Esophageal Atresia and Tracheoesophageal Fistula (EA-TEF)

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

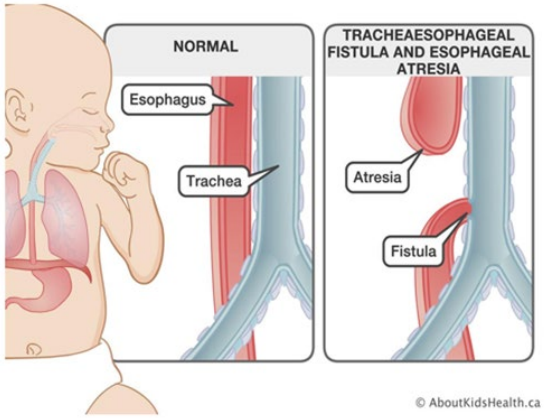
Overview

Esophageal atresia (EA) and Tracheoesophageal fistula (TEF) are rare conditions that develop before birth. They often happen together and affect the development of the esophagus, trachea, or both. These conditions can be very serious and need surgery shortly after birth. EA-TEF is rare, occurring in about one infant for every 4,000 births in the USA. The exact cause is not known, but there may be a genetic component.

"What is the esophagus and the trachea?"

The esophagus is part of the digestive system and is a muscular tube that carries food from the mouth into the stomach. The trachea is part of the respiratory system and is a windpipe that moves air from the mouth into the lungs.

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



(This image was created by The Hospital for Sick Children and is shared with permission from the AboutKidsHealth Team. Image can be accessed at https://www.aboutkidshealth.ca/Article?contentid=470&language=English.)

During pregnancy, the esophagus and trachea start as one tube and then separate into two completely separate tubes. EA, with or without TEF, happens when the separation does not happen properly. This can make the upper part of the esophagus a dead-end pouch, blocking swallowed food or spit from reaching the stomach.

"What are the different kinds of EA-TEF?"

While EA may happen alone, most infants with EA will also have a TEF. This is an abnormal connection between the esophagus and the trachea, which can cause breathing problems since food may enter the lungs. There are many types of EA based on the presence and location of a TEF.

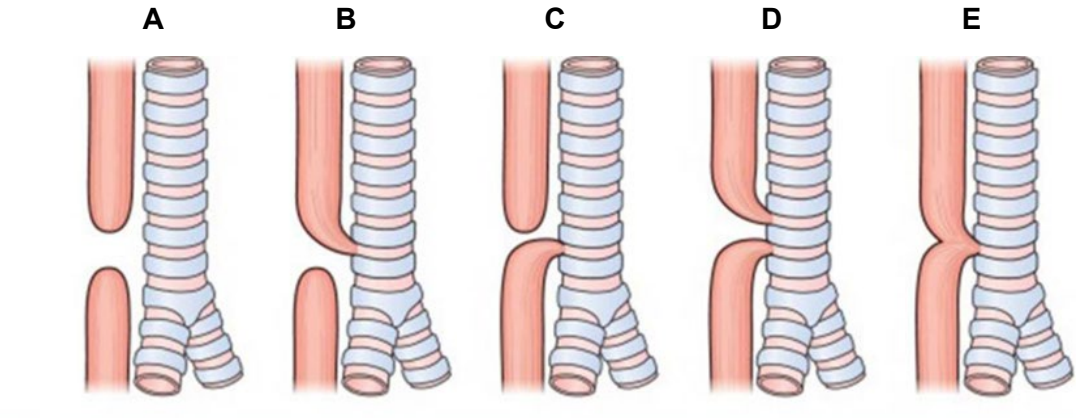
Type A: EA without a TEF, also called "pure" EA (~10%)

Type B: EA with a TEF on the upper pouch (~1%)

Type C: EA with a TEF on the lower pouch (**most common ~85%**)

Type D: EA with TEFs on both the upper and lower pouch (~<1%)

Type E: TEF without EA or "H-type fistula" (~4%)



(Image citation: Krishnan U. Eosinophilic Esophagitis in Esophageal Atresia. *Front Pediatr.* 2019 Nov 29;7:497.)

