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NEONATAL IMAGING

Neonatal diagnosis of biliary atresia: a practical review and update

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Abstract
Biliary atresia is challenging to diagnose because many of the clinical and imaging features of this condition overlap with those of other causes of cholestasis in newborns. When jaundice persists beyond 2 weeks of age, the neonate should be evaluated for cholestasis, and biliary atresia — the most common cause of neonatal cholestasis — should be considered. It is critical to diagnose biliary atresia early because failure to treat can result in hepatic fibrosis and death in less than 1 year. In this paper, we review the current diagnostic imaging methods, differential considerations and treatment options for biliary atresia.

Keywords Bile duct · Biliary atresia · Cholestasis · Infants · Neonates · Magnetic resonance imaging · Ultrasound

Introduction

Newborn, persistent cholestatic jaundice caused by hepatobiliary dysfunction is a relatively common condition, occurring in 1 in 2,500 live births [1]. Cholestasis is the result of reduced bile formation or flow, which causes bile to remain within the liver rather than be excreted. When jaundice and conjugated hyperbilirubinemia persist for more than 2 weeks after birth, several potential causes must be considered [2]. The most common causes include biliary atresia (25–40%) and genetic disorders (25%); less commonly, the condition might be related to parenteral nutrition, idiopathic or multifactorial causes [1]. Neonatal hepatitis is another potential cause of conjugated hyperbilirubinemia and jaundice, typically caused by neonatal infections, metabolic disorders or genetic disorders [1]. In this article, we address biliary atresia, the most common cause of neonatal cholestasis.

Biliary atresia: background

The biliary system develops primarily during the first trimester in utero. The first anlagen of the bile ducts and liver begin developing at 3–4 weeks and continue development for up to 8 weeks. Finely tuned interactions between the epithelium and mesenchymal tissues allow for the appropriate development and remodeling of the biliary system, forming a double cell layer that leads to the formation of the ductal plate. The ductal plate extends from the hilum of the liver toward the periphery along the portal vein branches [3, 4]. The ductal plate consists of portal veins surrounded by hepatic cell precursors, which remodel and form the intrahepatic biliary tree. Several conditions are linked to aberrant formation of the ductal plate, including biliary atresia, biliary cysts, Caroli disease, biliary hamartomas and congenital hepatic fibrosis (as seen in autosomal-recessive polycystic kidney/liver disease [ARPKD] and autosomal-dominant polycystic kidney disease [ADPKD]) [4].

The incidence of biliary atresia varies by international region, ranging from 1 in 6,000 live births in Taiwan to 1 in 19,000 live births in Canada. In the United States, the incidence of biliary atresia is approximately 1 in 12,000 live births [1]. The etiology of this condition is unknown but is thought to be related to genetic defects in the formation of the ductal plate and bile ducts or to in utero ductal inflammation from viral or autoimmune causes [1]. Biliary atresia leads to progressive fibrosis and destruction of the intrahepatic and extrahepatic biliary ducts, resulting in obliterative cholangiopathy [5]. Numerous patterns of biliary atresia have been described [6, 7]. The most common pattern, seen in approximately 66% of patients, involves complete atresia of the extrahepatic duct and gallbladder [6]. Also noted are cystic forms of biliary atresia with a large cyst at the liver hilum as well as differing

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degrees of atresia of the gallbladder and bile ducts. All types of biliary atresia involve abnormal intrahepatic bile ducts [6].

Clinically, children with biliary atresia might present with jaundice, acholic (pale) stools and dark urine, and they are more commonly female [8]. Nonsyndromic and syndromic forms of biliary atresia exist; nonsyndromic is the more common form, occurring in 80% to 90% of patients [6, 9]. The less common syndromic form is associated with heterotaxy, congenital heart disease, polysplenia, a preduodenal portal vein, and inferior vena cava (IVC) anomalies [6].

Diagnosing biliary atresia is challenging because no single method provides a definitive diagnosis. Additionally, clinical, laboratory and imaging features of this condition overlap with those of other causes of cholestasis. Results of liver function tests are commonly abnormal in these children; in a study by Robie et al. [2], as well as others [10], an elevated level of serum gamma-glutamyl transpeptidase (GGT) was demonstrated to be strongly associated with biliary atresia. Abnormal GGT levels are useful, but alone they are not diagnostic of biliary atresia. Additionally, a new biomarker, matrix metalloproteinase-7 (MMP-7), has demonstrated promising results, with >95% sensitivity and specificity for biliary atresia, and might become useful in the diagnosis [11]. Currently, screening for biliary atresia typically includes measurement of serum GGT levels as well as abdominal US and other imaging modalities.

