Case Snippets
Congenital intrahepatic porto-systemic venous shunt with galactosemia

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We report a 51-day-old infant with congenital intrahepatic porto-systemic venous shunt associated with galactosemia, who presented with cholestatic jaundice. He was treated with ursodeoxycholic acid, calcium supplements and galactose-free diet. The child was asymptomatic six weeks later. [Indian J Gastroenterol 2007;26:87-88]

Intrahepatic porto-systemic venous shunts (IPSVS) are rare, and are caused either by a congenital vascular malformation or by a collateral pathway secondary to cirrhosis of liver and portal hypertension. Its association with galactosemia is rare.

A 51-day-old male infant, weighing 4.5 Kg, presented with complaints of cholestatic jaundice since day 5 of life. The infant was born after hormonal induction and was born at full term with uneventful vaginal delivery. The child cried immediately after birth and had been on breast feeds since day 1 of life. There was no history of fever, seizures, abnormal movements, or bleeding. Prenatal sonographic findings had been normal.

Physical examination revealed the child was moderately built, active, and accepted feeds normally. Head circumference was normal. He had icterus, but there was no cyanosis, clubbing or lymphadenopathy. The liver was palpable 4 cm below the right costal margin (span 8.5 cm). Spleen was not palpable. The other systems were normal.

Serum bilirubin was raised (14 mg/dL), but the rest of the liver profile, serum electrolytes, urea and creatinine levels were within normal limits. Chest X-ray revealed mild cardiomegaly. Echocardiography was normal. Urine routine and culture tests were normal, but urine tested positive for galactose. Plasma ammonia levels were raised (176 µmol/L (normal 53-88). Galactose-1-phosphate uridyl transferase levels were low (0.275 units/g hemoglobin; normal 12-45), suggestive of galactosemia.

HIDA scan showed good hepatocyte function with slow biliary clearance into intestine. Ultrasonography and color Doppler (Fig) revealed a large vessel connecting the right branch of portal vein to the inferior vena cava. Spectral Doppler also showed turbulent flow in the right branch of portal vein. The child was managed with ursodeoxycholic acid, multivitamin, calcium supplementation and galactose-free diet. Six weeks later he was symptom-free.

Early diagnosis of IPSVS is important as it can lead to symptomatic hyperammonemia and hepatic encephalopathy. Percutaneous liver biopsy is a common cause and should be excluded before diagnosing congenital IPSVS.

Congenital anomalies of the portal venous system result from abnormal coalescence of the vitello-umbilical venous plexus during embryogenesis. These rare anomalies include cavernous transformation of the portal vein, preduodenal portal vein or peribiliary portal vein, duplication of portal vein, and portal vein atresia. A unique consequence of some of these anomalies is congenital porto-systemic shunting of the splenic venous circulation. Color Doppler imaging combined with pulsed Doppler examination is highly sensitive for the diagnosis of IPSVS and obviates the need for angiography.

IPSVS may be associated with congenital cardiac defects or abnormalities of the hepatobiliary system, including abnormal lobulation of the liver, hepatoblastoma and extrahepatic biliary atresia.

The choice of treatment of IPSVS is controversial. The vascular anomaly may regress spontaneously during infancy. In patients with large shunts causing encephalopathy, therapeutic intervention should be considered. This can be in the form of surgical banding, or angiographic intervention by coil embolization. Co-malformation of hepatic veins and severe anomalies such as cardiac defects exclude surgical intervention.

