: ALT 729 AST 646 GGTP 247 Alkaline phosphatase 144 T/D bilirubin 2.3/1.4 Initial HCV RNA 9040084 IU/ml HCV genotype 1a Isoniazid was discontinued, but the ALT continued to rise to a high of ALT 1248 two weeks later. Work-up for autoimmune and infectious causes and for Wilson's disease were negative. Liver biopsy revealed moderate /marked portal inflammation in all portal areas seen. Sofosbuvir 400mg and Simeprevir 150 mg were started in early February 2014. No side effects were noted during the 12 weeks of therapy. Liver enzymes levels started to fall immediately and by week six, liver enzymes were normal and HCV RNA was undetectable. Long-term follow-up has shown a sustained viral response, but the patient was found to have a mild persistent unconjugated hyperbilirubinemia. Gene testing showed the patient to have the UGT1A1*28 promotor variant for Gilbert's syndrome. For the poster presentation, liver histology and a detailed timeline including liver tests and viral titers will be presented. Conclusion: We present a 17-year-old boy with fulminant acute hepatitis C who was treated successfully with Sofosbuvir and Simeprevir within weeks after their approval for chronic hepatitis C.

334 *ENDOCRINOPATHIES ASSOCIATED WITH NEONATAL CHOLESTASIS.* M. middelhof, P. Mohanty, Pediatric Gastroenterology and Nutrition , University of Rochester, Rochester, New York, UNITED STATES|.

Introduction Neonatal cholestasis secondary to these hormone deficiencies is uncommon and generally not well recognized. A high index of suspicion can lead to prompt referral to the pediatric endocrinologist, facilitating timely diagnosis and treatment with hormone replacement therapy (HRT). We present a rare case of neonatal cholestasis secondary to congenital adrenal hyperplasia, and resolution of cholestasis with HRT. Case report A 2-week-old full term boy was evaluated for direct hyperbilirubinemia that started on sixth day of life. The neonatal course was complicated by pulmonary hypertension with mild mitral valve insufficiency, moderate pulmonary valve regurgitation with hypoxic respiratory failure requiring mechanical ventilation. His newborn metabolic screen was positive for mildly elevated 17-hydroxyprogesterone (158.3; normal 10-800 ng/ml). On thirteen day of life, progressive direct hyperbilirubinemia (5.3 mg/dl) with transaminitis (aspartate aminotransferase-166 U/L, alanine aminotransferase- 151 U/L) was noted. Further work-up for direct hyperbilirubinemia was negative for infectious (negative TORCH), structural (negative abdominal ultrasound, HIDA scan), normal alpha one antitrypsin level and phenotype. Repeated 17-hydroxyprogesterone levels (DOL 14) were markedly elevated (6920 ng/ml, normal 10-800 ng/ml) with hyponatremia and hyperkalemia. Further work-up was positive for elevated aldosterone (60-400 ng/dL, normal 20-50 ng/dL) and elevated renin level (164.5 ng/ml/hr), which were consistent with Non-virilizing 21-hydroxylase deficiency. He was then started on hydrocortisone and fludrocortisone supplementation with resolution of jaundice within 3 weeks of starting treatment with HRT. Discussion CAH is characterized by deficiency of all adrenal and gonadal steroid hormones, increased adrenocorticotropin hormone (ACTH) secretion, and

marked adrenal hyperplasia with progressive accumulation of cholesterol esters. Hypoglycemia and electrolyte disturbances in the presence of neonatal cholestasis should be considered for a possible congenital adrenal hyperplasia as a main pathology. Conclusion Congenital endocrinopathies must be considered in the differential diagnosis of neonatal conjugated hyperbilirubinemia. Early detection of cholestatic jaundice and accurate diagnosis are important for successful treatment and a favorable prognosis. Earlier the introduction of HRT, the faster is the remission.

339 *INFANT WITH BILIARY ATRESIA AND VACTERL*. N. Nguyen, S. Sundaram, Gastroenterology & Hepatology, Children's Hospital of Colorado, Aurora, Colorado, UNITED STATES|J. Bruny, Pediatric Surgery, Children's Hospital of Colorado, Aurora, Colorado, UNITED STATES|.

Introduction VACTERL association is defined as a non-random co-occurrence of congenital malformations including vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula (TEF), renal anomalies, and limb abnormalities. Biliary atresia, a progressive fibro-inflammatory process that results obstruction of the biliary tract, is the most common indication for liver transplant in pediatrics. We present a unique case of an infant affected by both VACTERL and biliary atresia. Clinical case A 9 day old female was transferred to our NICU from a community hospital after being diagnosed with multiple congenital anomalies, including an H-type TEF, tethered cord, anorectal malformation with vaginal fistula, left superior vena cava with interrupted inferior vena cava, and horseshoe kidney. Her labs were notable for cholestasis (total bilirubin 14.6 mg/dl, direct bilirubin of 4.9 mg/dl, GGT 658 U/L), with normal aminotransferases and normal synthetic liver function. She had an abdominal ultrasound with a non-visualized gallbladder and an anechoic tubular structure located in the expected region of the common bile duct, concerning for an atretic common bile duct. A HIDA scan revealed normal hepatic activity with no excretion of tracer into the biliary tree or bowel. Evaluation of neonatal cholestasis included a negative urine culture and urine CMV, normal alpha1 antitrypsin phenotype and level, normal thyroid studies, negative newborn screen for galactosemia and cystic fibrosis, and no choledochal cyst or gallstones on abdominal ultrasound. Alagille syndrome remained a possibility, given her horseshoe kidney and cardiac anomalies, although TEF, tethered cord and anorectal malformations have not been reported in Alagille syndrome. The compilation of an absent gallbladder, concern for atretic common bile duct on ultrasound, interrupted IVC, high GGT, and abnormal HIDA scan result raised clinical suspicion for biliary atresia, therefore she underwent cholangiogram and liver biopsy. An intraoperative cholangiogram only filled the gallbladder, with no filling of bile ducts. Based on these findings, she was diagnosed with biliary atresia and underwent Kasai portoenterostomy. She had normalization of total and direct bilirubin by post-operative day 7. Liver biopsy revealed extensive cholestasis with bile duct proliferation and plugs and expansion of portal tracts with inflammatory infiltrate. Discussion Biliary atresia is an isolated abnormality in most infants, however, there are a subgroup of patients who have other vascular and visceral abnormalities. Biliary atresia with splenic malformation is a well-recognized association. These patients have splenic malformations including polysplenia, double spleen, or asplenia. In addition, they may have other congenital malformations including situs inversus, intestinal malrotation, annular pancreas, and cardiac anomalies. They may have vascular anomalies including IVC abnormalities, as in our patient, however, this is usually accompanied by splenic malformations. To our knowledge, there is no known association between VACTERL and biliary atresia. However, the association of these disease states in this patient may suggest commonalities in developmental pathogenesis.

340 *"PRIMARY HEPATOCELLULAR CARCINOMA IN A PREVIOUSLY HEALTHY CHILD"*. D. Laman, Pediatric Gastroenterologist, Children's at Erlanger, Chattanooga, Tennessee, UNITED STATES|J. Niklinska, Pediatrics Resident, Children's at Erlanger, Chattanooga, Tennessee, UNITED STATES|C. Koontz, Pediatric Surgery, Children's at Erlanger, Chattanooga, Tennessee, UNITED STATES|.

Introduction: Hepatocellular carcinoma (HCC) is one of the ten most common adult cancers encountered worldwide. However, malignant tumors of the liver are very uncommon in childhood, accounting for approximately 0.5-2% of childhood neoplasms. The annual incidence of HCC in children is approximately 0.5 cases

per million. Children with hepatitis B virus infection and underlying metabolic diseases are the two main high risk groups of those with HCC, mainly occurring in the Western countries of Africa and the Orient. Case presentation: A 10 year old Caucasian male, previously healthy, presented to the gastroenterology (GI) clinic with nausea, vomiting, diarrhea, epigastric pain and a 10 lbs weight loss. An upper endoscopy revealed mild gastritis. Symptoms did not improve, thus a Hepatobiliary Iminodiacetic Acid Scan (HIDA) scan and an abdominal ultrasound were ordered to look for gallbladder issues, partially due to a familial history of gallbladder disease. The HIDA scan revealed a relatively photopenic area in the left upper quadrant, where a mass within the left lobe of the liver could not be excluded. The ultrasound follow up showed a 5.4 x 6.0 x 6.2 heterogeneous echogenic solid lesion within the left hepatic lobe medially, prompting an MRI that confirmed a 6.3 x 7.2 x 5.7 cm hepatic lobe mass on segments 2 and 3. This readied urgent consultation with Pediatric Surgery and the mass was resected with optimal post-op recovery. Prior to surgery the patient had a normal carcinoembryonic antigen, normal alpha-fetoprotein level, and a normal beta-HCG. PT and PTT were normal at 12.2 and 31 seconds respectively. Mild elevation of AST and ALT occurred only post surgery. Liver function tests were essentially all normal prior to surgery. Pathology report revealed primary hepatocellular carcinoma of the fibrolamellar type. The left lobectomy showed complete resection with the tumor approaching to within 0.3cm from the nearest parenchymal margin. Postoperative CT of the chest, abdomen, and pelvis to evaluate for lymphadenopathy and metastatic disease were negative. The patients bone scan was negative. An extensive list of labs showed all normal levels of alpha-1-antitrypsin, serum copper, anti-smooth muscle antibody, anti-liver kidney muscle antibody, hepatitis B panel, hepatitis C panel, CMV PCR, CMV antinuclear antibody, antimitochondrial antibody, base active peptide, and angiotensin. On further follow ups with the GI clinic, it was noticed that his appetite improved, the vomiting settled, and mild diarrhea occurred on average once a week. Off and on abdominal pain was treated with nexium and symptoms resolved. Liver function tests and alpha-fetoprotein are being followed every 6 months by the GI team, as well as scheduled appointments with the hematology/oncology team. Conclusion: Primary hepatocellular carcinoma is extremely rare and frequently lethal in pediatrics if not diagnosed and treated early. It is important to provide aggressive treatment for a successful recovery.

341 UNPREDICTABLE DISEASE PROGRESSION IN CHILDREN WITH NEONATAL SCLEROSING CHOLANGITIS. A.

Bobarnac, F. Alvarez, M. Paganelli, Pediatric Gastroenterology, Hepatology and Nutrition, CHU Sainte-Justine, Université de Montréal, Montreal, Quebec, CANADA | E. Bequet, F. Gottrand, Pediatric gastroenterology, CHRU de Lille, Lille, FRANCE | E. Sokal, Pediatric gastroenterology and hepatology, Cliniques Saint-Luc, Université catholique de Louvain, Brussels, BELGIUM |.

Background: Only 27 patients with neonatal sclerosing cholangitis (NSC) have been described so far in 5 reports. Very little is known on the disease course and long-term outcome. Aim: To describe the natural history of NSC in a new multicentric cohort. Methods: Medical records of all children with diagnosis of NSC seen at 3 referral centers for pediatric hepatology were reviewed. NSC was defined as presentation with neonatal cholestasis and cholangiographic evidence of patent extrahepatic biliary tree, irregular intrahepatic bile ducts with strictures and segmental dilations, and rarefaction of secondary branches. Results: Ten infants (6 girls) were identified having NSC between 1992 and 2014. Median age at diagnosis was 2 months (range 1-20). All patients presented neonatal jaundice, 5 had acholic stools, 1 presented with ascites, and 3 had failure to thrive. Hepatomegaly and splenomegaly at ultrasound were noted in 8 and 6 children, respectively. None had asplenia/polysplenia and all had normal gallbladder. All children showed elevated total ($119 \pm 77 \mu\text{mol/L}$) and conjugated ($68 \pm 35 \mu\text{mol/L}$) bilirubin levels, with normal liver function tests. ALT and GGT levels were elevated in 9 patients (83 ± 69 and $308 \pm 205 \text{ UI/L}$, respectively). Cholangiography (6 percutaneous, 4 laparoscopic) showed irregularities of ducts walls in 7 patients, dilatations in 6 and rarefaction of secondary branches in 3. Liver biopsy, performed in all children, showed portal fibrosis in 5, inflammation in 3, ductular proliferation in 7 and intracellular cholestasis in 3. Ursodeoxycholic acid was empirically administered to all at standard dose. Jaundice resolution was noted in 6 patients at a median age of 6 months (1-30). All subjects are alive after a median follow-up of 73 months (12-240). Nine patients developed secondary biliary cirrhosis. Five of them developed esophageal varices (4 had variceal

bleeding). Five children progressed to chronic liver failure. Three underwent liver transplantation (LT) at 1, 2 and 4.5 years, respectively, with no post-LT disease recurrence. One patient developed PTLD and tacrolimus-induced kidney failure, and underwent kidney transplantation 12 years post-LT. The other 2 patients are well with normal liver biochemistry 1.5 and 12 years after LT. Two patients (15 and 23 years old) are still waiting for LT. One girl, diagnosed when 2 months old, is asymptomatic at 6 years of age, with normal liver biochemistry, absence of fibrosis or inflammation at liver biopsy, but persisting signs of sclerosing cholangitis at cholangiography. No factors explaining the variability in disease progression were identified. Conclusions: We confirm in the second largest series of children with NSC ever described that differential diagnosis with biliary atresia is cumbersome, half of the patients presenting neonatal cholestasis with acholic stools and liver histopathology being aspecific in all cases. Although half of the children progressed to chronic liver failure before the 18th year of age, the clinical course was extremely variable. No predisposing factors for rapid progression and unfavorable outcome were found. A bigger multicentric study is needed to identify such factors.

344 PLASMAPHARESIS, INTRAVENOUS IMMUNOGLOBULIN AND RITUXIMAB SUCCESSFULLY TREAT RECURRENT PROGRESSIVE FAMILIAL INTRAHEPATIC CHOLESTASIS TYPE 2 (PFIC2) AFTER LIVER TRANSPLANTATION . . M.

SHAGRANI, M. Burdelski , T. ALGOUFI, F. Abaalkhail, D.C. Broering , H. Elsiesy, Department of Liver & Small Bowel Transplantation & Hepatobiliary/Pancreatic Surgery, King Faisal Specialist Hospital & Research Center, Riyadh , Riyadh, SAUDI ARABIA|.

Background: Allo-immune mediated Bile Salt Export Pump(BSEP) dysfunction may occur after liver transplantation in PFIC2 patients leading to a PFIC2 like phenotype. The IgG antibodies are reactive toward a canalicular epitope of BSEP, are of high affinity, and inhibited transport activity of BSEP, thus causing severe cholestasis. This phenomenon was first described in 2009, since then, few cases of PFIC-2 recurrence were reported with mixed results Methods: We report on two patients who developed recurrent normal GGT cholestasis mimicking primary (BSEP)disease, after liver transplantation. A 14 years old boy and his 19 years old sister who had received cadaveric liver transplantation at the United States in 2011. In January 2014 they presented with severe itching, high bilirubin, high AST/ALT, high serum bile acid with persistently low GGT. Virology, Autoimmune screen, Abdominal CT Scan, ERCP and liver biopsy were negative. Immunosuppressions were maximized with no improvement. A repeat biopsy of the 14 year-old boy on May 2014 showed recurrence of PFIC2, His Anti-BSEP came positive with a very high serum titer 1:1200, Treatment regimen for him started on June 2014, he received a course of 5 sessions of plasmapheresis each session followed by IV immunoglobulin (IVIG) , then received first dose of I.V. Rituximab 375/m2. The second course of Plasmapheresis where modified by doing 5 sessions of plasmapheresis every other day with an exchange volume of 1.5, followed by 3 days of IVIG to avoid washing out the IVIG by plasmapheresis, followed by the second dose of IV Rituximab 375/m2. His sister's liver biopsy on July 2014 showed PFIC2 recurrence. She started treatment for recurrence started on September 2014, using the modified protocol. Finished the third course of Plasmapheresis, IVIG and IV Rituximab . Results: Currently, both patients improved clinically and biochemically with a mean follow up period of 6 months .Conclusions: PFIC-2 recurrence after liver transplantation occur through an antibody mediated reaction against BSEP receptors on canalicular membrane mostly in stop codon type of mutation of (PFIC2) and in our two cases can successfully be treated with plasmapheresis, IVIG and rituximab obviating the need for re-transplantation

345 NEONATAL HEMOCHROMATOSIS DETECTED BY GALACTOSEMIA AT NEWBORN SCREENING. K. takashima, Y. maruo, T. yanagi, Y. takeuchi, Department of Pediatrics, Shiga University of Medical Science, Otsu, Shiga, JAPAN|M. kasahara, Transplantation Center, National Center for Child Health and Development, Setagaya-ku, Tokyo, JAPAN|.

Introduction Neonatal hemochromatosis (NH) is a congenital alloimmune disease that is clinically defined as severe liver failure associated with extrahepatic iron deposition. NH is usually diagnosed during management for premature birth, small for gestational age, perinatal hypoglycemia, coagulopathy, edema, and jaundice owing to liver and multi-organ failure. Many opportunities are currently present for the detection of NH owing to the

development of inspection techniques, such as newborn screening. Here we report the case of an infant who was detected with NH after the diagnosis of galactosemia at newborn screening. Although no report has been presented on the detection of NH after the diagnosis of a disorder of carbohydrate metabolism, in the present case, NH was suspected, the infant received early intervention, and good outcomes were obtained. Case report The patient was an 18-day-old Japanese boy who was admitted to our hospital because of galactosemia at newborn screening. His parents were not consanguineous, and his mother was gravida 1 para 1. He was born at term (weight, 2,970 g) by normal vaginal delivery after an uncomplicated pregnancy. A newborn screening program at 4 days of life showed that his serum galactose level was above 10.1 mg/dL. At admission, a physical examination revealed jaundice and hepatomegaly. His laboratory examination results revealed liver failure (albumin, 2.2 g/dL; aspartate aminotransferase, 290 U/L; alanine aminotransferase, 87 U/L; total/direct bilirubin, 15.0/8.0 mg/dL), coagulopathy (prothrombin time-international normalized ratio, 1.71; activated partial thromboplastin time, 61.7 s), and a serum ferritin level of 1082.1 ng/mL. He was treated with lactose-free formula for galactosemia. However, his liver failure worsened. Abdominal magnetic resonance imaging at 23 days of life showed low intensity on T2-weighted imaging of the hepatic parenchyma. Oral mucosal biopsy could not be performed because of marked coagulopathy. At 24 days of life, he was transferred to the National Center for Children Health and Development (Tokyo, Japan) for liver transplantation. At 34 days of life, living donor liver transplantation was performed with his father as the donor. His liver function returned to normal after the transplantation. Macroscopically, the explanted liver was atrophic and irregularly nodular, with fibrosis. Microscopically, berlin blue stain confirmed a marked increase in iron deposition within ductular hepatocytes and regenerative nodules. Based on the pathological findings, he was diagnosed with NH. Discussion This is the first report of the detection of NH after the diagnosis of galactosemia at newborn screening. In this case, the elevation of serum galactose was considered to have resulted from metabolic failure caused by NH. Liver transplantation is relatively successful for NH. Early recognition of NH is important for the early selection of a donor and transfer to a center specializing in pediatric liver transplantation, which will help improve survival. When galactosemia is diagnosed at newborn screening, it is important to consider the presence of NH if the infant presents with chronic end-stage liver failure.

349 RAPIDLY-PROGRESSING HEPATOCELLULAR CARCINOMA IN A 7 YEAR OLD BOY WITH FRUCTOSE 1,6-BISPHOSPHATASE DEFICIENCY WITH ALDOLASE-B DEFICIENCY-LIKE PHENOTYPE - A CASE REPORT. J.S. Whatley, Pediatrics, Helen DeVos Children's Hospital, Grand Rapids, Michigan, UNITED STATES|J.S. Whatley, Pediatric Residency Program, Grand Rapids Medical Education Partners, Grand Rapids, Michigan, UNITED STATES|H.A. Conrad, Pediatric Gastroenterology, Helen DeVos Children's Hospital, Grand Rapids, Michigan, UNITED STATES|D.S. Dickens, Pediatric Hematology/Oncology, Helen DeVos Children's Hospital, Grand Rapids, Michigan, UNITED STATES|J.M. Steinke, Pediatric Nephrology, Helen DeVos Children's Hospital, Grand Rapids, Michigan, UNITED STATES|.

Introduction Although hepatocellular carcinoma (HCC) is a well-established occurrence in the setting of cirrhosis, HCC ultimately resulting from chronic liver injury due to fructose 1,6-bisphosphatase deficiency (FBPD) has not been well described in literature. Here, we present a case of FBPD involving cirrhosis with portal hypertension and Fanconi syndrome on initial presentation, a clinical course complicated by chronic non-compliance, and the eventual development of rapidly-progressing hepatocellular carcinoma. **Case Description** A 7 year old boy with history of FBPD, ADHD, behavioral issues, chronic non-compliance, cirrhosis, and Fanconi syndrome presents with three-week history of worsening diffuse abdominal pain, nausea, feeding intolerance, weight loss, abdominal distension, diarrhea, restlessness, and mild shortness of breath. On physical exam, he had a markedly distended, tense abdomen with diffuse tenderness and guarding, hepatomegaly to the level of the umbilicus, and no jaundice. Laboratory studies showed interval elevation of liver enzymes. Abdominal ultrasound revealed a focal hypervascular hepatic mass in the left lobe, increased liver size from previous studies, splenomegaly, portal hypertension, and gallbladder varices. An alpha-fetoprotein level was markedly elevated at 60,500, which was dramatically increased from 1039 six months prior. An abdominal MRI revealed a widespread heterogeneous appearance to the liver and an infiltrative process consistent with hepatocellular carcinoma in the left lobe of the

liver. A liver biopsy of the lesion was performed and showed moderately differentiated hepatocellular carcinoma. Three months prior, a liver ultrasound showed no focal lesions. One month prior, an ultrasound-guided liver biopsy was normal. Immediately following the initial diagnosis of FBPD at age three, his primary interventions were strict dietary restriction of fructose-containing foods, maintaining normal glucose levels with continuous G-tube feedings at night, and electrolyte supplementation for co-occurring Fanconi syndrome. The next four years were complicated by worsening dietary non-compliance secondary to growing behavioral issues and the separation of his parents with resulting home life instability. In the five months prior to the diagnosis of HCC, he had been hospitalized on three occasions for non-compliance. During each hospitalization, he had improvement with a strict fructose-free diet in an inpatient setting. Intervention Given the extent of the HCC and the lack of viable treatment options, he entered hospice comfort care. He succumbed to the disease one week after diagnosis. Discussion This patient had a dramatic presentation of FBPD with presence of cirrhosis, portal hypertension, and Fanconi syndrome, all of which are more characteristic of aldolase-B deficiency. HCC is described as a serious complication of other meabolic diseases. Though this was a unique case of FBPD, this report suggests that HCC can occur in the setting of FBPD and screening for HCC is warranted in patients with FBPD, especially if the disease process is poorly controlled.

Saturday, October 10, 2015

POSTER SESSION III

Exhibit Hall

12:00pm – 2:00pm

Presenters at posters 12:30pm – 2:00pm

ENDOSCOPY/QI/EDUCATION

384 *MULTIPLE GIANT MID-THORACIC ESOPHAGEAL DIVERTICULA IN A 14-YEAR-OLD MALE.* M. Cho, J. Lim, C. Snyder, D. Rivard, Pediatric, Children's Mercy Hospital, Kansas City, Missouri, UNITED STATES| .