Imaging

Abdominal US is the primary imaging modality used to evaluate biliary atresia and biliary abnormalities. Nuclear medicine hepatobiliary iminodiacetic acid (HIDA) scans and magnetic resonance (MR) imaging/MR cholangiopancreatography (MRCP) are also used, although less commonly. The gold standard for diagnosing biliary atresia is an intraoperative cholangiogram with concurrent liver biopsy. CT is typically not useful for diagnosing biliary atresia except in occasional complicated cases.

Ultrasound

Initial imaging for infants with biliary atresia is typically performed with abdominal US. This modality is portable, widely available and effectively demonstrates the biliary system.

The gallbladder in infants with biliary atresia might be small, measuring less than 15–19 mm when measurements are obtained in the fasted state [6]; a healthy gallbladder should measure 15–30 mm in infants younger than 1 year and should have a thin, defined wall [7, 8]. It is important to ensure the child has been fasting 3–4 h before US to minimize possible obscuring bowel gas and allow full distention of the gallbladder [5]. Nonvisualization of the gallbladder is a finding with 100% specificity for biliary atresia [7]; however, nonvisualization does not confirm a diagnosis of biliary atresia, and so in this instance, further evaluation should be performed. Other gallbladder abnormalities include the "gallbladder ghost triad" or "pseudo gallbladder," which refers to a small gallbladder (<19 mm) with an indistinct wall and irregular contour (Fig. 1) [6, 12, 13]. In addition to an absent or abnormal, small and irregular gallbladder, features of biliary atresia on US might include the following: triangular cord sign, nonvisualization/absence of the common bile duct (CBD), presence of microcysts or macrocysts near the porta hepatis, increased hepatic artery diameter >1.5 mm, peripheral arterialization and polysplenia. Measurement of the hepatic artery can be challenging because of its small size; therefore, a non-oblique image with a high-frequency US probe is important [6]. Measurements can be obtained from the mid anterior wall to the mid posterior wall of the right proximal hepatic artery where it is parallel to the right portal vein. According to Kim et al. [14], when hepatic artery diameter measures 1.5 mm or greater, there is 92% sensitivity, 87% specificity and 89% accuracy for biliary atresia. The physiological basis for hepatic artery enlargement in biliary atresia remains unknown; however, hypotheses include compensatory blood supply, vascular malformation and possible association with cirrhosis [14].

The triangular cord sign refers to a fibrous remnant of the obliterated biliary duct adjacent to the anterior right portal vein wall, demonstrated on US as echogenic thickening of greater than 3–4 mm near the bifurcation of the main portal vein in the transverse plane (variable because there is no consensus on the best measurement site in the literature) [6, 15] (Fig. 2). The echogenic thickening, which might be tubular or triangular in

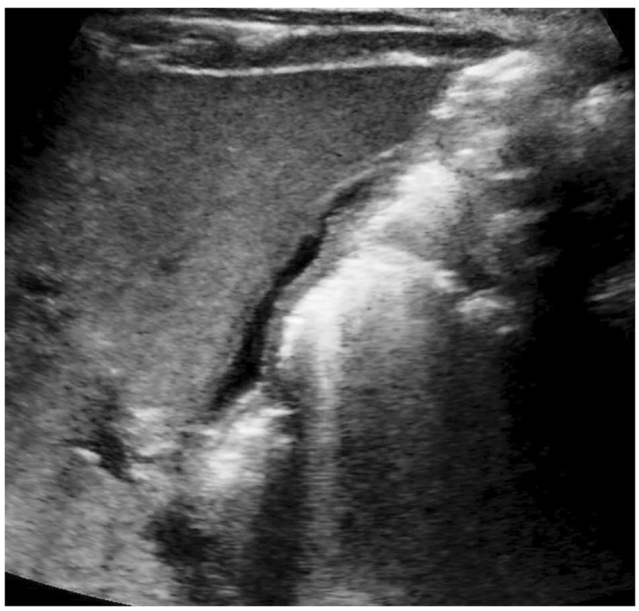

Fig. 1 Gallbladder ghost triad in a 6-week-old girl with conjugated bilirubinemia and biliary atresia. Transverse US image demonstrates a thin, long and irregular gallbladder, the gallbladder ghost triad

