PUBLICATIONS BY ISPGHAN MEMBERS IN PUBMED INDEXED JOURNALS (July 2021 – September 2021*)

*updated as on 25th September, 2021

JULY 2021

[1.]Yadav R, Bolia R, Bhat NK. Inappropriate Prescribing of Proton Pump Inhibitors in Children: Insights from a Survey. Indian J Pediatr. 2021 Oct; 88(10):1055. doi: 10.1007/s12098-021-03854-x. Epub 2021 Jul 1.

Authors have evaluated *the prevalence and appropriateness of proton pump inhibitors* (PPI) *use in the pediatric population* presenting to in a tertiary referral hospital over 2 months.All admitted children who had been prescribed PPIs by their referring physicians were identified and the appropriateness of the prescription was compared with FDA-approved indications. They evaluated 250 children, of which, 101 (40.4%, mean age 11 ± 6 y, 57 male) had been prescribed PPIs prior to admission (mean duration 25 ± 9 d). In only 14 (13.8%), PPIs were given for FDA-approved indications. Authors have found that 86.2% children were prescribed PPIs inappropriately with co-prescriptionwith steroids or NSAIDs (*n* = 29, 28.7%) being the commonest reason and concluded that inappropriate prescription of PPIs is highly prevalent in children.

[2.] Singh SP, Ahuja V, Ghoshal UC, Makharia G, Dutta U, Zargar SA, Venkataraman J, Dutta AK, Mukhopadhyay AK, Singh A, Thapa BR, Vaiphei K, Sathiyasekaran M, Sahu MK, Rout N, Abraham P, Dalai PC, Rathi P, Sinha SK, Bhatia S, Patra S, Ghoshal U, Poddar U, Mouli VP, Kate V. Management of Helicobacter pylori infection: The Bhubaneswar Consensus Report of the Indian Society of Gastroenterology. Indian J Gastroenterol. 2021 Jul 5. doi: 10.1007/s12664-021-01186-4. Epub ahead of print.

The Task Force constituted by Indian Society of Gastroenterology (ISG) which included pediatric gastroenterologist achieved a consensus on the 39 statements on *H. pylori infection* in Indians.*Statement 12* deals with *children*: Children with epigastric or upper abdominal pain, and with endoscopic findings of PUD (gastric or duodenal), should be tested and treated for H. pylori. Children with recurrent abdominal pain suggestive of functional pain should not be tested or treated for H. pylori.

[3.]Seetharaman J, Yadav RR, Srivastava A, Sarma MS, Kumar S, Poddar U, Yachha SK. Gastrointestinal bleeding due to pseudoaneurysms in children. Eur J Pediatr. 2021 Jul 14. doi: 10.1007/s00431-021-04201-0. Epub ahead of print.

In this retrospective study authors have described the etiology, clinical presentation, and outcome of radiological intervention in children with *pseudoaneurysm (PSA) of celiac or superior mesenteric artery branches*. Eleven children with PSA (5 boys, 11 [7–17] years) were analyzed . Ten (91%) patients were symptomatic: abdominal pain (10, 91%), hematemesis/ melena (9, 81%), and Quincke's triad (1, 9%). One child with pancreatic pseudocyst was diagnosed incidentally on imaging. Doppler ultrasound identified PSA only in 3 cases, while computed tomography angiography (CTA) picked all cases. Etiology was liver abscess (*n* 4), abdominal trauma (*n* 3), pancreatitis (*n* 3), and indeterminate in 1 case. Radiological embolization was done in 9 (81%) cases (coil 6, glue 2, both 1), without any complications or failure. One case resolved spontaneously and 1 died pre-intervention. Nine intervened cases were asymptomatic on follow-up [6 (1–24) months].Authors concluded that liver abscess, abdominal trauma and pancreatitis are common causes of celiac artery and superior mesenteric artery branch PSA in children and CTA has high sensitivity in identifying these pseudoaneurysms. Minimally invasive radiological angio-embolization, in the hands of trained radiologists, is a safe and successful modality of treatment in children.

