CONDITION

Biliary atresia

Learn how biliary atresia affects an infant's bile ducts, including early signs, diagnostic tests, the Kasai procedure, liver transplant, and follow-up care.

WHAT IT IS
Serious infant liver and bile duct condition

BODY SYSTEM
Digestive system

MAIN SYMPTOMS
Persistent jaundice, pale stools, dark urine, poor growth

WHAT CAUSES IT
Unknown, possibly immune and developmental

HOW IT IS DIAGNOSED
Cholangiogram, often with liver biopsy

MAIN TREATMENT
Surgery, sometimes liver transplant

CAN IT BE CURED?
No, surgery does not cure it

IS IT AN EMERGENCY?
Sometimes, vomiting blood or severe bleeding

LONG-TERM OUTLOOK
Progressive liver damage may eventually require transplant

Overview

Biliary atresia is a serious liver and bile duct condition that begins in infancy. The bile ducts outside the liver become inflamed, damaged, or blocked. In some babies, ducts within the liver are also affected.

Bile is a fluid made by the liver. It helps the body digest fats and remove bilirubin, a yellow substance produced when old red blood cells break down. Normally, bile travels through small ducts from the liver to the intestine. In biliary atresia, bile cannot drain properly. It builds up in the liver and can cause scarring, called fibrosis. Advanced scarring is known as cirrhosis.

Most affected babies appear healthy at birth. The first sign is often jaundice, which means that the skin or whites of the eyes look yellow. Jaundice is common during a newborn's first days, but jaundice caused by biliary atresia does not improve as expected. Pale stools and dark urine

are especially important clues.

Biliary atresia requires prompt assessment by a pediatric liver specialist. Surgery can improve bile drainage, particularly when performed early, but it does not cure the underlying disease. Some children eventually need a liver transplant even after successful initial surgery.

Symptoms

Symptoms usually develop during the first weeks of life. They can be difficult to distinguish from more common causes of newborn jaundice.

Possible signs include:

Normal infant stool is usually yellow, green, or brown. A stool color card or photograph may help a clinician assess a concerning color, but it does not replace an examination.

Pale or white stools, dark urine, or jaundice that persists beyond the early newborn period needs prompt medical assessment. These findings do not prove that a baby has biliary atresia, but testing should not be delayed.

Seek urgent medical care if a child with known or suspected liver disease develops fever, increasing jaundice, a newly swollen abdomen, repeated vomiting, poor feeding, or unusual sleepiness. These can be signs of infection or worsening liver function.

Call emergency services for difficulty breathing, a seizure, vomiting blood, black or bloody stool, severe bleeding, or a child who is difficult to wake.

- Yellow skin or eyes that persist or become more noticeable
- Stools that are pale, gray, white, or clay-colored
- Urine that is unusually dark and stains the diaper
- An enlarged or firm liver
- A swollen abdomen
- Slow weight gain or poor growth
- Irritability or reduced interest in feeding

Causes and risk factors

The exact cause of biliary atresia is not fully understood. Researchers believe that bile duct development, immune activity, inflammation, and possible environmental triggers may contribute. No single infection, medicine, or exposure has been shown to cause most cases.

Biliary atresia is not contagious. It is not caused by something a parent did during pregnancy or after birth. In most families, there is no history of the condition, and it is not considered a straightforward inherited disorder.

Most babies have biliary atresia without other major birth differences. A smaller group has additional conditions involving the spleen, heart, intestines, or the usual placement of organs in the chest and abdomen. This pattern may begin earlier during fetal development.

Clearly established, preventable risk factors are not known. There is also no standard prenatal test that can reliably rule biliary atresia in or out. The condition is generally identified after birth based on symptoms and medical testing.

Diagnosis

Evaluation usually begins with a physical examination and blood tests. The clinician will ask about the timing of jaundice, urine color, stool color, feeding, growth, and birth history.

A bilirubin blood test is especially important. Bilirubin is measured as total bilirubin and direct, or conjugated, bilirubin. A raised direct bilirubin level suggests cholestasis, meaning that bile is not flowing normally. Cholestasis has several possible causes and requires timely evaluation.

Other blood tests may measure liver enzymes, proteins made by the liver, blood sugar, and the blood's ability to clot. Tests may also look for infections, genetic conditions, or metabolic disorders that can resemble biliary atresia.

An abdominal ultrasound can examine the liver, gallbladder, bile ducts, spleen, and blood vessels. Ultrasound findings may support the diagnosis, but a normal or unclear scan does not always exclude it.

Depending on the situation, additional testing may include:

A cholangiogram directly shows the bile ducts after contrast material is placed into the biliary system. In infants with suspected biliary atresia, it is often performed during surgery. If the ducts are blocked or absent, the surgeon may proceed directly to treatment.

Because no single blood test or scan confirms every case, diagnosis often depends on the combined results of several tests. Early referral to a center experienced in pediatric liver disease and surgery is important.

- A nuclear medicine scan to assess whether bile reaches the intestine
- A liver biopsy, in which a small tissue sample is examined under a microscope
- Genetic or metabolic testing for other causes of infant cholestasis

- Heart imaging or other studies when associated birth differences are suspected

Treatment and management

The main initial treatment is the Kasai portoenterostomy, often called the Kasai procedure. During this operation, the surgeon removes the damaged bile ducts outside the liver. A section of small intestine is then connected to the liver's surface so bile can drain through any remaining microscopic channels.

