Publications by ISPGHAN members
(September – November, 2019)

September

1. *Sood V, Lal BB, Gupta E, Khanna R, Siloliya MK, Alam S. Hepatitis A Virus-related Pediatric Liver Disease Burden and its Significance in the Indian Subcontinent. Indian Pediatr. 2019 Sep 15;56(9):741-744.*

Hospital records of 431 children (age <18 y) diagnosed to be suffering from acute HAV infection during 2011 to 2018 were extracted and analyzed. Additionally, a seroprevalence study was done on 2599 participants (696 children and 1903 adults). The authors concluded that HAV infection is the major contributor the overall pediatric liver disease burden. A significant proportion of subjects remain susceptible to HAV infection even after 10 years of age. Population-based studies are required to further delineate the epidemiology of HAV infection in India for deciding introduction of HAV vaccine in the national immunization schedule

2. *Das MC, Srivastava A, Yadav RK, Yachha SK, Poddar U. Optic nerve sheath diameter in children with acute liver failure: A prospective observational pilot study. Liver Int. 2019 Sep 24. doi: 10.1111/liv.14259. [Epub ahead of print]*

Early detection of raised intracranial pressure (ICP) improves outcome in acute liver failure (ALF). The authors evaluated the feasibility of bedside, ultrasound-guided measurement of optic nerve sheath diameter (ONSD) in normal and ALF children and correlation of ONSD with grade of hepatic encephalopathy (HE), international normalized ratio (INR) and blood ammonia (BA). Forty-one ALF and 47 healthy children (5-18 years) were prospectively enrolled and 12 hourly clinical evaluations was done. The authors concluded that ONSD can be safely and easily measured in ALF children and correlates with HE grade, INR and BA. Normal ONSD in children (>4 years) is <4.5 mm and value of >5.1 mm in ALF requires urgent attention.

3. *Tripathi PR, Poddar U, Yachha SK, Sarma MS, Srivastava A. Efficacy of Single Versus Split Dose Polyethylene Glycol for Colonic Preparation in Children: A Randomized Control Study. J Pediatr Gastroenterol Nutr. 2019 Sep 20. doi: 10.1097/MPG.0000000000002511. [Epub ahead of print]*

Polyethylene glycol (PEG) is the most effective colon cleansing agent but volume related adverse effects are common. Though split-dose PEG is used in adults, no pediatric study so-far has compared split-dose with single-dose PEG. The authors aimed at comparing the efficacy and tolerability of split-dose versus single-dose PEG for bowel preparation in children. In this study consecutive children (1-18 years) were randomized into either single-dose or split-dose PEG. Single-dose group received 4000 mL/1.73mPEG solution day before colonoscopy while split-dose group received half dose day before and the remaining half on the day of colonoscopy. The conclusion drawn was that split-dose PEG is more effective than single-dose regimen for bowel preparation with better tolerability and improved sleep quality in the pediatric population.

4. *Maji P, Malik R, Lodha R, Bagga A. Sickle Cell Intrahepatic Cholestasis with Acute Liver Failure and Acute Kidney Injury: Favourable Outcome with Exchange Transfusion. Indian J Pediatr. 2019 Sep 13. doi: 10.1007/s12098-019-03071-7.[Epub ahead of print]*

Sickle cell disease may present with a life threatening complication of intrahepatic cholestasis. Bilirubin may be very high (> 50mg/dL) in this condition. Exchange transfusion maybe life-saving.

5. *Neupane N, Krishnamurthy S, Jagadisan B, Dhodapkar R. Hepatitis B Seroprotection in Pediatric Nephrotic Syndrome. Indian Pediatr. 2019 Aug 15;56(8):659-662.*

The authors concluded that children with nephrotic syndrome, in general, and steroid-resistant nephrotic syndrome in particular, show poor seroprotection with Hepatitis B vaccination.

6. *Menon J, Kumar A, Vaiphei K, Lal S. An interesting cause of chronic abdominal pain in a child. Trop Doct. 2019 Sep 17:49475519876422. doi:10.1177/0049475519876422. [Epub ahead of print]*

Chronic abdominal pain without red flag signs is usually functional abdominal pain and does not require investigation. The authors encountered an eight-year-old boy who was subsequently diagnosed with a retroperitoneal ganglioneuroma. In view of the rarity of this tumour and its presentation, the authors were prompted to report this case.