"What other conditions can occur with EA-TEF?"

More than half of the infants born with EA-TEF will also have abnormalities of one or more of the following body systems: Vertebral, Anorectal, Cardiovascular, Renal (kidney), and Limbs (VACTERL association). You can learn more about these associated conditions in the "VACTERL Association" document on our Patient and Parent Education page.

"What symptoms of EA-TEF will my child have?"

While developing in the uterus, an infant with EA-TEF may have too much amniotic fluid (polyhydramnios). After birth, your child may have trouble swallowing, drool a lot, or have frothy white bubbles coming from the mouth. Other signs include choking and coughing when trying to feed. Breathing problems (aspiration pneumonia) may occur if spit enters the trachea and lungs. Most infants born with EA-TEF are diagnosed soon after birth when symptoms first appear.

"What tests are done to confirm that my child has EA-TEF?"

To diagnose EA-TEF, medical providers will try to pass a tube from the nose or mouth into the stomach. If the tube doesn't pass easily, there might be a blockage in the esophagus. Chest and belly X-rays can show where the blockage is and if there's gas in the stomach, indicating a

connection between the esophagus and the trachea (windpipe). A special tube called a Replogle may be used to clear out spit until surgery.

"What surgery will be done to fix the EA-TEF and make my child better?"

Surgery is needed to repair EA-TEF. The type of surgery depends on the type of EA-TEF, the distance between the ends of the esophagus, and your child's overall health. The goals of surgery are to separate the esophagus and trachea and connect the two ends of the esophagus so food can be swallowed normally. Infants born premature and/or with heart or lung problems may need these other problems treated first, before they are safe to have EA-TEF surgery.

Surgery is done either through a single large incision between the ribs of the chest (thoracotomy) or several small incisions with long instruments and a camera (thoracoscopy). Some infants need only one surgery ("primary repair") and others need multiple surgeries ("staged repair").

Primary Repair**:** This type of surgery is done when the ends of the esophagus are not far apart and can be connected in one surgery. The area where the two ends are connected or sewn together is called an "anastomosis". In some cases, your child's surgeon may recommend a "delayed primary repair" by waiting a period of time before surgery to let the pouches of the esophagus grow closer together to allow for primary repair.

Staged Repair**:** If the two ends of the esophagus are too far apart to connect (long-gap EA) in a single surgery, the surgery is separated into multiple stages or steps to bring the ends of the esophagus together. This may involve helping to stretch the ends of the esophagus to bring them closer together to be connected.

"What care is needed after surgery while my child heals?"

After surgery, your child will need time to heal. They will return to the NICU for recovery. Recovery includes different care depending on the type of surgery. Primary repair tends to have a shorter recovery period than staged repair.

Temporary paralysis and breathing support: Your child may need sedation (medicine to make them very sleepy), temporary paralysis (medicine to make them unable to move their own body), and help breathing with a breathing tube and ventilator (breathing machine).

Lines and Tubes: Your child will be connected to machines to monitor the heart and lung function. They will likely have a chest tube (small tube into the side of the chest) that drains air and fluid from the chest after surgery. They may also have a urinary catheter (small tube that goes into the bladder).

Medications: Your child will have an IV (small tube into a blood vessel) for extra hydration (fluids), medications to treat pain, and antibiotics to prevent infection.

Nutrition: Your child may need special nutrition called total parenteral nutrition (TPN). This is given through a special type of IV called a peripherally inserted central catheter (PICC) or a central venous catheter (CVL). In some situations, a special feeding tube, called a gastrostomy tube (G-tube), is placed through the belly wall and into the stomach at the time of EA-TEF surgery.

"What testing and procedures may be needed after surgery?"

