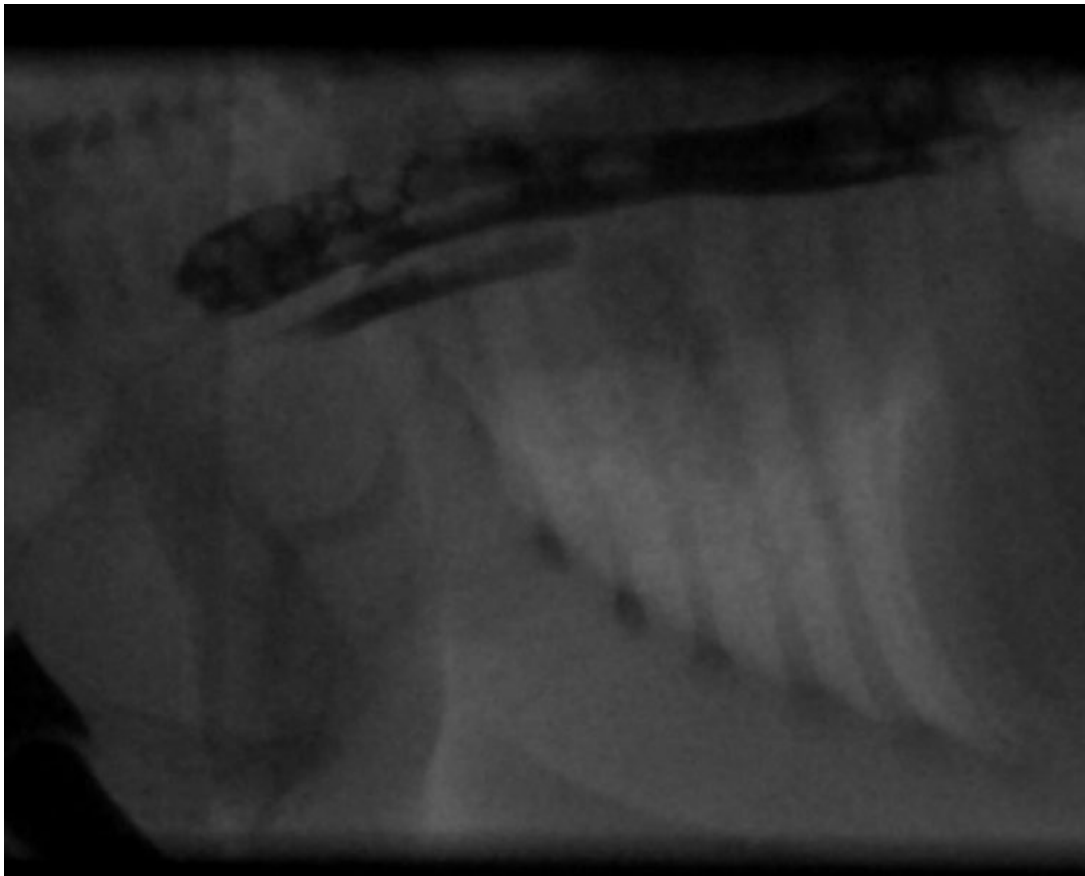


Tracheoesophageal Fistula (TEF) — Symptoms and Treatment

[See online here](#)

Congenital tracheoesophageal fistulas are a common congenital anomaly that is usually diagnosed prenatally or during the neonatal period. The condition is usually characterized by an esophageal atresia associated with a communication between the trachea and the esophagus. Maternal polyhydramnios is commonly seen in cases of esophageal atresia. The diagnosis can be confirmed prenatally by ultrasonography or postnatally by a chest X-ray or a computed tomography scan. Early surgical repair is recommended in all healthy infants.