Esophageal diverticula in the pediatric population are a rare occurrence and multiple esophageal diverticula are an even more rare phenomena. The etiologies and presentations can vary widely which can makes it difficult to diagnose without imaging or direct visualization. We are reporting a case of a 14-year-old male with a history of asthma that presented with progressive chest and back pain over a period of five months. Coughing and movement without back support exacerbated pain. He also experienced night sweats and poor appetite with a subsequent fifteen-pound weight loss. He was seen multiple times in the Emergency Department and pain was initially believed to be secondary to his asthma. He endorsed no symptoms of dysphagia. A chest x-ray showed a widened superior mediastinum. A chest computerized tomography was obtained and showed osteomyelitis of the T5 and T6 vertebra and findings concerning for mediastinal abscesses. Magnetic resonance imaging later revealed esophageal diverticula with mediastinal phlegmon extending to the affected vertebra. Bone cultures showed *Parvimonas micra*, *Capnocytophaga* species, and *Streptococcus intermedius*. Initial treatment included IV antibiotic therapy, bypassing the affected area with a nasogastric tube, and a cervical thoracic orthosis brace for spine stability. As his osteomyelitis resolved, esophagogastroduodenoscopy was safely performed in which multiple approximately 5cm esophageal diverticula and a small hiatal hernia were discovered. There were no strictures or fistula visualized and the mucosa appeared normal. Biopsies obtained from the esophagus, hiatal hernia, gastric antrum, and duodenum were normal with the exception of mucosal eosinophilia in the gastric antrum. With resolution of his osteomyelitis, he is back at his baseline without pain or dysphagia. Currently, the patient is being closely followed with no surgical intervention planned.

386 *SMALL BOWEL HEMANGIOMA PRESENTING AS GASTROINTESTINAL BLEEDING IN A TEENAGE PATIENT.* X. Liu, Pathology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES|P. Conjeevaram Selvakumar, A. Anani, B. Maksimak, V. Hupertz, Pediatric Gastroenterology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES|.

Background: Hemangiomas of the small bowel are rare contributing to 0.05 % of all intestinal neoplasms. Most of them usually present as occult or massive gastrointestinal (GI) bleeding requiring blood transfusion in a significant proportion of patients. Herein we report a case of ileal hemangioma presenting as GI bleeding in a teenage patient. Case: A 13 year old male was admitted with a 3 day history of painless melena, preceded by a month's history of intermittent epigastric pain. Prior to presentation he was seen by Pediatrician and was started on Omeprazole with no response. He had regular, normal consistency stools prior to admission. He did not have cutaneous lesions, musculoskeletal abnormalities, mucosal lesions, facial dysmorphism and his abdomen was soft, non-tender, non-distended without organomegaly. The remainder of his examination was also normal. His labs on admission revealed Hemoglobin (Hb) of 11.3, White blood cell count of 14.5, Platelet count of 361,000 and Erythrocyte Sedimentation Rate of 7. His liver function panel, renal function panel and coagulation studies were normal. One day after admission, his Hb dropped to 8.7 requiring packed red blood cell transfusion despite no hematemesis or melena. Due to this significant drop in Hb he underwent an Esophagogastroduodenoscopy (EGD) and colonoscopy. His EGD was normal but his colonoscopy revealed a 5 mm mass containing an adherent clot in the terminal ileum. He had a wireless capsule endoscopy which revealed multiple sessile lesions in the ileum with no bleeding. He had another significant episode of GI bleed, so he underwent laparoscopic resection of this ileal lesion during which prominent mesenteric nodes were noted. The histology of this lesion revealed lobular capillary hemangioma with no significant endothelial atypia or multilayering and the mesenteric lymph nodes demonstrated benign lymphoid hyperplasia. Discussion: Hemangiomas are benign vascular tumors which can be classified histologically as capillary, cavernous and mixed types. GI hemangiomas are more commonly found in small intestine (jejunum being the most commonly reported) than colon and rectum. They can be solitary or multiple. Other clinical presentations include obstruction, intussusception and perforation. They can be seen in association with Maffucci syndrome, Klippel-Trénaunay syndrome, disseminated neonatal hemangiomatosis, Proteus syndrome and Blue rubber bleb nevus syndrome. The rarity of occurrence and location in small bowel make them difficult to diagnose. Usual diagnostic methods include imaging such as CT, MRI, angiography or scintigraphy and endoscopic techniques such as EGD, Colonoscopy or capsule endoscopy. Sometimes patients might need laparoscopy or even exploratory laparotomy for diagnosis and location of small bowel hemangiomas. Medical management of small bowel hemangiomas can include corticosteroids, propranolol and interferon alpha-2a but the response to medical management is variable. Endoscopic methods such as snare polypectomy and cauterization are also reported. Surgical resection might be needed if there is no response to medical management or persistent GI bleeding.

387 *USE OF AMINOCAPROIC ACID IN A PATIENT WITH MULTIFOCAL LYMPHANGIOENDOTHELIOMATOSIS WITH THROMBOCYTOPENIA.* L. Fahey, E. Semeao, Division of Gastroenterology, Hepatology and Nutrition, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES|.

Multifocal lymphangioendotheliomatosis with thrombocytopenia (MLT) is a very rare multiorgan vascular disorder which presents in infancy. This disease was first described in 2004, and there have been 20-30 additional cases reported, commonly associated with severe gastrointestinal bleeding. This case reviews an 8 week old female who presented with anemia, thrombocytopenia, and hematochezia as well as red, blanching, macular lesions on her back. Her bowel ultrasound suggested vascular malformations in bowel loops in both the right lower quadrant and left upper quadrant of the abdomen. CT abdomen and pelvis enterography showed two vascular lesions in the ileum and jejunum. She was clinically diagnosed with MLT, which is characterized by the presence of multiple red to brown macules, papules and plaques on the skin as well as throughout the gastrointestinal tract from the esophagus to the sigmoid colon. The number of lesions can vary from as few as five to as many as several hundred lesions. Lesion size ranges from a few millimeters to five centimeters. MLT is associated with gastrointestinal bleeding, significant thrombocytopenia and anemia. The thrombocytopenia is due to platelet destruction within

the vascular lesions. Thrombocytopenia and gastrointestinal bleeding is most significant in the first year of life. In addition to the gastrointestinal tract, patients with MLT can have involvement of the lung, brain, spleen, bone and muscle as well. This patient had a biopsy of one of her skin lesions which showed dilated superficial dermal capillaries which represented capillary malformation. Immunohistochemical staining for lymphatic markers monoclonal antibody D2-40 and lymphatic vessel endothelial receptor-1 are typically positive in MLT, however cases have been reported based on clinical symptoms alone. This patient's staining was negative. In addition to skin biopsy, confirmation of this diagnosis can be made by performing an endoscopic biopsy of the gastrointestinal lesions. This patient has not had an endoscopy to date since she is clinically stable. Treatment of MLT is aimed at controlling the progression and subsequent bleeding of vascular lesions. There is currently no specific standard therapeutic regimen. Previous treatments that have been trialed include propranolol, systemic steroids, interferon alpha-2a, intravenous immunoglobulin, thalidomide, vincristine, bevacizumab, and argon plasma coagulation. This patient is currently undergoing treatment with aminocaproic acid as well as ranitidine, with no gross gastrointestinal bleeding since initiating this therapy. This case report suggests that aminocaproic acid should be considered as a potential treatment option for this rare disease.

388 *THE SAFE AND EFFECTIVE USE OF SIROLIMUS TO TREAT GASTROINTESTINAL BLEEDING IN BLUE RUBBER BLEB NEVUS SYNDROME.* L.J. Febo, A. Fifi, A. Langshaw, Pediatrics, University of Miami/ Jackson Memorial Hospital, Holtz Children's Hospital, Miami, Florida, UNITED STATES].

Blue Rubber Bleb Nevus Syndrome (BRBNS) is a rare condition characterized by growth of venous malformations on various tissues, most commonly the skin and gastrointestinal (GI) tract. There are only around 200 cases reported in literature. Significant morbidity and mortality depend mostly on the extent of GI involvement since these lesions can range from mild bleeding to severe GI hemorrhage and death. Other complications may include joint pain, central nervous system hemorrhage, epistaxis, among others. Currently there is no established treatment protocol for BRBNS, therefore it is important to be aware of available treatment options. We describe a case of a 6-year-old boy with BRBNS treated with Sirolimus 0.05 mg/kg/day. At birth he had a lesion on abdomen that was surgically removed, then later diagnosed at age 1 when blebs continued to grow on his skin, mostly on his left lower extremity. For the next 4 years these lesions had increased bleeding and became more painful which made it difficult for patient to ambulate and required him to use crutches. Since most of the patients with BRBNS have GI involvement, routine endoscopies/colonoscopies were performed yearly to monitor for the presence of GI lesions. At the age of 5, actively bleeding lesions were found at the sigmoid colon. Given progression of disease it became obvious that patient required medical intervention. We decided to start patient on Sirolimus, an immunosuppressant with antiangiogenic properties that has been used to treat other vascular lesions due to its potential to decrease the production of vascular endothelial growth factor. Before starting the medication, patient was first screened for immunodeficiencies, latent TB, EBV, and CMV, all of which were negative. Due to the adverse side effects Sirolimus may cause, patient was started on a low dose of 0.015 mg/kg and was slowly increased to 0.05 mg/kg over 8 months. Target blood levels were increased from 3-6 ng/mL to a level of 7-9 ng/mL. No other antiangiogenic drugs were trialed before starting Sirolimus. As dose of Sirolimus was increased the patient's skin blebs markedly decreased in size, were no longer bleeding, or tender on palpation. He had no further GI bleeding. Some of the side effects of Sirolimus include hepatic and renal damage, bone marrow suppression, and increased susceptibility to infections, for which patient is taking prophylactic TMP/Sulfa. Our patient has experienced only minimal side effects, including appetite changes and increased irritability, now resolved. There is another pediatric case report where BRBNS was treated with Sirolimus but at a lower target blood level due to fear of significant side effects. For our case we were able to use increased target blood levels of Sirolimus, which reduced morbidity and improved overall quality of life without any major side effects. We recommend Sirolimus be considered as a treatment option for severe, difficult to control BRBNS at a low dose and slowly be increased as needed to achieve the desired therapeutic effect. Patients should be screened prior to starting therapy to make sure there is no underlying immunodeficiency or latent infection.

389 *"IS THAT A TUMOR!?"*: A RARE CASE OF A PEDIATRIC GASTRIC XANTHOMA. D. Garcia, H. Sadek, A. Soroush, Pediatrics, Jersey Shore University Medical Center, Neptune, New Jersey, UNITED STATES|T. Matulewicz, Pathology, Jersey Shore University Medical Center, Neptune, New Jersey, UNITED STATES|.

Objectives: 1. Understand the endoscopic and microscopic make-up of xanthomas. 2. Consider xanthomas in the differential diagnosis of masses of the gastric mucosa. 3. Understand the role of EGDs in the differentiation of gastric xanthomas, ulcers and malignancies. Case Summary: A 16 year old white female was admitted to the pediatric floor with a history of ALL and AML (in remission for 11 years), diabetes mellitus Type 1, monosomy 7, mixed hyperlipidemia, chronic intermittent abdominal pain, excessive gassiness and intermittent constipation. She presented with 5 days of sharp, right-sided abdominal pain, vomiting and decreased oral intake. Physical exam was significant for tenderness of the right upper quadrant area. No rebound tenderness, Murphy's sign, hepatosplenomegaly or masses were appreciated. There were no xanthomas or xanthelasmas present on her skin. CBC, CMP, Amylase, Lipase and celiac profile were normal. Gallbladder and pelvic ultrasound showed no significant findings. KUB revealed non-obstructive bowel gas pattern. The patient was treated for constipation and started on a PPI which provided some symptom relief. Stool H. Pylori Ag was positive. Lipid panel obtained was as follows: Cholesterol 214, TG 168, LDL 133. An EGD and biopsy was performed, and she was found to have two duodenal ulcers, bile reflux in the stomach, gastritis and a small hiatal hernia. Three nodular lesions were seen in the body and antrum of the stomach, all less than 1 cm in size with a whitish, yellow surface. Biopsy was obtained from largest of these lesions with the tissue noted to be friable but ceased bleeding without any further intervention. Microscopic exam revealed duodenal ulcerations, gastric metaplasia in the duodenum, chronic gastritis and scattered organisms compatible with H. Pylori seen in the gastric biopsies and the xanthoma. Clo test was found to be positive for H. pylori. Nodular lesions were found to be gastric xanthomas with inflammatory changes and H. pylori. There was a collection of large cells with eccentric nuclei and an abundant amount of surrounding cytoplasm that had a foamy appearance filled with lipid (xanthoma cell). There were no malignant cells. She had decreased levels of duodenal disaccharides. Conclusion: Gastric xanthomas are benign tumor-like lesions made up of clusters of lipid containing foamy histiocytes located mainly within the antrum and pylorus region of the stomach. Although gastric xanthomas are seen frequently in elderly, it is rarely reported in pediatric population. Multiple etiologies have been proposed in the pathogenesis of gastric xanthomas. Our patient had multiple contributing factors including hypercholesterolemia, H.Pylori and bile gastritis. The differential diagnosis includes granular cell tumor, atypical mycobacteria infection and mucinous cell-type epithelial neoplasm. An upper gastrointestinal endoscopy and histologic examination is essential in the diagnosis of gastric masses for both the differentiation of not only benign vs malignant neoplasms but also in preventing the unnecessary use of acid blocking agents in attempt to treat incorrectly diagnosed gastric ulcers.

390 *PEDIATRIC GASTRIC SPINDLE CELL NEOPLASM/PLEXIFORM FIBROMYXOMA PRESENTING WITH FAILURE TO THRIVE, ENDOSCOPIC FINDINGS AND DIAGNOSTIC APPROACH.*. M. Haight, R. Gugig, Pediatric Gastroenterology, University of California San Francisco- Fresno , Fresno, California, UNITED STATES|M. Haight, M. Favreau, Oregon Health & Sciences University, Portland, Oregon, UNITED STATES|.

Spindle cell cancer neoplasms have an extensive differential diagnosis requiring extensive immunohistochemical testing. These tumors can occur in the stomach, and are quite rare. An 11 year old boy presented with cachexia and severe weight loss. He was found on CT imaging to have a heterogeneous enhancing mass protruding submucosally into the gastric lumen in the antrum, pylorus and partially into the duodenum. On endoscopy, the mass was partially obstructing the pylorus in a ball valve" manner. Using endoscopic ultrasound, the mass was biopsied after opening the gastric mucosa with an endoscopic cautery knife. The incision was closed with endoscopic clips. The lesion was subsequently removed surgically with good margins and with full recovery of nutrition status and gastric function. Immunohistochemical staining with AB, SMA and CD10, along with morphology confirmed that the low grade plexiform spindle cell neoplasm was most consistent with the rare entity known as plexiform fibromyxoma or plexiform angiomyxoid myofibroblastic tumor. This is a benign tumor. GIST (gastrointestinal stromal tumor) was excluded due to CD117 and DOG1 negativity. Sarcomatoid carcinoma was

excluded due to pankeritin negative staining. Fibromatosis was excluded due to no nuclear beta-catenin staining. Other tumors were also excluded due to negative staining: neural tumors (S100, SOX10), smooth muscle tumors (Desmin, Caldesmon), IFP sarcoma (HHV8, CD34), PeComa or melanoma (HMB45), IMT (ALK, CD30, Desmin, no plasma cells on CD138) and low grade fibromyoid or sclerosing epithelioid fibrosarcoma (MUC4)."

391 *GASTRIC ADENOCARCINOMA IN A TEENAGER WITH RECURRENT H. PYLORI ULCERATION*. M. Haight, R. Quan, R. Gugig, Pediatrics, UCSF, Fresno, California, UNITED STATES | M. Haight, M. Favreau, Oregon Health and Sciences University, Portland, Oregon, UNITED STATES | .

Gastric adenocarcinoma is rare in pediatric patients. We encountered a teenager who initially presented with gastric ulcers associated with H. pylori, who then returned with persistent H. pylori ulcerations despite treatment. Subsequent endoscopy confirmed the adenocarcinoma. A 15 year old previously healthy female presented initially with endoscopic findings of a lesser curvature gastric ulcer with associated H. pylori. Antibiotic and proton pump therapy was completed with transient improvement in symptoms. She then presented with severe abdominal pain, anemia and hematemesis. Follow up endoscopy revealed a large ulcer of the lesser curvature with improved gastritis, persistent H. pylori. Ulcer margins had unusual hypertrophied mucosa that came off in large clumps when biopsied. Further work up revealed possible mesenteric involvement (abdominal cytology negative). liver lesions (adenocarcinoma metastasis) and thickened gastric wall. Review of the initial endoscopic biopsies confirmed the presence of H. pylori, gastritis with ulceration, but no neoplastic changes. Follow up endoscopy had few H. pylori with CLO test positivity, gastritis with ulceration, and immunohistochemistry positive staining for pankeritin, HER2 negative, and moderate to poorly differentiated adenocarcinoma. She is currently on her 5th round of chemotherapy with no complications.

396 *THE FEASIBILITY OF ENDOFLIP IN PEDIATRIC EOSINOPHILIC ESOPHAGITIS PATIENTS*. R.A. Lirio, J.R. Lightdale, Pediatric Gastroenterology/Hepatology/Nutrition, UMASS Memorial Children's Medical Center, Worcester, Massachusetts, UNITED STATES | J. o'dea, Crospon, Galway, IRELAND | .

Eosinophilic esophagitis (EoE) in children can lead to marked inflammation and eventual stricturing of the esophagus, leading to dysphagia and food impactions. Currently, diagnosis is made via pathology and the eosinophilic count noted per high power field in the mid and distal esophagus. EndoFLIP is a novel technology that integrates multiple adjacent impedance planimetry electrodes into a balloon catheter as a means of precisely measuring esophageal dimensions and sphincter pressures without the need for fluoroscopy. In a previous case report, we reported using this system for successfully dilating an esophageal stricture in a 17 year old with EoE. Aim: To test the technical feasibility of EndoFLIP for the measurement of esophageal luminal and sphincter pressures in pediatric EoE patients. Methods: We obtained informed consent/assent to use the EndoFLIP EF-325 balloon catheter (Crospon, Galway, Ireland) to evaluate 2 patients with suspected or known EoE (Table 1), ages 13 and 3 years old. The balloons were introduced under endoscopic visualization, and approximately centered within the esophagus, as well as the upper and lower esophageal junctions. The endoscope was then retracted to remain in the upper esophagus for the duration of the procedure as pressures were measured. Our goals were to obtain precise measures of (1) the esophageal lumen and (2) upper and lower esophageal sphincters. Results: The EndoFLIP balloon catheter was safely deployed without complication. The pressures of the proximal esophagus were measured in both patients and noted to be 37.0 mm Hg, 57.8 mm Hg in the 13 y/o and 3 y/o, respectively. This correlated well with the sensation of a rather tight esophageal sphincter on passing the scope in the 3 y/o. Just proximal to the LES, measurements were obtained noting pressures of 36.1 mm Hg and 47.5 mm Hg in the 13 y/o and 3 y/o, respectively. At the LES, the 13 y/o had a pressure of 15.3 mm Hg and the 3 y/o, 25.1 mm Hg. Approximate diameters were also measured using the EndoFLIP system. (Table 2) Conclusion: Our case series documents the first use of EndoFLIP in pediatric EoE patients for measurement of pressures and luminal diameter without the need for fluoroscopy and radiation. This new diagnostic modality may provide a new pathway for assessing risk for strictures in this patient population, and at the same time, if needed, can also serve as a dilating/therapeutic device (EsoFLIP). Reference values using this technology will still need to be validated, but we

believe the precision afforded by this technology combined with the benefit of real time esophageal measurements represents a valuable and novel approach to the management and diagnosis of children with EoE and is worthy of further investigation.

398 *AN UNCOMMON CAUSE OF HEMATEMESIS: DUODENAL PERFORATION FOLLOWING NON-ACCIDENTAL TRAUMA*. T. Ratchford, B. Lewis, J.P. Middleton, Pediatrics, University of Virginia, Charlottesville, Virginia, UNITED STATES|.

Introduction: Hematemesis is relatively common problem in children, with endoscopy playing a role in the diagnosis and management of acute upper gastrointestinal bleeding. Duodenal perforation is a rare cause of abdominal pain and vomiting in children most often due to blunt or penetrating trauma.¹ Non-accidental trauma (NAT) is the most common cause of duodenal perforation in children under the age of five.² Diagnosis of duodenal perforation is most often made with computed tomography (CT) imaging. We present a case of acute hematemesis from duodenal perforation following non-accidental trauma diagnosed by upper endoscopy. Presentation: An otherwise healthy sixteen-month-old Hispanic male presented with decreased oral intake and multiple episodes of vomiting with hematemesis and bile stained fluid. Examination revealed a well-grown toddler with fever, tachycardia and non-tender, non-distended abdomen. Initial laboratory evaluation demonstrated mild transaminitis (aspartate transaminase 49, alanine transaminase 83 U/L), normal hemoglobin (10.7 g/dL) and normal white blood cell count (13.5 k/uL). Imaging with abdominal x-ray and ultrasound was unremarkable. The patient was taken for upper endoscopy the following morning after his hemoglobin dropped 2 g/dL. Endoscopy revealed normal appearing esophageal and gastric mucosal with a large blood and bile stained ulceration in the second portion of the duodenum. A CT abdomen confirmed duodenal perforation with retroperitoneal air and leakage of contrast. A NAT work-up demonstrated a liver laceration and a subacute left distal humerus fracture. Following resection of the distal duodenum with duodenojejunal anastomosis and placement of jejunal tube, the patient recovered fully and had his jejunal tube removed after two months. Despite a full investigation, the circumstances surrounding the injury were unclear and the child remains in custody of his biologic parents who removed him from his daycare center. Discussion: More than 670,000 children are victims of child abuse or neglect annually with 1520 children dying of NAT in 2013. Abdominal trauma from NAT is unusual and accounts for only 8% of the duodenal injuries described in a large pediatric trauma series.¹ Because the duodenum lies in the retroperitoneal space, abdominal imaging must include CT since retroperitoneal air will not be seen on an abdominal x-ray. Although abdominal pain and bilious emesis are common in children with duodenal perforation, to our knowledge, our case is the first to describe duodenal perforation from NAT presenting with acute hematemesis and diagnosed with endoscopy. ¹Desai KM, Dorward IG, Minkes RK, Dillon PA. Blunt Duodenal Injuries in Children. J Trauma 2003; 54: 640-646. Gaines BA, Shulz BS, Morrison K, Ford HR. Duodenal Injuries in Children Beware of Child Abuse. J Pediatr Surg 2004; 39: 600-602. U.S. Department of Health and Human Services, Administration for Children and Families, Administration on Children, Youth and Families, Children's Bureau. (2015). Child maltreatment 2013.

399 *ABDOMINAL WALL THICKENING AND HENOC SCHÖNLEIN PURPURA - AN ATYPICAL "RELATIONSHIP"*. W. Ruan, A.S. Patel, Pediatric Gastroenterology, UT Southwestern Medical Center, Dallas, Texas, UNITED STATES|W. Ruan, A.S. Patel, Children's Health, Dallas, Texas, UNITED STATES|.

Henoch Schönlein Purpura (HSP), also called immunoglobulin A vasculitis, is a systemic vasculitis that commonly affects children. Its typical presentation involves palpable purpura, joint pain, kidney disease, and abdominal pain. There is no definitive diagnostic laboratory or imaging test for this disease. What complicates the diagnosis of HSP further is that abdominal pain can precede the classical skin findings in up to two thirds of the cases of HSP. We describe a case of an 8 year old female who presented to our hospital with abdominal and joint pain. Eventually, she was diagnosed with atypical HSP; however, her diagnosis was complicated by the lack of palpable purpura and the histopathological findings on initial esophagogastroduodenoscopy of foveolar hyperplasia secondary to Ménétrier's disease versus a reactive healing process. She also initially had bowel wall thickening noted on a CT

scan prior to the EGD that was attributed to gastritis instead of HSP. Given the complexity of the diagnosis of her atypical HSP, we conducted a review of HSP patient encounters at our institution over the last 5 years. Specifically, their medical records were examined for any imaging performed. A total of 110 diagnosed HSP patient charts were examined. Of those, 60 patients had abdominal imaging of which 17 patients had CT scans. 11 of those patients had diffuse bowel wall thickening noted on their CT scans. 15 out of 53 patients with abdominal ultrasounds had abdominal wall thickening noted as well. Given these numbers, it appears that imaging may be useful in the diagnosis of atypical HSP. It was concluded that abdominal imaging in children can assist in the diagnosis of HSP especially if they do not present with classical palpable purpura. Many children with HSP who had abdominal imaging were discovered to have signs of abdominal wall thickening. This finding is not commonly used to aid in the diagnosis of HSP; however, in the setting of other symptoms and laboratory tests, it can be useful towards diagnosing atypical HSP. Given this prevalence and the difficulty in diagnosing HSP, other modalities, specifically noninvasive imaging, should be considered to aid in the diagnosis. When abdominal wall thickening is found on imaging performed on patients with abdominal pain in the context of other symptoms, HSP should be considered in the differential diagnosis.

