CONDITION

Extrahepatic biliary atresia

Extrahepatic biliary atresia is an infant condition in which scarred bile ducts prevent bile from draining out of the liver.

WHAT IT IS
Blocked, scarred bile ducts beginning in infancy

BODY SYSTEM
Liver and digestive system

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

WHAT CAUSES IT
Usually unknown; likely inflammatory injury around birth

HOW IT IS DIAGNOSED
Cholangiography, supported by liver biopsy and blood tests

MAIN TREATMENT
Early bile-drainage surgery and sometimes liver transplantation

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

IS IT AN EMERGENCY?
Yes - suspected cases need urgent specialist assessment

LONG-TERM OUTLOOK
Many need transplantation; others keep their native liver

Overview

Extrahepatic biliary atresia, usually called biliary atresia, is a condition that begins in early infancy. The bile ducts outside the liver become inflamed, scarred and blocked. Smaller ducts inside the liver may also be affected.

Bile is a digestive fluid made by the liver. It normally travels through the bile ducts into the intestine, where it helps digest fat. When the ducts are blocked, bile builds up in the liver. This causes jaundice and can gradually damage liver tissue.

Biliary atresia needs prompt specialist assessment and surgery. Treatment works best when it begins early, before extensive liver scarring develops. Surgery can improve bile drainage, but it does not create normal bile ducts or remove the need for long-term monitoring.

This condition is not cancer, and it is not contagious.

Symptoms

Symptoms usually appear during the first weeks of life. A baby may initially look well and feed normally even while liver damage is developing.

The main signs are:

Normal baby stools may be yellow, green or brown. Very pale stools happen because little or no bile is reaching the intestine. Stool colour can sometimes change from one nappy to the next, so take a photograph or keep the nappy if you are concerned.

Other possible symptoms include a swollen abdomen, an enlarged liver or spleen, easy bleeding and irritability. These may become more noticeable as liver damage progresses.

Jaundice is common in newborns and often has a harmless cause. However, jaundice that lasts beyond the early newborn period, especially with pale stools or dark urine, needs prompt testing. Breastfeeding itself does not explain pale stools.

- Yellow skin or eyes, called jaundice, that persists or becomes more noticeable
- Pale, grey, white or putty-coloured stools
- Dark yellow or brown urine that stains the nappy
- Slow weight gain or poor growth

Causes and risk factors

The exact cause of most biliary atresia is unknown. Researchers think injury to the bile ducts triggers inflammation and scarring before or shortly after birth. Possible triggers under study include abnormal immune responses, problems in bile-duct development and certain infections. No single infection has been established as the usual cause.

Most babies have biliary atresia without other birth differences. A smaller group has differences involving organs such as the spleen, heart or intestines. This is sometimes called syndromic biliary atresia.

Biliary atresia is not usually considered an inherited condition. Having one child with it does not mean another child will necessarily be affected. Families concerned about associated birth differences or recurrence can ask about genetic counselling.

Nothing a parent ate, did or failed to do during pregnancy is known to cause biliary atresia.

Diagnosis

No symptom alone can confirm biliary atresia. Because several liver and metabolic conditions can cause infant jaundice, testing aims to identify blocked bile flow quickly while checking for other explanations.

A blood test measures total bilirubin and direct, or conjugated, bilirubin. Bilirubin is a yellow waste substance processed by the liver. A raised direct bilirubin level suggests that bile is not flowing normally. Other blood tests assess liver inflammation, blood clotting, protein levels and general health.

An abdominal ultrasound looks at the liver, gallbladder, bile ducts and nearby organs. It may show features that support the diagnosis, but a normal-looking ultrasound does not completely rule out biliary atresia.

A liver biopsy may be performed. A small sample of liver tissue is examined under a microscope for bile blockage, inflammation and scarring. The result can strongly support the diagnosis and help separate biliary atresia from other liver conditions.

Cholangiography is the test that shows whether bile can travel through the ducts. Contrast dye is placed into the biliary system, and images are taken. This is often done during an operation so that, if the ducts are blocked, the surgeon can proceed directly to treatment.

A nuclear medicine bile-flow scan is sometimes used, but it cannot reliably confirm biliary atresia by itself.

Treatment and management

Care is usually coordinated by a specialist children's liver and surgical team. Treatment addresses the blocked ducts, nutrition and complications of liver disease.

The first operation is usually a Kasai portoenterostomy. During this procedure, the surgeon removes the scarred ducts outside the liver and connects a loop of intestine directly to the area where tiny bile channels leave the liver. The aim is to give bile another route into the intestine.

The Kasai procedure is not a complete cure. Some babies develop good bile drainage and their jaundice improves. Others have limited or no drainage. Outcomes tend to be better when surgery is performed promptly, which is why suspected biliary atresia is assessed urgently.

After surgery, care may include:

Families should use vitamins, medicines and feeding products only under the specialist team's

direction. Children with poor bile flow can absorb medicines and nutrients differently from other children.

