Publications by ISPGHAN Members in PubMed-indexed Journals (1st September–30th November, 2022)

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Annals of Pediatric Gastroenterology and Hepatology ISPGHAN (2022): 10.5005/jp-journals-11009-0116

Source: Anushree N, Lal SB, Rana SS, Saxena A, Venkatesh V, Sharma AK, Dayal D, Verma S. Morphological and functional recovery following acute and acute recurrent pancreatitis in children: a prospective sequential 2-point evaluation. Pancreatology 2022;22(6):698–705. DOI: 10.1016/j.pan.2022.06.008

In this study, authors have aimed to characterize the morphological and functional changes in the pancreas following acute pancreatitis (AP) and acute recurrent pancreatitis in children. They enrolled 73 children with AP who were followed prospectively and assessed at two time points, at least 3-month intervals, with the initial assessment at least 3 months after the AP episode. Exocrine and endocrine functions were measured using fecal elastase and fasting blood sugar/hemoglobin A1c levels, respectively. The morphological assessment was done using endoscopic ultrasound (EUS) and magnetic resonance imaging and magnetic resonance cholangiopancreatography (MRCP). They have concluded that more than one-quarter of children have evidence of altered glucose homeostasis and biochemical exocrine pancreatic insufficiency following an episode of AP. Similarly, morphological features of chronicity (identified in EUS and MRCP) seen in some of the children suggest that a fraction of subjects may develop chronic pancreatitis on longer follow-up.

Source: Amatya P, Kapalavai SK, Deep A, Sankaranarayanan S, Krupanandan R, Sadasivam K, Ramachandran B. Pediatric acute liver failure: an experience of a pediatric intensive care unit from resource limited settings. Front Pediatr 2022;10:956699. DOI: 10.3389/fped.2022.956699

In this retrospective study done at a single tertiary care children's hospital in South India from January 2014 to December 2019, the authors have aimed to identify the etiologies, outcome, and prognostic factors of acute liver failure and the validity of the existing liver transplantation (LT) criteria [King's College Hospital criteria (KCHC) vs International normalized ratio (INR) >4 criteria] to predict the outcome of pediatric acute liver failure (PALF) by enrolling 125 children aged 1 month–18 years. The main etiologies were infections (32%) (dengue was the commonest infection), indeterminate (23%), paracetamol toxicity (21%), metabolic (13%), and others (11%). Of 125 patients, 63.2% ($n = 79$) had spontaneous regeneration which was higher in paracetamol-induced (92.3%) compared to nonparacetamol-induced acute liver failure (55.5%). Of 38 children meeting KCHC for LT, 57.9% had spontaneous regeneration and 36.8% died. Of 74 children meeting INR >4 criteria, 54% ($n = 40$) had spontaneous regeneration and 43.2% died. They have concluded that infections are the commonest cause of PALF, with dengue fever being the predominant infectious cause, in our population. Children with paracetamol-induced ALF had a better chance of spontaneous regeneration. Patients with high peak alanine aminotransferase, those requiring inotropes, and advanced hepatic encephalopathy (HE) with a high likelihood of death could be potentially considered for LT. The KCHC or INR >4 criteria alone may not be enough to predict outcomes in PALF with different etiologies.

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How to cite this article: Sobhan PK. Publications by ISPGHAN Members in PubMed-indexed Journals (1st September–30th November, 2022). Ann Pediatr Gastroenterol Hepatol 2022;4(4)78–80.

Source of support: Nil

Conflict of interest: None

Source: Poddar U, Samanta A. Foreign body ingestion in children: the menace continues. Indian Pediatr 2022;59(9):716–717. DOI: 10.1007/s13312-022-2601-7

Through this communication, authors have discussed the changes and challenges in the management of foreign body ingestion in children which evolved over the past 5 decades. They have compared the pattern of foreign body ingestion half a century ago (metallic coin ingestion) with the current scenario (button battery ingestion) by rightly pointing out this continuing menace. Advances in endoscopic equipment, accessories, and technique in the last 50 years have also been discussed.