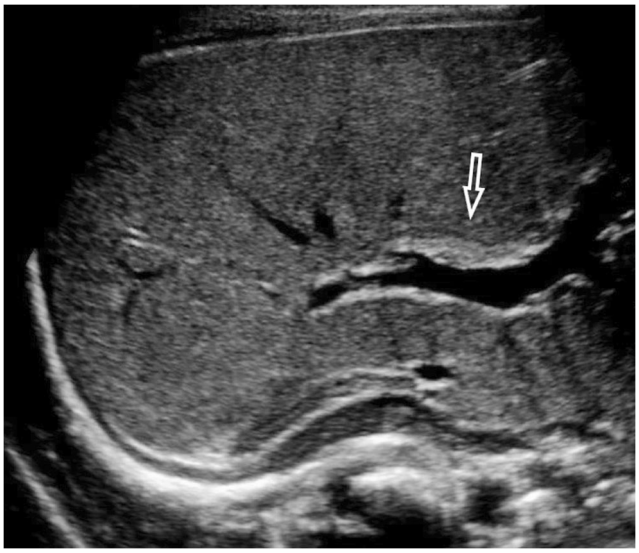

Fig. 2 Triangular cord sign in a 4-week-old boy with biliary atresia. Transverse US image demonstrates echogenic thickening near the bifurcation of the portal vein (*arrow*), compatible with the triangular cord sign

configuration on US images, can be difficult to identify in the early stages of the disease because of the plane of imaging and evolution of the finding over time [15–17]. When identified, the combination of triangular cord sign and a gallbladder abnormality is 84% sensitive and 100% specific for biliary atresia [6].

Other imaging findings of biliary atresia include microcysts and macrocysts. The presence of cysts near the porta hepatis, in the absence of a normal CBD, is a poorly sensitive but highly specific indicator of biliary atresia. Microcysts measure approximately 2–3 mm and should be evaluated with Doppler

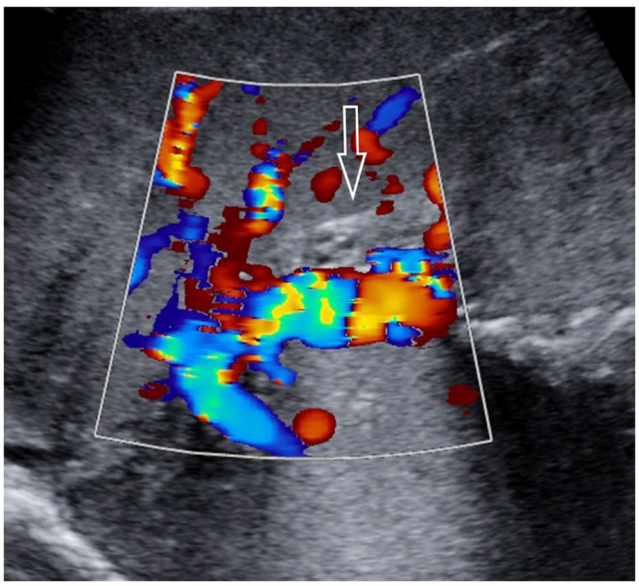

Fig. 3 Microcysts. In a 3-month-old girl with cholestasis and biliary atresia, transverse Doppler US image demonstrates microcysts (*arrow*) at the porta hepatis measuring 2 mm

imaging to confirm they are cysts rather than small vessels (Fig. 3). Macrocysts are defined as cysts greater than 5 cm in diameter without biliary duct dilation [7] (Fig. 4). A large cyst within the hepatic hilum in a child with biliary obstruction and cholestasis could be caused by cystic biliary atresia with the primary differential diagnosis of a choledochal cyst [7].

Polysplenia seen with cholestasis has a low sensitivity (10%) but high specificity (100%) for biliary atresia. This is seen in the less common syndromic form of biliary atresia in 4–14% of cases and is typically found in combination with other sonographic signs such as intestinal malrotation, preduodenal portal vein, absent inferior vena cava (IVC), aberrant hepatic artery and abdominal heterotaxy [5–7].

Sonographic elastography (acoustic radiation force impulse [ARFI]) can be a helpful tool in the evaluation of neonatal cholestasis because this modality offers a noninvasive, low-cost evaluation of liver stiffness or fibrosis, an indirect indicator of biliary atresia. ARFI might demonstrate marked abnormality early in life without hepatic parenchymal gray-scale changes, thus providing another tool for diagnosis. Shear wave measurements and liver stiffness values with ARFI might be higher in children with biliary atresia than in those with other causes of liver disease, such as Alagille syndrome and neonatal hepatitis [18, 19]. In a study by Leschied et al. [18], mean hepatic shear wave speeds were shown ranging from 1.9–2.3 m/s in a biliary atresia group versus 1.1–1.4 m/s in a non-biliary atresia group.

