Publications by ISPGHAN Members in PubMed Indexed Journals

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Annals of Pediatric Gastroenterology and Hepatology ISPGHAN (2023): 10.5005/jp-journals-11009-0118

Source: Mittal R, Kumar K, Malhotra S, Sibal A. Pediatric liver transplantation for autoimmune liver disease: ten-year experience from a liver transplant center in India. Indian J Gastroenterol 2022;41(6):634–642. DOI: 10.1007/s12664-022-01282-z

In this retrospective analysis of prospectively collated data between July 2010 and May 2020, the authors have aimed to describe the single-center experience of living donor liver transplantation (LT) for pediatric autoimmune liver disease (AILD). A total of 13 liver transplants were performed for AILD out of total 244 children transplanted during the study period. The mean (standard deviation) age at LT was 12 (±3.84) years. Indications for LT were decompensated liver disease (61.5%), acute-on-chronic liver failure (23.1%), acute liver failure (ALF) (7.7%), and recurrent cholangitis and growth failure (7.7%). The mean pediatric end-stage liver disease (PELD) score/model for end-stage liver disease score and the international normalized ratio at presentation were 24 (±12.81) and 2.48 (±1.54), respectively. (4/13) of the subjects had no postoperative complications, and three subjects died post-LT. The 1-year and 5-year patient survival rates were 76.9 and 70%, respectively. The authors have concluded that despite higher PELD scores on referral, graft and patient survival rates were encouraging for LT in pediatric AILD.

Source: Poddar U, Aggarwal A, Jayalakshmi K, Sarma MS, Srivastava A, Rawat A, Yachha SK. Higher prevalence of monogenic cause among very early onset inflammatory bowel disease in children: experience from a tertiary care center from northern India. Inflamm Bowel Dis 2023:izac254. DOI: 10.1093/ibd/izac254

In this retrospective analysis of a prospectively collated database of pediatric-onset inflammation between January 2010 and July 2021 from a North Indian tertiary care hospital, the authors have analyzed the data on very early onset inflammatory bowel disease (VEOIBD) and compared the clinical features and outcome of monogenic VEOIBD and non-monogenic VEOIBD. Of 200 children with inflammatory bowel disease (IBD), 48 (24%) were VEOIBD, and 15 (32%) of them had monogenic VEOIBD. The causes of monogenic VEOIBD included disorders of the immune system (including interleukin-10 receptor mutations) in 12 and epithelial barrier dysfunction in three. Clinical features that differentiated monogenic from non-monogenic VEOIBD were neonatal onset, perianal disease, history of consanguinity and sibling death, wasting, stunting, and IBD unclassified phenotype ($p < 0.05$). The authors conclude that in resource constraint countries, using the above clinical features will help to narrow down the phenotype who may likely have monogenic cause for their IBD and benefit most by next-generation sequencing.

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How to cite this article: Sobhan PK. Publications by ISPGHAN Members in PubMed Indexed Journals. Ann Pediatr Gastroenterol Hepatol 2023;5(1):20–22.

Source of support: Nil

Conflict of interest: None

Source: Poddar U, Umesh Reddy DV. Management of Hepatitis C in children - a new paradigm. Indian Pediatr 2023;60(1):55–62.

In this review, the authors have summarized the various treatment options for hepatitis C virus (HCV) infection in children, highlighting the changes in the management in the past decade. They have discussed the various regimens of direct-acting antivirals (DAAs) which have been approved in children above 3 years of age with documented evidence of high efficacy (sustained virologic response-12 of 92–100%) and excellent safety profile compared to treatment with interferon and ribavirin combination. They have concluded that with the advent of DAAs, which are the current standard of care, there has been a paradigm shift in the management of HCV infection in children in the past decade.

Source: Suchismita A, Ashritha A, Sood V, Lal BB, Khanna R, Kumar G, Alam S. Study of adherence to medication in pediatric liver diseases ("SAMPLD" Study) in Indian children. J Clin Exp Hepatol 2023;13(1):22–30. DOI: 10.1016/j.jceh.2022.10.006

In this cross-sectional study, authors have aimed to assess adherence to medications in Indian pediatric liver disease patients (including post-LT recipients) and to identify variables affecting its occurrence. Structured tools using prevalidated questionnaires (Medication adherence measure and the Child and Adolescent Adherence to Medication Questionnaire) were used to collect data related to nonadherence prevalence (based on missed and late doses) and factors influencing adherence. A total of 152 children were included in the study (Wilson Disease 39.5%, AILD 32.9%, and post-LT 27.6%). Authors have concluded that nonadherence was seen in around one-sixth of the patients, with the least nonadherence seen in post-LT recipients (0–2.4%). The older age of the patient, rural place of stay, and personal barriers like hard-to-remember/forgetfulness and bad medication taste were identified as factors independently leading to nonadherence.

