



An Unusual Cause of Cholestasis in an Infant: Biliary Atresia Type IIB

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Abstract

Keywords

- biliary atresia
- cholangiopathy
- common hepatic duct

Biliary atresia (BA) is a progressive destructive cholangiopathy of unknown etiology that presents in early infancy. It has a worldwide frequency of 1:8,000–1:15,000 and is common in Asia than in the west. Based on the level at which the lumen of the extrahepatic duct is obliterated, BA is classified into three types. Type III is the commonest (~85%) type and has the most proximal level of obstruction in the porta hepatis, while type II in which the atresia is at the level of the common hepatic duct, is the least common (~2.5%) and has been rarely reported. Here, we report the imaging features of an infant with type IIB biliary atresia.

Introduction

Biliary Atresia (BA) is an inflammatory progressive cholangiopathy that affects the intrahepatic and extrahepatic biliary tree. It is categorized into three types, of which type III in which the biliary tree is completely atretic is the most common.^{1,2} Type II is the rarest where the atresia is at the level of the common hepatic duct. Here, we describe a 4-month-old infant who presented with cholestatic jaundice. Ultrasound and MRCP were suggestive of type IIB BA that was confirmed with a percutaneous transhepatic cholangiogram (PTC) and liver biopsy.

Case History

A 4-month-old female patient presented with complaints of jaundice with acholic stools and high colored urine since day 5 of life. On general examination, she was alert and had pallor and icterus. Abdominal examination showed firm

hepatomegaly, splenomegaly, and moderate ascites. Laboratory investigation showed conjugated hyperbilirubinemia—total/direct bilirubin (14.03/9 mg/dL) and raised gamma-glutamyl transpeptidase (GGT) activity 724 (<40 IU/mL). Serology for CMV (IgM) was negative.

Clinically, a possibility of a biliary obstructive pathology was considered and the child was evaluated. Ultrasound (LOGIQS8, GE, UK) was performed after withholding feeds for 3 to 4 hours using a curvilinear (4–5 MHz) and a high-frequency linear transducer that revealed hepatomegaly (9 cm) with a coarsened echotexture and reduced echogenicity. There was a surface irregularity with a nodular outline, suggestive of chronic liver disease (CLD). There was moderate bilobar intrahepatic biliary radical dilatation (IHBRD) with inspissated secretions within the dilated biliary radicles. Gall bladder (GB) and common bile duct (CBD) were not visualized on repeated scans 24 hours apart. A triangular cord sign was evident (►Fig. 1C); however, a hepatic artery was not prominent. The subcapsular flow

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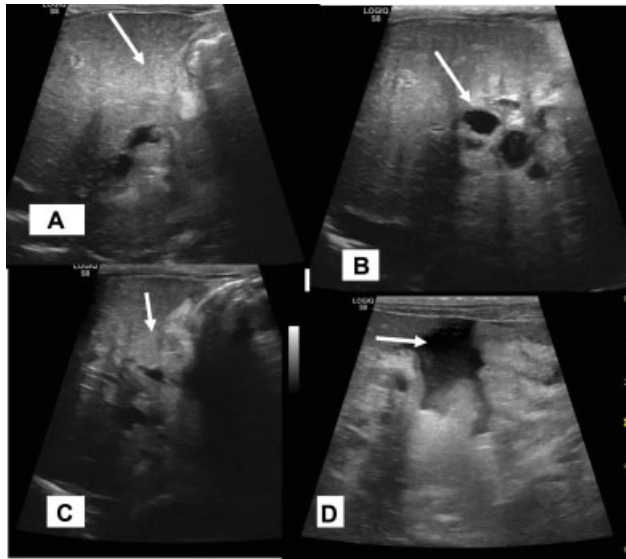


Fig. 1 (A–D) Transverse grayscale ultrasound using curvilinear and linear probes show nonvisualization of the gall bladder with empty GB fossa (arrow in A). There were dilated biliary radicles in both lobes (arrow in B) with coarsened echotexture of the liver. There was increased echogenicity seen anterior to the portal vein, and CBD was not visualized at porta, which was suggestive of a triangular cord sign (arrow in C). Free fluid was also seen in the peritoneal cavity (arrow in D). CBD, common bile duct; GB, gall bladder.

was also not evident. The spleen was enlarged and there was free fluid in the peritoneal cavity (►Fig. 1). MRI abdomen with MRCP revealed moderate central bilobar IHBRD with an abrupt cutoff at the level of the common hepatic duct (CHD). Primary confluence was patent. Cystic duct, CBD, and GB were not visible on MRCP. There was crossed fused ectopia, and the right kidney was lying on the left side below the left kidney (►Fig. 2).