Overview

- A fistula is an abnormal communication between two epithelial-lined surfaces.
- Atresia refers to complete obliteration or absence of an orifice of a hollow organ.
- A stenosis is change in diameter of a hollow organ to become narrower. It can

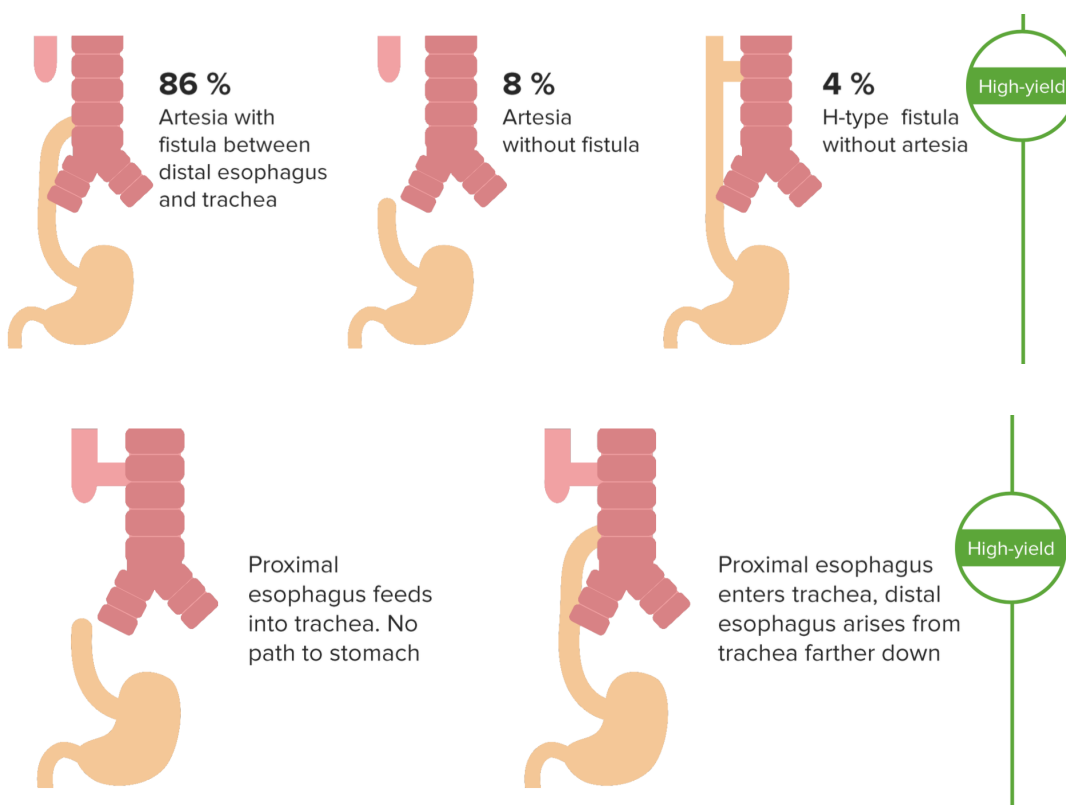
also be called stricture.

- A tracheoesophageal fistula, as the name implies, is an abnormal **communication between the trachea and esophagus**. Congenital tracheoesophageal fistulas are usually diagnosed at birth or soon thereafter.

Classification

Congenital tracheoesophageal fistulas can be classified into **five distinct types** based on the site of the communication as follows:

- **Esophageal atresia with distal tracheoesophageal fistula** is seen in up to 86% of the cases. It is the most common type.
- **Isolated esophageal atresia** without tracheoesophageal fistula. This type is seen in approximately 8% of the cases.
- **Isolated tracheoesophageal fistulas** come in third place in frequency and are diagnosed in 4% of the cases.
- **Esophageal atresia with proximal communication** represents 1% of tracheoesophageal fistulas.
- **Esophageal atresia with proximal and distal communications** represents 1% of tracheoesophageal fistulas.



Epidemiology of Congenital Tracheoesophageal Fistulas

The estimated incidence of congenital tracheoesophageal fistulas is **around 1 in 2,000 to 4,000 live births**. There are no racial differences and the incidence is equal in both genders.

