Biliary Atresia

Biliary Atresia is a condition in which the bile ducts outside and inside the liver are scarred (atretic) and blocked. It presents as conjugated (direct) hyperbilirubinemia within the first 4 weeks of life.

Conjugated hyperbilirubinemia is always pathogenic and defined as a conjugated bilirubin concentration greater than 2 mg/dL or more than 20% of total bilirubin. Incidence is 1 in 2,500 live births. Early identification (<2 months of life) of biliary atresia is important, allows for timely intervention and leads to improved outcomes.

The initial step in the evaluation of jaundice should focus on distinguishing between unconjugated and conjugated hyperbilirubinemia. CHLA recommends obtaining fractionated results of total hyperbilirubinemia by at least 4 weeks of age.

Work up	
Initial evaluation	- History including questions regarding prenatal care, maternal infections, recent infections and family history of liver diseases. - Physical exam with special attention to growth, organomegaly, heart murmur, and stool color. - Labs: complete blood count, reticulocyte count, liver testing (AST, ALT, total bilirubin, conjugated bilirubin and GGT) and coagulation profile (PT/INR)
Follow up evaluation	- Repeat labs in 5-7 days and continue close follow up if total/conjugated bilirubin are rising - Imaging: complete abdominal ultrasound with Doppler (If possible, otherwise will be done by GI team) **When To Refer to Hepatology/Gastroenterology** - Persistently elevated or rising conjugated bilirubin - Persistently elevated or rising AST/ALT

URGENT…if patient is < 2 months of age with conjugated hyperbilirubinemia and pale stools.
Call (323) 660-2450 and ask to speak to the Hepatologist on call

Referral Checklist

1) Clinic Notes: Initial and most recent clinic notes relevant to referring diagnosis including growth chart
2) Lab Work
3) Imaging Tests

Reviewed 7/14/2025