References
Reversal of severe hepatopulmonary syndrome in congenital hepatic fibrosis after living-related liver transplantation

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We report a 5-year-old girl with congenital hepatic fibrosis who presented with clubbing and cyanosis. Partial pressure of oxygen was 40 mmHg with oxygen saturation of 70% on room air, which improved to 128 mmHg and 92% on inhalation of 100% oxygen. Macroaggregated albumin scan showed 58% shunting to the brain, suggestive of severe hepatopulmonary syndrome. Echocardiogram and pulmonary angiogram ruled out pulmonary hypertension. Four weeks after living-related liver transplantation, she had normal blood gases and reduction in shunting to 7% on macroaggregated albumin scan. [Indian J Gastroenterol 2007;26:88-89]

Hepatopulmonary syndrome has also been described in patients with non-cirrhotic causes of portal hypertension.1,2 A five-year-old girl had presented at the age of 3 1/2 years with variceal bleed. She had large esophageal varices with severe portal hypertensive gastropathy. Clinical examination revealed a malnourished child with muscle wasting, moderate hepatosplenomegaly, grade III clubbing and cyanosis. She did not have icterus, edema of feet or ascites. She underwent multiple sessions of endoscopic variceal band ligation over the next 1 1/2 years. She also developed bleeding per rectum and was found to have large rectal varices. She had stopped going to school on account of dyspnea on exertion and easy fatiguability. There was history of abdominal distention at 6 months of age, which was attributed to hepatomegaly and ascites, which resolved in a few months. Liver biopsy then was reported as showing congenital hepatic fibrosis.

Investigations: total protein 6.3 g/dL (albumin 3.3), normal AST, ALT, bilirubin and alkaline phosphatase. Prothrombin time was 14 s (control 12), INR 1.1. HBsAg, anti-HCV, ANA were negative. Serum ceruloplasmin and serum ferritin levels were normal. Abdominal ultrasonography showed nodular, enlarged liver with splenomegaly and portal-systemic collaterals, with portal vein of 5 mm and splenic vein of 3 mm. Doppler examination revealed portal hypertension but the inferior vena cava and hepatic veins were normal.

Arterial blood gas showed oxygen saturation of 75%, PaO2 40 mmHg, which increased to 92% and 128 mmHg, respectively, after inhalation of 100% oxygen. Technetium-labeled macroaggregated albumin scan showed shunting in the lung (Fig) with 58% uptake by the brain. 2D echocardiogram was normal, with no evidence of pulmonary hypertension and pulmonary artery peak systolic arterial pressure of 25 mmHg. Conventional pulmonary angiogram did not reveal any pulmonary A-V fistulae and the mean systolic arterial pressure was 22 mmHg.

She was evaluated for living-donor liver transplantation. Her 33-year-old father of the same blood group (A+ve) was considered appropriate as the prospective donor. A left-lateral-segment graft was harvested by the standard technique. After recipient hepatectomy with preservation of the inferior vena cava, the graft was implanted. The graft left hepatic vein was anastomosed after creating a triangular orifice on the recipient cava. The donor left portal vein was anastomosed to the recipient portal vein and the donor left hepatic artery to the recipient common hepatic artery. Segments 2 and 3 bile ducts on the cut surface of the left lateral segment were anastomosed to a Roux loop of jejunum.

Fig: Technetium-labeled macroaggregated albumin scan before (left) and four weeks after liver transplantation
Her liver explant pathology showed presence of nodules containing central veins and fibrosis. The hepatic plates and bile ducts were normal, suggestive of congenital hepatic fibrosis. Postoperatively she was treated with tacrolimus and steroid-based immunosuppressive regimen. She required ventilation for 14 days and subsequently oxygenation was maintained on a rebreathing mask and nasal O2. By 3 weeks her SaO2 was 95% with PO2 of 80 mmHg off oxygen therapy. She is now 7 months post transplantation and is back to normal activity. She has gained 4 Kg weight. Macroaggregated albumin scan at 6 months is normal.

Severe pulmonary hypertension is a contraindication to transplant, but severe hepatopulmonary syndrome may be reversible with liver transplant. Portosystemic shunting in the form of TIPPS or surgical shunts have been used for the treatment of hepatopulmonary syndrome. However, both these procedures would be difficult in a small child. Also, surgical shunting may not necessarily reverse the process. There are reports of reversal of hepatopulmonary syndrome in patients with Budd-Chiari syndrome after cavoplasty was performed, or granulomatous hepatitis treated with corticosteroids, where the underlying disease was treatable. There also have been reports of liver transplantation being used for non-cirrhotic causes of hepatopulmonary syndrome.