[4.]Mantoo MR, Malik R, Das P, Yadav R, Nakra T, Chouhan P. Congenital Diarrhea and Enteropathies in Infants: Approach to Diagnosis. Indian J Pediatr. 2021 Jul 22. doi: 10.1007/s12098-021-03844-z. Epub ahead of print.

In this clinical brief authors have collated their experience of diagnosis and management of 4 cases of *congenital diarrhea and enteropathies* (CODEs) at their centre and suggested a *stepwise approach to the diagnosis and management* of these disorders in the Indian context.They have described 2 cases of congenital tufting enteropathy (CTE) and 1 case each of microvillous inclusion disease (MVID) and tricho-hepatoenteric syndrome (THES). Age at onset varied from 3 to 38 d of life. Light microscopy and electron microscopy of duodenal and rectal endoscopic biopsies were consistent with the diagnosis. Genetic evaluation was possible in 3 cases diagnosing causative mutations and highlight that with the availability of commercial next generation genetic testing, we are now better able to classify and manage these disorders. They have reported that 2 children (CTE and MVID) were alive at last follow-up.

[5.]Lal BB, Sood V, Khanna R, Alam S. Novel Variations in MYO5B Presenting as Isolated Intrahepatic Cholestasis: Long-Term Outcome after Partial Internal Biliary Diversion. Indian J Pediatr. 2021 Oct; 88(10):1052. doi: 10.1007/s12098-021-03848-9. Epub 2021 Jul 22.

In this correspondence authors have reported on 2 siblings with novel compound heterozygous variations in *MYO5B* presenting as isolated cholestasis with excellent long-term response after partial internal biliary diversion (PIBD). Both these children presented with *infantile onset cholestasis and low GGT* closely mimicking progressive familial intrahepatic cholestasis (PFIC) types 1 or 2.

[6.]Lal BB, Sood V, Rastogi A, Mukund A, Khanna R, Sharma MK, Alam S. Safety, Feasibility, Yield, Diagnostic and Prognostic Implications of Transjugular Liver Biopsy in Children and Adolescents. J Pediatr Gastroenterol Nutr. 2021 Jul 23. doi: 10.1097/MPG.0000000000003249. Epub ahead of print.

In this retrospective study authors have aimed to evaluate the indications, feasibility, complications and clinical implications of *transjugular liver biopsy* (TJLB) in children.102 children, including 5 with acute liver failure underwent TJLB with technical success in 101 (99%). The most common indications for TJLB in this cohort were elevated INR >1.5 (64.7%), ascites (45.1%) and thrombocytopenia (platelet count < 60,000/cu.mm) (41.2%). Mean size of the tissue received was 14.5 ± 5.6 mm with an average of 10.2 ± 4.7 portal tracts. Only 1 child developed major (category D) complication (hemobilia) and 12 (11.8%) developed minor complications post-procedure. Etiological diagnosis could be made in a total of 64 (63.9%) children undergoing TJLB, the most common diagnosis being autoimmune hepatitis (n = 31), non-cirrhotic portal fibrosis (n = 16) and drug-induced liver injury (n = 4).Authors have concluded that TJLB is safe, feasible and helps make a diagnosis in close to 64% children allowing timely medical and/or surgical intervention. It is especially useful for diagnosis of autoimmune liver diseases, DILI and NCPF.

[7.]Sen Sarma M, Gopan A. Chyle, not bile, from the major papilla. J Paediatr Child Health. 2021 Jul 27. doi: 10.1111/jpc.15652.

In this case report authors have described *tubercular sequelae of chyle leak from pancreatobiliary* tract in a 13-year-old boy, a treated case of multibacillary drug-sensitive abdominal tuberculosis (peritoneum, colon and lymph nodes) who presented with new-onset progressive anasarca 6 months after the completion of anti-tubercular therapy and responded to monthly intramuscular depot octreotide preparation (20 mg), high-dose fat-soluble vitamins and dietary therapy consisting of high protein (3 g/kg), high medium-chain triglycerides (30% of total calories) and low long-chain triglycerides (5% of total calories).