A liver transplant may be needed if the Kasai procedure does not provide enough drainage or if serious liver complications develop. Reasons can include worsening liver function, repeated bile-duct infections, poor growth or severe pressure in the veins around the liver.

Transplantation replaces the affected liver, but it requires lifelong follow-up and medicines that prevent rejection of the donated liver.

- Extra nutritional support and higher-calorie feeds
- Special fats that are easier to absorb
- Vitamins A, D, E and K, which need bile for absorption
- Medicines to support bile flow in selected children
- Antibiotics like amoxicillin or doxycycline when needed to prevent or treat bile-duct infection
- Regular blood tests, growth checks and liver imaging

Complications

Ongoing bile blockage can cause fibrosis, meaning scar tissue within the liver. Extensive scarring is called cirrhosis. Cirrhosis can interfere with liver function and restrict blood flow through the liver.

This restricted flow may cause portal hypertension, or high pressure in the vein carrying blood from the digestive organs to the liver. It can lead to an enlarged spleen, low platelet counts, fluid in the abdomen or swollen veins in the food pipe and stomach. These veins, called varices, can bleed.

Cholangitis is an infection involving the bile channels, particularly after a Kasai procedure. Symptoms may include fever, worsening jaundice, pale stools, dark urine, irritability or poor feeding. It needs urgent medical assessment and usually hospital treatment.

Poor bile flow can also make it difficult to absorb fat and fat-soluble vitamins. Without adequate nutritional support, a child may have poor growth, weak bones or problems with blood clotting.

Prevention

Biliary atresia cannot currently be prevented. There is no established action during pregnancy or infancy that stops it from developing.

Early recognition cannot prevent the condition, but it can reduce delays in treatment. Checking stool colour and arranging prompt assessment for persistent jaundice, pale stools or dark urine can help babies reach specialist care sooner.

Living with it and what to expect

The months after a Kasai procedure focus on whether bile drainage improves, whether jaundice clears and whether the baby grows. Blood tests and stool colour help the liver team assess progress.

Even when the operation works well, the liver is not considered completely normal. Scarring, portal hypertension or infections may develop later. Regular follow-up is therefore needed throughout childhood and into adult life.

Some children keep their own liver for many years. Others need a transplant in infancy, childhood or adulthood. It is not always possible to predict the course from the first few weeks after surgery.

Follow-up may include blood tests, ultrasound scans, nutrition reviews and checks for an enlarged spleen or varices. Appointments may become less frequent during stable periods, but they remain important even when a child feels well.

Many children attend school, play and take part in family life. Energy, diet and activity may need adjustment during periods of illness or when liver complications are present. After a successful transplant, many children have good day-to-day function, although anti-rejection medicines and transplant monitoring continue for life.

Keeping a written care plan can help families know which symptoms require emergency care, which hospital to attend and whom to contact after hours.

When to get medical help

Arrange urgent same-day medical assessment if a baby has pale, grey or white stools, dark urine, or jaundice that is persisting or worsening. These signs do not prove biliary atresia, but direct bilirubin testing should not be delayed.

After a Kasai procedure, seek urgent medical advice for fever, worsening jaundice, newly pale stools, poor feeding, vomiting or unusual sleepiness. These can be signs of cholangitis or

another infection.

Seek emergency care now for:

A baby younger than 3 months with a fever also needs prompt emergency assessment, particularly after liver surgery.

Routine follow-up is appropriate for stable concerns such as feeding-plan adjustments, vitamin monitoring, gradual growth changes or help managing scheduled medicines. Do not wait for the next routine appointment if jaundice, stool colour, fever or your child's overall condition changes suddenly.

- Vomiting blood or passing black, tar-like or visibly bloody stools
- Severe breathing difficulty, collapse or difficulty waking
- A rapidly enlarging or painful abdomen
- Significant uncontrolled bleeding
- Signs of severe dehydration, such as very few wet nappies and marked drowsiness

Common questions

Is biliary atresia genetic?

Most cases are not inherited in a clear pattern. A minority occur alongside other differences present from birth. Genetic assessment may be offered when these features suggest a broader condition.

Did I cause my baby's biliary atresia?

No parental action is known to cause it. It has not been linked to a particular food, medicine or routine activity during pregnancy.

Can my baby still breastfeed?

Breast milk may remain part of feeding, but babies with poor bile flow often need extra calories, special fats or vitamin supplements. A liver dietitian can create a feeding plan based on growth and blood tests.

Does the Kasai operation cure biliary atresia?

No. It creates another route for bile to drain and may protect liver function for years. Liver scarring can still progress, so long-term monitoring remains necessary.

Will my child definitely need a liver transplant?

Not every child needs one at the same age or for the same reason. Many eventually require

transplantation, while others live for years with their own liver. Bile drainage, growth, infections and signs of liver damage help guide the timing.

Can pale stools return after surgery?

Yes. Pale stools may mean that bile flow has decreased, especially when accompanied by fever, dark urine or worsening jaundice. Contact the liver team urgently rather than waiting for a routine appointment.

Sources

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