Most newborns with congenital tracheoesophageal fistulas are diagnosed during the

neonatal period because of the emergence of **severe complications**. In fact, the diagnosis of a congenital tracheoesophageal fistula is most commonly made at birth.

The prognosis of infants who had a congenital tracheoesophageal fistula surgically repaired is excellent with a 100% survival rate. If early surgical repair is not possible, the survival rate can drop by 10 to 20%.

Despite an excellent survival rate, infants are at risk of developing several morbidities after the repair of the tracheoesophageal fistula. **Esophageal dysmotility**, **gastroesophageal reflux** and **dysphagia** are commonly seen.

Recurrent pneumonia is another common complication of tracheoesophageal fistulas. **Tracheomalacia** has also been reported to be more common in infants who had a tracheoesophageal fistula.

Etiology of Congenital Tracheoesophageal Fistulas

Certain genetic disorders and **environmental exposures** have been linked to an increased risk of congenital tracheoesophageal fistulas. **Trisomies 18, 13 and 21** have been associated with tracheoesophageal fistulas.

Additionally, the use of the **decongestant imidazoline** in the first trimester has been linked to an increased risk of congenital tracheoesophageal fistulas. In most cases, however, a single etiology cannot be identified.

Pathophysiology of Congenital Tracheoesophageal Fistulas

Tracheoesophageal fistulas are usually associated with multiple anomalies that can affect the survival of the child.

As many as 30% of infants with esophageal atresia or tracheoesophageal fistula have other congenital defects; the most common association is described by the acronym **VACTERL complex** and includes:

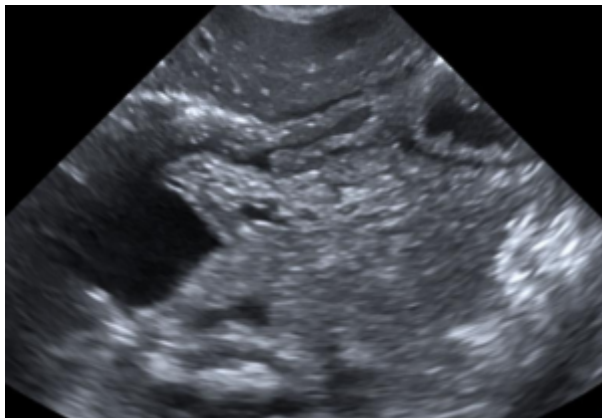
- Anomalies of the vertebrae (similar to those of spondylocostal dysplasia, 10%),
- Anal atresia (10 to 15%),
- Cardiac malformations (patent ductus arteriosus, atrial septal defect, or ventricular septal defect in 25%). They are commonly seen in children with congenital tracheoesophageal fistulas.
- Tracheoesophageal fistula.
- Renal anomalies (urethral atresia with hydronephrosis, renal agenesis, horseshoe kidney, polycystic kidney).
- Limb anomalies (hexadactyly, humeral hypoplasia, radial aplasia, and proximally placed thumb).

Tracheoesophageal fistulas can also be associated with a **duodenal atresia**, **malrotation of the gut**, **Meckel diverticulum** and **annular pancreas** as well. These associations suggest a common pathophysiology or etiology between these different malformations.

This association is closely linked with errors in the embryonic period as the respiratory system outpouches from the primitive foregut.

During **embryogenesis**, the **primitive foregut** gives rise to a **ventral diverticulum** which evolves into the trachea. This event usually starts happening in 4 to 6 weeks after conception and is also characterized by the **fusion of the tracheoesophageal folds** to form a **ventral laryngotracheal tube** and a **dorsal esophagus**.

This process is usually impaired in children with a tracheoesophageal fistula as the septum between the two tubes fails to form at some point.



