Publications by ISPGHAN members in Pubmed Indexed Journals (July-Sep 2020)

JULY 2020

1.) **Ujjal Poddar, Surender K. Yachha, Narendra Krishnani, Niraj Kumari, Anshu Srivastava and Moinak Sen Sarma.** ***Solitary Rectal Ulcer Syndrome in Children: A Report of 140 Cases.*** **JPGN 2020 July; 71: 29–33**

Solitary rectal ulcer syndrome (SRUS) is a chronic, benign but difficult to treat condition, described mainly in young adults. The common presentations are rectal bleeding, passage of mucus, excessive straining during defecation, a sense of incomplete evacuation, constipation, and sometimes rectal prolapsed.

Authors have aimed to describe their experience in a large cohort of children with solitary rectal ulcer syndrome (SRUS) with regard to clinical presentations, endoscopic appearance, and treatment outcome. Clinical, endoscopic and histopathologic data of children (18 years or younger) diagnosed with SRUS between 2000 and 2018 were retrieved and analyzed. They have concluded that majority of cases of SRUS presented in second decade with rectal bleeding and features of dyssynergic defecation. Ulcer was noted in 72% cases. The outcome of medical treatment with behavioural modification and local therapy was modest. Two thirds of children without overt rectal prolapsed showed clinical response to behavioural modification along with corticosteroid enema.

2.) **Moinak Sen Sarma and Surender Kumar Yachha. Applebite colon in a child. Digestive Liver Disease.2020 July 6.** doi**: 10.1016/j.dld.2020.07.001** [Epub ahead of print]

The authors have reported on 10 year old girl who presented with frequent painless bleeding per rectum and moderate splenomegaly for 18 months. There was no history of variceal bleeding. Systemic examination was normal. Hemogram was suggestive of hypersplenism (hemoglobin: 9.7 g%, total leucocyte count 4000/mm^3, platelet count 85,00 0/mm3) with normal liver function tests. Colonoscopy showed large deep punched out discrete ulcers with everted edges and white exudates in rectosigmoid and normal areas in the rest of the colon. Histopathology and tissue cultures were not suggestive of inflammatory bowel disease, tuberculosis or immunodeficiency. Stool examination for opportunistic pathogens was normal. Esophagogastroduodenoscopy showed small esophageal varices. Magnetic resonance portovenography showed a portal cavernoma (including right and left branches) with congested inferior mesen- teric vein, patent splenic and superior mesenteric vein. A diagnosis of extrahepatic portal venous obstruction (EHPVO) with portal colopathy was considered. Child underwent a central end-to-side splenorenal shunt with splenectomy. Repeat colonoscopies at 3 and 6 months showed rapid healing of the ulcers. At follow-up of 5 years, child continued to be asymptomatic.

Portal colopathy is seen in 36% of EHPVO but rarely symptomatic. Other than anorectal varices, small mucosal lesions are usually reported. Large colonic ulcers are a rare and unusual manifestation of portal colopathy as a primary presentation in EHPVO.

3.) **Jagadeesh Menon, Naresh Shanmugam, Kinisha Patel, Abdul Hakeem, Mettu Srinivas Reddy and Mohamed Rela.** ***Awareness and concerns about novel Coronavirus disease 2019 (COVID-19) among parents of pediatric liver transplant recipients.*** **Pediatric Transplantation. 2020 July 07. DOI: 10.1111/petr.13805** [Epub ahead of print]

Authors undertook a web-based survey among parents of post–liver transplant pediatric patients to assess knowledge and concerns regarding COVID-19 pandemic and impact of social media on them.

This cross-sectional online survey was conducted between March 21 and March 26, 2020. A 19-item questionnaire was sent to 172 parents of post–liver transplant children. 106 (62%) of parents responded. 100% of parents understood concept of social distancing. Television followed by newspapers was the main source of their information; though over 40% claim to regularly receive information through social media. 87% would consult their doctor if the child had flu-like symptoms rather than modify immunosuppression or try alternative medications. Parental concerns mainly revolved around early recognition of symptoms, queries on unconventional treatments circulating over social media, and supply of medications during the lockdown period. Authors concluded that majority of parents had basic understanding of COVID-19 pandemic and social media appeared to be an important source of information.

