



Children's Liver
Disease Foundation

fighting childhood
liver disease

Choledochal Cyst

**(commonly known as a Choledochal Anomaly or
Choledochal Malformation)**

A Guide



An explanation of what choledochal
cysts are, their symptoms, diagnosis
and treatment

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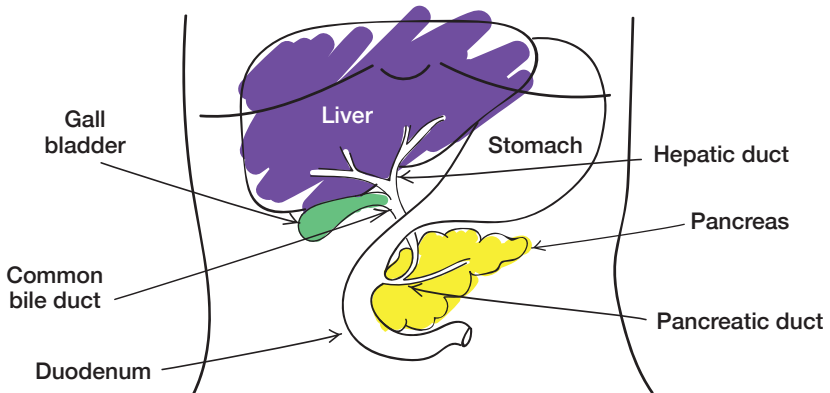
Introduction

This information should be read in conjunction with:

- An Introduction to Liver Disease

Before explaining what a choledochal cyst is it is helpful to know a little about what the bile ducts are, where they are and what they do. See diagram below.

Normal bile duct anatomy



Our liver produces a secretion known as “bile” which contains useful products. Bile has many functions which help in food absorption from the gut. Bile also gets rid of waste products that are not needed by the body. Bile ducts are tubes which carry bile from the liver to the gall bladder and then to the gut when required.

What is a choledochal cyst?

The term choledochal cyst can be broken down into the following:

- chole ~ relating to bile
- dochal ~ containing or receiving
- cyst ~ fluid collection

A choledochal cyst is a swelling / widening or dilatation of the bile ducts.

The condition usually affects the part of the bile ducts outside the liver (the common bile duct and the hepatic ducts) but sometimes it also affects those inside the liver (the intrahepatic ducts).

We don't know the exact cause of a choledochal cyst but there are a few theories.

One explanation is that it happens due to the bile duct not forming properly when the baby is developing in the womb in early pregnancy. This concept is supported by the fact that in some cases, the cyst is first seen during a routine ante-natal ultrasound scan of the baby in early pregnancy.

Another theory is that the junction of the bile duct with the pancreatic duct is not normal and forms an abnormally long 'common channel'. Currently there are arguments in support of both of these.

Is the condition rare?

Yes, it is rare. It affects about 1 in 50,000 children in the West. Choledochal cysts are three times as common in girls as boys and rarely run in families.

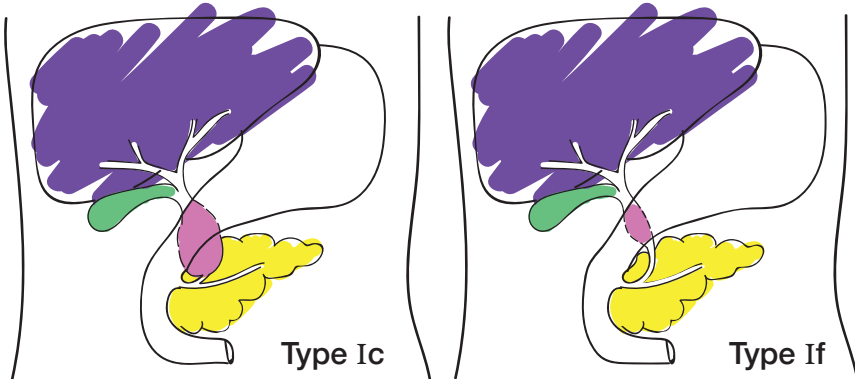
What are the different types of choledochal cyst?

There are several types of choledochal cyst. Their positions and sizes may differ.

Type I cysts are the most common. The ducts inside the liver look normal but the common bile duct may be swollen up like a small balloon (type Ic) or a fat cigar (type If).