7. *Valamparampil JJ, Reddy MS, Shanmugam N, Vij M, Kanagavelu RG, Rela M. Living donor liver transplantation in Alagille syndrome-Single center experience from south Asia. Pediatr Transplant. 2019 Sep 30:e13579. doi: 10.1111/petr.13579.[Epub ahead of print]*

To analyze the clinical characteristics and the outcomes of living donor liver transplantation in children with Alagille syndrome (AGS). Clinical data of children with AGS who underwent liver transplantation between July 2009 and May 2019 in the authors unit were retrospectively analyzed. Primary end-points were patient and graft survival. Ten children with AGS underwent living donor liver transplantation at a median age of 28 months (range, 12-84 months). The most common indication for transplantation was severe pruritus with poor quality of life. They reported 100% patient and graft survival at a mean follow-up of 32 months (range 3-72 months). This is the first series of LDLT for Alagille syndrome in the Indian sub-continent. They report excellent post-transplant outcomes in contrast to outcomes reported from Western literature.

8. *Ravindranath A, Sen Sarma M, Yachha SK. Bile acid synthetic defects: Simplified approach in a nutshell. Hepatobiliary Pancreat Dis Int. 2019 Sep 12.pii: S1499-3872(19)30181-X. doi: 10.10 16/j.hbpd.2019.09.003. [Epub ahead of print]*

In this review the authors present a simplified approach to bile acid synthetic defects.

October

1. *Snehavardhan P, Lal BB, Sood V, Khanna R, Alam S. Efficacy And Safety Of Sodium Benzoate In The Management Of Hyperammonemia in Decompensated Chronic Liver Disease of the Childhood- A Double Blind Randomised Controlled Trial. J Pediatr Gastroenterol Nutr. 2019 Oct 22. doi: 10.1097/MPG.0000000 000002521. [Epub ahead of print]*

It was a prospective, interventional, double-blinded randomized controlled trial conducted from August'2017 to December'2018. The objective was to evaluate the efficacy and safety of sodium benzoate in the management of hyperammonemia and hepatic encephalopathy in decompensated chronic liver disease (CLD). The authors concluded that the addition of sodium benzoate significantly reduced the ammonia levels on the first 2 days of therapy but the effect was not sustained till day 5. The effect of sodium benzoate would probably be more sustained, if higher dosage (400 mg/kg/day) could be used under monitoring of benzoate levels. There was no effect on resolution of HE. Sodium benzoate caused an increasing trend of adverse events with no effect on short-term survival.

2. *Ravindranath A, Sen Sarma M, Yachha SK, Lal R, Singh S, Srivastava A, Poddar U, Neyaz Z, Behari A. Outcome of portosystemic shunt surgery on pre-existing cholangiopathy in children with extrahepatic portal venous obstruction. J Hepatobiliary Pancreat Sci. 2019 Oct 25. doi: 10.1002/jhbp.692. [Epub ahead of print]*

This study was performed to assess the effect of porto-systemic shunt surgery (PSS) on portal cavernoma cholangiopathy (PCC) in children with EHPVO. Children with EHPVO and PCC (unfit for Meso-Rex shunt) underwent Magnetic resonance cholangiogram (MRC) and Magnetic resonance portovenogram (MRPV) before non-selective PSS. Those with patent shunt were re-evaluated at least 6 months after surgery with MRC, MRPV and compared with pre-shunt images. The authors concluded that non-selective PSS decompresses esophago-gastro-splenic venous circuit effectively but fails to ameliorate cholangiopathy and

peribiliary collaterals. Peristence of cholangiopathy is attributable to SMV block.

