

# Fact Sheet

## Biliary Atresia



### What is Biliary Atresia?

Biliary Atresia is a rare disease of the liver that destroys the bile ducts, which carry bile from the liver to the intestine. This disease involves the progressive destruction of these bile ducts, especially outside the liver, sometimes inside the liver as well. Because the bile is unable to drain, it builds up in the liver and damages the liver.

### 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 own immune system. It is 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.

### Symptoms of Biliary Atresia

Symptoms or signs of biliary atresia typically appear within the first two weeks to 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 of excess bilirubin throughout the body as the liver isn't able to clear it properly.
- **Urine** - Appears very dark yellow or brown: this is due to the increased bilirubin in the bloodstream, which then passes to the kidneys.
- **Stools** - Appear pale or clay-coloured. This happens because there is little or no bile reaching the intestine to colour the bowel movements.
- **Enlarged Liver** - Feels larger and harder than normal.
- **Poor Weight Gain** - Infants with biliary atresia often do not gain

weight. Bile is required to digest and absorb most types of fat.

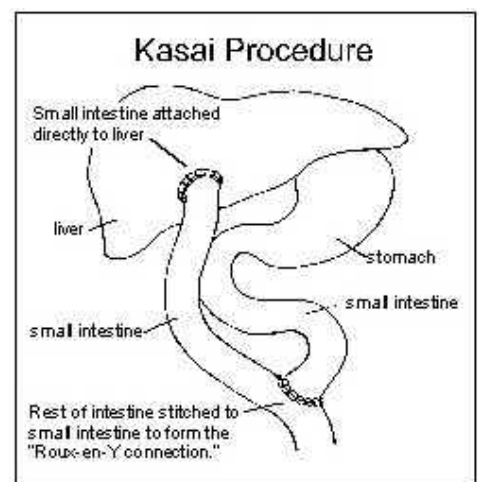
### Diagnosis

Several other liver diseases can give the same symptoms seen in biliary atresia. Therefore a series of tests are required to work out the cause of these symptoms. These tests include various blood tests, urine tests, ultrasound scan of the liver, tests to look at flow of bile through the liver 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.

### Treatment of Biliary Atresia

One particular type of operation, the Kasai Procedure, can be done to help re-establish bile flow from the liver to the intestine. This operation involves connecting the liver directly to the small intestine with a loop of the small intestine. This bypasses the blocked ducts, letting bile flow from the small bile ducts inside the liver straight into the intestine.

An early operation can improve the outcome of biliary atresia. 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 will require liver transplantation. Liver transplantation is the only cure for biliary atresia and most children needing a transplant do very well long-term.



# Fact Sheet

## Biliary Atresia



### Complications of Biliary Atresia

- **Cholangitis** - An inflammation of the bile ducts caused by bacteria moving up from the bowel. This is a common problem after the Kasai procedure, because bacteria are present in the loop of the intestine used to drain the bile. Symptoms include irritability, fever, increased jaundice and poor appetite. Medical treatment should be sought immediately to treat the infection and prevent further liver damage. Cholangitis is treated with IV antibiotics in hospital.
- **Failure to Thrive** - Lack of bile salts in the intestine, which are needed for fat digestion, result in poor growth and fat-soluble vitamin deficiency. 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** - is when scarring to the liver occurs causing irreversible damage. Once cirrhosis develops the functions of the liver start to be interrupted and liver transplantation is considered.
- **Portal Hypertension** - As the liver becomes scarred, veins 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) leading to a big spleen and fluid problems (such as ascites).
- **Ascites** - Ascites is a complication of portal hypertension. This occurs when there is a build up of fluid in

the space between the lining of the abdominal wall and the lining of the organs.

- **Varices** - Another complication of portal hypertension is varices. Varices occur when the increased pressure causes blood to flow through smaller veins, which leads them to weaken and swell. Varices can lead to bleeding. This bleeding can cause dark or black-coloured stools and blood-stained vomit. Immediate medical attention is necessary.

### Long term outlook

There are many factors that can affect the long term progress and outcome of children with biliary atresia after their Kasai operation. These factors include the extent of bile duct damage, the child's age at the time of operation, the extent of overall liver damage that has occurred and the nutritional health of the child.

### Useful Websites

[www.liverkids.org.au](http://www.liverkids.org.au) - Liver Kids Organisation

[www.childrenliverdisease.org](http://www.childrenliverdisease.org) - Children's Liver Disease Foundation (UK)

[www.classkids.org](http://www.classkids.org) - Children's Liver Association for Support Services (USA)

### Remember

**Your specialist, together with your GP, will manage your child with Biliary Atresia.**

This fact sheet is for education purposes only. Please consult with your doctor or other health professional to make sure this information is right for your child.

This document was reviewed on Thursday, 7<sup>th</sup> August 2008.

the children's hospital at Westmead

[www.chw.edu.au](http://www.chw.edu.au)

 **SYDNEY CHILDREN'S HOSPITAL RANDWICK**

[www.sch.edu.au](http://www.sch.edu.au)

 **Kaleidoscope**  
HUNTER CHILDREN'S HEALTH NETWORK

[www.kaleidoscope.org.au](http://www.kaleidoscope.org.au)