Although some authors initially suggested severe hepatopulmonary syndrome to be too high a risk for liver transplantation, other studies suggest otherwise. The reversibility of hypoxemia with inhalation of oxygen pre-operatively was considered an indicator of reversibility of hepatopulmonary syndrome postoperatively in our patient. Post-transplant, patients with hepatopulmonary syndrome have a longer ventilatory requirement as compared to other patients, and it can take up to 14 months for the syndrome to reverse. Our patient showed reversal of hepatopulmonary syndrome within 3 weeks of transplant in spite of severe disease.

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Wegener’s granulomatosis – an etiology of acute pancreatitis
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Wegener’s granulomatosis is a systemic disease that usually involves the upper respiratory tract and kidneys. We report a 47-year-old man with Wegener’s granulomatosis that presented as acute pancreatitis. [Indian J Gastroenterol 2007;26:89-90]

Either gallstone or alcohol causes acute pancreatitis in 80% of cases. We report a patient who presented with pancreatitis, and was later diagnosed to have Wegener’s granulomatosis.

A 47-year-old man was admitted with complaints of two episodes of severe abdominal pain in 2 months. Pain was continuous with intermittent exacerbations, aggravated by meals, and radiating to the flanks and back. He had mild fever during each attack. There was no history of vomiting, diarrhea or jaundice during the attack. During the second episode he also had pain in both ears and nose, reddening of eyes, throat congestion, mouth ulcers and frothy urine. He had pain and swelling with early morning stiffness of large joints. He had history of recurrent rhinitis, nasal crusting and burning in the eyes for 2½ years, and had had nasal polypectomy 25 years earlier. He was a smoker since 25 years and never consumed alcohol.

On examination the patient was stable hemody-
namically, and had moderate tenderness in the epigas-
trium. Bowel sounds were normal. The ophthalmolo-
gist diagnosed episcleritis.

**Investigations:** hemogram normal, ESR 49 mm in
first hour, random blood sugar 256 mg/dL, renal and
liver profile normal; urine showed trace albumin and
few granular casts. Serum amylase was 874 units, li-
pase 1294 units. X-ray chest and abdomen was nor-
mal; ultrasonography showed bulky body and tail of
pancreas with altered echotexture. CT scan showed
bulky and edematous pancreas.

Treatment with broad-spectrum antibiotics, IV fluids
and other supportive measures was initiated. The im-
munologist suspected Wegener’s granulomatosis. An-
tinuclear cytoplasmic antibody (ANCA) immuno-
fluorescence showed a cytoplasmic pattern, and
proteinase-3 (ELISA) was 156.2 U/mL (normal less than
5). Nasal endoscopy showed severe deviation of nasal
septum to the left with spur to the right. Multiple granu-
lations were present on the right middle turbinate and
over the right side of the septum. Biopsy from the granu-
lations showed inflammatory infiltrate composed pre-
dominantly of neutrophils and large areas of necrosis
surrounded by histiocytes and granulomatous reaction
along with Langhan’s type of giant cells (Fig). It
confirmed Wegener’s granulomatosis.

The patient was subsequently started on corti-
costeroids and cyclophosphamide. He responded with
marked improvement in arthralgia, nasal and ocular con-
gestion and abdominal pain. Six months later, he was
asymptomatic except for occasional redness of eyes.
He was continued on oral prednisolone 10 mg/day.