401 *A RARE CAUSE OF RECURRENT IRON DEFICIENCY ANEMIA.* S. Sankararaman, J. Moses, Department of Pediatrics, Pediatric Gastroenterology division, Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES|H. Shojaei, R.W. Raymond, Department of Pathology, University Hospitals Case Medical Center, Cleveland, Ohio, UNITED STATES|M. Dingeldein, Department of Pediatric Surgery, Rainbow Babies and Children's Hospital, Cleveland, Ohio, UNITED STATES|.

Gastrointestinal stromal tumors (GIST) are rare neoplasms of the gastrointestinal (GI) tract. GIST are mesenchymal tumors with strong expression of kit protein (CD 117 or CKIT). About 1.4 - 2.7% of all GIST occur in children. Pediatric GIST differ from their adult counterparts in several ways: they are more common in females, predominantly occur in the stomach, lack CKIT or PDGFRA mutations, and generally have a protracted course even in cases with lymph node metastases. Chronic anemia from gastrointestinal bleeding is the most common symptom and surgical resection with negative tumor margin is usually curative. Our patient is an 11-year-old male with history of hydrocephalus admitted with ventriculoperitoneal shunt malfunction. He also had history of severe iron deficiency anemia of unknown etiology for at least four months. His anemia persisted despite treatment with blood transfusion and oral iron therapy. Two weeks prior to this admission, he had an episode of coffee ground emesis. On admission, he was pale and physical examination was otherwise unremarkable. His Hb was 5.5 g/dl and serum albumin was 3.2 g/dL. He required blood transfusion. Esophagogastroduodenoscopy revealed a 4 cm wide, smooth, vascular mass in the antral region and computer tomogram of his abdomen revealed a hypoattenuated mass. He underwent diagnostic laparoscopy followed by open resection of the gastric mass. Histopathological examination revealed a multilobulated tumor arising in the muscularis propria of the gastric body with focal extension to mucosa and serosa. Histology showed a mixed epithelioid and spindle cell pattern with low mitotic rate, minimal atypia, and no areas of tumor necrosis which are typical features of GIST. Immunohistochemistry showed strong expression of CD117 (CKIT). Genetic analysis revealed no abnormalities in the commonly mutated exons of CKIT and PDGFRA. He is currently under follow up and has not had any further episodes of GI bleeding or anemia.

402 *RECTAL BLEEDING - HOW COMMON IS AN INTESTINAL LYMPHOMA IN CHILDREN?* S. Sood, L. Cukaj, S.H. Berezin, Pediatrics, Division of Pediatric Gastroenterology, Hepatology and Nutrition, New York Medical College, Valhalla, New York, UNITED STATES|.

We present the case of a 4 year old previously healthy male admitted with two days of painless rectal bleeding, described as large volume, bright red blood with clots mixed with dark formed stool and Hb: 8.5 g/dL- normocytic anemia. Past medical history was significant for chronic constipation. Physical exam was remarkable for fullness in the suprapubic and right hemi-abdomen and an anal fissure at 6'o clock position. Initial abdominal X-ray showed fecal impaction. Patient was given a Golytely clean out. Meckel's scan was unremarkable. Infectious colitis was

ruled out. Upper endoscopy and colonoscopy was remarkable for a large polypoidal mass in the cecum obscuring about two thirds of the lumen, friable, no stalk was identified, ileocecal valve was deviated to the right and incompletely visualized. CT abdomen and pelvis with Iv contrast showed a large soft tissue mass in the pelvis and lower abdomen predominantly on the right measuring approximately 7.8 x 4.6 x 5.6 cm extending from the rectovesicular pouch to the inferior tip of the liver with a dumbbell configuration. Prominent vessels were seen overlying the periphery of the mass which are SMA mesenteric branches to the terminal ileum and the right colon. The mass extended into the cecum and ascending colon. There was associated mesenteric adenopathy with the index node measuring up to 1.6 x 2.4 cm. Given the location and appearance of the mass the likely diagnosis was a non-Hodgkin's type lymphoma originating in mesenteric lymph nodes or in bowel wall. On exploratory laparotomy, the tumor was resected in entirety along with the cecum, the ascending colon and part of the terminal ileum. Primary re-anastomosis was performed. Bone marrow aspirate, CSF and pericardial fluid was negative for malignant cells. Histopathology: H and E sections through the ileocecal mass extending to the appendix revealed proliferation of monomorphic lymphoid cells in the background of a starry sky pattern with high nucleus to cytoplasmic ratio, negative EBV stain. Based on morphologic, immunophenotypic and FISH study this was a high grade, aggressive diffuse large B-cell lymphoma. Post-operatively, patient tolerated full enteral feeds and has received chemotherapy with Vincristine, Cyclophosphamide, Doxorubicin and Rituximab. Discussion: Primary tumors of the gastrointestinal (GI) tract are rare in children and represent less than 5% of all pediatric neoplasms. Diffuse Large B cell lymphomas are very similar to Burkitt's or Burkitt-like lymphomas and represent 10-20% of all pediatric Non-Hodgkin's Lymphomas. Commonly seen in the second decade of life, with primary mediastinal involvement. In the GI tract, ileocecal junction is the most frequent site. Clinically, they present with symptoms ranging from abdominal mass to acute abdominal emergency caused by intussusception, abdominal pain, vomiting, constipation, diarrhea or intestinal obstruction. These tumors are chemosensitive and can be treated effectively with primary chemotherapy with or without debulking surgery depending on the location and stage of the disease. In conclusion, this case is unique in its presentation of a primary intestinal lymphoma with painless rectal bleeding.

403 MESENTERIC VENOUS BYPASS PROCEDURE : EFFECTIVE AND SAFE TREATMENT FOR DUODENAL VARICES CAUSED BY SUPERIOR MESENTERIC VEIN THROMBOSIS. K.T. Toor, Y. ZHENG, Gastroenterology, Childrens Hospital Los Angeles, Los Angeles, CA, UNITED STATES MINOR OUTLYING ISLANDS].

Introduction: This is a case of a child presenting with progressively worsening large duodenal varices due to superior mesenteric vein (SMV) thrombosis. He was safely and effectively treated with mesenteric venous bypass procedure. Case Description : Our patient is a 9 year old, ex 32 weeker, african american male. His history is significant for necrotizing enterocolitis (NEC) in neonatal period with gut perforation requiring surgical repair with minimal small bowel resection. He also underwent Ladd's procedure for malrotation and volvulus at 2 years of age. He developed chronic diarrhea of unknown etiology after his second surgery leading to extensive workup including endoscopic evaluation. His first esophagogastroduodenoscopy (EGD) at 2 years of age showed mild duodenal varices which were mistakenly identified as thickened duodenal folds. EGD repeated at 7 years of age showed significant interval change. Multiple large varices were seen in the second portion of the duodenum extending into the proximal jejunum. Abdominal imaging confirmed SMV thrombosis and mesenteric varices. Banding ligation was not a therapeutic option in this case given the thin wall of the duodenum and the extent of duodenal varices. After extensive discussion with general surgery, vasular anomaly team, intervention radiologist, advanced gastroenterology procedurist and transplant surgeons, the decision was made to proceed with mesenteric venous bypass at 9 years of age. Pt underwent extensive lysis of adhesions. His jejunal vein was found to be thrombosed and fibrosed leading to drainage of 30- 40cm of small bowel via duodenal varices. A 3 mm anastomosis was created connecting the jejunal vein with a venous tributary from proximal jejunum. Repeat EGD two months after bypass surgery showed near complete resolution of the varices. Discussion : Our patient had history of umbilical line placement as a neonate and surgical repair of NEC as well as malrotation. Both procedures put him at increased risk of SMV thrombosis. SMV thrombosis is a well known entity more commonly seen in hypercoagulable states in conjunction with surgery. Clinical presentation is variable based on the extent of thrombosis and vessel

involvement. Chronic mesenteric venous thrombosis presents with complications of portal-vein or splenic-vein thrombosis such as esophageal variceal. Here, we are reporting that duodenal varices are an extremely rare complication of SMV thrombosis. They can be safely and effectively treated with mesenteric venous bypass procedure.

405 *CONGENITAL ESOPHAGEAL STENOSIS: A TALE OF TWO INFANTS*. R. Venkatesh, A. Katz, Pediatric Gastroenterology, MGH, Boston, Massachusetts, UNITED STATES|.

Purpose: The aim of this study was to review two cases of congenital esophageal stenosis (CES). Congenital esophageal stenosis (CES) is a rare clinical entity with an incidence of approximately 1: 25,000-50,000 live births. It is characterized by intrinsic circumferential narrowing present at birth, though may not necessarily symptomatic. Our first patient is a 7 month old with Trisomy 21 presenting with inspiratory stridor with feedings and emesis shortly after episodes of stridor. He underwent a triple scope (laryngoscope, bronchoscopy, endoscopy) to further evaluate and the endoscope was not able to pass through the distal esophagus with tight narrowing noted at the distal esophagus. He was treated with Savary dilation and has clinically improved. Our second patient is a 3 month old infant presenting with poor feeding and growth. An upper GI was notable for distal esophageal narrowing that has responded to Savary dilation. In a review of the literature, CES is often it is associated with esophageal atresia, but can also be an isolated finding as in our two patients. As in our cases, symptoms may range from dysphagia, food impaction to vomiting and malnutrition often presenting with the introduction of solid foods. Therapeutic management can vary with surgical approaches versus endoscopic dilation. We have found favorable outcomes with serial Savary dilation although long-term follow-up will be necessary.

406 *FAMILIAL ADENOMATOUS POLYPOSIS; SUCCESSFUL USE OF SIROLIMUS*. H.A. yuksekkaya, M. Gumus, A. Yucel, pediatric gastroenterology, N. Erbakan University, Meram Medicine of Faculty, Konya, TURKEY|H. Toy, Patology, N. Erbakan University, Konya, TURKEY|.

Background: Familial adenomatous polyposis (FAP) is characterized by the early onset of colonic polyposis. Unless these are removed, colorectal cancer develops inevitably. It is essentially a systemic disease, and it has also extra-colonic manifestations. It has recently been demonstrated that the mTOR pathway is activated in intestinal polyps; prophylaxis by sirolimus markedly inhibits the development of APC mutation-related polyposis in mouse models. Case: A 13-year-old boy has been presented with a 1-year-history of rectal bleeding and weakness. On the physical examination, height and weight were both in the 10 percentile. He has paleness and a nonspecific systolic cardiac murmur which was attributed to anemia. The rest of the physical examination was unremarkable. There was a family history in which his mother, grandmother and uncle had died from FAP. Laboratory examination revealed severe iron deficiency anemia. Hemoglobin level was 7.2 g/dL, ferritin was 3.7 µg/L, tumor markers were normal except CA19- 9:78 (normal range: 0-35), and other biochemical tests were normal. Endoscopic examination showed thousands of polyps in whole colon and three polyps in periampullary region in duodenum. In histological examination, colonoscopic polypectomy showed that adenomatous polyposis, severe and moderate dysplasia and duodenal polyps showed moderate dysplasia. His father was informed about colectomy, but he did not accept it. The family was informed about the side effects of sirolimus. We then received approval from the family to begin low-dose sirolimus as an anticancer agent. Polypectomy and pathologic evaluation were repeated again on 6,12, 24 month (from the largest of eight polyps and different region). After treatment with sirolimus, small polyps resolved completely in the duodenum and the size of colonic polyps decreased but did not disappear. Also, colonic high-grade dysplasia improved the moderate and mild dysplasia, and the level of hemoglobin increased from 7.2 g/dL to 14 g/dL by iron supplementation. Stool occult blood test and CA19-9 level were normalized. There was no drug adverse reaction at 30-month follow-up. To the knowledge, this is the first report related to the use of sirolimus in a patient with FAP. Results: This case has demonstrated that sirolimus has powerful antitumor effects in a patient with FAP. It has beneficial effects on both the size of colonic and duodenal polyps and on the severity of the dysplasia although further experience is required in that context.

FUNCTIONAL/MOTILITY

407 *MALIGNANT GASTROINTESTINAL PERIVASCULAR EPITHELOID CELL TUMOR PRESENTING AS COLONIC INTUSSUSCEPTION*. S. Taboada, Pathology, North Shore LIJ, New Hyde Park, New York, UNITED STATES | R. Ahlawat, J. Markowitz, Pediatric Gastroenterology, North Shore LIJ, New Hyde Park, New York, UNITED STATES | .

Perivascular epitheloid cell tumors (PEComas) are mesenchymal neoplasms defined by the presence of histologically and immunohistochemically distinctive perivascular epitheloid cells. They have been reported in various anatomic sites and usually occur in young adult females, although are rarely seen in children. Gastrointestinal (GI) PEComas are exceptionally rare and only a few case reports involving the colon, small intestine, rectum and stomach have been described. GI PEComas typically present as painless mass and occasionally with abdominal pain. We describe an unusual presentation of GI PEComa as intussusception. A 18 yo female with past medical history significant for constipation and rectal bleeding presented to the emergency department (ED) with 2 month history of recurrent abdominal pain. She had visited the ED almost 2 weeks prior for the pain and noted to have a benign abdomen. Abdominal x-ray revealed gas and stool in the colon suggesting constipation. Treatment with laxatives did not improve the pain. Subsequent laboratory evaluation showed anemia (Hgb 9.3 g/dL) and elevated inflammatory markers (ESR 42 mm/hr, CRP 1.39 mg/dL) raising concern for possible inflammatory bowel disease. EGD and colonoscopy was scheduled, however due to acutely worsened pain, she was referred to ED for further evaluation and imaging. On exam, abdomen was tender with palpable mass in left lower quadrant. CT abdomen and pelvis revealed colocolic intussusception involving mid to distal transverse colon with submucosal mass-like hyperenhancement at the distal end of the intussusceptum. An exploratory laparotomy with intussusception reduction and wedge resection of the distal transverse colon was performed. An approximately 4 x 3 cm, firm, nodular polypoid mass causing an intussusception was removed. Histopathology of the mass revealed tumor cells arranged in sheets and nests around thin-walled blood vessels. The tumor cells showed prominent and vesicular nucleoli, with granular eosinophilic to clear cytoplasm. On Immunohistochemical (IHC) staining, tumor cells were positive for HMB-45 and MelanA, and negative for S100, vimentin, SMA, desmin, AE1/AE3, and CD 117. The combined morphologic and IHC findings were consistent with a PEComa. The degree of pleomorphism, mitotic rate, necrosis and infiltrative border favored a malignant tumor, although due to rarity of this tumor, there is no firm criterion for malignancy. PET scan showed no evidence of metastasis. As signs of malignancy were exhibited, patient will undergo wide marginal resection of transverse colon and surgical staging, based on which further management will be decided. Although PEComas have been described in different organs and are ubiquitous, malignant GI PEComas are extremely rare. To our knowledge, this is the first case of GI PEComa presenting as colonic intussusception. Currently, surgical resection with a wide margin seems to be the mainstay of treatment. The benefit of adjuvant chemotherapy, radiation and immunotherapy has not yet been established. Due to their relative rarity, diagnostic criteria, optimal treatment strategies, and prognostic factors needs to be further investigated.

410 *RECTAL PROLAPSE AND CONSTIPATION AS THE PRESENTING FACTORS FOR A RECTAL DUPLICATION CYST: A CASE REPORT*. K. Bittar, S. Safder, Pediatric Gastroenterology , Orlando Health, Orlando, Florida, UNITED STATES | C. Bessett, R. Holcomb, Pediatrics, Orlando Health, Orlando, Florida, UNITED STATES | .

Uncommon presentation of a rectal duplication cyst (RDC) identified as the cause of recurrent rectal prolapse (RP) in an otherwise healthy toddler with known chronic constipation (CC). Physical findings on digital rectal exam (DRE) lead to further investigation with imaging that confirmed presence of rectal cystic mass. The clinical findings, diagnosis, and management of this patient are discussed in this case report. INTRODUCTION: RP in children is often thought to be secondary to an underlying condition. The most common causes are CC, diarrhea, cystic fibrosis, Hirschsprung's disease, neuromuscular and pelvic nerve disorders. There have been only 2 cases in the literature on the underlying etiology for RP being a RDC; both had a physical finding of posterior or posteriolateral (PL) mass on DRE, similar to our case. This case is not only a rare finding, but emphasizes the importance of DRE on children with RP. CASE PRESENTATION: A 20-month-old male, previously healthy, presented to an outpatient clinic with a chief complaint of recurrent RP since 8 months of age with a history of CC. It was described as a painless, non-bleeding mass that protruded during defecation and reduced spontaneously afterwards. Parents visualized

stool passing around the protrusion. The child was diagnosed with CC and was put on a regimen of polyethylene glycol. The prolapsing persisted despite softened stools. A referral was made to our pediatric gastroenterology clinic for evaluation. In the clinic, he had RP that was difficult to reduce. DRE revealed non-tender slightly mobile PL 2-3cm mass palpable 3cm inside rectum. He was admitted for further evaluation. Upon admission, a pelvis MRI revealed a rounded cystic mass in the retrorectal space consistent with RDC. Surgical excision (SE) was performed. Pathology confirmed diagnosis of RDC. The remainder of hospital course was uncomplicated. **DISCUSSION:** Alimentary tract duplications (ATD) can appear anywhere from mouth to anus. Less than 5% of them are RDC. About two-thirds of ATD present before the age of 2. ATD has a variety of presentations depending on location, most commonly nausea, vomiting and palpable abdominal mass. The most common presentation of a RDC is CC, rectal bleeding, or peri-rectal abscess. RDC is rarely presented as RP. It is important to suspect rare underlying pathology such as duplication cysts as the cause of RP in children. While CC is a common, stand-alone cause for prolapse, more serious pathology should be considered before this is determined. In the cases reported previously, as well as in our case, DRE revealed palpable rectal mass and was a key factor in diagnosing the RDC. Early diagnosis is key so that prompt SE can be made to prevent complications such as perianal abscess or fistula formation. In addition, cases of malignant transformation of rectal duplication cysts are reported in adults. **CONCLUSION:** DRE should be completed on every child seen with RP, and may lead to earlier detection of underlying etiology ultimately resulting in earlier intervention. While rare, RDC should be considered in the differential diagnosis of underlying etiology for RP especially in the setting of CC.

411 *A RARE PRESENTATION OF ACHALASIA IN AN OTHERWISE HEALTHY INFANT: A CONGENITAL FINDING?*. K. Bittar, S. Safder, Pediatric Gastroenterology, Orlando Health, Orlando, Florida, UNITED STATES | M. Maximos, Pediatric Gastroenterology, University of Florida, Gainesville, Florida, UNITED STATES |.

Introduction: Achalasia is a rare disorder in the pediatric population. There have been few cases in this age group, however, patients either had cricopharyngeal (CP) disease or the achalasia was associated with a syndrome or was post infectious such as Chagas disease (CD). We present a case of a young infant with Achalasia type 2 (AT2). **Presentation:** The patient is an 11-month-old Bahamian female who was born at full term. Symptoms started early in infancy with large volume emesis of breast milk. She was treated with gastroesophageal reflux (GER) precautions for 1 month until she was admitted to the local hospital for failure to thrive (FTT). During her initial hospitalization, she had an upper gastrointestinal series, which showed a narrowing of the distal esophagus with otherwise normal anatomy. An upper endoscopy confirmed this narrowing and she underwent esophageal balloon dilatation (EBD). She was also placed on anti-GER treatment with famotidine. She experienced relief of symptoms for 2 months until they progressively returned. She was subsequently referred to our hospital for evaluation. An Esophagram reviewed with radiologist confirmed positive bird's beak sign, highly suspicious for achalasia. Ph probe with impedance study was unremarkable. Esophageal manometry revealed failure of the lower esophageal sphincter (LES) relaxation with pressurization of the esophagus with repeated swallows. This pattern of esophagogastric junction obstruction and non-peristaltic pressurization in esophagus is concerning for AT2 pattern. Narrowing of pylorus was also seen. She subsequently underwent EBD up to 12 mm. In addition, due to the FTT (weight < 5 percentile), percutaneous endoscopic gastrostomy (PEG) tube placement was performed to assist in feeding needs. Botulinum toxin was also administered to the pylorus muscle secondary to noted pyloric channel narrowing. She consequently tolerated feeds via tube and was discharged to home with the plan of repeat EBD until she gains appropriate weight to undergo surgical intervention such as Heller myotomy. **Conclusion:** Achalasia, meaning "failure to relax", is characterized by impaired relaxation of the LES. It is rare in the pediatric population. Many of the reported cases are associated with Allgrove's syndrome, CD or are CP in nature. We believe our patient to be one of the few infants with no underlying medical problems who presented with this rare problem. Given patient was symptomatic since early infancy; we suspect this to be a congenital finding in the absence of a clearly identifiable syndrome.

414 *N OF 1 SPONTANEOUS TRIAL DEMONSTRATES EFFICACY OF SERUM BOVINE IMMUNOGLOBULIN FOR EMESIS (POTENTIAL CYCLIC VOMITING) AND GASTROINTESTINAL SYMPTOMS IN AN AUTISTIC PATIENT.* B.P. Burnett,

Medical Affairs, Entera Health, Inc., Cary, North Carolina, UNITED STATES | M. Dave', Gastroenterology, Texas Digestive Disease Consultants, Plano, Texas, UNITED STATES |.

Gastrointestinal (GI) dysfunction in children with autism spectrum disorders (ASDs) is unusually high compared to other children with reported rates of symptoms of up to 41% abdominal discomfort, 45% constipation, 77% diarrhea and 19% persistent diarrhea. Cyclic vomiting may also be linked to mitochondrial defects in autistic patients. While it is unknown whether there is a direct link with GI conditions and ASDs, safe, effective therapies are needed. Serum-derived bovine immunoglobulin/protein isolate (SBI) is a prescription medical food product for management of GI conditions. SBI has a multifaceted mechanism of action which includes binding to microbial components, maintaining GI immune balance, managing gut barrier function and improving nutrient utilization. A 12-year-old Caucasian female patient (46.7kg, 165cm, BMI 17.15) diagnosed with ASD, sensory processing disorder (SPD) and anxiety was referred to this practice on 10/2014 with vomiting, abdominal pain, and constipation that began 2 years prior. Previous therapies included rifaximin for possible small intestinal bacterial overgrowth, erythromycin for gastric motility, cyproheptadine for gastric accommodation, magnesium for constipation, and PPIs for possible gastric reflux. Digestive enzymes, amitriptyline, VSL#3 and various other probiotics, as well as B complex supplements were also attempted without success. The patient had prior normal EGD/Colonoscopy with biopsies, gastric emptying scan, and blood work as well as multiple admissions for nasogastric cleanouts due to constipation. Upon presentation, this patient still had 4-10 episodes of non-bilious, non-bloody emesis every night with weight loss. The presentation of the vomiting was not classical cyclic vomiting, but was considered as a possibility. During the day, she complained of nausea, abdominal pain and hard, painful bowel movements (BMs) with incomplete defecation despite daily polyethylene glycol 3350 and magnesium. Cholescintigraphy with cholecystokinin in 10/2014 was normal. SBI 5 g QD was added to VSL#3, B vitamins, polyethylene glycol 3350 and magnesium. Within 1 month, vomiting resolved completely and constipation improved to soft comfortable daily BMs. An N of 1 test for the effect of SBI spontaneously ensued: when the patient stopped SBI after 1 month, all her symptoms returned, and upon re-challenge with SBI, her symptoms resolved again. When she stopped SBI therapy a second time after another month, the symptoms again returned. The patient has now been on SBI daily since 2/2015 and is doing well with her condition fully managed. The patient has had a 4.5 kg weight gain from 10/2014 to 4/2015, normal appetite and no nausea. She also has had a remarkable improvement in her quality of life for the first time in years, able to attend school and participate in extracurricular activities. She continues therapy for her anxiety and SPD. More investigation is needed into the exact mechanism that may manage vomiting in this patient. This N of 1 trial suggests that SBI may be a safe option in patients with ASDs for management of GI conditions.

