What happens to children with biliary atresia?

If left untreated, the blocked bile flow will lead to further liver damage such that few children with biliary atresia survive beyond the age of two. When bile flow is only partly restored by surgery, the complications of cirrhosis – a condition in which the liver is scarred and damaged, will gradually develop. If the Kasai procedure is successful in draining bile from the liver with complete disappearance of the jaundice, children may live for many years and grow normally. Some may never need a liver transplant.

Sometimes, despite successful surgery, slow and progressive damage to the liver still continues. When progressive cirrhosis occurs, it would eventually result in the need for a liver transplant.

Liver transplantation plays an important role in the long-term treatment of children with biliary atresia. Paediatric liver transplantation has evolved into a highly successful treatment and now offers significant hope for all children with biliary atresia.

Other Instructions

About the Khoo Teck Puat – National University Children's Medical Institute (KTP-NUCMI)

The KTP-NUCMI is the paediatric arm of the National University Hospital and comprises the Departments of Paediatrics, Paediatric Surgery and Neonatology. We provide comprehensive and specialised medical and surgical services for newborns, children and adolescents. NUH is the only public hospital in Singapore that offers paediatric kidney and liver transplant programmes. Through a generous gift from the Estate of Khoo Teck Puat, we have set up an integrated outpatient facility with medical, diagnostic and rehabilitation services.

For more information about us, visit www.nuh.com.sg/ktp-nucmi.

Contact Us

24-hour Children's Emergency
Location: NUH Main Building, Zone F, Level 1
General Enquiry: +65 6772 5000

KTP-NUCMI
Location: NUH Main Building, Zone E, Level 2
(Accessible via Kent Ridge Wing, Zone C, Level 2, Lift Lobby C)
Operating Hours: 8.30am – 5.30pm (Mon to Fri), 8.30am – 12.30pm (Sat)
General Enquiry: +65 6772 5736 Appointment Line: +65 6908 2222
Email: ktpnucmi_appt@nuhs.edu.sg

9a Viva-University Children's Cancer Centre
Location: NUH Medical Centre, Zone B, Level 9
Operating Hours: 8am – 5.30pm (Mon to Fri)
Appointment Line: +65 6772 5030 Fax: +65 6872 4314
Email: cbccappt@nuhs.edu.sg

NUH Children's Urgent Care Clinic @ Bukit Panjang
Location: Junction 10, 1 Woodlands Road, #01-22, Singapore 677899
Operating Hours: 9am – 11pm daily (including public holidays)
General Enquiry: +65 6219 1538 Email: childrenucc@nuhs.edu.sg
Website: www.nuh.com.sg/ucc

NUH Child Development Unit @ JMC
Location: Jurong Medical Centre,
60 Jurong West Central 3, Level 2, Singapore 648346
Operating Hours: 8.30am – 5.30pm (Mon to Fri)
Appointment Line: +65 6665 2530/2531 Fax: +65 6665 0158
Email: cdu@nuhs.edu.sg Website: www.nuh.com.sg/cdu

NUH Child Development Unit @ Keat Hong
Location: Keat Hong Community Club,
2 Choa Chu Kang Loop, #03-01, Singapore 689687
Operating Hours: 8.30am – 5.30pm (Mon to Fri)
Appointment Line: +65 6769 4537/4637 Fax: +65 6665 0158
Email: cdu@nuhs.edu.sg Website: www.nuh.com.sg/cdu

National University Hospital
Location: 5 Lower Kent Ridge Road, Singapore 119074
Tel: +65 6908 2222 Email: contactus@nuhs.edu.sg
Website: www.nuh.com.sg

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Information is correct at the time of printing (October 2022) and subject to revision without prior notice.

National University Hospital

Biliary Atresia

Khoo Teck Puat – National University Children's Medical Institute

What is biliary atresia?

