BILIARY ATRESIA

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OBJECTIVES

- Recognize the pathogenesis and likely clinical presentation
- Understand the different laboratory and diagnostic imaging modalities used to evaluate for biliary atresia
- Discuss the management and postoperative considerations

PATHOGENESIS

- Cause unknown but some etiologies have been hypothesized
  - Viral/Toxic: studies have failed to identify associations to viral infections such as CMV, rotavirus, reovirus or specific toxins
  - Genetic
    - Not a causative factor in a majority of cases but may play a role in subgroup with biliary atresia splenic malformations
    - Some suspect prenatal genetic factors may influence development of biliary tree
  - Immunologic
    - Some studies implicate immune dysregulation as a contributing factor (primary or secondary to infectious/genetic trigger)

CLINICAL FEATURES

- Jaundice
- Acholic stools
  - Study from Japan showed how stool color cards completed by parents had sensitivity of 76.5 percent and specificity of 99.9 percent to identify biliary atresia
- Dark urine
- Firm, enlarged liver and splenomegaly

DIAGNOSTIC TESTING

- Lab work

Finding	Implications
Initial tests for all infants	
▪ Comprehensive metabolic panel	
• Total and conjugated bilirubin	To evaluate for conjugated hyperbilirubinemia (cholestasis) versus unconjugated hyperbilirubinemia.
• ALT and AST	To assess for hepatocyte injury.
• Alkaline phosphatase and GGTP	To assess for biliary injury. Furthermore, several genetic/metabolic disorders can be divided into high and low GGTP categories*.
• Total protein and albumin	To assess hepatocyte function. Low albumin suggests poor nutrition, renal losses, or poor hepatic synthetic function.
• Electrolytes, bicarbonate, glucose	To assess for metabolic disease. Abnormalities in these results are often seen in infants with metabolic disease.
▪ CBC with differential	To assess for infection and/or splenic sequestration. Elevated WBC is suggestive of infection. Low WBC and platelet count could indicate portal hypertension (with splenic sequestration).
▪ PT/INR and PTT	To assess hepatocyte function and/or vitamin K deficiency. Abnormal results indicate impaired liver synthetic function and/or vitamin K deficiency.

Source: Update, Biliary atresia, retrieved Apr 2020

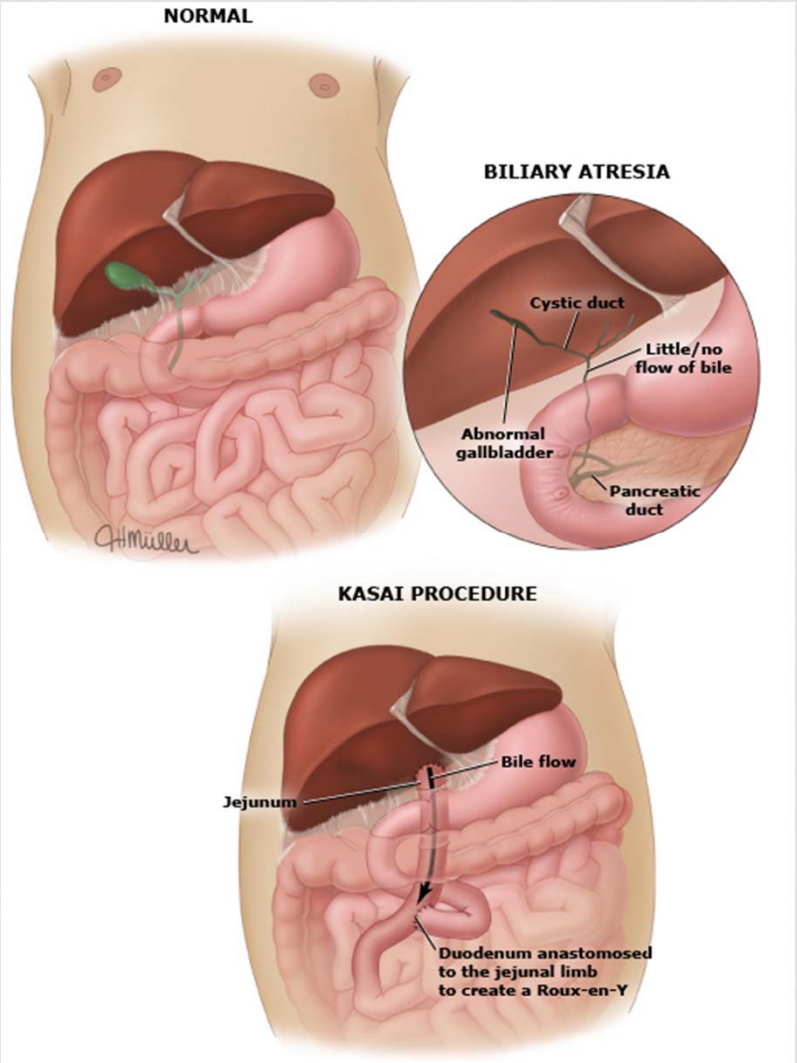
- Lab findings: elevated direct bilirubin, aminotransferases, and gamma-glutamyl transpeptidase
  - ELEVATED DIRECT BILIRUBIN IS NEVER PHYSIOLOGIC