Nuclear medicine

Hepatobiliary iminodiacetic acid scans can be used to assess the anatomy and function of the biliary system, allowing for evaluation of the liver's ability to excrete bile. In these scans, a hepatobiliary agent is injected, and the radiotracer is taken up by the hepatocytes to be cleared and excreted along with the bile. When no radiotracer is excreted within 24 h, this is suggestive of but not definitive for biliary atresia with a sensitivity of 98.7% and specificity of 70.4%, and further evaluation should be considered [20] (Fig. 5). Other etiologies with poor hepatocellular function and biliary excretion (as in neonatal hepatitis) can also lead to a lack of excretion of radiotracer into small bowel. Pretreatment with ursodeoxycholic acid or phenobarbital for several days increases biliary enzyme secretions through the stimulation of hepatocytes [8, 17] and can help prevent false positives. If radiotracer excretion into the small bowel is visualized within 24 h, biliary atresia is excluded (Fig. 5).

Magnetic resonance imaging

Magnetic resonance imaging and MRCP are less commonly used to evaluate biliary atresia because of the potential need for sedation with MR, and the effectiveness of US in

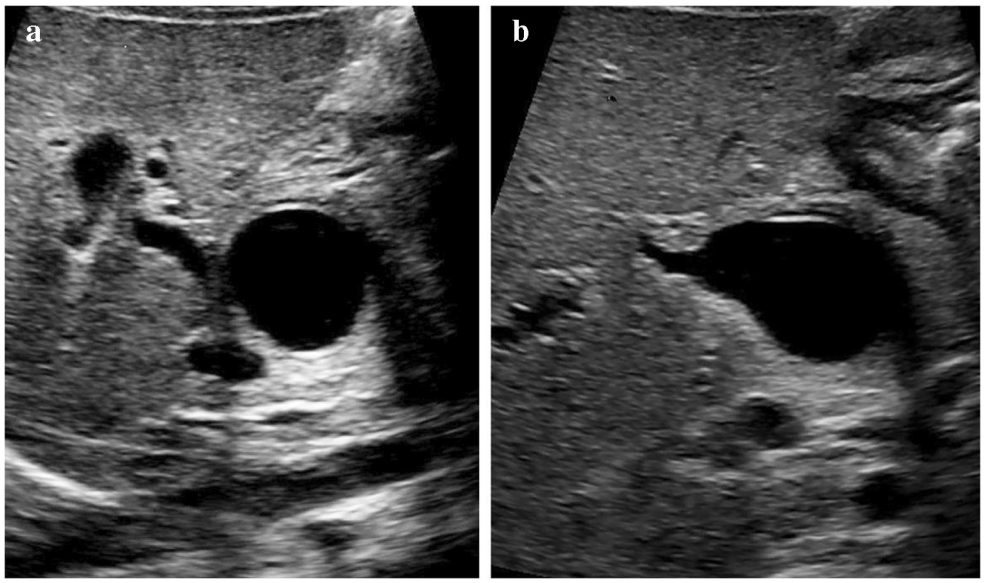

Fig. 4 Macrocysts in a 3-week-old girl with biliary atresia. **a** Transverse US image at the hepatic hilum demonstrates a large cyst (macrocyst) with absence of a normal gallbladder. **b** Longitudinal US image at the hepatic hilum demonstrates a large cyst (macrocyst) with absence of a normal gallbladder. Both cases courtesy of Dr. Rama Ayyala, Cincinnati Children's Hospital Medical Center

evaluation of the biliary system. When MR and MRCP are used, they can demonstrate absence of the extrahepatic duct, presence of an abnormal gallbladder, and the MR equivalent of the triangular sign — triangular cord thickness. On MR imaging, the triangular cord thickness measurement is taken at the site of greatest thickness on MR axial T2 images rather than anterior to the right portal vein, as on US. Triangular cord thickness should measure greater than 5.1 mm in abnormal cases [21]. Nonvisualization of the CBD on MR images can also indicate biliary atresia. Alternatively, if the extraheptic CBD is visualized on MRCP, biliary atresia can be excluded. One study found that the combination of an abnormal gallbladder and nonvisualization of the CBD on MR images was indicative of biliary atresia in 100% of cases [15, 21]. Most commonly, however, MR imaging is used to exclude other causes of cholestasis rather than as the primary diagnostic modality for biliary atresia.

