CHAPTER 81
BILIARY ATRESIA

Sanjay Krishnaswami
Richa Lal
Katrine Lofberg
Alaa Fayez Hamza

Introduction

Biliary atresia (BA) is a neonatal disease characterised by the inflammatory and sclerotic obliteration of part or all of the extrahepatic biliary tree, with varying involvement of the intrahepatic bile ducts. Although seen in only a small percentage of jaundiced neonates, this disorder is one of the more common structural causes of neonatal cholestatic jaundice.

If left untreated, biliary atresia is almost uniformly fatal. Hepatic failure, infection, or bleeding secondary to portal hypertension causes death by the age of 1 to 2 years in the vast majority of patients.[1,2] In the current era, despite the diagnostic difficulties and technical challenges that BA poses, early recognition and the proper performance of a Kasai portoenterostomy procedure can be life saving. When available, liver transplantation can serve as the ultimate therapy for those who have end-stage disease.

Demographics

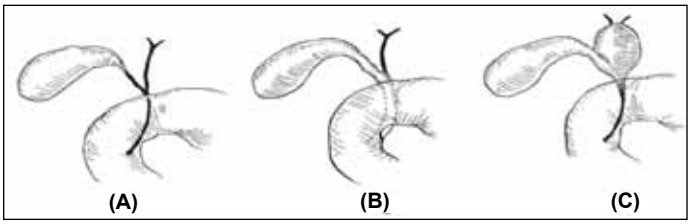
Based mostly on Western series, the incidence of biliary atresia is between 1:8,000 and 1:17,000 live births, with an overall female preponderance of up to 1.7:1.[3] In most of Africa, incidence data are not available, but individual institutional reports suggest that up to five children with this disorder are encountered yearly in many centres.[4–6] Therefore, although cholestasis secondary to infectious causes is more common, BA is encountered fairly frequently in Africa.[4,7,8] No specific genetic factors are associated with the disease, but associated congenital malformations occur in 11–20% of the cases.[3] The most commonly associated anomaly is polysplenia; other associated anomalies include intestinal atresias, abdominal situs inversus, malrotation, and genitourinary and cardiac anomalies.

Aetiology/Pathology and Classification

Many aetiologies for the disease have been proposed, including genetic factors, congenital developmental causes such as a failure of recanalisation or antenatal ischaemia, and viral or other infectious causes. None have been proven, however, and the pathogenesis remains unknown. Because two groups of patients are affected—those with other congenital anomalies and those with a late foetal or perinatal anomaly that appears to occur in isolation—there may in fact be an interplay of these various aetiologies in the development of BA in any individual patient.

Microscopic examination of the biliary system reveals fibrosis of the ductules with varying degrees of inflammation. Early in the disease, bile duct proliferation, biliary plugs, and mild portal fibrosis are present. Later in the disease, this fibrosis extends intrahepatically and will manifest as bridging fibrosis of the biliary structures.

Classification of biliary atresia is generally based upon the macroscopic location of the fibrotic biliary cord remnants (Figure 81.1). The most common type is complete fibrosis of the entire extrahepatic biliary system, seen in up to 70–80% of cases. Gallbladder and common bile duct patency with obliteration of the porta and hepatic duct is the next most frequent (12–20%). Absence of portions of the biliary system with fibrosis of the remaining portions as well as distal fibrosis with proximal hilar cyst variants can also be seen. Macroscopically, these mucus-containing cyst variants can be confused for a fibrotic hilar cone or for a choledochal cyst. As these cysts do not communicate with the biliary tree, all such structures need to be resected. Failure to recognise this variant can lead to a nonfunctional anastamosis with the sequestered cyst.[3]



Source: Karrer FM, Pence JC. Biliary atresia and choledochal cyst. In: Ziegler MM, ed. Operative Pediatric Surgery. McGraw-Hill, 2003. Reproduced with permission from the McGraw-Hill Companies.

Figure 81.1: Illustration of common types of biliary atresia: (A) complete extrahepatic biliary obliteration (most common, affecting about 80%); (B) proximal obliteration, distal patency; and (C) common hepatic duct cystic dilatation with portal and distal common bile duct obliteration.

Clinical Presentation

Most children with biliary atresia appear normal at birth but become increasingly jaundiced after 3 to 6 weeks of age. Additionally, in those children with physiologic jaundice from birth that does not resolve spontaneously, BA should be suspected because persistence of jaundice after 2 to 3 weeks of life should always be considered pathologic until proven otherwise. This sign, coupled with the findings of progressively acholic stools and dark urine, is suggestive of BA.

