

Biliary Atresia

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Abstract

Biliary atresia is a rare neonatal condition characterized by absent or obstructed bile ducts. It can have lifelong consequences, leading to cirrhosis, liver failure, and a liver transplant. Early diagnosis is important and leads to improved outcomes. The US is often the first imaging study to image infants with conjugated hyperbilirubinemia. It can show gallbladder abnormalities, the triangular cord sign, and cysts in the porta hepatis. If biliary atresia is suspected, a liver biopsy should be performed. Once the diagnosis is confirmed, early intervention, particularly through the Kasai procedure, is essential for improving outcomes. Even with surgery, the long-term prognosis remains variable.

Keywords: neonatal, hepatobiliary, progressive

Case Summary

A 2-month-old, 26-week premature infant presented with persistent jaundice and acholic stools. At the laboratory evaluation, liver enzymes, including alanine aminotransferase, aspartate aminotransferase, gamma-glutamyl transferase, alkaline phosphatase, direct bilirubin, and total bilirubin levels, were all elevated.

Imaging Findings

US of the liver (Figure 1) revealed an absent gallbladder and common bile duct. In addition, there was a thickened, echogenic cord adjacent to the portal vein. An intraoperative cholangiogram (Figure 2) was performed. A structure in the expected location of the common bile duct was cannulated. Contrast filled the structure to the hepatic duct bifurcation. Intrahepatic ducts were not

able to be opacified, despite the injection of additional contrast.

Diagnosis

Biliary atresia

The differential diagnosis includes various conditions causing conjugated hyperbilirubinemia, including Alagille syndrome, alpha-1 antitrypsin deficiency, Toxoplasmosis, Other, Rubella, Cytomegalovirus, and Herpes Simplex virus infections, Caroli disease, choledochal cyst, idiopathic neonatal hepatitis, lipid metabolism disorders, and total parenteral nutrition-associated hepatitis.

Discussion

Biliary atresia is a rare and devastating neonatal condition characterized by progressive obstruction or absence of the bile ducts. It ultimately leads to cirrhosis and liver failure if not promptly

addressed.¹⁻³ Biliary atresia remains a significant challenge for pediatricians, neonatologists, and radiologists due to its complex clinical presentation. Early diagnosis and intervention are pivotal to improving outcomes and preventing irreversible liver damage.

The disease emerges in infancy with complete choledochal blockage and progresses to involve the intrahepatic biliary epithelium. Pathologically, the disease is associated with arterial middle layer thickening, indicative of vascular remodeling, and a decreased number of interlobular bile ducts. Hypoxia/ischemia processes correlate with biliary proliferation and medial layer thickening.^{2,4}

Distinguishing biliary atresia from other entities in the differential diagnosis requires a comprehensive diagnostic approach involving serological, radiological, and histopathological assessments. Currently, liver biopsy is used to distinguish biliary atresia from other causes of cholestatic jaundice. Findings of biliary atresia on biopsy include bile duct proliferation, bile plugging, multinucleated giant cells, focal liver parenchymal necrosis, extramedullary hematopoiesis, and inflammatory cell infiltrate. Notably, bile duct proliferation stands out as the most sensitive and specific feature among these features.⁵

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Disclosures: The authors have no conflicts of interest to disclose. None of the authors received outside funding for the production of this original manuscript, and no part of this article has been previously published elsewhere.

Figure 1. Longitudinal US image of the liver without and with color Doppler flow shows a thickened, triangular-shaped echogenic structure (arrow) adjacent to the portal vein. The gallbladder and common bile duct were not definitely identified.