Intraoperative cholangiogram and liver biopsy

Cholangiogram is the gold standard for the diagnosis of biliary atresia. This procedure can be performed by laparotomy or percutaneously. When possible, contrast agent should be injected into the gallbladder. Findings indicative of biliary atresia include absence of the gallbladder lumen, presence of a small atretic gallbladder or absence of contrast agent in the

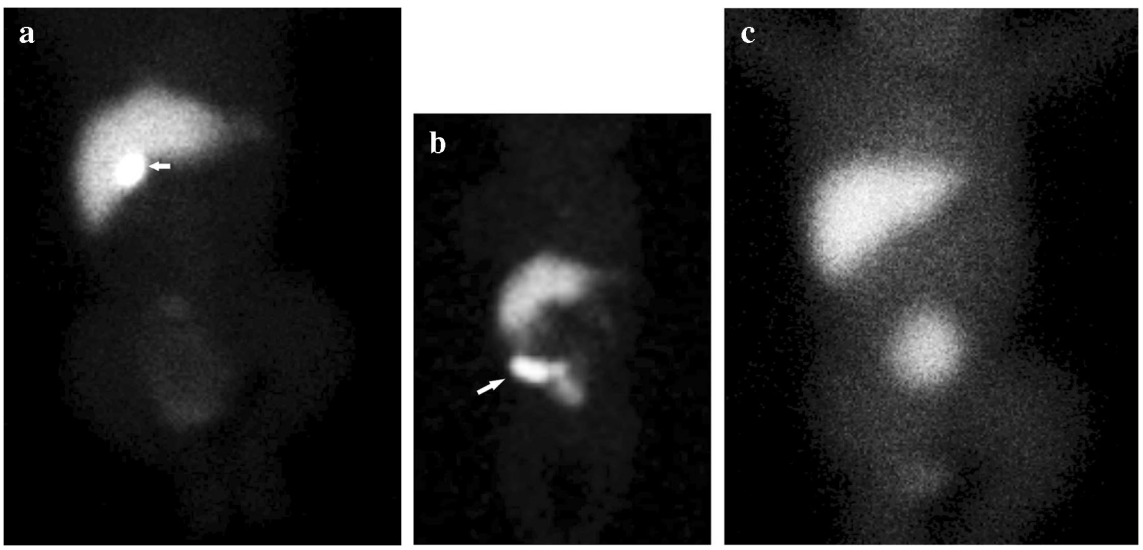

Fig. 5 Hepatobiliary iminodiacetic acid (HIDA) scan to assess the biliary system. **a** In a 6-week-old girl with cholestasis, normal HIDA scan at 4 h shows expected radiotracer uptake in the gallbladder (*arrow*). **b** Normal HIDA scan at 24 h, with radiotracer uptake now evident in the duodenum (*arrow*) in the same girl. **c** In a different girl, 7 weeks old with biliary atresia, HIDA scan at 24 h demonstrates absence of radiotracer uptake in the gallbladder or small bowel

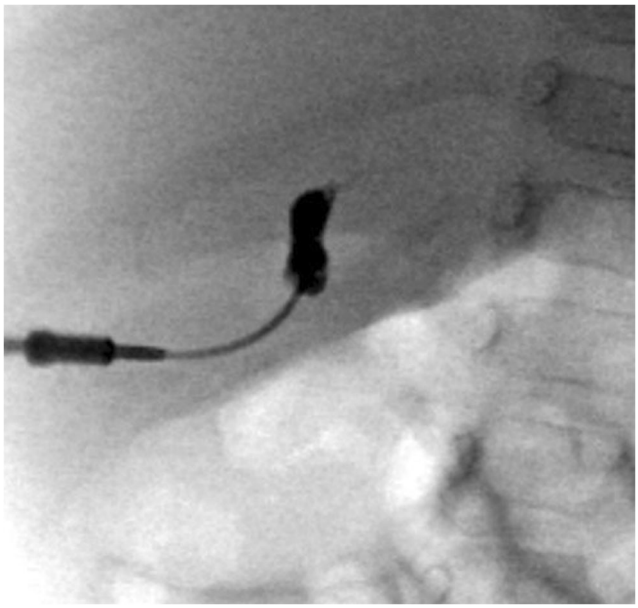

Fig. 6 Single anteroposterior image from an intraoperative cholangiogram demonstrates absence of biliary duct filling and an atretic gallbladder filled with contrast agent in an infant with confirmed biliary atresia. Case courtesy of Dr. Greg Tiao, Cincinnati Children's Hospital Medical Center

biliary ducts and small bowel (Fig. 6) [8, 22]. Liver biopsy can be performed in the operating room or by US guidance. Findings on pathological analysis include bile duct proliferation from biliary obstruction, bile plugs, a paucity of bile ducts and possible ductal plate malformations [23].

