PEDIATRICS

S1473 Presidential Poster Award

Metatranscriptomic Analysis of Pediatric Esophageal Epithelium and Microbiome Response to Proton-Pump Inhibitors

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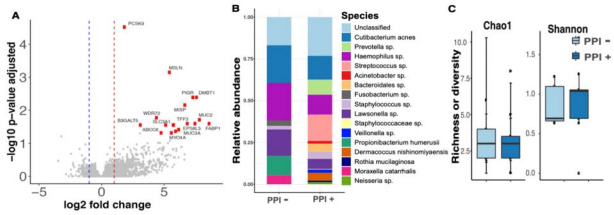
Introduction: Proton-pump inhibitors (PPIs) are empirically used to treat children with esophageal symptoms even when there are no histologic abnormalities (normal esophagus). Little is known about the impact of PPIs on the molecular profile and microbiome of a normal pediatric esophagus. Our aim was to investigate the effects of PPIs on esophageal epithelial transcriptional and active microbiota responses in children with a normal esophagus. We hypothesized that the PPIs alter the epithelial transcriptional profile and active esophageal microbiome simultaneously in children with normal esophagus.
Methods: Distal esophageal biopsies obtained from 20 children (6-18 years) with esophageal symptoms and without any histologic abnormalities were included in the analysis. Seven children were not on a PPI (PPI−) and 13 were on a PPI (PPI+). They were not on any concurrent medications or antibiotics. Metatranscriptomic methods were used to capture host transcriptional and microbial profiles from the biopsies. Transcripts mapped to the human genome were used for gene expression analysis. Microbial transcripts were used to profile active microbial abundance, alpha diversity and beta diversity.
Results: The median (IQR) age of the cohort was 14 (12-15) years. Compared to PPI−, the PPI+ children showed upregulation of 19 genes and no genes were downregulated (Figure 1A). The prominent mucin genes (MUC2 and MUC3), DMBT1 (deleted in malignant brain tumor 1) - a regulator of mucosal homeostasis possibly through the linking of mucosal defense and regeneration, FABP1 (Fatty Acid-Binding Protein 1) involved in the binding, transport and metabolism of long-chain fatty acids, and Trefoil factor 3 (TFF3) involved in the maintenance and repair of the intestinal mucosa were upregulated. Microbial transcripts revealed Haemophilus sp. Lawsonella sp. and Propionibacterium sp. were significantly abundant in PPI−, whereas Prevotella sp. and Streptococcus sp. were highly abundant in PPI+ (Figure 1B). There was no differences in the alpha and beta diversity between the 2 groups (Figure 1C).
Conclusion: Metatranscriptomic analysis of normal pediatric esophageal samples revealed that PPIs increase the expression of genes involved in esophageal epithelial cell function and mucosal homeostasis, and also leads to alterations in the esophageal microbiome simultaneously. Studies are underway to confirm our results in a larger group of children.



[1473] **Figure 1.** (A) Volcano plot showing differentially expressed host genes between children on PPI (PPI+) and children not on PPI (PPI−). We used a threshold of $\log_2$ fold change >1 and adjusted $P < 0.05$ to call the genes that are up- or downregulated. The upregulated genes that satisfy the threshold are shown in red dots. (B) A color-coded bar plot shows the relative abundance of esophagus microbiome in PPI− and PPI+ groups. (C) Richness and alpha diversity of the esophagus microbiome. Alpha diversity and richness (measured by Shannon and S.chao1 index) are compared between the PPI− and PPI+ groups.

S1474

Real World Ustekinumab Use in Pediatric Patients With Crohn's Disease

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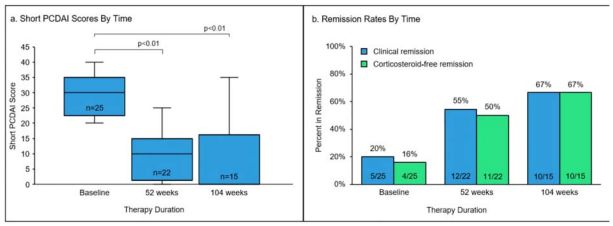
Introduction: Ustekinumab (UST), a humanized IgG1κ monoclonal antibody antagonist of interleukin-12 and interleukin-23, is currently approved for patients (pts) 18+ years with moderate-to-severe Crohn's disease (CD). Promising clinical results have been reported in the literature with UST in pediatric CD pts. The purpose of this study was to evaluate the real-world experience of UST in pediatric CD pts treated at large gastrointestinal private practices.
Methods: This was a retrospective observational analysis of CD pts aged ≤18 years who initiated UST therapy. Pts were followed for a minimum of 1 year, and additional analyses were conducted for those with 104 weeks of follow-up data. Demographics, disease characteristics, previous therapy, UST utilization, and adverse events data were collected from electronic medical records. The short pediatric Crohn's disease activity index (sPCDAI) was used to assess disease activity at UST initiation, 6 weeks, 24 weeks, 52 weeks, and 104 weeks. Clinical remission was based on sPDCAI scores < 15. Corticosteroid (CS) use was evaluated at each time point.
Results: There were 25 eligible pts (48% female) who initiated UST therapy during the study period. The mean (SD) age and disease duration was 16.5 (2.3) years and 3.3 (2.8) years, respectively. The median [IQR] weight was 62.1 [45-69] kg, and the majority (52%) used a UST intravenous induction dose of 390 mg. All pts subsequently received UST 90 mg subcutaneously every 4-8 weeks. Most (88%) were biologic-experienced having failed one (n = 8), 2 (=13), or 3 (n = 1) biologic therapies previously. Three pts were biologic-naïve. Of the 25 pts, 22 (88%) had follow-up data available at 52 weeks and 15 pts (60%) at 104 weeks. Significant reductions in sPCDAI scores compared to baseline is show in the Figure 1. At 52 weeks, 12 out of 22 pts (55%) had reached clinical remission, and 11 of 22 pts (50%) were in CS-free remission. By week 104, the 10 out of 15 pts (67%) who reached clinical remission were also CS-free. Within the first 52 weeks, 3 pts discontinued UST due to tolerability (n = 1 at induction) or the lack or loss of response (n = 2).
Conclusion: Pediatric pts experienced significant improvement in disease activity scores at both 52 and 104 weeks. Our long-term UST data shows good rates of clinical and CS-free remission in pediatric CD pts. Further studies in this population are warranted.



[1474] **Figure 1.** Disease Activity Scores and Remission Rates in Pediatric Crohn's Disease Patients on Ustekinumab.