4.) **Saalim Nazki, Ravi Prakash Kanojia, Monika Bawa, Vineet Binu, Sadhna Lal, Ashwani Sood and Ram Samujh. Robotic Excision of Choledochal Cyst with Hepaticoduodenostomy (HD): Report of HD Technique, Initial Experience, and Early Outcome. Eur J Pediatr Surg.**2020 July 15.doi.org/ 10.1055/s-0040-1713933. [Epub ahead of print]

Minimal access surgical approach to choledochal cyst (CC) is becoming a standard of care in pediatric age group. Robotic-assisted excision of CC is increasingly being practiced at centers which have access to the system.

Authors have presented their experience with robotic excision of CC and technique of hepaticoduodenostomy (HD) in this retrospective study (August 2017 and March 202). Over all initial experience, short-term outcomes and complications are also presented and discussed. Patients with active cholangitis, liver dysfunction, and perforated CC were excluded for robotic procedures. They underwent excision of CC with HD. The duodenal anastomosis was done after limited mobilization and emphasis was laid on anastomosing the distal D2 part to the common hepatic duct. This prevents bile

reflux into stomach. The follow-up evaluation was done for these patients. Hepatobiliary iminodiacetic acid (HIDA) scan for duodenogastric reflux (DGR) was done only if patients reported symptoms related to it. A total of 19 patients (10 females) were studied. The mean age was 84 months. Type 1b was present in 12 patients and the rest were type IVb. Complete cyst excision with HD was done in all patients except conversion to open in one patient. The mean surgical time was 170 _ 40 minutes with console time of 140 _ 20 minutes. Median follow-up duration is 2.5 years (range: 3.5–0.5 years). HIDA scan was done in five patients who had reported epigastric pain. Of these five, one patient had a positive DGR. He is on conservative management. Authors have concluded that robot-assisted CC excision with HD is feasible as proven by the outcome of 19 patients presented in this series. HD is to be done away frompylorus in distal part of down curving D2. This particular step prevents DGR and is themost important point of technique in doing HD.

5.) **Rupjyoti Talukdar, Mohsin Aslam, D. Nageshwar Reddy, Zaheer Nabi, Upender Shava, V. V. Ravikanth, Steffie Avanthi and B. Govardhan. Pancreas Divisum Increases the Risk of Recurrent Acute Pancreatitis in Patients with *rs12338* Polymorphism in the Cathepsin B Gene.** Digestive Diseases and Sciences.2020 July 22.doi.org/ 10.1007/s10620-020-06517-7 [Epub ahead of print]

Pancreas divisum (PD) as a cause of pancreatitis has been debated.

In this prospective study conducted at a high-volume academic centre from May 2015 to September 2016, the authors have reported the association of multiple gene polymorphisms on the risk of recurrent acute pancreatitis in the presence of PD. This study enrolled 687 individuals (167 idiopathic recurrent acute pancreatitis [IRAP], 276 idiopathic chronic pancreatitis [ICP] and 244 unrelated healthy controls) including children under 18 years of age. Patients were divided into those with/without PD. Associations between the significantly prevalent single-nucleotide polymorphism (SNPs) and IRAP/ICP in the presence of PD were evaluated. Cathepsin B Gene [CTSB] *(rs12338)* polymorphisms were significantly associated with IRAP [OR (95% CI) 2.44 (1.41–4.22); $p = 0.001$] among patients with PD. No association was observed with ICP. Authors have concluded that risk of RAP due to PD increases in patients with rs12338 polymorphism in the cathepsin B gene.

6.) **Neelam Mohan and Shivani Deswal. *Corona Virus Disease (COVID-19) Fecal-oral transmission: Is it a potential risk for Indians?* Indian Journal of Gastroenterology.2020 July 24.** doi.org/10.1007/s12664-020-01072-5 [Epub ahead of print]

Given the potential for fecal–oral transmission of the SARS-CoV-2 virus, authors have commented that public education regarding individual hygienic measures like frequent hand washing, covering pot after defecation, flushing, cleaning with bleach, avoiding open defecation and restricted use of public toilets need to be advocated proactively along with social distancing especially in a country like India where fecal-oral transmission of diseases is a major public health problem due to lesser degree of sanitation practices.

