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The American Liver Foundation is a nonprofit, national voluntary health organization dedicated to the prevention, treatment, and cure of hepatitis and other liver diseases through research, education, and advocacy.

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What is Biliary Atresia?

Biliary Atresia

Biliary atresia is a serious disease that affects young infants. It is the most common indication for liver transplantation in young children. The cause of biliary atresia is unknown.

What is biliary atresia?

Biliary atresia is a serious disease that occurs in young infants. It results in inflammation and obstruction of the ducts which carry bile from the liver into the intestine. Since bile cannot flow normally, it backs up in the liver (a situation called "biliary stasis"). This results in jaundice, or a yellowing of the skin, and causes cirrhosis, in which healthy liver cells are destroyed and replaced with scar tissue. This scarring interferes with blood flow through the liver, causing more cell damage and scarring.

What are the first signs of biliary atresia?

The symptoms of biliary atresia are usually evident between two and six weeks after birth. The baby will appear jaundiced, and may develop a large, hardened liver and a swollen abdomen. The stools are usually pale grey and the urine may appear dark.

Some babies may develop intense itching, or pruritus, which makes them extremely uncomfortable and irritable. The exact cause of this itching is not yet known, although researchers have found a connection between it and the backup of bile.

What causes biliary atresia?

The cause of biliary atresia has not yet been discovered.

The disease affects approximately one infant in every 15,000 live births. Girls are affected slightly more often than boys, but no racial or ethnic group appears to be more affected than any other.

Biliary atresia is not known to be a hereditary condition. Many parents experience feelings of guilt, but they should be reassured that nothing they have done caused their child's illness.



How is biliary atresia diagnosed?

There are many liver diseases that cause symptoms similar to those of biliary atresia. Consequently, many tests may have to be performed before biliary atresia can be diagnosed conclusively.

Every effort should be made to search for any of the causes of jaundice, which might be confused with biliary atresia. This involves blood and urine tests, liver tests, blood counts, and tests for clotting function. A painless examination using ultrasound is often done to study the liver and determine the size of the bile ducts and gallbladder.

Other tests, which are often used, are specialized X-ray techniques or radioactive scans of the liver, which can be helpful in focusing on the true abnormality. A liver biopsy, in which a tiny sample of the liver is removed with a needle, allows the physician to examine the liver tissue microscopically.

What about treatment?

There is no cure for biliary atresia. The most successful treatment for biliary atresia to date is a type of surgery that allows drainage of bile from the liver when the ducts have become completely obstructed. This operation is called the Kasai procedure (hepatopertoenterostomy) after Dr. Morio Kasai, the Japanese surgeon who developed it.

In the Kasai procedure, the surgeon removes the damaged ducts outside of the liver (extrahepatic) and replaces them with a length of the baby's own intestine, which acts as a new duct.

The aim of the Kasai procedure is to allow excretion of bile from the liver into the intestine via the new duct. The operation accomplishes this about 50% of the time.

In those who respond well, jaundice usually disappears after several weeks.

In the remaining 50% of cases where the Kasai procedure does not work, the problem often lies in the fact that obstructed bile ducts are "intra-

hepatic" or inside the liver, as well as outside. No procedure has yet been developed to correct this problem except for liver transplantation.

What happens after surgery?

The aim of treatment after surgery is to encourage normal growth and development. If bile flow is good, the child is given a regular diet. If bile flow is reduced, a modified fat diet is recommended, since the absorption of fats and vitamins is impaired. Multiple vitamins, vitamin B complex, and vitamins E, D, and K should be given as supplements.

Is the Kasai procedure a cure for biliary atresia?

Unfortunately, despite bile flow, the Kasai procedure is not a cure for biliary atresia. For reasons that are still unknown, liver damage often continues and, eventually, cirrhosis and its complications may appear.

What are the complications?

Patients with cirrhosis have changes in blood flow through the liver that may produce abnormalities, such as easy bruising of the skin, nosebleeds, retention of body fluid, and enlarged veins, called varices, in the stomach and esophagus. Increases in pressure in these veins can make them "leaky" and internal bleeding or vomiting of blood results. In some cases, a procedure may be required to stop the bleeding, whereby a hardening (sclerosing) agent is injected into these veins to prevent bleeding.

What can be done about these complications?

Following the Kasai operation, infection in the bile ducts (cholangitis) is common. This is usually treated using antibiotics.

If retention of body fluid occurs, it can be treated with diuretics and potassium replacement.

Itching can be treated with medications such as ursodeoxycholic acid.

What is the outlook for a baby with biliary atresia?

The extent and type of liver damage differ in each baby with biliary atresia. Some infants respond to the Kasai procedure; others do not. If bile continues to flow, long-term survival is possible. However, it is presently impossible for a physician to determine in advance which baby is likely to respond to treatment.

Is liver transplantation the solution?

Liver transplantation is an option that is becoming increasingly useful for certain liver diseases. The survival rates for transplant recipients have increased dramatically with improved surgical techniques and the development of drugs that help overcome the problem of organ rejection.

In children with biliary atresia, liver transplantation is generally not attempted until the Kasai procedure has been performed. If this operation is not successful, and before complications of the resulting cirrhosis become severe and life threatening, liver transplantation may be attempted. As in all organ transplantation, success depends greatly upon the timely availability of suitably matched organs for donation. The use of reduced-size and living-related transplants are aiding in the timing and availability of suitable donor organs.

What can the family do?

Watching a young infant suffer from biliary atresia is a devastating experience. It can also be frustrating because so little is known about the disease. Feelings of anger and helplessness are not uncommon.

Many parents have found it helpful to learn as much as they can about the disease. Talk to your physician, inquire about specialists, and request any literature on the subject.

Perhaps the biggest comfort for parents is to discuss the problem with others who have or are going through a similar experience. Finding out that they

are not alone, that others feel the way they do, and learning how other parents are coping with their child's disease, is often a great comfort.

Where can parents turn for support?

The American Liver Foundation recognizes that parents of children with biliary atresia need help in coping with the immense strain of this chronic illness. To meet this need, the American Liver Foundation is continually organizing and coordinating mutual help groups through its chapters to provide emotional support for families, making referrals to specialists where appropriate, and keeping people aware of the latest research developments.

Will there ever be a cure for biliary atresia?

There can be no cure for biliary atresia until the cause of the disease can be determined. Researchers are focusing on trying to find this cause, but a great deal of work still needs to be done. More research into how the liver works is also vital.

Research is the key that will unlock this mystery. The American Liver Foundation is the only national voluntary health agency dedicated to funding research and helping people understand more about the liver and liver disease.