Types of choledochal cyst

Type 1:

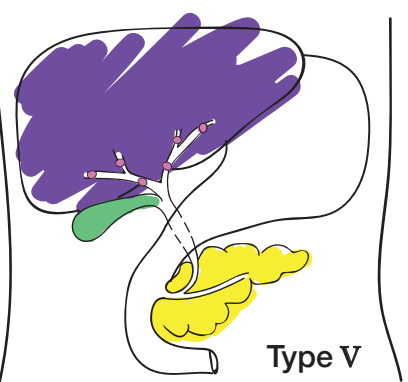
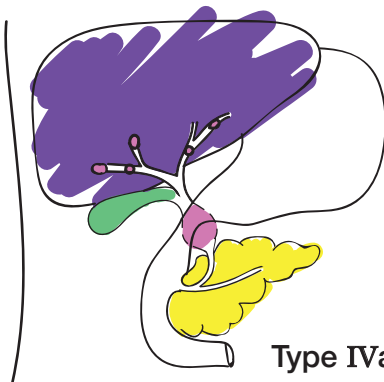
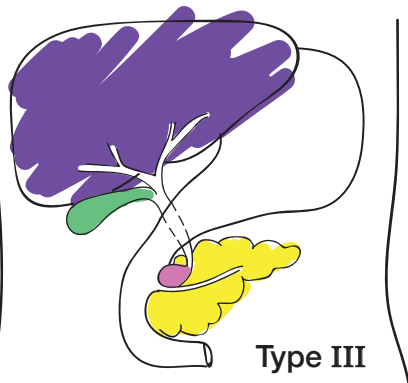
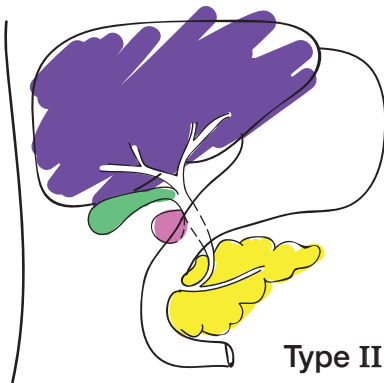


In these types of choledochal cyst, the lower end of the common bile duct often joins up with the pancreatic duct. This is not normal and forms an abnormally long 'common channel'.

This causes the pancreatic juice and the bile to mix together very easily before they enter the duodenum. This early mixing can result in weakening of the bile duct wall leading to ballooning. In addition it can lead to complications such as pancreatitis (inflammation of the pancreas) or cholangitis (inflammation of the biliary system).

Other types of choledochal cysts are less common.

Other types of choledochal cysts:



Apart from these types of choledochal cyst there is another known as the “forme fruste” type of choledochal anomaly. Although this involves an abnormality of the junction between the bile duct and pancreatic duct there is no widening (dilation) of the ducts. This kind of abnormality can also give rise to serious complications.

What are the symptoms and signs of a choledochal cyst?

Some of these cysts are found before birth on the antenatal scan. Sometimes, a choledochal cyst is found when a child is having an abdominal ultrasound scan for an unconnected reason like pain in the abdomen. In some cases the child may have symptoms such as jaundice, a mass in the abdomen, pancreatitis or cholangitis. Most choledochal cysts are detected in childhood. A child with a choledochal cyst may have some or possibly all of the following symptoms:

- Jaundice
This is due to blockage of bile drainage. This might be prolonged jaundice (lasting more than two weeks) in a newborn baby or intermittent jaundice (jaundice which comes and goes) in an older infant or child. Usually, the jaundice is associated with pale coloured stools and dark urine.
- Intermittent abdominal pain (abdominal pain which comes and goes) which can be severe.
- Cholangitis (an infection of the bile duct causing fever, jaundice, and sometimes shaking attacks called rigors).
- Peritonitis (a severe inflammation within the abdomen) if the cyst bursts or leaks.
- A lump or swelling in the belly.
- Pancreatitis.
- An inflammation (irritation, swelling and damage) in the pancreas.

How is a diagnosis made?

The diagnosis is generally made using a combination of the following:

- History
- A physical exam of the child
- Tests including blood tests and an ultrasound scan

A choledochal cyst is most often suspected if an abdominal ultrasound scan shows a dilated bile duct in a baby or child with jaundice or if the child has abdominal pain. Once suspected some other tests are required to get more information. For example, is there any blockage in the duct by a stone or is there an abnormal junction of the bile duct with pancreatic duct (a long common channel)?

These extra tests are carried out to get more information about the type of choledochal cyst present which helps the surgeon to get a 'road map' to plan the operation.