[8.]Venkatesh V, Aneja A, Seetharaman K, Anushree N, Rana SS, Lal SB. A Novel Cohesinopathy Causing Chronic Intestinal Pseudo Obstruction in 2 Siblings and Literature Review. J Neurogastroenterol Motil. 2021 Jul 30; 27(3):436-437.

In this letter to the editor the authors have reported on 2 siblings of the same family who presented constipation, intermittent episodes of pain, and distension of the abdomen of long duration, affected with *cohesinopathy* causing *chronic intestinal pseudo obstruction* (CIPO) and identified to harbor mutation in the *Shugoshin like-1 (SGOL1)* gene on genetic analysis which is a very entity. A proportion of primary CIPO in children is familial. SGOL1 gene encodes for a part of cohesin complex which is involved in cell division and is expressed in the intestinal wall; both smooth muscle and enteric nervous system. Authors concluded that this is the first report from Asia of this gene mutation leading to CIPO.

[9.]Nabi Z, Lakhtakia S, Chavan R, Asif S, Basha J, Gupta R, Yarlagadda R, Reddy PM, Kalapala R, Reddy DN. Diagnostic utility of EUS-guided tissue acquisition in children: A tertiary care center experience. Endosc Ultrasound. 2021 Jul-Aug; 10(4):288-293. doi: 10.4103/EUS-D-20-00203.

In this study, authors aimed to evaluate the feasibility, safety, and diagnostic utility of *EUS-FNA/FNB in children* with various gastrointestinal diseases. Sixty-seven children (32 - boys, 14.8 ± 2.9 years, range 8-18 years), underwent EUS-guided tissue acquisition procedures using standard therapeutic echoendoscope during the study period. The indications included solid pancreatic lesions in 29 (43.3%), mediastinal or abdominal lymphadenopathy in 30 (44.7%), cystic pancreatic lesions in 5 (7.5%), subepithelial lesions in 2 (3%), and retroperitoneal mass in 1 (1.5%). EUS-FNA and-FNB were performed in 42 and 25 children, respectively. All the procedures could be successfully performed and there was no major procedure-related adverse event. Minor adverse events included self-limiting throat pain (10) and abdominal pain (3), self-limited bleeding at puncture site (3), and transient fever (1). EUS-FNA/FNB provided a histopathological diagnosis in 59 (88.1%) children. **Authors** have concluded that EUS-guided tissue acquisition using standard echoendoscope is feasible and safe in the pediatric age group. EUS-FNA/FNB establishes diagnosis in majority of the children when performed for appropriate clinical indication.

AUGUST 2021

[10.]Madhusudan M, Sankaranarayanan S, Ravikumar T. Enterokinase Deficiency: A Case of Pancreatic Insufficiency. Indian J Pediatr. 2021 Aug; 88(8):825.

In this scientific letter, authors have reported a case of genetically confirmed *enterokinase deficiency* in a two month old female infant, 2nd born to non-consanguineous marriage who presented with passage of frequent oily stools with no blood or mucus since 2 weeks of age. The authors have highlighted that this case is being reported, as it is closely mimicks cystic fibrosis and there has been a drastic improvement in clinical features upon initiating treatment (pancreatic enzyme and fat-soluble vitamin supplementation, following which, her stools improved and she started gaining weight.)

[11.] Chaubal G, Hatimi H, Nanavati A, Deshpande A, Andankar P, Biradar V, Gupte P, Hanchnale P, Bhalerao S, Tambe S. Pediatric living donor intestine transplant following an atypical complication of COVID-19: A unique case report from India. Am J Transplant. 2021 Aug 13:10.1111/ajt.16798. doi: 10.1111/ajt.16798. Epub ahead of print.