7.) **Upender Shava, Anshu Srivastava, Amrita Mathias, Narvesh Kumar, Surender Kumar Yachha, Sanjay Gambhir and Ujjal Poddar. *Functional dyspepsia in children: a study of pathophysiological factors.* J Gastroenterol Hepatol.2020. July 25.doi: 10.1111/jgh.15193**[Epub ahead of print]

Authors have evaluated children with functional dyspepsia (FD) for abnormality of gastric accommodation and emptying, psychological stressors (PS), *Helicobacter pylori* (HP) infection and post-infectious FD. Diagnosis of FD was based on ROME III criteria. Clinical evaluation including dyspeptic symptom scoring and assessment for psychologic stressors (PS) was done. Satiety drink test (SDT) for gastric accommodation, gastroscopy with biopsy for helicobacter pylori (HP) infection and solid meal gastric emptying (GE) by scintigraphy were done. 67 healthy children were enrolled for assessing PS and SDT. 55 FD children (33 boys, age 12[6–18] years) with symptoms for 4(2-48) months and dyspeptic score of 5(1-13) were enrolled. PS were more common in FD than in controls. Median satiety drink volume (SDV) was 360 ml (180-1320 ml), no patients had SDV of <5th centile of healthy children. The frequency and severity of postprandial symptoms (PPS) was higher in FD than in controls. Delayed GE was present in 6.5%, HP infection in 11% and post-infectious FD in 13% cases. Etiological factor was identified in 87%children, with 20% having multiple factors. Authors have concluded that abnormality of gastric sensorimotor function is seen in one fourth of FD cases. HP infection and post-infectious FD are present in 11% and 13% cases respectively

8.) **Rishi Bolia and Anshu Srivastava. *Natural history of pancreatic fluid collections: are children different from adults?* Pancreatology.2020 July 27. doi.org/10.1016/j.pan.2020.07.403** [Epub ahead of print]

Authors have written this article as a comment to the article published in Pancreatology by Lal et al. summarized as item (2) under March 2020.

In the cohort by Lal et al, 28 % (11/39) mature PFCs were pseudocysts suggesting that unlike adults, pseudocysts continue to be seen in children. An MRI or EUS were done only in 12 children subjected to drainage. The remaining (n-27) were classified as pseudocyst or walled-off necrosis (WON) only on the basis of USG (or maybe CT in some) which has a poor sensitivity and raises concerns about the accuracy of classification. The authors have pointed out that while studying any natural history "timelines" are important. The information about day of pancreatitis when child was admitted was missing in the article by Lal et al. Similarly, total follow–up time after drainage (especially after stent removal), imaging protocol followed before stent removal and information about recurrence was also lacking. They have concluded that pediatric pancreatic fluid collections (PFCs) are different from those in adults with regards to the etiology, type and outcome. Further, age dependant limitations of radiologic testing (EUS, MRI etc) govern the choice of evaluation. There is a need of modifying adult definitions and guidelines suitable for children.

AUGUST 2020

1.) **Mridul Chandra Das, Moinak Sen Sarma, Anshu Srivastava, Surender Kumar Yachha and Ujjal Poddar. *Effect of chelation therapy in pediatric Wilson's disease: liver and endoscopic outcome.* J Hepatobiliary Pancreat Sci.2020. August 03. doi:10.1002/JHBP.812**[Epub ahead of print]

This study was aimed to evaluate of efficacy of chelation on hepatocellular function and portal hypertension in pediatric

Wilson's disease (WD). Clinical, biochemical, endoscopic and treatment data of children with WD from June 2007 to June 2018 (n=111) were extracted and analyzed. All patients were started on D-penicillamine and supportive therapy. Zinc was added on if there was failure of clinical improvement to D-penicillamine within 4 weeks of starting therapy. Only asymptomatic patients with mild liver inflammation were started on zinc monotherapy. In case of drug intolerance at follow-up, chelators were changed accordingly. Patients with ≥9 months of follow-up (n=65) were evaluated for response to chelation therapy in the following categories: a) complete remission, b) partial remission c) progression of disease; d) drug toxicity. Favourable outcome (complete and or partial remission), progression of disease and drug toxicity were seen in 71%, 29% and 10.8% respectively. Two-third had esophageal varices which did not show progression. The indicators of poor outcome were PELD score (cut-off >9.45) and Nazar score (cutoff >3.5) with fairly reliable ROC with AUC of 0.71 and 0.68 respectively. Authors have concluded that despite severe liver disease, majority of the pediatric hepatic WD can be managed on D-penicillamine monotherapy. PELD score and Nazar score effectively determines the outcome