DIAGNOSTIC IMAGING

- Abdominal ultrasound
  - To exclude other anatomic causes of cholestasis (ie, choledochal cyst)
  - In BA, gallbladder either absent or irregular in shape
    - "triangular cord" sign: a triangular echogenic density seen just above the porta hepatis
    - Absence of the common bile duct
    - Enlarged hepatic hilar lymph node
- Hepatobiliary scintigraphy
- Liver biopsy
- Intraoperative cholangiogram – gold standard

MANAGEMENT

- Kasai procedure – hepatoportoenterostomy
- Restores bile flow from the liver to the proximal small bowel by creating a Roux-en-Y loop of bowel with anastomoses to the hilum of the liver



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POSTOPERATIVE MANAGEMENT

- Choleretics: ursodeoxycholic acid (ursodiol) once taking food by mouth
- Nutrition
  - Caloric intake must be increased; malnutrition is a risk
  - Deficiency in fat soluble vitamins
- Infection
  - Cholangitis is a frequent complication
    - Recurrent infection may lead to scarring of the liver and liver transplantation
  - Evidence for prophylaxis with either trimethoprim-sulfamethoxazole or neomycin
  - Interestingly, one study showed that probiotics may be as effective as neomycin

REFERENCES

- Takamizawa S, Zaima A, Muraji T, Kanegawa K, Akasaka Y, Satoh S, Nishijima E. Can biliary atresia be diagnosed by ultrasonography alone? J Pediatr Surg. 2007;42(12):2093.
- Erlichman J, Loomes KM. Biliary Atresia. In: UpToDate, Rand, EB(Ed), UpToDate, Waltham, MA, 2020.
- Zallen GS, Bliss DW, Curran TJ, Harrison MW, Silen ML. Biliary Atresia. *Pediatr Rev*. 2006;27(7):243 LP - 248. doi:10.1542/pir.27-7-243.
- Bu LN, Chen HL, Chang CJ, Ni YH, Hsu HY, Lai HS, Hsu WM, Chang MH. Prophylactic oral antibiotics in prevention of recurrent cholangitis after the Kasai portoenterostomy. J Pediatr Surg. 2003;38(4):590.
- Lien TH, Bu LN, Wu JF, Chen HL, Chen AC, Lai MW, Shih HH, Lee IH, Hsu HY, Ni YH, Chang MH. Use of Lactobacillus casei rhamnosus to Prevent Cholangitis in Biliary Atresia After Kasai Operation. J Pediatr Gastroenterol Nutr. 2015 May;60(5):654-8.