As the liver becomes progressively obstructed, it grows in size and firmness. Malabsorption of fat-soluble vitamins resulting from hepatic obstruction and failure can lead to diarrhoea, anaemia, and malnutrition. Overall growth of the infant may therefore appear normal in the first few weeks or months after birth, but most patients develop failure to thrive once liver failure is more significant. Prolonged bleeding can be seen from the umbilical stump in such cases. Finally, splenomegaly, bleeding oesophageal varices, and other signs of portal hypertension can also be significant parts of the examination in patients who present with advanced illness.[9] By recent African reports, up to two-thirds of patients present late in the illness after advanced signs of liver failure are present.[10]

The value of the experienced paediatrician in recognising the jaundiced baby with persistently acholic stools cannot be overstated. It is important to recognise these and other physical findings early because successful outcomes after surgery for biliary atresia are tied closely to age at performance of Kasai portoenterostomy.[11] Note, however, that the physical signs and symptoms of BA described above overlap with myriad other causes of jaundice in infancy (discussed in the next section), and none of these findings should be considered as conclusive for BA. Rather, early recognition of signs of persistant jaundice should prompt swift referral to centres capable of completing the diagnosis and instituting definitive surgical intervention, if warranted.

Investigation

The differential diagnosis of neonatal jaundice is prodigious and includes such surgically correctable obstructive lesions as biliary atresia and choledochal cyst as well as inborn metabolic errors, congenital infectious causes, and other causes, as listed here:

- *physiologic:* immaturity of glucuronyl transferase (resolves quickly);
- *breast milk feeding*;
- *hematologic:* Rh/ABO blood group incompatibility, hemolytic diseases (spherocytosis);
- *infectious:* the TORCH (TOxoplasmosis, Rubella, Cytomegalovirus (CMV), Herpes) complex; syphillis, hepatitis, and others;
- *genetic/metabolic:* α1-antitrypsin deficiency, galactosaemia, tyrosinaemia, cystic fibrosis (CF), hypothyroidism, Gaucher's disease, iron storage disease;
- *hepatocellular dysfunction:* Gilbert's disease, Crigler-Najjar syndrome;
- *neonatal hepatitis*;
- *total parenteral nutrition (TPN)-related cholestasis*; and
- *extrahepatic processes:* BA, choledochal cyst, Alagille syndrome (arteriohepatic dysplasia), inspissated bile plug syndrome, bile duct stenosis or stricture, spontaneous perforation of the bile duct.

Testing

The main goal of early testing is to rapidly differentiate jaundice due to obstruction from that due to other causes. A proper combination of serology testing, imaging, and biopsy prior to or during an operative cholangiogram leads to an accurate diagnosis in >95% of patients.[12] Conventional serum liver function tests are nonspecific. Hyperbilirubinaemia, usually in the 5–12 μg/L range, is typical of the early stage of the disease. However, bilirubin fractionation is not useful in distinguishing obstructive jaundice due to BA from the more common intrahepatic cholestatic diseases (e.g., neonatal hepatitis) because there is elevation of indirect, unconjugated as well as direct, conjugated bilirubin in both diseases. Similarly, alanine transaminase (AST) and aspartate transaminase (ALT) are also often elevated in both cases, although extreme elevations of these are unusual in BA. In contradistinction to this, alkaline phosphatase and gamma-glutamyl transpaptidase (GGT) are often very elevated, and low levels of these may suggest an alternate diagnosis. Alterations in hepatic function tests, such as prothrombin time, partial thromboplastin time, and albumin, are typically seen only in advanced cases of BA. Serum testing should also be done to screen for both inborn metabolic errors and classic infectious causes such as TORCH and syphilis, among others. An exhaustive hunt to rule out all these possibilities can take many weeks, so one should not wait for the results of testing prior to imaging, which may help confirm or eliminate the possibility of an obstructive, mechanical cause of jaundice.[1,9]

Imaging

The two most useful imaging modalities in further differentiating surgically amenable causes from other causes of cholestasis are ultrasound (US) and nuclear hepatobiliary imaging. Although operator-dependant, US is available in many African centres and offers a safe and noninvasive way to evaluate the jaundiced neonate. It can be used to assess the size of the gallbladder and intra- and extrahepatic biliary ducts and to visualise gallstones. Although the presence of a cystic extrahepatic or intrahepatic ductal dilatation typically rules out BA, the disease is suspected if there is a very small or absent gallbladder, extrahepatic ducts are not at all visualised, or the cone-shaped fibrotic portal plate is seen.[13] The presence of polysplenia or pre-duodenal portal vein lends further support to a diagnosis of biliary atresia. Doppler use helps to correctly interpret the adjacent hepatic artery as a vascular rather than a biliary duct structure.

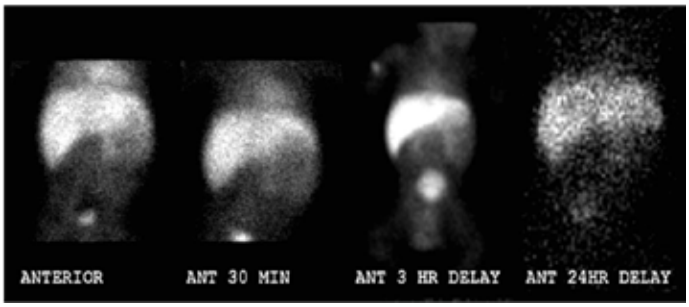


Figure 81.2: Nuclear hepatobiliary scan demonstrating prompt uptake of technecium tracer with no excretion into the gut after 24 hours. This image is suspicious for biliary atresia, but could still be consistent with neonatal hepatitis. Excretion into the gut at any time rules out biliary atresia.