2.) **Rupjyoti Talukdar, Mohsin Aslam, D. Nageshwar Reddy, Zaheer Nabi, Upender Shava, V. V. Ravikanth, Steffie Avanthi, B. Govardhan.** ***Pancreas Divisum Increases the Risk of Recurrent Acute Pancreatitis in Patients with rs12338 Polymorphism in the Cathepsin B Gene*****. Digestive Diseases and Sciences.2020. August 04. doi.org/10.1007/s10620-020-06517-7** [Epub ahead of print]

In this study, authors have reported the association of multiple gene polymorphisms on the risk of recurrent acute pancreatitis (RAP) in the presence of Pancreas Divisum (PD). They have enrolled 687 individuals including children [age <18years] {167 idiopathic recurrent acute pancreatitis (IRAP), 276 idiopathic chronic pancreatitis (ICP), and 244 unrelated healthy controls) from May 2015 to September 2016. Patients were divided into those with/without PD. Associations between the significantly prevalent SNPs and IRAP/ICP in the presence of PD were evaluated. Thirty-three (19.8%) and 82 (29.7%) patients with IRAP and ICP, respectively, had PD. CTSB *(rs12338)* polymorphisms were significantly associated with IRAP [OR (95% CI) 2.44 (1.41–4.22); $p = 0.001$] among patients with PD. No association was observed with ICP. They have concluded that risk of RAP due to PD increases in patients with rs12338 polymorphism in the cathepsin B gene.

3.) **Jaya Agarwal.** ***Pancreatobiliary endoscopic interventions for pediatric pancreatic pathology.*** **Digestive Diseases and Sciences. 2020. August 05.doi.org/10.1007/s10620-020-06513-x** [Epub ahead of print]

In this invited review, the author has collated the current literature regarding diagnostic and therapeutic uses, benefits, limitations, and clinical outcomes of endoscopic ultrasound (EUS) and endoscopic retrograde cholangiopancreatography (ERCP) in pediatric pancreatology. ERCP and EUS have clearly established their safety and efficacy in children primarily based on outcomes from retrospective studies. Author concludes by stating that there is also an unmet need for trained pancreaticobiliary pediatric endoscopists whose familiarity with unique pediatric physiology and disease processes may expand the horizons of this novel area.

4.) **Anshu Srivastava, Malathi Sathiyasekharan, Barath Jagadisan, Rishi Bolia, Maya Peethambaran, Geetha Mammayil, Bhaswati Acharya, Rohan Malik, Srinivas Sankaranarayanan, Vishnu Biradar, Smita Malhotra, Mathew Philip, Ujjal Poddar, Surender Kumar Yachha.** ***Paediatric inflammatory bowel disease in India: a prospective multicentre study.*** **Eur J Gastroenterol Hepatol. 2020. August 10. doi: 10.1097/MEG.0000000000001859** [Epub ahead of print]

This multicentre study prospectively evaluated the demographics, clinical phenotype and outcome of pediatric inflammatory bowel disease (PIBD) from India. Data of children (≤18 years) with PIBD were collected using a proforma containing details of demographics, clinical profile, extraintestinal manifestations (EIM), investigations, disease extent and treatment and analyzed. Three hundred twenty-five children (305) [Crohn's disease: 65.2%, ulcerative colitis:28.0%, IBD unclassified (IBDU): 6.7%, median age at diagnosis: 11 (interquartile range 6.3) years] were enrolled. 6.9% children had family history of IBD. Pancolitis (E4) was predominant in ulcerative colitis (57.8%) and ileocolonic (L3, 55.7%) in Crohn's disease. Perianal disease was present in 10.9% and growth failure in 20.9% of Crohn's disease cases. Steroids were the initial therapy in 84.2%, 5-amino salicylic acid in 67.3% and exclusive enteral nutrition (EEN) in 1.3% cases. Overall, immunomodulators and biologics were given to 84.3 and 17.9% cases, respectively, and 2.9% cases underwent surgery. Very early onset IBD (VEOIBD) was seen in 60 (19.2%) children. IBDU was commoner in the VEOIBD than the older-PIBD (18/60 vs 4/253; $P < 0.001$). VEOIBD-Crohn's disease patients more often had isolated colonic disease than the older Crohn's disease (45.4% vs 11.8%;$P < 0.001$). Prevalence of perianal disease, EIM, therapeutic requirements and outcome were not different between VEOIBD and older-PIBD. Authors have concluded that disease location and phenotype of PIBD in Indian children were similar to the children from the west. VEOIBD accounted for 19.2% of PIBD. The therapeutic options of EEN, biologics and surgery were underutilized.

