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duodenal atresia

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What is duodenal atresia?

During normal fetal development the duodenum — the upper part of the small intestine — remains unobstructed, allowing contents from the stomach to flow freely through the baby's digestive tract (Figure 1). Duodenal atresia (DWAH-de-nal ah-TREE-zha) is a condition that occurs when a portion of the duodenum doesn't form. This condition results in a blockage (atresia) that stops food or fluid from leaving the baby's stomach (Figure 2).

Duodenal atresia can be an isolated condition (which means no other birth defect or condition occurs with it), but is also more common in infants with Down syndrome (trisomy 21). About one in three babies born with duodenal atresia has Down syndrome.

What causes duodenal atresia?

Duodenal atresia is a congenital condition, which means it develops before birth. Exactly what causes the condition is unknown, although genetics may play a role in rare cases.

How is duodenal atresia diagnosed?

Duodenal atresia is diagnosed by ultrasound, but not usually at the routine 20-week screening ultrasound. That's because signs of the condition tend not to be visible by ultrasound until later in the pregnancy.

The ultrasound that leads to the diagnosis usually occurs through one of the following two pathways:

1. If genetic screening or other diagnostic testing determines the baby is at an increased risk for Down syndrome, an ultrasound will be performed to screen for duodenal atresia.
2. In pregnancies with no increased risk for Down syndrome, an ultrasound will be ordered if the uterus measures large for dates in the third trimester. An enlarged uterus is sometimes caused by excessive amounts of amniotic fluid, a condition known as polyhydramnios. Extra amniotic fluid can accumulate when the unborn baby is having difficulty swallowing — a difficulty that can result from the presence of a duodenal atresia.

The diagnosis is further established if the ultrasound image shows the classic sign of duodenal atresia: a “double bubble” in the baby’s abdomen. (Figure 3.) One “bubble” is the fluid-filled stomach; the other is the fluid-filled duodenum. Those “bubbles” mean that, due to the atresia (blockage), there is fluid in the stomach and in part of the duodenum, but not further down the intestinal tract.

How is duodenal atresia managed before birth?

The prenatal management of babies with duodenal atresia starts with acquiring as much information about the condition as early as possible. Tests will also be done to help determine if Down syndrome or a heart-related birth defect are present. To gather all of that information, we use several different techniques, including high-resolution fetal ultrasonography, fetal echocardiography and amniocentesis.

When will my baby have an operation?

Treatment for duodenal atresia requires an operation to remove the blockage (atresia) and repair the duodenum. The surgery is not considered an emergency, and is typically done when the baby is two or three days old.

Although there are several subtypes of duodenal atresia (Figure 4), the surgical procedure is basically the same for all of them. The surgeon opens up the blocked end of the duodenum and then connects it to the rest of the small intestine (Figure 5). The surgeon will also pass a

tube from your baby’s mouth through the stomach and into the small intestine. This “feeding tube” will be used for the first few weeks after surgery.

The surgery is done under general anesthesia. Afterward, your baby will be returned to the NICU. For several days, your baby may need a machine (ventilator) to help with breathing.

Figure 4 - Incomplete obstructions (A) are known as duodenal “webs” because of the web-like membrane that forms inside the duodenum at the point of the obstruction (B). Complete duodenal atresia occurs when a segment of the duodenum is absent (C).

Figure 5 - During surgery to repair duodenum atresia, the surgeon opens up the blocked ends of the duodenum (A) and then sutures them together (B).

What is my baby’s prognosis?

The prognosis for babies with isolated duodenal atresia is excellent when the condition is diagnosed and treated promptly.

Will my baby require long-term follow-up?

Most babies with isolated duodenal atresia do not require long-term follow-up. Such follow-up may be necessary, however, for babies with the condition who also have Down syndrome and/or a heart defect. These children may also require more surgery and hospitalization.

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