Thrombotic phenomena associated with COVID-19 infection can affect larger vessel-like superior mesenteric artery leading to small bowel gangrene. Authors have reported on a 9-year-old child after recovering from mild COVID-19 infection developed small bowel gangrene due to superior mesenteric artery thrombosis requiring resection of entire necrotic .small bowel along with caecum causing ultra-short bowel syndrome. A *living donor intestinal transplant* was performed using 200 cm of ileum donated by the patient's father after being on individualized parentral nutrition for 3 months. He could be weaned off completely from parenteral nutrition by postoperative day 21. The donor had an uneventful recovery and was not complicated with thrombosis, infection, reactivation of latent COVID-19 or rejection. Six month follow-up was satisfactory with the child achieving complete enteral autonomy.

[12.]Menon J, Shanmugam N, Valamparampil JJ, Hakeem A, Vij M, Jalan A, Reddy MS, Rela M. Liver Transplantation: A Safe and Definitive Alternative to Lifelong Nitisinone for Tyrosinemia Type 1. Indian J Pediatr. 2021 Aug 16. doi: 10.1007/s12098-021-03826-1. Epub ahead of print.

Authors have aimed to analyze the clinical data of children with *tyrosinemia type 1* (TT-1) who underwent living donor liver transplantation (LT) between July 2009 and May 2020 retrospectively . Data included pre-LT nitisinone therapy,

graft type, post-LT complications, HCC incidence, and graft/patient survival. Nine children were diagnosed with TT-1 at a median age of 12 mo (6-54 mo). Nitisinone was started in 6 patients at a median age of 15 mo (6-42 mo), but all had frequent interruption of therapy due to logistics with drug procurement including its cost. Median age at transplantation was 5 y (2-11 y). Explant liver showed HCC in 5 patients (55% of total cohort). The graft and patient survival are 100% with median follow-up of 58 months (24-84 months). Authors have concluded that LT is curative for TT-1 and excellent results can be obtained in experienced centers. They further added that LT is especially favorable in countries with limited resources where the cost of medical therapy is highly prohibitive, with lifelong diet restrictions and unclear long-term risk of HCC.

[13.]Manjunath S, Mahajan R, De D, Handa S, Attri S, Behera BN, Bhasin SL, Bolia R. The severity of malnutrition in children with epidermolysis bullosa correlates with disease severity. Sci Rep. 2021 Aug 19; 11(1):16827. doi: 10.1038/s41598-021-96354-z.

In this study authors have aimed to assess nutritional aspects of Indian children suffering from *epidermolysis bullosa* (EB) and to evaluate the effect of severity of EB on the severity of malnutrition. In this single center, prospective longitudinal study, 57 pediatric EB patients were evaluated prospectively for baseline nutritional status using anthropometric parameters and WHO growth charts, and its correlation with disease severity using instrument for Scoring Clinical Outcomes for Research of Epidermolysis Bullosa-*iscorEB*. In second phase, an individualized diet chart was given to meet the energy, protein and micronutrients needs and its effects were observed after 6 months. The median age of participants was 3 years (IQR-9). Malnutrition was seen in 40.35% patients (22.81%-moderate and 17.54%-severe), and significantly correlated with iscorEB ($r = 0.45$, $p < 0.0001$). On bivariate regression analysis, iscorEB was independently associated with moderate-to-severe malnutrition ($p = 0.047$; OR 1.038, CI 1.011-1.066).Authors have concluded that the severity of malnutrition in EB children significantly correlates with disease severity, and is an independent predictor of moderate-to-severe malnutrition.

[14.]Bolia R, Sherwani P, Garnaik DK. Stercoral Colitis in an Adolescent. Clin Gastroenterol Hepatol. 2021 Aug 20:S1542-3565(21)00902-2. doi: 10.1016/j.cgh.2021.08.022. Epub ahead of print.