Differential considerations

The differential diagnosis for cholestasis is primarily based on clinical and laboratory findings, with imaging findings aiding in the evaluation. Choledochal cyst is the primary imaging differential consideration for the cystic variants of biliary atresia. Cystic biliary atresia is a rare form of biliary atresia, occurring in 5–10% of cases, and it is difficult to distinguish from choledochal cyst on prenatal imaging [1, 24]. Choledochal cysts are congenital dilations of the bile ducts. The cause of these cysts is unknown; possible etiologies include ductal plate malformation or an anomalous junction between the pancreatic duct and distal CBD, resulting in reflux of pancreatic enzymes leading to inflammation, weakening, stricture and dilatation of the biliary system (Fig. 7) [8, 25]. In children with cystic biliary atresia, US and MRCP demonstrate an atretic or irregular gallbladder, usually small cysts at the hepatic hilum, no intrahepatic ductal dilation, and a lack of sludge or stones [8]. In contradistinction, choledochal cysts are large and demonstrate intrahepatic ductal dilation.

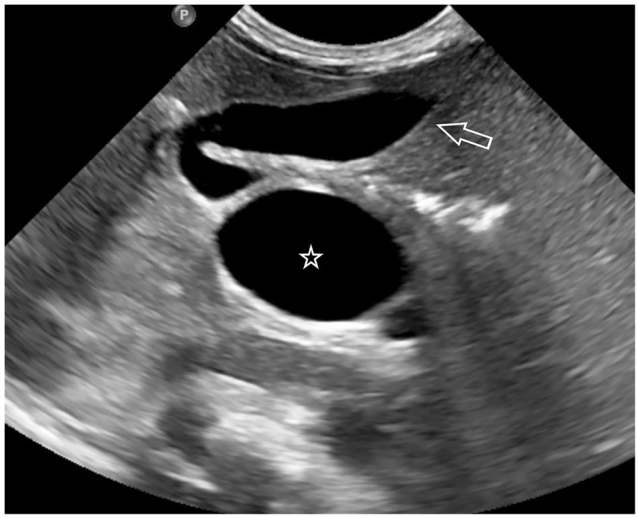

Fig. 7 Choledochal cyst. Transverse US image demonstrates a normal gallbladder (*arrow*) with an adjacent large choledochal cyst (*star*) in a 1-day-old boy

Causes of cholestasis other than biliary atresia include multisystem diseases such as Alagille syndrome and cystic fibrosis, and hepatocellular diseases such as alpha-1 antitrypsin deficiency, infantile sclerosing cholangitis, Byler disease and other forms of progressive familial cholestasis [5]. Alagille syndrome is an autosomal-dominant genetic disorder in which affected children have a paucity of intrahepatic bile ducts with other characteristic clinical criteria, such as cardiac and facial abnormalities. This disorder can be diagnosed based on clinical findings, imaging findings, laboratory results and confirmed with genetic testing. The imaging findings for Alagille syndrome are nonspecific, but affected children might demonstrate a normal gallbladder, a lack of ductal dilation on abdominal US and vertebral segmentation anomalies on conventional radiographs (Fig. 8) [5, 26]. Cystic fibrosis might present with liver enzyme abnormalities, as a result of mild biliary obstruction,