Source: Sodhi KS, Maralakunte M, Bhatia A, Lal SB, Saxena AK. Utility of the New Faster Compressed SENSE MRCP at 3 Tesla MRI in Children with Pancreatitis. Indian J Pediatr 2023. DOI: 10.1007/s12098-022-04403-w

In this study, authors have aimed to compare the acquisition time, diagnostic efficacy, and image quality of the newer compressed sensing and sensitivity encoding three-dimensional (3D) magnetic resonance cholangiopancreatography (MRCP) (CS-3D MRCP) with conventional 3D MRCP (C-3D MRCP) in children with pancreatitis. A total of 24 children (2–17 years) diagnosed with pancreatitis were enrolled. The children underwent CS-3D MRCP and C-3D MRCP sequences. A two-fold decrease in the acquisition time of CS-3D MRCP (~148 ± 61 seconds) was seen, compared to C-3D MRCP (~310 ± 98 seconds), $p < 0.001$. The median scores for overall image quality on CS-3D MRCP and C-3D MRCP, respectively, were 2.05 ± 0.52 and 2.21 ± 0.53 ($p = 0.18$). Authors have concluded that the application of CS-3D MRCP in children with pancreatitis results in a two-fold reduction in acquisition time with acceptable image quality. This may help in reducing the need for long sedation in children requiring anesthesia support for the MRCP and potentially help in reducing motion artifacts.

Source: Samanta A, Srivastava A, Mandal K, Sarma MS, Poddar U. MPV17 mutation-related mitochondrial DNA depletion syndrome: a case series in infants. Indian J Gastroenterol 2023. DOI: 10.1007/s12664-022-01281-0.

In this case series, authors have reported on pathogenic mitochondrial protein 17 (MPV17) mutations which can cause mitochondrial deoxyribonucleic acid depletion syndrome, which has myriad presentations with neurological, muscular, and hepatic involvement. They have discussed the clinical presentation and outcome of four infants from four separate families with pathogenic, homozygous MPV17 mutations. All had predominant hepatic involvement with cholestasis, lactic acidosis, and hypoketotic hypoglycemia. Three of them had presented with liver failure. Two of the four cases died in infancy, while the other two improved on conservative management with a medium-chain triglyceride-based diet, vitamin supplements, co-enzyme Q, and carnitine. The two surviving children are alive at 12 and 25 months of age with native liver with normal to mildly deranged liver function and no neurological dysfunction. Authors have proposed mutation testing for the MPV17 gene during the evaluation of indeterminate infantile liver failure, especially those with hypoglycemia and raised plasma lactate.

Source: Sahoo B, Kumar K, Malhotra S, Sibal A. Enteric fever masquerading as Crohn's disease in a child with abdominal tuberculosis. BMJ Case Rep 2023;16(2):e249531. DOI: 10.1136/bcr-2022-249531

Through this illustrative case report of an adolescent boy who had a long history of abdominal pain, bleeding per rectum, documented weight loss, and acute onset fever [which was masquerading as inflammatory bowel disease (Crohn's disease)], authors have described the challenges of diagnosing *Mycobacterium tuberculosis* with a superadded *Salmonella typhi* infection in tropical countries like India.

Source: Dogra S, Kumar K, Malhotra S, Jerath N, Sibal A. Acute Liver Failure in Dengue: A common but overlooked entity in pediatric patients in tropical countries. J Pediatr Gastroenterol Nutr 2023;76(2):149–153. DOI: 10.1097/MPG.0000000000003646

In this retrospective, observational study from a North Indian tertiary care center, authors have aimed to study the incidence of ALF in dengue infection over a period of 5 years between 2016 and 2021, understand the demographic and biochemical profiles, and identify prognostic factors associated with mortality. A total of 30 children with dengue infection were identified to have developed a during the ALF study period, which was 29.1% (30 of 103) of ALF admissions. A total of 189 children with dengue infection needed admission during the same period, and 15.8% (30 of 189) of them developed ALF. The mean duration of onset of ALF was 5.4 days after fever onset. A total of 22 patients (73%) survived, and eight patients expired. High creatinine, low albumin level, and multisystemic involvement were identified as poor prognostic markers in those patients who died. Authors have concluded that dengue should be considered a common cause of pediatric ALF in tropical countries, and ALF is common in severe dengue patients.

