Esophageal Atresia *and* Tracheoesophageal Fistula

Information for parents and families

L'Hôpital de Montréal pour enfants
The Montreal Children's Hospital
Centre universitaire de santé McGill
McGill University Health Centre

What is Esophageal Atresia (EA) and Tracheoesophageal Fistula (TEF)?

The esophagus is the tube that carries food and saliva from the mouth to the stomach.

EA is a malformation of the digestive system, which happens when the esophagus does not develop properly. The esophagus does not connect to the stomach.

A fistula is an abnormal passageway between two structures that normally are not connected. Therefore, a tracheoesophageal fistula (TEF) is an abnormal link between the trachea (where air goes in) and the esophagus (where food goes in).

There are several types of EA/TEF:

Type A: The upper esophagus simply ends and does not connect with the lower esophagus and stomach. No fistula associated.

Type B: As in type A, but there is a fistula at the upper portion of the esophagus.

Type C: EA along with a fistula connecting to the lower portion of the esophagus (this is the most common form of EA).

Type D: EA with a fistula on both the upper and lower segments of the esophagus (this is rare).

Type E: A fistula is present without an EA (also called an H-type fistula).

What causes EA/TEF?

EA/TEF is a congenital defect, which means it happens before birth. In most cases, this defect occurs randomly. A small percentage of cases are linked to chromosomal abnormalities, or a genetic disorder. One in every 2,500 to 3,000 babies is affected by EA/TEF.

What are the signs of EA/TEF?

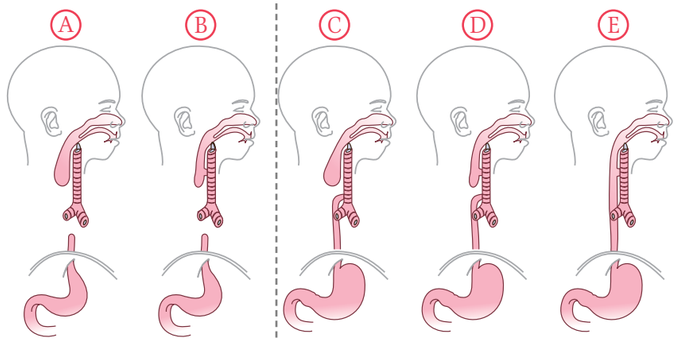
Some possible signs include:

- During pregnancy, there may be a large amount of amniotic fluid around the fetus (also known as polyhydramnios).
- Once born, the baby cannot swallow his/her saliva. Bubbly saliva may be seen coming out of the baby's mouth.
- The newborn may have difficulty breathing and have episodes of coughing and choking.
- The baby may appear cyanotic (have a blue tint to his/her skin).
- The baby may have an enlarged abdomen due to air building up in his/her stomach.

Are there other problems associated with EA/TEF?

Yes, other medical problems occur in about 50 per cent of children with this malformation. Other malformations can be involved at the same time including (these are often referred to as the VACTERL association):

- Vertebral (spine)
- Anorectal (anus)
- Cardiac (heart)
- Tracheoesophageal
- Renal (kidneys)
- Limb defects (arms, hands, feet)



Isolated EA: 8% | EA with proximal TEF: 1% | EA with distal TEF: 87% | EA with double TEF: 1% | Isolated TEF: 4%

How is EA/TEF treated?

- Initially, a tube is inserted into the upper portion of the esophagus to suction the accumulation of saliva.
- After the baby is admitted to the Neonatal Intensive Care Unit (NICU) and stabilized, an operation will be done.
  - If an atresia is present, a surgery called an anastomosis is done. This includes sewing the upper and lower portions of the esophagus together.
  - Sometimes the two portions of the esophagus are too far apart to allow a surgeon to sew them together. In this case, the surgery may have to be delayed until your baby is bigger. In the meantime, your child will be fed through a gastrostomy tube (tube directly inserted into the stomach).
  - If a tracheo-esophageal fistula is present, surgery is performed to close the fistula.

What are the long-term complications?

Reflux is a common complication. This happens when the contents of the stomach are brought back up into the esophagus. This is usually treated with medication and sometimes requires surgery.

The trachea is also usually abnormal in patients with EA and TEF. It tends to be softer and more prone to collapse, leading to a condition called tracheomalacia. This results in a "barking" cough. Sometimes it may result in severe "blue spells," which would require further investigation and possibly additional surgery.

Following the surgery of the esophagus, the area where the repair was done may become narrow (stenosis). This area may need to be stretched (dilation).

Breathing problems such as bronchitis, pneumonia or asthma are common in infancy and tend to improve with time. These problems are usually well treated with medication.

Food and liquids may move more slowly down the esophagus (dysmotility), but most children adapt very well with time.

The EA/TEF Multidisciplinary Team of The Montreal Children's Hospital of the MUHC follows all children diagnosed with EA/TEF. The team meets regularly to determine what form of treatment and follow-up would ensure the best quality of care for your child.

Team members include:

Gastroenterologist • Surgeons • Pediatrician • Respirologist • Otorhinolaryngologist (ENT) • Occupational Therapist • Nurse • Dietician • Others as needed including: Geneticist • Cardiologist • Orthopedic Surgeon • Psychologist

Other resources:

www.afao.asso.fr
www.tofs.org.uk
www.eatef.org
www.aqao.org

Contact us for more information:

514.412.4400, ext. 23435
www.thechildren.com

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This brochure was prepared by the Esophageal Atresia–Tracheoesophageal Fistula Clinic team at the Montreal Children's Hospital of the McGill University Health Centre.
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