5.) ***John Matthai, Malathi Sathiasekharan, Ujjal Poddar, Anupam Sibal, Anshu Srivastava, Yogesh Waikar, Rohan Malik, Gautam Ray, S Geetha and SK Yachha for the Indian society of Pediatric Gastroenterology, Hepatology and Nutrition; Pediatric Gastroenterology chapter of Indian Academy of Pediatrics. Guidelines on Diagnosis and Management of Cow's Milk Protein Allergy.*** **Indian Pediatrics.2020 August 15; 57; 533-535.**

This is the recommendations on Cow's milk protein allergy (CMPA) by a group of experts on behalf of the ISPGHAN and Pediatric Gastroenterology Chapter of Indian Academy of Pediatrics which are summarized below.

CMPA is most common in the first year of life. Gastrointestinal manifestations are usually non-IgE mediated and therefore skin prick test and specific IgE levels are not useful in diagnosis. Clinical response to elimination diet followed by a positive oral food challenge is diagnostic. In patients with only gastrointestinal manifestations, sigmoidoscopy and rectal biopsy may be considered as an alternative. Management involves strict avoidance of all forms of bovine milk protein. For infants who are artificially fed, an extensively hydrolyzed formula is the first choice. Soy formula is an alternative in those above six months of age. Since most infants outgrow the allergy, elimination diet is only for a limited period and re-evaluation should be done periodically.

6.) ***Rishi Bolia, Raksha Ranjan and Nowneet Kumar Bhat. Recognising the Gastrointestinal Manifestation of Pediatric Coronavirus Disease 2019. The Indian Journal of Pediatrics.2020 August 21***. doi: 10.1007/s12098-020-03481-y [Epub ahead of print]

In this scientific letter, the authors have reported 2 children diagnosed with COVID 19 who had gastrointestinal symptoms viz. nausea and non-bilious vomiting as the presenting features in the absence of respiratory symptoms. The authors have concluded by stating that the spectrum of GI manifestations of pediatric COVID-19 may vary from mild non-specific symptoms as seen in their patients to severe symptoms mimicking a "surgical" abdomen, which may occur even in the absence of respiratory symptoms. Further, it is important for pediatricians to be aware of these clinical presentations and maintain a high index of suspicion for COVID-19, especially in those who have been exposed to a COVID-19 patient.

SEPTEMBER 2020

1.) ***Arghya Samanta, Anshu Srivastava, Sonali Verma, Moinak Sen Sarma and Ujjal Poddar. White Serum – A Clue to Diagnosis of Acute Recurrent Pancreatitis. The Indian Journal of Pediatrics.2020 September 4.*** doi: 10.1007/s12098-020-03488-5 [Epub ahead of print]

Severe hypertriglyceridemia [serum triglyceride (TG) >1000 mg/dl] is a rare cause of acute recurrent pancreatitis (ARP).

Authors have reported a case of ARP in a 4 year old girl caused who had white serum with markedly elevated TG (2451 mg/dl) and total cholesterol of 343 mg/dl. Genetic testing by exome sequencing confirmed homozygous c.644G > A (p.Gly215Glu) LPL (lysosomal protein lipase) pathogenic variant. She was successfully treated with insulin infusion during the acute phase with disappearance of pain within 24 h and reduction of TG (885 mg/dl at 24 h, 354 mg/dl at 96 h). She was subsequently given omega-3 fatty acid, fenofibrate and niacin along with low-fat diet. At 4 months follow- up, serum TG was maintained below 400 mg/dl, without recurrence of pancreatitis. Authors conclude that LPL deficiency should be suspected in pancreatitis patients with white colored serum.

