

FACTSHEET

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Biliary atresia

What is biliary atresia?

Biliary atresia is a rare disease of the liver that scars and blocks the bile ducts. Because the bile is unable to drain, it builds up in the liver and damages the liver.

Bile ducts are tubes inside and outside the liver which carry bile to the intestine. Bile is a green brown fluid that helps with digesting food.

Why does biliary atresia occur?

The cause of biliary atresia is not known. Suggested causes include viral infections or an over-response of the body's immune system. It is usually not a hereditary disease and is unlikely to occur more than once in a family. Biliary atresia only occurs about once in every 15,000-20,000 births worldwide. In a small percentage of patients it occurs in association with other abnormalities of the heart or abdominal organs.

Symptoms of Biliary Atresia

Symptoms or signs of biliary atresia typically appear in the first two months of life. These may include:

- **Jaundice** - A yellow appearance of the skin and whites of the eyes (sclera) can be present in many newborn babies. In biliary atresia, this jaundice does not improve within 1 to 2 weeks. The jaundice is due to the build-up in the body of a substance called bilirubin which is normally removed from the body via the bile.

- **Urine** – Increased bilirubin in the bloodstream can make the urine appear very dark yellow or brown.
- **Stools** – Because bile and bilirubin does not reach the intestine, the stools can appear pale or clay-coloured.

Diagnosis

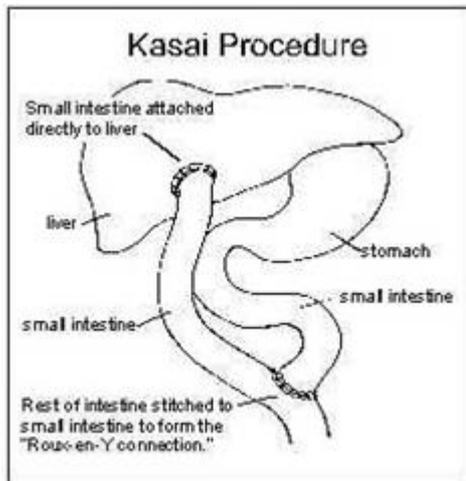
Several other liver diseases can give the same symptoms as biliary atresia. Therefore a series of tests are needed to work out the cause of these symptoms. These tests include blood tests, urine tests, an ultrasound scan of the liver, tests to look at flow of bile through the liver (nuclear medicine scan or cholangiogram) and tests to look directly at the liver tissue (a biopsy). Biliary atresia is usually diagnosed after checking the results of many of these tests. The investigation of an infant with suspected biliary atresia should be undertaken in a children's hospital.

Treatment of biliary atresia

Babies with biliary atresia need surgery. Usually an operation called a portoenterostomy (sometimes called a Kasai procedure) is performed. This operation involves removing the blocked ducts and connecting the liver directly to the small intestine so that bile may be able to drain away from the liver.

An early operation can improve the outcome of biliary atresia. About 25% of infants will have good bile flow after surgery while 50% will have some bile flow. The remaining 25% will have little or no bile flow and the only option then is liver transplantation. Some infants initially improve but then continue to develop worsening liver disease and need liver transplantation. All infants who

have biliary atresia need to be monitored closely by specialist doctors and teams through childhood, even if they are doing well. Biliary atresia is the most common reason for liver transplantation in children, and long term survival is currently very good.



Complications of biliary atresia

- **Cholangitis** – This is an infection of the tiny bile ducts in the liver caused by bacteria moving up from the bowel into the liver. It is a common problem after the Kasai procedure and can be life threatening as infants can become very sick very quickly. Symptoms include irritability, fever, increased jaundice and poor appetite. Any child who has had a Kasai procedure who develops any of these symptoms needs to seek medical treatment. Cholangitis is treated with intravenous antibiotics in hospital.
- **Failure to thrive** – Bile is needed for fat digestion in the intestine. So poor bile drainage results in poor growth and deficiency of certain vitamins. Vitamins A, D, E and K can be given orally to prevent deficiencies of these vitamins. Special infant formulas are usually needed to help with growth.
- **Cirrhosis** – This is scarring of the liver that results from long term damage. **Cirrhosis itself can also impair growth and physical development of the child.**
- **Portal hypertension** - As the liver becomes scarred, blood vessels travelling through the liver become constricted. This impairs the flow of blood and increases the pressure in these veins, specifically in the portal vein (a major vein of the liver system). Increased pressure in the veins connected to the portal vein lead to an enlarged spleen and possibly to ascites or varices (see below).
- **Ascites** – Excess fluid in the abdominal cavity which can cause the child's tummy to be enlarged.
- **Varices** – These are swollen veins in the gastrointestinal tract that form due to portal

hypertension. Varices can lead to bleeding. This bleeding can cause dark or black-coloured stools and blood-stained vomit. If this occurs, the child should be taken to the nearest hospital immediately.

Long term outlook

Many children with biliary atresia will have a good response to surgery and will not need major medical interventions in childhood and adolescence. They do need follow up in a specialist clinic to make sure their growth and nutrition is normal and that any complications are detected early. Some infants and children with biliary atresia develop progressive liver disease and will need to have a liver transplant in childhood.

Useful Websites

Liver Kids Organisation

www.liverkids.org.au

Children's Liver Disease Foundation (UK)

www.childliverdisease.org

Children's Liver Association for Support Services (USA)

www.classkids.org

Remember

- Your specialist, together with your GP, will care for your child with Biliary Atresia.