Choledochal Cyst

Patient Information Series – What you should know, what you should ask.

What is choledochal cyst?

Choledochal cysts (CC) are rare congenital dilations of the biliary tract, which include the organs and ducts that produce and store bile, a fluid made by the liver to help digest fats and release it to the intestine. They can be found along the biliary system, either outside the liver or within the liver.

How does a choledochal cyst happen?

Choledochal cysts are rare with a reported incidence of 1 in 100,00-150,000 births in the western population, with a higher incidence in Asian populations. It is thought that choledochal cysts begin at an abnormal bile duct junction into the pancreas. Pancreases perform the important function of producing enzymes that are vital to digestion of food we eat. This abnormality of the location of the choledochal cysts allows for mixing of the pancreatic enzymes which causes local destruction and dilation of the bile duct along its normal pathway.

Should I have more tests done?

Choledochal cysts are most isolated abnormalities. However, they can be associated with other fetal problems and so a detailed ultrasound of your baby, including a fetal echocardiogram, a test like ultrasound that uses sound waves that “echo” off the structures of the fetus's heart, should be performed.

What are the things to watch for during the pregnancy?

Babies with choledochal cysts should have additional ultrasound examinations throughout the pregnancy to assess change in size of the cyst.

What does it mean for my baby after it is born?

After birth babies may develop a yellow colour to the eyes and skin, pain in upper-right belly, pale or clay-colored stools, nausea, vomiting and fever and feeding problems. These symptoms can be a sign that the baby has a problem. If your baby has any of these symptoms, you should promptly call your pediatrician. If left untreated, choledochal cysts may cause infections of the bile duct and gallbladder. Occasionally, they rupture. Surgical removal is generally required. Surgery is typically performed before the baby is 6 months old.

The good news about choledochal cysts is that knowing about the cyst during pregnancy allows for early monitoring and appropriate follow-up after your baby is born. Earlier diagnosis that leads to surgical repair at early stages results in fewer complications and a better prognosis.

Will it happen again?

Choledochal cysts are rare, and most parents will not have a second child with a choledochal cyst. There are rare reports of certain types of choledochal cysts that may be genetically linked. Consultation with a Maternal Fetal Medicine specialist and/or a geneticist may be helpful. Early diagnosis and surgical removal of the cyst with Roux-en-Y hepaticojejunostomy (before the age

of 1 month) dramatically lowers the risk of developing malignancy and achieves the best prognosis.

What other questions should I ask?

- Are there any other defects found?
- How often will I have ultrasound examinations done?
- Where will the baby receive the best care after it is born?
- Will I be able to deliver at my scheduled hospital?
- Is surgery available after I deliver in my area?
- Can I meet in advance the team of doctors that will be looking after my baby when it is born?

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