S1475

Endoscopic Ultrasound-Guided Liver Biopsy (EUS-LB) in Pediatric Non-Alcoholic Fatty Liver Disease (NAFLD): A Single Center Study

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Introduction: Endoscopic ultrasound – guided liver biopsy (EUS-LB) is a well-established technique for liver biopsy in adult patients due to direct visualization and real-time guidance, feasibility in obese patients, patient tolerance and comfort, and decreased recovery time. The applicability in pediatric patients is still underreported.
Methods: This is a single center retrospective review to assess the safety and utility of EUS-LB in the evaluation of pediatric NAFLD at Community Regional Medical Center in California's Central Valley. An analysis of the biopsy sample including quality, features, and interpretability was performed. To fully critique the utility of EUS-LB in assessing liver steatosis and fibrosis, we compared other forms of hepatic assessment including laboratory values and imaging.
Results: A total of 15 pediatric patients (7 female) ages ranging from 7 – 18 years old (median age 16 years) were included in the review. Twelve patients underwent only transgastric fine needle biopsy (FNB), with an average of 1.92 (range 1-3) passes. Two patients only underwent transduodenal FNB passes with an average of 1.33 (range 1-2) passes. One patient underwent both transgastric and transduodenal FNB passes. The shortest specimen length was 0.7 cm and the longest was 2 cm in length with a median of 1.3 cm. All biopsies performed were technically successful. All biopsy specimen was adequate for histopathologic evaluation and diagnostic yield was 100%. By diagnostic criteria for NAFLD, 86.67% (46.15% female, 92% Hispanic) were diagnosed with NAFLD and of those, 61.54% of patients found to have NASH. No major or minor post-procedural complications were identified or reported.
Conclusion: Noninvasive investigation such as labs and imaging may be useful for screening and identifying patients with likely diagnosis of NAFLD, however noninvasive methods have limitations. Accurate staging, histologic review, and prognostication for NAFLD are best characterized by liver biopsy. EUS-LB is a efficacious and safe procedure that should be considered to be the primary method when pursuing liver biopsy in pediatric patients. We recognize limitations of the study, including small sample size and largely homogenous patient population.

S1476

Efficacy and Clinical Outcomes of Endoscopic Variceal Ligation With N-butyl-2-cyanoacrylate in Pediatric Patients in a Tertiary Care Center—A Retrospective Study

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Introduction: Acute variceal bleeding (AVB) from gastroesophageal varices is a life-threatening complication in patients with portal hypertension. Bleeding from gastric varices is less common and treatment options are not standardized. Endoscopic variceal obliteration (EVO) with N-butyl-2-cyanoacrylate glue or balloon-occluded retrograde transvenous obliteration (BRTO) are the 2 most common treatment modalities and information on these are limited to adult series. There is paucity of information about gastric varices and its treatment in children.
Methods: We conducted a retrospective cohort study among pediatric patients (1-18 years) who presented to our center with AVB from gastric varices. Between 2011-2021, we identified 538 among 4539 cases (12%) of portal hypertension with bleeding from gastric varices. Of these only 14 cases (2.6%) had a diagnosis of gastric variceal bleeding in children. Details of 13 patients were extracted with non-availability of records of 1 patient. Cyanoacrylate glue in 0.5-1 ml were injected in to the gastric varices. Patients were evaluated for efficacy, safety and complications associated with the treatment. Endoscopy was repeated after 3 weeks for assessment of variceal size and need for repeat glue injection.
Results: The mean age of the patients was 13.3±5.2, with males (54%) and females (46%). The mean age at first presentation to hospital with gastrointestinal bleeding was 8.7±3.6, with hematemesis (62%) being the most common presentation. On endoscopy, 8 patients had type 1 gastro-esophageal varices and 3 patients had type 2 gastroesophageal varices. All 13 patients had stigmata of recent gastric variceal bleeding. The most common cause of portal hypertension was extra hepatic portal venous obstruction (EHPVO) (54%). The mean age at glue therapy treatment was 11.3±5.3. While immediate hemostasis was achieved in all 13 patients (100%) with gastric variceal bleeding, re-bleeding was seen among 3 patients, which occurred after 1 month of glue therapy. 1-3 sessions were required for obliteration of gastric varices. There was one case of death among the 13 children: a patient with decompensated Wilson's disease. We did not encounter complications like anaphylactic shock, treatment-associated infection, gastric perforation, and distant emboli.
Conclusion: The experience from our series suggests that endoscopic vascular obliteration using cyanoacrylate glue injection is safe and effective in children with gastric variceal bleeding.

S1477

Effect of a Detailed Educational Intervention on Pre-Endoscopy Anxiety Levels in Children and Young Adults

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Introduction: Invasive medical procedures such as endoscopy are a significant source of anxiety for patients. Published data show that higher levels of pre-endoscopy anxiety are associated with worse clinical outcomes. Studies in adults have shown that pre-endoscopy interventions such as educational videos or brochures are associated with decreased pre-endoscopy anxiety. Similar literature in pediatrics is limited. We sought to evaluate whether an educational video effectively reduces pre-endoscopy anxiety in a pediatric cohort of patients and to identify the most common concerns surrounding endoscopy.
Methods: This is a prospective randomized controlled trial of patients ages 8-21 years requiring gastrointestinal endoscopy with their parents participating when appropriate. The intervention group was shown an educational video created by the study investigators. Subjects were asked to report anxiety using a Numerical Rating Scale (NRS) and the State-Trait Anxiety Inventory (STAI, STAI-C). They also reported their anxiety levels surrounding specific components of the procedure.
Results: 160 subjects have been recruited and the study remains open. 82 (51%) and 78 (49%) subjects were randomized to the intervention and control group, respectively. Most subjects identified as Hispanic (26%) or African American (15%) and 28% had Medicaid. 35% of subjects had a history of prior endoscopy. Baseline anxiety trait was not significantly different between control and intervention groups. NRS and STAI/STAI-C scores were comparable in intervention and control groups amongst all-comers. However, for the cohort of patients recruited prior to the day of endoscopy, NRS scores were lower in the intervention group (mean score of 3.4 vs 5.6, $P = 0.01$). Subjects receiving the intervention prior to the day of endoscopy had lower STAI-C state scores than those receiving the intervention the day of endoscopy. Regarding concerns surrounding endoscopy, patients and parents were most worried that some disease might be found, followed by the need for anesthesia.
Conclusion: Our results suggest that an educational intervention may attenuate pre-endoscopy anxiety, but that this may be contingent on the timing of delivery. Our results also highlight the factors surrounding endoscopy that are most anxiety-provoking for patients and parents, providing valuable information which may help further define topics that should be addressed not only by educational interventions but also in physician-patient discussions regarding endoscopy.

