

## Duodenal Atresia — Symptoms and Treatment

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**Duodenal atresia is one of the most common inborn defects of the digestive system, namely of the intestine, which is often associated with other congenital malformations and genetic pathologies (21 trisomy). The frequency of occurrence of this disorder is affected by the hypoxia of the fetus in the first trimester of pregnancy and is caused by severe chronic respiratory and cardiovascular diseases, stress, diabetes, drug, and alcohol abuse during pregnancy. Nowadays, duodenal atresia is one of the most common congenital defects in the neonates. Moreover, it is well-diagnosed and curable in most of the cases, if timely detected and not neglected.**



### Definition and Background

Atresia is a **complete** obstruction of the lumen of a hollow viscus.

**Duodenal atresia** is a **congenital defect of the gastrointestinal tract** characterized by the complete absence of the lumen of the duodenum (the first part of the small intestine). This makes the absorption and passage of food through the digestive canal impossible, causing duodenal obstruction.

In **duodenal stenosis**, there is partial narrowing (stenosis) of the duodenal lumen. It is

associated with variable and delayed manifestations, followed by concealed symptoms at the beginning, and may last up to adulthood.

## Etiology & Pathophysiology of Duodenal Atresia

The exact etiology of duodenal atresia is not known as the most cases are sporadic. However, it is suggested that during the early weeks of gestation (6th and 7th week), the endodermal epithelium proliferates, and the portions of the gastrointestinal tract lumen become occluded. The recanalization normally occurs during 8 - 10 weeks of gestation, and the lumen patency is restored. The failure to recanalize due to any reason results in atresia. One of the suggested reasons for the recanalization failure is **hypoxia** of the fetus during the early stages of embryogenesis (ischemic episode).

## Epidemiology of Duodenal Atresia

Duodenal atresia is a relatively common congenital anomaly of the gastrointestinal tract occurring in 1 in 2,500-40,000 live births. One-Third of these cases are associated with Down syndrome (trisomy 21). There is no racial or gender propensity, with equal incidence in both males and females.

There is a pattern for intrinsic inborn duodenal obstructions:

- Duodenal atresia, 40-60%
- Duodenal web, 35-45%
- Annular pancreas, 10-30%
- Duodenal stenosis, 7-20%

## Presentation of Patients with Duodenal Atresia



**Image:** "Duodenal atresia in a neonate," by Kinderradiologie Olgahospital Klinikum Stuttgart - Own work. License: [CC BY-SA 3.0](https://creativecommons.org/licenses/by-sa/3.0/)

In 85% of the cases of complete duodenal atresia, **bilious** vomiting occurs during the **first hours of life**. The color of the vomitus is yellowish-green, sometimes with brown granular matter. In the rest of the cases (15%), the vomiting may be non-bilious as the

atresia occurs proximal to the ampulla of Vater.

On physical examination, the abdomen is sunken (scaphoid abdomen). There may be epigastric fullness due to distension of the stomach and proximal duodenum. Although the bowel movements are absent due to high intestinal blockage, the meconium passage is not affected during the first 24 hours after birth. The affected newborn may be small for his/her gestational age.

The congenital intestinal defects that are associated with bilious vomiting and the time of manifestation are:

- **Duodenal atresia**, a few hours after birth, no distention
- **Malrotation with volvulus**, at 3 to 7 days, rapid deterioration with volvulus
- **Jejunoileal atresia**, during the first hours of birth, abdominal distention

**Partial duodenal atresia (stenosis) symptoms** depend on the severity of the defect. The illness may not show any symptoms for months or even years and usually manifests with long-lasting vomiting, constipation, food aspiration and failure to thrive. The constipation is unresponsive to treatment.

The duodenal atresia or stenosis may be an isolated finding, but most commonly they are associated with other congenital defects such as:

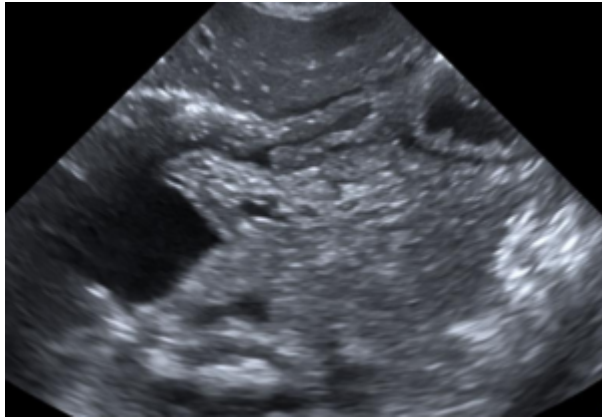
- Down syndrome (most common and present in 25-40% of the cases)
- VACTERL association
- Annular pancreas
- Anal atresia
- Ileal atresia
- Jejunoileal atresia