416 *ESOPHAGEAL DYSMOTILITY FOLLOWING INTESTINAL TRANSPLANTATION FOR MEGACYSTIS MICROCOLON HYPOPERISTALSIS SYNDROME (MMIHS): AN EXPANDED CLINICAL SPECTRUM FOR THIS ENTITY.* M.L. Goldschmidt, S. Kocoshis, J. Nathan, G. Tiao, M. Alonso, A. Kaul, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, UNITED STATES |.

Background: MMIHS is characterized by megacystis, abnormal urodynamics, microcolon, and a dilated, aperistaltic stomach and small intestine. Without bowel transplantation, mortality is high. A few patients reported after multivisceral transplant (MVTX) purportedly tolerate enteral nutrition, but no description of their esophageal motility has emerged. We follow 4 patients with MMIHS who underwent small intestinal or MVTX. When one developed dysphagia 5 years post-transplant, we studied in detail her (and the others') clinical symptomatology and esophageal function. Methods: Retrospective analysis of esophageal symptomatology and prospective esophagoscopy, esophageal histology, contrast radiography, and esophageal manometry. Results: These female patients are currently 10, 8, 7, and 6 years of age. Transplants were performed at ages 40, 10, 21, and 6 months of age, respectively. Three underwent antrectomy, gastrojejunostomy, and composite liver, pancreas, small bowel, and colon transplant. One underwent MVTx, including stomach, liver, pancreas, small bowel, and colon. All 4 retained native esophagus and lower esophageal sphincter (LES). Dysmotility was diagnosed at 54, 117, 68, and 100 months of age, respectively, and only 2 were dysphagic at diagnosis. The other two, endoscoped because of

acute diarrhea, had dilated, aperistaltic esophagi containing retained food. Contrast radiography and endoscopy showed esophageal dilation, dysmotility, and slow clearance of fluid in all. Prior to the discovery of dysmotility, all had eosinophilic esophagitis that improved on swallowed budesonide, but budesonide did not improve manometric, radiographic, or endoscopic findings. Manometry on all 4 showed aperistaltic esophagi, but normal LES pressure and complete LES relaxation with swallows. Therapy consisted of elevation of the head of the bed, proton pump inhibitors, and small, frequent meals ending at least 4 hours before bedtime. Despite normal LES manometry, Botox injections were tried on two with minimal effect. Conclusions: Esophageal dysmotility should be considered part of MMIHS, but it may not be clinically, radiographically, or endoscopically evident until the patient is several years of age. Eosinophilic esophagitis is associated, much as it is in achalasia, but successful treatment of it fails to improve the dysmotility. Recommendations: Thorough esophageal evaluation should be conducted as part of the pre-transplant evaluation of MMIHS patients. Patients with esophageal dysmotility should undergo rapid sequence induction when anesthetized. Therapy should not include Botox or other therapies directed toward lowering LES pressure. Patients should be instructed to elevate the head of their bed, to eat small meals, and to refrain from eating for several hours before bedtime.

417 *VOMITING WITH A TWIST: CASE REPORT OF ANTRAL WEB AND MALROTATION*. M. Gribbons, Pediatrics , Children's Medical Center, Dallas, Texas, UNITED STATES|S. Megison, Surgery, UT Southwestern, Dallas, Texas, UNITED STATES|M. Sathe, Gastroenterology , UT Southwestern, Dallas, Texas, UNITED STATES|.

Congenital antral webs are seen in less than 1:100,000 births and are more rare anomaly than duodenal webs. While duodenal atresia, stenosis, and webs are associated with malrotation, there are no current reports of pyloric webs coexisting with malrotation. An 11-month-old Hispanic female initially presented with a one-month history of constipation associated with abdominal distention, frequent belching, and intermittent emesis. Her medical history was only notable for two prior hospitalizations for bronchiolitis. She had no physical or developmental abnormalities and was growing along the 90th percentile. Abdominal radiograph revealed large stool burden and she underwent a successful bowel prep at home. The patient's symptoms persisted for six months, prompting an upper endoscopy, which showed a pinpoint opening to her pylorus. An ultrasound revealed a dilated stomach with complex contents, but no suggestion of pyloric stenosis. An upper GI study was non-diagnostic, but showed persistently dilated stomach concerning for a bezoar, and no evidence of malrotation. Due to high clinical suspicion for eosinophilic gastritis with outlet obstruction, she was treated with balloon dilation and steroids. However, two months after her dilation, the patient's symptoms persisted even after repeat dilation. An exploratory laparotomy was performed with intraoperative upper endoscopy. The patient was found to have both a pyloric web and a duodenum that did not cross the midline. She successfully underwent a Heineke-Mikulicz pyloroplasty and Ladd procedure. This case illustrates a clinical picture of intermittent vomiting, abdominal distension, and constipation resulting from two rare congenital obstructive processes: an antral web and malrotation. While, upper GI is diagnostic in 90% cases of antral webs and sensitive for malrotation in 93%, the patient's condition was only diagnosed intraoperatively. This case serves as a reminder that the existence of one rare congenital condition does not exclude the existence of a second. This is the first reported case to our knowledge of an antral web with malrotation.

418 *ANAL STENOSIS AS A GASTROINTESTINAL MANIFESTATION IN TWO PEDIATRIC PATIENTS WITH NEWLY REPORTED YORK PLATELET SYNDROME*. S. Hommeida, I. Absah, Pediatric Gastroenterology and Hepatology, Mayo School of Graduate Medical Education, Rochester, Minnesota, UNITED STATES|M. Ishitani, Department of Surgery, Division of Pediatric Surgery, Mayo School of Graduate Medical Education, Rochester, Minnesota, UNITED STATES|.

Background: York platelet syndrome is a rare autosomal dominant syndrome. There are 7 reported cases in the literature with 4 cases reported in one family. It is characterized by thrombocytopenia and ultrastructural platelet abnormalities including giant opaque organelles, multilayered target bodies, increased tubular network and deficiency of platelet Ca²⁺ storage in delta granules. No gastrointestinal manifestations have been reported. Case

presentation: Two pediatric patients (15 months and 3 years old) with York platelets syndrome were evaluated for refractory constipation that did not respond to usual medical therapy. Both patients underwent extensive evaluation including TSH, free T4, celiac serology, electrolytes and lead level which were all normal except for thrombocytopenia (which is a feature of this syndrome). Digital rectal examination revealed anal stenosis and spastic anal sphincter that did not relax in response to the digital examination in both patients. Barium enema showed a large amount of colonic fecal material without a transition zone and anorectal manometry showed normal rectosphincteric reflex abdomen. Both patients underwent anal dilation by Hegar dilators up to 15 French resulting in resolution of their constipation. Their father who has York platelet syndrome also reported a similar problem with long standing refractory constipation since childhood. Discussion: The presence of constipation in two generations of this family with York platelet syndrome suggests that constipation could be a gastrointestinal manifestation of York platelet syndrome. In patients with York platelet syndrome and constipation, careful assessment and physical examination should be considered to exclude the presence of anal stenosis.

419 *BRIEF COGNITIVE AND BEHAVIORAL TREATMENT OF RUMINATION SYNDROME IN A TYPICALLY DEVELOPING 8-YEAR-OLD BOY.* P.L. Huston, Helen DeVos Children's Hospital, Grand Rapids, Michigan, UNITED STATES|P.L. Huston, Pediatrics & Human Development, Michigan State University, East Lansing, Michigan, UNITED STATES|.

OBJECTIVE: To describe the successful treatment of a typically developing boy with rumination syndrome using motivational interviewing and behavioral psychology techniques in order to highlight the importance of multidisciplinary treatment. BACKGROUND: Rumination syndrome is one of the least understood functional GI disorders, especially in typically developing children (Chitkara et al., 2006). Due to an extremely low prevalence rate and rumination being a diagnosis of exclusion, children and adolescents often undergo costly and time-consuming invasive testing and treatment prior to proper diagnosis (Chial et al., 2003). In a large study (N=147) of childhood rumination, average time from symptom onset to diagnosis was 27 months (Chial et al., 2003). Furthermore, many providers are unaware that the most effective treatment for childhood rumination is a behavioral approach (Chitkara et al., 2006). CASE: An 8-year-old boy sought treatment due to a 6-month history of rumination diagnosed by his GI physician. Data collected using daily logs throughout treatment indicated between 3-17 episodes of regurgitation per day on intake (see chart). Of note, the patient reported no distress from the rumination and no initial motivation to address the symptoms. Treatment consisted of motivational interviewing (MI) and behavioral (diaphragmatic breathing and progressive muscle relaxation) treatments in 4 sessions across 6 weeks. MI was used to build the patient's motivation to change, while a form of habit-reversal training, consisting of training the patient to engage in competing behavior during the times when he would typically have regurgitation, was the primary treatment method. RESULTS: Regurgitation was reduced significantly upon initiation of diaphragmatic breathing at home following each meal. The patient noted increased motivation to change as a result of discussions about things he was missing out on due to his regurgitation (sleepovers, meals with friends, etc.). In the 2 weeks after discharge, no more than 1 episode of regurgitation was reported per day. At 6 months post-treatment, no regurgitation was reported within the prior two weeks. CONCLUSION: Behavioral therapy techniques are the current gold standard treatment for rumination syndrome. Symptoms can be reduced/eliminated within a short time (4 sessions in this case) once identified. The addition of MI, a technique for increasing patient motivation to change, addressed this patient's lack of desire to eliminate his rumination. Due to early detection and treatment (within 7 months), this patient avoided more invasive diagnostic procedures and has maintained his improvements for 6 months post-treatment. This case can serve as a standard for ideal care for a typically-developing child with rumination syndrome.

430 *A CASE OF NON-FUNCTIONAL DYSSYNERGIC DEFECATION.* R. Medina-Centeno, M. Semrin, R. Sanghavi, University of Texas Southwestern, Dallas, Texas, UNITED STATES|R. Medina-Centeno, M. Semrin, R. Sanghavi, Children's Health, Dallas, Texas, UNITED STATES|.

Background: Dyssynergic defecation, also known as functional outlet disorder or pelvic floor dyssynergia, is defined as paradoxical contraction or failure of pelvic floor muscles to relax during defecation. This causes the anorectal

angle to narrow and increases the pressures of the anal canal so that evacuation is less effective. Most providers treat this with biofeedback and other functional therapies, and do not pursue further testing for constipation. We report a case of dyssynergic constipation in a pediatric patient, who also had an underlying colonic transit issue. Case: 17 y/o female with past medical history of long standing constipation not responsive to aggressive bowel regimen including osmotic and stimulant laxatives. She had multiple episodes of left sided abdominal pain, distension and bloating. She would have one bowel movement a day, 3-4 in the Bristol stool scale, associated with significant straining. She underwent an anorectal manometry which showed normal IAS resting pressure (84 mmHg), IAS contraction with attempted defecation with only 4% relaxation of anus, normal squeeze pressure and duration, present RAIR, consistent with dyssynergic defecation. She was recommended biofeedback and continued bowel regimen. She then presented with persistent pain and bloating; hence underwent imaging which showed persistent gaseous distension of a segment of transverse colon and splenic flexure. Due to this, she underwent a colonic manometry, which showed persistent absent motility in the right colon pre and post stimulation. Given this information, patient underwent left sided partial colectomy with resolution of symptoms. Discussion: Dyssynergic defecation is thought to be a functional disorder that usually responds to aggressive bowel regimen and/or biofeedback if available, with surgical options considered for significant structural abnormalities and/or second line treatment. However, our case highlights the need for continued manometric studies if symptoms fail to improve or worsen. Once again, we also demonstrate the utility of colonic manometry in treating refractory constipation by guiding surgery.

431 *MEDIAN ARCUATE LIGAMENT SYNDROME: THINKING BEYOND FUNCTIONAL ABDOMINAL PAIN.* R. Mittal, C. Fraga-Lovejoy, Pediatric GI, OUHSC, Edmond, Oklahoma, UNITED STATES | A. Agrawal, Internal Medicine, Indiana University, Indianapolis, Indiana, UNITED STATES | S. Prabhu, Pediatric Radiology, OUHSC, Oklahoma City, Oklahoma, UNITED STATES | .

Introduction: Median arcuate ligament syndrome (MALS) is a vascular compression syndrome with symptoms that overlap chronic functional abdominal pain (CFAP). Case Presentation: We present a case of 13 Y F with history of post prandial periumbilical pain for 4 months lasting 15-20 min. Review of system was positive for nausea and about 15 lbs. weight loss. Initial workup including CBC, CMP, amylase, lipase, celiac panel, UGI and EGD with biopsies was unremarkable. No response to omeprazole, hyocyanine, dicyclomine, probiotics and amitriptyline was noted. US abdomen with Doppler showed a marked decrease in velocity within the celiac axis on changing position from supine to upright, and there was increase in velocity to celiac axis upon expiration. CT angiogram revealed focal narrowing at the origin of the celiac axis and an impression of the medial arcuate ligament to this portion of celiac artery. Discussion: MALS is also known as Celiac Artery Compression Syndrome. Characteristic symptoms of post-prandial abdominal pain, nausea, occasional diarrhea and weight loss, overlap with those of CFAP. MALS is felt to be caused by compression of celiac vessels by diaphragmatic crura, leading to decreased flow, a steal phenomenon causing postprandial abdominal pain; also the neurogenic compression may lead to the clinical symptoms. Objective measurement of vessel flow velocity and alterations in vascular architecture by high definition duplex ultrasound scan and CT or MR angiography allows vascular occlusive diseases such as MALS to be more readily diagnosed. The gold standard for diagnosis of MALS is invasive angiography with measurements of pressure gradients. Surgical management entails division of the median arcuate ligament with or without celiac artery reconstruction. Laparoscopic release of the median arcuate ligament overlying the celiac artery can be performed safely in the pediatric population with the release of the artery, resulting in significant improvement in the hemodynamics, symptoms, and overall quality of life. In conclusion, in children with the diagnosis of CFAP, MALS may be a treatable etiology with minimally invasive treatment. A high index of suspicion and increased awareness of the diagnosis of MALS can lead to earlier diagnosis and possible treatment of MALS in patients with diagnosis of CFAP. Continued study of these patients following surgery is needed to better determine long-term effects.

433 *THE USE OF PYRIDOSTIGMINE IN A PEDIATRIC PATIENT WITH PSEUDO OBSTRUCTION.* N. Sainath, K. Fiorino, Gastroenterology, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, UNITED STATES | .

A 10 year old girl with a history of constipation since infancy, eructation, and flatulence transferred care in 2014. Prior workup included a normal spine MRI and a rectal biopsy with sparse ganglion cells. After her initial evaluation, a contrast enema showed marked colonic distension up to 12 cm and considerable redundancy with no transition zone. Two months later, she presented with massive abdominal distention and sigmoid volvulus. She underwent reduction of the volvulus, partial colonic resection, fecal disimpaction, and colostomy formation. Two weeks later, she had a small bowel obstruction secondary to a left paraduodenal hernia. Following repair, she developed significant gastric distension and gastric outlet obstruction. Her obstruction was attributed to congenital bands and a colostomy stricture resulting in a dilated transverse colon that compressed the antrum. She had a revision of the colostomy, lysis of bands, placement of an ND tube, and G-tube placement to improve orientation of the stomach. Postoperatively, upper GI revealed delayed gastric emptying without obstruction. She tolerated advancement of ND feeds, but continued to have significant gastric output of 1-2 L per day. She also had poor colostomy output, requiring daily irrigations. Erythromycin and metoclopramide were trialed without effect. Further operations during her hospital stay included placement of a jejunostomy tube and roux limb creation to prevent jejunal feeds from backing up towards the stomach. Diagnostic workup included a small bowel biopsy with normal enteric plexi, skeletal muscle biopsy without evidence of mitochondrial myopathy, and a normal serum gastrin level. Anorectal manometry showed a normal anal canal resting pressure and relaxation of the internal anal sphincter with balloon distension. Antroduodenal manometry showed an absence of migrating motor complexes in the fasting and post-erythromycin periods, suggesting antral hypomotility and intestinal pseudoobstruction. There was absence of propagated contractions in both the fasting and post bisacodyl states on colonic manometry, consistent with colonic myopathy. She continued to have high volume NG tube output. She was started on colace and senna daily to improve ostomy output, but continued to require frequent ostomy irrigations. Despite the gastric output, she was able to advance to goal J-tube feeds. She was started on pyridostigmine and regular dulcolax irrigations via rectum and colostomy, after which, her gastric output decreased. About a month later, she received botox injections to the pylorus. Following this, her G tube was clamped and her sump was later removed. She was able to tolerate jejunal feeds and a regular diet, in addition to her normal gastric secretions. Pyridostigmine is a cholinesterase inhibitor that can improve gastrointestinal motility. It has been used in patients with pseudoobstruction and autoimmune dysmotility to increase colonic motility. This patient has significant pseudoobstruction with high output gastric drainage and colonic myopathy. The addition of pyridostigmine allowed her pass her gastric contents and tolerate enteral feeds.

435 *MEDIAN ARCUATE LIGAMENT SYNDROME IN A PEDIATRIC POPULATION: A CASE SERIES.* L. Small, M. Rose, S. Toberman, A. Meltzer, R. Zarnegar, A. Solomon, T. Ciecieroga, New York Presbyterian Hospital - Weill Cornell Medical Center, New York, New York, UNITED STATES |.

Introduction: The median arcuate ligament is a fibrous arch which passes superior to the origin of the celiac trunk to connect the diaphragmatic crura on either side of the aortic hiatus. In 10-24% of patients, the ligament passes anterior to the celiac trunk and may cause compression of the celiac artery. The majority of these patients remain asymptomatic, however in some patients compression causes significant hemodynamic changes resulting in clinical symptoms. Median arcuate ligament syndrome (MALS) is characterized clinically by chronic epigastric and/or generalized abdominal pain, often postprandial, weight loss, nausea, vomiting and bloating. MALS is diagnosed using radiographic modalities including vascular duplex abdominal ultrasound, CT angiography (CTA) and/or MR angiography. CTA demonstrates a characteristic focal narrowing in the proximal celiac axis with a hooked appearance of the artery. Vascular ultrasound evaluates the peak systolic velocity (PSV) at the celiac trunk during inspiration and expiration, demonstrating increased PSV during expiration in patients with MALS. This case series reports four pediatric cases of MALS. Patients: Four pediatric patients were diagnosed with MALS between April 2014 and April 2015 at a single tertiary care center. Three were females and one was male. Age at diagnosis ranged from 11y 4m to 18y 2m (mean 15y 1.5m). Time to diagnosis from onset of symptoms ranged from 11m to 6y (mean 2y 7m). All patients presented with abdominal pain. Three patients complained of post-prandial pain with one describing pain after exercise. Three patients had nausea. All patients had normal abdominal ultrasound and upper endoscopies. Other normal studies included colonoscopy in 2/4 pts, MRE in 2/4 pts, gastric emptying

study in 2/4 pts and pH studies in 2/4 pts. Two patients were referred to psychiatry. Past medical history was significant in one patient for asthma and food allergies. Results: All patients underwent vascular duplex abdominal ultrasound which were consistent with the diagnosis of MALS. Ultrasound demonstrated increased celiac artery PSV on expiration. All patients subsequently underwent confirmatory CTA. Measurement of the celiac artery at the area of stenosis ranged from 2.5-5.9mm (mean 4.45mm) with post-stenotic dilation ranging from 5.3-8.4mm (mean 7.03mm). Two patients had a characteristic hook-like appearance of the celiac artery on CTA. Three patients underwent surgical median arcuate ligament release and two of these patients had complete resolution of their symptoms post operatively. Conclusions: The diagnosis of median arcuate ligament syndrome should be considered in children presenting with abdominal pain after eating or exercise without obvious alternate explanation. In patients with symptoms consistent with MALS consideration should be given to obtaining a vascular ultrasound of the abdomen to assess the celiac artery velocities. This may be more cost conscious if suspicion is high enough to avoid more extensive, expensive and potentially unnecessary evaluations including labs, endoscopy and additional imaging studies.

INFLAMMATORY BOWEL DISEASE

440 *AN ELEVATED SERUM IL-10 LEVEL LEADS TO AN IL-10 RECEPTOR DEFICIENCY DIAGNOSIS IN A PATIENT WITH VERY EARLY ONSET IBD.* K. Dallalzadeh, N. Mendez, B. Sahn, Division of Gastroenterology and Nutrition, North Shore LIJ, Cohen Children's Medical Center, New Hyde Park, New York, UNITED STATES|A. Jongco, S. LaBarba, V. Bonagura, Division of Allergy and Immunology, North Shore LIJ, Cohen Children's Medical Center, New Hyde Park, New York, UNITED STATES|.

A 3 year-old male with a history of cortical blindness, partial central diabetes insipidus, and milk protein allergy presented with bloody diarrhea and intermittent fevers for 2 months after a course of antibiotics for oral stomatitis and gingivitis. He is the product of a consanguineous marriage, his parents being first cousins of Turkish descent. Physical examination was significant for aphthous stomatitis and several perirectal fissures and ulcers. Laboratory studies: WBC 17.2 K/ μ L, hemoglobin 10.6 g/dL, and albumin 2.9 g/dL. Stool cultures for bacteria, rotavirus, *C. difficile*, and parasites were all negative. Upper endoscopy showed an erosion in the esophagus, erythematous mucosa in antrum and body of the stomach, and an aphthous ulceration in the duodenum. Colonoscopy was significant for scattered deep ulcerations, exudates, with friable mucosa throughout, and normal terminal ileum. Abdominal MRI was significant for pancolitis without evidence of active small bowel inflammation. Histopathology was negative for viral cytopathic effect or granulomas. Additional testing for tuberculosis, chronic granulomatous disease, and leukocyte adhesion deficiencies were all negative. Flow cytometry showed normal IgA, IgG, IgM, T-cell, B-cell, and NK cell populations. A serum interleukin-10 (IL-10) level was 26.9 pg/mL, a marked elevation above normal range (<2.0 pg/mL). While his immunologic evaluation was ongoing, he clinically deteriorated, developing severe malnutrition with hypoproteinemia, hypophosphatemia, and zinc deficiency with associated acrodermatitis. He was stabilized over many weeks on total parenteral nutrition, antibiotics, and nothing by mouth except water. Due to his elevated serum IL-10 level, an immunopathy in the IL-10 pathway was suspected. A functional assay showed that lipopolysaccharide stimulation of the patient's white blood cells resulted in release of pro-inflammatory TNF α not inhibited by the addition of IL-10, suggestive of IL-10 receptor (IL-10R) deficiency. Further genetic evaluation identified a homozygous point mutation in exon 3 of IL-10RA, the gene encoding the IL-10 receptor alpha subunit, resulting in an amino acid exchange at position 101 from arginine to tryptophan. This was a previously described mutation in the literature, thus confirming the diagnosis in our patient with very early onset inflammatory bowel disease (VEO-IBD). IL-10 typically down-regulates excessive immune responses by inhibiting the secretion of pro-inflammatory cytokines such as TNF α , IL-12, and IFN γ . Serum IL-10 levels have not been reported previously in IL-10R deficient patients and may be potentially used as a screening tool for IL-10R deficiency in the future. Additional genetic testing to better explain our patient's entire phenotype is ongoing. VEO-IBD due to IL-10R deficiency has been noted in the literature to be refractory to immunosuppressive treatments such as corticosteroids, thiopurines, and infliximab, so these were not attempted. The patient is now at home awaiting bone marrow transplantation, which is potentially curative.

441 *INFLIXIMAB-INDUCED INTERSTITIAL PNEUMONITIS IN A PEDIATRIC PATIENT WITH ULCERATIVE COLITIS*. T. Danialifar, H. Naon, Pediatrics, Children's Hospital Los Angeles, Los Angeles, California, UNITED STATES | S. Zhou, Pathology, Children's Hospital Los Angeles, Los Angeles, California, UNITED STATES |.