Biliary atresia is a condition where the bile ducts drainage system of the liver becomes progressively blocked. It typically presents in the first few weeks of life. When this happens, the bile that is trapped causes damage and scarring to the liver.

On average, there is one case of biliary atresia out of every 15,000 live births. Females are affected slightly more often than males.

What causes biliary atresia?

The cause of biliary atresia is unknown. Auto-immune mechanism may be partly responsible for the progressive process that takes place. Research also suggests that a viral infection may be linked with biliary atresia. About 10% of cases have other associated defects in the heart, blood vessels, intestine, or spleen.

Although the cause is uncertain, it is known that biliary atresia affects only newborns and it is not hereditary, not contagious and unpreventable. Parents can be assured that biliary atresia is not caused by anything the mother did during pregnancy.

How is biliary atresia diagnosed?

The usual history is a full-term infant who appears normal at birth but develops jaundice subsequently. The infant will have yellow eyes and skin, light-coloured stools and dark urine caused by build-up of bilirubin in the blood. The abdomen may be swollen with a firm and enlarged liver. Weight loss and irritability may develop as the level of jaundice increases.

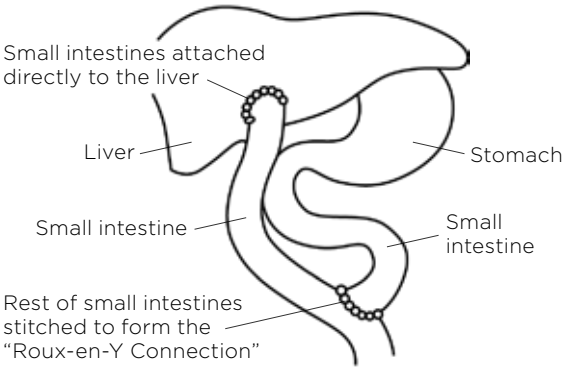
Several tests are needed to make a diagnosis of biliary atresia or to detect any underlying cause for persistent jaundice in an infant. These may include:

- Blood tests
- Ultrasound scan of the liver and gall bladder
- HIDA scan (radioactive scan of the biliary system)
- Liver biopsy
- A surgical procedure (to examine the bile ducts)

How is biliary atresia treated?

The treatment for biliary atresia is an operation called the “Kasai” procedure. Essentially, the surgeon will remove the blocked bile ducts and connect a loop of small intestine directly to the liver. This allows bile to flow from the liver directly to the intestine. This segment of intestine that connects to the liver forms a Y-shaped connection and is called a “Roux-en-Y hepatoportojejunostomy”.

Kasai Procedure



Early diagnosis of biliary atresia is important as the success rate of surgical intervention is higher. Better results have been found when Kasai procedure is performed in children less than 100 days old. Some children would still experience no bile flow even after the Kasai procedure. These patients would then require a liver transplant.

What complications should I take note of?

The most common complication that can occur after the Kasai procedure is infection of the bile duct system within the liver. This is called “cholangitis”. Bacteria normally found in the small intestine can migrate up the Roux-en-Y causing infection. Signs of cholangitis include fever, increased jaundice and lighter colouration of stools. Antibiotics are needed to prevent this from occurring, or to treat it when it happens.

Adequate bile flow is needed for the digestion and absorption of dietary fats and fat-soluble vitamins, including vitamins A, D, E and K. When bile flow is reduced, poor growth and malnutrition may occur. Special formulas containing medium-chain triglycerides (an easily digested form of dietary fat) and water-soluble vitamin supplements are often prescribed to maximise the child’s growth and development.

Formation of scar tissue within the liver may cause hardening of the liver and a condition known as portal hypertension. Portal hypertension refers to increased pressure in the veins connecting the intestines and spleen to the liver. Complications of portal hypertension include problems with bleeding and clotting; enlarged weak veins in the oesophagus and stomach; and accumulation of fluid in the abdominal cavity called ascites. When these complications can no longer be treated effectively with medication, the child is referred for liver transplantation.