As the child had features of decompensated liver disease, a surgical drainage procedure was not deemed feasible, and percutaneous transhepatic biliary drainage was done as a temporary measure to relieve jaundice through the right biliary system. Percutaneous transhepatic cholangiogram (PTC) revealed bilobar IHBRD with an abrupt cutoff at the CHD. Primary confluence was patent; however, there was no passage of contrast into distal CHD/CBD (►Fig. 3). As the stricture was not negotiable, the child was put on external drainage. In view of PTC findings, hepatobiliary iminodiacetic acid (HIDA) scan was not performed and a radiological diagnosis of BA type IIB was made. Subsequently, an ultrasound-guided liver biopsy was done. The portal tracts showed fibrous portal tract expansion and bile duct proliferation. There was the presence of intrahepatic and intracanalicular biliary cholestasis and portal to central fibrosis with nodule formation (►Fig. 4). Features were suggestive of cirrhosis secondary to BA. Upper gastrointestinal endoscopy showed small esophageal varices and the major duodenal papilla was not visualized in the duodenum.

A liver transplant was planned. Unfortunately, while undergoing evaluation, the child developed spontaneous bacterial peritonitis and died of severe sepsis.

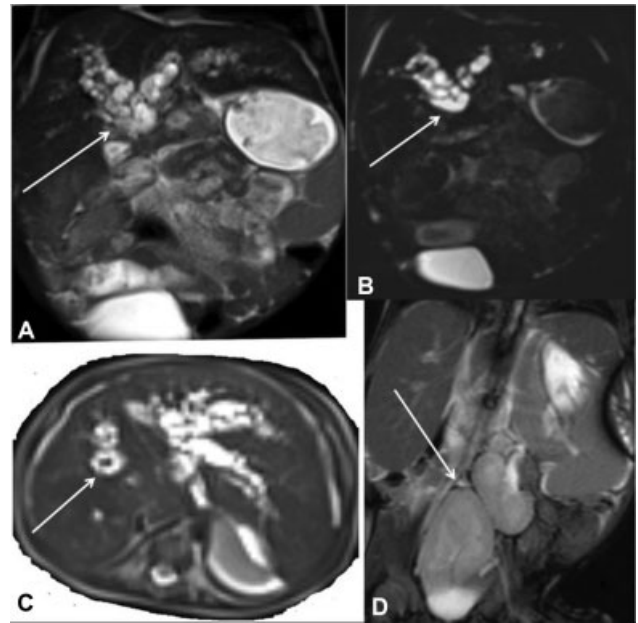


Fig. 2 (A–D) T2W FS coronal image showing bilobar intrahepatic biliary radicles dilatation (arrow in A) with patent primary confluence and abrupt cutoff at the level of the common hepatic duct (arrow in B). Cystic duct, common bile duct, and gall bladder are not visualized. T2 hypointense inspissated secretions/plugs are seen in the dilated biliary radicles (arrow in C). Crossed fused renal ectopia is also seen. The right kidney is seen on the left side at the inferior aspect of the left kidney which is also malrotated (arrow in D). T2W FS, T2-weighted fat saturated.

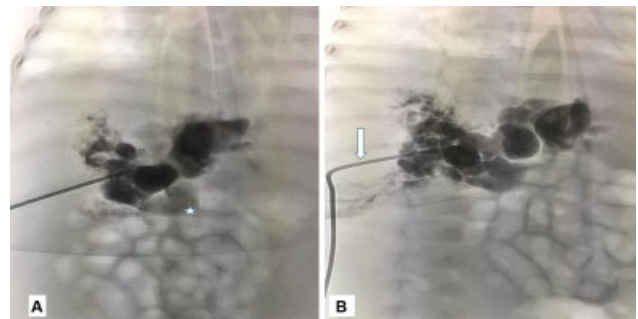


Fig. 3 (A) Percutaneous cholangiogram using the right ductal approach showing patent primary confluence with dilated bilobar ducts and abrupt cutoff at the CHD level (*). No passage of contrast seen in distal CHD, CBD, or duodenum. (B) An external drainage catheter was inserted for decompression and drainage of the biliary system (arrow). CBD, common bile duct; CHD, common hepatic duct.