S1478

An Assessment of Pediatric ERCP Long-Term Quality of Life Outcomes

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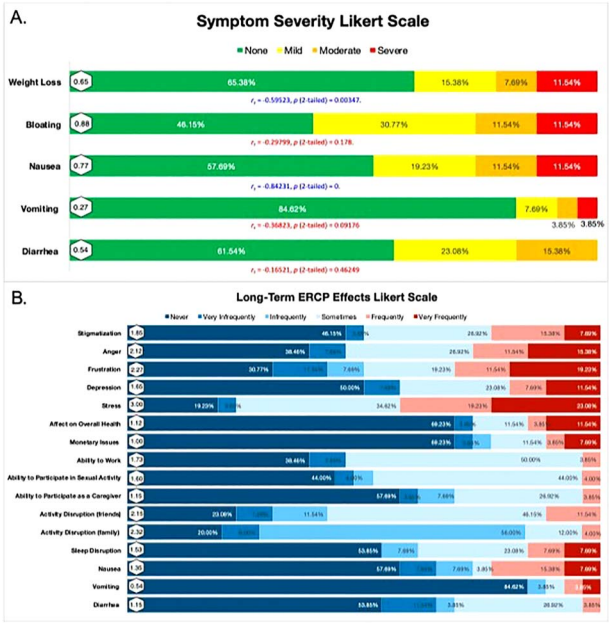
Introduction: Due to its proven efficacy, endoscopic retrograde cholangiopancreatography (ERCP) is an increasingly used modality in pediatric populations, with utility extending to recurrent pancreatitis (the most common indication in children), chronic pancreatitis, and gallstone disease. However, little is known about long-term ERCP outcomes in pediatric patients. The goal of this project was to assess long-term quality-of-life outcomes in pediatric ERCP using a questionnaire previously validated for patients with chronic pancreatitis.
Methods: Adult patients who had an ERCP performed as a child at one of our 3 academic centers were contacted to assess their willingness to participate in the survey. A telephone script was utilized to reduce bias in the posing of questions. The questionnaire incorporated parts of pancreatitis quality of life instrument (PANQOLI), which has primarily been validated for chronic pancreatitis, as well as unique questions designed to evaluate distinctive problems posed by pancreatic and bile duct manipulation.
Results: 26 patients completed the questionnaire, with 46.15% reporting complete satisfaction with their health. Though only 23.08% recounted that their presenting symptoms were cured by ERCP (**Table 1**), the majority of patients (69.23%) perceived no effect of the prior ERCP on their overall health as adults. 34.61% of patients needed at least one additional ERCP, and a diagnosis of chronic pancreatitis was described by 42.31% of surveyed patients. Spearman correlation testing identified that increasing severity of nausea ($r_s = -0.84231$) and weight loss ($r_s = -0.59523$) correlated with decreases in patient health satisfaction (**Figure 1A**). 23.08% of patients continued to require pancreatic enzyme replacement into adulthood, and 4 of the 26 patients had prescriptions for opiate pain medication related to chronic abdominal pain. Very frequent stress and very frequent frustration were reported by 23.08% and 19.23% of patients, respectively (**Figure 1B**).
Conclusion: This study offers a glimpse into the long-term physical, social, and emotional states of patients who underwent ERCP in childhood. Though we found that chronic symptoms related to ERCP contribute to decreased health satisfaction, further study is needed to assess risk factors for long-term complication in pediatric ERCP. A limitation of this study is the questionnaire's reliance on patient recall, considering that many of these procedures were performed during childhood many years prior.

Table 1. Survey Results

	Not at all	Slightly	Unsure	Mostly	Completely	
Overall Health Satisfaction	15.38%	11.54%	11.54%	15.38%	46.15%	
Cured by ERCP	11.54%	30.77%	19.23%	15.38%	23.08%	
	Unsure	No	Yes	Yes, multiple		
Additional ERCP	7.69%	57.69%	7.69%	26.92%		
Subsequent Surgery	0.00%	65.38%	23.08%	11.54%		
Acute Pancreatitis	0.00%	50.00%	15.38%	34.62%		
Chronic Pancreatitis	0.00%	57.69%	42.31%	0.00%		
	None	Mild	Moderate	Severe		
Diarrhea	61.54%	23.08%	15.38%	0.00%		
Vomiting	84.62%	7.69%	3.85%	3.85%		
Nausea	57.69%	19.23%	11.54%	11.54%		
Bloating	46.15%	30.77%	11.54%	11.54%		
Weight Loss	65.38%	15.38%	7.69%	11.54%		
	Never	Very Infrequently	Infrequently	Sometimes	Frequently	Very frequently
Diarrhea	53.85%	11.54%	3.85%	26.92%	3.85%	0.00%
Vomiting	84.62%	0.00%	3.85%	3.85%	3.85%	3.85%
Nausea	57.69%	7.69%	7.69%	3.85%	15.38%	7.69%

Of the 26 patients that completed our survey, only 23.08% reported that ERCP completely cured their presenting condition. 42.31% of patients reported chronic pancreatitis as a diagnosis after their ERCP, with 26.92% reporting an additional ERCP needed to address their presenting condition. The most common persistent symptoms were nausea, bloating, and weight loss.



[1478] **Figure 1.** Likert Scale showing response rates for reported symptom severity (A) and long-term effects from prior ERCP (B). Symptom severity scale (A) includes Spearman correlation coefficients which identified a correlation between increasing severity of nausea and weight loss and decreasing overall health satisfaction. A sizable number of patients reported stigmatization, anger, frustration, depression, or stress attributable to their prior disease course. There were also post-procedure limitations in the ability to work, participate in sexual activity, participate as a caregiver, and spend time with friends or family.

S1479

HIDA Way to Go? The Predictability of the HIDA Scan for Resolution of Symptoms Post Cholecystectomy in Pediatric Patients

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Introduction: A HIDA (hepatobiliary iminodiacetic acid) scan is the most relied upon study for determining the benefit of a cholecystectomy in adult patients. A HIDA ejection fraction (EF) of 35% or lower in adults is considered abnormal; there is no standardization in pediatrics at this time. The purpose of this study was to examine whether specific patient profiles are associated with clinical response to cholecystectomy among a pediatric population in rural east Tennessee.
Methods: This retrospective cohort included 100 pediatric patients without cholelithiasis who had a cholecystectomy between 2014-2017 at a rural children's teaching hospital. Hospital and outpatient clinic records were reviewed for preoperative symptoms, laboratory and radiologic investigations, and postoperative outcomes. Analysis included frequencies of symptoms, cross tabulations between HIDA scan EF, obesity, presence of postoperative symptoms (including abdominal pain, nausea, or vomiting), and work up prior to the procedure, and chi-square tests of association.
Results: Of the 100 patients, 78 had a documented HIDA scan reported; 55 of which had an abnormal EF (<35%) and 23 of which had no resolution of symptoms postoperatively, ($P < 0.05$). Of the documented 41 obese patients, 23 had an abnormal EF and 9 had persistent symptoms ($P < 0.058$). Of the 55 with an abnormal EF, 30 patients had a documented trial of proton pump inhibitors (PPIs) prior to the HIDA scan, of which 50% had no resolution of symptoms post cholecystectomy ($P < 0.066$). 18 patients had an EGD and a trial of PPIs, of which 10 had persistent symptoms ($P < 0.001$). 10 patients had documented abdominal Xray (KUB) consistent with constipation, with no statistical significance related to resolution of symptoms or EF.
Conclusion: This study was limited by lack of documentation in the electronic health record. Despite having an abnormal EF, 42% of patients had persistent symptoms post cholecystectomy. Therefore, the HIDA scan is not diagnostic and should not be the sole diagnostic criteria regarding biliary dyskinesia in children. Our results indicate that regardless of HIDA EF, BMI, KUB, EGD findings, or a trial of PPI, one cannot predict the resolution of symptoms postoperatively. While the HIDA scan is standardized in the adult population, further research is needed to assess gallbladder motility in children. Hence, before recommending a cholecystectomy, clinicians should investigate other potential differentials for abdominal pain.

