Tracheoesophageal Fistula and Esophageal Atresia

What is tracheoesophageal fistula?
The word fistula means “abnormal connection.” Tracheoesophageal fistula (TEF) is a condition that occurs when a baby’s trachea (windpipe) and esophagus (food tube) are connected when they are born. The trachea and esophagus connect for a time during pregnancy but then separate. We do not know why they stay connected in some babies.

What is esophageal atresia?
Esophageal atresia (EA) occurs when the esophagus, the tube that connects the mouth to the stomach, does not completely form during pregnancy. Usually, this means the tube leading down from the mouth and the tube leading up from the stomach do not meet. Instead, each section of tube has a closed end (atresia). When this happens, there is no way for food to travel from the mouth to the stomach.
Do children with TEF/EA have other health problems?

About half of all children born with TEF/EA have other health problems. This is called the VACTERL association. Each letter stands for a problem that may occur with TEF/EA:

- **V** = vertebral, problems with the spine
- **A** = anal atresia, a problem with the way a baby’s anus or rectum has formed
- **C** = cardiac, heart problems
- **TE** = tracheoesophageal fistula
- **R** = renal, or kidney problems
- **L** = limb, problems with arms and legs

If your child has TEF/EA, they will need a series of tests, to find out if they have any of these other problems.

How can you tell if my baby has TEF or EA?

Some babies are diagnosed during pregnancy (prenatally). The most common sign of TEF after birth is severe coughing or choking when your baby tries to swallow. Some babies may have a hard time breathing (respiratory distress) from the stomach contents getting into the airways (aspiration). Esophageal atresia may be suspected at birth if your baby is not swallowing their saliva, but instead is drooling a lot or has frothy mucus in the mouth or nose. We will test your baby by placing a tube down their throat. If we cannot pass the tube from your baby’s nose or mouth into the stomach this usually confirms that they have EA.

We will give your baby more tests to see if there are other problems. They may include:

- X-rays of your baby’s spine and belly (abdomen).
- An echocardiogram (eh-ko-KAR-dee-uh-gram). This test uses sound waves to create a picture of the heart that we can show on a monitor. This is called an “Echo.”
- Ultrasound of the kidneys.
- Physical exam to look for any problems with your baby’s arms, legs and anus.

If an X-ray of your baby’s belly shows air in their stomach, this confirms TEF as well as EA.

What is the treatment for TEF/EA?

Surgery is the only treatment for TEF/EA. The type of surgery babies have depends on their condition. In many cases, surgeons correct the problem with one surgery. Sometimes doctors need to correct the problem in stages. Your baby’s surgeon will go over the possibilities with you.

If your baby has TEF/EA, we will close the fistula, the abnormal connection between the trachea and esophagus, thru a cut usually on side of their chest.
If the ends of the esophagus are close enough to each other, the surgeon will repair the tube by sewing the ends together. If the two ends of the esophagus are too far apart, the doctor may wait to reconnect the esophagus until they grow longer. The wait may be up to 12 weeks or more. In this case, we will put a feeding tube (called a gastrostomy or g-tube) into your child’s stomach to provide their feedings. In some children, the ends of the esophagus are very far apart and the esophagus may need to be reconstructed using the stomach, colon or small bowel. This is a much more complex surgery and your surgeon will discuss it with you if needed.

**What can I expect after surgery?**

After surgery for combined TEF and EA, your baby may need a machine to help with breathing (ventilator). They may need to use the ventilator for several days after surgery until they have healed enough to breathe on their own. We will care for your baby in our Neonatal Intensive Care Unit (NICU) after the surgery.

**Feeding your baby**

Your baby will not be able to eat for a few days after surgery. They will get their nutrition through an IV (tube in a vein).

About 7 days after your baby’s surgery, we will have your baby swallow a liquid called barium and take X-rays (barium swallow test). This helps us check for leaks at the spot where we repaired the esophagus. If there are no leaks, your baby can be fed through their mouth. For most babies getting used to oral feedings takes time, often days, weeks or even months.

If your baby was on tube feedings before surgery, they may still need to use the g-tube until they learn how to eat by mouth for the first time. Sometimes babies will go home taking some of their feedings by mouth and some by a feeding tube. We will remove their feeding tube once your baby is taking all their feedings by mouth. Your surgeon will follow your baby in the hospital and then in clinic to help make these decisions.

**Pain and comfort**

We partner with you to prevent and relieve pain as much as possible. You know your child best. We encourage you to take an active part in your child’s recovery by talking with your care team about options for your child. After surgery, your child is likely to have some pain and discomfort. Your child will have an IV after surgery through which the nurses can give pain medicines to help keep your child comfortable. We will work with you to create a plan that, in addition to medicine given for pain while in the hospital, encourages coping activities to treat pain and provide support. No matter the level of your child’s pain, we join you to assess and respond right away. Help your child get better, faster with good pain management.
Will my child have any problems as they get older?