Wegener’s granulomatosis is believed to be a
hypersensitivity or autoimmune response to an
inhaled infectious or other environmental agent.
It is characterized by the triad of i) necrotizing
granulomatous inflammation involving the upper
respiratory tract, ii) necrotizing vasculitis affect-
ing small to medium-sized vessels all over body and
iii) renal disease, especially focal necrotizing
glomerulonephritis. Approximately 80%-85% of pa-
tients show some signs of renal involvement but
less than 20% show renal impairment. 2

Pancreas rarely gets involved in this disease
as a part of generalized vasculitis. Only three cases
have been reported showing Wegener’s granulo-
matosis as a cause of acute pancreatitis, 3,4,5 one
among them being diagnosed at autopsy. 5 Cases
presenting as pancreatic tumor 6 and with pancre-
atic exocrine insufficiency have also been reported. 7

Most patients require immunosuppression. Cy-
clophosphamide is highly effective, though 50% of
patients relapse during treatment with it. Eighty
percent of patients die within 1 year if left un-
treated.

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**Hepatic calcification following dengue virus-induced fulminant hepatic failure**

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Hepatic calcification can be seen with various infec-
tious and neoplastic conditions. We report a 32-year-

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**Fig:** Lymphocyte-poor inflammatory infiltrate with histio-
cytes, granulomatous reaction and Langhan’s type of giant
cells (H&E, 40X)
A 32-year-old man presented with high-grade fever with chills and rigors and progressive jaundice of 6 days’ duration. He developed anuria and altered behavior 24 hours prior to presentation. Past medical history was unremarkable. On clinical examination, he was febrile (38.9°C), icteric and tachypneic (respiratory rate 24/min). His pulse rate was 112/min and blood pressure was 130/80 mmHg. There was no lymphadenopathy, skin rashes, mucosal bleed or pedal edema. Abdominal examination revealed liver span of 14 cm, moderate ascites and normal bowel sounds. Chest and cardiovascular system examination was normal. The patient was drowsy, responding to verbal commands, and had asterixis.

Serum biochemistry showed total bilirubin 7.1 mg/dL (conjugated 4.2), AST 13,000 IU, ALT 5,600 IU, alkaline phosphatase 727 IU, proteins 7.0 g/dL (albumin 3.6), creatinine 7.3 mg/dL and phosphorus 5.1 mg/dL; prothrombin time was 24 s (control 13). A diagnosis of acute liver failure with acute renal failure was made. A possibility of underlying chronic liver disease was also considered.

Markers for hepatotropic viruses (IgM anti HAV, HBsAg, IgM anti HBC, anti HCV antibodies and IgM anti HEV) were negative. Autoimmune markers ANA, ASMA, and anti-LKM were also negative. Twenty-four-hour urinary copper and serum ceruloplasmin levels were within the reference range. Serology for leptospirosis, herpes simplex virus I and II, cytomegalovirus and Epstein-Barr virus was negative. Repeated blood cultures were sterile. Peripheral smear for malarial parasite and malaria antigen were negative. IgM antibody against dengue virus was strongly positive. Serology for HIV I and II was negative. CPK was normal and urine for myoglobinuria and hemoglobinuria tested negative. Urine culture was sterile, Ultrasonography revealed ascites and liver span of 14 cm with diffusely increased echogenicity; hepatic margins were smooth. Gall bladder, common bile duct and pancreas were normal. Chest X-ray revealed minimal right-sided pleural effusion.

The patient was managed conservatively in the ICU with intravenous antibiotics, fluids, and other supportive treatment. He underwent twelve hemodialysis sessions during hospital stay and was later on maintenance hemodialysis for a month after discharge. His hepatic and renal function gradually improved and normalized after two months.

