Choledochal Cyst

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

A choledochal cyst (CC) is a rare abnormality of the bile ducts. The bile ducts and its branches (hepatic ducts) are structures through which bile travels from the liver to the intestine (Figure 1). Bile is a body fluid made by the liver and stored in the gallbladder. In response to a meal, the gallbladder releases bile released into the small intestine to help in breaking down (digestion) of foods.

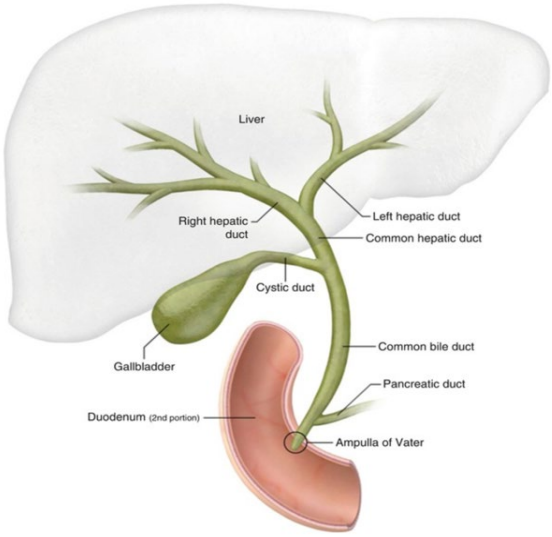

Figure 1: Bile duct anatomy
Image courtesy of Radiology Key.com

Normally the bile ducts and its branches are thin channels (a few millimeters in size). Very small bile ducts start in the liver and join together within the liver to create larger branches, and then right and left hepatic ducts exit the liver and join to become the main (common) hepatic duct. In choledochal cysts, there is enlargement or expansion of parts of the bile duct system.

The exact cause (etiology) of CC is unknown. Some experts think that infants are born with this condition. In 30 to 70% of CCs, there is abnormal anatomy where the bile duct enters the small intestine relative to where the pancreas enters (this is called a “common channel”). Experts think that when this happens, juice from the pancreas can go up the bile duct causing the bile duct to get damaged and expand.

Choledochal cyst can occur at any age, but 80% are diagnosed in infancy. The incidence of CC in Western countries is 1 in 100,000 – 150,000 individuals, while in Japan it is 1 in 1000 - 13,000 individuals. The problem is three to four times more common in females.

Pediatric surgeons classify CCs based on what part of the biliary tree is dilated. How the CC is treated is also based on the location of the abnormality. In some cases, the expansion of the bile duct is only localized to the part outside the liver. These cysts can be removed completely by surgery. Some types have the abnormality within the small intestine. These cysts may be able to be removed using endoscopy. In some cases, the ducts are abnormal everywhere—inside and outside the liver (Figure 2).

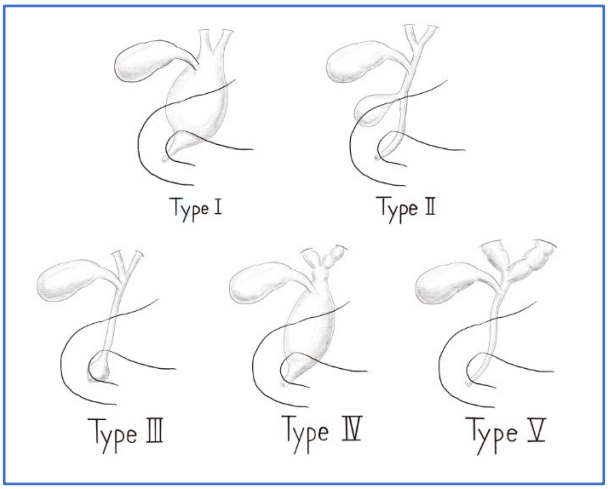

Figure 2: Different types of choledochal cysts
Illustration courtesy of Mark Mazziotti, MD

A surgeon will recommend removal of the choledochal cyst to prevent future problems. Cancer can grow in the cyst over time. The patient may develop infection of the bile ducts (*cholangitis*) or inflammation of the pancreas (*pancreatitis*). In addition, stones and sludge can form in the cyst and cause abdominal pain or inflammation of the gallbladder (cholecystitis).

Signs and Symptoms - "What symptoms will my child have?"

Early signs: A choledochal cyst is usually diagnosed in during in infancy or childhood. It can also be diagnosed as an adult. Abdominal pain, jaundice (yellow tinge to the skin and white of eyes), or a bump in the upper part of the belly may be noted. The patient may also have no symptoms at all.

Later signs/symptoms: When stones and sludge form in the cyst and/or gallbladder, inflammation of these structures can cause more severe pain or fever. Rarely, choledochal cysts can rupture resulting in a belly pain and swelling, as well as fever.

Infection of the bile ducts (cholangitis) and inflammation of the pancreas (pancreatitis) can occur. Studies suggest that the risk of cancer after 10 years of follow-up is 10-15% if the cyst is not resected.

Conditions that mimic choledochal cyst: gallstones, sclerosing cholangitis (a progressive scarring liver disease), pancreatic pseudocyst (cyst of the pancreas usually resulting form inflammation or trauma), biliary hamartoma (a growth of the bile duct).

Caroli's disease (Type 5 CC): The risk of cancer is less than 7%. Surgery is sometimes needed for infection of the bile ducts or liver complications.

Diagnosis - "What tests are done to find out what my child has?"

- **Physical exam** by a doctor to evaluate for jaundice, abdominal tenderness and abdominal masses.
- **Ultrasound**: Any gallbladder or bile duct abnormality is usually first assessed with ultrasound. Using sound waves, the bile duct and its branches are imaged.
- ***MRCP****: (Magnetic Resonance CholangioPancreatography)*. This study is a type of MRI scan where magnets are used to get pictures of the organs in the belly, with detailed images of the bile ducts. No radiation is used, unlike a CT scan. Small children may need sedation (medicine to make them sleepy) or anesthesia to remain still during this painless study.