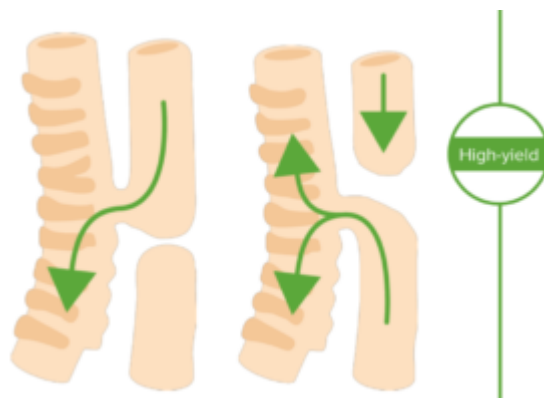
[Image](#): "Duodenal atresia neonate ultrasound double bubble." by Kinderradiologie Olgahospital Klinikum Stuttgart - Own work. License: [CC BY-SA 3.0](#)

Clinical Presentation of Congenital Tracheoesophageal Fistulas

Esophageal atresia, a key component in most types of tracheoesophageal fistulas, can be diagnosed prenatally. **Maternal polyhydramnios** is usually evident in most cases and **prenatal ultrasonography** can reveal the stomach's gas absence.

During the neonatal period, the most common presentation is the development of **copious amounts of white frothy bubbles** in the mouth and nose. These bubbles recur despite repeated suctioning. Infants with congenital tracheoesophageal fistulas usually present with **respiratory distress symptoms** and **recurrent pneumonia**.

Coughing, **choking** and **cyanosis** can be seen in an infant with a congenital tracheoesophageal fistula, especially during feeding.



Most TEF present almost immediately after birth.

- Food cannot get to the stomach, resulting in **immediate emesis** and **rapid dehydration**.
- Gastric contents may enter the lungs, resulting in **respiratory distress**.

The main goal of a physical examination in an infant with tracheoesophageal fistula is to exclude any of the previously-mentioned anomalies that can be associated with this congenital malformation.

Diagnostic Workup for Congenital Tracheoesophageal Fistulas

Laboratory investigations are not useful in establishing the diagnosis of a congenital tracheoesophageal fistula. **Prenatal and postnatal imaging studies** are the main diagnostic tools to establish the diagnosis of a tracheoesophageal fistula.

Prenatal ultrasonography can reveal **polyhydramnios**, the **absence of gas in the stomach**, a **distended esophageal pouch** and **in-utero growth retardation**. These findings are indicative of esophageal atresia and not a tracheoesophageal fistula per se, but, because esophageal atresia is seen in most cases of tracheoesophageal fistulas, establishing the diagnosis on the confirmation of an esophageal atresia is reasonable.

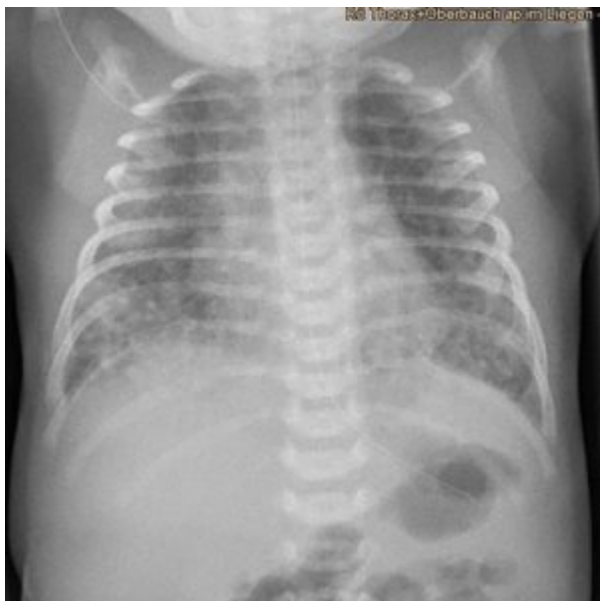


Image: "Chest X-ray of a female neonate with meconium aspiration." by Kinderradiologie Olgahospital Klinikum Stuttgart - Own work. License: [CC BY-SA 4.0](https://creativecommons.org/licenses/by-sa/4.0/)

Neonates or infants with a congenital tracheoesophageal fistula should undergo **chest X-ray imaging** to exclude complications such as **aspiration pneumonia** and to have more clues to confirm the diagnosis. The most common findings on a chest X-ray are **tracheal compression and deviation**, or the **absence of a gastric bubble**.