3. *Ravindranath A, Srivastava A, Yachha SK, Poddar U, Sarma MS, Saraswat VA,Mohindra S, Yadav RR, Kumar S. Childhood pancreatic trauma: Clinical presentation, natural history and outcome. Pancreatology. 2019 Oct 31. pii: S1424-3903(19)30762-8. doi:10.1016/j.pan. 2019.10.008. [Epub ahead of print]*

The objective of this study was to study the presentation, management strategies and long-term natural history of children with pancreatic trauma. Children admitted with pancreatic trauma were analyzed for their presentation, management and outcome. 36 children [29 boys, age 144 (13-194) months] presented at 30 (3-210) days after trauma. Management consisted of various combinations of nasojejunal feeds [n = 17,47.2%], TPN [n = 5,13.8%], octreotide [n = 13,36%], pseudocyst drainage [radiological (n = 18,50%), endoscopic (n = 3,8.3%)] and ERCP [n = 12,33.3%]. Surgical intervention was done in 2 (5.5%) cases. Of the 32 cases in follow-up, 19 (59.3%) recovered and 13 (40.6%) developed CP, with half (6/13) of them being symptomatic with recurrent pain. The authors concluded that multi-disciplinary non-operative management is effective for managing pancreatic trauma in 94.4% of children, with 75% requiring radiological or endoscopic intervention. 40% developed structural changes later but only half were symptomatic.

4. *Karunakaran P, Kochhar R, Lal S, Nampoothiri RV, Varma N, Varma S, Malhotra P. High Prevalence of Celiac Disease in Patients with Immune Thrombocytopenia. Indian J Hematol Blood Transfus. 2019 Oct;35(4):722-725*

Celiac disease (CD) is known to be associated with several autoimmune disorders. The authors studied the prevalence of subclinical CD among patients with immune thrombocytopenia (ITP) as compared to general population. Four patients of primary ITP (4/79) were positive for both serology as compared to 2 (2/316) healthy controls [odds ratio 8.37 (CI 1.50-46.47, $p < 0.005$)]. Among the ITP cases only one had clinical symptoms of CD while none of the healthy controls had symptoms of CD. There is a significantly higher prevalence of subclinical CD in patients with ITP.

5. *Menon J, Shanmugam N, Vij M, Reddy MS, Rela M. Cutaneous Leishmaniasis Presenting As Macrocheilitis In A Post Liver Transplant Pediatric Patient. J Pediatr Gastroenterol Nutr. 2019 Oct 29. doi: 10.1097/MPG.0000000 000002545. [Epub ahead of print]*

A 3-year-old boy, who underwent living donor liver transplantation at 1 year age for biliary atresia presented during his routine follow with painful perioral swelling of two months duration. Considering cellulitis, he was treated with intravenous antibiotics for 5 days, but with little improvement. Kaposi sarcoma was considered in immunosuppressed patient and lip biopsy was done which suggested Leishmaniasis

6. *Ahlawat R, Parikh NS, Jhaveri A. Triple Diagnosis of Crohn's Disease, Celiac Disease, and Eosinophilic Esophagitis in a Child With Siderius-Hamel Syndrome. WMJ. 2019 Oct;118(3):140-142*

The authors present a case of a child with Siderius-Hamel syndrome who had characteristic findings of all these conditions - Crohn's disease, celiac disease, and EoE-an occurrence that to our knowledge has not been reported previously.

November

1. *Shankar S, Bolia R, Foo HW, D'Arcy CE, Hardikar N, Wensing M, Hardikar W. Normal Gamma Glutamyl Transferase Levels at Presentation Predict Poor Outcome in Biliary Atresia. J Pediatr Gastroenterol Nutr. 2019 Nov 14. doi:10.1097/MPG.0000000000002563. [Epub ahead of print]*

Gamma-glutamyl transferase levels (GGT) are typically elevated in biliary atresia (BA), but normal GGT levels have been observed. This cohort of 'normal GGT' BA has not been described nor has the prognostic value of GGT level on outcomes in BA. Infants diagnosed with BA between 1991 - 2017 were

retrospectively analysed. Outcomes were defined as survival with native liver, liver transplantation (LT) and death. Patients were categorised into normal (<200IU/L) or high GGT groups based on a mean of three consecutive GGT values done prior to Kasai portoenterostomy (KPE). Baseline parameters, age at surgery, clearance of jaundice and outcomes were compared between the two groups. The authors concluded that 12.3% of infants with BA had normal GGT levels at diagnosis. Low GGT levels at presentation in biliary atresia was associated with a poorer outcome.

