CONDITION

EHBA

EHBA is a condition in babies where damaged bile ducts outside the liver cannot drain bile normally.

WHAT IT IS
Blocked or absent bile ducts outside the liver

BODY SYSTEM
Liver and digestive system

MAIN SYMPTOMS
Yellow skin and eyes, pale stools, dark urine, poor growth

WHAT CAUSES IT
Unknown; not usually inherited

HOW IT IS DIAGNOSED
Blood tests, imaging, liver biopsy, operative cholangiography

MAIN TREATMENT
Early bile-drainage surgery, supportive care, sometimes liver transplantation

CAN IT BE CURED?
Sometimes - liver transplant replaces the affected liver

IS IT AN EMERGENCY?
Sometimes - fever after surgery may mean cholangitis

LONG-TERM OUTLOOK
Early surgery helps, but many eventually need liver transplantation

Overview

EHBA stands for extrahepatic biliary atresia. "Extrahepatic" means outside the liver, "biliary" refers to the bile ducts, and "atresia" means a normal passage is missing or blocked.

Bile ducts carry bile from the liver to the small intestine. Bile helps digest fats and removes waste products from the liver. In EHBA, the ducts outside the liver become damaged and scarred around the time of birth. Bile cannot drain normally and builds up in the liver.

This causes jaundice, a yellow colour in the skin and whites of the eyes. It can also cause liver scarring, or fibrosis, which may progress to cirrhosis and liver failure without effective treatment.

EHBA only occurs in babies and needs prompt assessment because surgery generally works

better when performed early. Treatment usually begins with an operation to create a new route for bile drainage. Some children later need a liver transplant.

Symptoms

Newborn jaundice is common and usually settles within the first two weeks. With EHBA, jaundice continues or becomes more noticeable. It may be harder to see on brown or black skin, so check the whites of the eyes and gums in good natural light.

Important symptoms include:

Pale stools are an especially important sign. Normal baby stools may be yellow, green, orange, or brown. White, chalky, or putty-coloured stools suggest that little or no bile is reaching the intestine. A photograph can help a healthcare professional assess the colour.

Babies with EHBA may initially feed well and appear otherwise healthy. Over time, poor bile flow can reduce the absorption of fats and vitamins, contributing to poor growth.

- Yellow skin or eyes after two weeks
- Very pale, cream, grey, or white stools
- Dark yellow or brown urine
- A swollen or firm abdomen
- Slow weight gain or poor growth
- Irritability or unusual sleepiness

Causes and risk factors

The exact cause of EHBA is unknown. Abnormal bile duct development and inflammation before or shortly after birth may both contribute. No single infection or immune trigger has been confirmed.

EHBA is not caused by anything a parent did during pregnancy. It is not contagious and is not usually inherited.

Most affected babies have no other major condition. A smaller group has developmental differences involving organs such as the spleen, heart, intestines, or major blood vessels.

There is no reliable pregnancy test that can rule EHBA in or out. It is occasionally suspected on prenatal ultrasound but is usually recognised after birth.

Diagnosis

A baby with persistent jaundice or pale stools needs prompt testing. Blood tests measure bilirubin, a yellow waste product processed by the liver. EHBA increases direct, or conjugated, bilirubin because bile cannot drain properly.

Other blood tests assess liver inflammation, clotting, nutrition, and alternative causes of jaundice. An abdominal ultrasound may examine the liver, gallbladder, bile ducts, spleen, and nearby blood vessels. A normal-looking ultrasound does not always exclude EHBA.

Further tests may include:

Diagnosis is usually confirmed by cholangiography. Dye is placed into the bile system so the ducts can be seen on an X-ray, often during an operation. If the ducts are blocked or absent, the surgeon may proceed directly to treatment.

No single test should be interpreted alone. A specialist children's liver and surgical team assesses the symptoms and combined results.

- A liver biopsy, in which a small tissue sample is examined under a microscope
- A nuclear medicine scan that tracks a tracer through the liver and bile system
- Tests for infections, genetic conditions, or other causes of poor bile flow

Treatment and management

The main initial treatment is a Kasai portoenterostomy. The surgeon removes the damaged ducts outside the liver and connects a loop of small intestine to the area where tiny bile channels leave the liver, creating a new drainage pathway.

The operation does not rebuild normal ducts or remove existing liver damage. It aims to improve bile flow, reduce jaundice, support growth, and delay or avoid transplantation. Outcomes are generally better when surgery happens early, before extensive scarring develops.

After surgery, treatment may include medicines to support bile flow and antibiotics like amoxicillin or doxycycline to reduce the risk of bile channel infection. Babies may need high-energy feeds, special dietary fats, or supplements containing vitamins A, D, E, and K, which are harder to absorb when bile flow is poor.

