Biliary Atresia

Why is the liver important?
The liver is the second largest organ in your body and is located under your rib cage on the right side. It weighs about three pounds and is shaped like a football that is flat on one side.

The liver performs many jobs in your body. It processes what you eat and drink into energy and nutrients your body can use. The liver also removes harmful substances from your blood.

What is biliary atresia?
Biliary atresia is a rare disease of the bile ducts that affects infants. Bile is a liquid that is made in the liver. It travels through the bile ducts to the small intestine, where it helps break down fats. In babies with biliary atresia, the bile ducts become blocked soon after birth. This blockage causes bile to stay in the liver, where it starts to destroy liver cells and cause cirrhosis (scarring of the liver).

What causes biliary atresia?
The cause of biliary atresia is unknown. It is not a genetic disease. Biliary atresia does not run in families. In some infants, biliary atresia is present at birth (congenital). About one in ten babies with biliary atresia have other congenital conditions.

What are the symptoms of biliary atresia?
Symptoms of biliary atresia may include:
- Jaundice (yellowing of the skin and whites of the eyes)
- Dark urine
- Pale stools
- Swollen abdomen

Symptoms of biliary atresia usually appear between two and six weeks after birth.

What are the complications of biliary atresia?
Complications of biliary atresia may include:
- Ascites (fluid in the abdomen)
- Cirrhosis (scarring of the liver)
- Enlarged spleen
How is biliary atresia diagnosed?
Biliary atresia is diagnosed using different tests which may include:

<table>
<thead>
<tr>
<th>Test</th>
<th>Checks for:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blood test</td>
<td>Liver abnormalities</td>
</tr>
<tr>
<td>Ultrasound</td>
<td>Enlarged liver</td>
</tr>
<tr>
<td></td>
<td>Anything blocking the bile ducts</td>
</tr>
<tr>
<td>Liver biopsy (a small piece of liver tissue is removed and studied in the lab)</td>
<td>Biliary atresia</td>
</tr>
</tbody>
</table>

How is biliary atresia treated?
The Kasai operation is usually the first treatment option for babies with biliary atresia. During the Kasai operation, damaged bile ducts are removed and a new duct from the liver to intestine is created to improve bile flow.

The Kasai procedure is most successful when it is performed before a baby reaches two to three months of age. If successful, jaundice and other symptoms usually disappear after several weeks. Doctors also may recommend a special diet and vitamin supplements.

The other treatment option is a liver transplant. In children with biliary atresia, a liver transplant generally is not done unless the Kasai procedure has been tried. About 75% of children with biliary atresia may eventually need a liver transplant.