Nuclear imaging is performed with intravenous administration of technecium-99m, which is taken up by the liver. In neonatal hepatitis, there is delayed technecium uptake due to hepatocellular dysfunction and delayed or absent excretion into the gut. In BA, uptake is usually prompt (especially early in the disease) but there is no gut excretion (Figure 81.2). Therefore, visualisation of tracer in the intestine rules out BA, whereas absence of tracer visualisation in the intestine could be due to either process. In resource-poor locations without nuclear imaging techniques, it may be judicious to proceed straight to either percutaneous biopsy or operative biopsy and cholangiogram via a laparoscopic or open technique.

Management

If, after initial investigation, the diagnosis of BA is still suspected, prompt operative intervention is warranted to definitively determine whether the patient has BA and, if so, to perform a Kasai portoenterostomy procedure.

Preoperative Considerations

In addition to routine preoperative care for any abdominal procedure, a dose of vitamin K (1 mg/kg) can be given several days prior to surgery. Coagulation factors (prothrombin time/international normalization ratio/partial thromboplastin time, or PT/INR/PTT) should be checked to ensure suitability for operation, and type-specific blood should be available. Routine bowel preparation is not necessary, but a dose of preoperative broad-spectrum antibiotic is given prior to incision.

Initial Intraoperative Considerations: Cholangiogram and Liver Biopsy

Traditionally, the operation commences with a small right upper quadrant transverse or oblique incision. However, if available, laparoscopy can also be utilised for the initial portion of the procedure. If a gallbladder is visualised and is patent, an operative cholangiogram is the next manoeuvre, wherein the tip of the gallbladder is cannulated with an angiocatheter and contrast is instilled under fluoroscopy. Contrast in the duodenum and continuity with the intrahepatic ducts rules out BA (Figure 81.3)

If biliary atresia is ruled out and a biopsy has not previously been done, a generous wedge biopsy of the liver is performed and the incision(s) is (are) closed. An "adequate" liver biopsy specimen that has at least five portal tracts, reported by an experienced pathologist done after 4–6 weeks of age, should have an overall accuracy of 96%. The biopsy not only helps to differentiate BA from neonatal hepatitis, but also points towards specific aetiologies of neonatal hepatitis as well as α_1-antitrypsin deficiency and storage disorders such as Niemann-Pick disease. In addition to being sent for pathologic exam, a portion of the biopsy specimen should be sent for viral and bacterial cultures.

If no gallbladder or a fibrotic gallbladder is present, or if cholangiogram reveals a lack of either the proximal or distal extrahepatic ducts (Figure 81.4), the right upper quadrant incision should be widened or the procedure converted to open from laparoscopic in preparation for hepatic portal exploration and the Kasai procedure.

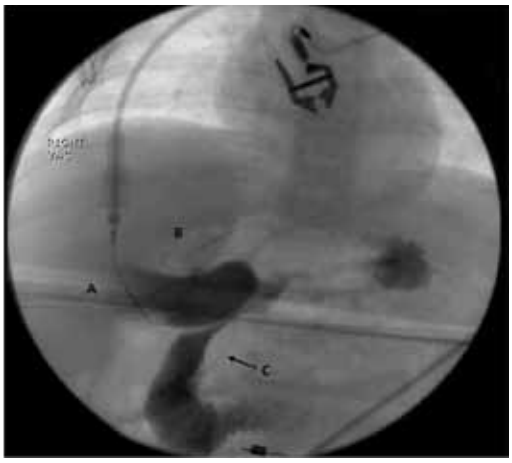


Figure 81.3: Intraoperative cholangiogram with contrast filling hypoplastic proximal and distal duct system with emptying into duodenum: (A) contrast-filled catheter within gallbladder; (B) left and right hepatic ducts; (C) intrapancreatic common bile duct. Patient had Allagille syndrome, not biliary atresia.

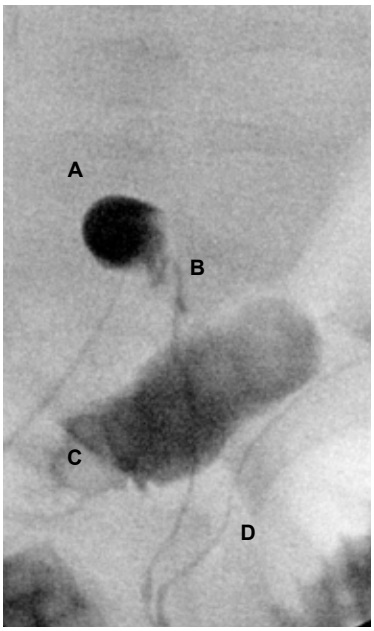


Figure 81.4 : Intraoperative cholangiogram demonstrating emptying of contrast into the duodenum with no proximal duct filling (proximal biliary atresia): (A) contrast being administered through a small but patent gallbladder; (B) stenotic but patent common bile duct passing intrapancreatic and emptying into the duodenum; (C) contrast in the duodenum; (D) retrograde filling of the pancreatic duct.

