Introduction
Biliary atresia (BA) is a destructive inflammatory obliterative cholangiopathy of neonates that affect varying lengths of both intrahepatic and extrahepatic bile ducts. The degree of hepatic synthetic dysfunction is variable, manifesting in varying degrees of coagulopathy which can lead to bleeding manifestation at neonatal period. A raised portal pressure has been recorded in 60-70% of affected infants at the time of first operation and biliary atresia is now second to portal vein thrombosis as the most common cause of portal hypertension and esophageal varices in children. However, bleeding from collaterals is a late manifestation in biliary atresia and most commonly occurs after 2 years of age. We present a child with biliary atresia who presented with retroperitoneal hematoma and collateral vein formation at 3 months of age which is rarely reported in literature.

Case Report
A 3month old male infant presented with jaundice associated with high colored urine and clay-colored stools since day 18 of life. He was born full term with birth weight of 2.5 kg out of non-consanguineous marriage. There was no illness in mother during pregnancy. On examination at 3 months of age his weight and length were 4.3kg and 52cm respectively. The baby had pallor, jaundice and splenohepatomegaly. Other systemic examination was normal. Investigations showed bilirubin 15.2 mg/dl with direct fraction of 7.9 mg/dl, serum glutamic oxaloacetic Transaminase (SGOT) 658 IU/l, Serum Glutamic Pyruvic Transaminase (SGPT) 416 IU/L, Gamma-glutamyl transpeptidase (GGTP) 350 IU/L, total protein-7.4 g/l, albumin - 3.5 g/l, alkaline phosphatase 254 IU/l, prothrombin time (PT) more than 2 minutes and partial thromboplastin time (PTT) more than 2 minutes. Hemoglobin was 6.6 g/dl and white cell count was 24500 cells/cumm. Thyroid function test was normal (free T3-3 pg/ml, free T4-1.47 ng/dl, TSH-2.96 microIU/ml) and cytomegalovirus (CMV) IgM was reactive with titre of 2.32 (Positive > 1.09). Ultrasound (USG) abdomen showed hepatosplenomegaly with heterogenous hyperechoic lesion measuring 6.3x2.4x3.3 cms in right psoas muscle with mild ascites and gall bladder was not visualized. CT scan abdomen was suggestive of hematoma in right iliac psoas muscle. Echocardiography was normal. The baby was taken for surgery and intra-operatively the surgeons discovered a right sided retroperitoneal hematoma with cirrhotic liver and atretic gall bladder along with common bile duct (CBD) with collaterals near porta and spleno-renal collaterals. Liver biopsy was taken and atretic gall bladder was removed. The procedure was abandoned without doing a Kasai surgery due to high risk of complications and risk of bleeding. Liver histology showed preserved architecture, widening of portal tract with bile ductular proliferation and focal areas of cholangitis. Hepatocytes showed mild ballooning with intrahepatic and intracanalicular bile stasis. Gall bladder showed flattened mucosa with sparse chronic inflammatory infiltrate in the submucosa and muscle coat. The cystic duct and distal bile duct radicle showed marked narrowing of lumen with sparse inflammatory infiltrate, suggestive of biliary atresia. (Figure1) The baby received multiple packed cells transfusions and fresh frozen plasma. He was discharged on ursodeoxycholic acid, fat soluble vitamins, cotrimoxazole and propranolol.

Discussion
Biliary atresia or neonatal obstructive cholangiopathy is characterized by fibrosclerosing obliteration of the extrahepatic bile duct. The great majority of these children develop chronic end-stage liver disease, which leads to portal hypertension and its consequences, such as massive bleeding from rupture of esophageal or gastric varices, ascites, hepato-renal syndrome, and hepatic encephalopathy. Portal hypertension has been documented in 68% of these children. In portal hypertension, portosystemic collaterals decompress the portal circulation and give rise to varices. The major sites of collaterals are: 1. rectum, where the systemic inferior mesenteric vein connects with the portal pudendal vein and results in rectal varices; 2. umbilicus, where the vestigial umbilical vein communicates with the left portal vein and gives rise

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to prominent collaterals around the umbilicus (caput medusa); 3. retroperitoneum, where collaterals communicate between ovarian vessels and iliac veins and ; 4. distal esophagus and proximal stomach, where gastroesophageal varices form major collaterals between the portal venous system and the systemic venous system. Varices form only when the hepatic vein portal pressure gradient (HVPG) exceeds 10 mm Hg and bleed only when the HVPG exceeds 12 mm Hg. Not all patients who have a HVPG greater than 12 mm Hg bleed. Larger varices at sites of limited soft tissue support, notably the gastroesophageal junction, are at greater risk for variceal rupture and bleeding in patients who have portal hypertension.

Yoshitoshi et al found that varices started at a median age of 1.6 years for patients without drainage surgery. Bleeding occurred at a mean age of 1.45 years for patients without drainage surgery and 2.98 years for patients with drainage surgery. The majority of deaths among the children in this study without drainage was before the end of the first year. As most of them didn’t submit to an endoscopy before their death, they probably lost the diagnosis of varices among them. Grunert et al found portal hypertension with hepatofugal collaterals in the first year of life. Yokoyama et al found splenoretroperitoneal shunt as early as 1 month of age associated with neonatal hepatitis. Portosystemic shunt (PSS) is most commonly associated with liver cirrhosis, but also occurs in patients without a cirrhotic liver and may be due to congenital malformation. It has been proposed that extrahepatic PSS originate with the persistence of subcardiohepatic anastomoses with the vitelline veins. The presumed basis for intrahepatic PSS is a persistent communication between the vitelline veins of the omphalomesenteric system and sinus venosus due to a focal absence of sinusoid formation. Similar findings were seen in our patient where the liver architecture was relatively preserved on histology, but the baby had lienorenal collaterals and collaterals at the porta which may have led to the retroperitoneal bleed in the baby. In our patient, though there was presence of collaterals, we were not able to establish whether the baby had developed those due to portal hypertension or were due to denovo shunts as previously reported by Yokohomo. Also, the infant had coagulopathy which could have led to the bleeding, but the baby did not have bleeding in any other area except the area of the collaterals suggesting that apart from the coagulopathy, the collateral shunts were also an important factor for the hematoma. However, the baby did have a retroperitoneal bleed suggesting that due to limited soft tissue support, these collaterals tend to bleed. This kind of porto-systemic collaterals in biliary atresia have not been reported earlier and we report this case for its rarity and its difficulty to operate for Kasai surgery.

**Conclusion**

Early collateral porto-systemic shunts in biliary atresia is a rarity and may suggest vascular malformation.

**Compliance with Ethical Standards**

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Conflict of Interest: None

**References**


