

13. What is successful Surgery and unsuccessful surgery?

At the time of the operation the surgeon will not be able to tell if the Kasai operation has been successful. Usually we wait for few weeks and check the bilirubin in blood.

Failure to clear jaundice after 3 months of surgery is deemed to be unsuccessful and these children require liver transplantation

Even if the jaundice clears after a Kasai operation some children will develop complications due to liver damage. Therefore, all children should continue to be seen for regular checkups.

14. Why the surgery can fail?

If the bile drainage system inside the liver is not formed properly the surgery might not help in jaundice clearance and there is no way we can predict this before surgery.

Dr. Rela Institute & Medical Centre is a multi-specialty quaternary care hospital located in Chennai, India.

The Institute is within the campus of Sree Balaji Medical College and Hospital, which is spread across 36 acres. It has 14 operating theatres with 450 beds, inclusive of 150 critical care beds.

The Institute is conveniently located 10 minutes from the Domestic and International Airport.

The hospital is designed to provide highly specialized care in various departments with a focus on multi-organ transplantation. Prof. Mohamed Rela, a world renowned surgeon in the field of Liver surgery and transplantation is the Chairman and Managing Director of the Institute.

In addition to quaternary & quality care, is also committed to provide day to day primary and secondary care to the local population, with facilities of international standards.

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**Diagnosis & Management
of Biliary Atresia**

Introduction

Bile is a yellow colour liquid produced by liver which is secreted into the intestine through a small tube called bile duct. Bile contains waste products and other products needed to digest food. The bile mixes with food in the intestine and gives the yellow colour of stools. In absence of bile stool will be white or clay colour.

1. What is biliary atresia?

Biliary atresia is a medical condition, where the bile ducts are not formed properly, resulting in stasis of bile in the liver. This in turn causes scarring (fibrosis) in the liver.

2. What causes biliary atresia?

The cause of biliary atresia is unknown. There is no evidence to suggest that biliary atresia is hereditary (can be passed on to children by their parents). Children with biliary atresia will not pass the condition on to their own children.

3. What are the signs of biliary atresia?

Presence of jaundiced beyond two weeks of birth (three weeks if pre-term) needs proper investigation. A simple blood test of split bilirubin could give a clue to this condition

The other important signs of pathological jaundice are:

■ Urine colour

Normally the urine of a newborn baby is colourless. If a baby's urine is persistently yellow or dark in colour this can be an indication of liver disease.

■ Pale Stools

Normally the stools of a baby are green or yellow. If the stools of a baby are grey, white, fawn or pale then this can be an indication of liver disease.

4. How is biliary atresia diagnosed?

If a baby has the signs and symptoms above then they may be admitted to hospital for further investigation.

A number of investigations will need to be carried out in order to make a diagnosis.

These may include:

- Blood tests
 - Urine test
 - An ultrasound
 - Some children will need a liver biopsy and/or a small operation to confirm or rule out a diagnosis.
- These investigations are completed over the course of a few days.

5. How can biliary atresia be treated?

If the tests show that there is a strong indication that your baby has biliary atresia, he/she will require surgery. This will involve carrying out an investigation called an operative cholangiogram under a general anaesthetic to confirm the diagnosis. If biliary atresia is confirmed then this will be followed by a "Kasai procedure", usually done under the same general anaesthetic at the same time. The aim of the operation is to help bile drain from the liver into the gut. The surgeon will join a loop of intestine to the under surface of the liver. The name of this procedure is Kasai operation (Kasai portoenterostomy).

6. When should the surgery performed?

Ideally the surgery should be performed before 8 weeks of life and beyond that the doctors can access the feasibility of surgery upto 16 weeks. Beyond that the success of surgery is practically nil. Such children should be assessed for liver transplantation directly.

7. How long will the operation last?

Usually the surgical time is around 4-5 hours

9. What happens immediately after the Kasai operation?

Usually the baby is kept in ICU for two days. The feeding is withheld for three to four days during which the baby would get nutrition through an intravenous drip.

Fluids are given in order to keep the sugar, salt and water levels at the right level in the body. Blood tests will be carried out to check these levels so that they can be changed if needed. All fluids required by your baby will be given through the drip until feeding is re-started.

10. When will my baby be able to leave hospital and what happens next?

If there are no problems after the surgery then your baby will be allowed to go home, usually around seven to ten days after the operation, once you and the medical team are happy with your baby's condition.

11. What should I do when I am back at home with my baby?

When you get home you should try and treat your baby as normally as possible. Obviously you will be worried at times and this is quite normal. Babies with biliary atresia get common illnesses just like any other baby. It is often helpful for your baby to be weighed on a weekly basis to monitor weight gain. It is important that your child receives their immunisations and to discuss timing and additional immunisations with their medical team.

Symptoms for which you should seek advice from your GP or hospital are:

- A raised temperature of 37.8°C or more, or non-specific illness with a raised temperature
- More jaundice with paler stools and darker urine
- The child is generally unwell

12. What medicines may my baby need and why are they given?

Some commonly used medicines are:

■ Antibiotics

After the operation antibiotics are given by a drip at first and then by mouth. They are used to reduce the risk of infection in the bile ducts (cholangitis). Further antibiotics may be given as needed.

■ Vitamins

Additional vitamins are necessary because poor bile flow can reduce the absorption of vitamins.

■ URSO (Ursodeoxycholic acid)

This drug is given after restarting feeding after the Kasai operation. It aims to promote the flow of bile.

■ Colestyramine

■ Ranitidine

■ Steroids

Steroids are also given to reduce inflammation