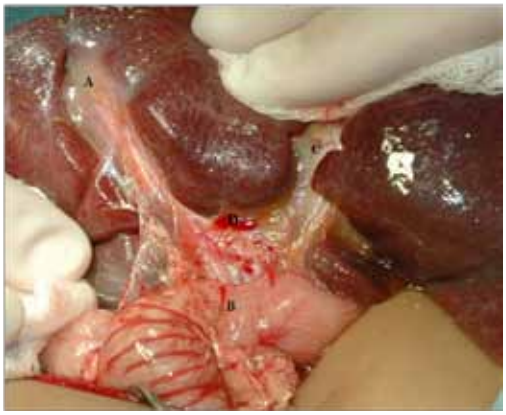


Figure 81.5. Biliary atresia: view of porta hepatis and hepatoduodenal ligament prior to dissection: (A) fibrotic gallbladder; (B) duodenum; (C) falciform ligament; (D) approximate location of fibrotic endplate.

Kasai Roux-en-Y Portoenterostomy

The portoenterostomy procedure, first described by Kasai and Suzuki in 1959,[14] uncovers patent biliary microductules proximal to the level of extrahepatic biliary fibrosis and allows these structures to drain directly into a segment of defunctionalised intestine.

The procedure begins with separation of the gallbladder from its liver bed down to the junction of the cystic and common bile ducts.

1. The peritoneum over the hepatoduodenal ligament is opened, exposing the biliary and hepatic arterial structures (Figure 81.5). The fibrous common duct is dissected and transected at its margin with the duodenum.

2. The entire gallbladder and fibrous common duct is placed on traction in a superior direction. The cystic artery is ligated, being careful to definitively identify it from the right hepatic artery.

3. The ductal remnants are then dissected from the adjacent hepatic artery and portal vein until the right and left branches of the portal vein are identified (Figure 81.6).

4. Further dissection reveals the widening of the fibrotic duct into a cone-shaped mass entering the liver superiorly. This fibrotic hepatic endplate is then transected at the point at which it is flush with the liver edge. In order to identify this level, small branches of the portal vein to the central portion of the fibrotic endplate must often be divided such that the most superior dissection is actually behind the portal vein. (Figure 81.7).

5. If there is discontinuity of the proximal biliary tree, meticulous exploration over the bifurcation of the portal vein will lead to identification of the fibrotic cone. Once the endplate is divided, cautery on this portion of the liver should be avoided, and a moist gauze should be placed here while attention is turned to the construction of a Roux-en-Y intestinal limb. The completed portal dissection is shown in Figure 81.8.

6. A 40-cm Roux-en-Y limb is constructed by dividing the bowel 10–15 cm after the ligament of Treitz. The distal cut edge is advanced in a retrocolic fashion up to the level of the liver. A handsewn end-to-end or side-to-side jejunojejunostomy re-establishes intestinal continuity. The mesenteric defect at the jejunojejunostomy is closed, and the Roux limb is tacked to the colonic mesentery at its retrocolic window to prevent internal hernia and excessive tension on the portoenterostomy (Figure 81.9).

7. The portoenterostomy is performed in a single layer, end-to-side or end-to-end fashion, using long-term absorbable sutures in an interrupted or running fashion, being careful to take meticulous full thickness bites of only the periphery of the porta hepatis (Figures 81.10 and 81.11). This ensures that the portoenterostomy incorporates the entire portion of tissue that may contain biliary channels and that risk of microductular structure compromise is minimised.

8. The completed portoenterostomy is shown in Figures 81.12 and 81.13. A closed-suction drain can be placed posterior to the portoenterostomy, and the abdomen is closed in layers.

To limit potential postoperative cholangitis, many modifications of the original portoenterostomy have been proposed over the years since Kasai first described the technique. These have included initial externalisation of the biliary conduit via a stoma and an antireflux intussusception valve within the Roux limb, among others. However, as none of these have been successful in preventing cholangitis, and in fact have been associated with unique complications of their own, their use should be discouraged.

Postoperative Considerations

Nasogastric drainage is continued for several days after the operation until gut function resumes. At that time, an oral diet is given as tolerated. Perioperative intravenous antibiotics, such as a cephalosporin, are administered by most surgeons for at least the first 12–24 hours. Many

surgeons administer long-term oral antibiotics (e.g., trimethoprim/sulfa, 2 mg/kg per day) as well as choleretic agents, such as ursodeoxycholic acid (10–15 mg/kg per day) upon discharge in the hope of lessening the incidence of cholangitis following a Kasai procedure.[11]