2.) ***Moinak Sen Sarma, Anshu Srivastava, Surender Kumar Yachha and Ujjal Poddar. Ascites in children with extrahepatic portal venous obstruction: Etiology, risk factors and outcome. Digestive Liver Disease.2020 September 6.*** doi: 10.1016/j.dld.2020.08.018 [Epub ahead of print]

Ascites in extrahepatic portal venous obstruction (EHPVO) is uncommon. The authors have studied the etiology and natural history of ascites and risk factors of post-bleeding ascites in children with EHPVO. All admitted EHPVO patients with clinically detectable ascites and/or gastrointestinal (GI) bleeding in previous 6 weeks were analysed. Subjects with ascites were classified as post-bleeding ascites and de-novo ascites (no GI bleeding) Post-bleeding ascites group was compared with controls (GI bleeding without ascites) for risk factors of developing ascites. Of the total 307 analysed EHPVO patients, 26% (n=79) had ascites. Majority (n=66, 83%) were post-bleeding ascites and 17% (n=13) had de-novo ascites due to secondary causes. Risk factors of ascites in post-bleeding ascites (n=56) versus controls (n=188) were younger age of disease onset, lower height z-scores and greater reduction in serum protein, albumin and hemoglobin from baseline non-bled state. 32%, 39%, and 29% of patients with post-bleeding ascites had ascites resolution with salt restriction, additional diuretics and large volume paracentesis respectively. They have concluded that majority of ascites in EHPVO children is after GI bleeding where early age of disease onset, large volume of blood loss and poor nutritional status are risk factors. Overall outcome of EHPVO with ascites was favourable.

3.) ***Aathira Ravindranath, Gautham Pai, Moinak Sen Sarma, Anshu Srivastava et al. Gastrointestinal Basidiobolomycosis: A Mimic of Lymphoma. Journal of Pediatrics. 2020 September 7.*** doi: 10.1016/j.jpeds.2020.09.009 [Epub ahead of print]

Gastrointestinal basidiobolomycosis is rare and may occur in individuals who are immunocompromised. Presentation may mimic other diseases, such as inflammatory bowel disease and malignancies.

In this case report on a 7 year old boy who presented with fever, abdominal pain, constipation, and abdominal distension of 15 days' duration. He had pallor, no peripheral lymphadenopathy, and a firm mass in the periumbilical and left lumbar areas. Laboratory investigation revealed anemia (haemoglobin 7.8 g/dL) and eosinophilia (absolute eosinophil count 3300). A computed tomography scan revealed a heterogeneously enhancing conglomerate necrotic mass encasing the jejunum, terminal ileum, ascending colon, and transverse colon. Colonoscopy identified an ulceroproliferative growth in the ascending colon and cecum with luminal narrowing. An ultrasonography-guided percutaneous biopsy specimen obtained from the mass and endoscopic mucosal biopsy specimens showed granulation tissue and eosinophilia. The patient subsequently developed acute intestinal obstruction that mandated laparotomy. The mass was unresectable on laparotomy because it involved the distal half of the small bowel and 80%of the large bowel. Thus, a venting ileostomy was performed. Surgical biopsy specimens showed dense eosinophils, giant cells, and nonseptate fungal elements. A fungal culture grew *Basidiobolus ranarum* after 4 days of incubation. An immunodeficiency workup was negative. The child had history of pica, which could have contributed to infection of the gastrointestinal tract by the fungus. He received intravenous voriconazole (4 mg/kg/day) for 6 months, followed by oral voriconazole for 12 months. Fever resolved in 2 days, and eosinophilia resolved in 2 months. The mass resolved at 15 months, after which closure of ileostomy was performed. At a 2-year followup, the child was asymptomatic, with normal abdominal imaging.

4.) **Vikrant Sood, James E Squires, George V Mazariegos, Jerry Vockley, Patrick J McKiernan Living related Liver Transplantation for Metabolic Liver Diseases in Children. J Pediatr Gastroenterol Nutr 2020 Sep 22.** doi: 10.1097/MPG.0000000000002952 [Epub ahead of print]

Metabolic liver diseases (MLDs) are a heterogenous group of inherited conditions for which liver transplantation can provide definitive treatment. The limited availability of deceased donor organs means some who could benefit from transplant do not have this option. Living related liver transplant (LrLT) using relatives as donors has emerged as one solution to this problem. This technique is established worldwide, especially in Asian countries, with shorter waiting times and patient and graft survival rates equivalent to deceased donor liver transplantation. However, living donors are underutilized

for MLDs in many western countries, possibly due to the fear of limited efficacy using heterozygous donors. Authors have reviewed the published literature and shown that the use of heterozygous donors for LT is safe for the majority of MLDs with excellent metabolic correction. They have concluded the use of LrLT should be encouraged to complement DDLT for treatment of MLDs in this invited review.

