



The Childhood Liver Disease Research and Education Network strives to provide information and support to individuals and families affected by liver disease through its many research programs.

## **BILIARY ATRESIA**

### **What is biliary atresia?**

Biliary atresia is an inflammation of the large bile ducts outside the liver that irreversibly blocks bile flow from the liver to the small intestine. The cause of biliary atresia is unknown. It is estimated that biliary atresia affects 1 in 8000 to 15,000 live births worldwide. Biliary atresia is the principal cause of chronic cholestatic liver disease in children, and a common reason for liver transplantation in children.

### **What are the symptoms?**

Clinical features of biliary atresia include infants with jaundice, acholic stools, dark urine, hepatomegaly and splenomegaly. Other symptoms can include pruritis and ascites.

### **How is it diagnosed?**

Early identification of biliary atresia is crucial as long term outcome is dependent upon age of treatment. There are a number of tests which suggest the diagnosis of biliary atresia but there is only one 'gold standard' and that is the intraoperative cholangiogram. Before this is done the infant goes through a few days of blood tests and x-rays and often a liver biopsy, all of which help point to or away from the diagnosis of biliary atresia. Please see Frequently Asked Questions for a detailed description of these tests.

### **How is treated?**

Upon diagnosis of biliary atresia, surgical treatment known as the Kasai procedure or hepatoportoenterostomy is performed. The obstructed bile ducts are removed and a section of the small intestine is directly connected to the liver using a Roux-en-Y method to initiate bile flow directly from the liver to the small intestine.

If the hepatoportoenterostomy is successful, jaundice disappears and bilirubin levels return to normal. The stool color returns to the normal yellow, green or brown and the pale color resolves. After hepatoportoenterostomy, antibiotics are given to prevent infection of the liver from the small intestine. Fat soluble vitamins, ursodeoxycholic acid (Actigall) and specialized predigested formulas are given to ensure proper nutrition continues.



## **What is the outlook for infants diagnosed with biliary atresia?**

The chance of a successful Kasai procedure is best if done before 2 months of age. Almost all infants diagnosed with biliary atresia will undergo a Kasai procedure. The only exception is an infant who has advanced liver disease at the time of diagnosis. Such infants are usually not diagnosed until after 3 - 4 months of age and are often listed for liver transplantation without undergoing a Kasai. However, even with early intervention, scarring of the liver can continue, resulting in cirrhosis. Before the Kasai was developed, no children survived a diagnosis of biliary atresia. A liver transplant will be required if the scarred liver's function fails. About half of infants who undergo a Kasai procedure will still need liver transplantation by three years of age. About one quarter will clear their jaundice initially but will probably need liver transplantation by the teenage years because of slowly progressive cirrhosis. Only about one quarter of those initially undergoing the Kasai will survive to their 20's without needing liver transplantation. Although liver transplantation is a major surgical procedure requiring long term immunosuppression, this life-saving procedure has markedly improved survival rates of infants and children with biliary atresia.