Steroids are purported to benefit biliary function after a Kasai procedure through stimulation of salt-independent bile flow in addition to their marked anti-inflammatory, immunosuppressive, and scar-limiting properties. They were originally proposed for use along with broad-spectrum antibiotics in cases of postoperative cholangitis and in cases of sudden cessation of bile flow in a previously well-functioning portoenterostomy.[1,14] More recently, the routine use of steroids in the postoperative period has become standard for some surgeons. Although its use remains controversial, especially in light of reports showing equivocal benefits, this practice is widespread. The evidence for steroids comes from multiple studies reporting improved short- and long-term outcomes after standard postoperative steroid use as reflected by decreased bilirubin level several months after the Kasai procedure, decreased mortality at one year postoperatively, and improved jaundice-free survival with native liver at four- to five-year follow-up.[15–17] Although many of these studies involved small numbers of patients and many potentially confounding variables exist, none of the studies noted significant adverse reactions from steroid use, such as infectious complications or wound issues. The optimal dose and length of steroid treatment to achieve positive effects is unknown and varied across the studies cited. One author of this chapter prefers a three-week tapering course of oral prednisone (2 mg/kg per day) beginning at hospital discharge.

Complications

Although the usual operative complications, such as severe bleeding and anastomotic leak, can occur, these are rare—even in patients with fairly advanced disease. The most common issues postoperatively are cholangitis, nutritional deficiencies, and portal hypertension.

Cholangitis occurs in one-third to two-thirds of patients; its incidence is highest within the first several years after a Kasai procedure. It is thought that bacteria in the Roux-en-Y conduit and bile stasis combine to cause this problem. The onset of cholangitis is heralded by fever, leukocytosis, and an increase in bilirubin. Most cases are responsive to supportive treatment with fluids and early institution of broad-spectrum antibiotics covering intestinal flora. In refractory cases, one can consider a short burst of intravenous corticosteroids, as previously described. If not already being used prior to the first episode of cholangitis, chronic oral suppressive antibiotics, ursodeoxycholic acid, or oral steroids may be of benefit to prevent recurrent episodes.[18] Reoperation to prevent cholangitis (by creation of an antireflux valve or lengthening of the Roux limb) is generally unsuccessful.[3]

Portal hypertension is the most serious delayed complication, seen in up to 50–70% of long-term survivors.[19] It can occur even in patients who initially had a successful Kasai procedure and usually manifests as ascites, variceal bleeding, or hypersplenism. Treatment with diuretics, beta blockers, or variceal banding and splenectomy or splenic embolisation can be successful. However, without the possibility of liver transplantation, many of these patients will succumb to this portal hypertension.

Severe nutritional deficiency can accompany liver disease secondary to BA. Without adequate bile flow for intestinal fat absorption, children can develop essential fatty acid deficiency. Until adequate bile flow is achieved, it is prudent to administer formula feeds with a high percentage of medium-chained triglycerides that can be absorbed directly without the assistance of bile salts. Additionally, affected neonates can develop deficiencies of the fat-soluble vitamins (K, E, A, and D), which can result in diseases such as rickets (vitamin D deficiency) or severe coagulopathy (vitamin K deficiency). Monitoring vitamin levels and/or supplementation of these vitamins as available can prevent significant morbidity.

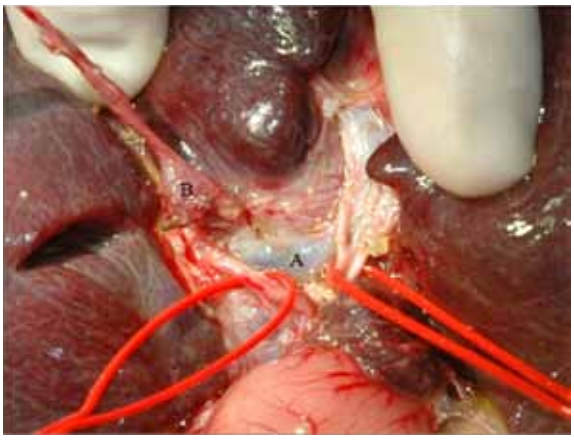
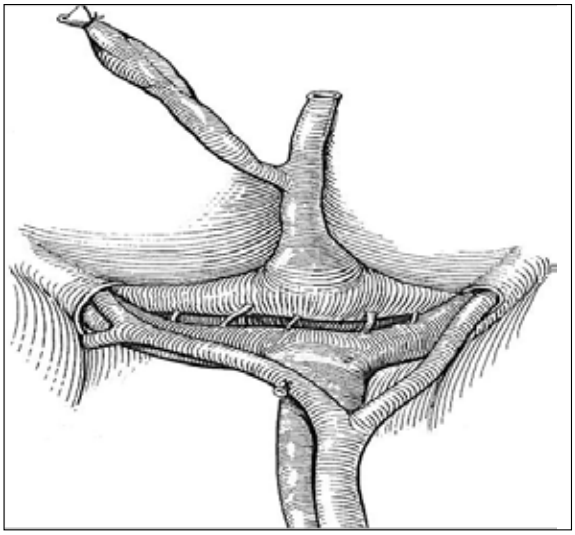


Figure 81.6: Porta after isolation of hepatic arteries and partial dissection of fibrotic endplate vessel loops around hepatic arteries: (A) portal venous confluence; (B) fibrotic ductal structures (medial portion of fibrous endcone already divided and reflected laterally).