Background: Development of interstitial pneumonitis has been described in adult rheumatologic and IBD patients on anti-TNF therapy (1-5). Presence of Human Anti-Chimeric Antibody (HACA) has never been reported in this setting. Case Presentation: 12-year-old female was diagnosed with severe ulcerative colitis (UC) with pancolitis. She received induction dose of infliximab 5mg/kg with minimal clinical response and received another dose of infliximab 10mg/kg after 4 days. Prior to initiation of infliximab she had a normal chest radiograph and negative QuantiFERON®-TB Gold. Given insufficient response she was started on methylprednisolone 20 mg intravenously twice daily and transitioned to oral prednisone 20 mg twice daily with good clinical response. She received subsequent 10mg/kg infliximab doses at weeks 6, 10, 14, and 22. Prednisone taper was initiated at week 6 and completed by week 14. The patient remained in clinical remission. Three weeks following the final dose of infliximab she developed shortness of breath with exertion. She was hospitalized 3 weeks later with dyspnea at rest and hypoxia with oxygen saturation of 87% at room air. She had no fever or cough and her UC was in clinical remission with pediatric ulcerative colitis activity index (PUCAI) score 0. Chest radiograph demonstrated severe diffuse interstitial disease bilaterally with patchy opacities at bilateral lung bases. Chest CT (Fig. 1) demonstrated patchy opacities, interstitial disease, and minor early honeycombing peripherally with minimal bronchiectasis. Pulmonary function testing (PFT) was consistent with restrictive lung disease. Infectious causes were ruled-out by laboratory assessment and bronchoalveolar lavage. Rheumatologic evaluation was unremarkable. Human Anti-Chimeric Antibody (HACA) level assessed 6 weeks following last dose of infliximab was positive at 15.1 U/mL with undetectable serum infliximab level. Lung wedge biopsy (Fig. 2) demonstrated a diffuse inflammatory process within the interstitium composed of predominantly lymphocytes admixed with plasma cells, plasmacytoid cells and histiocytes. The airspaces appeared consolidated by increased collection of foamy macrophages. There was septal and pleural fibrosis. Infectious stains were negative. The final diagnosis was interstitial pneumonitis with extensive accumulation of foamy alveolar macrophages. She was treated with 1g Solumedrol daily for 3 days with improvement in symptoms, reduced oxygen requirement, improvement in appearance of chest radiograph, and significant improvement in PFT's. The family refused any further immunosuppression. At time of last follow-up 8 months following hospitalization the patient was asymptomatic, off oxygen, and PFT's had normalized. She had normal chest radiograph. She has maintained clinical remission with mesalamine. Discussion: This is the first reported case of interstitial pneumonitis secondary to infliximab therapy in a pediatric patient and the first report to demonstrate presence of HACA. This may represent a type of infliximab hypersensitivity and is supported by the complete resolution of pulmonary symptoms with short steroid course and discontinuation of infliximab.

452 *EXTRACORPOREAL PHOTOPHERESIS GIVEN AS A SHORT COURSE MONOTHERAPY FAILS TO IMPROVE REFRACTORY CROHN'S DISEASE*. S. Khan, L. Su, B. Pasternak, Phoenix Children's Hospital, Phoenix, Arizona, UNITED STATES |.

Background: Crohn's disease is a chronic inflammatory bowel disorder resulting from an aberrant T cell response to bacteria present in the gastrointestinal tract. Treatment to induce remission includes steroids and/or nutrition, and biologics and immunosuppressive drugs are used for maintenance. Occasionally medical therapy is futile necessitating surgery. Extracorporeal photopheresis (ECP) is another option in refractory cases. The mechanism of action involves development of regulatory T cells, decreased CD8 cells, and a balance restoration of TH1/TH2 cells. ECP has been used in cutaneous T cell lymphoma treatment, GVHD, as well as transplant allograft rejection. It has been used in autoimmune diseases, including Crohn's disease, but limited data is available. Several reports have shown promise with decreased need for steroids and/or clinical improvement. Case Presentation: An 18 year old male diagnosed with Crohn's disease refractory to multiple treatments, was ultimately trialed on ECP. He presented with chronic diarrhea, blood in his stools, and fatigue. He was started on 5ASA and 6-MP; but due to lack of response, he was transitioned to Remicade. Following induction, he had a significant flare requiring hospitalization with bowel rest and Solumedrol. At this time, he was changed to Humira. He continued to have

symptoms of bloody stool, abdominal pain, nausea and fatigue, despite induction. Family sought a second opinion where he developed a sepsis-like picture and possible toxic megacolon. He underwent colectomy with end ileostomy. He was not placed on post-operative prophylactic therapy. After returning, he had a flare requiring hospitalization, so was started on budesonide and transitioned back to 6-MP. Subsequently, the family traveled to another facility and was started on rifampin, ethambutol, and clarithromycin for mycoplasma treatment. Due to lack of efficacy with continued mucosal disease, elevated inflammatory markers, and weight loss, he was re-induced with Humira and enteral nutrition. The patient had another flare and due to failure of various treatments, ECP and TPN was initiated. He received 8 ECP treatments over 4 weeks. He had improved cramping, and less blood per rectum. Follow-up capsule endoscopy showed active disease with 15 cm of active ileitis, and he continued to have microcytic anemia and elevated CRP and ESR. ECP was discontinued and Ustikinimab was started. He continues to have cramping, difficulty with oral intake, and active disease on repeat imaging. He is on TPN/IL currently, and has been referred to surgery to consider terminal ileum resection. Discussion: In this patient with Crohn's disease, multiple treatments for maintenance therapy were ineffective leading to clinically active disease. ECP was trialed as a monotherapy, however was not effective. The course was limited to 4 weeks due to insurance barriers and costs. In other reports, patients received ECP as a combination therapy over a longer course, ranging from 27 weeks to 6 months, which may explain the lack of clinical response in this case. Continued investigation is required to understand the role of ECP in Crohn's disease treatment.

454 *ENTEROCOLITIS AND IMMUNODEFICIENCY AS A FIRST PRESENTATION OF TELOMERE MEDIATED DISEASE.* S. Ogholikhan, H. Hamandi, A. Guerrero, Pediatric Gastroenterology, The Johns Hopkins University, Baltimore, Maryland, UNITED STATES | M. Armanios, H. Lederman, D. Valle, N. Sobriera, C.L. Wagner, The Johns Hopkins University, Baltimore, Maryland, UNITED STATES | .

Clinical Vignette: Caucasian male born at 37 weeks gestational age after a pregnancy complicated by IUGR and oligohydramnios. A postnatal ultrasound showed hydronephrosis and prophylactic antibiotics were started. Weight and height were 3 percentile, but at 1 month, the wt/ht was the 50 percentile. Head circumference was originally normal, but his head grew poorly, and he subsequently became microcephalic with craniofacial disproportion. An MRI was performed at 2 days for poor feeding showing mild prominence of the parieto-occipital subarachnoid spaces bilaterally, but a follow up head CT and MRI showed normal cerebral brain volume and parenchyma. He initially had chronic, loose stools ascribed to the antibiotics. Feeding improved and he was gaining weight appropriately. At 4 months a swallow study showed silent aspiration, therefore gastrostomy tube placement and Nissen fundoplication were performed. He presented at 5 months with worsening, now bloody, diarrhea. Laboratory evaluation consistently showed a normal WBC with normal/elevated absolute neutrophil count and persistently low absolute lymphocytic count. T cell subsets were normal with a normal mitogen response, while B cells were low, but present. His skin, nails, and oral mucosa were unremarkable. An upper endoscopy showed small intestinal mucosa with prominent crypt apoptosis and absent lamina propria plasma cells. Goblet cells and paneth cells were present. A flexible sigmoidoscopy showed cryptitis with crypt dropout and distortion, and reduced, but present, plasma cells. Parenteral nutrition was initiated and treatment included high dose steroids, infliximab and sirolimus to which there was minimal, if any, response. Whole exome sequencing was performed and revealed a DKC1 mutation (p.A308G). Telomeres were measured as abnormally short (delta= -4.5 kb from the median). A third head MRI showed generalized brain volume loss. His feeding intolerance persisted and there were several episodes of culture negative sepsis. He eventually developed respiratory failure and care was electively withdrawn. He expired at 9 months of age. Discussion: Telomeres function to protect chromosome ends from deterioration and defects in telomere maintenance genes cause their shortening leading to apoptosis. Infants with telomere mediated disease present with Hoyeraal-Hreidarsson (HH) syndrome, a syndrome marked by developmental delay, intrauterine restriction and immunodeficiency. In children, telomere-mediated disease typically presents with dyskeratosis congenita. Recent more detailed characterization of telomere mediated phenotypes has revealed enterocolitis may also be a first manifestation in infancy presumably due to high cellular turnover of gastrointestinal tissue. Typically, there is severe crypt dropout, increased apoptosis and absent plasma

cells; and this condition appears refractory to immunosuppression. Telomere length measurement is an inexpensive, rapid screening tool for the infant or child with enterocolitis, immunodeficiency, and lymphopenia.

455 *NEUTROPHIL DYSFUNCTION AND FAILURE TO THRIVE ATTRIBUTED TO MUTATION IN G6PC3*. J.A. Picoraro, K.G. Margolis, S. Kinberg, Division of Pediatric Gastroenterology, Hepatology and Nutrition, Columbia University Medical Center, New York, New York, UNITED STATES|C.W. Glasser, Division of Pediatric Hematology, Oncology and Stem Cell Transplantation, Columbia University Medical Center, New York, New York, UNITED STATES|C. Patel, P. Jain, M. Mansukhani, Department of Pathology and Cell Biology, Columbia University Medical Center, New York, New York, UNITED STATES|A. Iglesias, Division of Pediatric Clinical Genetics, Columbia University Medical Center, New York, New York, UNITED STATES|.

Disruption in neutrophil function is associated with inflammatory bowel disease (IBD). Disorders of the glucose-6-phosphatase (G6Pase) / glucose-6-phosphate transporter (G6PT) complex have classically included glycogen storage disease 1a (GSD 1a; caused by mutations in G6Pase- α) and GSD 1b (caused by mutations in G6PT), both involving disruption of glucose homeostasis with resultant hypoglycemia and hepatomegaly. GSD 1b is additionally associated with neutropenia and IBD. Recently, G6Pase- β (also known as glucose-6-phosphatase catalytic subunit 3 (G6PC3) has been implicated in severe congenital neutropenia and, less commonly, IBD, but without features of classic GSD 1. Two unrelated children with overlapping gastrointestinal manifestations exhibited recurrent neutropenia. Patient 1, a 12 year old boy born to third-cousin parents, presented with failure to thrive with recurrent diarrhea and mouth sores. Colonoscopy revealed normal gross appearance and histology. Neutropenia with absolute neutrophil count (ANC) trend from 300-500 to <100 was not associated with serious infection. Patient 2, a 17 year-old boy born to non-consanguineous parents, presented with ileal stricture associated with arteriovenous malformation and failure to thrive. He exhibited recurrent diarrhea and persistent elevation of serum inflammatory markers. Oral and intestinal ulcerations were characterized histologically by inflammation without granulomas or transmural involvement. Recurrent neutropenia, with ANC trend from 400-700 to >200, was associated with gastrostomy site cellulitis and C difficile colitis. Whole exome sequencing of both probands and both mothers identified a homozygous canonical splice site mutation (c.218+1G>A) in G6PC3 and was confirmed by conventional di-deoxy DNA sequencing. In both cases the variant is inherited from the mother, who is heterozygous and unaffected; the carrier status of the father is undetermined. The variant was not found in 1000 Genomes database or in NHLBI Exome Sequencing Project. The variant is predicted to cause loss of the canonical splice site and changes it to a stop codon, leading to a truncated protein predicted to have altered or loss of function. Patient 1 was treated with GCSF therapy with a robust response in ANC, resolution of gastrointestinal symptoms and appropriate growth. Patient 2 was treated with systemic steroids and infliximab with moderate improvement in weight gain. The response to GCSF therapy is pending. The identification of patients with gastrointestinal manifestations associated with mutations in G6PC3 provides insight into the nature of IBD caused by disturbances in neutrophil function. Loss of G6PC3 function disrupts glucose metabolism leading to endoplasmic reticulum stress. This leads to increased neutrophil apoptosis and may affect neutrophil function through an as yet unidentified process. The association of increased GCSF antibodies with ileitis and the response of patients with IBD to GCSF suggest the role of neutrophil dysfunction in some forms of IBD. Further studies in patients with G6PC3 mutations associated with gastrointestinal manifestations will help delineate this important subtype of IBD.

458 *ENTEROURACHAL FISTULA AS AN INITIAL PRESENTATION IN A TEENAGER WITH CROHN'S DISEASE*. R. Sabe, S. Sankararaman, T.J. Sferra, A. Khalili, Pediatric Gastroenterology, Hepatology, and Nutrition, University Hospitals Rainbow Babies & Children's Hospital, Cleveland, Ohio, UNITED STATES|D. Weinert, Pediatric Radiology, University Hospitals Rainbow Babies & Children's Hospital, Cleveland, Ohio, UNITED STATES|.

Background: Crohn's Disease (CD) in children can present as complicated disease (stricturing/penetrating). In penetrating forms of CD, fistulas may form between bowel loops and other organs. Rarely an infected urachal cyst has been reported as a result of fistulizing Crohn's disease. Our patient presented with recurrent anterior abdominal wall abscesses and dysuria in the context of a patent urachus, and was found to have a fistulous

connection from the terminal ileum (TI) resulting in those recurrent abscesses. Case: 17-year-old male with a history of recurrent anterior abdominal wall abscesses first presented to our institution with abdominal pain and dysuria. He first developed an abdominal wall abscess 5 months prior to this presentation. The abscess resolved with antibiotic therapy, but recurred 2 months later and required surgical drainage. He underwent a colonoscopy at an outside institution during the interval between the two infections. He was diagnosed with terminal ileitis and prescribed a 5-aminosalicylic acid. He was non-compliant with this therapy. At the time of presentation to our institution he reported poor oral intake, a 40 lb weight loss, abdominal pain, and dysuria. He had regular, non-bloody stools. His laboratory evaluation demonstrated a normal hemoglobin, platelet count, albumin, and urinalysis, but was significant for elevated inflammatory markers. Computed tomography (CT) demonstrated an abscess tracking from the umbilicus to the superior ventral urinary bladder wall and an adjacent abscess lateral to the right rectus abdominis muscle that was contiguous with the anterior abdominal wall abscess. A CT scan done at an outside hospital demonstrated an open urachus at the same location. He was placed on antibiotic therapy. He then underwent esophagogastroduodenoscopy and colonoscopy which were significant for severe ileitis without colitis. Magnetic resonance enterography (MRE) confirmed the CT scan findings and demonstrated a fistula from the TI to the urachal cyst abscess. A diagnosis of fistulizing Crohn's disease was made. Therapy for Crohn's disease including corticosteroids and immunomodulator was initiated. The abscess was drained and he completed a full course of intravenous antibiotic therapy. Repeat MRE showed improved inflammation, resolved abscesses, but did show a blind-ended fistula from the TI. After colorectal surgical evaluation, he underwent an ileocectomy with primary anastomosis. Summary: Fistulizing disease can be the initial presentation in children with Crohn's disease. An enterourachal fistula is a rare occurrence in Crohn's disease. This case highlights the importance of recognizing children with signs and/or symptoms of complicated Crohn's disease, especially when patients have an underlying congenital abnormality such as a patent urachus.

460 *ELEVATED LIPASE AT INITIAL PRESENTATION OF IBD: DO WE CHECK FOR IT ?*. P.D. SENGUPTA, M. Van Arsdall, J. Rhoads, Pediatric Gastroenterology, University of Texas Health Science Center at Houston, Houston, Texas, UNITED STATES].

Background: Pediatric IBD represents more extensive inflammation with predilection for colitis. Pancreatitis and hyperlipasemia without pancreatitis are uncommon features of IBD. There have been very limited data on the incidence of elevated lipase in pediatric IBD. Case Report: A 13-year old female presented with hematochezia; loose stools with blood clots for the preceding 4-6 months. She denied abdominal pain, fever, vomiting, weight loss and had minimal urgency/tenesmus. Labs showed elevated calprotectin 225 $\mu\text{g/g}$ and an elevation in pancreatic enzymes; amylase 102 u/l, lipase 1053 u/l (reference range 73-393 u/l). Ultrasound revealed no pancreatic abnormality and normal biliary system. Endoscopy revealed left sided colitis. Pathology confirmed chronic active colitis with cryptitis and crypt abscesses. IBD-7 panel was consistent with UC. She was started on balsalazide with some improvement but ultimately required steroid suppositories for complete resolution of rectal bleeding. Her lipase continued to rise, peaking at 1223 u/l, > 3 times the upper limit of normal. After 3 months of treatment her lipase showed a downward trend, approaching normal values. This correlated with clinical improvement and resolution of bleeding. Discussion: Several potential pathophysiological processes for hyperlipasemia with or without pancreatitis in IBD have been suggested: abnormal passage of pancreatic enzymes from the gut lumen into the blood due to increased permeability of inflamed mucosa, direct involvement of pancreas as part of the spectrum of inflammation, effect of inflammatory mediators and cytokines released from the inflamed gut on the pancreas, pancreatic duct abnormalities. Pancreatic involvement in IBD may also be a part of a common immune disorder whereby the target cells of the gut epithelium and pancreas share a similar molecular structure, lending themselves to antigenic mimicry. Finally, a remote possibility is cryptogenic hyperlipasemia without pancreatitis, wherein macrolipase or immunoglobulin-linked enzymes are suggested to be induced by autoimmune mechanisms resulting in reduced glomerular excretion of the large macrolipase molecules, thereby increasing the serum half-life of lipase. Conclusion: Subclinical or silent pancreatitis may be masked by symptoms attributed to IBD, and hence underreported. If undetected, it may lead to subsequent chronic pancreatitis or exocrine pancreatic insufficiency, which has been reported in 50% of IBD patients with a history of acute pancreatitis in adult

studies. This case shows that checking pancreatic enzymes during initial presentation may be important in determining if pancreatic involvement has resulted from the inflammation in IBD or as an adverse effect of therapy. One should use caution in considering change or cessation of a particular treatment regimen in IBD with pancreatic involvement, as treatment of the underlying inflammation in IBD may be the most important management for resolution of pancreatitis. Longitudinal observation as with our patient and follow-up pediatric studies are needed to see if subclinical pancreatitis or asymptomatic hyperlipasemia results in clinically significant pancreatic disease in IBD patients later in life.

463 PYOGENIC LIVER ABSCESS IN A PATIENT WITH PERIANAL STRICTURING CROHN'S DISEASE - RELATED TO FREQUENT ANAL DILATATIONS. V. Uppal, H. Al-Atrash, F.J. DelRosario, Gastroenterology, Nemours A I duPont Hospital for Children, Wilmington, Delaware, UNITED STATES|.

Background: Liver abscess is a rare extra-intestinal manifestation of Crohn's disease (CD). The incidence of liver abscess in patients with Crohn's disease is 10–15 times higher than that of the general population. There are few cases of liver abscess reported complicating CD. We hereby report a case of pyogenic liver abscess in an 18-year-old female with perianal stricturing CD. Clinical history: An 18 year old female with Crohn's disease of 6 years duration, presented with one-week history of intermittent fever ($T_{max} 103^{\circ}F$) and right-sided lower chest pain that was exacerbated with breathing. Her CD affected her esophagus, ileocecal valve, ileum, and rectum. She failed treatment with Pentasa, Remicade and was now on weekly Humira and methotrexate. She developed anal strictures about one year ago. Initially she required fortnightly surgical dilatations but for the past 5 months she was doing daily self-dilatations. Laboratory findings: WBC 22.9 K/UL; Hemoglobin 11.1 g/dl; Platelet count 475 K/UL, Serum albumin 4.9 g/dl, Total bilirubin 1.7 mg/dl; ALT 44 U/L; AST 17 U/L; GGT 332 U/L; ESR 77 mm/hr; CRP 38.7 mg/L; blood culture – no growth; PT 13.4 sec. Imaging studies: Ultrasound of the right upper quadrant showed a heterogeneous lesion with possible small cystic component in the right lobe of liver. MRI revealed a liver abscess measuring 10.4 x 8.8 x 7.7 cm. Treatment: Ultrasound guided drainage of the abscess was performed. Microbiology culture of the abscess showed growth of *Streptococcus intermedius*. Her initial empirical antibiotic therapy included Zosyn and Vancomycin, which was later changed to Ceftriaxone. Discussion: Liver abscess is a rare but known complication of Crohn's disease. The various predisposing factors for liver abscess formation in CD include: intraabdominal abscesses, fistulous complications, treatment-related immune suppression, malnutrition, use of metronidazole and rarely surgical manipulation, such as colonoscopy. Rarely liver abscess can be initial manifestation of CD. *Streptococcus*, especially *Streptococcus milleri*, is the most frequent cause of liver abscess in patients with Crohn's disease. Frequently, the clinical presentation of liver abscess in these patients can masquerade as an exacerbation and may result in delayed diagnosis. In the current case, it is likely that repeated anal stricture dilatation in the setting of immunosuppression and malnutrition led to seeding of the liver and later abscess formation. Conclusion: The diagnosis of liver abscess should be suspected in a patient with CD with unexplained persistent fever. Liver abscess can be managed with drainage and antibiotics. Liver abscess in a pediatric patient with stricturing perianal Crohn's disease requiring frequent anal dilatations has not been reported.

464 USE OF THALIDOMIDE IN SEVERE PERIANAL CROHN'S DISEASE – RESISTANT TO CONVENTIONAL THERAPY. V. Uppal, A. Maheshwari, F.J. DelRosario, Z. Molle Rios, Gastroenterology, Nemours A I duPont Hospital for Children, Wilmington, Delaware, UNITED STATES|.

Introduction: Refractory Crohn's disease (CD) account for 30% of cases of pediatric CD and these patients are at increased risk of permanent impairment and higher health care costs. Clinical History: An eleven-year-old female, diagnosed with ileocolonic Crohn's disease at the age of 6 years was initially treated with Imuran and Pentasa. A year later, she developed an anal fissure, which progressed to perianal and rectovaginal fistulae. An endoscopy and colonoscopy confirmed progression of the disease with ulcerations in the esophagus, duodenum, terminal ileum and worsening of colonic disease. Her therapy was escalated to Infliximab, which was stopped due to an anaphylactic reaction. She was subsequently treated with Humira. She developed pseudotumor cerebri so Humira

was discontinued. She was then treated with subcutaneous Methotrexate. Due to worsening perianal disease, she underwent examination under anesthesia and a loop Seton was placed. She was also placed on a prolonged course of Ciprofloxacin and Metronidazole. Her symptoms improved transiently, but worsened again necessitating repeat endoscopy and colonoscopy, as well as an examination under anesthesia (EUA). Perianal examination showed a wound measuring 5 x 6 x 7.5 cm, requiring a diverting ileostomy. Post surgery, she required prolonged hospitalization to treat the large perianal wound. She also developed a poorly healing wound at the ileostomy site. Given the appearance of the wounds and their refractory nature to treatment, pyoderma gangrenosum was suspected complicating both the stoma site and the perianal wound. Due to lack of response to Imuran, Pentasa and Methotrexate, and a history of significant side effects with Infliximab and Humira, she was started on Thalidomide. She required TPN to optimize calories and the dose of Thalidomide was optimized to 100 mg/day over the next few weeks. A wound vac was also utilized on the large perianal wound site. Within weeks of intensive wound care and treatment with Thalidomide, her wound started healing. An upper endoscopy and colonoscopy were performed after three months, which showed near complete resolution of esophageal erosions and ulcers and significant improvement in the colonic findings. She has tolerated Thalidomide well for the past 15 months and her perianal wound has significantly improved. Discussion: Thalidomide is recommended as a single drug immunosuppressive therapy and not as combination therapy. Thalidomide has three mechanisms of actions: inhibition of TNF- α synthesis, inhibition of angiogenesis and interference with cell adhesions. Peripheral neuropathy is the main side effect. There are a few studies in pediatric patients supporting efficacy of Thalidomide in severe refractory and fistulizing CD, who failed anti TNF α therapy. Conclusion: We are reporting a case, where a combination of unconventional therapy (Thalidomide monotherapy), surgery (diverting ileostomy) and intensive wound care resulted in significant resolution of severe, penetrating Crohn's disease. Thalidomide has maintained this young girl in almost complete remission.