PEDIATRICS

S3346

To Bead or Not to Bead: Use of Ultrasound and a Conservative Approach to Water Bead Ingestion in an Infant

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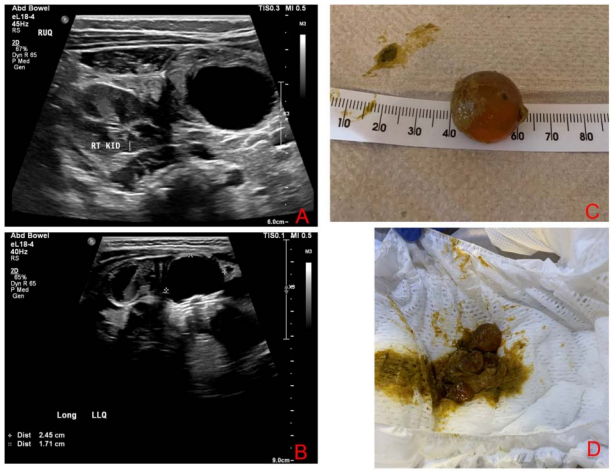
Introduction: Water beads are expanding, non-toxic toys made of water-absorbing polymer. Ingestion of such beads has the potential to cause bowel obstruction due to its ability to continually expand. There are few case reports of ingestion of water beads in infants, however, many required surgical intervention. Here, we describe an infant with water bead ingestions whose symptoms resolved with conservative management, using ultrasound as a supportive imaging modality.
Case Description/Methods: A 7-month-old male presented to the emergency department with gagging and poor intake after playing with water beads approximately 16 hours prior to presentation. Physical exam was normal. No foreign body was noted on nose-to-rectum Xray. Pediatric gastroenterology was consulted for esophagogastroduodenoscopy (EGD). Upon intubation, there was concern for foreign body in the right main bronchus due to decreased breath sounds. No foreign body was visible on either rigid bronchoscopy or subsequent EGD. An ultrasound confirmed a circular anechoic structure in the right upper quadrant, likely the small bowel. He was admitted for observation and made NPO due to the potential for developing bowel obstruction. A second ultrasound 16 hours later revealed an increase in size from 2.2cm to 2.5cm and progression to the left lower quadrant, unclear if located in the colon or small bowel. With a benign abdominal exam, the decision was made to resume breastfeeding. Soon after, he passed two stools containing multiple fragments of ruptured water beads and one fully intact bead measuring 2 cm. He was discharged home with no further reported problems. (Figure)
Discussion: The gastrocolic reflex from breastfeeding most likely contributed to our patient's ability to pass the water bead without further medical or surgical intervention. The use of ultrasound, while helpful, is not a widely recognized modality for detection of ingested water beads because it may not accurately depict the number, size, or location. Clinicians must consider the patient's clinical presentation, timing post-ingestion, diet, and suspected size and number of water beads in the management. Future considerations in the management include the utilization of osmotic agents, such as gastrografin or polyethylene glycol, to osmotically impact the bead's ability to enlarge further and thus avoid surgical intervention.



[3346] **Figure 1.** A. Initial ultrasound with circular anechoic structure in right upper quadrant. B Repeat ultrasound with circular anechoic structure now in left lower quadrant, slightly larger in size. C. Intact water bead found in stool. D. Water beads with one intact bead and multiple fragments.

S3347

The Thirteenth Case of a Congenital Portosystemic Shunt in a Down Syndrome Patient

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Introduction: Down syndrome is caused by non-disjunction (Trisomy 21), translocation, or mosaicism at chromosome 21 and is characterized by intellectual disability, dysmorphic facial features, and congenital malformations, including gastrointestinal abnormalities. Congenital portosystemic shunts (CPSS) are rare vascular malformations that form aberrant connections between a portal and systemic vein within the liver, diverting portal flow from the liver to the systemic venous system. The etiology of these malformations is poorly understood. Some hypotheses suggest dysfunction in angiogenic factors, which may play a role in the development of features already seen in Down syndrome, including placental hypo-vascularity, increased fetal nuchal fold thickness, and a tendency to develop pulmonary hypertension. There is an increasing appreciation of congenital portosystemic shunts in Down syndrome patients.
Case Description/Methods: Our patient presented with hyperammonemia, altered mental status, and choreiform movements. A computed tomography angiography of the abdomen and pelvis demonstrated a connection between the right portal vein and inferior vena cava, consistent with an intrahepatic congenital portocaval fistula. The lower lungs showed dilated pulmonary artery branches of the peripheral system, suggesting developing hepatopulmonary syndrome (HPS). Interventional radiology placed an 18 mm, Amplatzer PFO closure device within the congenital fistula. No residual flow was seen in the portal vein from the inferior vena cava post-procedure. Clinical improvement was noted following the procedure. His ammonia levels normalized. His mental status returned to baseline with the resolution of choreiform movements. A follow-up abdominal ultrasound demonstrated no residual or recurrent portocaval fistula. At the two-year follow-up, the patient had no recurring issues related to the fistula or closure device placement.
Discussion: Herein we describe the third case of a CPSS in a Down Syndrome patient and the 13th overall case of a CPSS with our approach to work up and treatment highlighted in the context of a literature review of similar cases.

S3348

Attraction-Not so Attractive: Case Series of Endoscopic Removal of Buckyballs from Colon via Magnetism of Insertion Tube

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Introduction: Neodymium magnets, also known as buckyballs, are powerful toy magnets that, when separated, can attract to one another with formidable force. Retrieval of these magnets requires endoscopy, oftentimes emergently, to prevent pressure necrosis of bowel wall between 2 magnets. Here, we describe two pediatric patients with magnet ingestion where the magnets were difficult to visualize in the distal small bowel/colon due to their attraction to the insertion tube of the colonoscope.
Case Description/Methods: A 14-year-old female accidentally swallowed 4 buckyball magnets. At presentation, the magnets were seen connected in a linear fashion in the stomach on Xray; however, the magnets had moved before endoscopy could be performed. Repeat X rays over a 54-hour period showed persistence of the magnets in the right lower quadrant despite the use of laxatives. CT abdomen

confirmed the magnets' position in the distal ileum or cecum. During colonoscopy and ileoscopy, no magnets were visualized. On careful withdrawal of the scope, the 4 magnets were seen attracted to one another in the rectum. The magnets were then removed with a Roth net. An 11-year-old male accidentally swallowed 2 buckyball magnets. The 2 attracted magnets moved to the right lower quadrant on X ray where they remained for 54 hours. During colonoscopy, no magnets were visualized in the cecum. However, during an attempt at ileoscopy, the connected magnets appeared in the cecum and the magnets were then moved with a Roth net. (Figure)

Discussion: For these patients, the magnets were not visualized initially during colonoscopy. It is our theory that due to their powerful magnetic force, the magnets were attracted to the magnetic insertion tube, proximal to the non-magnetic bending section of the scope and approximately 15 cm from the lens. Therefore, the endoscopist was not able to visualize the magnets until they were physically separated from the insertion tube. Endoscopists should be aware that the magnets may adhere to the insertion tube, which is not in the field of vision and can lead to confusion during colonoscopy. When considering this possibility, we recommend retroflexion of the bending segment if in an approximate location of the magnets per imaging. Additionally, we recommend re-examining the rectum as the magnets may disconnect from the scope as the insertion tube is withdrawn past the anal sphincter, leaving the magnets behind in the rectum to be visualized by the non-magnetic lens and bending section.

[3348] **Figure 1.** A. Buckyball magnets with no attraction to the bending segment of colonoscope B. Buckyball magnets attracted to insertion tube portion of colonoscope.