Source: Karrer FM, Pence JC. Biliary atresia and choledochal cyst. In: Ziegler MM, ed. Operative Pediatric Surgery. McGraw-Hill, 2003. Reproduced with permission from the McGraw-Hill Companies.

Figure 81.7: Fibrotic extrahepatic bile ducts with branches of portal vein to endplate.

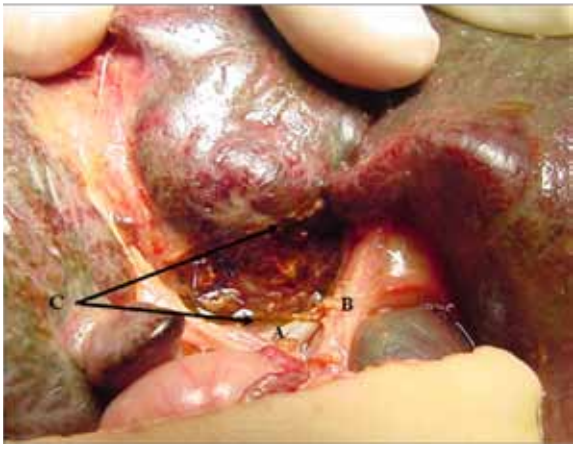


Figure 81.8: Intraoperative photo of completed dissection of porta prior to portoenterostomy in another patient: (A) portal vein confluence; (B) right hepatic artery; (C) portal plate after resection of fibrous cone; arrows mark anterior and posterior edges.

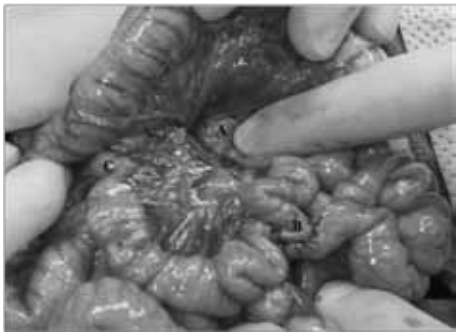


Figure 81.9: Completed jejunojejunostomy of Roux limb: (A) proximal jejunum at ligament of Treitz; (B) end-to-side jejunojejunostomy; (C) Roux limb to liver passing retrocolic, secured to transverse colon mesentery.

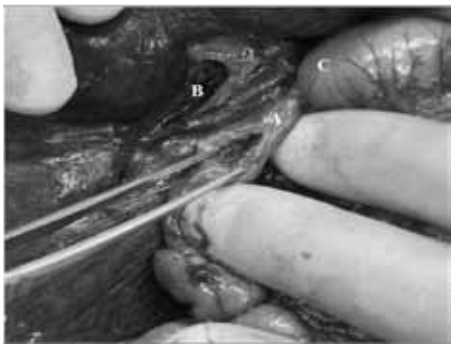


Figure 81.10: Distal Roux limb (superior to its retrocolic passage) being opened on its antimesenteric aspect in preparation for end-to-side anastomosis with porta: (A) side of Roux limb; (B) portal endplate; (C) pylorus posterior and adjacent to tip of Roux limb.

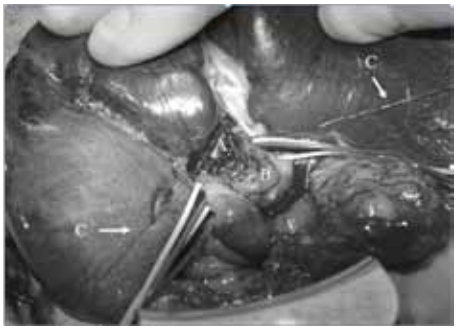


Figure 81.11: Posterior row of portoenterostomy anastomosis complete. Vessel loops around portal venous and hepatic arterial branches posterior to Roux limb: (A) inferior border of portal plate; (B) mucosal edge of jejunum ready for anterior row sutures; (C) lateral sutures leading to edge of posterior portion of anastomosis.

Figure 81.12 : Completed Kasai procedure: (A) Roux-en-Y end-to-side portoenterostomy anastomosis; (B) stomach and pylorus; (C) transverse colon with retrocolic Roux limb behind it.

Outcomes

Given that biliary atresia was previously a uniformly fatal disease, the Kasai operation has dramatically improved the survival rates of infants with this disease. However, the results are far from perfect. In general, by Japanese and Western reports, 60–80% of patients will achieve initial improvement in jaundice and early success with portoenterostomy, defined by some as normalisation of serum bilirubin to less than 2 mg% at 3 months following the procedure. Of these, one-half will have permanent relief and the other one-half will have progressive liver failure. The patients who never achieved initial improvement go on to experience progressive liver failure. The best five-year survival (up to 60%) has been reported by the Japanese Biliary Atresia Registry. Rates in most African reports are significantly lower, likely due to the patient's advanced age at the time of diagnosis and morbidities of advanced disease.[6,10]

Prognostic Factors

The most important long-term prognostic factors in predicting outcome after the Kasai procedure appear to be age at presentation, achievement of primary biliary drainage, experience of the centre, and occurrence of cholangitis.