Source: Menon J, Thapa BR, Kumari R, Puttaiah Kadyada S, Rana S, Lal SB. Efficacy of oral psyllium in pediatric irritable bowel syndrome: a double-blind randomized control trial. J Pediatr Gastroenterol Nutr 2023; 76(1):14–19. DOI: 10.1097/MPG.0000000000003622

In this double-blind randomized controlled trial, authors have aimed to evaluate the efficacy of psyllium husk as compared to placebo in pediatric irritable bowel syndrome (IBS) patients. A total of 43 children were assigned to the psyllium arm (group I) and 38 to the placebo arm (group II). Severity was assessed at baseline and after 4 weeks of treatment using the IBS severity scoring scale (IBS-SSS) and classified into mild, moderate, and severe categories. At baseline, type, severity, and parameters (IBS-SSS) of IBS were equally distributed in two groups. There was a significant reduction in the median (interquartile range) of total IBS-SSS in psyllium vs

placebo at 4 weeks. Similarly, 43.9% in group I vs 9.7% in group II attained remission. Authors have concluded that psyllium husk is effective for the therapy of pediatric IBS when compared with a placebo in the short-term.

Source: Semwal P, Bolia R, Rajvanshi N, Phulware RH, Bhat NK. Gastric outlet obstruction due to cytomegalovirus infection in an immunocompetent child. Indian J Gastroenterol 2022. DOI: 10.1007/s12664-022-01243-6

Through this case report published as an image, authors have reported the case of a 4-year-old immunocompetent boy with recurrent nonbilious vomiting and abdominal pain, who was diagnosed to have a solitary deep ulcer due to cytomegalovirus (CMV) infection with a clean base at the prepyloric location causing gastric outlet obstruction. He responded to antiviral therapy and graded endoscopic balloon dilation. They have also highlighted that the depth of the biopsy is an important consideration since CMV is more commonly found at the ulcer base for clinching histopathological diagnosis through this illustrative case report.

Source: Prasad BS, Yachha SK. Congenital glucose-galactose malabsorption in a child. Indian Pediatr 2022;59(10):811–812. DOI: 10.1007/s13312-022-2627-x

In this clinical case letter, authors have reported on the case of a novel mutation of solute carrier family 5 members 1 gene in a 6-month-old infant with congenital glucose-galactose malabsorption (cGGM) for the first time from India. He had presented with recurrent episodes of explosive watery diarrhea with perianal excoriation from 1st week of life and had failure to thrive with features of malabsorption. His loose stools would worsen with oral rehydration solutions and he had evidence of medullary nephrocalcinosis and distal renal tubular acidosis for clinically suspecting the possibility of cGGM. He improved symptomatically and responded to a carbohydrate-free formula supplemented with fructose powder with significant weight gain at 21 months of follow-up.

Source: Sen Sarma M, Tripathi PR. Natural history and management of liver dysfunction in lysosomal storage disorders. World J Hepatol 2022;14(10):1844–1861. DOI: 10.4254/wjh.v14.i10.1844

In this mini-review, authors have discussed the major natural history and management of lysosomal storage diseases (LSDs) that cause liver dysfunction [Gaucher disease (GD) and Niemann–Pick disease] and lysosomal acid lipase deficiency [cholesteryl ester storage disease and Wolman disease (WD)]. Unexplained organomegaly, portal hypertension, and fatty liver are important presentations. Neonatal cholestasis and ascites are rare presentations in infants. Those presenting with neonatal or early-onset of liver disease have a universally poor prognosis. The diagnosis of all LSD is based on enzymatic activity, tissue histology, and genetic testing. Enzyme replacement is possible in GD and Niemann–Pick types A and B though there are major limitations in the outcome. Those that progress invariably require LT with variable outcomes. The prognosis of Niemann–Pick type C and WD is universally poor. Enzyme replacement therapy has a promising role in cholesteryl ester storage disease.

Source: Sivasankaran M, Ramesh V, Sankaranarayanan S, Munirathnam D. Gastrointestinal manifestations in children with primary immune deficiencies: a case series. Indian J Gastroenterol 2022;41(5):513–518. DOI: 10.1007/s12664-022-01273-0

The authors have presented a series of five children who presented predominantly with gastrointestinal (GI) manifestations of primary immune deficiency (PID), not attributable to infections. Noninfectious GI manifestations such as allergic, autoimmune, and inflammatory disorders can be the predominant manifestations of PIDs. Next-generation sequencing led to the underlying genetic diagnosis. Early diagnosis and hematopoietic stem cell transplantation could be life-saving in these children.

