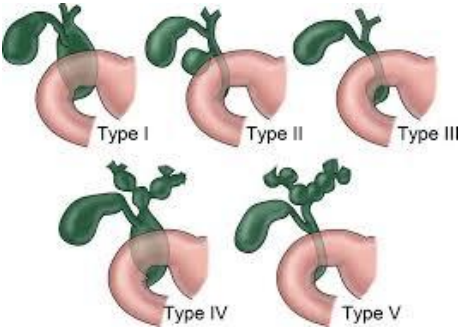
Indian Association of Pediatric Surgeons

Patient Information Sheet

CHOLEDOCHAL CYST



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What is choledochal cyst ?

Choledochal cyst is a condition in which there is single or multiple dilatations in the bile drainage system (ducts) which channels the flow of bile from liver into small intestine. These dilatations may be intrahepatic (within the liver), extrahepatic (outside the liver) or both intra and extra hepatic. Depending on these variables, the disease is classified into various types. And depending on the type, the management also differs.

What causes this problem and how common is it ?

It is thought to be due to localised weakness in the wall of the bile ducts which causes these dilatations. The weakness is speculated to be due to reflux of pancreatic enzymes into biliary system which normally does not happen. In some cases of abnormal pancreatic development, where the pancreatic and biliary systems are not separated adequately, reflux of pancreatic enzymes leads to this condition. It is seen in about 1 in every 1,00,000 live births and 4 times more common in a female child.

What are the symptoms ?

Children with choledochal cysts may have no major symptoms. The cyst may be discovered at an ultrasound scan of the child done usually for evaluation of pain or an antenatal ultrasound scan of the mother. Classic symptoms include pain, swelling in the upper abdomen, fever and jaundice.

When to see your doctor ?

Children with jaundice outside the normal period (up to 21 days after birth) will need to be evaluated by your doctor. Along with jaundice, any other symptoms like pain or swelling in the upper abdomen, fever or pale stools then this warrants detailed evaluation.

How is it diagnosed ?

A few blood tests and radiological tests are required. Blood tests include complete hemogram, liver function tests,

serum amylase and lipase. Radiological tests include an ultrasonography of abdomen followed by a CT scan or Magnetic Resonance Cholangio-pancreatography (MRCP) which will delineate the biliary drainage system and identify the site and extent of the disease. Some cases may require Endoscopic Retrograde Cholangio-pancreatography (ERCP) to define the presence of abnormal union of pancreatic and biliary systems.

What are the treatments available ?

Available treatment options are surgical excision of the abnormal dilated extrahepatic biliary tree and establishing the bile flow into intestine using a segment of small intestine. In cases where there only intrahepatic part of biliary tree is dilated, in such cases liver transplantation may be necessary.

Are there any alternatives to surgery ?

In some cases, ERCP and placement of stent may be needed to tide over the crisis. But these children will also require complete surgical excision of the cyst, subsequently.

What does the operation involve ?

The operation is a major procedure, which involves complete removal of dilated or cystic extra-hepatic biliary system including the gall bladder up to the intestines. Then small portion of small intestine is used as a conduit to re-establish the connectivity between liver and intestines. A small piece of liver may be removed for biopsy at the same time.

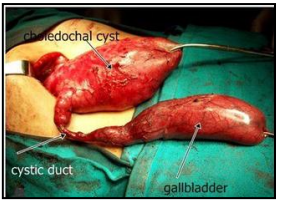
What are the possible complications / what happens after the operation ?

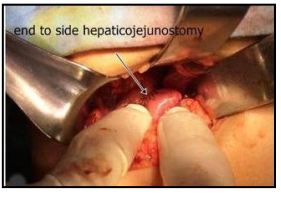
Post procedure, the child will be nursed in a Pediatric Intensive Care Unit (PICU). There will be tubes going in and coming out of the child to help her/him recover. Depending on the progress towards recovery, these tubes will be removed and shifted out to ward. Gradually oral intake will be started and escalated. Complications include bile leak and cholangitis (infection of the biliary system), which may require the tubes

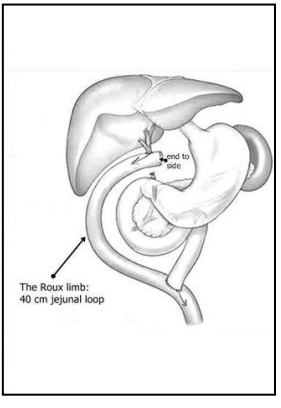
and PICU stay for a longer time.

What is the outlook or future of these children ?

Children with extra-hepatic choledochal cysts who are diagnosed and treated surgically promptly, fare much better and are mostly cured for their life. However, the ones with intrahepatic involvement need monitoring and may need further surgeries. If the liver is excessively damaged, liver transplant may be required.







Surgery for choledochal cyst