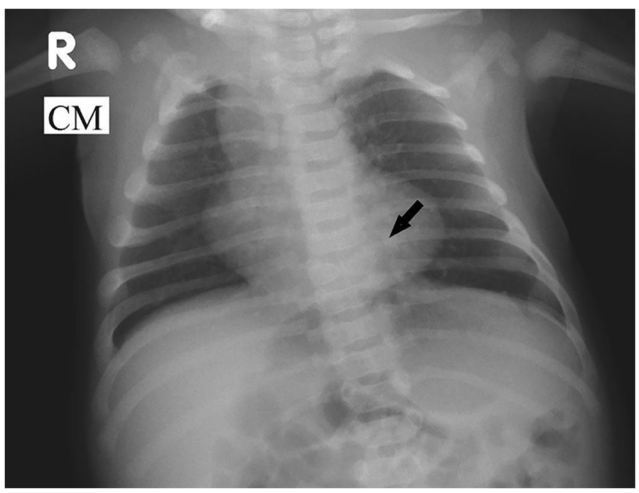

Fig. 8 Alagille syndrome in a 2-month-old girl. Anteroposterior radiograph of the chest demonstrates a butterfly vertebra of the mid thoracic spine (*arrow*)

and might demonstrate nonspecific heterogeneous liver parenchymal changes and cirrhosis on US. These imaging findings typically occur later in childhood or adulthood [18]. Alpha-1 antitrypsin deficiency is the most common inherited cause of neonatal cholestasis; these children might demonstrate absence of biliary excretion on HIDA and, in some cases, features compatible with cirrhosis on abdominal US [1]. Infantile sclerosing cholangitis is a rare idiopathic disorder that can result in inflammatory biliary fibrosis involving the intrahepatic and extrahepatic ducts. MRCP in these children might reveal beading and irregular ductal dilation. Byler disease and other forms of progressive familial cholestasis, inherited conditions in which cells in the liver cannot release bile, also present clinically with cholestasis. On US imaging, Byler disease and other forms of progressive familial cholestasis might present with a distended gallbladder and increased periportal echogenicity, which can be confused with the triangular cord sign seen in biliary atresia [12].

Bile plug syndrome is another differential consideration for cholestasis and jaundice in the neonate; this condition is caused by mechanical obstruction of the biliary ducts by inspissated bile or sludge. This obstruction can result from the use of total parenteral nutrition, administration of diuretics, prematurity, the presence of congenital heart disease, cystic fibrosis or hemolysis. US imaging findings in these children include sludge in the gallbladder or ducts, gallbladder wall thickening and biliary obstruction. These children can be treated conservatively, with biliary lavage, retrograde cholangiopancreatography or percutaneous drainage (Fig. 9) [5, 8].

Treatment

Early treatment of biliary atresia is essential. A treatment delay of more than 60 days can result in accelerated hepatic fibrosis and liver failure; if not treated, the child can die within 1 year of birth [1]. The primary treatment for biliary atresia is a Kasai hepatic portoenterostomy with resection of the biliary remnant and creation of a Roux-en-Y intestinal anastomosis at this site to restore some bile flow. If, during intraoperative cholangiogram and biopsy, no open ducts are identified, the surgeon will perform the Kasai procedure. The bile formed in the liver can then drain directly into the small bowel. If this procedure is performed within 60 days after birth, approximately 70% of infants will have bile flow; if the procedure is performed more than 90 days after birth, less than 25% of infants will have resultant bile flow [1]. Complications of the Kasai procedure that might be seen on follow-up imaging include recurrent cholangitis, intrahepatic cysts, Roux loop obstruction, biliary leak and anastomotic narrowing [8].

If the Kasai procedure fails or cirrhosis develops, the infant will require a liver transplantation. Biliary atresia is the most common indication for liver transplantation in children [22]. Although not currently the standard of care, some centers attempt revision of the Kasai procedure in select infants to delay transplantation; others do not, because they believe that attempted revisions can later complicate liver transplants because of the presence of adhesions [8, 27]. Indications for transplantation in these infants include cirrhosis, liver failure, gastrointestinal bleeding from portal hypertension, growth retardation, pruritus, hepatopulmonary syndrome and repeated cholangitis (Fig. 10). Research has shown that performing the Kasai procedure followed by liver transplantation in infants with liver failure leads to improved survival; however, early failure of the Kasai procedure is associated with increased mortality after liver transplantation [28].