Children with TEF/EA may have more problems eating than other children. Your child may cough, gag or choke if they try to eat too quickly, if they do not chew their food thoroughly, or if their food is not cut up into small pieces. Your child may have to eat more slowly so that they can chew thoroughly. Avoid giving your child foods that they cannot chew well such as peanuts and other small hard foods. They can have them once your child is old enough to reliably chew them. Here are the common problems that may happen with TEF/EA, most are related to feeding.

**Dysmotility (slow movement of food):** Normally after food is swallowed, wave-like muscle movements (contractions) in the esophagus help move the food to the stomach. If your child had EA, these contractions may be weak, so that food moves more slowly or in the wrong direction.

**Reflux (Gastroesophageal reflux, GER):** GER is a condition where acidic stomach contents come back up into the baby’s throat or mouth. It may be caused by the weak contractions in the esophagus. The acidic stomach contents irritate the throat and cause a wheeze-like reaction in the lungs. Positioning, diet, and medicines will usually help to decrease reflux. Anti-reflux surgery may be needed if the GER is severe.

**Strictures (slight narrowing of the food tube from scar tissue):** Many children with EA will have slight narrowing in the esophagus where the two ends were connected during surgery. If the scar area becomes too tight, liquids and solids cannot pass through. If this happens, your doctor may need to insert a tube down your child’s throat to gently stretch this area. This is called balloon dilation. Some children will need many dilations early in life, but most outgrow the need for it.

**Impacted foreign bodies (stuck food):** Because a child with EA may have strictures and/or dysmotility, large pieces of food or other objects that are swallowed may get stuck in the food tube and need to be removed by the doctor. This usually happens when a child starts to eat new foods and textures. You will learn which foods and textures are best for your child and which ones to avoid. Eating small bites of food, chewing thoroughly and drinking plenty of liquid is always a good idea.

**Tracheomalacia (soft trachea):** The trachea (windpipe) may be slightly soft and this may cause your child to have a “barky” cough. Some children also have more trouble with colds, respiratory infections, and wheezing. Most children will outgrow this problem as cartilage develops. Some may need treatment later on, including surgery, if it is severe.

**Anastomotic leak:** Some children develop a leak where the esophagus was surgically reconnected (the anastomosis). Depending on how bad the leak is, they may be able to be treated with or without surgery. This would only happen during the first 1-2 weeks after surgery and would not be an issue once you and your baby went home. Surgeons closely monitor your baby for this problem as the esophagus is healing.
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**To Learn More**
- General Surgery 206-987-2794
- Hospital operator for after-hours 206-987-2000
- Ask your child’s nurse or doctor
- www.seattlechildrens.org

**Free Interpreter Services**
- In the hospital, ask your child’s nurse.
- From outside the hospital, call the toll-free Family Interpreting Line 1-866-583-1527. Tell the interpreter the name or extension you need.

**Recurrent Fistula:** Sometimes the fistula can return or there can be a second fistula that was not seen at the time of the first surgery. Children with this usually have coughing when drinking liquids. If your child has this symptom tell your surgeon and they will do tests to see if your child has this problem.

**Laryngotracheal cleft:** This problem occurs in the womb and is related to the development of TEF. It is an abnormal connection between the trachea and esophagus but higher up than the usual TEF. Children with this also have coughing or gagging when drinking liquids. A test, called a bronchoscopy, is done to find out if your child has this.

**How common are TEF and EA**
TEF and EA typically occur together, but sometimes one or the other occurs alone. About 1 in 4,000 children is born each year with TEF and/or EA. There are 5 different types of TEF/EA. In the most common type, the upper part of the esophagus has a closed end and the lower part of the esophagus connects to the trachea (see the illustration of esophageal atresia with distal tracheoesophageal fistula on page 1). About 85% of children with TEF/EA have this type.

**Where can I get more information about EA/TEF?**
- Ask your doctor or nurse about support groups, newsletters, or other written information.
- Ask your dietitian about feeding concerns.

Seattle Children’s offers interpreter services for Deaf, hard of hearing or non-English speaking patients, family members and legal representatives free of charge. Seattle Children’s will make this information available in alternate formats upon request. Call the Family Resource Center at 206-987-2201.

This handout has been reviewed by clinical staff at Seattle Children’s. However, your child’s needs are unique. Before you act or rely upon this information, please talk with your child’s healthcare provider.

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