S3349

Colo-Colonic Intussusception Caused by a Sigmoid Duplication Cyst in a 6-Year-Old Child

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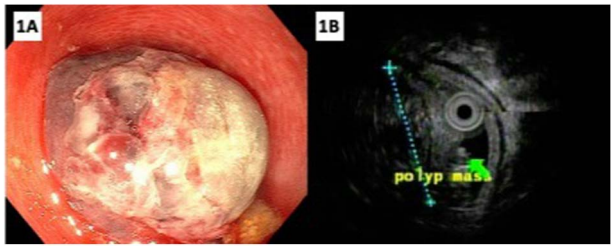
Introduction: Colo-colonic intussusception is rare with a prevalence of 0.2-4.4% and usually associated with colonic polyps and malignancy in adults. Distal colo-colonic intussusception is extremely rare in children and those caused by enteric duplication cysts as lead points are unusual. While enteric duplication is more commonly seen in children, the most common location is in the ileum and only 6.8 to 13% occur in the hindgut. It is usually diagnosed with ultrasound or CT and treated with surgery for removal.

Case Description/Methods: We present a previously healthy 6-year-old female who had abdominal pain for two days and bloody stools for one day. Abdominal pain was colicky and intermittent, and located in left lower quadrant. When examined, she was tender to palpation in the left lower quadrant but otherwise unremarkable. She had an abdominal ultrasound which showed intussusception in the left lower quadrant. CT of the abdomen and pelvis showed rectosigmoid intussusception without pneumatosis or free air. There was a round structure with a slender rim in the mid-low abdomen at the region of rectosigmoid junction with complicated fluid, suggesting possible gastrointestinal duplication cyst or a polyp. She underwent flexible sigmoidoscopy, which showed 2 cm x 3 cm intraluminal sigmoid colon duplication cyst that was sessile and ischemic (Figure 1A). Mucosa around the duplication cyst was friable, erythematous, and had loss of vascularity. Endoscopic ultrasound interrogation of the duplication cyst showed loss of mucosal differentiation, unclear involvement of muscular propria, and hypoechoic foci without calcification (figure 1B). It was not amenable for endoscopic removal. She subsequently underwent exploratory laparoscopy, which showed a ruptured duplication cyst on the mesenteric sidewall of the sigmoid colon. She then had laparoscopic partial sigmoidectomy with anastomosis. Pathology of resected duplication cyst showed serosal fibrosis with acute to chronic inflammation. She was discharged with return of bowel function and tolerating oral diet.

Discussion: The presented case is unique given the less common location of sigmoid duplication cyst and rare presentation of colo-colonic intussusception in a pediatric patient. This case also highlights the application of endoscopy and specifically EUS not only to diagnose and further characterize enteric duplication cysts, but also to potentially intervene therapeutically.



[3349] **Figure 1.** 1A. Flexible sigmoidoscopy showing duplication cyst. Figure 1B. Endoscopic ultrasound interrogation of the duplication cyst.

S3350

Compassionate Use of an Intermittent Colonic Exo-Peristaltis Device for a Handicapped Minor to Treat Chronic Constipation

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Introduction: A nurse professionally dedicated to her daughter (S) used to give S manual colonic massage which usually provoked her tendinitis due to daily administration and force needed. The pediatric gastroenterologist (EJH; Child.Hosp.Colorado), at the request of the mother asked the FDA for the compassionate use permission of MOWOOT device. This medical device automatically gives an intermittent colonic exo-peristaltis (ICE) that has been demonstrated to be safe and effective in a previous clinical trial (doi:10.14309/ctg.0000000000000267).

Case Description/Methods: S. was a 12 year-old girl, with anatomic proportions of a 6yr (22kg), with Trisomy 21, severe autism, mixed receptive-expressive language disorder, moderate to severe vision impairment in both eyes, self-aggressive crises, and severe chronic constipation from NBD origin. Prior treatments for constipation included PEG laxatives, extra fluids, extra fiber, and enemas. S received nutrition via a G-tube but frequently pulled out the tube. S lacked intellectual capacity for behavioural modification programs. She was placed an additional J-tube for nutrition. The G-tube was maintained for medication. The next treatment option was a surgically placed cecostomy tube for ante-grade enemas. Due to S behavioural issues, an additional tube was not likely to be successful, and S was at high risk of infection. Here the compasionate use started. For 45 days S. received 20min treatment daily. The mother registered in a bowel diary the number of bowel movements, fecal consistency, quantity of evacuated

feces, and dose of laxative taken. Before and after the study the bowel function was assessed on a visual analogue scale (VAS, 0:No problems; to 10:A lot of problems to defecate). To use the ICE device, both G and J tubes had to be disconnected. The active pneumatic elements of the belt directly worked onto the occluded osteomas. (Figure)

Discussion: Despite the challenges, the treatment worked well from and there was no adverse event reported. S enjoyed the treatment from the first day and reduced her self-aggressiveness. After 4 weeks of use, the bowel movements increased by 2/week, while the dose of laxatives decreased. The defecatory function ameliorated from 8.5 to 2.5 (VAS, d1 to d45). The improvement progressed over time. Three months later, the patient had gained 8 Kg weight and was notably calmer.The safety, tolerability, and effectiveness of the ICE system was demonstrated. The patient's and family's whole quality of life was significantly increased.

[3350] **Figure 1.** S. at home using the ICE medical device. Photograph sent by her mother.

Table 1. Results of the 45 day follow-up. The "minus" sign indicates a reduction at the end of treatment (w6) respect baseline (w1). Slope is shown as mean (SD)

	Week 1	Week 3	Week 6	Diference (w6 - w1)	Linear Regression		
					R^2	slope	P
Feces (g/w)	3150	3500	3600	450	0.80	-0.22 (0.14)	0.0405
Laxatives (cc/w)	280	245	185	-95			

S3351

Fulminant Hepatitis Becomes the Initial Presentation of X-Linked Lymphoproliferative Disease

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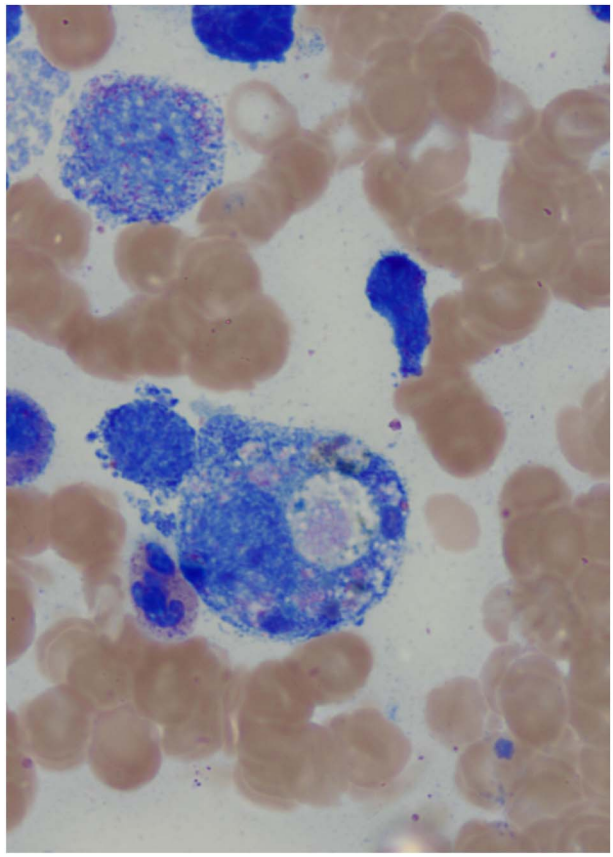
Introduction: Fulminant liver failure in the pediatric population remains an evolving topic due to its high impact on morbidity and mortality. Although, numerous causes have been proposed, 45% of cases remain cryptogenic. This abstract raise awareness of an incidence of fulminant hepatic failure secondary to Epstein Barr virus (EBV) infection in the presence of an unknown underlying rare immunological phenomenon, X-Linked Lymphoproliferative Syndrome (XLP).

