Publications by ISPGHAN members
March – May, 2019

March

1. *Janwadkar A, Shirole N, Nagral A et al. Citrullinemia Type 1: Behavioral Improvement with Late Liver Transplantation. Indian J Pediatr. 2019 Mar 8. [Epub ahead of print]*

Citrullinemia Type 1 (also known as classic citrullinemia) is a rare autosomal recessive urea cycle disorder due to reduced activity of argininosuccinate synthetase 1; characterized by hyperammonemia leading to neurological damage. The authors report a case of an 8-y boy who was diagnosed with Citrullinemia Type 1 at birth which was anticipated prenatally due to family history. His diagnosis was confirmed as a homozygous mutation (Exon 15: c.1168G > A (p.G390R)) of ASS gene. Inspite of being on a protein-free diet and ammonia scavenging treatment; the patient developed recurrent episodes of encephalopathy and seizures; complicated with behavioral issues. The patient underwent living related liver-transplantation from his mother (heterozygous carrier of the same mutation) resulting in the reversal of neuro-behavioral changes. It is important to consider liver transplantation in citrullinemia Type 1 as it corrects the genetic deficiency of ASS

2. *Kadyada SP, Thapa BR, Bhatia A et al. Role of Diagnostic Endoscopic Ultrasound in Idiopathic Acute Pancreatitis and Acute Recurrent Pancreatitis in Children. Pancreas. 2019 Mar;48(3):350-355*

Endoscopic ultrasound (EUS) is a minimally invasive pancreatic imaging modality. The authors evaluated children with idiopathic acute pancreatitis (IAP) and acute recurrent pancreatitis (ARP) for changes of chronicity (Rosemont criteria). Diagnostic yield of simultaneously performed transabdominal ultrasonography (TUS) was compared with EUS. In this prospective observational study. Patients underwent EUS and TUS after 2 months of a pancreatitis attack. Forty-five (18 IAP, 27 ARP) patients were evaluated. It was found that applying EUS, changes of chronicity and risk factors were noted only in ARP. Endoscopic ultrasound performed better than TUS in detecting chronicity.

3. *Alam S, Lal BB, Sood V et al. AARC-ACLF score: best predictor of outcome in children and adolescents with decompensated Wilson disease. Hepatol Int. 2019 Mar 19. [Epub ahead of print]*

Doubts have been raised about efficacy of New Wilson's index (NWI) in predicting Liver Transplant (LT) or mortality in decompensated Wilson Disease (WD) patients. APASL ACLF Research Consortium (AARC) has introduced a new score (AARC-ACLF) which has not been studied in children. In this study, sixty-six confirmed cases of decompensated WD were evaluated. Thirty-nine (59%) improved on medical management and 27 (41%) either died (20) or were transplanted (7). AARC-ACLF had the best predictive score for mortality at 90 days with AUROC of 0.939. For every unit increase in AARC-ACLF score, there was a likelihood of 66% increase in 90-day mortality. The optimal cut-off for the AARC-ACLF score to predict mortality was 11 or more.

4. *Pamecha V, Vagadiya A, Sinha PK, et al. Live donor liver transplantation for acute liver failure - Donor safety and recipient outcome. Liver Transpl. 2019 Mar 12.. [Epub ahead of print]*

In countries where deceased organ donation is sparse, emergency live donor liver transplantation (LDLT) is the only lifesaving option in select patients with acute liver failure(ALF). The aim of the current study was to evaluate live liver donor safety and recipient outcomes following

LDLT for ALF. The authors found that outcome of emergency live liver donation was comparable to elective donors. Post-operative worsening of cerebral edema, preoperative SIRS, sepsis predicted outcome after LDLT for ALF

5. *Cherukuru R, Reddy MS, Shanmugam NP et al. Feasibility and Safety of Split-Liver Transplantation in a Nascent Framework of Deceased Donation. Liver Transpl. 2019 Mar;25(3):450-458*

Split-liver transplantation (SLT) is a valuable option for optimizing the use of good-quality deceased donor grafts. It is not routinely reported outside the West because of limited deceased donor numbers, technical and organizational constraints, lack of experience, and a predominant living donor liver transplantation (LDLT) practice. The authors report their experience of SLT and compare outcomes with pediatric and adult LDLT recipients.

They found that SLT is an effective technique with outcomes comparable to living donor grafts for adult and pediatric recipients. Using SLT techniques at centers with limited deceased donors optimizes the use of good-quality whole grafts and reduces the gap between organ demand and availability.

