# Biliary Atresia

# 26

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Biliary atresia is a cholangiodestructive disease affecting all parts of biliary tract. A number of variants can be identified among which the most important are those with other anomalies (e.g., BASM), those with cystic change and those related to CMV infection.

It is invariably fatal if untreated.

## 26.1 Epidemiology

- 1 in 10,000—Japan and China
  - ↓ Incidence of BASM and associated cardiac anomalies
- 1 in 16–20,000—the UK, USA, and Europe
- F > M (slight in isolated BA, marked in BASM and Cystic BA).

## 26.2 Embryology

A primordial bile duct emerges from the distal end of the foregut at about 20 days post-fertilization. This becomes engulfed by the developing liver anlage, which at this stage contains few hepatoblasts and is mainly mesenchyme and a source of blood cell precursors. The critical point of extrahepatic bile duct development is coincident (5–6 weeks gestation) with the determination of abdominal situs, spleen

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formation, final development of portal vein, and vena cava (hence the association with BASM).

Biliary epithelial cells differentiate from their hepatoblast precursors to form intrahepatic biliary ductules from about the seventh week of gestation and undergo a process of selection/deletion with progressively fewer but larger remaining ducts. Bile is secreted into the GI tract by 12 weeks emphasizing biliary continuity.

As the etiology of BA is largely unknown, hypotheses abound. Two of them have a degree of evidential support.

- *Developmental—primary failure to form lumen or sufficiently functional intrahepatic bile ducts.*
  - BASM (polysplenia or asplenia; situs inversus; absence of IVC; pre-duodenal portal vein; cardiac anomalies) ~2% will have gene mutations (*PKD1L1*).
  - Cystic BA— > 60% detected on antenatal US scan.
  - Genetic predisposition—so-called susceptibility genes (e.g., *ADD3*) in Han Chinese.
- *Perinatal acquired*—i.e., a normally developed bile duct is damaged later, with secondary loss of luminal continuity. May be due to:
  - Viral insult—e.g., CMV (N.B. IgM+ BA can be seen in ~10% in the UK). Rotavirus-induced experimental BA in mice.
  - Immunological overreaction.

## 26.3 Classification

Figure 26.1 illustrates classification with the most proximal level of obstruction determining the type. About 5% will have cystic change within otherwise obliterated biliary tract **(cystic biliary atresia).** These need to be differentiated from a *choledochal malformation*, which even if obstructed should connect to a smooth, progressively distended intrahepatic duct system.

## 26.4 Clinical Features

Conjugated jaundice, pale stools, dark urine, and failure to thrive. Nil specific.

Look for situs, polysplenia, and cardiac anomalies of BASM.

### 26.4.1 Investigations

See Chap. 24 for details.

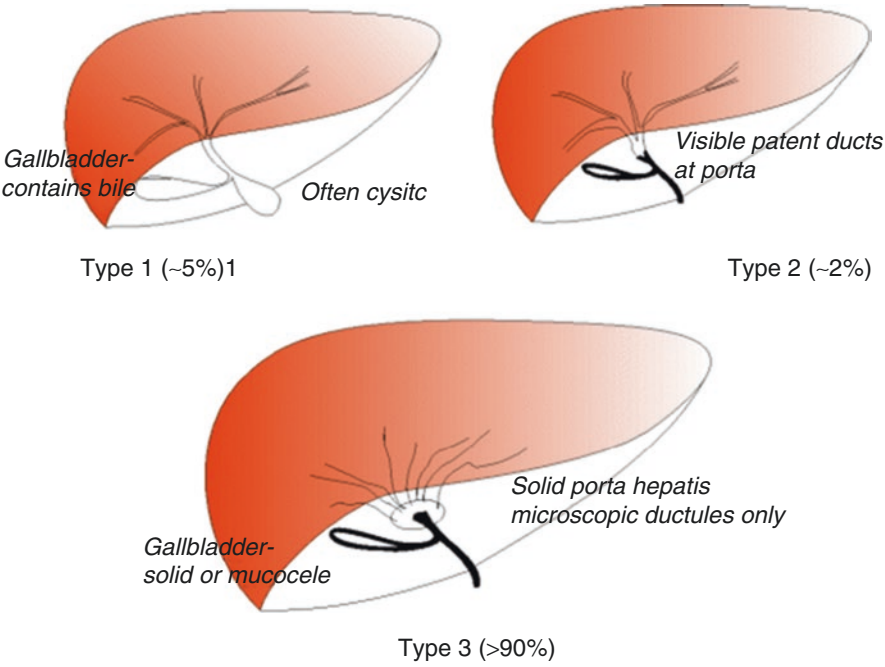


**Fig. 26.1** Classification of biliary atresia

## 26.5 Surgery

- Correct coagulopathy (vitamin K).
- ~5–10 have associated cardiac anomalies that may need correction *before* KPE.

