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Biliary atresia

What is biliary atresia?

Biliary atresia is a condition of uncertain cause where part, or all, of the extrahepatic bile ducts are obliterated by inflammation and subsequent fibrosis, leading to biliary obstruction and jaundice. It is fatal if untreated.[1]

A viral aetiology has been proposed although the association with other congenital anomalies in some cases suggests a possible developmental abnormality.

How common is biliary atresia? (Epidemiology)

- Biliary atresia occurs in 1 in 8,000–18,000 live births and accounts for the most paediatric liver transplants worldwide.[2]
- Approximately 20% have co-existing congenital anomalies, most commonly involving the heart, abdomen and genitourinary tract. There may be associated situs inversus or polysplenia/asplenia with or without other congenital anomalies.[3]

Biliary atresia symptoms[2] [4]

The natural history of biliary atresia starts with a latent preclinical phase with abnormalities that are often unrecognised, such as high fractionated bilirubin levels. Visible symptoms of cholestasis develop after the first weeks of life, followed by rapidly progressing liver injury and end-stage liver disease in the first year of life.

- Clinically, biliary atresia presents with persistent jaundice, pale stools and dark urine in term infants with normal birth weights. All term infants who remain jaundiced after 14 days (and preterm infants after 21 days) should be investigated for liver disease, initially with simple measurement of the conjugated fraction of bilirubin.

- Normal meconium is passed initially and the stools may be bile-coloured for a short period afterwards, but pale stools are the rule. Screening with a stool colour card may be a cost-effective and simple screening method for biliary atresia in neonates.[5]
- Splenomegaly is not usually a feature unless presentation is late (aged more than 3 months) and it is thus a sign of portal hypertension.
- Failure to thrive is a result of poor absorption of long-chain fats and the catabolic state.

Classification

This is according to the site of atresia in the extrahepatic biliary system:

- Type I: common bile duct atresia with patent proximal ducts.
- Type II: common hepatic duct atresia with cystic structures in the porta hepatis.
- Type III: right and left hepatic duct atresia to the level of the porta hepatis (most common).

Investigations[6]

- LFTs are abnormal with a conjugated hyperbilirubinaemia. Gamma-glutamyltransferase (GGT) is usually higher in biliary atresia than in other causes of neonatal cholestasis. Serum cholesterol might be raised but triglycerides are within the normal range.
- Ultrasound is recommended as the initial imaging strategy. Ultrasound and hepatobiliary scintigraphy (technetium-^{99m}) can be used to help differentiate atresia from neonatal hepatitis, intrahepatic biliary hypoplasia and extrahepatic obstructive lesions.
- Liver histology (obtained by percutaneous biopsy) is the usual diagnostic method of choice.[7]
- Endoscopic retrograde cholangiopancreatography to visualise the biliary tract is occasionally needed when the diagnosis is unclear, but it is technically difficult in infants and use is confined to large centres.

Differential diagnosis

- Other causes of obstructive jaundice - choledochal cyst, cholelithiasis and spontaneous perforation of the bile duct can all occur in the neonatal period.
- Cystic fibrosis.
- Lipid storage disorders.
- Idiopathic neonatal hepatitis.
- Congenital infections.
- Alpha-1-antitrypsin (A1AT) deficiency.

Biliary atresia treatment

The Kasai procedure and liver transplantation represent the only therapeutic options for patients with biliary atresia.[8]

Surgical

- Provided there is no cirrhosis and the patient presents early, the primary treatment for biliary atresia is the Kasai portoenterostomy or one of its variants.
- In the unmodified operation the atretic extrahepatic tissue is removed and a Roux-en-Y jejunal loop anastomosed to the hepatic hilum. It may restore bile flow and clear jaundice.
- Kasai portoenterostomy directly connects the intestines to the liver to restore bile flow. A critical factor predicting outcomes after this procedure is the time at which the operation is performed. Before 30-45 days of life provides the greatest chance of delaying or avoiding liver transplant.[2]
- Portoenterostomy remains as the first-line operative treatment in biliary atresia while liver transplantation serves as a salvage treatment when portoenterostomy fails or liver function gradually deteriorates after initially successful establishment of bile flow.[9]
- Early liver transplantation appears to be beneficial in cases with an available liver for transplantation.[10]

Complications

- Ascending cholangitis can develop in the first few months after surgery, with recurrence of jaundice, acholic stool and abdominal pain. Sometimes sepsis is severe and requires resuscitation and intensive care.
- Recurrent or late cholangitis might suggest an obstruction of the Roux-en-Y loop as it passes through the mesocolon. Bile lakes can develop in the liver at any time after surgery and could be a source of recurrent infection.
- Cirrhosis, portal hypertension and liver failure.[11]
- Hepatocellular carcinoma.
- Osteomalacia or biliary rickets.

Prognosis[12]

Lower degree of biliary fibrosis, bile ductular proliferation, absence of ductal plate malformation, large ducts more than 150 µm and younger age were found to be associated with better long-term outcome.

The long-term survival with native liver is significantly lower; supporting liver transplantation is the preferred choice for biliary atresia management. Adult outcome studies in biliary atresia patients quote the survival with native liver at 20% in adults 20 years post Kasai, and 10% among those who are 30 years post Kasai.

Further reading

- British Liver Trust
- Children's Liver Disease Foundation
- Lendahl U, Lui VCH, Chung PHY, et al; Biliary Atresia - emerging diagnostic and therapy opportunities. EBioMedicine. 2021 Dec;74:103689. doi: 10.1016/j.ebiom.2021.103689. Epub 2021 Nov 12.

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