Massive hepatic calcification is rare. Hepatic calcification can be seen with granulomatous and other infections and in liver tumors.1,2

Parenchymal hepatic calcification may be intracellular or interstitial, the latter occurring in the majority of cases.1 It is usually associated with infectious or neoplastic processes and is focal or multifocal in appearance.1,2 A more diffuse pattern has been described in patients with severe parasitic infections.3

Calcification of the liver is usually dystrophic and occurs in dead tissue following hepatocyte necrosis due to infection or ischemia. Hepatic calcification has been reported as a sequel to shock liver or ischemic hepatitis secondary to systemic hypotension.4,5

Fulminant hepatic failure associated with viral hepatitis does not lead to any sequelae and the hepatocytes regenerate completely. The only case of dengue virus-induced hepatic calcification reported till date is that of a 15-year-old patient who developed acute liver failure after a week of 'flu-like illness and jaundice. However, in contrast to our case, hepatic calcification in this patient was distributed throughout the liver and was detected on follow up 10 years later.6 We could not find an explanation for the isolated calcification in our case. Theoretically, such type of calcification may occur as a result of lobar or segmental vascular compromise.

References
Non-resolving liver abscess with Echinococcus cross-reactivity in a non-endemic region

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A 40-year-old woman presented with high fever with chills and rigors. Imaging studies revealed multiple liver abscesses with hepatospleno-megaly and gallstones. Ultrasound-guided aspirate revealed pus that was negative on Gram and acid-fast staining and for amebic trophozoites. ELISA for echinococcus was strongly positive, but she did not respond to albendazole therapy. At surgery, Fasciola hepatica was detected and she responded well to bithinol postoperative. [Indian J Gastroenterol 2007;26:92-93]

Fasciola infection is endemic in North and South America, Europe, Africa, Asia and the Middle East. In India, fascioliasis is one of the most common parasitic infections of cattle; in contrast, human fascioliasis has rarely been reported. The adult worms of Fasciola hepatica primarily reside in and damage the biliary system but can occasionally involve the liver as well. In the latter situation it can cause local hepatic parenchymal necrosis, hemorrhage, and abscess formation.

A 40-year-old woman presented with complaints of high fever with chills and rigors for 4 months. In addition, she had a dull aching pain in the right upper quadrant for 3 months. There was no history of jaundice. Earlier, she was admitted elsewhere where she had received antibiotics for 2 weeks without improve-
(ranging from 5-25 mm in diameter), or as a single complex mass, as in our case. Serological diagnosis is by demonstration of fasciola antigen extracted from adult *F. gigantica* and *F. hepatica*. However, sera in many diseases, e.g., schistosomiasis, hydatidosis, echinococcosis and amebic liver abscess may cross-react with crude fasciola antigen. Partial purification of crude fasciola antigens is a suitable method to avoid cross-reactivity. The most specific test is counter immunoelectrophoresis using partially purified fasciola antigen.

**Case Snippets**

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**Pancreatic epidermoid cyst**

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Epidermoid cyst of intrapancreatic spleen is described but epidermoid cyst occurring in the pancreas itself is not documented. We report a 36-year-old man who presented with abdominal pain radiating to the back. On ultrasonography, a multilocular cyst was seen in the head of the pancreas. Fine-needle aspiration cytology suggested a diagnosis of epidermoid cyst, which was confirmed on histology of the resected specimen. [*Indian J Gastroenterol* 2007;26:93-94]

**Congenital cysts of the pancreas are rare and include a) simple or solitary cyst, b) multiple cysts associated with inherited polycystic disease, c) pancreatic and parapancratic lymphoepithelial cysts, and d) dermoid cysts.** Earlier reports of pancreatic epidermoid cysts were of cysts located in intrapancreatic accessory spleen. A 36-year-old man complained of abdominal pain radiating to the back for 2 years with acute exacerbation in the last 2 months. There was no associated vomiting. On examination a pulsatile mass was felt in the epigastrium and the left hypochondrium. Sonography showed a multilocular cystic mass in the left hypochondrium between the stomach and left lobe of the liver. The other intra-abdominal organs were normal. Guided fine-needle aspiration cytology showed flakes of keratin and groups of squamous cells, suggesting an epidermoid cyst.