Age at the time of the Kasai procedure is perhaps the most important prognostic indicator. Most reports state that the best outcomes, in both initial success and long-term survival, occur in patients younger than age 60–70 days at the time of operation (68% 10-year survival). The Japanese Biliary Atresia Registry extends such rates to patients up to 90 days of age, with a precipitous drop in success after that point.[3,11,20] Most series report performance of a Kasai procedure in infants after 70 days of age as a risk factor for failure; however, some do not agree with this.[21–23]

Although it is a very important postoperative prognostic factor, primary biliary drainage is not a guarantee of the long-term success of a portoenterostomy. Rather, it is a *requirement* for success in that nearly all patients who do not achieve primary biliary drainage will never do so. Surgical treatment via a second (redo) Kasai procedure for patients who did not achieve primary drainage as well as for those who had initial drainage but subsequent cessation, was met with initial interest secondary to case reports and small series of successes. Recent reports, however, suggest that redo portoenterostomy is not a useful strategy in the vast majority of patients.[24]

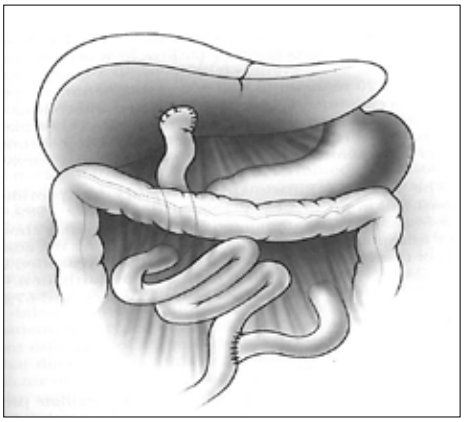
Role of Liver Transplantation

Liver transplantation, where available, is principally useful in cases of inadequate biliary drainage after portoenterostomy and in cases of progressive hepatic dysfunction or refractory portal hypertension despite initial portoenterostomy. Liver transplantation centres in North America, Europe, and Japan now report up to 85% 5-year survival after transplant for biliary atresia.[25] Given these excellent results, some in the West have advocated for primary liver transplantation in the neonate with BA. However, the long-term success of portoenterostomy in a significant percentage of patients, combined with the risks of long-term immunosupression beginning early in life (infectious and malignant), the expense of liver transplant, the shortage of donors, and the need for intense follow-up, make this a less attractive initial option in these patients. These issues are magnified in the African setting. In those regions where transplantation is an option, it should be noted that performance of an initial Kasai procedure has not been shown to affect the success of subsequent transplantation.[26]

Conclusion

The Kasai procedure remains the mainstay of initial treatment for biliary atresia throughout the world. In addition, it may offer the only hope for children with BA in most of Africa, where liver transplantation may not be available.

Despite the fact that many African reports state a poor outcome for even those BA patients who undergo portoenterostomy, likely due to the extremely late presentation of patients for the procedure, this should not discourage the use of the Kasai procedure in general.[6,10] It is therefore important for the surgeon to advocate to the community for early referral of patients with neonatal jaundice who may have biliary atresia. Time is of the essence, so accepting centres should have expertise in the rapid work-up of such patients. Subsequent thorough preoperative resuscitation and preparation and meticulous technique in the performance of Kasai portoenterostomy are crucial to the successful outcome of the patient who presents with potentially salvageable liver disease.



Source: Altman RP, Buchmiller TL. The jaundiced infant—biliary atresia. In: Grosfeld JL, O'Neill JA, Coran AG, Fonkalsrud EW (eds.). Pediatric Surgery, 6th ed., Vol. 2. 2006, Pp 1609.

Figure 81.13 : Illustration of completed Kasai procedure.

Key Summary Points

1. Although not as common as infectious aetiologies of neonatal cholestasis, biliary atresia is seen regularly in centres throughout Africa.
2. Persistent jaundice after 3 weeks of age is pathologic until proven otherwise.
3. The differential diagnoses for cholestasis in the newborn are large and include genetic/metabolic disorders, infectious agents, hepatocellular dysfunction, neonatal hepatitis, and extrahepatic obstructive disorders.
4. Early referral to a centre that can rule out extrahepatic biliary obstruction is vital to a successful outcome if biliary atresia is found.
5. A combination of ultrasound, nuclear imaging (if available), and operative cholangiogram with or without biopsy will definitively determine whether a patient has biliary atresia.
6. Kasai portoenterostomy is the cornerstone of therapy for biliary atresia.
7. Modifications of the Roux-en-Y portoenterostomy, such as externalisation of the biliary conduit and intussusception valves within the Roux limb, offer no benefit.
8. Cholangitis, the most frequent postoperative complication, is usually responsive to broad-spectrum antibiotics.
9. Advanced age at the time of the Kasai procedure is one of the most important indicators of poor outcome.