A **nasogastric tube** might be inserted to confirm the diagnosis of esophageal atresia. When esophageal atresia is present, the nasogastric tube will coil in the mediastinum and one would visualize the radiopaque line of the catheter.

Multidetector-row computed tomography scans can be used to confirm the

diagnosis of a tracheoesophageal fistula without the use of a contrast medium. Because of the non-invasiveness of the procedure, computed tomography scans become preferable by most pediatric surgeons to plan the repair procedure for tracheoesophageal fistulas.

Treatment of Congenital Tracheoesophageal Fistulas

It is essential to transfer any neonate or infant who is diagnosed with a tracheoesophageal fistula to a **pediatric hospital** that has **experience in the surgical repair of tracheoesophageal fistulas**. The early repair of the fistula has been associated with an excellent survival rate.

Definitive treatment:

Surgical repair can be delayed in neonates with **very low birth weight** or **aspiration pneumonia**. All other healthy infants should be provided a primary surgical repair of the fistula within the first few days of life; however, this may not be possible depending on the nature of the lesion (distance between the segments) and the clinical status of the neonate, thus, several surgical approaches have been adopted.

- Primary ligation of the fistula and anastomosis of the esophageal segments if the distance is not too great.
- Two-stage procedure in instances where the ends of the esophagus cannot be brought together.
 - The initial surgery entails fistula ligation, esophageal stoma, and a gastrostomy fashioning. The neonate should be breastfed, and the milk let out through the esophageal stoma (sham feeding) to allow for the baby to learn how to breastfeed while actual feeding is done through the gastrostomy tube.
 - The second surgery involves anastomosis of the esophageal segments after growth in length. Gentle stretching of the esophageal segments before later anastomosis may be beneficial, or repair by interposing a segment of colon between the esophageal segments may be required.

Echocardiography should be performed prior to surgery to rule out a right-sided aortic arch which would lead to preference of a left-sided thoracotomy instead of a median sternotomy approach.

A **cuffed endotracheal tube** should be placed distal to the fistula to prevent aspiration during the operation.

Once the fistula is repaired, care must be taken to **prevent gastroesophageal reflux and prolonged mechanical ventilation** as they were associated with an increased risk of recurrent tracheoesophageal fistulas.

Supportive treatment:

- The tube in the proximal pouch should be placed on continuous suction to drain secretions and prevent aspiration.
- The head of the bed should be elevated to prevent reflux of gastric contents through the distal fistula into the lungs.

- Intravenous glucose and fluids should be provided, and oxygen administered as needed.

Use of a **tracheostomy if the difficulty in breathing and cyanosis is not alleviated by the above measures.**

Prognosis

The prognosis of infants who had a congenital tracheoesophageal fistula surgically repaired is excellent with a 100% survival rate. If early surgical repair is not possible, the survival rate can drop by 10 to 20%.

Complications

Despite an excellent survival rate, infants are at risk of developing several morbidities after the repair of the tracheoesophageal fistula.

- **Esophageal dysmotility**, [gastroesophageal reflux](#) and **dysphagia** are commonly seen.
- **Recurrent pneumonia** is another common complication of tracheoesophageal fistulas.
- **Tracheomalacia** has also been reported to be more common in infants who had a tracheoesophageal fistula.

References

Sharma S. [Tracheoesophageal Fistula](#). Background, Pathophysiology, Etiology. <http://emedicine.medscape.com/article/186735>. Published February 7, 2017. Accessed May 1, 2017.

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