Surgical exploration revealed a 2 cm x 1 cm x 1 cm mass arising from the head and body of the pancreas, adhering to the stomach. The mass was excised. Cut surface showed a multilocular cyst containing cheesy keratinous material. Its wall varied in thickness and was yellowish brown in color. Histologically, the cyst wall was partially lined by squamous epithelium and partly by markedly thickened fibrous tissue with mild chronic inflammation (Fig). There were some dilated duct-like structures lined by flattened to cuboidal cells in the wall. The cyst contained keratinous material. A diagnosis of epidermoid cyst of pancreas was made.

Epidermoid cysts occur in the 4th decade of life. They range in size from 2 cm to 6.5 cm, and arise exclusively in the intrapancreatic accessory spleen within the tail region. These lesions should be differentiated from other cystic neoplasms. According to Burrig, epidermoid cyst is a variant of mesothelial cyst with focal squamous metaplasia. However, Tateyma at al feel that epidermoid cysts might develop from pancreatic ducts protruding into accessory spleen located in the

**Fig:** Cyst lined by stratified squamous epithelium with fibrous wall (H&E, 400X)
pancreas. The present case is unique in that it is a pancreatic epidermoid cyst originating from the head of the pancreas.

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Portal hypertension associated with sickle cell disease

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We report a 12-year-old girl with sickle cell disease who presented with pain in abdomen, fever, joint pain and hematemesis. On examination she had mild jaundice and splenomegaly. Upper GI endoscopy showed esophageal varices. She was treated with variceal band ligation, and is well on folic acid supplements and propranolol. [Indian J Gastroenterol 2007;26:94]

Sequelae of sickle cell disease (SCD) include invasive infections, painful episodes, acute chest syndrome, strokes, and chronic pulmonary hypertension.

A 12-year-old girl presented to the emergency services with left upper quadrant abdominal pain, polyarthalgia, and low-grade fever for three days. There was no history of vomiting or melena. Her eight-year-old sister was diagnosed as having sickle cell disease (SS type) three years earlier.

On examination she was conscious, oriented, but irritable. Her pulse rate was 100 per minute and blood pressure 110/70 mmHg; she was pale and icteric. Abdomen was soft and non-tender. Spleen was palpable, non-tender and firm. There was no free fluid. Rest of the systemic examination was unremarkable.

Investigations: hemoglobin 6.5 g/dL; leukocyte and platelet counts were 2100 and 70,000 per cubic mm, respectively. Peripheral smear showed sickled cells; hemoglobin electrophoresis confirmed sickle cell disease SS type. She had unconjugated hyperbilirubinemia (3.7 mg/dL, conjugated 1.2), with AST 97 U/L, ALT 145 U/L, alkaline phosphatase 145 IU/L. HBsAg was negative. Chest X-ray was normal.

We treated her as SCD with vaso-occlusive and probable sequestration crisis. She was started on intravenous fluids and pentoxyfylline. On the second day she was started on oral NSAIDs for her joint pains. Her vitals remained stable and pain gradually subsided. On the third day she vomited about 1.5 liters of blood. She was shifted to the ICU and was resuscitated.

Abdominal imaging revealed splenomegaly (18 cm). Portal vein was dilated (12.5 mm); liver was normal in size and echotexture. Doppler study revealed normal inferior vena cava, and a small thrombus in the splenic vein. Upper GI endoscopy revealed grade 3 esophageal varices, and a superficial erosion in the gastric antrum. The patient underwent band ligation of varices, and was discharged on oral folic acid supplements and propranolol. She was doing well four weeks later.

SCD is seen mainly in the tribal belts of central India; our patient belonged to the same area.

SCD is remarkable for its clinical heterogeneity. The sickled cells block blood flow, resulting in lung tissue damage, pain episodes, stroke and priapism. In one report, a 16-year-old black male with hemoglobin SS disease presented with recurrent abdominal pain and hematemesis. Endoscopic examination revealed gastritis, and biopsy showed *Helicobacter pylori* infection.

In our patient, the splenic vein thrombosis leading to portal hypertension could be related to SCD. We have not found esophageal varices in any other patients with SCD in our institute.

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