Source: Ashritha A, Gautam V, Lal BB, Mukund A, Vijay P, Khanna R, Sood V, Alam S. Percutaneous cholecystocholangiography-a tool to conclusively exclude biliary atresia. Indian J Pediatr 2022; 89(11):1144–1147. DOI: 10.1007/s12098-022-04354-2

In this retrospective chart review authors have aimed to evaluate the feasibility, safety, and diagnostic accuracy of percutaneous cholecystocholangiography (PCC) in cases of conjugated hyperbilirubinemia in which biliary atresia (BA) could not be diagnosed or ruled out based on clinical, radiological, and histopathological findings. PCC was performed *via* the transhepatic route using a 23-g needle. The patency of both the proximal and distal biliary trees was assessed. PCC was technically feasible in 12/13 (92.3%) of infants without any procedure-related complications. PCC demonstrated proximal and distal biliary patency in 7/12 (58.3%) infants, thereby avoiding unnecessary laparotomy in them. PCC failed to demonstrate biliary patency in five infants; of which, four were confirmed as cases of BA on laparotomy. Authors have concluded that PCC can correctly differentiate BA from non-BA cases of conjugated hyperbilirubinemia preoperatively, reducing the negative laparotomy rates.

Source: Menon J, Shanmugam N, Valamparampil J, Vij M, Kumar V, Munirathnam D, Hakeem A, Rammohan A, Rela M. Outcomes of liver transplantation in children with Langerhans cell histiocytosis: experience from a quaternary care center. Pediatr Blood Cancer 2023;70(1):e30024. DOI: 10.1002/pbc.30024

In this review of a prospectively collected database of children with Langerhans cell histiocytosis (LCH) and liver disease from a single center over 7 years, authors have analyzed the outcome. Patients with LCH and hepatic involvement were divided into two categories based on the severity of liver involvement to guide appropriate chemotherapy. Category 1—patients with clinical, radiological, or histological evidence of chronic liver disease or cirrhosis with bilirubin less than 3 mg/dL (N: 0.6–1.2 mg/dL) and no signs of decompensation (absence of ascites, HE, or GI bleed). Category 2—patients with decompensated liver disease; that is, with ascites, variceal bleed or HE, and bilirubin >3 mg/dL, and/or acute on chronic liver failure; acute on chronic liver failure (bilirubin more than 5 mg/dL with INR above 1.5 along with the onset of ascites and/or HE within 4 weeks of the onset of jaundice). Category 1—patients were treated with a standard chemotherapy regimen for high-risk LCH. Category 2—patients who were less likely to tolerate the standard chemotherapeutic regimen were started on a modified chemotherapy regimen. Of the eight (five male) patients during the study period, six (75%) underwent LT (four and two for compensated and decompensated cirrhosis, respectively). On a median follow-up of 30.5 (10.5–50) months, all post-LT patients were alive with stable graft function and showed no disease recurrence. Authors have concluded that an algorithmic approach, along with newer chemotherapeutic agents, results in excellent outcomes in LCH patients with liver involvement.

Source: Bolia R, Goel AD. Systematic review and meta - analysis of the frequency and re-classification trends of pediatric inflammatory bowel disease - unclassified. Arq Gastroenterol 2022; 59(4):531–539. DOI: 10.1590/S0004-2803.202204000-92

Inflammatory bowel disease-unclassified (IBDU) is used when an individual has chronic colitis but cannot be subtyped into ulcerative colitis (UC) or Crohn's disease (CD) on the basis of the

clinical, endoscopic, imaging, and histopathological features. Through this systematic review, authors have concluded that IBDU comprises 7.1% of PIBD at initial diagnosis. Half of these children are reclassified into UC or CD on follow-up with a higher likelihood of reclassification to UC as compared to CD.

Source: Singh SK, Sarma MS. Hereditary fructose intolerance: a comprehensive review. World J Clin Pediatr 2022;11(4):321–329. DOI: 10.5409/wjcp.v11.i4.321

In this mini review, the authors have discussed the major natural history and management of hereditary fructose intolerance. Most commonly children are affected with GI symptoms, feeding issues, aversion to sweets, and hypoglycemia. Liver manifestations include an asymptomatic increase of transaminases, steatohepatitis, and rarely liver failure. Renal involvement usually occurs in the form of proximal renal tubular acidosis and may lead to chronic renal insufficiency. For confirmation, a genetic test is favored over the measurement of aldolase B activity in the liver biopsy specimen. The cornerstone of hereditary fructose intolerance management lies in the absolute avoidance of foods containing fructose, sucrose, and sorbitol.