LIVER

472 *VARICEAL BLEEDING REQUIRING EMERGENT MESOCAVAL SHUNT PLACEMENT*. S. Batra, J. Webster, I. Leibowitz, J.B. Piper, Inova Hospital, Arlington, Virginia, UNITED STATES |.

Introduction: Esophageal and gastric variceal bleeding is a well-known, life threatening complication of portal hypertension. Initial presentation is commonly with signs of significant hemodynamic compromise, at which point urgent stabilization and definitive treatment is often necessary. Determining the best surgical management in an emergent situation is vital for the survival of the patient. We present such a case of variceal bleeding which ultimately required emergent mesocaval shunt placement. Case: 9 year old male with history of MTHFR gene mutation, cavernous transformation of the portal vein with secondary portal hypertension and esophageal varices presented to the emergency department with syncope. Patient was seen in the Emergency Department two weeks prior for melena and received a packed red blood cell transfusion. He had a subsequent endoscopy one week later for variceal banding and sclerotherapy. On the day of presentation, the patient developed several episodes of hematemesis in the Emergency Department and clinical deterioration with bradycardia and significant hypotension. Patient was immediately taken to the Operating Room at which time he had massive amounts of bleeding from his mouth and nose. Ventilation became impossible secondary to his rapidly distending abdomen and an emergent exploration was started prior to intubation. After abdominal decompression, the patient was intubated and underwent gastrostomy with evacuation of clot, esophageal transection with EEA stapling device, oversewing of gastric varices, mesocaval shunt placement and gastrostomy tube placement. Patient received a total of 21 units of packed red blood cells, 3 units of cryoprecipitate, 4 units of platelets, and 8 units of fresh frozen plasma. His abdomen was left open due to elevated peak airway pressures intraoperatively, requiring closure on day 2 of admission. He was awake and communicative 2 days postoperatively. He spent 12 days in the hospital without further complication. Although he had a successful mesocaval shunt, he has gone on to have recurrence of his gastric varices requiring balloon dilation Discussion: This case provides an example of the devastation that can result from gastric and esophageal varices in patients with portal hypertension. The choice of surgical intervention was vital in providing him with such a successful outcome. Mesocaval shunts have been shown to work well in

emergent situations and are ultimately a simpler shunt option. The immediate resuscitation efforts and subsequent emergent shunt placement in this patient were lifesaving.

474 TRANSVENOUS COIL EMBOLIZATION TREATMENT OF CONGENITAL EXTRAHEPATIC PORTOSYSTEMIC SHUNT: REPORT OF TWO CASES IN CHILDREN. Y. Cho, D. Tokuhara, H. Shintaku, Pediatrics, Osaka City University Graduate School of Medicine, Osaka, JAPAN | A. Yamamoto, Radiology, Osaka City University Graduate School of Medicine, Osaka, JAPAN |.

Background: Congenital porto-systemic shunt (CPSS), which is a major cause of neonatal hypergalactosemia without galactose-metabolizing-enzyme deficiency, is known to cause brain manganese deposition, pulmonary hypertension, and hyperammonemia. Among CPSS, extrahepatic congenital portosystemic shunt (ECPSS) rarely regresses spontaneously, thus an effective therapeutic approach is needed for shunt closure. Objectives: We report two cases of transvenous coil embolization (TCE) for ECPSS in children. Methods: Two patients (boys aged 6 and 15 years), one with direct communication of the inferior mesenteric vein with the internal iliac vein and the other with direct communication of the left renal vein with the splenic vein, were treated by transvenous embolization with platinum coils. The clinical outcomes were then evaluated. Results: In the 6-year-old, blood levels of ammonia and total bile acids normalized immediately after TCE: the fasting serum total bile acid level declined from $46.5 \pm 17.5 \mu\text{mol/L}$ (mean \pm SD) to $4.9 \pm 1.8 \mu\text{mol/L}$ and the fasting blood ammonia level declined from $75 \pm 6 \mu\text{g/dL}$ (mean \pm SD) to $32 \pm 10 \mu\text{g/dL}$. Brain MRI showed reduced manganese deposition in the basal ganglia 3 months after TCE. The shunt index, as calculated by per-rectal portal scintigraphy, declined from 52.9% to 24.3%. Recovery was complicated by portal vein thrombosis discovered 1 day after TCE, but it was treated successfully by antithrombotic therapy. In the 15-year-old, blood ammonia and total bile acid levels normalized immediately after TCE: the fasting serum total bile acid level declined from $51.9 \pm 18.3 \mu\text{mol/L}$ (mean \pm SD) to $17.2 \mu\text{mol/L}$ and the fasting blood ammonia level declined from $72.8 \pm 6.4 \mu\text{g/dL}$ (mean \pm SD) to $44 \mu\text{g/dL}$. The shunt index declined from 42.7% to 15.9%. The patient had no complications. Conclusion: TCE is an effective therapy for shunt closure in ECPSS, but care must be taken to watch for post-TCE portal vein thrombosis.

478 CRACKING THE CASE OF IRRITABILITY IN INFANT WITH LIVER DISEASE. W. Elfar, R. Abell, Pediatric GI, University of Rochester, Mendon, New York, UNITED STATES |.

Introduction: Biliary atresia is one of the leading causes of cholestatic liver disease in infants and a cause of hepatic osteodystrophy. Bone disease is seen in chronic cholestasis, largely in part due to incomplete absorption of fat soluble vitamins such as Vitamin D. Due to low Vitamin D levels, there is malabsorption of calcium and magnesium. It has been reported that 80% of jaundiced biliary atresia patients have osteoporosis versus 13.6% in non-jaundiced patients. This severe skeletal demineralization can result in fractures in the absence of trauma in most cases. Case Report: We present the case of a 12 month old female with Biliary Atresia s/p Kasai procedure at 30 days of life who had persistent irritability, especially at night. Her course was complicated by portal venous thrombosis, portal hypertension, ascites, hepatosplenomegaly, hypoalbuminemia, hypocalcemia, clubbing, intermittent acrocyanosis and a period of failure to thrive. Despite receiving 50,000 IU of vitamin D daily, she had severe hypovitaminosis D with a Vitamin D level of 7. Her irritability was initially attributed to her recurrent viral infections, reflux, and pruritis, but did not seem to improve after multiple interventions. She fell down while in the arms of her dad and was seen in the emergency room where she was found to have a buckle fracture of her right femur. Because the fall was not from a significant height and she had metabolic bone disease, the decision was made to obtain a bone scan which showed multiple fractures of different ages in her left radius, ulna and wrist. Her parents denied any other significant falls or trauma that they could recall. After casting and pain medication, she was able to rest comfortably. Discussion: Bone disease is well documented in cholestatic diseases such as Biliary Atresia. Despite supplementation of vitamins and minerals, levels often do not reach normal levels until after liver transplant. Occult fractures without significant trauma have been reported in 91% of cases. Irritability or refusal to use a limb should prompt investigation for possible fracture in these children. Early diagnosis and preventative measures to minimize bone demineralization is imperative as well.

480 *CHRONOLOGIC RELATIONSHIP BETWEEN EBV INDUCED ACALCULOUS CHOLECYSTITIS AND INFECTIOUS MONONUCLEOSIS*. S. Kassabian, M. Wyneski, R. Garcia, Pediatric Gastroenterology, Akron Children's Hospital, Akron, Ohio, UNITED STATES|G. Nagendra, Northeast Ohio Medical University, Rootstown, Ohio, UNITED STATES|.

Background and case presentation: In children, acute acalculous cholecystitis (AAC) can account for 30-50% of cholecystitis cases, and it occurs secondarily to a wide range of extracholecystic conditions. Surveillance is the treatment of choice, and the condition is associated with a more favorable prognosis than in adults. On ultrasound (US), AAC can present as thickening of the gallbladder wall. However, this finding should also prompt the consideration of infectious mononucleosis (IM) from Epstein-Barr Virus (EBV) as the primary etiology. While literature has documented previous cases of EBV induced AAC we present a rare case where the clinical signs of IM present after AAC diagnosis, indicating that EBV workup should still be completed in patients who present with AAC but without clinical signs of IM. Results: Literature review of previous reported cases showed that 9 patients (39%) progressed from clinical IM to AAC over a mean 5.8 days with an average gallbladder wall thickening (GWT) of 8.9 mm at time of their AAC diagnosis. Four (17%) proceeded from AAC to IM over an average 6.3 days with a mean GWT of 11.2 mm at time of AAC diagnosis. The remaining ten (43%) patients had concomitant IM and AAC, and at the time of their AAC diagnosis, their average GWT was 8.6 mm. Conclusions: Either clinical IM or AAC presents within one week after signs of the other develop, with a majority of cases presenting concomitantly or progressing from IM to AAC. Ninety one percent of all cases occurred in females, and 96% recovered with conservative management. We continue to recommend nonsurgical treatment for previously healthy patients who present with EBV AAC, and note that significant elevation of liver enzymes in cases of IM may be a predictor for an impending AAC.

481 *BURKHOLDERIA MULTIVORANS SEPTICEMIA IN A PEDIATRIC LIVER TRANSPLANT RECIPIENT*. S.S. Ho, B. Kamath, Gastroenterology, Hepatology and Nutrition, The Hospital for Sick Children, Toronto, Ontario, CANADA|N. Nashid, V.J. Waters, Division of Infectious Diseases, The Hospital for Sick Children, Toronto, Ontario, CANADA|J.J. LiPuma, Department of Pediatrics and Communicable Disease, University of Michigan Medical School, Ann Arbor, Michigan, UNITED STATES|A. Otley, Division of Gastroenterology & Nutrition, IWK Health Centre, Dalhousie University, Halifax, Nova Scotia, CANADA|J.E. Zlosnik, Department of Pediatrics, University of British Columbia, Vancouver, British Columbia, CANADA|Y.C. Yau, Division of Microbiology, Department of Pediatric Laboratory Medicine, The Hospital for Sick Children, Toronto, Ontario, CANADA|.

Burkholderia multivorans is a species within the *Burkholderia cepacia* complex (Bcc), a group of several species of opportunistic Gram-negative bacteria that cause respiratory tract infection in persons with cystic fibrosis and have been described in outbreaks of infection in intensive care and oncology patients. This is the first reported case of *B. multivorans* septicemia in a pediatric liver transplant recipient. A 23-month old Caucasian male, post-liver transplant was transferred from a provincial hospital to our transplant center for persistent fever of unknown cause. He had biliary atresia and Kasai surgery at 58 days old. His newborn screening and sweat test were normal. He received a living related liver transplant from his father at 6 months of age with standard immunosuppression of steroids and tacrolimus. At 12 months of age, he was diagnosed with intestinal post-transplant lymphoproliferative disorder (PTLD) and treated with rituximab. Otherwise, he remained well apart from recurrent perianal abscesses. The current illness began with 4 days of fever presenting to a local hospital. He was treated with piperacillin/tazobactam and tobramycin before his blood culture (BC) became positive for Bcc. He was treated with ceftazidime and ciprofloxacin to which Bcc was susceptible to for 2 weeks following negative BC. A central venous line (CVL) was inserted due to difficult intravenous access. On the last day of the antibiotic course, he had a single episode of fever that was attributed to a possible viral infection. He was treated with piperacillin/tazobactam and tobramycin again for 48 hours until BC was negative. His CVL line was removed prior to discharge. Within 5 days, his fever recurred and he was readmitted. With the sole symptom of fever, serially negative BCs and recent measurable serum EBV PCR, recurrence of PTLD was considered to be the likely diagnosis. His tacrolimus levels were between 2 and 3ug/L and it was discontinued. Endoscopic assessment and luminal biopsies were normal. Another CVL was inserted. A chest x-ray performed prior to transfer to our institution

showed patchy opacities in both lung fields. At our institution, he continued to be febrile. BCs were repeated. Meropenem and vancomycin were commenced empirically on arrival. Within a day, he developed septic shock. Peripheral and CVL BCs became positive for Bcc. The CVL was removed after the onset of septic shock. Despite adding amphotericin B, ciprofloxacin and sulfamethoxazole/trimethoprim, his condition worsened and he subsequently died from disseminated intravascular coagulation, multi-organ failure and secondary hemophagocytic lymphohistiocytosis. Postmortem examination revealed necrotic abscesses in both lungs and most organs, hemophagocytosis in the liver and a large intracerebral hemorrhage. Lung tissue cultured positive for Bcc. Molecular typing identified the Bcc to be *B. multivorans*, an uncommon multi-locus sequence type, ST355. This case illustrated an unusual opportunistic organism, *B. multivorans*, occurring in a liver transplant recipient. Although the incidence of Bcc septicemia in transplant patients is unknown, early recognition and appropriate treatment is crucial.

484 *CHOLEDOCHAL CYST RUPTURE - CASE REPORT*. S. Kumar, C.P. Vanderpool, Division of Pediatric Gastroenterology, Hepatology and Nutrition, Indiana University School of Medicine, Riley Hospital for Children, Indianapolis, Indiana, UNITED STATES].

Choledochal cyst is a rare congenital anomaly characterized by cystic dilation of the biliary ductal system. This condition may present during early infancy or later into childhood and can result in significant morbidity and mortality. Incidence is geographically dependent, being more common in Asia (1/1000) than in western countries (1/100,000 to 1/150,000). Classic symptomatic triad of jaundice, pain and palpable mass is seen in only 10-25% of cases at initial presentation. Spontaneous perforation of the choledochal cyst with biliary peritonitis has been reported, however this is a very uncommon initial presentation of this rare condition. We report the case of a 7-month-old previously healthy female presenting with five day history of vomiting and irritability. Physical exam was significant for diffuse abdominal tenderness and distension. Laboratory evaluation revealed elevated bilirubin and lipase levels. Initial imaging showed large amount of ascites with uniform dilation of the extrahepatic common bile duct, consistent with a diagnosis choledochal cyst. Hepatobiliary scan revealed extravasation of radiotracer into the peritoneal cavity, confirming the diagnosis of a ruptured choledochal cyst with resulting biliary peritonitis. A laparotomy along with cyst excision, cholecystectomy and Roux-en-Y hepaticojejunostomy was performed. The patient did well after the surgery. Spontaneous perforation is a rare complication of choledochal cyst, but leads to serious complications including biliary peritonitis. Ruptured choledochal cysts can be difficult to diagnose; clinical presentation may be nonspecific and imaging may not clearly delineate cystic dilation of the bile duct if cyst is decompressed following rupture. However, this diagnosis should be considered in children presenting with peritoneal signs along with new onset ascites. A high index of suspicion, appropriate investigation, such as Magnetic Resonance Cholangiopancreatography and hepatobiliary scan can help in reaching an early diagnosis and reducing morbidity and mortality associated with this rare condition.

487 *SPONTANEOUS PERFORATION OF CHOLEDOCHAL CYST WITH BILE PERITONITIS*. W. Elfar, P. Mohanty, Pediatric gastroenterology, University of Rochester Medical Center, Rochester, New York, UNITED STATES].

In infants, one of the rare complications associated with choledochal cyst is spontaneous perforation and subsequent bile peritonitis. The most common site of choledochal cyst rupture is the junction of the cystic and common bile duct. We report a case of a 12 month old previously healthy girl who presented with one day history of inconsolability, non-bloody non bilious emesis, decreased activity and poor oral intake. Her physical examination was remarkable for abdominal distention, right upper quadrant tenderness, guarding and rigidity. Ultrasound showed distended gall bladder with debris and wall thickening. The common bile duct was distended and measured approximately 6 mm. There was intra-abdominal free fluid present. Her labs showed WBC 12.8 THOU/uL, AST 470 U/L, ALT 637 U/L, GGT 277 U/L, INR 1.4, and normal bilirubin. On emergent laparotomy, she was found to have a large choledochal cyst involving the common hepatic duct. A perforation was noted just distal to the confluence of the right and left hepatic ducts with diffuse bile peritonitis. She underwent resection of the extrahepatic biliary tree with hepaticoduodenostomy. Her postoperative course was uneventful. In this report, we

describe a case of bile peritonitis and acalculous cholecystitis after spontaneous perforation of the choledochal cyst and emphasize an unusual site of perforation not previously described in the literature.

488 *ELEVATED LIVER ENZYMES IN THE OVERWEIGHT CHILD - SHOULD WE BE LOOKING FOR ZEBRAS?. I.M. Monteiro, W. Attia, Pediatrics, Rutgers New Jersey Medical School, Newark, New Jersey, UNITED STATES |*

Introduction: Nonalcoholic fatty liver disease (NAFLD) is increasing in overweight and obese children in the US. Serum alanine aminotransferase (ALT) can be used as a screening tool, though the gold standard is a liver biopsy. We present the case of an overweight male with elevated liver enzymes, diagnosed with Niemann-Pick Disease (NPD) Type B on liver biopsy. Case report: 9.5 year old (y) overweight Hispanic male presented with history of elevated liver enzymes noted on routine testing since 8y. Initial labs revealed AST-90, ALT-103, Cholesterol-243, Triglycerides-151. His hepatitis panel and autoimmune markers were negative. Alpha-1-antitrypsin (A1AT), copper and ceruloplasmin were normal. Abdominal ultrasound showed mild splenomegaly. At 10y he had a liver biopsy that revealed lipidosis type storage disease with multiple macrophages filled with delicate material. Stains for A1AT, copper, iron and trichrome were negative. Pathological diagnosis was NPD vs Gauchers disease. Genetic workup revealed NPD Type B. Family screening revealed both parents and both the siblings are carriers of NPD. His liver enzymes have continued to stay high with minimal fluctuations. His cholesterol and triglycerides have increased. He developed some shortness of breath and pulmonary evaluation revealed interstitial lung disease. He had a tonsillectomy for obstructive sleep apnea. No cherry red spot was noted on eye exam. Echocardiogram was normal. Over time he gradually developed mild hepatomegaly. Discussion: NPD is an autosomal recessive disorder due to the deficiency of a lysosomal enzyme, acid sphingomyelinase, leading to accumulation of sphingolipids, especially reticuloendothelial cells. NPD has been described as type A,B,C and D. Types A and B result from deficient activity of acid sphingomyelinase, encoded by a gene located on chromosome 11. NPD type A, the infantile form, is a fatal disorder of early childhood characterized by failure to thrive, hepatosplenomegaly, and a rapidly progressive neurodegenerative course that leads to death by 2-3y. NPD type B, more common in Spanish ancestry, is a milder, non-neuropathic form with later onset and longer survival, into adulthood. Splenomegaly appears first, followed by hepatomegaly as in our patient. Lung involvement leads to breathlessness and respiratory infections. Types C and D, are rarer forms of NPD with impaired intracellular cholesterol trafficking, symptoms appear after 2y, with gradual loss of speech. Organomegaly is not as prominent as in types A and B and death occurs by adolescence. Management includes supportive care and genetic counselling. Enzyme replacement and transplantation may prove promising in the future. Conclusions: Elevated liver enzymes in the overweight or obese child are usually secondary to NAFLD. Without a liver biopsy NPD would have been missed in our patient. However the cost effectiveness and benefits of doing a liver biopsy in all overweight/obese children with elevated liver enzymes still needs to be determined

504 *A RARE CASE OF A LARGE UNRUPTURED HEPATIC ANGIOMYOLIPOMA IN A PATIENT WITH TUBEROUS SCLEROSIS AND POLYCYSTIC KIDNEY DISEASE. V. Uppal, K.N. Furuya, Gastroenterology, Nemours A I duPont Hospital for Children, Wilmington, Delaware, UNITED STATES | G. Gregory, Hematology-Oncology, Nemours A I duPont Hospital For Children, Wilmington, Delaware, UNITED STATES | C.J. LaRosa, Nephrology, Nemours A I duPont Hospital For Children, Wilmington, Delaware, UNITED STATES |*

Background: Angiomyolipoma (AML) is a benign mesenchymal tumor that is frequently found in the kidney and, rarely, in the liver. AML belongs to a group of perivascular epithelioid cell tumors called PEComa (tumor showing perivascular epithelioid cell differentiation). The tumor is composed of blood vessels, smooth muscle, and adipose cells (1). Unlike renal angiomyolipomas, only a few cases of small hepatic angiomyolipomas have been reported in patients with tuberous sclerosis. Clinical History: 11 year old female diagnosed in-utero with tuberous sclerosis, who was also found to have autosomal dominant polycystic kidney disease (presumably due to TSC2/PKD1 contiguous gene syndrome) and seizures. She was incidentally found to have hepatic lesions on renal ultrasound at the age of 9 years. She subsequently had an ultrasound and MRI of the abdomen. The lesions were felt to be hepatic angiomyolipomas based on their typical radiological appearance in that the lesions appeared

heterogeneous and demonstrated early enhancement (Figure 1). Several of the lesions contained multiple increased areas of T1 signal which did not suppress on the in/out phases and represented areas of hemorrhage and thus was most consistent with lipid poor angiomyolipomas. She had multiple lesions in the liver, the largest one of which was 8.5 x 8.1 x 4.7 cm. Of note her alanine aminotransferase, aspartate aminotransferase, gamma glutamyl transpeptidase, albumin, bilirubin, prothrombin time and alpha-fetoprotein levels were normal. Repeat imaging over next two years demonstrated increasing size and number of the hepatic lesions. Therapy with sirolimus was considered in order to control lesion growth, but due to concerns of side effects and drug interactions, the family refused treatment. Discussion: Angiomyolipomas may occur in the liver but are very rare and often occur in the context of tuberous sclerosis. Patients may present with abdominal pain or discomfort, malaise, fever, or anorexia. Many cases are asymptomatic, diagnosed as an incidental finding on imaging studies. Angiomyolipomas are thought to arise from a perivascular epithelioid cell. Studies have reported higher frequency of multiple hepatic angiomyolipomas in females with tuberous sclerosis (2). There are rare case reports of hepatic angiomyolipoma with spontaneous rupture and hemorrhage. In angiomyolipomas occurring in the kidneys, the tumors size ≥ 4 cm is considered to be a predictor of tumor rupture. However, the size at which hepatic lesions are at increased risk for rupture has not been delineated. Some authors have recommended that hepatic lesions greater than 4 cm in diameter should be monitored every 3 months (3). Here, we report a liver lesion of 8.5 cm that has not ruptured.

505 *ACUTE HEPATITIS E INFECTION IN A US TEENAGER WITH CHRONIC HEPATITIS B*. G.B. Winberry, L. Gillis, M.R. Nicholson, Pediatric Gastroenterology, Vanderbilt, Nashville, Tennessee, UNITED STATES|.

Case: The patient is a 16yo male with a history of congenitally acquired chronic hepatitis B, who presented with 5 days of acute onset abdominal pain, vomiting, and jaundice. There was no history of travel in either the patient or his close contacts. Exam was notable for faint jaundice and right upper quadrant tenderness. Labs obtained were notable for an aspartate aminotransferase (AST) of 1308, alanine aminotransferase (ALT) of 1635, total bilirubin of 4.7mg/dL, conjugated bilirubin of 3.5mg/dL, and an international normalized ratio (INR) of 1.5. His baseline liver enzymes were an AST of 42 and an ALT of 67 with normal synthetic liver function, obtained 14 months prior to presentation. He underwent extensive workup for his acute trasaminitis and mild synthetic liver dysfunction, including autoimmune, infectious, toxic, and metabolic studies. Ultimately, his serum hepatitis E IgM antibody resulted positive, while his serum IgG antibody was negative. His stool HEV polymerase chain reaction is pending at the Centers for Disease Control and Prevention. Given his clinical presentation, these antibody findings are highly suggestive of acute hepatitis E infection. His synthetic function normalized, and his liver enzymes returned to baseline over the next 8 weeks with supportive care. Discussion: Acute hepatitis E infection is recognized as a common cause of hepatitis in developing countries. Cases of sporadic hepatitis E are becoming increasingly recognized in developed countries over the past decade. In fact, based on recent population data, it is estimated that the incidence of hepatitis E in the general US population may be as high as 0.7%, which suggests nearly 2 million cases per year in the US. Additionally, recent testing done on CDC samples of patients with acute hepatitis that are seronegative for acute hepatitis A/B, suggest that many of these cases are occurring in non-travelers. However, given this data, there are a startling low number of autochthonous (locally acquired) cases of hepatitis E reported in the US. Part of the under-reporting may be the lack of commercially available hepatitis E testing methods in the US, as well as the fact that zoonotic transmission of genotypes 3 and 4 are often asymptomatic. Regardless, given that clinically significant hepatitis leading to morbidity (especially in immunosuppressed patients or those with chronic liver disease) can occur secondary to acute hepatitis E infection, US clinicians should consider this on the differential for acute hepatitis, even in non-travelers."