These tests may include:

- A more detailed ultrasound scan to look not only at the bile ducts but also to look at the blood vessels going to the liver which run alongside the bile ducts.
- MRCP (Magnetic Resonance Cholangio-Pancreatography). This type of detailed scan is carried out in a large machine. It does not require X-rays but the patient does need to lie flat and still and so babies and small children often require a general anaesthetic. The bile ducts and the pancreatic duct, are shown better on an MRI scan.
- ERCP (Endoscopic Retrograde Cholangio-Pancreatography). This is a specialised procedure which is not often needed. It is done under a general anaesthetic. In this procedure a flexible tube with a small camera on the end (an endoscope)

is passed through the mouth down into the intestine. A tiny plastic tube is then passed through this endoscope into the opening of the bile duct and dye is injected to outline the choledochal cyst and the pancreatic duct. This helps to give information about the junction of bile duct with pancreatic duct (long common channel) and if there is any blockage in the bile duct due to stones etc.

This investigation can sometimes stir up inflammation in the pancreas (pancreatitis) causing temporary abdominal pain and vomiting afterwards. It is rarely done as it is an invasive investigation.

- Rarely a HIDA (liver scan) becomes necessary, if there is only a single cyst in the liver and the MRCP cannot confirm it to be a choledochal cyst/anomaly.
- The exact nature of the choledochal cyst sometimes only becomes clear at the time of surgery when dye is injected into the bile duct system (an operative cholangiogram).

What is the treatment for a choledochal cyst?

The treatment of choledochal cysts is surgery. This should be done in a centre which has experience of looking after children who have choledochal cysts and surgeons experienced with this kind of operation. The operation can be performed as key hole surgery (using a small opening through the skin) or as an open procedure (a larger opening in the skin). Both techniques achieve good results.

In the common variety, the cyst is completely removed. This means removing most of the bile ducts (tubes carrying bile) outside the liver together with the gallbladder. The hepatic ducts (tubes from the liver) coming out of the liver are then attached to a loop of the child's bowel so that the bile can drain into the bowel.

This loop of bowel is a specially created loop as shown in the picture on the next page. Removing the gallbladder has no important long-term effects that we know of. A liver biopsy may also be taken at the time of surgery to see if the liver is healthy or damaged.

In babies with a cyst discovered by ultrasound scan but who have no symptoms, there is debate about the best time to perform surgery. Children's surgeons who specialise in liver and bile duct surgery often recommend early surgery, generally at about six months of age.

Normal Anatomy

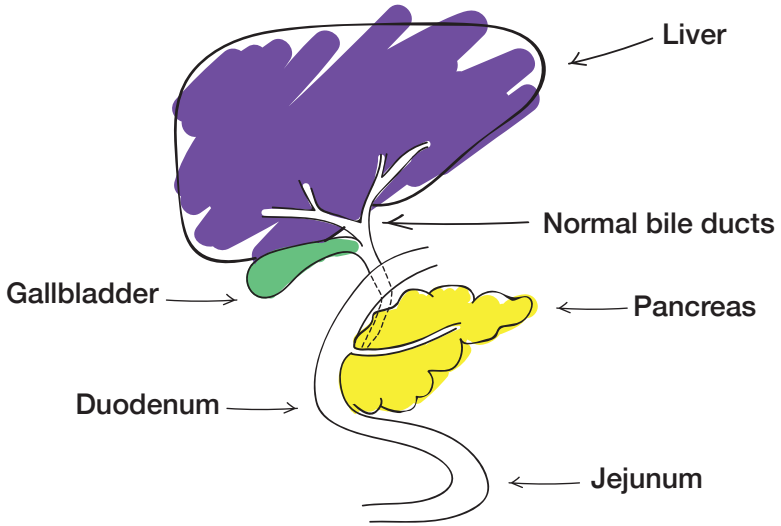
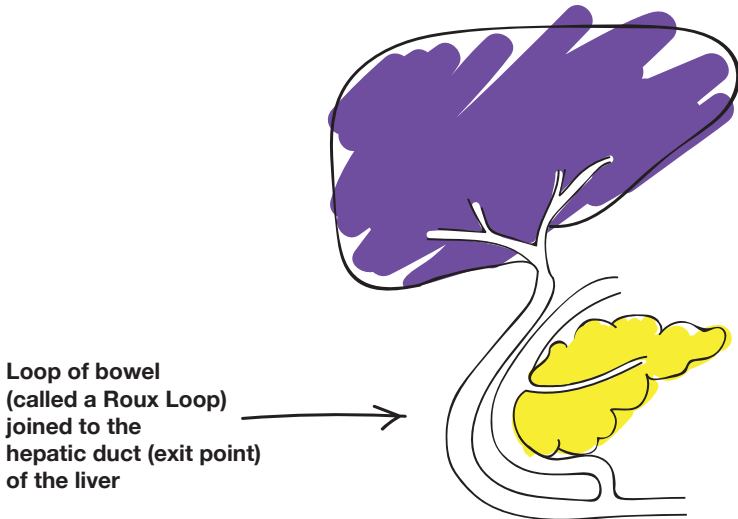


Diagram of Surgery



Are there any complications of surgery?