6. *Bhatnagar S, Srivastava G , Ansari A : Bowel Habits of Healthy Indian Children Less Than Two Years of Age. Trop Gastroenterol 2018;39(1):17-21 (In Print in March 2019)*
   DOI : http://dx.doi.org/10.7869/tg.455

Background: The bowel habits of children less than two years are quite varied and there is no definite hard data on stool pattern of Indian children particularly less than two years of age. Aim: To define the normal frequency and consistency of stools of healthy Indian children between 0-24 months of age. Methods: Parents of children aged up to 24 months were asked on a three day recall basis about their child's usual bowel habits and dietary history. Bowel habit was recorded in terms of number of stools the child passes per day, stool consistency, the age at which night bowel movements stopped, and the age of commencement of regular bowel movements. Feeding type was recorded as exclusive breast feed, mixed milk feeds or solid feeds. The bowel habits were correlated with the age and type of feeding. Result: Children in their first six months of life had stools which were predominantly "pasty" and "runny like cream" with high and variable frequency. Beyond six months consistency was "solid" and "pasty" stools. On analyzing the combined effect of the type of milk feed and age on bowel frequency and consistency, children beyond one month of age either on exclusive breast feed or on mixed milk feed had similar frequency and consistency of stools. By one year of age more than 90% children attained regular stool pattern with no night time bowel movements. Conclusion: This is the first report from India which describes the stool pattern of normal healthy children less than two years of age.

April

1. *Bolia R, Rajanayagam J, Hardikar W et al. Impact of Changing Treatment Strategies on Outcomes in Pediatric Ulcerative Colitis. Inflamm Bowel Dis. 2019 Apr 19. [Epub ahead of print]*

In recent years, treatment strategies for ulcerative colitis have evolved with an early step-up approach, the availability of biologicals, and therapeutic drug monitoring. This study was carried out to evaluate the effect of these changes on disease outcomes.

The authors found that a reduction in 2-year colectomy rates has been observed in patients with pediatric ulcerative colitis since biologics have become available for its treatment. However, the numbers of disease-flares rates and hospital admissions remain unchanged.

2. *Prasad D, Srivastava A, Tambe A, et al. Clinical Profile, Response to Therapy, and Outcome of Children with Primary Intestinal Lymphangiectasia. Dig Dis. 2019 Apr 26:1-9. [Epub ahead of print]*

Intestinal lymphangiectasia (IL; primary or secondary) is an important cause of protein-losing enteropathy. The authors evaluated the clinicolaboratory profile, response to therapy, complications, and outcome of children with primary IL (PIL). Twenty-eight children with PIL (16 boys, age at symptom onset-12 [1-192] months and at diagnosis 8 [1-18] years) were studied. Pedal edema (93%), chronic diarrhea (78.6%), and recurrent anasarca (64%) were the common presentations.Presence of chylous ascites suggests severe disease in children with PIL. Majority of PIL children respond to dietary therapy; only 20% need additional therapy. Long-term follow-up is essential to monitor for symptoms relapse and complications.

3. *Jain V, Sangdup T, Malik R et al. Abernethy malformation type 2: varied presentation, management and outcome. J Pediatr Surg. 2019 Apr;54(4):760-765.*

The purpose of this study was to study the varied presentations and the outcomes in children with Type 2 Abernethy malformation following shunt ligation. Five patients were included with a median age of 6 years. Hepatopulmonary syndrome was the presentation in 4 patients while one patient presented with liver tumor. At the median follow up at 14 months, good intrahepatic portal flow was seen in all patients. All patients demonstrated improvement/ resolution of symptoms.

May

1. *Phulware RH, Gahlot GPS, Malik R et al. Microvillous Inclusion Disease as a Cause of Protracted Diarrhea. Indian J Pediatr. 2019 May 2. doi:10.1007/s 12098-019-02963-y. [Epub ahead of print]*

Microvillous inclusion disease (MVID), also known as congenital microvillus atrophy, was first described by Davidson et al. in 1978. Till date, only a handful of cases with MVID have been described in English literature. These patients usually present with intractable secretory diarrhea in early days of life. The pathognomonic findings of MVID are villous atrophy along with the formation of intracellular microvillous inclusions on electron microscopy. Till date, no curative therapy exists.
Herein, authors describe a case of intractable diarrhea with MVID diagnosed in a 3-mo-old male child who presented with intractable diarrhea.

2. *Sood V, Khanna R, Alam S, Rawat D, Bhatnagar S, Rastogi A. Ductal paucity and Warkany syndrome in a patient with congenital extrahepatic portocaval shunt. World J Hepatol. 2014 May 27;6(5):358-62.*

The authors describe an eleven-year-old clinically dysmorphic and developmentally retarded male child presenting with complaints of 5 episodes of recurrent cholestatic jaundice since 3 years of age was evaluated. Imaging revealed features consistent with congenital extrahepatic portocaval shunt (Abernethy type 1b), multiple regenerative liver nodules and intrahepatic biliary radical dilatation. The presence of ductal paucity and trisomy 8 (Warkany syndrome) were identified on liver biopsy and karyotyping. In this article the authors have proposed an explanation for these unusual and previously unreported features.