MICROBIOLOGY/INFECTIONS/PROBIOTICS

507 *TWO CASES OF HERPES SIMPLEX VIRUS (HSV) ESOPHAGITIS IN IMMUNOCOMPETENT HOSTS- A RARE PRESENTATION*.. A. Akalonu, A. Maheshwari, S.E. Shaffer , E. Kutsch , Gastroenterology, Hepatology and Nutrition, Alfred I. duPont Hospital for Children/Nemours, Wilmington, Delaware, UNITED STATES|A. Akalonu, A.

Maheshwari, S.E. Shaffer , E. Kutsch , Thomas Jefferson Medical School, Philadelphia, Pennsylvania, UNITED STATES|.

Case 1:A previously healthy 12 Y/M, presented with 4 days of chest pain, epigastric pain, vomiting and dysphagia. He had upper respiratory symptoms 1 week prior to presentation. Exam-mild epigastric tenderness. Laboratory results-WBC: 9.2 k/UL, 75% neutrophils, 16% lymphocytes. Inflammatory markers, hemoglobin, liver function tests, lipase and amylase were normal. Upper endoscopy (EGD): severe ulcerative esophagitis in the middle and lower third of the esophagus. Pathology-acute herpetic esophagitis with ulceration. Immunohistochemical stains- HSV-1 positivity with weak positivity for HSV-2. Tissue culture- positive for HSV-1. He was discharged on acid suppression and carafate with good response. We opted not to treat him with an antiviral as he did not have an underlying immunosuppressive condition. Case 2:A previously healthy 11 Y/M, presented with 10 days of progressively worsening dysphagia and odynophagia to both solids and liquids. Exam- moderate dehydration. Laboratory results: ESR: 24mm/hr. Hemoglobin, WBC, liver function tests, lipase and amylase were normal. EGD-multiple irregular deep ulcerations in whole esophagus. Pathology-acute esophagitis with reactive epithelial atypia. Immunohistochemical staining-positive for HSV-1, negative for HSV-2, CMV and EBV. Tissue culture- positive for HSV-1. He was started on IV acyclovir given the severity of his symptoms and discharged on oral valacyclovir and carafate. Post discharge, the patient did well and his immunodeficiency workup was negative. Discussion:Herpes esophagitis (HE) is common in immunosuppressed patients, but has rarely been reported in immunocompetent individuals. We describe 2 new cases of HE in otherwise healthy patients seen in our hospital within the last 3 years. HE has been documented during periods of immunosuppression in patients infected with HIV, underlying malignancy, or in patients who have been treated with radiation, steroids, or chemotherapy. HSV-1 is the most common cause of infectious esophagitis after candidiasis. HE is rare in immunocompetent patients. It may represent the reactivation of a latent infection, but more often is due to a primary infection, probably by local spread of the virus from an orolabial or pharyngeal focus. There is evidence suggesting that herpes virus may infect any traumatized tissue, thus several factors that disrupt the esophageal mucosal integrity, such as gastroesophageal reflux or esophageal instrumentation have been involved in the pathogenesis of HE in the immunocompetent host. Esophageal infection may result from contact of such disrupted mucosa with swallowed saliva that contains the virus. Only a few small case series and several isolated case reports of immunocompetent patients with well documented HE exist in the literature. HE in immunocompetent individuals is usually a self limited disease. It typically resolves spontaneously within 1 to 2 weeks, and only rarely may be complicated by gastrointestinal bleeding or esophageal perforation. Acyclovir is a well established treatment for HE in the immunocompromised host, but its efficacy in immunocompetent hosts is limited and there are no controlled trials evaluating it.

510 *ORIGIN OF CHROMOSOMALLY INTEGRATED HHV6B AFTER LIVER TRANSPLANT IN A CHILD WITH POST-TRANSPLANT LYMPHOPROLIFERATIVE DISORDER.* J. Barry, Pediatrics, Cleveland Clinic Children's, Cleveland, Ohio, UNITED STATES|J. Goldfarb, Pediatric Infectious Disease, Cleveland Clinic Children's, Cleveland, Ohio, UNITED STATES|M. Kay, Pediatric Gastroenterology, Cleveland Clinic Children's, Cleveland, Ohio, UNITED STATES|B. Yen-Lieberman, Molecular Virology, Serology, and Cellular Immunology, Cleveland Clinic, Cleveland, Ohio, UNITED STATES|P. Pellett, Immunology and Microbiology, Wayne State University School of Medicine Detroit, Detroit, Michigan, UNITED STATES|.

Introduction Human herpesvirus-6b (HHV-6b) is a viral pathogen that infects nearly all children by age 2, remaining latent in adulthood. Reactivation of this virus has been documented following cord blood transplantation with the majority of cases being self limited and a minority developing active disease (notably encephalitis). Recent studies show a 16% HHV-6 PCR detection rate among pediatric patients post liver transplant, with the rate of active infection at 2.6%. In 1-2% of cases, HHV-6b can be chromosomally integrated (ciHHV-6b), leading to vertical transmission of the virus, and is suspected when elevated copy numbers of the virus (6 copies/ml whole blood) are persistently detected by PCR. Studies have investigated genomic integration of HHV-6b from donor to recipient in stem cell transplants, but this has yet to be studied in pediatric patients post liver

transplant. Post-transplant lymphoproliferative disease (PTLD) in pediatric liver transplant is a significant source of morbidity and mortality (incidence of 6-20%), thought to be caused by the interplay between immunosuppression and Epstein Barr virus (EBV) infection. The role of ciHHV-6b in PTLD among solid organ transplants is unknown. We propose that ciHHV-6b is analogous to EBV's role in PTLD by its presence in our serum EBV negative patient diagnosed with PTLD following liver transplant. Methods We investigated the case of a 10 year old boy with Caroli disease (serum EBV negative donor/recipient status) who developed EBV chromogenic in situ hybridization (EBER-CISH) positive PTLD with low serum EBV titers (452 copies-Low (500 copies) 5 months post liver transplantation, in the setting of extraordinarily high levels of HHV-6b that emerged after liver transplantation. To determine the origin of chromosomally integrated HHV-6b, parental and organ donor tissue, and whole blood were examined using PCR, along with fluorescence in situ hybridization (FISH) analysis of chromosomes from white blood cells. Results At the time of PTLD diagnosis, serum specimens collected from the patient were positive for HHV-6b DNA, and over a period of 2 months, levels increased from 10^3 copies/ml to 10^6 copies/ml, evidence of the patient lymphocytes harboring the HHV-6b virus. PCR results of donor and recipient liver showed the patient's native liver to have elevated levels of ciHHV-6b compared to negative levels in the donor liver tissue specimens. The PTLD tissue tested showed significantly elevated levels of HHV-6b. Conclusions Tissue from the patient and liver donor allowed us to conclude that native reactivation of ciHHV-6b likely resulted in PTLD in this EBV serum negative patient. In the setting of low (or absent) EBV serum titers, this clinical scenario illustrates the diagnostic complexities associated with ciHHV-6b in liver transplant recipients, and the need for further investigation of this viruses potential role in the development of PTLD.

513 *KLEBSIELLA SEPTICEMIA AFTER FECAL MICROBIOTIA TRANSPLANT IN A PEDIATRIC PATIENT.* A. CHOE, L. Boamah, Pediatrics, Naval Medical Center San Diego, San Diego, California, UNITED STATES | C.R. Salinas, University of California School of Medicine, San Diego, California, UNITED STATES | .

Introduction: Fecal Microbiota Transplant (FMT) of healthy donor stool is a rapidly emerging treatment for restoring the intestinal microbiota of patients with refractory *Clostridium difficile* (*C. diff*) infection. Studies comprised largely of case reports, case series, and a small number of RCTs support FMT efficacy and safety using various routes of administration. Pediatric patients have been shown to tolerate the procedure, with known adverse events limited to immediate post-transplant vomiting and mucoid stools (1). No serious complications in pediatric patients have been reported. Case: A 17-month-old male (BF) with Hirschsprung's disease and recurrent *C. diff* enterocolitis requiring multiple hospital admissions and recent placement of a central line presented with two days of fever, vomiting and copious foul-smelling diarrhea. The patient was admitted with presumed recurrent Hirschsprung's-associated enterocolitis versus central line infection. He was treated with bowel rest, intravenous fluids, and broad spectrum intravenous antibiotics while continuing his home regimen of sulfasalazine and oral vancomycin for a recent bout of *C. diff* enterocolitis. Initial stool studies and blood cultures were negative. Fever and vomiting resolved, but the patient could not tolerate oral intake due to persistent diarrhea. Because of the refractory nature of his disease, BF was determined to be a candidate for FMT. Mother was found to be a suitable donor after thorough screening per American Blood Bank Association guidelines (1). On hospital day 6, FMT was performed via nasogastric tube in accordance with the protocol recommended by the FMT working group (2). Informed consent was obtained from parent. At 17 hours post-FMT, BF acutely decompensated due to septic shock. The patient was treated appropriately and he had full recovery with noted dramatic decrease in stool output. Repeat blood culture from the central line grew *Klebsiella oxytoca*. Discussion: This is the first report of septic shock following FMT in a pediatric patient. FMT has recently been shown to be well-tolerated in pediatric patients given by nasogastric route and has even been used to successfully treat severe diarrhea and sepsis (3). Reported adverse events in children are immediate post-transplant vomiting and mucoid stools (1). Reported adverse events in adult patients are bacteremia, systemic inflammatory response syndrome, peritonitis, microperforation, norovirus infection, and aspiration pneumonia-related death attributed to anesthesia (4,5,6). Our literature review did not identify septic shock in any pediatric patients following FMT. This case of septic shock as a serious adverse event of FMT supports the need for caution and further studies in children.

514 *SUCCESSFUL TREATMENT OF RECURRENT D-LACTIC ACIDOSIS WITH FECAL TRANSPLANTATION.* Z. Davidovics, K. Vance, N. Etienne, J. Hyams, Pediatric Gastroenterology, Hepatology, & Nutrition, Connecticut Children's Medical Center, Hartford, Connecticut, UNITED STATES|Z. Davidovics, J. Hyams, Pediatrics, University of Connecticut School of Medicine, Farmington, Connecticut, UNITED STATES|.

Background: D-lactic acidosis can occur in patients with short bowel syndrome (SBS) when excessive malabsorbed carbohydrate (CHO) enters the colon and is metabolized by colonic bacteria to d-lactate. D-lactate can be absorbed systemically and increased serum levels are associated with central nervous system toxicity manifested by confusion, ataxia, and slurred speech. Current therapy, usually directed toward suppressing intestinal bacteria and limiting ingested carbohydrate, is not always successful. Fecal transplantation, the infusion of donor feces into a recipient's intestinal tract, has been used for decades to treat recurrent *C. difficile* infection, and case reports document its use in the successful treatment of constipation, diarrhea, and abdominal pain. The mechanism of action is unknown, but it is surmised that the alteration of the intestinal microbiome, and reintroduction of potentially beneficial microbes, helps mitigate disease. We present a child with SBS and recurrent, debilitating d-lactic acidosis, which was successfully treated with fecal transplantation. Case Report: A 15 year old male with SBS secondary to malrotation and mid-gut volvulus at 3 years of age, resulting in 25cm of remaining small intestine, intact ileocecal valve, and gastrostomy tube dependence, presented with 4 episodes of severe d-lactic acidosis (3.84-9.22mmol/L; normal range 0 – 0.25) over a span of 3 months. With each episode, he developed progressive fatigue, confusion, slurred speech, and ataxia, requiring hospitalization during which fluid resuscitation and CHO restriction improved his symptoms. These episodes continued despite restriction of dietary CHO using Ketocal and multiple rotating courses of antibiotics including metronidazole, gentamicin, and rifaximin. After receiving IRB approval on a compassionate care basis and informed consent from the patient and family, we identified the patient's father as a suitable donor, and screened the patient and his father according to FDA guidelines for fecal transplantation. Two days prior to the procedure all antibiotics were stopped and the patient was started on a proton pump inhibitor, and the day before the procedure the patient was given a PEG-3350 bowel preparation. On the day of the procedure, 2 tbsp of the father's fresh stool was mixed with 75ml of normal saline, and infused via the patient's gastrostomy tube. Following the procedure the patient had one week of increased abdominal pain and fecal urgency which has since subsided. The patient has gained 3 kg in body weight and is eating an ad lib diet along with 5 cans of Peptamen daily. He has developed a rancid smell to his feces and breath that has prevented him from attending school. At 2 months post fecal transplant he has had no recurrence of d-lactic acidosis while receiving no oral antibiotics. Serum d-lactate is now 0.23 mmol/L. Discussion: This is first case report of fecal transplantation used to successfully treat d-lactic acidosis. The mechanism of action is possibly due to the introduction of an intestinal microbiome that is less likely to produce d-lactate. Further studies, including microbial sequencing analysis, may further elucidate the process.

515 *CAREGIVER EDUCATION REDUCES THE INCIDENCE OF COMMUNITY ACQUIRED CLABSI IN THE PEDIATRIC INTESTINAL FAILURE PATIENT.* N. Channabasappa, Pediatric Gastroenterology, UT Southwestern, Dallas, Texas, UNITED STATES|B. Drews, Pediatric Gastroenterology, Children's Health , Dallas, Texas, UNITED STATES|M. Macaluso, Infection Prevention and Control, Children's Health, Dallas, Texas, UNITED STATES|H.G. Piper, Pediatric Surgery, UT Southwestern, Dallas, Texas, UNITED STATES|.

Pediatric patients with intestinal failure (IF) often require central venous catheters (CVCs) for extended periods of time for parenteral nutrition, blood sampling, and medication administration during intestinal adaptation, increasing morbidity, mortality and costs. Translocation of bacteria and yeast from the gastrointestinal tract has been presumed to account for a large proportion of central line infections in the IF population, however we have observed that many bloodstream infections occur due to lapses in line care practices. We aim to show that patient, family, caregiver and home health nurse education of standardized central line care does significantly reduce community acquired central line associated blood stream infections (CLABSI) in the IF patient and this reduction is sustainable. From 2005 to 2007, we previously reported a community acquired catheter related blood stream infection (CRBSI) rate of 7.0 per 1,000 catheter days in 45 IF patients (Drews 2009). During and after the study

period, the nursing department made a concerted effort to reduce community acquired infection rates with standardized CVC care teaching of the patient/family caregivers, as well as the home health nurse with success in reducing community acquired central line infection rates. Data Collection: The CLABSI data were collected prospectively for our outpatient intestinal failure patients from January 2009 to December 2014. With the implementation of standardized care guidelines and CVC care education, the yearly community acquired rate of CLABSI infection went from 4.8 to 2.9 per 1,000 catheter days in 80 IF patients from January 2009 through December 2014. This is a significant decrease from our reported baseline rate of 7.0 per 1,000 central line days in 2007.

518 *PRESUMED ALLERGIC PROCTOCOLITIS RESOLVES WITH LACTOBACILLUS GG, BOTH AS MONOTHERAPY AND IN CONJUNCTION WITH DIETARY MODIFICATION: A CASE SERIES.* V.J. Martin, Q. Yuan, Pediatric Gastroenterology and Nutrition, Massachusetts General Hospital for Children, Boston, Massachusetts, UNITED STATES|W. Shreffler, Pediatric Allergy and Immunology, Massachusetts General Hospital for Children, Boston, Massachusetts, UNITED STATES|.

Introduction Allergic diseases have been dramatically rising in the United States and other developed nations over recent decades. Recent hypotheses have suggested that Western lifestyle alters exposure to microbes, causing perturbations in the colonization of the intestinal mucosa and affecting mucosal immune system development, thus setting the stage for allergic and autoimmune diseases. Allergic proctocolitis (AP) is among the earliest and most common food allergic diseases of infancy and yet its pathophysiology is not well understood. There is scant evidence that adjunctive therapy with probiotic *Lactobacillus rhamnosus* GG (LGG) may hasten symptom resolution from AP when combined with hydrolyzed formula[1]. There are no reports of probiotic monotherapy in these infants. Case Series Over the past 3 years, in both our primary care pediatrics and pediatric gastroenterology clinic settings, we describe 3 cases of clinically diagnosed AP whose symptoms resolved entirely on LGG monotherapy, and 11 cases initially treated with dietary modification, whose symptom resolution seemed to accelerate with the subsequent addition of LGG. Of these 14 cases, the mean age at diagnosis of AP was 71 days (median 72, range [21-130]). There were 9 females and 5 males. Case presentation Patient A is a term female conceived via IVF and born via C-section for post-dates pregnancy. She had no exposure to antibiotics and was exclusively breastfed (with an unrestricted maternal diet). She presented at 3 months of age to her pediatrician with a 1-week history of persistent grossly bloody stools. There was no evidence of fissures nor of possible infectious causes. A clinical diagnosis of allergic proctocolitis was made. Her family was resistant to dietary modification and she was started empirically on LGG (*Lactobacillus* GG, Culturelle for Kids). Her bloody stools completely resolved over the following 10 days and she was guaiac negative after 2 weeks on LGG. Her mother continued breastfeeding throughout infancy while eating an unrestricted diet. At her well child visits at 4, 6, and 9 months of age, the infant remained on LGG and had guaiac negative stools. She tolerated standard introduction of solid foods, including dairy products. She had no recurrence of any symptoms of AP. The two other cases of successful LGG monotherapy followed similar courses (resolution in less than 30 days). Discussion: To our knowledge these are the first case reports of children with allergic proctocolitis whose symptoms resolved entirely with probiotic monotherapy. Additionally, we add to the literature a group of infants whose symptoms seemed to resolve more quickly with the addition of LGG to their dietary modification. These cases may suggest an important role for the infant intestinal microbiome in the development of gastrointestinal mucosal food allergies such as AP. Prospective investigation of the intestinal microbiome in infants with AP may further our understanding of this disease's pathogenesis. Use of probiotic monotherapy in the treatment of AP also warrants further investigation. [1] M. E. Baldassarre et al, *J Pediatr* 156, 397-401 (2010).

519 *FIDAXOMICIN: A NEW TREATMENT OPTION FOR RECURRENT CLOSTRIDIUM DIFFICILE INFECTION IN CHILDREN.* A.S. Patel, Pediatric GI, UT Southwestern Medical Center, Dallas, Texas, UNITED STATES|P. Luu, A.S. Patel, Children's Health, Dallas, Texas, UNITED STATES|.

Clostridium difficile (CD) is a growing problem in the pediatric population. CD, a common-cause of antibiotic-associated diarrhea caused by a gram positive, toxin producing anaerobic bacillus has become increasingly prevalent in the pediatric community. Of concern is the increase in incidence among pediatric patients in both the community and hospital setting, a population previously believed to be at low risk. The incidence in the pediatric population has doubled in the past decade. Much of the changing epidemiology parallels the emergence of hypervirulent strains of CD. The treatment options for *Clostridium difficile* infection (CDI) must change to meet the emerging trend of more resistant strains of CD and higher recurrence rate as well as morbidity and mortality especially in children. Despite the increases in the incidence, severity and financial burden of care related to CDI in the past decade, treatment options lag behind epidemiological trends. While patients generally respond well to standard treatment with metronidazole and vancomycin, the rate of recurrence (27% and 24% respectively) and treatment failures are high (14% and 22% respectively). The rate of recurrence in children, up to 25%, is similar to adults. Fidaxomicin has been FDA approved for the treatment of recurrent CDI in adults, but pediatric indication has not been evaluated. Fidaxomicin inhibits the RNA synthesis by RNA polymerase. Fidaxomicin is highly effective against CD (MIC 0.25 mcg/mL) including the hypervirulent strains. Clinical cure rates were similar between vancomycin and fidaxomicin (92.1% vs 89.8% respectively); however, fidaxomicin had lower recurrence rates (13.3% vs 24%), lower spore counts after treatment and better ability to preserve the normal gut microbiome during and after treatment. We report our clinical experience treating CDI in children using fidaxomicin, a macrocyclic antibiotic approved for the treatment of CDI in adults. A retrospective chart review was conducted from 1/2013 to 1/2015 at CMC Dallas/UTSW which identified 5 patients treated with fidaxomicin 200 mg twice a day for 10 days following treatment failure with FDA approved therapy. All five patients tested positive by PCR for CD toxin and presented with diarrhea, abdominal pain, dehydration, nausea, vomiting and fever. In our subjects, the risk factors for CDI include antibiotic exposure, acid suppression, and immunosuppressant therapy. One patient developed community-acquired CDI while the remaining four had hospital-acquired CDI. Multiple drugs have been studied for the treatment of CDI in adults, but therapeutic options for the pediatric indication are limited. Yet over the past decade, the incidence of recurrent CDI has doubled in the pediatric population. Similar to findings in adults, we found that fidaxomicin had a more sustained and durable resolution of disease in the 5 pediatric cases we studied. Treatment with fidaxomicin led to eventual clearance of CDI with durable response and was well tolerated without complications. Fidaxomicin may have a role in the treatment of recurrent CDI in children and its potential application must be explored given the changing epidemiology of CDI.

520 SAFETY AND EFFICACY OF FECAL MICROBIOME TRANSPLANTATION BY NASOJEJUNAL AND/OR ENDOSCOPIC ROUTE IN PEDIATRIC PATIENTS WITH RECURRENT CLOSTRIDIUM DIFFICILE INFECTION. P.A. Patel, D. Keljo, A. Goyal, Pediatric Gastroenterology, Children's Hospital of Pittsburgh of UPMC, Pittsburgh, Pennsylvania, UNITED STATES |.

BACKGROUND & AIMS: *Clostridium difficile* infection (CDI) is difficult to eradicate despite antibiotics, and recurrence increases likelihood of persistence. Fecal microbiome transplantation (FMT) cures CDI in up to 91% of adults. Pediatric literature on FMT for CDI is sparse. We aimed to report the first comparative analysis of the tolerability, safety, and efficacy of nasojejunal (NJ) versus endoscopic FMT in pediatric patients with recurrent CDI. **METHODS:** This study was an Institutional Review Board-approved retrospective chart review of children receiving FMT per institutional protocol for recurrent CDI at a tertiary care children's hospital. Twenty-eight patients (mean age 12.3+ 6.8) who received 31 FMTs for recurrent CDI were included of whom 75% had inflammatory bowel disease (IBD) flare and 25% had primary CDI. Of the 31 FMTs, 17 were by NJ and 14 by endoscopic route. Mean follow-up was 11.6+ 5.6 months (range 3-22). **RESULTS:** Overall, 90% of the FMTs had resolution of CDI. Resolution of recurrent CDI was seen in 94% (16/17) of NJ FMTs and 86% (12/14) of endoscopic FMTs (P = 0.58). Only three patients with IBD and recurrent CDI failed to respond to FMT (1 NJ, 2 endoscopy). Of the patients with IBD flare, 67% had symptomatic resolution of flare, 4% had a partial response, and 24% remained active despite negative CDI. **Adverse events:** During NJ infusion, 3/17 patients experienced transient headache, dizziness, abdominal pain, and vomiting. Early gastrointestinal symptoms (<48 hours) occurred in one patient with NJ and 2 with endoscopic FMT. Two of these patients needed brief hospitalization for fever, abdominal pain, vomiting, and diarrhea. Intermediate symptoms (>48 hours to <7 days) of self-limited diarrhea were seen in 1/17 patient with NJ FMT.