In this case report presented as electronic image of the month authors have described *stercoral colitis* in a 15-year-old adolescent boy with cerebral palsy having chronic constipation for 5 years. Sigmoidoscopy demonstrated significant stool in the rectosigmoid region with diffuse erythema, edema, and ulceration, biopsies showing necrotic granulation tissue. The child responded to conservative management with aggressive bowel cleansing and intravenous antibiotics.

SEPTEMBER 2021

[15.] Tripathi PR, Sen Sarma M, Yachha SK, Aggarwal A, Bhatia V, Kumar A, Srivastava A, Poddar U. Relative Adrenal Insufficiency in Decompensated Cirrhotic Children: Does It Affect Outcome? Am J Gastroenterol. 2021 Sep 10. doi: 10.14309/ajg.0000000000001486. Epub ahead of print.

In this study authors have aimed to prospectively study the presence and outcome of *relative adrenal insufficiency (RAI) in children with decompensated cirrhosis* over 180 days. 63 hemodynamically stable children with decompensated cirrhosis were sampled for serum basal cortisol and peak cortisol (after 30 minutes of 1-μg intravenous Synacthen) at day 1 and day 21. RAI was diagnosed as peak cortisol <500 nmol/L.Patients with RAI at baseline (D1-RAI) developed higher complications at follow-up as compared to the normal adrenal function group (53% vs 24%, $P = 0.02$). The PELD score (odds ratio 1.08, confidence interval 1.05-1.12, $P < 0.01$) and D1-RAI (odds ratio 3.19, confidence interval 1.32-7.73, $P = 0.01$) were independent predictors of follow-up complications. The PELD-delta cortisol model (area under the receiver operating curve 0.84, $P < 0.001$, 92% sensitivity; 60% specificity) predicted morbidity better than isolated PELD or Child-Turcotte-Pugh scores. Authors have concluded that RAI is a risk factor for development of complications in pediatric cirrhosis over short-term follow-up. The PELD-delta cortisol score is a promising prognostic model for predicting follow-up complications.

[16.] Mohanty N, Dheivamani N, Mane S, Acharyya B, Kamale V, Poddar S, Khobragade A, Thomas W, Prabhudesai S, Choudhary A, Mitra M. Effect of *Saccharomyces boulardii* CNCM-I 3799 and *Bacillus subtilis* CU-1 on Acute Watery Diarrhea: A Randomized Double-Blind Placebo-Controlled Study in Indian Children. Pediatr Gastroenterol Hepatol Nutr. 2021 Sep; 24(5):423-431.

In this multi centre randomized double-blind placebo-controlled study authors have aimed to assess the effect of combination probiotic *Saccharomyces boulardii* CNCM-I 3799 and *Bacillus subtilis* CU-1 in outpatient management of acute watery diarrhea in children. 180 participants aged six months to five years with acute mild to moderate diarrhea were enrolled from six centers across India and centrally randomized to receive *S. boulardii* CNCM-I 3799 and *B. subtilis* CU-1 or a placebo along with oral rehydration salts and zinc supplementation. Each participant was followed up for three months to assess recurrence of diarrhea.Authors have concluded that *S. boulardii* CNCM-I 3799 and *B. subtilis* CU-1 combination was effective in reducing the duration of diarrhea when administered within 48 hours of diarrhea onset. Similarly, it reduced recurrence of diarrhea and its intensity in the subsequent three months.

MISSED INADVERTENTLY IN PREVIOUS ISSUES

JANUARY 2021

[1.] Srivastava A, Saini N, Mathias A, Arya A, Jain S, Yachha SK. Prevalence and predictive factors of undernutrition and low bone mineral density in children with chronic pancreatitis. Pancreatology. 2021 Jan; 21(1):74-80. doi: 10.1016/j.pan.2020.11.009.