Source: Jebaying Y, Kumar K, Malhotra S, Sibal A. Novel mutation in the HSD3B7 gene causes bile acid synthetic disorder and presents as recurrent liver failure in early childhood. BMJ Case Rep. 2023 Feb 7; 16(2):e245852. DOI: 10.1136/bcr-2021-245852

Through this illustrative case report, the authors have described a young child who presented with recurrent episodes of ALF. In the first episode, both coagulopathy and encephalopathy improved on supportive treatment, but the aetiological evaluation was inconclusive. During the second presentation, whole-exome sequencing was sent, identifying a compound heterozygous novel mutation in the 3-β-hydroxysteroid dehydrogenase type 7 gene leading to bile acid synthetic defect.

Source: Acharyya BC, Mukhopadhyay M, Chakraborty H. Changing trend in the spectrum of upper gastrointestinal bleeding in children-a multicentre experience. Indian J Gastroenterol 2023. DOI: 10.1007/s12664-022-01306-8

In this retrospective analysis of prospectively collated data from May 2011 to May 2018, authors from two centers in Eastern India have aimed to study whether there is any change in the trend in the etiology and outcome of pediatric upper gastrointestinal bleed (UGIB). A total of 180 children were evaluated, including 30 (16.7%) infants. The predominant cause of UGIB was a gastroduodenal ulcer and erosions (60%), followed by variceal bleeding (19.4%). Vascular lesions were detected in four (2%). The hyperplastic antral polyp was an unusual etiology in three (1.7%) infants. Various endotherapies were needed in 28% of cases. No mortality was noted. Melena, hemoglobin below 8 gm%, the need for volume replacement, and packed red blood cell transfusion on admission were associated with significant endoscopic lesions, which needed endotherapies. Authors have concluded that this study has found that there is a change in the etiology of UGIB in children with a negative endoscopic yield of just 4% compared to the previous studies that identified variceal bleeding as the major aetiology.

Source: Dhanasekhar Kesavelu and Pramod Jog. Current understanding of antibiotic-associated dysbiosis and approaches for its management. Ther Adv Infect Dis 2023;10:1–18 DOI: 10.1177/ 20499361231154443

In this review, authors have discussed the pathologic role of increased exposure to antibiotics during early childhood as a risk factor for antibiotic-associated dysbiosis, which is associated with reduced diversity of gut microbial species and abundance

of certain taxa, disruption of host immunity, and the emergence of antibiotic-resistant microbes. The disruption of gut microbiota and host immunity in early life is linked to the development of immune-related and metabolic disorders later in life, which calls for judicious and rational use of antibiotics among neonates and young children to prevent the detrimental effects on gut health.

Source: Sharma S, Sinha A, Malik R, Bagga A. gastrostomy tube feeding in Indian children with advanced chronic kidney disease. Indian J Pediatr 2023;90(4):400–402. DOI: 10.1007/s12098-023-04499-8

In this retrospective report, authors have reviewed the impact of gastronomy tube (G-tube) feeding on nutritional intakes and anthropometric parameters over a 1-year follow-up in five children with chronic kidney disease stage five-dimensional managed at one tertiary care center in Northern India. Authors have observed that gastrostomy feeding facilitated significant increments in caloric and protein intake and was easy and safe. However, G-tube feeding led to additional expenses, and the changes in growth parameters were variable in the short term.

Source: Venuthurimilli AK, Gupta R, Singhal S, Madaan V, Kumar P, Singh A, Sah R, Rastogi H, Vohra S, Sahni R, Bharadwaj R, Kumar K, Malhotra S, Jerat N, Sibal A, Goyal N. Intraoperative portal vein stenting through umbilical vein approach: an innovative salvage procedure for portal vein thrombosis in pediatric liver transplant. Pediatr Transplant 2023;27(1):e14427. DOI: 10.1111/petr.14427

Authors have retrospectively analyzed their single experience of opened umbilical vein approach to place the stent in the portal vein intraoperatively (IPVS) and deliver anticoagulation through a catheter by retrieving the prospectively collated database of 150 pediatric transplantation between January 2017 to February 2022 age, weight, PELD score, diagnosis, portal vein diameter on preoperative computed tomography (CT), portal flow after stenting, and decrease in spleen size after stenting in follow-up CT were extracted from the database and reviewed. Eight patients underwent IPVS following living donor liver transplantation (mean age 10.6 ± 2.2 months, mean weight 8.1 ± 1.6, mean PELD score 32.7 ± 7.3). The mean PV diameter on preoperative CT scan was 3.6 mm (range 2.7–5.6 mm). The mean portal flow following stenting was 718.75 cc/minute. The percentage reduction in the size of the spleen was 26.35% beyond 2nd postoperative week. No patient had recurrent PV thrombosis following IPVS, and all maintained an adequate portal flow throughout the immediate postoperative period. Two patients had in-hospital mortality secondary to septic complications. Authors have concluded that the umbilical vein approach is technically feasible, easy to manipulate the stent, and catheter placement after stenting helps to deliver anticoagulants locally.