Case Description/Methods: 17-year-old male with a past medical history of B Cell Lymphoma with colonic involvement at the age 3 presented with a 2-week history of fever, right upper quadrant abdominal pain, and jaundice associated with vomiting, diaphoresis, and weight loss. Pertinent positive labs included albumin 2.9 g/dL, T Bili 5.7 mg/dL, Alk Phos 806 U/L, AST 675U/L, ALT417 U/L, GGT 381 U/L, INR 1.58, Triglycerides 334 mg/dL, Ferritin 26,000 ng/mL, Fibrinogen 97 mg/dL. EBV PCR and flow cytometry confirmed an acute EBV infection. CT abdomen/pelvis showed hepatosplenomegaly, proctocolitis, and widespread lymphadenopathy. Liver U/S revealed gallbladder sludge without biliary dilatation. The intractable fevers, hepatosplenomegaly and labs indicating coagulopathy, lactic acidemia, hypoglycemia, and hyperammonemia were concerning for acute liver failure requiring organ transplant. Pancytopenia, agammaglobulinemia, thrombocytopenia and hyperferritinemia suggested Hemophagocytic Lymphohistiocytosis (HLH) confirmed by bone marrow biopsy. Despite antibiotics, antifungals and immunosuppressive therapy—steroids, IVIG, Rituximab, Alemtuzumab administration, hepatic encephalopathy ensued leading to death. Post-mortem genetic panel was positive for UNC13D and SH2D1A gene mutation. (Figure)

Discussion: EBV is commonly a self-limiting virus. In rare cases, EBV produces an alteration in NK and Cytotoxic T Cells which leads to the inability to properly eliminate macrophages and CD8+ T cells causing the immune dysregulation known as HLH. Suspicion for inherited HLH was supported by UNC13D gene mutation, hemophagocytes on bone marrow biopsy (Figure 1) and decreased levels of NK and T cells. This notion was challenged when genetic panel revealed a mutation in SH2D1A gene unique to XLP, which idiopathically causes sensitivity to EBV. XLP is linked with Non-Hodgkin's B Cell Lymphomas manifesting extra-nodal in the intestinal tract. EBV-associated hepatitis and agammaglobulinemia w/ or w/o B cell malignancy should raise suspicion for XLP and prompt early initiation of treatment.



[3351] **Figure 1.** Illustrates the numerous hemophagocytes seen on bone marrow biopsy supporting the diagnosis of Hemophagocytic Lymphohistiocytosis.

S3352

Gut Microbiome 6 Months After Fecal Microbiota Transplant (FMT) in an Adolescent With Autism Using Familial Donor

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Introduction: Autism affects 1/40 to 1/100 children in America and worldwide. Autism Spectrum disorder (ASD) is a varied set of neurobehavioral changes characterized by distinct, repetitive behaviors along with impaired socialization and communication. Several studies point to gut dysbiosis present in these children, leading to intestinal microbiota transplant as a therapeutic option. Studies by Kang, Adams, and Borody et al. showed a 50% regression of neurological symptoms in children 2 years post-transplant. Cases show more improvement in younger patients, suggesting early treatment with a better donor, such as a family member, especially in the older ASD patients.
Case Description/Methods: Fecal matter from a healthy sibling (15 year old sister as donor) was infused via colonoscopy into the subject's gastrointestinal tract following a protocol approved by FDA as an Investigational New Drug. This study was IRB approved. Fecal samples were collected at baseline (patient and donor) and then from the patient at week 5, and month 3 and 6. Metagenomic Next Generation Sequencing was performed on fecal samples, where DNA samples were extracted and normalized for library downstream fabrication using Shotgun methodology. There were marked improvements in the patients' gut microbiome post-transplant. Compared to baseline, there was a decrease in relative abundance of phylum Proteobacteria from 49% to 1.3%. Over the 6 months a significant increase was noted post-transplant in phylum Actinobacteria (0.012% to 2.5%) and specifically genus *bifidobacteria* (0.0% to 1.9%). Species *Lactobacillus animalis*, absent in the donor, disappeared in the recipient (from 58% to 0.0%) after transplant. Shannon Index of bacterial diversity significantly improved (2.2 to 6.2) almost matching donor who was 6.7. Clinically, the patient progressed from aggressive to calm and said his first 2 words "mama" and "baba" after 1 month. Childhood Autism Rating Scale (CARS) testing is underway, but given recent nature of the implant and subject's age (19 yrs.) substantial changes are not yet expected.
Discussion: This is the first case demonstrating implantation of microbes from a familial donor (sister) to an adolescent patient (brother) with Autism. Although early in clinical assessment, this case demonstrates disappearance of an overabundant microbe (*L. Animalis*) and restoration of phyla and species missing or in low numbers in the autistic patient when compared to his sister.

S3353

Improvement in Paediatric Autoimmune Neuropsychiatric Disorders Associated With Streptococcal Infections (PANDAS) Syndrome Following Treatment With a Combination of Antibiotics and Fecal Microbiome Transplantation (FMT)

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Introduction: Paediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal Infections (PANDAS) syndrome is a condition that impairs normal neurological brain function and is triggered by a streptococcal infection. It is characterised by an obsessive-compulsive disorder (OCD) tic disorder, or both and usually occurs suddenly between the age of 3-12 years. Other symptoms include mood changes, anxiety, attention deficit/hyperactivity disorder, trouble sleeping, and joint pain. Metagenomic profiles of those with PANDAS syndrome show an altered gut bacterial community. Commonly treatment involves antibiotics for streptococcal infection. Here, we report on the effects of fecal microbiota transplantation (FMT) on a set of identical twins with concomitant PANDAS syndrome.
Case Description/Methods: Case 1: A male (16yrs) presented with slow transit type constipation with a colonic transit half clearance time of 70minutes. His medical history also showed he had PANDAS syndrome and later tested positive for Clostridium Difficile (C. difficile). The patient was treated with metronidazole before receiving a single dose of FMT which was transfused colonoscopically. At 1-month the patient tested negative for C. difficile. Post treatment he reported significant improvement in abdominal pain, bloating and constipation that lasted 2 years. Simultaneously he reported symptom improvement in anxiety, depression, and Rheumatoid Arthritis which are associated with PANDAS syndrome. Case 2: A male (16yrs) presented with severe constipation with a normal colonic transit half clearance time of 50minutes. Constipation prompted a worsening of symptoms of concomitant PANDAS syndrome which had previously been treated with steroids. After further testing he was found to have C. difficile for which he was prescribed metronidazole and vancomycin before receiving a single dose of FMT by colonoscopic infusion. At 1-month post treatment eradication of C. difficile was confirmed. Post treatment he reported

experiencing improvement in constipation, diarrhoea, flatulence, and vomiting which lasted for 2 years. He also reported symptom improvement in Autism, OCD, and Rheumatoid Arthritis which are linked to PANDAS syndrome.
Discussion: To the best of our knowledge these are the first cases of improvement in PANDAS symptoms after FMT treatment. Further study is required into the effects of FMT on PANDAS syndrome before FMT can be confirmed as a potential treatment.