Discussion

BA is a rare congenital cause of biliary obstruction. The etiology is still elusive although several viral, genetic, and immune-mediated theories have been postulated.² There is inflammatory damage to both intrahepatic and extrahepatic bile duct leading to sclerosis, narrowing, and complete obliteration of the biliary tree, which leads to progressive fibrosis of the liver and eventually cirrhosis as seen in our child. The clinical presentation consists of jaundice, acholic stools, and dark urine, and hepatomegaly. They generally

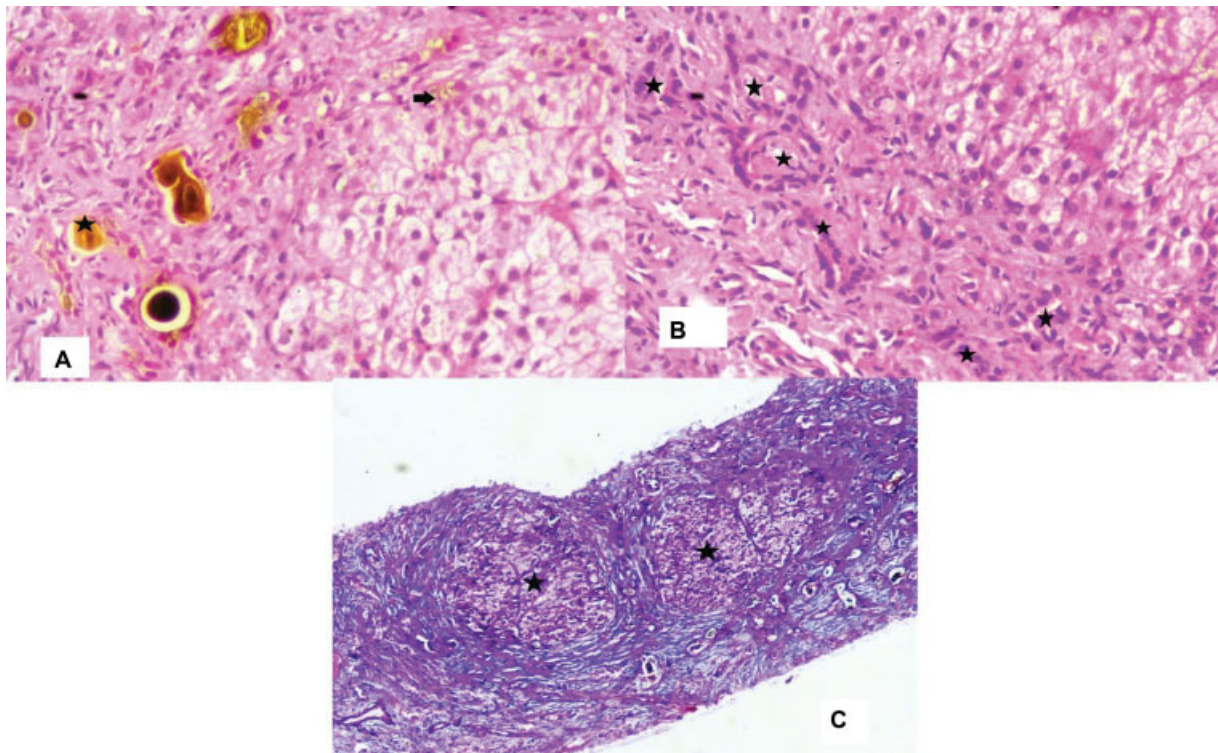


Fig. 4 (A) Microphotograph showing hepatocytes with ballooning. Degeneration (triangle). Intracanalicular (star) and intrahepatic (arrow) cholestasis is also noted. (HE x400). (B) Microphotograph showing bile duct proliferation (star). (HE x400). (C) Magnified view showing nodule formation (arrows), with surrounding fibrosis (Masson's trichrome $\times 100$). HE, hematoxylin and eosin.

appear well initially and have normal growth despite their jaundice, which leads to a delayed referral as seen in our child. Laboratory analysis includes direct hyperbilirubinemia and elevated GGT.³

Two forms of BA, syndromic or biliary atresia splenic malformation (~10%), associated with other congenital anomalies and non-syndromic (~90%) in which BA is the isolated anomaly are described.² Our child had associated crossed fused ectopia of the kidneys along with BA.

The Japanese Association of Pediatric Surgeons is the most widely used classification of BA, which is based on the level of atresia and is as follows.⁴

Type I: Atresia at the level of CBD (12%), also known as distal BA. Gall Bladder and hepatic ducts are patent. Type II: Atresia at the level of CHD (2.5%)-Rarest form. Type 2 is divided into IIA and IIB. CBD, cystic duct, and GB are patent in Type IIA, while obliterated in Type IIB.

Type III: Atresia at the porta hepatis (85%), also known as complete BA.

Diagrammatic Representation of Classification of EHBA

The described infant was Type IIB BA, which is the rarest. The most commonly encountered BA is complete or Type III BA in which there is non-visualization of bile ducts and cystic duct and gall bladder. In other subtypes, bile ducts and gall bladder can be seen depending on the level of atresia. Due to limited literature available on other subtypes, the status of biliary radicles proximal to the atresia in other types is not

described previously. In our patient, intrahepatic biliary radicles were dilated, so other differentials such as congenital stricture or web and inspissated bile duct syndrome were also considered.