In most cases, an attempt to preserve native liver using portoenterostomy is a better strategy than primary liver transplantation. However, latter should be considered in "old" infants (>100 days), especially those with obvious cirrhosis (ascites, portal hypertension).

### 26.5.1 Kasai[1] Portoenterostomy (Fig. 26.2)

RUQ—muscle-cutting—extended across midline.

1. Confirm diagnosis ± cholangiogram.
2. Porta hepatis dissection—facilitated by extra-abdominal delivery of liver.

[1] Morio Kasai (1922–2008) Japanese pediatric surgeon who reported initial technique in 1959.

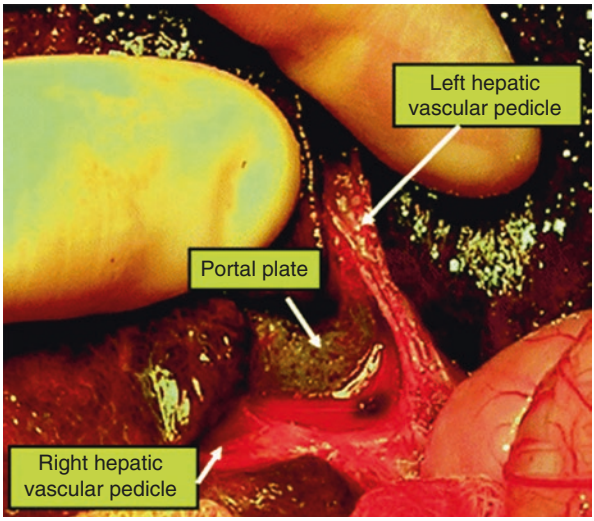


**Fig. 26.2** Kasai portoenterostomy—close up of porta hepatic. Transected bile ductules are visible in this case (unusual)

3. Excision of all extrahepatic remnants to the level of liver capsule, facilitated by retraction of portal vein confluence. Clearance proceeds from bifurcation of right vascular pedicle to insertion of umbilical vein on left portal vein.
4. Roux loop[2] (~40 cm) reconstruction and portoenterostomy (6/0 PDS).

**Adjuvant Therapy**

1. High dose steroids (e.g., prednisolone 4–5 mg/kg/day—tapered for 5 weeks).
2. Ursodeoxycholic acid (5 mg/kg tds).
3. Prophylactic antibiotics (e.g., Septrin™, trimethoprim, cefalexin).

## 26.6 Complications

- Cholangitis (40%).
  - Gram −ve organisms (usually).
  - Treated with IV antibiotics (Tazocin and Gentamicin).
- Portal hypertension.
  - Splenomegaly.
  - Esophageal, gastric, and anorectal varices.
- Hepatopulmonary syndrome.
  - Hypoxia, cyanosis, dyspnea, and clubbing due to the development of pulmonary arteriovenous shunts.
  - ↑ Incidence with BASM, diagnosed with saturation monitoring.
  - Reversed after liver transplantation.

[2] Cesar Roux (1857–1934) Swiss surgeon. Designed "Roux loop" for drainage of the stomach when obstructed but has been used for many indications since.

- Inguinal Hernia.
  - ↑ ascites and intrabdominal pressure.
- Malignancy
  - HCC, hepatoblastoma, and cholangiocarcinoma (rare ~2% native liver survivors).

## 26.7 Outcome

Success post-KPE can be gauged by:

1. Proportion to clear jaundice (ideally <20 μmol/L).
   (a) 50–60% is achievable.
2. Proportion to survive with own liver.
   (a) 50% after 5 years is achievable.

Prognosis post-KPE can be affected by:

1. Age at surgery—there are no "cut-off" values however.
2. Experience of surgeon/center.
3. BASM—prognosis reduced by presence of cardiac anomalies.

## Further Reading

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