S3354

Ménétrier's Disease: A Rare Cause of Edema in Childhood

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Introduction: Ménétrier's disease (MD) is a rare protein-losing gastropathy characterized by enlarged gastric rugal folds. Adult MD is associated with a high degree of morbidity and mortality due to potential malignant transformation and need for gastrectomy. Pediatric MD, in contrast, is typically an acute, self-limited process, frequently associated with cytomegalovirus (CMV) infection of the stomach.
Case Description/Methods: A previously healthy 2-year-old boy was admitted for generalized edema, which had been preceded by a week of diarrhea. Growth and development had been normal. Initial laboratory evaluation was notable for serum albumin of 1.3 g/dL. Other labs were largely unremarkable. Cardiac, renal, and liver etiologies were excluded. Esophagogastroduodenoscopy showed a normal gastric antrum, but the remainder of the stomach had enlarged rugae with areas of ulceration and nodularity. The duodenum and esophagus appeared normal. Histology was notable for hyperplastic gastric pits and foveolae. Immunohistochemistry for CMV and *Helicobacter pylori* were negative, and viral culture of a gastric sample was also negative for CMV. CMV serologies showed elevated IgM and IgG levels. Additionally, a viral enteric pathogens panel was positive for sapovirus. The patient received 2 doses of albumin and antisecretory agents during admission, but his edema and hypoalbuminemia improved without treatment of CMV.
Discussion: In a toddler presenting with edema and hypoalbuminemia, it is necessary to consider protein-losing gastroenteropathies on the differential. Endoscopic evaluation with biopsy is essential for diagnosis of MD. Histology may show foveolar hyperplasia with cystic dilation of pits of the gastric body with relative sparing of the gastric antrum. Full-thickness biopsy may be required to detect classic histologic findings. Evaluation for infection should be done as one-third of pediatric cases are associated with CMV. Management of pediatric MD is primarily supportive due to the self-limited nature of the condition. Treatment with ganciclovir can be considered for severe or refractory cases associated with CMV.

S3355

Postnatal Cytomegalovirus Infection as a Potential Trigger for Very Early Onset Crohn's Disease in an Immunocompetent Infant

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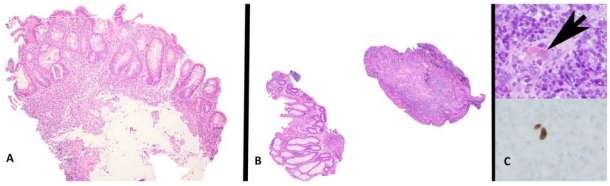
[1]Oakland University William Beaumont School of Medicine, Rochester, MI; [2]Beaumont Health, Royal Oak, MI; [3]Beaumont Children's Hospital, Troy, MI; [4]Beaumont Children's Hospital, Royal Oak, MI.

Introduction: Intestinal complications of congenital and postnatal cytomegalovirus (CMV) infections include bloody diarrhea, necrotizing enterocolitis, and perforation. CMV infection triggering inflammatory bowel disease (IBD) is rarely recognized, with most cases reported in adults (>60% ulcerative colitis). Proposed mechanisms include alteration of intestinal immunity with viral proteins upregulating inflammation. IBD in infancy is rare and often associated with monogenic pathogenic mutations. We describe a girl with colitis onset at 2 weeks of age in whom postnatal CMV infection appears to have been the event triggering the recognition of Crohn's disease (CD).
Case Description/Methods: A 15-day-old term breast-fed girl with nonsyndromic anomalies and negative urine CMV PCR on day 2 of life presented with frequent bloody/mucousy stools. Milk-protein allergy was suspected. With amino acid-based formula, she had fewer bowel movements, but mucous/blood persisted. Sigmoidoscopy at 4 months revealed moderately congested mucosa with erosions throughout the sigmoid colon and rectum (Figure 1A); methylprednisolone was started. Biopsies showed chronic moderate to severe colitis with negative CMV by immunostaining; urine CMV PCR was positive. CMV IgG and IgM were markedly elevated. Endoscopy 3 days later showed chronic inactive duodenitis, mild chronic gastritis, chronic inactive ileitis, and chronic colitis with rare CMV-positive cells in the descending colon and rectum. She was treated with valganciclovir (VAL) for CMV colitis and azathioprine (AZA) + prednisolone for possible CD, with partial response. Immune deficiency work up was negative (Table 1). On day 229 of life, colonoscopy revealed some disease improvement; CMV-positive cells were more abundant (Figure 1B/1C). She was switched to IV ganciclovir for 6 weeks followed by oral VAL; AZA was discontinued and steroids weaned. Symptoms persisted and she was treated with bowel rest and parenteral nutrition. Biopsies on day 266 of life showed CD pathology with absence of CMV. At 9 months, she started infliximab (INF) and demonstrated clinical improvement. VAL was stopped after her third INF dose. Table 1 details event chronology.
Discussion: This case illustrates how postnatal CMV infection in a seemingly immunocompetent infant without CD-associated gene mutations can lead to earlier IBD onset. Treatment of CMV colitis is needed in conjunction with immunosuppressive therapy for CD. The frequency of CMV infection as a trigger for infantile CD needs further study.

Figure 1. Tissue histopathology with CMV immunostaining



A) Crypt architectural distortion, acute cryptitis, and crypt abscess (day 138 of life)
B) Tissue fragment to the left shows some mild crypt architectural distortion and the one to the right shows complete ulceration with granulation tissue and abundant mixed acute and chronic inflammation (day 229 of life)
C) High-power images of tissues biopsied on day 229 of life showing on top a small-sized vessel with a cytoplasmic inclusion in an endothelial cell (arrow) and positive CMV staining (brown staining) in tissue (bottom image)

[3355] **Figure 1.** See above caption.

Table 1. Chronology of Procedures, Diagnoses and Treatment Modalities

Day of Life	Clinical Event
2	Urine CMV by PCR negative; done for ventriculomegaly, sensorineural hearing loss, and secundum atrial septal defect
15	Blood and mucous in stool, milk-protein allergy suspected
138	Sigmoidoscopy, moderate-to-severe chronic colitis, CMV immunostain negative
139	Methylprednisolone 0.5 mg/kg/dose IV BID started
140	Urine CMV PCR positive, CMV IgM 80.9 AU/mL (positive if ≥35) and CMV IgG >10 U/mL (positive if ≥0.70), increased methylprednisolone dose
141	EGD/Colonoscopy, chronic colitis, ileitis, duodenitis, gastritis, IV ganciclovir started 5 mg/kg/dose BID, continued corticosteroids; rare-CMV positive cells on immunostaining in descending colon and rectum
150	Oral Valganciclovir started at 16 mg/kg/dose BID

Table 1. (*continued*)

