American Pediatric Surgical Nurses Association Inc.
Safety and Excellence in the Surgical Care of Children

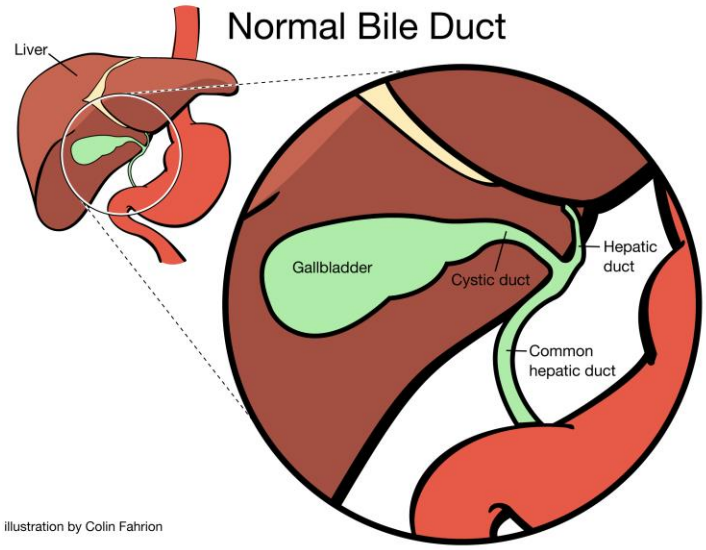
PATIENT AND FAMILY INFORMATION SHEET

Biliary Atresia

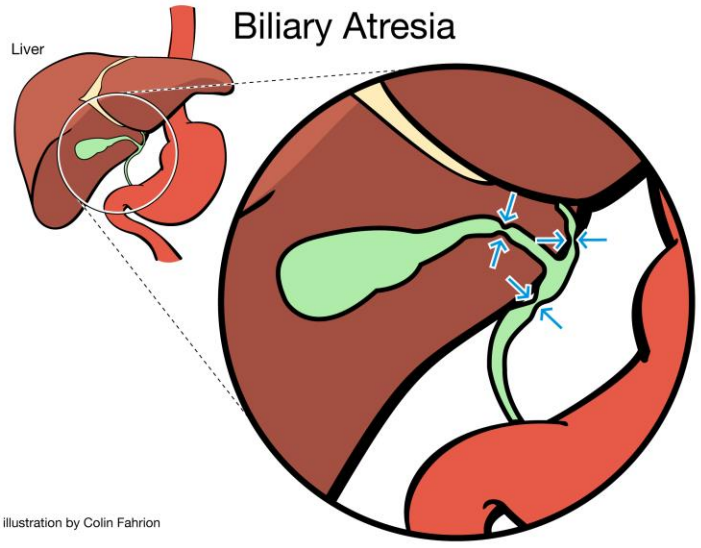
What is Biliary Atresia?

Biliary atresia (BA) is a rare liver condition in infants. It affects the biliary system. This system includes the liver, bile ducts, and gallbladder. The liver makes bile. Bile is a green fluid that helps digest the fat in food. Bile travels through bile ducts into the gallbladder, where it is stored, and then drains into the intestine. In biliary atresia, the bile ducts are not formed normally or may be missing. Because of this bile cannot drain out of the liver into the intestine. Bile that does not drain builds up in the liver. This causes damage to the liver.

Biliary atresia occurs during a baby's development but is not found until after a baby is born. While Biliary atresia may be caused by a combination of factors, no cause is known at this time. Babies found to have biliary atresia are often healthy full-term babies. They slowly develop a yellow color to the skin and the white part of the eyes. This is called jaundice. This happens when bilirubin from bile builds up in the liver, enters the bloodstream. Jaundice is one of the first signs of biliary atresia.



Normal bile ducts in the liver



Note areas of narrowed bile ducts and small gallbladder

Biliary Atresia

How do I know if my child has biliary atresia?

Here are some of the symptoms your child may have:

Jaundice or yellowing of the skin and eyes usually starts to appear after birth and gets worse as the infant over the first weeks of life. It rarely appears after the infant is 8 weeks of age or older. Jaundice causes urine to be very dark. Infants can have pale, white, or what medical experts call, acholic stools (poop). This means that the stool lacks color. Bile is what makes the stool yellow/green. When there is blockage of bile flow into the intestine, the stool becomes more pale and eventually white in color.