References

1. Karrer FM, Lilly JR. Corticosteroid therapy in biliary atresia. J Pediatr Surg 1977; 90:736.
2. Adelman S. Prognosis of uncorrected biliary atresia: an update. J Pediatr Surg 1978; 13:389–392.
3. Nio M, Ohio R, Miyano T, et al. Five- and 10-year survival rates after surgery for biliary atresia: a report from the Japanese Biliary Atresia Registry. J Pediatr Surg 2003; 38:997.
4. Obanafunwa JO, Elesha SO. Childhood liver diseases in Jos, Nigeria: a retrospective histopathological study. East Afr Med J 1991; 68:702–706.
5. Motala C, Ireland JD, Hill ID, Bowie MD. Cholestatic disorders of infancy—aetiology and outcome. J Trop Pediatr 1990; 36:218–222.
6. Mabogunje OA. Biliary atresia in Zaria, Nigeria: a review. Ann Trop Paediatr 1987; 7:200–204.
7. Muthuphei MN. Childhood liver diseases in Ga-Rankuwa Hospital, South Africa. East Afr Med J 2000; 77:508–509.
8. Akang EE, Osinusi KO, Pindiga HU, Okpala JU, Aghadiuno PU. Congenital malformations: a review of 672 autopsies in Ibadan, Nigeria. Pediatr Pathol 1993; 13:659–670.
9. O'Neill JA, Jr., Grosfeld JL, Fonkalsrud EW, Coran AG, Caldamone AA, eds. Principles of Pediatric Surgery. 2nd ed. Mosby, 2004.
10. Mshelbwala PM, Sabiu L, Lukong CS, Ameh EA. Management of biliary atresia in Nigeria: the ongoing challenge. Ann Trop Paediatr 2007; 27:69–73.
11. Altman RP, Lilly JF, Greenfeld J, et al. A multivariable risk factor analysis of the portoenterostomy (Kasai) procedure for biliary atresia: twenty-five years of experience from two centers. Ann Surg 1997; 226:348; discussion 353.
12. Zerbini MC, Gallucci SD, Maezono R, et al. Liver biopsy in neonatal cholestasis: a review on statistical grounds. Mod Pathol 1997; 10:793–799.
13. Sera Y, Ikeda S, Akagi M. Ultrasonographic studies for the diagnosis of infantile cholestatic disease. In: Ohi R, ed. Biliary Atresia. Professional Postgraduate Services, 1987.
14. Kasai M, Suzuki H, Ohashi E, et al. Technique and results of operative management of biliary atresia. World J Surg 1978; 2:571–580.
15. Dillon PW, Owings E, Cilley R, et al. Immunosuppression as adjuvant therapy for biliary atresia. J Pediatr Surg 2001; 361:80.
16. Meyers RL, Book LS, O'Gorman MA, et al. High-dose steroids, ursodeoxycholic acid, and chronic intravenous antibiotics improve bile flow after Kasai procedure in infants with biliary atresia. J Pediatr Surg 2003; 38:406.
17. Muraji T, Tusagawa C, Nishijima E, et al. Surgical management for intractable cholangitis in biliary atresia. J Pediatr Surg 1997; 32:1103; discussion 1106.
18. Bu LN, Chen HL, Chang CJ, et al. Prophylactic oral antibiotics in prevention of recurrent cholangitis after the Kasai portoenterostomy. J Pediatr Surg 2003; 38:509.

19. Lilly JR, Weintraub WH, Altman RP. Spontaneous perforation of the extrahepatic bile ducts and bile peritonitis in infancy. Surgery 1974; 75:664.
20. Grosfeld JL, Fitzgerald JF, Predaina R, et al. The efficacy of hepatoportoenterostomy in biliary atresia. Surgery 1989; 106:692; discussion 700.
21. Davenport M, Kerkar N, Mieli-Vergani G, et al. Biliary atresia: the King's College Hospital experience (1974–1995). J Pediatr Surg 1997; 32:479.
22. Maksoud JG, Fauza DO, Silva MM, et al. Management of biliary atresia in the liver transplantation era: A 15-year, single-center experience. J Pediatr Surg 1998; 33:115.
23. van Heurn LW, Saing H, Tam PK: Portoenterostomy for biliary atresia: Long-term survival and prognosis after esophogeal variceal bleeding. J Pediatr Surg 2004; 39:6.
24. Hasegawa T, Kimura T, Sasaki T, et al. Indication for redo hepatic portoenterostomy for insufficient bile drainage in biliary atresia: Re-evaluation in the era of liver transplantation. Pediatr Surg Int 2003; 19:256.
25. McDiarmid SV. Current status of liver transplantation in children. Pediatr Clin North Am 2003; 50:1335.
26. Vacanti JP, Shamberger RC, Eraklis A, Lillehei CW. The therapy of biliary atresia combining the Kasai portoenterostomy with liver transplantation: a single center experience. J Pediatr Surg 1990; 25:149.