Duodenal Atresia/Stenosis

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

Overview - "What is it?"

The *duodenum* is the first portion of the intestine that comes right after the stomach. The duodenum is where the bile and pancreatic juice drains. When babies are born with obstruction of the duodenum, there are several types of blockages. There is either a complete obstruction (atresia), a thin obstructing membrane (a web), or a very narrowed segment (stenosis).

- Atresia is a term used to describe a condition where an opening or passage in the body is closed or absent. Duodenal atresia means that there is a blockage between one part of the duodenum to another. Because of the blockage, the portion of duodenum before the blockage is enlarged (dilated) while the part after the blockage is normal or smaller than normal.
- Stenosis is a term that describes a partial narrowing of an opening or passage. In duodenal stenosis, the narrowing causes slowing of flow of food and fluids through the duodenum. The cause of any of these obstructions is likely multifactorial, with genetic and developmental factors.

Duodenal atresia is the most common cause of intrinsic duodenal obstruction in the newborn. It occurs in 1 in 7,000-40,000 live births. Trisomy 21 (Down Syndrome) occurs 30% of cases of duodenal atresia.

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

Early signs of duodenal atresia may be seen on a regularly scheduled ultrasound performed on the pregnant mother. Because there is blockage of the duodenum, the fluid that the baby swallows builds up, resulting in enlargement of the stomach and the duodenum. After delivery, the newborn will often start vomiting within a few hours of birth and not tolerate feeds. Most often babies will have bilious (green/yellow) emesis.

Later signs/symptoms can occur in the case of duodenal web or stenosis. Infants may be able to tolerate some feedings but as the baby eats more, the food is hung up at the area of narrowing, leading to vomiting and problems gaining weight.

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

Physical examination should be performed by an experienced physician.

Blood tests are usually not done unless a test is sent to look for Down Syndrome.

Prenatal ultrasound shows dilated stomach and duodenum and too much fluid in the uterus.

Abdominal X-ray may show air in the dilated stomach and duodenum. Most of the time, the plain X-ray of the abdomen is enough to make a diagnosis. However, in a patient with a narrowing (instead of complete) blockage, other studies may be needed to confirm the diagnosis.

Upper Gastrointestinal (GI) Study: A contrast is placed in the stomach and duodenum to check the anatomy of these structures and the flow of food through the system.

If the baby has Trisomy 21, an **echocardiogram** is needed prior to surgery because there is an association of heart problems in these patients.

Conditions that mimic this condition include malrotation (abnormal twist of the bowel), duodenal duplication, or blockage caused by organs compressing the duodenum such as *annular pancreas* (pancreas is abnormally shaped and encircles the duodenum), preduodenal portal vein (where the main vein entering the liver lies in front of the duodenum).

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

Medicine: There is no medicine to treat the obstruction, only surgery.

Preoperative care: Prior to surgery a tube (orogastric or nasogastric) is placed in the stomach to remove the fluid in the obstructed stomach and duodenum. The tube decreases the risk of vomiting and aspiration (vomit fluid gets in the lungs).

Surgery: An operation is usually performed once the baby has stabilized from delivery and all testing has been completed. Laparoscopic or open techniques may be used. Laparoscopy is an approach where multiple small incisions are made on the belly to insert a telescope and small instruments. The open technique is an approach where one larger cut is made on the belly to complete the surgery. The surgeon will identify the area of blockage. The intestine before the blocked segment is sewn on to the intestine beyond the blockage, bypassing the obstruction. If the dilated segment is really big, then it may need to be narrowed (duodenoplasty) so it can

work better. During the operation, the surgeon will make sure that there are no other areas of atresia/stenosis in small and large intestine.

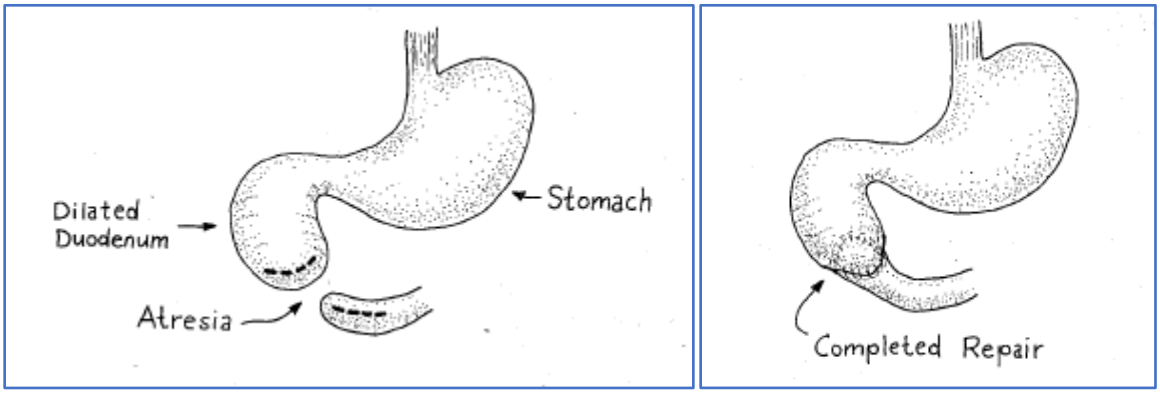

Illustration credits: Mark Mazziotti, MD

Postoperative care: The nasogastric (NG) tube will continue to suction. The dilated stomach and duodenum usually take days, even weeks to function normally so the baby usually needs IV (intravenous) nutrition (TPN) while the stomach and duodenum recover. Feeding orally will start when there is only a little fluid that comes from the NG tube. Increasing feeds will be done gradually. Medicines for pain such as acetaminophen (Tylenol) or ibuprofen (Motrin or Advil) or something stronger like a narcotic may be needed to relieve pain for a few days after surgery.

Risks from the surgery include leak from area where the duodenum is put back together, narrowing of the bypass, wound infection, and slow recovery of the stomach and dilated intestine.

Benefit of surgery is that when the duodenum is put back together, the child is eventually able to eat**.**

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

Diet: Normal for age.

Activity: Normal for age. Ask surgeon when "tummy time" can resume.

Wound care: Can wash incision with soap and water. May not be able to submerge under water for about a week after surgery. By the time the baby is discharged, it should be fine to resume baths.

Medicines: By the time the baby is discharged, pain medication is likely not needed.

What to call the doctor for: Call your doctor for redness, warmth, or drainage from incision, vomiting, or fever.

Follow-up care: Your child should follow up with his or her surgeon 2-3 weeks after surgery to ensure proper postoperative healing. Later follow up may be needed if vomiting is a problem.

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

Survival of babies with duodenal atresia approaches 95%, with most of the deaths coming from associated anomalies (problems other than the duodenal atresia). In some patients the dilated duodenum may have abnormal function and may not move well. This may require reoperation with revision of intestinal connection and/or tapering of dilated duodenum.

Patients may have problems with gastroesophageal reflux, but this is usually treated medically. Patients can also have bowel obstruction from adhesions (scar tissue), as can any patient who has had abdominal surgery. This can be treated medically with NG tube but may need surgery to divide scar tissue

Updated 10/2021
Author: Grace Mak, MD
Editors: Patricia Lange, MD; Marjorie J. Arca, MD; Mark V. Mazziotti, MD, MEd