The Kasai procedure does not create normal bile ducts or remove all liver injury. Its goal is to restore enough bile flow to reduce jaundice, support growth, and delay further liver damage. In general, outcomes are better when diagnosis and surgery occur early in infancy.

After surgery, the care team monitors stool color, bilirubin levels, liver tests, growth, and signs of infection. Some centers use medicines intended to support bile flow or reduce the risk of infection. Practices vary, and these treatments must be selected and monitored by the child's specialists.

Nutrition is a central part of care. Poor bile flow makes it harder to absorb fat and the fat-soluble vitamins A, D, E, and K. Some infants need higher-calorie feeding, specialized formulas containing easier-to-absorb fats, vitamin supplements, or feeding through a tube. A pediatric liver dietitian can adjust the plan based on growth and blood tests.

Cholangitis is an infection and inflammation of the bile ducts. It can occur after the Kasai procedure and may cause fever, worsening jaundice, pale stools, irritability, or poor feeding. It needs urgent medical evaluation and is usually treated with antibiotics like amoxicillin or doxycycline .

A liver transplant may be recommended if the Kasai procedure does not establish useful bile flow or if progressive liver disease causes serious complications. Reasons can include poor growth despite nutrition support, repeated cholangitis, severe portal hypertension, fluid buildup, impaired liver function, or reduced quality of life.

Transplant evaluation is a detailed process rather than an emergency operation in most cases. The team assesses the child's liver disease, nutrition, heart and lung health, infections, and family support needs. A liver may come from a deceased donor or, in some situations, a healthy living donor who gives part of the liver.

Children who receive a transplant need lifelong specialist care and medicines that reduce immune rejection. These medicines also increase susceptibility to some infections, so monitoring and vaccination planning are important.

Complications

Biliary atresia can continue to affect the liver even when bile drainage improves after surgery. The pattern and pace vary from child to child.

Possible complications include:

Vitamin K deficiency or reduced liver function can interfere with blood clotting. Families should report unexplained bruising, prolonged bleeding, blood in stool, or vomiting blood promptly.

Portal hypertension can develop even when a child otherwise seems well. This is one reason that regular specialist visits and planned testing remain important.

- Cholangitis: Infection involving the bile drainage system
- Cirrhosis: Extensive scarring that changes the liver's structure and function
- Portal hypertension: High pressure in the vein carrying blood into the liver
- Enlarged spleen: Often related to portal hypertension and sometimes associated with low blood cell counts
- Varices: Enlarged veins, usually in the esophagus or stomach, that can bleed
- Ascites: Fluid buildup in the abdomen
- Poor growth: Caused by increased nutritional needs and difficulty absorbing nutrients
- Vitamin deficiencies: Especially deficiencies of vitamins A, D, E, and K
- Itching: A possible effect of substances retained when bile flow is reduced
- Progressive liver failure: Reduced ability of the liver to perform essential tasks

Follow up

Follow-up is usually coordinated by a pediatric hepatologist, a doctor who specializes in children's liver conditions. Surgeons, transplant specialists, dietitians, pharmacists, nurses, and primary care clinicians may also be involved.

Routine follow-up may include:

Families should keep all scheduled follow-up visits, even if jaundice has improved. Changes in blood tests, growth, or portal pressure may appear before obvious symptoms.

The care team should provide a clear plan for symptoms that need urgent attention. Fever, worsening jaundice, pale stools, dark urine, a rapidly enlarging abdomen, repeated vomiting, or a sudden change in feeding or alertness should be reported promptly. Any fever in a young

infant needs timely medical guidance.

Emergency care is needed for vomiting blood, black or bloody stool, major bleeding, breathing difficulty, seizure, or severe difficulty waking the child.

Routine childhood care also remains important. Vaccines, dental care, developmental screening, and age-appropriate activity should be discussed with the child's clinicians. Before giving prescription medicines, nonprescription products, vitamins, or herbal supplements, caregivers should check with the liver team because some products can affect the liver or interact with treatment.

- Measurement of weight, length or height, and head growth
- Blood tests for bilirubin, liver function, nutrition, and clotting
- Monitoring for vitamin deficiencies
- Ultrasound or other imaging of the liver, spleen, and blood vessels
- Assessment for portal hypertension and enlarged veins
- Review of medicines, supplements, feeding, and development
- Discussion of transplant evaluation when appropriate

Sources

- Fawaz R, Baumann U, Ekong U, et al. "Guideline for the Evaluation of Cholestatic Jaundice in Infants: Joint Recommendations of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition and the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition." Journal of Pediatric Gastroenterology and Nutrition . 2017;64(1):154-168.
- Bezerra JA, Wells RG, Mack CL, et al. "Biliary Atresia: Clinical and Research Challenges for the Twenty-First Century." Hepatology . 2018;68(3):1163-1173.
- Hartley JL, Davenport M, Kelly DA. "Biliary Atresia." The Lancet . 2009;374(9702):1704-1713.
- Sundaram SS, Mack CL, Feldman AG, Sokol RJ. "Biliary Atresia: Indications and Timing of Liver Transplantation and Optimization of Pretransplant Care." Liver Transplantation . 2017;23(1):96-109.
- National Institute of Diabetes and Digestive and Kidney Diseases. "Biliary Atresia." National Institutes of Health, Health Information. Accessed March 8, 2025.