Authors have studied the *nutritional status and bone mineral density (BMD) of children* with *chronic pancreatitis (CP)* and the factors predicting them.83 children (46 boys, 14[4.3-21] years) with CP were prospectively evaluated with a detailed questionnaire, anthropometry, 25-hydroxy vitamin D, fecal elastase and BMD [total body less head (TBLH), spine and hip] by dual energy x-ray absorptiometry. Body mass index (BMI) Z score of -1 to -1.9, -2 to -2.9 and <-3 was

taken as mild, moderate and severe malnutrition respectively. Low BMD and osteoporosis were defined as per International Society for Clinical Densitometry. They have concluded that 41% CP children have undernutrition with a majority having mild undernutrition(mild-37.3%, moderate-2.4%, severe-1.2%). Nearly 20% have low BMD, with osteoporosis in none. Subjects with low BMI have lower BMD and percentage body fat1.3[-1.9 to 0.34] vs 0.8 [-2.1 to 5.50; p = 0.03).

MARCH 2021

[2.] Bolia R, Srivastava Y, Yadav R, Sherwani P. Unusual cause of severe iron deficiency anaemia in a child: paraoesophageal hernia. BMJ Case Rep. 2021 Mar 4; 14(3):e237728. doi: 10.1136/bcr-2020-237728.

Authors have described a 5-year-old boy who presented with severe transfusion-dependent iron deficiency anaemia caused by a *paraoesophageal hernia*. Surgical repair of the hiatus hernia led to complete resolution of anaemia. They have concluded that hiatus hernia should be considered as a diagnostic possibility when evaluating a child with refractory iron deficiency anaemia.

[3.] Shabrish S, Kelkar M, Yadav RM, Bargir UA, Gupta M, Dalvi A, Aluri J, Kulkarni M, Shinde S, Sawant-Desai S, Kambli P, Hule G, Setia P, Jodhawat N, Gaikwad P, Dhawale A, Nambiar N, Gowri V, Pandrowala A, Taur P, Raj R, Uppuluri R, Sharma R, Kini P, Sivasankaran M, Munirathnam D, Vedam R, Vignesh P, Banday A, Rawat A, Aggarwal A, Poddar U, Girish M, Chaudhary A, Sampagar A, Jayaraman D, Chaudhary N, Shah N, Jijina F, Chandrakla S, Kanakia S, Arora B, Sen S, Lokeshwar M, Desai M, Madkaikar M. The Spectrum of Clinical, Immunological, and Molecular Findings in Familial Hemophagocytic Lymphohistiocytosis: Experience From India. Front Immunol. 2021 Mar 5;12:612583. doi: 10.3389/fimmu.2021.612583.

This study is a retrospective analysis of 101 molecularly characterized *familial hemophagocytic lymphohistiocytosis* (FHL) patients over the last 10 years from 20 different referral centers in India. FHL2 and FHL3 together accounted for 84% of cases of FHL in our cohort.This article sheds light on the current scenario of FHL in India. Molecular characterization of respective genes revealed 76 different disease-causing mutations including 39 (51%) novel mutations in *PRF1, UNC13D, STX11*, and *STXBP2* genes. Overall, survival was poor (28%) irrespective of the age of onset or the type of mutation in our cohort.This data reveal a wide genetic heterogeneity of FHL in the Indian population and confirms the poor prognosis of FHL.

JUNE 2021

[4.] Sen Sarma M, Yachha SK. Fever, ascites and rugged skin lesions. J Paediatr Child Health. 2021 Jun 28. doi: 10.1111/jpc.15624. Epub ahead of print.

In this case report published as image of the month, authors have described a 13-year-old girl from northern India who presented with fever,weight loss and ascites of 2 months. She also had non-pruritic,non-tender progressive skin lesions on the anterior aspect of the right thigh for 1 month Histopathology of skin nodules showed lymphoplasmacytic infiltration of subepithelium with presence of acid-fast bacilli and caseating granulomas. *Cutaneous lupus vulgaris* (tuberculosis of skin) was diagnosed. Anti-tubercular therapy was administered for a total of 6 months with complete resolution of all symptoms. Lupus vulgaris is a very rare association with peritoneal tuberculosis.

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