Treatment - "What will be done to make my child better?"

The treatment of CC depends on which segment of the bile duct is affected.

- ***Endoscopy*:** In some less common types of CC, where the abnormality is localized only to the segment of the bile duct within the small intestine, endoscopy may be used to remove the abnormality. Endoscopy means placing a flexible scope into the mouth of the child and navigating the end of the scope into the small intestine where the bile duct empties. In select cases, this approach can take care of the problem.
- **Surgery:** In the most common types of CC, where the abnormalities are in the bile ducts outside of the liver, surgery is required. Depending on the situation, the surgery may be done with a large incision (laparotomy, which is called open surgery) or multiple small incisions with a scope (laparoscopy, minimally invasive surgery).

The abnormal dilated part of the bile duct must be completely removed. In most cases, the gallbladder may need to be removed as well. If the main bile duct is removed, the flow of the bile from the liver to the intestine must be restored. This can be done by creating a connection between the remaining small amount of the common bile duct and the intestines (either the duodenum or jejunum).

If there are also abnormalities in the bile ducts within the liver, the surgery becomes more complicated and may require removal of parts of the liver. This type of CC can be hard to manage.

Benefits: Choledochal cysts can cause blockage of the flow of bile and infection of the bile ducts. If CCs are not removed, there is a risk that cancer can grow within the cyst. The risk of cancer increases with age if the CC is not removed.

Risks: Surgery has low risks, but includes bleeding, postoperative infection, small bowel obstruction from adhesions, bile leak, and the risks of anesthesia. All surgery has a risk of damage to internal structures and further operation may be needed to fix a problem.

Preoperative Care: The child will have nothing to eat for eight hours prior to operation to decreases the risk of stomach contents spilling over into the lungs (aspiration) as the child goes to sleep. The surgeon will check the child's blood count and blood type prior to surgery. Blood will be available for transfusion in case of intraoperative bleeding. In most cases, the child can come to the hospital on the morning of surgery and does not need to be admitted prior. Most of the time, a thorough bath or shower is recommended the day prior or the day of the surgery to decrease bacteria in the skin and risk of infection.

Informed consent: A consent form is a legal document that states the tests, treatments or procedures that your child may need and the doctor or practitioner that will perform them. Before surgery, your doctor should tell you what the operation is, the goal of the surgery and other possible treatment options that are available. Your doctor should explain the risks and benefits of the surgery. You give your permission when you sign the consent form. You can

have someone sign this form for you if you are not able to sign it. You have the right to understand your child's medical care in words you know. Before you sign the consent form, make sure all of your questions are answered. It is important to know that during surgery, there are things that can happen that your doctor may have not predicted before going in. He or she will explain these to you after the surgery.

Postoperative Care:

- **Activity:** Typically, the child is encouraged to walk around as soon as possible. To decrease lung complications, breathing exercises such as blowing bubbles, pinwheels or deep breathing is encouraged.
- **Diet:** Intestines are usually slow after surgery. Most children are not allowed to eat after surgery until there is return of intestinal function as seen by passing gas or having stool. Often, a tube is placed through the nose, with the tip in the stomach to suction stomach contents while the intestines are not working yet. This avoids having the child vomit and retch, which is uncomfortable especially with a new incision.
- **Medicines:** Your child may need anti-nausea medicine in addition to pain medicines.

Home Care - "What do I need to do once my child goes home?"

Diet: Your child will be discharged on their usual diet.

Activity: Your child should avoid strenuous activity and heavy lifting for the first 1-2 weeks after laparoscopic surgery, 4-6 weeks after open surgery.

Wound care: Surgical incisions should be kept clean and dry for a few days after surgery. The stitches used in children are absorbable and do not require removal. Your surgeon will give you specific guidance regarding wound care, including when your child can shower or bathe.

Medicines: Medicines for pain such as acetaminophen (Tylenol) or ibuprofen (Motrin or Advil) or something stronger like a narcotic may be needed to help with pain for a few days after surgery. Stool softeners and laxatives are needed to help regular stooling after surgery, especially if narcotics are still needed for pain.

What to call the doctor for: Call your doctor for worsening belly pain, fever, vomiting, diarrhea, jaundice, if the wounds are red or draining fluid.

Follow-up care: Your child should follow up with his or her surgeon 2-3 weeks after surgery to ensure proper post-operative healing.

Long Term Outcomes - “Are there future conditions to worry about?”

Prognosis is excellent after surgery. Medical studies report excellent survival rates of over 90% and survival without need for further intervention or surgery (event-free survival). Regardless of type, appropriate surgery has good results.

Although cancer is rare, excision of a choledochal cyst does not completely reduce the risk to zero, so long term follow-up is indicated. The physician may order periodic blood tests or ultrasound studies.

Unique subtypes: Type 4 choledochal cysts and cysts diagnosed in infancy require more detailed follow-up by a pediatric gastroenterologist, as they have a higher incidence of complications of the liver and complications after surgery.

The overall incidence of complications is approximately 5-9%. Bile leak from the surgery (where the remaining bile duct was sutured to the intestine) is noted to be 0-5% and cholangitis (an infection of the bile ducts and liver) is 2-3%. Pancreatitis (2.5%) can occur and is inflammation of the pancreas, which shares anatomy with the choledochal cyst. Small bowel obstruction (0-5%) is uncommon but more common with open (one large incision) compared to laparoscopic (several small incisions).

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Author: Joanne E. Baerg, MD
Editors: Patricia Lange, MD; Marjorie J. Arca, MD; Mark V. Mazziotti, MD, MEd