Intestinal Duplication Cyst

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

What is Intestinal Duplication Cyst?

Intestinal duplication cyst (IDC) is a rare congenital anomaly that occurs when there is an abnormal development of intestinal tissue that is attached to or adjacent to the intestines. It can occur anywhere from the esophagus to the rectum. They are characterized by an epithelial lining representing a part of the gut, a well-defined layer of smooth muscle, and a close approximation with some part of the gastrointestinal tract, such as sharing a common wall. The intestinal duplication cyst may be lined with normal intestinal mucosa but it may also contain tissues from other portions of the body, such as gastric or pancreatic mucosa, which can lead to complications like ulceration and bleeding.

How does an Intestinal Duplication Cyst happen?

Intestinal duplication cyst is rare, and the exact incidence of intestinal duplication cyst is unknown. It is estimated to occur in 1 in 4,500 births. Most are found within the first two years of life. It is not clear how it happens. Typically, it is an isolated finding, but it can be associated with spinal, lung, and heart issues. Intestinal duplication cyst is also seen twice as frequently in males than females.

Should I have more tests done?

Intestinal duplication cyst can be found on prenatal ultrasound, known for a characteristic "gut signature" or "double wall" sign. Once diagnosed, a test called amniocentesis (where a needle is used to take some of the fluid from the womb) can be used to look for genetic abnormalities with chromosomes. A fetal echocardiography, an ultrasound of the heart of the baby, can be done with a detailed ultrasound to assess for other associated abnormalities. Fetal MRI may also be useful for ruling out other common intra-abdominal lesions.

What are the things to watch for during the pregnancy?

Babies with intestinal duplication cysts are at risk of some issues during the pregnancy. Serial ultrasounds are recommended to assess fetal growth, amniotic fluid levels (fluid in the womb), or overall fetal condition. These babies may develop extra amniotic fluid around baby which is called polyhydramnios, fluid in the baby's abdomen secondary to cyst rupture, or bleeding within the cyst from growth of the lesion.

The size of the lesion and the presence of blockage of the intestines will determine if you should have vaginal delivery or a caesarean section. If the cyst is large enough, there is risk of the cyst rupturing during vaginal delivery or having difficulty removing the baby from the vagina safely. There is typically no reason for these babies to be delivered early based solely on the presence of the intestinal duplication cyst.

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

If identified prior to birth, the outcome for baby is typically excellent after surgical resection of the lesion (process that cuts out the lesion), especially if the lesion is isolated as resection, is curative.

These babies typically will need to be born at a hospital that has appropriate neonatology and pediatric surgeon teams that are able to facilitate early intervention with surgery.

If not identified prior to birth, most patients will experience symptoms that would occur within 1-2 years of life like pain, bleeding, respiratory complaints, and/or a palpable mass. These symptoms will lead to additional imaging and surgical intervention.

Babies who also have problems within the chromosomes or have other associated pulmonary, spinal, or heart defects may have even more issues after being born. These may lead to additional intervention with medications, surgery, or prolonged stay in the New Born Intensive Care Unit (NICU) depending on the type of problem that the baby has.

There are typically no long-term complications with intestinal duplication cysts. Nutrition and growth are not affected.

Will it happen again?

With complete surgical resection, intestinal duplication cysts rarely recur. The risk of intestinal duplication cysts happening again in another pregnancy depends on if there is a genetic reason associated with the lesion. Consultation with a specialist may be helpful to determine that risk. Further study is required to ascertain best practice for the management of asymptomatic IDCs, as the rare nature of this congenital malformation makes such studies challenging

What other questions should I ask?

- Are there any other defects found?
- Is the lesion associated with any genetic abnormalities?
- How often will I have ultrasound examinations done?
- What will be the best method of delivery based on the size of the lesion in my baby?
- Where will the baby receive the best care after it is born?
- Will I be able to deliver at my scheduled hospital with this lesion present in my baby?
- 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?
- Can you give an indication of the duration of the hospital stay after surgery?

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