Conclusion

Biliary atresia can be challenging to diagnose because it has many clinical and imaging features that overlap with those of

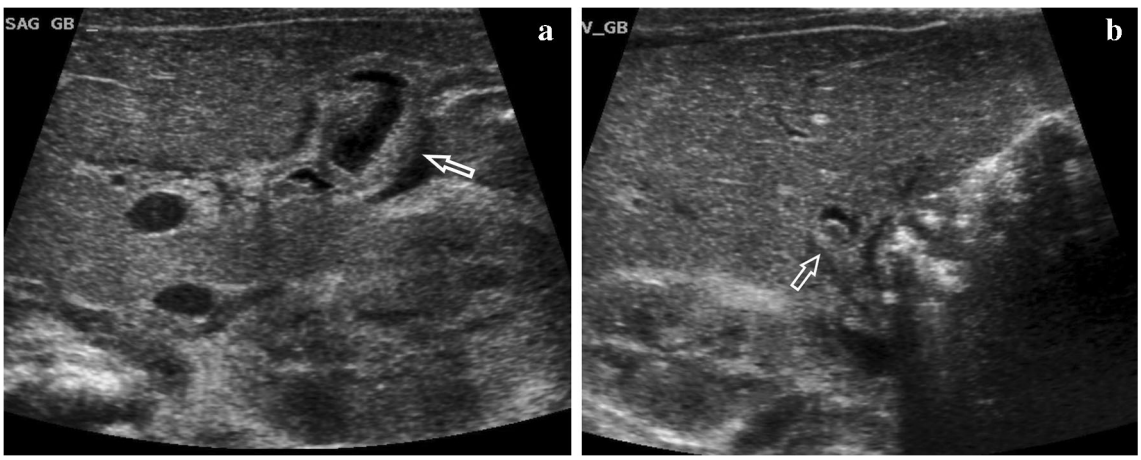

Fig. 9 Bile plug syndrome in a 6-week-old girl with direct bilirubinemia. **a** Sagittal US image shows a thickened gallbladder wall (*arrow*). **b** Transverse US image shows a sludge ball in the gallbladder neck (*arrow*)

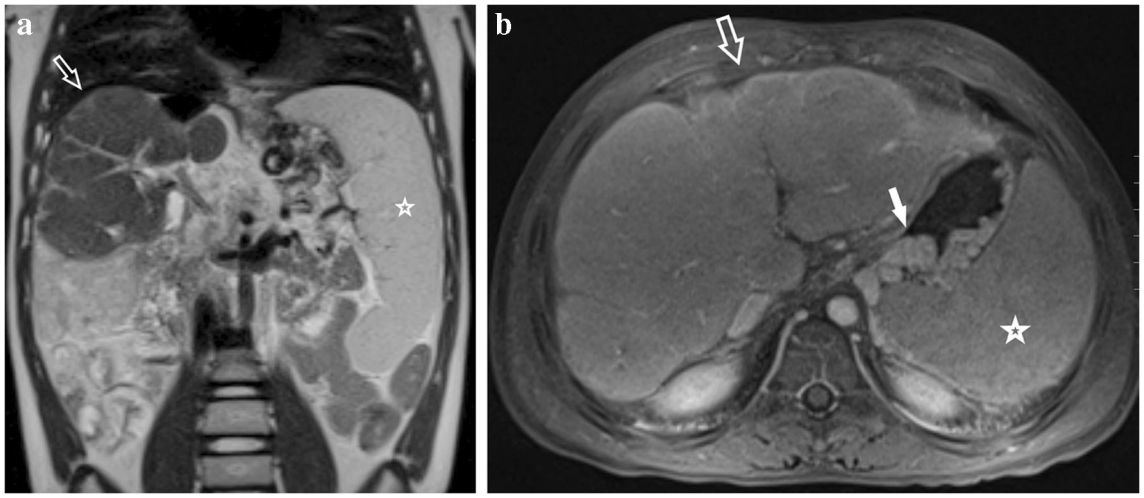

Fig. 10 Imaging in an 8-year-old boy with history of biliary atresia, post Kasai procedure. **a** Coronal half-Fourier acquisition single-shot turbo spin-echo (HASTE) MR image demonstrates a shrunken and nodular liver (*arrow*), compatible with cirrhosis, and splenomegaly (*star*). **b** Axial volumetric interpolated breath-hold examination (VIBE) post-contrast MR image further demonstrates the shrunken and nodular liver (*open arrow*), compatible with cirrhosis, along with splenomegaly (*star*) and varices (*closed arrow*)

other disorders that cause neonatal jaundice and hyperbilirubinemia. Biliary atresia must be identified and treated early to have a good outcome. In the evaluation of the neonate with persistent cholestasis, US is the primary imaging modality. Sonographic ARFI, hepatobiliary scintigraphy, and MRI/MRCP can be used to provide additional diagnostic information. When clinical and sonographic findings are suggestive of biliary atresia, intraoperative cholangiogram and liver biopsy remain the gold standard for diagnosis.

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Declarations

Conflicts of interest None

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