Publications inadvertently missed in previous issues
February 2020

1.) ***Shivani Deswal, Kumble Seetharama Madhusudhan, Sanjay Sharma and Rohan Malik. Balloon-occluded Retrograde Transvenous Obliteration (BRTO): A Treatment Option in Children with Gastric Varices. The Indian Journal of Pediatrics.2020 February 26***. doi: 10.1007/s12098-020-03265-4 [Epub ahead of print]

BRTO (ballon-occluded retrograde transvenous obliteration) involves occlusion of the outflow veins of the portosystemic shunt by a balloon and endovascular administration of a sclerosant to directly obliterate the gastric varices.

In this scientific letter, the authors have reported the utility of BRTO as a successful treatment option for gastric varices in a 7 year old boy with portal hypertension due to Caroli's syndrome with congenital hepatic fibrosis.

APRIL 2020

1.) **Anil C. Anand, Bhaskar Nandi, Subrat K. Acharya, Anil Arora, Sethu Babu, Yogesh Batra, Yogesh K. Chawla, Abhijit Chowdhury, Ashok Chaoudhuri, Eapen C. Eapen, Harshad Devarbhavi, RadhaKrishan Dhiman, Siddhartha Datta Gupta, Ajay Duseja, Dinesh Jothimani, Dharmesh Kapoor, Premashish Kar, Mohamad S. Khuroo, Ashish Kumar, Kaushal Madan, Bipadabhanjan Mallick, Rakhi Maiwall, Neelam Mohan, Aabha Nagral et al. The INASL Task-Force on Acute Liver Failure. Indian National Association for the Study of the Liver *Consensus Statement on Acute Liver Failure (Part 2): Management of Acute Liver Failure.* Journal of Clinical and Experimental Hepatology.2020 April 12. doi.org/** 10.1016/j.jceh.2020.04.011 [Epub ahead of print]

A roundtable discussion of the INASL task force on ALF which included experts from Pediatric Gastroenterology and Hepatology was held on 6th and 7th July 2019 to discuss, debate, and finalize the consensus statements on on management of ALF in India.

Acute liver failure (ALF) is not an uncommon complication of a common disease such as acute hepatitis. Viral hepatitis followed by antituberculosis drug–induced hepatotoxicity are the commonest causes of ALF in India. Clinically, such patients present with appearance of jaundice, encephalopathy, and coagulopathy. Hepatic encephalopathy (HE) and cerebral edema are central and most important clinical event in the course of ALF, followed by superadded infections, and determine the outcome in these patients. The pathogenesis of encephalopathy and cerebral edema in ALF is unique and multifactorial. Ammonia plays a crucial role in the pathogenesis, and several therapies aim to correct this abnormality. The role of newer ammonia-lowering agents is still evolving. These patients are best managed at a tertiary care hospital with facility for liver transplantation (LT). Aggressive intensive medical management has been documented to salvage a substantial proportion of patients. In those with poor prognostic factors, LT is the only effective therapy that has been shown to improve survival. However, recognizing suitable patients with poor prognosis has remained a challenge. Close monitoring, early identification and treatment of complications, and couseling for transplant form the first-line approach to manage such patients. Recent research shows that use of dynamic prognostic models is better for selecting patients undergoing liver transplantation and timely transplant can save life of patients with ALF with poor prognostic factors.

MAY 2020

1.) ***Sahana Shankar and Jeremy Rosenbaum. Chronic diarrhoea in children: A practical algorithm-based approach. Journal of Paediatrics and Child Health 2020 May 10.*** doi: 10.1111/jpc.14986 [Epub ahead of print]

In this review article the authors have exhaustively described an approach to evaluation of chronic diarrhoea in children based on its pathophysiologic mechanisms with focus on aetiology, investigation and management. It includes a brief description of normal fluid homeostasis in the gut and pathophysiology of diarrhoea. Relevant history, physical examination findings, first and second-line investigations which help in differentiating the different types of diarrhoea have been enlisted and an algorithmic approach to individual types of diarrhoea has been presented. Principles of management and recent advances in diagnostics and therapeutics of diarrhoea are briefly discussed.

2.) ***Charu Singh, Bhawna Sharma, Aradhana Aneja, Sadhna B. Lal, and Sumeeta Khurana. Coinfection with Hymenolepis nana and Hymenolepis diminuta infection in a child from North India: A rare case report.2020 May 20.*** doi: 10.4103/tp.TP_47_19: 10.4103/tp.TP_47_19 [Epub ahead of print]

Hymenolepiasis is considered the most common tapeworm infection throughout the world infecting 50–75 million people. *Hymenolepis diminuta* infection is not commonly reported in human beings as compared to *Hymenolepis nana* because it is primarily a parasite of rats and mice.