## Differential Diagnosis of Duodenal Atresia

| Atresia  | Anal<br>Esophageal<br>Small bowel  |
|--|--|
| Plain radiograph appearance                                  | Duodenal stenosis<br>Duodenal web<br>Intestinal malrotation<br>Midgut volvulus |
| Ultrasound appearance of a "double bubble type" (distal gas) | Fetal choledochal cyst<br>Fetal omental cyst<br>Fetal duplicated cyst          |

## Diagnosis of Duodenal Atresia

### Imaging studies



[Image](#): "Duodenal atresia neonate ultrasound double bubble," by Kinderradiologie Olgahospital Klinikum Stuttgart – Own work.  
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**Radiography** is an informative method of examination in duodenal atresia. It finds the dilated stomach and the first part of the duodenum ('double bubble') and the absence of air behind the second bubble. This finding of a **double bubble sign with no distal gas** strongly suggests duodenal atresia.

**Ultrasonography** is efficient in the prenatal diagnosis of duodenal atresia, apart from the specific appearance of the abdomen; there is **polyhydramnios**, which is a prominent symptom of the defect. Ultrasonography is preferred in esophageal and duodenal atresia.

**Barium enema examination** is an additional study that makes distant duodenal atresia, as well as malpositioned cecum, evident. However, in the diagnostics of malrotation and volvulus, barium enema is not that informative.

## Laboratory studies

No laboratory studies are diagnostic of duodenal atresia. However, laboratory studies help in the assessment of the infant's overall clinical condition.

- **Blood glucose** may be low in premature babies; they tend to develop hypoglycemia as there is a limited supply of glycogen.
- **Serum electrolytes** have to be determined as infants with duodenal atresia tend to lose fluids and electrolytes produced by the stomach, due to a large amount of gastric aspiration.
- **Karyotype analysis** is the appropriate method of examination in the case of **trisomy 21**.
- **Immunohistochemistry** of proximal and distal duodenal samples of affected infants reflects changes and transformation of neuronal cells, intestinal cell Cajal, hypertrophy of circular muscles. All of these witness about the persistent disorder of the duodenal movement after primary surgery.
- **Measurement of gastric aspiration.** Normally, it should not exceed 5 ml. Duodenal atresia is suspected when an infant's gastric aspiration is greater than 30 ml. The fullness of the epigastrium may be detected manually and visually when the food gets stacked above the narrowed lumen.
- **Rectal biopsy** conducted in order to exclude **Hirschsprung disease**, especially in patients with **Down syndrome**.
- **Hematocrit** has to be checked prior to [general anesthesia](#).
- **Blood type** and crossmatch.
- **Arterial blood gas measurements** are needed if the infant has aspirated or

shows signs of respiratory distress.

## Management of Duodenal Atresia

There is no conservative treatment available for this condition.

### Surgical intervention

| Type of duodenal obstruction | Preoperative management   | Surgical treatment  | Postoperative period   | Prognosis  |
|------------------------------|---|---|--|--|
| Duodenal atresia             | IV fluids and nasogastric suction 24-48 hours                                       | Diamond-shaped duodenostomy                                 | No feeding per os for 2-4 days after operation; nasogastric suction. | Benign, if there are no serious associated congenital defects. |
| Jejunioileal atresia         | IV fluids, STAT surgery for the patients with obvious symptoms; nasogastric suction | Ladd's procedure: sometimes a second laparotomy is required | No feeding per os; nasogastric suction                               | Benign, if there is no large bowel resection.                  |
| Jejunioileal atresia         | IV fluids and nasogastric suction for 24-48 hours                                   | Resection and anastomosis                                   | No feeding per os for 2-4 days after operation; nasogastric suction. | Benign, if there is no large loss of the bowel.                |

In spite of the benign prognosis after the surgery, 22% of babies may develop distant complications:

- Blind loop syndrome
- Megaduodenum with altered duodenal motility
- [Gastritis](#) with duodenal-gastric reflux
- [Peptic ulcer](#)
- Esophagitis and gastroesophageal reflux
- Pancreatitis
- Cholecystitis

## References

[Pediatric Duodenal Atresia and Stenosis Surgery](#) via medscape.com

[Duodenal Atresia or Stenosis](#) via rarediseases.org

[Fetal Duodenum](#) via fetalultrasound.com

[Duodenal atresia](#) via radiopaedia.org

[Bilious Vomiting in the Newborn: Rapid Diagnosis of Intestinal Obstruction](#) via aafp.org

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