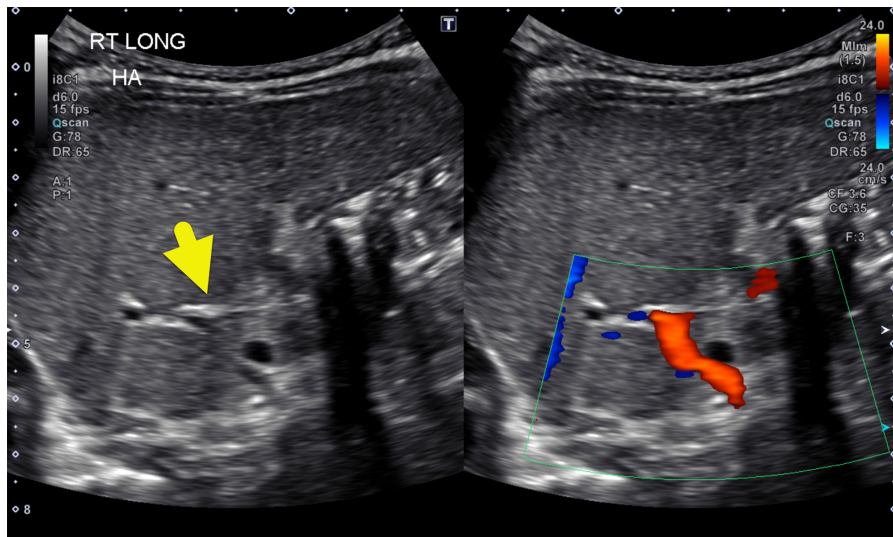


Figure 2. Intraoperative cholangiogram performed with injection at the hepatic hilum shows contrast extending towards the liver in the right upper abdomen. However, there are no visible intrahepatic bile ducts.



The US plays a crucial role in diagnosing biliary atresia. Gallbladder abnormalities, a triangular cord sign, and porta hepatis cysts are findings of biliary atresia. Gallbladder abnormalities include an absent or hypoplastic gallbladder, a fully filled gallbladder with a length <1.5 cm, the absence of smooth, complete mucosal lining, and the absence of postprandial contraction. The triangular cord sign signifies the presence of a solid proximal biliary remnant. It appears as echogenic

material located anterior to the portal vein bifurcation. This sign is recognized when the echogenic anterior wall of the right portal vein measures >2.0 mm (excluding the hepatic artery) or >4.0 mm (including the hepatic artery). A porta hepatis cyst occurs anterior to the right portal vein within the porta hepatis. In neonates with conjugated hyperbilirubinemia, findings of gallbladder abnormality or the triangular cord sign warrant surgical exploration or intraoperative cholangiography.⁶ On the intraoperative cholangiogram, there is an absence of an extrahepatic biliary system. In the absence of US findings, elastography and percutaneous cholecystocholangiography may be performed.⁷

Hepatobiliary scintigraphy is performed at some institutions using Tc99m iminodiacetic acid (IDA) analogs (HIDA scan). When performed, patients are pretreated with phenobarbital for 5 days to increase biliary secretions. Normally, the radiopharmaceutical is excreted via the biliary system and is present within the bowel. In patients with biliary atresia, there is no radiopharmaceutical within bowel loops at 24 hours. Unfortunately, this finding is not specific and can be seen in the setting of neonatal hepatitis.⁸

Because of the requirement for pretreatment and the lack of specificity, some institutions do not perform a HIDA scan. Instead, they perform a biopsy when the diagnosis of biliary atresia is suspected.³

The cornerstone of treatment remains the Kasai hepatoportoenterostomy procedure, which establishes an alternate biliary drainage pathway.⁹ Early intervention is crucial for improved outcomes, with studies showing enhanced success rates when the procedure is performed within the first 2 months of life.¹⁻³ However, despite surgical intervention, the long-term prognosis varies. Some patients achieve satisfactory bile flow and liver function, while others develop progressive liver cirrhosis necessitating liver transplantation. Currently, biliary atresia is the most common indication for pediatric liver transplantation. The need for transplantation arises when symptoms of end-stage liver disease manifest, including recurrent cholangitis, advancing jaundice, complications related to portal hypertension, ascites, diminished synthetic function, growth/nutritional deficiencies, or the development of hepatocellular carcinoma.¹⁻³

Conclusion

Biliary atresia is a rare neonatal condition characterized by absent or obstructed bile ducts. It can have lifelong consequences leading to cirrhosis, liver failure, and liver transplant. Early diagnosis is important and leads to improved outcomes. The US is often the first imaging study to image infants with conjugated hyperbilirubinemia. It can show gallbladder abnormalities, the triangular cord sign, and cysts in the porta hepatis. If biliary atresia is suspected, a liver biopsy should be performed. Once the diagnosis is confirmed, early intervention, particularly through the Kasai procedure, is essential for improving outcomes. Even with surgery, the long-term prognosis remains variable.

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