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Esophageal Atresia and Tracheo-Esophageal Fistula (EA and TEF)

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

Overview - “What is it?”

The esophagus is the muscular tube that allows food and liquid to pass from the mouth to the stomach. In esophageal atresia (EA), the esophagus does not form normally; instead, the upper part ends blindly in the lower neck or upper chest and is therefore blocked. There are different types of EA that may be associated with a tracheo-esophageal fistula (TEF)

Most of the time, EA is associated with a TEF, a condition where the lower part of the esophagus has an abnormal connection (fistula) to the trachea or windpipe, allowing stomach acid to enter the lungs. Other, less common forms include EA without a TEF (also known as “isolated esophageal atresia”) or “long gap esophageal atresia” or a TEF without an EA (also known as “H type fistula”).

The condition is rare, occurring in roughly one infant for every 4,000 births in the USA. More than half of 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, see below). For parents who have a child with EA/TEF, a subsequent child will have a higher risk than the general population of having EA/TEF. Having a parent with EA/TEF increases the likelihood of the offspring having EA/TEF.

Signs and Symptoms - “What symptoms will my child have?”

While developing in the uterus, an infant with EA/TEF may have too much amniotic fluid (polyhydramnios) surrounding it. After the birth, the first clues are often excess saliva at the mouth (not able to handle saliva) and choking and coughing with attempts to feed.

Diagnosis - “What tests are done to find out what my child has?”

If EA/TEF is suspected, an attempt is made to place a tube from the nose or mouth to the stomach. If there is resistance to passage of the tube, it may mean that there is a blockage in the esophagus. **Chest X-rays** show the tube coiled in the esophagus.

It is important to find out if the baby has abnormalities commonly found in patients with EA/TEF, so nothing is missed that can impact the baby’s health moving forward. The most common abnormalities are grouped into the “**VACTERL**” association (named after the first letter of the most common anomalies).

- **V**ertebral (spine): spine X-ray to look for abnormalities of the bones and ribs. Spine ultrasound or MRI may be obtained
- **A**nus: Baby has high imperforate anus/cloaca
- **C**ardiac (heart): An ultrasound of the heart (echocardiogram) is needed to check for problems such as abnormal holes, problems with valves, etc.
- **T**racheo-Esophageal fistula (TEF): Abnormal connection between airway (trachea) and esophagus (tube that connects mouth to stomach) and a blind-ending esophagus in neck.
- **R**enal (kidney): Ultrasound of kidneys to look for abnormalities
- **L**imbs (arms and legs): Examine arms and legs for deformity. Arms most common place for abnormal bones.

Ultrasound or MRI of the spine to look at the location of the spinal cord.

Cardiac echocardiogram: Will show whether the aorta (largest blood vessel from the heart) lies on the right or left side of the chest. Usually, the surgery is done on the side opposite to where the aorta is located.

Treatment - “What will be done to make my child better?”

Medicine: No medication can make this situation better.

Surgery: Surgery is the only option for this condition. The goal of surgery is to cut the abnormal connection (fistula) between the lower part of the esophagus and the trachea, close the hole in the trachea, and connect the upper and lower ends of the esophagus so that food can pass from the mouth to the stomach.

- The operation is done through a large incision between the ribs of the chest (thoracotomy), or thoracoscopy (surgery with a small video camera) where a few small cuts are made in the chest. For this thoracoscopy way of doing the surgery, small instruments are placed through the other small cuts to do the operation. The usual

number of incisions (cuts) for thoracoscopic surgery vary. Since the aorta is more commonly on the left side, the surgery is usually done on the right chest.

- When the two ends of the esophagus are too far to bring together in a single setting, a variety of techniques may be employed to accomplish the connection in a staged fashion and at a time when the baby is bigger. When these staged repairs are needed, a feeding tube is placed into the stomach (gastrostomy or G tube) to allow the infant to be fed directly into the stomach before the esophagus is reconnected. Rarely, attempts to connect the ends of the esophagus are abandoned and the esophagus is “replaced” by using a segment of large intestine, or the stomach is pulled up into the chest to be connected to the upper end of the esophagus. In these extreme cases, life-long difficulties with normal eating should be expected.
- In the rare cases where the esophagus is not blocked but has a connection to the trachea (“H-type” TEF), the operation is usually approached through the right side of the neck to divide the abnormal connection between the trachea and the esophagus.

Preoperative care: Studies to check the heart and kidneys are performed to make sure that the baby is ready for anesthesia and surgery. Antibiotics are given prior to starting the surgery to prevent infection.

Postoperative care: The child may need support for breathing with a breathing tube and a ventilator. They may have a drain (chest tube) to drain air and fluid that collect after surgery. A few days after surgery, antibiotics may be given and the baby will undergo a swallow study to check that the connection between the upper and lower parts of the esophagus is healed and is not leaking.

Benefits of surgery: Surgery will establish the continuity of the esophagus, allowing the food that the baby eats by mouth go through the esophagus into the stomach. Additionally, abnormal connections between esophagus and the windpipe are cut off, protecting the airway from stomach contents and/or swallowed food.

Risks of surgery: After the esophagus is reconnected, several later complications may occur. These include leakage at the connection or narrowing (stricture) due to scar tissue. Leakage usually will close on its own when given extra time.

Home Care - “What do I need to do once my child goes home?”

- After an infant is discharged home, the parents need to be aware of the symptoms that occur when certain situations happen that are common in TEF/EA patients.
- Narrowing at the area where the upper esophagus is sewn to the lower esophagus happens quite a bit. The baby may have problems swallowing, coughing and sputtering as the milk or formula gets caught up in the narrowed point (structure). A few weeks after surgery, it will be stretched out to allow for easier passage of food.

- Frequently, reflux of stomach acid into the esophagus (GERD) occurs. GERD may need to be treated with medication, or in severe cases, with surgery to prevent the acid from doing damage to the esophagus or lungs.
- When a child is born with EA/TEF, the esophagus will not have the normal ability to propel food into the stomach (poor motility) making certain foods difficult to eat. This problem may become more obvious when the baby starts eating solids.
- Also, the trachea may be floppy and collapse on itself with vigorous crying (tracheomalacia). This can inhibit air from moving in and out of the lungs. In severe cases of tracheomalacia, an operation may need to be done to keep the trachea from collapsing on itself.
- In addition, the connection between the trachea and the esophagus can reopen over time (recurrent TEF) and require another operation to disconnect them again.

Long Term Outcomes - “Are there future conditions to worry about?”

Your surgeon will let you know the specifics of the long-term conditions your child will need to be followed for, depending on their type of EA/TEF. Many children will be able to eat and swallow and have no problems with this. Some children may have problems with the motility of their esophagus (how the esophagus moves to push food into the stomach) which can last into their adult years.

In general, your child will need to be followed by their surgeon for reflux, and for changes to the esophagus that may require medications or other procedures/surgeries. Children born with EA/TEF can develop chronic irritation of the esophagus called esophagitis. There is also a long-term risk of developing cancer of the esophagus. Once your child becomes a teenager, their surgeon will help guide your family in the transition of care to adult doctors and surgeons, to continue providing thorough follow-up care.

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Author: John C. Bleacher, MD

Editors: Patricia Lange, MD; Marjorie J. Arca, MD; Janice Taylor, MD