There are risks as with any other surgery but these are not common. For example, there is a small risk of bleeding, and infection. There is also a risk of leakage and narrowing where the bile ducts are joined to the bowel.

The operation is generally very successful. Most of the time, children recover rapidly and are able to go home within one week after surgery. They are then seen in the outpatient clinic from time to time when a blood test (to check liver function) and an ultrasound scan may be performed.

What about the long term outlook after surgery?

The majority (90% or more) of children remain well and grow up to be healthy adults.

A small number (10% or less) may get problems such as cholangitis. Cholangitis is an infection of the bile ducts within the liver. If it occurs it generally happens in the first few months after surgery. If a child becomes unwell after surgery e.g. has a temperature, it is important to seek medical advice.

Some children require further surgery, for example, because the joint between the bile ducts and the bowel has narrowed down. In a very small number of patients, there is a small, long term risk of other complications including cancer of the bile ducts in adult life. This is extremely rare if the cyst has been removed properly (not leaving behind a portion of cyst or delaying the surgery for long time).

Rarely, the liver is very scarred (cirrhosis) when a choledochal cyst is first diagnosed. In such cases, medical monitoring is necessary after the cyst has been removed because further problems can develop and these need to be recognised early.

What happens if a choledochal cyst is left untreated?

If a choledochal cyst is left untreated, it may lead to major complications.

These can include:

- Liver damage (fibrosis and cirrhosis)
- Infection (cholangitis)
- Bile duct damage
- The cyst may burst or leak
- Problems with food absorption and growth
- Bleeding
- Stone formation (gallstones) in the cyst
- Pancreatitis (inflammation in the pancreas) and damage to the pancreas and its duct
- Risk of cancer developing within the bile tubes or in the gallbladder

Your specialist liver team will discuss the risks to your child if the cyst is left untreated.

CLDF produces a wide variety of information resources for children and young people up to the age of 25 with liver disease, their families and the healthcare professionals who look after them. This information can be downloaded or ordered from CLDF's website childliverdisease.org. For further enquiries regarding CLDF's information please contact us by email at info@childliverdisease.org or call **0121 212 3839**.

Thanks

The booklet has been written, edited and reviewed with the help of staff at each of the specialist paediatric liver centres: Birmingham Children's Hospital, King's College Hospital and Leeds Children's Hospital. Thank you to all of the staff involved who have made the production of this leaflet possible.

Disclaimer

This leaflet provides general information but does not replace medical advice. It is important to contact your/your child's medical team if you have any worries or concerns.

Feedback and Information Sources

Information within this leaflet has been produced with input from the three specialist paediatric liver centres in the UK. To provide feedback on this leaflet, for more information on the content of this leaflet including references and how it was developed contact Children's Liver Disease Foundation:
info@childliverdisease.org

This leaflet has been reviewed in December 2020. It is due to be reviewed by December 2023.

What is Children's Liver Disease Foundation (CLDF)?

CLDF is the UK's leading organisation dedicated to fighting all childhood liver diseases. CLDF provides information and support services to young people up to the age of 25 with liver conditions and their families, and is a voice for everyone affected.

Are you a young person up to the age of 25 with a liver condition or a family member? CLDF's Families and Young People's teams are here for you, whether you want to talk about issues affecting you, meet and share with others, or just belong to a group which cares, knows what it's like and is fighting to make a difference. You are not alone.

If you are a parent/carer or family member then get in touch with CLDF's Families Team:

Phone: 0121 212 6023 **Email:** families@childliverdisease.org

If you are a young person and want to find out more about CLDF's services you can contact CLDF's Young People's Team:

Phone: 0121 212 6024 **Email:** youngpeople@childliverdisease.org

CLDF have a social media platform called HIVE for young people with a liver disease/transplant to make new friends, connect and share stories.

childliverdisease.org/young-people/hive

Would you like to help us support the fight against childhood liver disease?

CLDF's work relies on voluntary donations and fundraising. Please help us to continue to support children, young people and families now and in the future. To find out more about fundraising and how you can join the fight against childhood liver disease you can visit www.childliverdisease.org/get-involved. Alternatively, you can email the Fundraising Team at fundraising@childliverdisease.org or call them on **0121 212 6022**.

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