

PUBLICATIONS BY ISPGHAN MEMBERS IN PUBMED INDEXED JOURNALS

(16th March, 2022 – 31st May, 2022)

Updated as on 31st May, 2022

APRIL 2022

[1.] Kumar K, Malhotra S, Sibal A. **Transient infantile hypertriglyceridaemia due to homozygous mutation in GPD1 presenting in childhood with hepatic adenoma.** *BMJ Case Rep.* 2022 Apr 1; 15(4):e248801. doi: 10.1136/bcr-2022-248801. PMID: 35365473; PMCID: PMC8977762.

In this case report authors illustrate on the development of adenoma in transient infantile hypertriglyceridaemia [HTGTI] in a child on follow up who presented with hypoglycaemia, hepatomegaly, high transaminases and hypertriglyceridaemia in infancy. Authors conclude that there are only a few cases reported worldwide and none has reported development of adenoma so far. This could be the first report of development of adenoma in transient HTGTI.

[2.] Sathiaseelan M, Bolia R, Barallon R, Alex G, Hardikar W, Rajanayagam J. **Impact of ulcerative colitis on liver-related outcomes of children with primary sclerosing cholangitis.** *J Paediatr Child Health.* 2022 Apr 4. doi: 10.1111/jpc.15954. Epub ahead of print. PMID: 35373867.

In this retrospective analysis of prospectively maintained data authors from a tertiary Pediatric liver unit have aimed to evaluate the effects of ulcerative colitis [UC] on liver related outcomes. 51 children with primary sclerosing cholangitis [PSC] during the study period from January 1998 to May 2016 were enrolled in the study. Outcomes studied included biliary complications, clinically significant portal hypertension, need for liver transplantation and post-transplantation recurrence. Thirty-seven (73%) patients had concurrent UC, of which 26 had their diagnosis confirmed prior to or within 6 months of PSC diagnosis (early-onset). PSC complications were more common in children with PSC-UC compared with PSC alone. Children with endoscopically mild or moderate UC at diagnosis showed a greater propensity for liver-related complications compared with children with severe UC. Children with late-onset UC had higher rates of clinically significant portal hypertension and liver transplantation. Children with PSC-UC had significantly higher rates of pancolitis, rectal sparing and milder colitis than those with UC alone.

[3.] Bhatia A, Bhatia H, Saxena AK, Lal SB, Sodhi KS. **Shear wave elastography of the spleen using elastography point quantification: stiffness values in healthy children.** *Abdom Radiol (NY).* 2022 Apr 18. doi: 10.1007/s00261-022-03519-z. Epub ahead of print. PMID: 35437707.

In this prospective study authors have aimed to evaluate the shear wave elastography (SWE) values of the spleen in

healthy children using Elastography Point Quantification (ElastPQ). SWE values were recorded in upper pole, mid pole, and lower pole of the spleen in 146 children and compared across the age groups ≤ 5 years, $> 5-10$ years and $> 10-15$ years. Authors have concluded that SWE values of the spleen in healthy children do not correlate with age, and no significant difference is there in the SWE values for boys and girls. There was a statistically significant difference in the SWE values of the spleen while comparing the groups based on the median splenic length.

MAY 2022

[4.] Nabi Z, Talukdar R, Lakhtakia S, Reddy DN. **Outcomes of Endoscopic Drainage in Children with Pancreatic Fluid Collections: A Systematic Review and Meta-Analysis.** *Pediatr Gastroenterol Hepatol Nutr.* 2022 May; 25(3):251-262.

In this systematic review and meta-analysis authors have aimed to analyze the outcomes of endoscopic drainage with or without endoscopic ultrasonography (EUS) guidance in children with pancreatic fluid collections [PFCs]. Fourteen studies (187 children, 70.3% male) were included in this review. The subtypes of fluid collection included pseudocysts (60.3%) and walled-off necrosis (39.7%). The pooled technical success rates in studies where drainage of PFCs were performed with and without EUS guidance were 95.3% and 93.9% respectively. The pooled clinical success after one and two endoscopic interventions were 88.7% and 92.3%, respectively. The pooled rate of major adverse events was 6.3%. The pooled rate of recurrent PFCs after endoscopic drainage was 10.4%. They have concluded that endoscopic drainage is safe and effective in children with PFCs and added that future studies are required to compare endoscopic and EUS-guided drainage of PFCs in children.

[5.] Dogra S, Kumar K, Malhotra S, Sibal A. **Novel mutation causing congenital disorder of glycosylation in a child with recurrent anasarca.** *BMJ Case Rep.* 2022 May 13; 15(5):e245884. doi: 10.1136/bcr-2021-245884. PMID: 35568419; PMCID: PMC9109037.

In this case report authors have reported on a child who presented with recurrent anasarca who was diagnosed to have congenital disorder of glycosylation harbouring a novel mutation. Through this case report authors emphasize that protein losing enteropathy should be suspected in children with chronic diarrhoea and peripheral oedema and worked up for definitive etiology.

[6.] Menon J, Shanmugam N, Vij M, Rammohan A, Rela M. Liver transplantation for Tight junction Protein 2 Deficiency associated Liver Disease. *Liver Transpl.* 2022 May 26. doi: 10.1002/lt.26515. Epub ahead of print. PMID: 35615909.

