Esophageal Atresia — Symptoms and Treatment

In most cases, medical students specializing in pediatrics may face esophageal atresia. Just a few key aspects are relevant examination points in this matter, but they have to be clarified. Here you can get an overview of the important basics which may help you later in clinical routine.

Definition of Esophageal Atresia

Esophageal atresia (or *oesophageal atresia*) describes a malformation of the *esophagus*, a congenital medical condition which affects the alimentary tract. Hereby, we may mention various forms of manifestation. Mostly, there is a discontinuity of the esophagus and a *tracheoesophageal fistula* between the esophagus and the trachea. Esophageal atresia is present in 1 in 3500 live births.

Syndromes associated to esophageal atresia

The disease is often (up to 50% of cases) associated with other malformations such as VACTERL syndrome and CHARGE syndrome, being a disorder that affects many body systems.

- Vertebral defects
- Anal atresia
- Cardiac defects (up to 30% of cases)
- Tracheo-esophageal fistula
- Renal abnormalities
- Limb abnormalities
And

- Coloboma
- Heart defects
- Atresia of the choanae
- Retardation of mental and/or physical development
- Genital hypoplasia
- Ear abnormalities

Pathophysiology of Esophageal Atresia

What causes esophageal atresia?

The esophagus develops in the embryonic period from the foregut. First, there appears a connection between the respiratory and digestive pathways which however will be closed in the course of development by the oesophagotracheal septum up to the proximal sector.

Disturbances in this development, during the fourth up to the sixth weeks of pregnancy, often cause the esophagus to end in a blind-ended pouch, rather than connecting normally to the stomach, frequently in combination with a tracheoesophageal fistula in the lower section.

Classification by Vogt

Classification of esophageal atresia

Esophageal atresia may be divided into types I-III according to Vogt:

- Vogt I (< 1 %): Esophageal aplasia, with no esophagus;
- Vogt II (about 10 %): Esophageal atresia without a tracheoesophageal fistula;
- Vogt III a (< 1 %): Esophageal atresia with a proximal tracheoesophageal fistula;
- Vogt III b (80 %): Esophageal atresia with a distal tracheoesophageal fistula;
- Vogt III c (5 %): Esophageal atresia with a proximal and distal tracheoesophageal fistula;

And finally, there is “H-fistula”: in this case tracheoesophageal fistula exist but there is no esophageal atresia.

**Note:** Esophageal atresia with a lower tracheoesophageal fistula (Vogt III b) represents at 80 % the most frequent form of esophageal atresia!

Symptoms of Esophageal Atresia

**Prenatal:**

- Intrauterine complication that may end with preterm labor and preterm birth is known as polyhydramnios. The fetus swallows amniotic fluid, which does not reach the gastrointestinal tract and cannot be absorbed due to the malformation, but it accumulates.

**Postnatal:**
Noticeable symptoms of the newborn babies are frequent bouts of coughing and rattling breathing with respiratory disorders up to cyanosis.

In the case of H-fistula, the symptoms appear in the later stage since there is no discontinuity of the esophagus. However, due to the existing fistula between the esophagus and the trachea, the baby often coughs and demonstrates aspiration from food at feeding time. The children often suffer from respiratory infections including pneumonia, due to high risk of aspiration.

Diagnosis of Esophageal Atresia

Diagnostic imaging

In some cases, the esophageal atresia can be diagnosis on a prenatal ultrasonography, especially in the 3th trimester.

In born, on suspicion of esophageal atresia, in the first step, physician may use a gastric tube which can provide the first indication: the gastric tube should be protruded 10 cm deep, but not longer, so that the examiner may encounter a resistance. Physician may inject air into the tube and check its location with a stethoscope: typical gurgles cannot be auscultated as usual over the stomach. Also, no stomach contents can be aspirated (except for the case with H-fistula).

A chest X-ray can help in diagnosis of esophageal aresia. The “blind sac” sign is show in the superior mediastinum in this study. The esophagus in its upper part is air-filled and demonstrates increased transparency up to its discontinuity. If contrast medium was used, the “blind sac” appears to look hyper-transparent. In the case of a tracheoesophageal fistula, an air-filled stomach is also evident due to the connection of the trachea and the esophagus.

Note: In the chest X-ray you may scan the “blind sac” and diagnose esophageal atresia. In endoscopy, a tracheoesophageal fistula can be represented particularly well.

Due to the known VACTERL and CHARGE syndrome, targeted further diagnostics with respect to the associated medical conditions should always be conducted.

Differential Diagnoses

Similar clinical patterns as in the case of esophageal
atresia

If a newborn baby has the symptoms described above, other medical conditions should be kept in mind along with esophageal atresia. So, with a cesarean section, a newborn may swallow a big amount of amniotic fluid and then produce frothy sputum. However, this should be self-adjusted after the correspondent expectoration. Furthermore, achalasia, esophageal stenosis or impaired swallowing reflex could be put in question.

**Therapy of Esophageal Atresia**

**Treatment options for esophageal atresia**

The operation is not an emergency procedure; however, it should take place within the first two days of life. Preoperatively, a baby must not be fed or intubated in order to avoid aspiration. By means of a gastric tube, salivary secretion can be derived from the upper esophageal section, parenteral nutrition as well as an adequate fluid and electrolyte balance is necessary. If you suspect of aspiration pneumonia, a course of antibiotics should be prescribed.

**Note:** In the case of suspicion of esophageal atresia, the baby should not be fed!

![Image: “Oesophageal atresia” by Spitz L – Orphanet J Rare Dis (2007), License: CC BY 2.0](image)

When performing an operation, it should be attempted to anastomose the two esophageal ends with each other; any existing fistula should be closed. If a direct end-to-end anastomosis is not possible, a gastric pull-up as an esophageal replacement with plastic tubes may be required.

**Prognosis of Esophageal Atresia**

The prognosis is usually very good, but it is also optionally dependent on the additionally malformations. Unfortunately, during the operation, there appear fairly frequent long-term complications, such as [GERD](https://www.ncbi.nlm.nih.gov/pubmed/19580554) a gastroesophageal reflux disease, or anastomotic stenosis.
Review Questions

The correct answers are below the references.

1. **The most frequent form of esophageal atresia is:**
   
   A. Vogt I: Esophageal aplasia
   B. Vogt III a: Esophageal atresia with a proximal tracheoesophageal fistula
   C. Vogt III b: Esophageal atresia with a distal tracheoesophageal fistula
   D. Vogt III c: Esophageal atresia with a proximal and distal tracheoesophageal fistula
   E. None of the above-mentioned points from A) to D).

2. **After protruding a gastric tube, there appears a suspicion of esophageal atresia in a newborn baby. In chest X-ray, a blind sack in the superior mediastinum and an air-filled stomach bubble become apparent. Which form of esophageal atresia is explicit?**
   
   A. This cannot be determined on the basis of X-ray findings.
   B. H-fistula
   C. Esophageal atresia with a proximal tracheoesophageal fistula
   D. Esophageal aplasia
   E. Esophageal atresia with a distal tracheoesophageal fistula

3. **A newborn definitely suffers from esophageal atresia and a tracheoesophageal fistula. Which of these additional defects would you least suspect?**
   
   A. Vertebral defects
   B. Cardiac defects
   C. Limb abnormalities
   D. Abnormal thymus
   E. Renal anomalies

References

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Correct answers: 1C, 2E, 3D

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