How is biliary atresia treated?

The treatment for biliary atresia is surgery.

What happens before surgery?

Here are common tests to find out if a child has biliary atresia.

Blood tests- These tests measure the bilirubin level and the different types of the bilirubin in the blood. These are direct and indirect bilirubin and conjugated and unconjugated bilirubin.
Liver function tests (AST, ALT, alkaline phosphatase, GGT) are also done. These and other blood tests are taken to help look for causes of jaundice other than biliary atresia.

A newer test called MMP-7 may be done. This can help your provider diagnose biliary atresia.

Ultrasound - This is a painless test that can scan the gallbladder, liver, and spleen for signs of biliary atresia. In biliary atresia the gallbladder may be small or missing.

HIDA scan - This test looks at the biliary system in the liver with a special dye. This can show if the biliary system is open allowing bile to flow into the intestine or if there appears to be a blockage of bile flow into the intestine.

Liver Biopsy - This is a procedure done in the operating room. A small piece of the liver is taken to be tested. A special doctor called a pathologist looks at the liver sample under a microscope to look for changes seen in biliary atresia.

Intraoperative cholangiogram - This is a test that is done in the operating room. A liver sample can be taken at same time. The cholangiogram is the best way to test for biliary atresia. The surgeon squirts a special dye into the gallbladder to see if it will flow into the biliary system and intestine. If the surgeon does not see dye flow into the intestine or if the gallbladder is missing or small the infant has biliary atresia.

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What happens during surgery?

Biliary atresia is treated with a surgery called a Kasai procedure or hepatoportoenterostomy. If the liver is too badly damaged from bile that has not drained well or if the infant has a normal intraoperative cholangiogram, the surgeon will end the procedure. The infant will go to the recovery room.

If the biliary system is not normal and blocks bile from draining into the intestine the infant has biliary atresia. If this is the case, the surgeon will do a Kasai hepatoportoenterostomy. The Kasai procedure will help the bile flow into to the intestines. The surgeon will use a piece of intestine to drain bile from the liver. This surgery will take several hours.

What happens after surgery?

- *When can I be with my child again?*

  As soon as your child wakes up after surgery, someone from the recovery room will call you so you can be with your child again.
- *Will my child have any pain?*
- Pain is a common side effect of surgery. Pain medication can be given to keep your baby comfortable and with help recovery. Sometimes the doctors will give spinal anesthesia in the operating room, similar to an epidural during childbirth. Before the surgery the doctors will talk to you about this option.

When can we go home?

- The time it takes to recover from surgery can vary. Your infant will probably be in the hospital for a week or longer.
- Some of the common discharge goals include eating well, tolerating medications and vitamins with good pain control.
- Specific vitamins, specialized formula and higher calories are often needed in the hospital and at home. A dietician will help make a nutrition plan for your infant. Some infants may need a feeding tube to help with feeding and growth.
- The Kasai procedure is the best way to help bile flow into the intestine. This may work for a long time or for a lifetime. However, it is also possible the Kasai may not work. In this case a liver transplant may be needed. For this reason, the liver team will help with your infant's care in the hospital and after discharge.

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How do I care for my child at home?

- Give all medications, vitamins and formula or breastmilk exactly as prescribed. Do not skip doses or stop giving them to your infant without talking to your surgical team and/or liver team.
- Follow all instructions given to you by the hospital providers.
- Attend all follow up appointments with the surgeon and the liver team.

When should I call the office?

If your child experiences any of the following, please call our office:

- Signs of a wound infection including redness around the area of the incision.
- Drainage (yellow or green) from the incision.
- Fever
- Stools/bowel movements that are white or grey
- Abdominal distension (full hard belly).
- Vomiting not related to another illness.
- Pain or discomfort that becomes worse and is not being helped by the pain medication

Your child will need to follow up with the surgeon. You will receive specific instructions for follow up when your child is discharged.

- Please don't hesitate to call our office if you have any problems or concerns.
- Surgical provider: _____
- Office Number: _____
- After hours number, if applicable: _____

Thank you for allowing us to care for your child.