After surgery, tests that may be done to make sure the esophagus has healed properly include:

Esophagram: This test is done about 1-2 weeks after surgery to see if the esophagus is fully healed. Contrast dye is given by mouth or through a tube inserted from the nose to the esophagus, while having an X-ray to see if the dye stays inside the esophagus or leaks outside into the chest. This test also shows if there is a stricture (a tightening caused by scar tissue) that needs further treatment.

Esophagogastroduodenoscopy (EGD) or Upper Endoscopy: An endoscope (long flexible tube with a camera on the end) is passed through your child's mouth or G-tube site while under anesthesia to check the esophagus for healing and to gently stretch any narrow areas.

"What are common problems after EA-TEF surgery?"

Doctors will monitor for problems that can occur after surgery, either while in the hospital or after discharge home.

Early/In-hospital Complications:

Esophageal leak: If the pouches of the esophagus do not heal tightly together, swallowed food or spit can move out of the esophagus and into the chest or lungs which can cause an infection. Signs of a leak are fussiness, high heart rate, fast breathing, or a fever. Older children may complain of chest pain. Treatment for a leak may be needed but many heal on their own.

Aspiration: Aspiration is when your child breathes liquid or food into the lungs while eating. Symptoms include coughing/choking or difficulty breathing/color changes with feeding. The management depends upon identifying and treating the underlying cause. Children may temporarily not be able to eat by mouth or may need their diet changed by thickening liquids to minimize the risk.

Late/At-home Complications:

Gastroesophageal Reflux Disease (GERD): Almost every child with EA has problems with acid reflux or GERD since the esophagus does not squeeze normally. Symptoms include frequent vomiting or spit-ups, fussiness or arching of the back while feeding, coughing while lying down, a sour taste in the mouth, tooth decay (cavities), and breathing problems. Treatment is with medications to prevent damage from the stomach acid.

Esophageal stricture: A stricture is a narrowing from scar tissue where the esophagus ends are connected which is a common complication after this surgery. Signs of a stricture include choking or having a hard time swallowing food, vomiting (spitting up) undigested foods, coughing while eating, and food getting stuck in the esophagus. Many strictures are treated with EGD and gentle stretching of the narrowed area but some may need surgery.

Feeding Aversion and Growth Issues: Some children may refuse to eat because they have not been able to eat. Many children will eventually learn to eat and drink everything by mouth over time. Some may need help from occupational therapists and feeding specialists. Children may be followed by a registered dietician or nutritionist to ensure normal growth and development.

Esophageal Dysmotility: Children may have esophageal dysmotility, or decreased contractions (squeezing) in the esophagus. This makes it difficult for the esophagus to propel food into the stomach. This problem may become more obvious when your child starts to eat solid food. Taking frequent sips of water after eating bites of food can help to push or "wash" the food down into the stomach. Further testing can be done to look for dysmotility and medications may be used for treatment.

Recurrent TEF: Sometimes after healing from surgery, the TEF can come back (recurrence). Symptoms are similar to aspiration (above). A recurrent TEF might be seen with an esophogram or a direct laryngoscopy bronchoscopy (DLB) or bronchoscopy, done under anesthesia, which uses thin tubes with cameras to check the airway for a recurrent TEF. A recurrent TEF would require another surgery to disconnect the esophagus from the trachea again.

"When can my child go home and what do I need to know?"

The time your child spends in the hospital may range from a few weeks to months after surgery depending on whether your child was born prematurely, if your child's esophagus was connected in one surgery, your child's lung health, and whether or not your child has other medical problems. Before discharge you will receive specific instructions on feeding, medications, and how to care for the surgical wound. You will also be taught how to recognize signs of complications and when to call the pediatrician or surgeon with concerns.

"How will EA-TEF affect my child's life long-term?"

With the help of their family and healthcare team, most children born with EA-TEF can recover well and live normal lives. Feeding and breathing problems are common in early childhood but can be managed with medical care. Once your child becomes a teenager, the healthcare team will help guide your family in the transition of care to adult doctors. Many patients benefit from ongoing routine monitoring by a Gastroenterologist for management of gastroesophageal reflux and related symptoms. Consistent long-term follow-up will ensure the best health possible.

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