2. *Nabi Z, Ramchandani M, Chavan R, Darisetty S, Kalapala R, Shava U, Tandan M,Kotla R, Reddy DN. Outcome of peroral endoscopic myotomy in children with achalasia. Surg Endosc. 2019 Nov;33(11):3656-3664*

Achalasia cardia is rare in children and optimum endoscopic management options are not well known. Peroral endoscopic myotomy (POEM) is a novel treatment modality for achalasia with excellent results in adult patients. The long-term outcomes of POEM are not well known in children. In this study, the authors evaluated the outcome of POEM in children with idiopathic achalasia. A total of 44 children (boys-23, girls-21) with mean age of 14.5 ± 3.41 years (4-18) were diagnosed with achalasia during the study period. POEM was successfully performed in 43 children (technical success-97.72%). Intra-operative adverse events occurred in 11 (25.6%) children including retroperitoneal CO2 (7), capno peritoneum (3), and mucosal injury (1). Clinical success at 1, 2, 3, and 4 years' follow-up was 92.8%, 94.4%, 92.3%, and 83.3%, respectively. Erosive esophagitis was detected in 55% (11/20) children. On 24-h pH study, GER was detected in 53.8% (7/13) children.

3. *Malik I, Bhatia V, Kumar K, Sibal A, Goyal N. Pediatric Hepatic Venous Outflow Tract Obstruction: Experience from a Transplant Center. Indian Pediatr. 2019 Nov 15;56(11):965-967.*

The authors carried out a review of case records of children diagnosed with hepatic venous outflow tract obstruction at their center in last 10 years. Out of 11 cases identified, 6 had variable blocks in the hepatic venous system and 4 had combined hepatic venous and inferior vena cava (IVC) block. One child with paroxysmal nocturnal hemoglobinuria (PNH) had isolated IVC involvement. Angioplasty was attempted in 3 patients; among them 2 had successful outcome. Seven children with advanced liver disease underwent transplantation, which was successful in six. With availability of modalities like interventional radiology and transplantation, the overall prognosis of hepatic venous outflow tract obstruction seems to be good when managed in a well-equipped center.

4. *Deswal S, Dewan V, Ahuja A, Singh S, Tiotia R, Vani Narayani K, Anwar S. Endo-gastric Teratoma - A Rare Cause of Upper GI Bleeding in an Infant! Indian J Pediatr. 2019 Nov 11. doi: 10. 1007/s12098-019-03097-x. [Epub ahead of print]*

The authors report a rare cause of upper gastrointestinal bleeding in an infant - An endo-gastric teratoma.

5. *Chang MH, Fischler B, Blauvelt B, Ciocca M, Dhawan A, Ekong U, Ni YH, Porta G, Sibal A, DAgostino D, Wirth S, Mohan N, Schwarz KB. Survey of Impediments to Prevention of Mother-to-infant Transmission of Hepatitis B Virus by International Societies. J Pediatr Gastroenterol Nutr. 2019 Dec;69(6):648-654*

Mother-to-infant transmission (MIT) is the leading cause of hepatitis B virus (HBV) infections globally. The aim of this international study was to assess the impediments to prevention of (MIT) of HBV. cross-sectional survey was developed by the Federation of the International Societies for Pediatric Gastroenterology, Hepatology and Nutrition(FISPGHAN). The survey was sent to HBV experts of the 5-member societies of FISPGHAN, and 63 of 91 countries/regions responded. Among the participating countries/ regions, 11% did not implement infant HBV immunization programs. The first dose of vaccine was given >24 hours in 36% of the total countries and 100% of African countries. The recommended birth dose was unavailable for outborn neonates in 45% of the total countries, including 92% of African and 50% of Latin American countries/regions. During pregnancy, 44% countries do not screen maternal viral markers, and 46% do not provide third trimester antiviral therapy for highly viremic pregnant mothers.