Blood tests, growth, stool colour, and signs of liver scarring are monitored regularly. Families should follow the specialist plan and not start vitamins, herbal products, or over-the-counter medicines independently. Some can harm the liver or interact with prescribed treatment.

A liver transplant may be needed if the Kasai operation does not establish enough bile flow or complications develop. Reasons include ongoing jaundice, repeated infections, poor growth, severe portal hypertension, or liver failure.

Transplantation replaces the affected liver but involves major surgery and lifelong follow-up. Anti-rejection medicines are required afterward.

Complications

Poor bile drainage can cause progressive liver scarring. Cirrhosis means extensive scar tissue has changed the liver's structure and blood flow.

Possible complications include:

Complications can occur even if jaundice improves after surgery, so regular long-term monitoring remains important.

- Cholangitis: infection in the bile channels, often causing fever, irritability, worsening jaundice, or pale stools
- Portal hypertension: increased pressure in the vein carrying blood to the liver
- Enlarged spleen: often related to portal hypertension
- Varices: swollen veins in the food pipe or stomach that may bleed
- Ascites: fluid collecting in the abdomen
- Poor growth: caused by reduced calorie and vitamin absorption
- Bleeding or bruising: linked to poor vitamin K absorption or reduced liver function
- Liver failure: when the liver cannot perform its essential functions adequately

Prevention

EHBA cannot currently be prevented. No proven action during pregnancy, feeding choice, vaccine, or supplement stops it from developing.

Early recognition cannot prevent EHBA, but it can reduce treatment delays. Persistent jaundice, dark urine, and especially pale or white stools need prompt assessment.

Living with it and what to expect

The course after a Kasai operation varies. In the following weeks and months, darker stools and falling bilirubin levels suggest improved bile drainage. Jaundice may fade gradually.

Some children maintain useful bile flow and their own liver for many years. Others continue to have jaundice or develop complications despite surgery. Even after a good early response, scarring and portal hypertension may progress later. Many people with EHBA eventually need a liver transplant, although symptoms alone cannot predict when.

Ongoing care usually includes blood tests, growth checks, nutrition review, imaging, and monitoring for infection or portal hypertension. Appointments remain important even when a child looks well.

Feeding and growth may require extra support in infancy. As health improves, many children can attend nursery or school, participate in suitable physical activity, and follow ordinary family routines. Activity advice may change if the spleen is enlarged because an abdominal impact could cause injury.

After transplantation, many children have improved liver function and growth. They still need lifelong specialist care, anti-rejection medicines, blood tests, and monitoring for rejection or medicine side effects.

The liver team can also help with nutrition, medicines, school planning, emotional support, and preparation for transplant if needed.

When to get medical help

Arrange prompt medical assessment if a baby:

These signs need timely investigation but do not confirm EHBA by themselves.

After a Kasai operation, contact the child's liver team urgently for fever, worsening jaundice, newly pale stools, unusual irritability, vomiting, or reduced feeding. These may indicate cholangitis, which often needs rapid treatment to protect the liver.

Seek emergency help if a child:

Continue routine follow-up even without new symptoms. Blood tests may detect changes before they are visible at home.

- Is still jaundiced after two weeks of age
- Develops new or worsening jaundice
- Passes pale, grey, white, or chalky stools
- Has dark urine that stains the nappy

- Is feeding poorly or not gaining weight
- Vomits blood or passes black, tar-like stool
- Is very difficult to wake, unusually floppy, or confused
- Has severe breathing difficulty
- Develops a rapidly increasing, tense abdomen
- Has uncontrolled bleeding
- Shows severe dehydration, such as very few wet nappies and marked drowsiness

Common questions

Is EHBA the same as newborn jaundice?

No. Common newborn jaundice usually improves during the first two weeks and does not cause pale stools. EHBA blocks bile drainage and increases direct bilirubin. Jaundice lasting beyond two weeks needs assessment.

What colour is an EHBA stool?

Stools may be cream, light grey, white, chalky, or putty-coloured because little bile reaches the intestine. Colour can vary between nappies. Take a photograph and seek prompt advice if a stool appears unusually pale.

Does the Kasai operation cure EHBA?

The Kasai operation may restore some bile flow, but it does not create normal ducts or remove existing liver scarring. Some children do well for years with their own liver. Others need a transplant sooner, and many eventually require one.

Will my child definitely need a liver transplant?

Not necessarily straight away, but transplantation is common. Its timing depends on bile drainage, growth, infections, portal hypertension, and liver function. Regular monitoring helps the team identify when transplantation offers more benefit than continuing with the existing liver.

Is EHBA genetic?

EHBA is not usually inherited in a predictable way, and most families do not have another affected child. Rare genetic or developmental factors may contribute, but no single gene explains most cases.

Can EHBA come back after a liver transplant?

The original blocked ducts do not return in the transplanted liver. However, transplantation can cause complications such as rejection, infection, and medicine side effects. Lifelong follow-up remains necessary.

Sources

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