In this brief report authors have presented a first series of preliminary post-liver transplant [LT] outcomes in progressive familial intrahepatic cholestasis [PFIC] type IV. Previously undescribed, certain interesting findings unique to this disease were noted in this series of 4 cases and have been highlighted. One of the four patients had a high-GGT cholestasis; one of the patients, developed recurrent pneumothorax observed on the 5th post-LT day and one of four patients had an incidental HCC on the explant liver. On a median follow-up of 18.5 months (range 5-58 months), all the recipients remained well with a normal graft function. Severely stunted growth was a striking feature of all patients except one, all of them had a remarkable improvement in their Z scores following LT. None of the patients developed any extrahepatic manifestations or disease recurrence.

MISSED INADVERTENTLY IN PREVIOUS ISSUES

2021

[1.] Vinayagamoorthy V, Srivastava A, Sarma MS. Newer variants of progressive familial intrahepatic cholestasis. *World J Hepatol.* 2021 Dec 27; 13(12):2024-2038. doi: 10.4254/wjh.v13.i12.2024. PMID: 35070006; PMCID: PMC8727216.

In this minireview authors have described the pathogenesis and clinical features of the newer variants of progressive familial intrahepatic cholestasis [PFIC]. PFIC manifests with a varying spectrum of clinical features, with some variants progressing rapidly into end stage liver disease. Recently, newer variants of PFIC have been described including PFIC 4 due to tight junction protein 2 (TJP2) mutation, PFIC 5 due to NR1H4 mutation and MYO5B related cholestasis also sometimes known as PFIC 6. TJP2 related PFIC has variable clinical presentations and carries risk for development of hepatocellular carcinoma. PFIC-5 patients usually have rapidly progressive liver disease with early onset coagulopathy, high alpha-fetoprotein and ultimately require a liver transplant. Subjects with MYO5 B-related disease can present with isolated cholestasis or cholestasis with intractable diarrhea resulting from microvillus inclusion disease [MVID]. These children are at risk of worsening cholestasis post intestinal

transplant (IT) for MVID, hence combined intestinal and liver transplant or IT with biliary diversion is preferred.

2022

[1.] Yachha SK, Sarma MS, Mohan N, Wadhwa N, Kumar Vr N, Srinivasan R, Shah D, Soans S, Yewale V, Basavaraja GV, Parekh BJ. Indian Academy of Pediatrics Consensus Guidelines for Probiotic Use in Childhood Diarrhea. *Indian Pediatr.* 2022 Mar 10:S097475591600411. Epub ahead of print. PMID: 35273131.

A national consultative group (NCG) was constituted by the Indian Academy of Pediatrics (IAP), consisting of subjects experts including representation from ISPGHAN fraternity to develop a guideline for the use of probiotics in children with diarrhea. The NCG suggested *Lactobacillus GG* as a conditional recommendation with low-to-moderate level evidence or *Saccharomyces boulardii* as a conditional recommendation with very low-to-low level evidence as *adjuvant therapy in acute diarrhea*. The NCG also recommended the *use of combination probiotics in neonatal necrotizing enterocolitis (NEC)*, as these reduce the risk of NEC stage II and above, late-onset sepsis, mortality and also time to achieve full feeds. *The NCG did not recommend the use of any kind of probiotics in the therapy of acute dysentery, persistent diarrhea, Clostridium difficile diarrhea and chronic diarrheal conditions such as celiac disease, diarrhea-predominant irritable bowel syndrome and inflammatory bowel disease in children.* The NCG recommended probiotics only in special situations of AAD. *L. rhamnosus GG* or *S. boulardii* may be used for the prevention of AAD. *VSL#3*, a combination probiotic, may be used as an adjuvant in active pouchitis, prevention of recurrences and maintenance of remission in pouchitis.

Compiled by

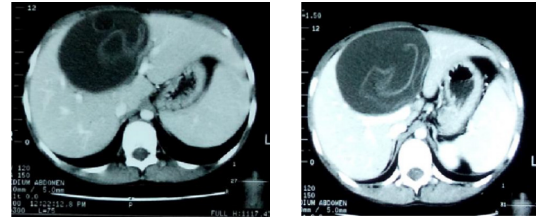
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A 14 year old girl presented with progressively enlarging lump and pain in right hypochondrium with intermittent fever for last 3 years. Complete blood picture and liver function tests were largely unremarkable. A contrast enhanced computerised tomography was performed.

What is the diagnosis and possible modalities of management (in brief)?



QUIZ 1 ANSWER

1. Diagnosis: Water lily sign, Hydatid cyst of liver.
2. Possible treatment modalities:
 - a) Long term albendazole,
 - b) Percutaneous puncture, aspiration, injection, re-aspiration (PAIR),
 - c) Excision surgery

Congratulations to the Winners of Quiz 1 APGH

Dr Siddartha Korampalli, Pune
 Dr Lata Jha, Ahmedabad
 Dr Aryan Gupta, Pune
 Dr Valapala Venkatesh, Visakhapatnam
 Dr Sherin PA, Latur
 Dr Ashok Goyal, New Delhi
 Dr Arghya Samanta, Lucknow

QUIZ QUIZ 2

A 1.5 month old baby was brought to the emergency with recurrent non-bilious vomiting and failure to gain weight. Complete blood picture and liver function tests were largely unremarkable. Abdominal sonology showed normal pyloric channel. A barium study was performed.

What is the diagnosis and the possible modality of management?



Send in your answers to pglj.ispghan@gmail.com by 15/07/2022 with your name, designation, affiliation and email id.

Answers will be revealed in the next issue | Winners will receive an e-certificate