Publications inadvertently missed in the previous issues –

1. *Meena DK, Akunuri S, Meena P, Bhramer A, Sharma SD, Gupta R. Tissue Transglutaminase Antibody and Its Association with Duodenal Biopsy in Diagnosis of Pediatric Celiac Disease. Pediatr Gastroenterol Hepatol Nutr. 2019 Jul;22(4):350-357*

This study aimed to evaluate a possible association between the anti-tissue transglutaminase antibody (anti-tTG) titer and stage of duodenal mucosal damage and assess a possible cut-off value of anti-tTG at which celiac disease (CD) may be diagnosed in children in conjunction with clinical judgment. The authors concluded that There is an association between the anti-tTG titer and stage of duodenal mucosal injury in children with CD. An anti-tTG value of 115 AU/mL (6.4 times the upper normal limit) had 76% sensitivity, 100% specificity, with a 100% PPV, and 17% NPV for diagnosing CD (95% CI, 0.75-1). This cut-off may be used in combination with clinical judgment to diagnose CD.

2. *Malik I, Kumar K, Hussain H, Bhatia V, Sibal A, Malhotra S. Celiac disease: What the Indian pediatricians know about the disease. Indian J Gastroenterol. 2019 Jun;38(3):263-267*

To ascertain the knowledge, awareness, and practices pertaining to celiac disease (CD) among the Indian pediatricians. A survey link containing a questionnaire was shared through electronic mail using a pediatric database. The survey was kept active for 6 months; all responses received at the end of the survey were analyzed. Two hundred and seventy one pediatricians out of more than 10,000 chose to respond to the survey. Most pediatricians agreed that more patients with CD are being diagnosed than earlier. Most pediatricians opined that clinical manifestations which prompted to a diagnosis of CD were failure to thrive (96.2%) and chronic diarrhea (81.4%). Knowledge about atypical manifestations of celiac disease was low. Though knowledge about the common association of CD with type 1 diabetes (62.1%) and autoimmune hepatitis (55.8%) was there, awareness about its association with other uncommon conditions was lacking. A trial of gluten-free diet (GFD) was thought to be a logical step if serology was positive by 31.3% of respondents. While 87.7% of pediatricians advocated lifelong adherence to GFD, 12.3% felt that GFD could be discontinued in the future. This web-based survey revealed that though pediatricians are seeing increasing number of celiac disease patients, there is a need to increase awareness regarding the disease, its associated conditions, the need for mucosal biopsy to confirm the diagnosis and the necessity of lifelong adherence to GFD.

3. *Nabi Z, Basha J, Lakhtakia S, Shava U, Pal P, Ramchandani M, Gupta R, Kalapala R, Darisetty S, Tandan M, Reddy DN. Disconnected Pancreatic Duct in Children With Walled OFF Necrosis: Impact on Outcomes of Endoscopic Drainage. J Pediatr Gastroenterol Nutr. 2019 Jul;69(1):116-119*

Disconnected pancreatic duct syndrome (DPDS) is frequently encountered in cases with walled off necrosis (WON). The impact of DPDS on the outcomes of pancreatic fluid collections (PFCs) is not well known. In this study, the authors aimed to evaluate the incidence of DPDS and its clinical impact on the outcomes of endoscopic ultrasound (EUS)-guided drainage of PFC in children. The authors concluded that majority of the children with DPDS do not develop a symptomatic recurrence of PFC after the removal of cystogastric stents. DPDS may be a risk factor for the development of new-onset diabetes. However, future prospective studies are needed.

4. *Ravindranath A, Srivastava A, Seetharaman J, Pandey R, Sarma MS, Poddar U,Yachha SK. Peritoneal Lymphomatosis Masquerading as Pyoperitoneum in a Teenage Boy. ACG Case Rep J. 2019 Jun 17;6(6):e00116.*

A 16-year-old boy presented with 1 month of fever, abdominal pain, and distension. The ascitic tap drained pus-like fluid, and ultrasonography showed diffuse thickening of the omentum and mesentery with echogenic ascites. The ascitic fluid appearance deceptively resembled pus, but further analysis revealed atypical lymphocytes. Omental and bone marrow biopsies confirmed Burkitt lymphoma. Awareness of this rare presentation is imperative for making a correct diagnosis.

Compiled by : Dr. Rishi Bolia