Ultrasound is the preliminary investigation and requires fasting for at least 6 hours. Depending on the type, numerous sonographic findings are seen that include either nonvisualization or rudimentary GB, absence of contraction after feeding, and presence of triangular cord sign in the vicinity of the portal vein in type III BA.⁵ GB is visualized in type I and IIA. Ultrasound will also detect the changes of CLD and portal hypertension as is seen in our case. Color Doppler findings include dilated hepatic artery and increased subcapsular flow that aid in the diagnosis.⁶ Our case showed IHBRD with nonvisualization of CBD and GB and superimposed CLD, suggesting type IIB BA.

MRI of the abdomen with MRCP is done to confirm the findings, for better depiction of biliary anatomy, level of atresia, as well as to look for any other associated congenital anomaly. The sensitivity of MRCP is 90 to 100% and specificity is 77 to 96%.⁷ Nonvisualization of the extrahepatic biliary tree and GB is the diagnostic hallmark of BA; however, the demonstration of only a normal CBD also does not necessarily exclude BA, as the disease may involve only the proximal extrahepatic biliary system. In our patient CBD, cystic duct and gall bladder were not visualized, and there were dilated biliary radicles with T2 hypointense-filling defects, suggestive of inspissated secretions. These imaging features were consistent with a diagnosis of type IIB BA, which was confirmed by the percutaneous transhepatic cholangiogram

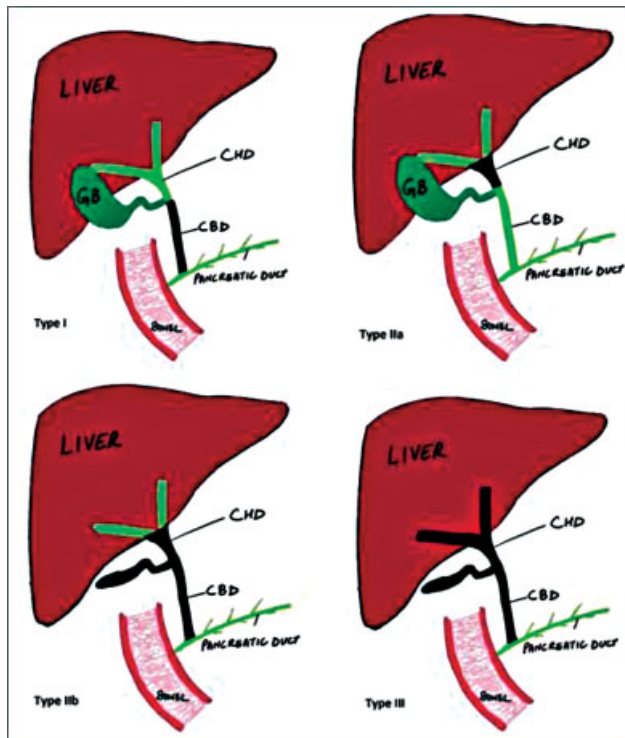


Fig. 5 Diagrammatic representation of the classification of biliary atresia.

(PTC) that demonstrated moderate dilatation of bilobar biliary radicles with a patent primary confluence. Just distal to the formation of CHD, there was an abrupt cutoff with non-passage of contrast distal to it. In children, inspissated bile duct syndrome may often mimic BA, and PTC cannot only help in establishing the diagnosis by showing ductal filling defects but an attempt at therapeutic lavage also can be made.⁸ A congenital stricture or web of the common hepatic duct is another rare condition that may present with features of biliary obstruction; however, the early clinical presentation and aggressive clinical course with features of established cirrhosis by 4 months of age would make it unlikely. An absent GB with non-visualization of the web, which is seen as a linear-filling defect on MRCP ruled out the diagnosis of the congenital web on imaging.⁹ The diagnosis of BA was further corroborated by the liver biopsy findings, which had a diagnostic accuracy of 90.1% for BA.¹⁰ A Kasai portoenterostomy is the treatment of choice; however, in children who

present late with features of advanced cirrhosis, as seen in our child, one has to proceed directly for liver transplantation. We planned for the same but unfortunately, the child died of sepsis while undergoing evaluation for the same (►Fig. 5).

To conclude, we describe the multimodality imaging features of a child with Type IIB BA which is the rare type. Various imaging findings of Type III EHBA, which is the commonest type, are described previously; however, the imaging findings of type IIB have not been described previously to the best of our knowledge.

Funding

None.

Conflict of Interest

None declared.

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