Transient mild abdominal pain +/- diarrhea and vomiting was also observed between 7 and 30 days post FMT in 3 patients (2/17 NJ, 1/14 endoscopy). Most of these adverse events including NJ tolerability and early, intermediate, and late gastrointestinal symptoms were self-limited. Unexpected adverse events seen in two patients included cholangitis and abdominal abscess (likely pre-dated FMT secondary to undiagnosed stricture). There was no significant difference in adverse events seen between both FMT routes ($P = 0.46$). CONCLUSIONS: Similar safety and efficacy were seen in NJ and endoscopic routes. The efficacy was comparable to adult data. Most adverse events were mild and transient. FMT can be considered in the management of pediatric IBD patients presenting with flare and concomitant CDI.

522 *RECURRENT SEVERE CLOSTRIDIUM DIFFICILE INFECTION IN A 27 MONTH- OLD MALE SUCCESSFULLY TREATED WITH FECAL MICROBIOTA TRANSPLANTATION: A CASE REPORT.* U. Rani, A. Imdad, M.R. Nicholson, Department of Pediatrics, Division of Gastroenterology, Nutrition and Hepatology, Vanderbilt University, Nashville, Tennessee, UNITED STATES|.

Clostridium difficile (*C.difficile*) is a gram-positive, spore forming anaerobic rod that can lead to severe gastrointestinal infections. However, severe illness in young children and infants is rare, and infants demonstrate a high rate of asymptomatic carriage. We present a case of a 27 month-old boy who presented with diarrhea, vomiting and dehydration and tested positive for a toxigenic strain of *Clostridium difficile* confirmed by DNA amplification testing. Sigmoidoscopy showed pseudomembranous colitis. His hospital course was complicated by hypoalbuminemia, ascites and pleural effusion. He developed toxic megacolon and rectal prolapse. The patient was treated with antibiotics and supportive care. He was discharged after complete recovery. However, 1 week post-discharge, he presented with similar clinical complaints and was found to again have *C.difficile* colitis. In addition to standard therapy for recurrent *C. difficile* infection, fecal microbiota transplantation was performed to prevent further recurrence of disease. On follow up, patient was asymptomatic, with no signs or symptoms of recurrent disease. This case report is novel due to the presence of severe *Clostridium difficile* infection in a young child, a group more commonly colonized and without severe disease, and the successful use of fecal microbiota transplantation for cure.

523 *FUNGAL ESOPHAGITIS AND GASTRITIS IN A FOUR WEEK OLD MALE WITH PYLORIC STENOSIS.* M. Stark, Pathology, LSU, Children's Hospital of New Orleans, New Orleans, Louisiana, UNITED STATES|N. Santucci, B. Keith, Pediatric Gastroenterology, LSU, Children's Hospital of New Orleans, New Orleans, Louisiana, UNITED STATES|.

We report of a case of fungal gastritis and fungal esophagitis in a four week old Hispanic male with a three day history of hematemesis, melena and extreme fussiness with feeds. The infant remained hemodynamically stable. Upper endoscopy showed easily washable white exudates extending throughout the esophagus, stomach and duodenal bulb with spontaneous friability, suggestive of severe esophagitis and gastritis. Biopsies were consistent with neutrophilic abscess in the esophagus and active neutrophilic and plasma cell infiltrate in the antrum and body of stomach. Focal PAS positive fungal forms most consistent with *Candida* were found in both esophagus and stomach. No immune deficiency has been identified to date despite an extensive infectious and immunologic workup. The infant was later (8 weeks of age) found to have pyloric stenosis when vomiting failed to resolve after treatment with antifungals and required a pyloromyotomy. Fungal antritis in infants has not been previously reported in literature. This unusual presentation of fungal antritis highlights the importance of infectious disease monitoring and the need for vigilant endoscopy in the pediatric population. The causal relationship of fungal antritis and pyloric stenosis is unclear.

524 *NOVEL THERAPY FOR RECURRENT C. DIFFICILE INFECTION.* H. Schwenk, Pediatric Infectious Diseases, Stanford University School of Medicine, Stanford, California, UNITED STATES|K. Harmann, Pediatric Liver and Intestinal Transplant, Lucile Packard Children's Hospital Stanford, Palo Alto, California, UNITED STATES|T.L. Piester, J. Fuentebella, K.T. Park, Pediatric Gastroenterology, Stanford University School of Medicine, Stanford, California, UNITED STATES|.

Background: Clostridium difficile is the most commonly recognized cause of infectious diarrhea in the health care setting and the incidence of C. difficile infection (CDI) is increasing. Although metronidazole and vancomycin remain the first-line treatments for CDI, up to 25% of patients will experience a relapse of their disease. Alternative therapies for recurrent CDI (rCDI), including fecal microbiota transplantation, have emerged as potentially efficacious strategies, but may be contraindicated in certain immunocompromised populations. We report a case series of response to plant phenols (LiveLeaf™) in a cohort of patients with rCDI. Methods: The Divisions of Pediatric Infectious Diseases and Gastroenterology have established a clinical protocol to assess patients with rCDI. Based on clinical judgment, a trial of LiveLeaf™, a commercially available blend of plant phenols, is offered to patients with rCDI. To date, 3 patients have participated, all of whom had a history of treatment failure with conventional antimicrobial agents. The patients were given LiveLeaf™ for at least 21 days and closely monitored for clinical improvement. Patients also underwent repeat CDI testing by real-time qualitative PCR. Results: Patient 1 is a 19 month old otherwise healthy female with CDI despite a 10 day course of metronidazole, 14 day course of vancomycin, and 12 week vancomycin taper. The patient was given LiveLeaf™ for 21 days with resolution of symptoms and no adverse events. A stool PCR 2 weeks after treatment was negative for C. difficile. Patient 2 is a 6 year old female with a history of PFIC type 1 requiring liver transplant. Her post-transplant course was complicated by relapsing, disseminated salmonella enterocolitis and rCDI. Following several weeks of antibiotics, including two courses of vancomycin, she was started on LiveLeaf™ for 3 months with complete resolution of symptoms and documented negative C. difficile PCR. She tolerated the product without side effects and has had no recurrences of her CDI for the last 6 months. Patient 3 is a 3yr old boy with a history of acute liver failure of unclear etiology requiring liver transplant. He was diagnosed with CDI approximately 6 months post-transplant and initially responded to a 10 day course of metronidazole. His course was complicated by two CDI recurrences, for which he received oral vancomycin. He developed rCDI shortly after stopping his medications and received a 6 week vancomycin taper. He was started on LiveLeaf™ midway through this course and completed 5 weeks of daily therapy. His diarrhea has resolved and he has not had any CDI recurrences for two months. Conclusions: We have demonstrated a clear pattern of symptomatic improvement and microbiologic clearance using plant phenol therapy for patients with rCDI. Given the substantial morbidity and cost related to rCDI, there has been renewed interest in therapeutic strategies for patients whose disease is poorly-controlled with currently available antimicrobials. Our data suggests that LiveLeaf™ may be highly effective in the management and prevention of rCDI and merits further investigation in a randomized controlled trial.

525 *FAILURE OF EMPIRIC BROAD-SPECTRUM ANTIBIOTICS TO PREVENT SMALL BOWEL BACTERIAL OVERGROWTH AND D-LACTIC ACIDOSIS IN PEDIATRIC INTESTINAL FAILURE: A CASE REPORT.* A. Tsou, C. Duggan, Gastroenterology, Hepatology and Nutrition, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES | D.A. Stamm, C. Duggan, Center for Advanced Intestinal Rehabilitation, Boston Children's Hospital, Boston, Massachusetts, UNITED STATES |.

Background: D-lactic acidosis related to small bowel bacterial overgrowth (SBBO) is a well known complication of intestinal failure. Reduced absorptive capacity and impaired motility of the intestine can lead to an abundance of carbohydrate in the lumen, as well as migration of colonic bacteria into the small intestine. This may foster overgrowth of D-lactate-producing bacteria, fermentation of unabsorbed carbohydrate to D-lactate, and development of metabolic anion gap acidosis. Standard medical therapy for SBBO includes rotating courses of broad-spectrum antibiotics. Methods: Retrospective chart review was performed on a pediatric intestinal failure patient with severe D-lactic acidosis. Results: A 2 year old girl with intestinal failure due to multiple intestinal atresias with foreshortened small bowel and a full colon presented with severe, symptomatic D-lactic acidosis despite antibiotic therapy. She had a prolonged course of parenteral nutrition (PN) with transition to full enteral feeds at 1 year of age. Contrast radiography showed diffusely dilated small bowel. She had a history of intermittent acidosis and elevated D-lactate levels prompting use of bicarbonate supplements and empiric broad-spectrum enteral antibiotics for treatment of presumed SBBO. At age 16 months she was receiving 3.8 mEq/kg/day of sodium bicarbonate, and cyclic ciprofloxacin and metronidazole for 3 weeks per month. She nonetheless developed an asymptomatic anion gap acidosis (CO₂=13, anion gap=21, D-lactate=2.1 (0.0-0.25 mmol/L)).

Bicarbonate supplements were increased to 5.8 mEq/kg/day, and she was referred for upper GI endoscopy, which revealed copious fluid throughout the duodenum and normal histopathology. Duodenal fluid culture exclusively grew *Lactobacillus* sp., sensitive to clindamycin, erythromycin, gentamicin and penicillin and resistant to vancomycin. Shortly after endoscopy, she presented with intermittent lethargy and clumsiness which was most noticeable in the morning after completion of overnight feeds. Her physical exam was notable for a respiratory rate of 40, lethargy, slurred speech and ataxia, and her labs showed an anion gap acidemia (pH=7.27, CO₂=10, anion gap=26). She was given intravenous Lactated Ringer's solution, and bowel rest for 1 night. Based on duodenal aspirate sensitivities, her antibiotic regimen was changed to continuous, alternating weekly courses of clindamycin and gentamicin. She presented to clinic 2 weeks later with resolution of anion gap acidosis, and complete resolution of signs and symptoms of D-lactic acidosis. Conclusions: Endoscopy with duodenal aspirate collection and culture facilitated appropriate treatment of a patient with severe D-lactic acidosis related to *Lactobacillus* sp. which developed despite aggressive empiric antibiotic therapy. D-lactic acidosis should be high on the list of differential diagnoses in intestinal failure patients with altered mental status and anion gap acidosis, and threshold for performing endoscopy with duodenal aspirate and quantitative cultures should be low.

526 *ESOPHAGEAL ASPERGILLOMA IN A THREE YEAR OLD WITH HLH AND MASSIVE GI BLEEDING*. J. Ramakrishna, K. Tran, G. Zella, Pediatric GI & Nutrition, Floating Hosp for Children/Tufts Med Ctr, Boston, Massachusetts, UNITED STATES].

Introduction: *Aspergillus* is a frequent cause of opportunistic infection in immunocompromised patients with high mortality. The gastrointestinal tract is a common location for dissemination and symptoms can include abdominal pain and gastrointestinal bleeding. In particular, esophageal *Aspergillus* is associated with poor outcomes. Case: A previously healthy 3-year-old female presented with fever of unknown origin and elevated LFTs that quickly progressed to sepsis and multi-system organ failure. She was diagnosed with Hemophagocytic Lymphohistiocytosis (HLH) based on pancytopenia, markedly elevated ferritin, liver dysfunction, and bone marrow biopsy. She was treated with an individualized HLH chemotherapy protocol and broad spectrum antibiotics including fluconazole prophylaxis. Repeat bone marrow done 3 weeks later showed aplastic marrow. Soon after, she had fevers and invasive *Aspergillus* was noted in right upper lobe on chest CT, while MRI of head and spine showed extensive sinus fungal disease. She was taken to the OR for sinus debridement and started on voriconazole. However, fevers persisted with deteriorating clinical condition. She then had a significant hemoglobin drop with multiple large bloody stools as well as bloody output from the nasogastric tube. EGD showed multiple 2-5 cm circular areas of denuded deep ulcers in various stages of bleeding throughout the stomach. In the mid-esophagus, a light green mass 3 cm in length was noted to project from a denuded circular ulcer-like lesion in the esophageal wall. This mass was connected to the ulcer via a thin stalk that was unable to be clearly visualized. Using a biopsy forceps, this mass was separated from its base and removed through the mouth. Histologic evaluation confirmed this mass as an aspergilloma with evidence of full-thickness esophageal necrotic tissue. She was started on octreotide IV drip, pantoprazole IV drip, sulcrafate, and continued on voriconazole. GI bleeding stabilized and patient underwent repeat EGD three weeks later which showed near complete resolution of previously seen gastric ulcers. There were no esophageal masses or lesions. Clinical improvement continued with no further significant GI bleeding and she was discharged with outpatient management of HLH. Discussion: Though *Aspergillus* can disseminate to the gastrointestinal system, reports of esophageal invasion are rare. Moreover, case reports of esophageal aspergilloma have been associated with fatal outcomes. In our patient, we theorize that the aspergilloma may have been continually seeding sites of gastric infection and endoscopically removing this fungal mass eliminated the key nidus of persistent infection. This may explain why the patient showed remarkable clinical improvement and resolution of gastric lesions on follow-up endoscopy while on the same anti-fungal therapy.

529 *CYSTIC ECHINOCOCCUS INFECTION IN A 10 YEAR OLD IRAQI REFUGEE*. J.H. Weitzner, J.L. Bilhartz, A.J. Freeman, Pediatric Gastroenterology, Hepatology, and Nutrition, Emory University, Atlanta, Georgia, UNITED STATES|J.F. Magliocca, Department of Surgery, Division of Transplantation, Emory University, Atlanta, Georgia, UNITED STATES].

Background Cystic echinococcus (CE) is an infection which primarily targets liver and lung and is caused by the larval form of the canine tapeworm *Echinococcus granulosus*. The disease is slow to progress and extremely rare in the United States with nearly all reported cases are involving immigrants who have arriving from endemic areas. Case Description A 10 year old boy presented to the ED at a tertiary medical center with 3 months of non-specific abdominal pain. Five years prior, he and his family fled a refugee camp in Mosul, Iraq where he had lived the first four years of his life. He has lived and traveled exclusively in the United States since. Upon presentation, an abdominal ultrasound was obtained which revealed multiple fluid filled sacs. MRI abdomen revealed 5 hydatid cysts in his liver, 3 involving his right lobe and 2, the left. The largest, superior to the dome, measured 11 cm and contained multiple daughter cysts. MRI of the chest and brain was normal. Echinococcus IgG was positive. The remainder of his investigation was unremarkable. He was started on albendazole 300mg BID for 4 weeks prior to surgery. In surgery, the cysts were identified. Two involving his left lobe were identified and resected in a non-anatomic fashion. Due to the size and involvement of the remaining cysts, the patient underwent right lobectomy with preservation of the middle hepatic vein. The patient tolerated the procedure well and preserved adequate liver function demonstrated by serial INR post operatively. He will continue albendazole for 3 months with aminotransferases and CBC monitored routinely. Repeat MRI to evaluate for eradication will be obtained after 3 months. Conclusion: Although endemic to certain regions of the globe, CE is a very uncommon diagnosis in the United States. Treatment is dictated by appropriate staging of the cysts and includes medical therapy with benzimidazoles, and interventional therapy with PAIR (Puncture-Aspirate-Injection-Reaspiration) and/or resection. Patients should be followed for at least five years post intervention. Although the liver is the most commonly affected organ, lung, skeletal, and CNS involvement is well described and must be considered prior to initiation of therapy. Complications of disease include compression of adjacent structures, embolism, and severe anaphylactic reaction to cyst contents in the event of rupture. The regions with the highest prevalence of Echinococcus include Asia Minor, the Middle East, and South Asia. As conflict in these areas continues, immigrants and refugees from these areas unknowingly infected with Echinococcus will seek treatment in American medical centers. Treatment requires a multidisciplinary approach with possible consultation/referral to centers with expertise in caring for this disease.

NUTRITION

533 *GIANT SPLENIC CYST: AN UNUSUAL CAUSE OF RECURRENT EMESIS IN AN ADOLESCENT*. N. Eltawil, L. Mahajan, R. Steffen, Pediatric Medicine, Cleveland Clinic Children's Hospital, Cleveland, Ohio, UNITED STATES | N. Bhesania, Department of Pediatric Gastroenterology and Nutrition, Cleveland Clinic Foundation, Cleveland, Ohio, UNITED STATES |.

A 12 year old male presented with recurrent emesis and mild epigastric abdominal pain of two months duration. He had missed almost 30 days of school due to the emesis. Despite symptoms, his appetite remained good and he had not lost weight. There was no recent history of trauma; however, the family reported he had fallen down a flight of stairs, landing on his abdomen two years prior. Pain had resolved after several days and no medical attention was sought. Physical exam revealed an obese male, BMI = 32 kg/m². Vital signs were normal. Abdominal exam revealed no tenderness. Obesity precluded exam for organomegaly or masses. Remainder of exam was normal. Labs were normal including CBC, serum aminotransferases, amylase and lipase. An upper GI series showed displacement of his stomach and upper small intestines to the right. Abdominal ultrasound showed a splenic cyst measuring 17.7 cm x 18.5 cm x 20.7 cm, Contrast enhanced abdominal CT scan showed the cyst to be well-defined, originating from the spleen, with a thin rim of calcification anteriorly in the capsule (Figure A). Patient was then given pre-operative pneumococcal prophylaxis. Laparoscopic single port partial splenectomy was performed following intra-operative aspiration of 2.2 liters of chocolate colored fluid from the cyst. Massive adhesions were identified between the cyst wall, the peritoneum and the diaphragm. The cyst wall was thick and leathery, requiring blunt dissection for removal. Normal salvaged spleen measured 3 cm x 7 cm x 4 cm. No malignant cells were identified in the fluid. Pathology showed a benign fibrous walled cyst, negative for neoplasm. Abdominal trauma accounts for 75% of splenic cysts causing hematoma formation with resultant resorption and serous fluid

collection. This etiology is supported in our patient by the remote history of trauma, massive adhesions extending outside the spleen, and the fibrous lining of the resected cyst. Surgical management is required for symptomatic splenic cysts or if the size reaches a diameter of >5cm. Our patient's cyst is the largest splenic cyst reported in a pediatric patient without a history of acute trauma. Due to the thick nature of the rind of our patient's splenic cyst, we speculate it had formed years earlier and only recently became symptomatic due to possible unrecognized re-injury. Treatment is intended to spare splenic tissue with limited resection as much as possible in order to avoid the risks of total splenectomy (overwhelming sepsis, thrombosis). Prophylactic anti-pneumococcal vaccination is recommended before surgery. With the increasing prevalence of obesity worldwide, the clinician needs to maintain a high index of suspicion for abdominal masses, as physical examination is often inadequate. Our patient's obesity likely precluded an earlier detection of this abdominal mass during routine physician visits.

534 *NUTCRACKER SYNDROME: WHAT THE PEDIATRIC GASTROENTEROLOGIST NEEDS TO KNOW*. N. Bhesania, L. Mahajan, Pediatric Medicine, Cleveland Clinic Children's Hospital, Cleveland, Ohio, UNITED STATES].

A 16 year old female presented with a six year history of progressively worsening abdominal pain, diarrhea, left sided costo-vertebral angle tenderness, and malnutrition. At age 10, she developed recurrent painless hematuria and was diagnosed with IgA nephropathy. Subsequently, she developed abdominal pain, diarrhea with intermittent hematochezia and progressive weight loss. Her symptoms acutely worsened over the last 1 year, prompting further evaluation at numerous tertiary care centers. She underwent extensive work up including laboratory evaluation, stool studies, EGD, colonoscopy, cystoscopy, gynecologic exam, multiple CT scans and X-rays which were unremarkable. Nutritional rehabilitation with psychological counseling was advised. On presentation to our facility, she reported daily post-prandial abdominal pain and diarrhea. Pain radiated to the left perineal region, was severe and debilitating. No emesis reported. On exam, she was malnourished with a BMI of 16.09 kg/m² (1st%). Abdominal exam was normal. Upper GI demonstrated possible narrowing of the third portion of the duodenum, prompting further evaluation. EGD and colonoscopy were normal. Magnetic resonance enterography revealed mild compression of the third part of the duodenum and left renal vein by the overlying superior mesenteric artery consistent with Nutcracker Syndrome. Urinalysis showed large hemoglobin. Referral to Vascular Surgery was made for left renal vein stenting. As the patient did not have emesis, concomitant superior mesenteric artery syndrome (SMA) was present. Nutcracker syndrome (NCS) refers to compression of the left renal vein (LRV), between the aorta and SMA. The syndrome is characterized by hematuria (secondary to rupture of the thin-walled septum between the small veins and collecting system of the renal fornix), proteinuria, flank pain, emotional disturbances, pelvic congestion in females and varicocele development in male patients. Diagnosis is based on history and physical examination, basic lab tests to exclude other causes of hematuria, cystoscopy and ureteroscopy to confirm unilateral hematuria. Diagnostic imaging evaluation includes Doppler ultrasound, CT or MR angiography. Review of the literature suggests that this condition is relatively more common in females, with most cases presenting in the 3rd or 4th decade of life. Few patients have presented in adolescence, making this an unfamiliar diagnosis to many pediatricians and subspecialists. The Pediatric Gastroenterologist must always consider this syndrome in any patient presenting with chronic abdominal pain, diarrhea or malnutrition in association with persistent hematuria of unexplained etiology.

557 *KWASHIORKOR IN UPPER MIDDLE CLASS SUBURBIA*. D. Francis, M. Tobin, A. Chawla, Pediatric Gastroenterology, Stony Brook Long Island Children's Hospital, Stony Brook, New York, UNITED STATES].

Background: Malnutrition, kwashiorkor, is a major cause of death worldwide among children below 5 years. Previously a disease of the developing country is now in patients in the developed world due to faddism, or presumed food allergy. We describe a patient with kwashiorkor fed rice milk. Case: An 11-month old Caucasian male with a 3-day history of progressively worsening generalized edema, decreased intake, and rash. He had not gained weight for 4 months. He drank Rice Dream and some pureed foods. Patient was exclusively breast fed for 6 months then took formula. He tried multiple formulas which caused gassiness and rash. He was seen by two allergists and diagnosed with dairy, soy, and sesame allergies. He was placed on rice milk. On physical exam patient

was irritable with a diffuse maculopapular rash, scattered vesicles and desquamation on the bilateral lower extremities and buttocks. A lacy erythematous papular rash was on the trunk. Patient had pitting edema of the extremities, scrotum, and periorbital region. He had hypoalbuminemia(1.4 g/dL), anemia(7.8g/dL), zinc (37) and copper deficient(43), and elevated liver enzymes(AST 144, ALT 448). Abdominal ultrasound demonstrated ascites with small bilateral pleural effusions. While hospitalized,he received Albumin and nasogastric feeds with Elecare, zinc supplementation and a multivitamin. His edema and rash improved. He was discharged home on hospital day 6. Eight weeks later showed an average weight gain of 11 grams daily. Discussion: Kwashiorkor is characterized by severe malnutrition, hypoalbuminemia, edema, irritability, and a rash. It is commonly reported in Africa and other developing countries and quite rare in the United States. Due to faddism, presumed food allergy or ignorance this condition is resurging, reported in infants receiving rice milk as their major source of nutrition. Rice milk is an alternative treatment for milk protein allergy(MPA). However, not all products have the same nutritional value. A recent study, Vandenplas et al. (2014) examined 39patients with MPA fed an extensively hydrolyzed rice protein based formula called Novarice (21.9 calories and 0.6grams of protein per ounce), compared to typical infants formulas (20calories and 0.5g of protein per ounce). Results revealed normal growth in these patients. Novarice is available in Europe. Patients on rice based milk in our country drink Rice Dream milk (15 calories and 0.13g of protein per ounce). Its nutritional composition does not resemble that of other formulas. Infants who ingest this milk are at high risk of developing kwashiorkor with other nutritional deficiencies including zinc and copper like our patient. It is clear from this case and others that consumers (parents and patients) and health care providers need educations on commercially available products as they are not nutritionally alike. Translating an evidence based study on the benefits of fortified rice milk into clinical practice needs to be performed cautiously since not all rice milk products are nutritionally alike.