In case report authors describe coinfection of *H. nana* and *H. diminuta* diagnosed by stool microscopy (saline and iodine mounts of unconcentrated and concentrated stools) in a 4-year-old boy from a semirural area of India who presented with acute and severe colitis. The child was given a single dose of praziquantel (20 mg/kg), and a second parasitological examination of stool was carried out 7 days following the treatment which was negative, and the child became asymptomatic. The stool was examined fortnightly for 1 month which yielded the negative results.

JUNE 2020

1.) ***Veena Raghunathan, Neelam Mohan. Maninder Dhaliwal, Prashant Bhangui, Vijay Vohra, Arvinder Singh Soin. Pediatric liver transplantation in severe hepatopulmonary syndrome and use of inhaled nitric oxide for post-transplant hypoxemia–a single center experiencePediatric Transplantation. 2020 June 22.*** doi: 10.1007/s12098-020-03265-4 [Epub ahead of print]

There are limited data on liver transplantation in severe hepatopulmonary syndrome (HPS) and outcomes in pediatric population.

In this retrospective study (2011 – 2017), the authors have studied the spectrum and outcomes of pediatric patients with HPS undergoing living donor liver transplantation (LDLT) and the role of inhaled nitric oxide (iNO) for post-LDLT refractory hypoxemia. 23/150 pediatric patients who underwent LDLT during this period had HPS. Refractory post-operative hypoxemia was treated with iNO by institutionally developed protocol. Biliary atresia was the most common underlying cause (52.2%). By oxygenation criteria, 6 (26.1%) had very severe (VS)-HPS. VS-HPS was associated with longer length of ICU stay (LOS) (p = .031) and prolonged oxygen requirement (p = .001) compared with other HPS patients. 4/6 patients with VS-HPS had pO2 < 45 mm Hg. Among these, 2 developed ICH post-operatively and 1 died. 3 developed refractory post-operative hypoxemia, successfully treated with iNO. Mean duration of iNO was 26.3 days. In the group of patients with HPS, the incidence of hepatic artery thrombosis (HAT) and portal vein thrombosis was 17.3% and 4.3%, respectively. One year post-LDLT survival of patients with HPS was similar to non-HPS patients (86.9% vs 94.4%; p = .88). The authors have concluded that, pediatric patients with VS-HPS, especially those with pre-operative pO2 < 45 mm Hg, have long and difficult post-LT course. Refractory postoperative hypoxemia can be successfully overcome with strategic use of iNO.

2.) ***Murali D, Dhua AK, Jain V, Bhatnagar V, Malik R, Yadav R. Giant gastric polyp in Peutz–Jeghers syndrome: Report of a case. J Indian Assoc Pediatr Surg 2020;25:236-8.***

Authors have reported a case of PJS, with an adenomatous giant gastric polyp in a 2-year-old boy who was incidentally diagnosed with anemia during evaluation for an episode of upper respiratory tract infection. There was no history of hematemesis, melena, or abdominal pain. Upper gastrointestinal endoscopy identified a large sessile gastric polyp that was not amenable to endoscopic resection. Physical examination was insignificant except for a few melanotic spots over the lower lip. Radiological imaging revealed a large hyper-enhancing mass lesion in the lumen of the stomach, measuring about 8 cm in greatest dimension along with multiple small intestinal polyps. Radiological imaging revealed a large hyperenhancing mass lesion in the lumen of the stomach, measuring about 8 cm in greatest dimension along with multiple small intestinal polyps. Laparotomy revealed a large sessile polyp involving the greater curvature and posterior wall of the stomach measuring about 10 cm × 8 cm along with multiple small sessile polyps involving the corpus of the stomach sparing the antrum and pylorus, requiring partial gastrectomy. There were two more polyps, one each in the duodenum and distal jejunum in their antimesenteric border and they could be excised by enterotomy. The child had an uneventful postoperative course. The histopathology revealed the features of tubulovillous adenoma in all the resected polyps. At 36 months of follow-up, the child is asymptomatic and disease free.

Authors have highlighted that adenomatous giant gastric polyp may be an extremely rare presentation of PJS through this case report.

Compiled by **Dr. Prasanth.K.S**