Day of Life	Clinical Event
157	Invitae Primary Immunodeficiency Panel (407 genes) and Comprehensive Deafness Panel (203 genes) showed two pathogenic mutations (hemizygous) in *CTNS* and *SLC26A3*, but no other known mutations associated with primary immune deficiency, including *NOD2*
174	Weaning oral steroids while on oral valganciclovir
181	Azathioprine 1 kg/mL day started for partial response to therapy
229	EGD/Colonoscopy, chronic colitis, increased CMV-positive cells on immunostaining in transverse, descending, and sigmoid colon as well as rectum, weaned steroids
231	IV ganciclovir started for a 6-week course
237	Azathioprine discontinued
246	CMV DNA undetectable in serum
254	TPN, prednisolone restarted, urine CMV by PCR negative
266	Colonoscopy, complete suppression of CMV but chronic inflammation still present, oral valganciclovir started
274	First infliximab infusion 10 mg/kg q3w, continued bowel rest and TPN
292	Gastrostomy
321	Third infliximab dose, clinical relapse, oral valganciclovir discontinued
330	Azathioprine restarted to decrease clearance of infliximab
363	Colonoscopy, rectal ulcer with positive CMV cells on immunostaining, improved or absent colitis elsewhere, oral valganciclovir re-started

Abbreviations: BID, Twice daily; CMV, Cytomegalovirus; EGD, Esophagogastroduodenoscopy; PCR, Polymerase chain reaction; IBD, Inflammatory Bowel Disease; IV, Intravenous; q3w, Every 3 weeks.

S3356

A Case of Eosinophilic Gastritis in a Pediatric Patient Effectively Treated With a Multimodal Approach

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Introduction: Eosinophilic gastrointestinal diseases (EGIDs) are a spectrum of rare, chronic, immune-mediated gastrointestinal disorders histologically characterized by eosinophilic-predominant tissue inflammation and GI symptoms. Here we describe a pediatric case of new-onset eosinophilic gastroenteritis with hyper-eosinophilic syndrome.
Case Description/Methods: Our patient is a 15 y/o male who presented with a 3-week history of intermittent episodes of non-bilious, non-bloody emesis and accompanying 13-pound weight loss over 3 months. He had no notable past medical history and lack of apparent trigger for the GI symptoms. Initial EGD showed several ulcerations of the esophagus and stomach and mild eosinophilia in the esophagus and duodenum. He was also urease positive. He was discharged on triple therapy for a likely H. Pylori infection. He presented again in one month with worsening abdominal pain and hematemesis. Repeat EGD was notable for eosinophils > 30/HPF in the duodenum and eosinophils > 100/HPF in the stomach and distal esophagus. His peripheral eosinophil count was 5370. He met criteria for hypereosinophilic syndrome with eosinophilic gastroenteritis and was initiated on parenteral steroids. In the outpatient setting, he followed up with pediatric allergy and GI. He was initiated on mepolizumab, steroids, PPI, gabapentin, and dairy-free diet and his symptoms appeared to be well-controlled. Repeat EGD showed stable to improved gastric eosinophilic inflammation. However, he was re-admitted in the following months for worsening symptoms including feeding intolerance and unintentional weight loss. His diet was restricted to elemental formula and eventually weaned to an oral diet with exclusion of dairy, wheat, soy, eggs, nuts, and seafood. He achieved complete remission of symptoms with adequate weight gain and improved feeding tolerance. Full regular diet was gradually reintroduced and medications were stopped, only requiring a monthly mepolizumab injection. He has recovered all lost weight and has no symptoms on mepolizumab monotherapy with pending endoscopy to assess for mucosal healing.
Discussion: Therapy for EGIDs is complicated and involves a multimodal approach including management of abdominal pain, steroids, biologic agents, and diet restrictions. This case demonstrates complete remission of symptoms related to eosinophilic gastritis in a pediatric patient following therapy with PPI, steroids, mepolizumab, elemental diet and maintenance treatment with mepolizumab alone.

S3357

Achalasia Cardia in Child With Severe Malnutrition: A Case Report

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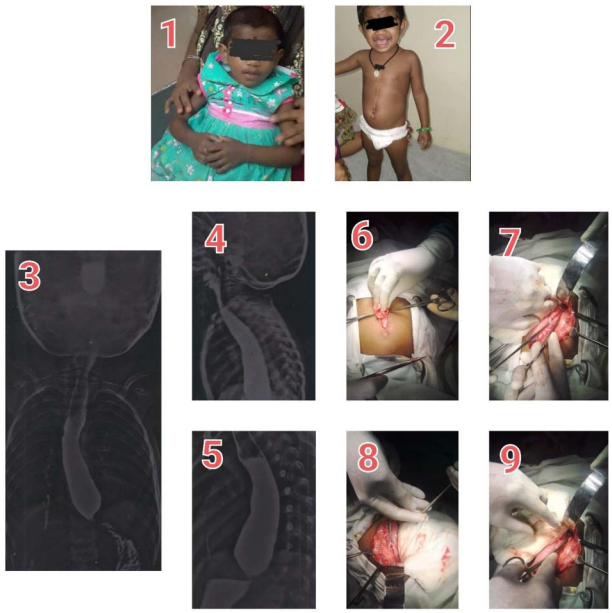
Introduction: Achalasia cardia is an esophageal motility disorder due to the failure of the lower esophageal sphincter to relax. It is extremely rare in the pediatric population with an incidence of 0.11 per 100,000 children. In children, it is usually associated with other conditions like Adrenal glucocorticoid deficiency and Down's syndrome. The symptoms include vomiting, regurgitation, and weight loss. Its exact etiology is still unclear. It is considered to be due to degeneration of the myenteric plexus that innervates the lower esophageal sphincter leading to difficulty with swallowing and absence of peristalsis.
Case Description/Methods: We present the case of a 3-year-old girl with a history of recurrent vomiting and poor weight gain. Her symptoms have been progressive for over 2 years. The child was initially treated at multiple local hospitals for Gastroesophageal reflux disease with no improvement. At presentation, she was severely malnourished and weighed 6 kgs. On evaluation, she was also found to be anemic. After admission to our pediatric unit, she underwent a barium study which showed tapering of the distal esophagus, representing the classic bird-beak sign, and the diagnosis of achalasia cardia was confirmed. She was treated surgically by Heller's Cardiomyotomy and Fundal Wrap. Her post-operative phase was uneventful and she was discharged with follow-up advice. She exhibited excellent recovery with good weight gain and resolution of her symptoms. Her weight at the 1-month follow-up visit was 9kgs and at 3 months it was 12kgs. (Figure)
Discussion: Achalasia cardia is a rare presentation in children but can lead to major health concerns if left untreated. Due to the nonspecific symptoms, it is mostly misdiagnosed as gastroesophageal reflux disease which leads to delayed diagnosis and severe malnutrition. Diagnosis is with barium swallow study and manometry. It is followed by endoscopic biopsy in adults to rule out malignancy, however, it is not a consideration in children. Medical management has often shown recurrence of symptoms, particularly in children, and is not considered an effective choice of treatment. Surgery is the definitive treatment and includes Heller's esophagomyotomy with or without anti-reflux procedures. Prompt diagnosis and treatment have an excellent prognosis. Peroral endoscopic myotomy is a promising new therapy in adults with achalasia but has limited studies in children. Persistent undiagnosed cases can lead to severe malnutrition and failure to thrive.



[3357] **Figure 1.** 1: At presentation, weight 6kgs; Fig2:At 3months, weight 12kgs; Fig.3,4,5: Barium Study; Fig6: Supraumbilical midline incision; Fig7:Cardiomyotomy; Fig8:Approximation of Crura; Fig9:Fundal Wrap.