

PATIENT AND FAMILY INFORMATION SHEET

Esophageal Atresia/Tracheoesophageal Fistula

What is Esophageal Atresia (EA)?

- The esophagus is a tube that connects the mouth/throat to the stomach.
- When a baby is forming in the womb, the esophagus does not form properly and it becomes separated into two parts. The upper part connects the mouth/throat to a blind (closed) end or pouch (proximal end) and the lower part connects the stomach to a blind end or pouch (distal end).
- Because the two ends do not connect to each other, feedings (breast milk or formula) and saliva cannot move from the mouth to the stomach.

What is Tracheoesophageal Fistula (TEF)?

- The trachea is also known as the windpipe. It is a tube in the chest that connects the throat to the lungs.
- When a baby is forming in the womb, the trachea and esophagus start out as one single tube.
- Around 4-8 weeks of your baby's development a wall forms in this tube and divides it into two separate tubes . They are no longer connected.
- If the trachea and esophagus don't separate into two tubes, a tracheoesophageal fistula (TEF) can occur. This means that there is an abnormal connection in one or more places between the esophagus and the trachea. There are many types of this defect (see pictures below).
- A baby usually has both conditions of TEF and EA, but sometimes the baby has one or the other.
- If a baby with TEF tries to swallow feedings or saliva the liquid can go into the lungs between one of the connections through the esophagus and trachea. This could cause lung infections.

How do I know if my baby has TEF/EA?

- Sometimes, TEF/EA is diagnosed by an ultrasound before the baby is born.
- After the baby is born, some signs and symptoms of TEF/EA are:
 - Coughing or choking; especially when feeding
 - Frothy, white bubbles from the mouth
 - Vomiting
 - Difficulty breathing
 - Very round, full belly
 - A tube will not pass from the baby's mouth to the stomach
- If there is concern that your baby has TEF/EA, a full physical examination along with X-rays of the chest and the belly will be done.

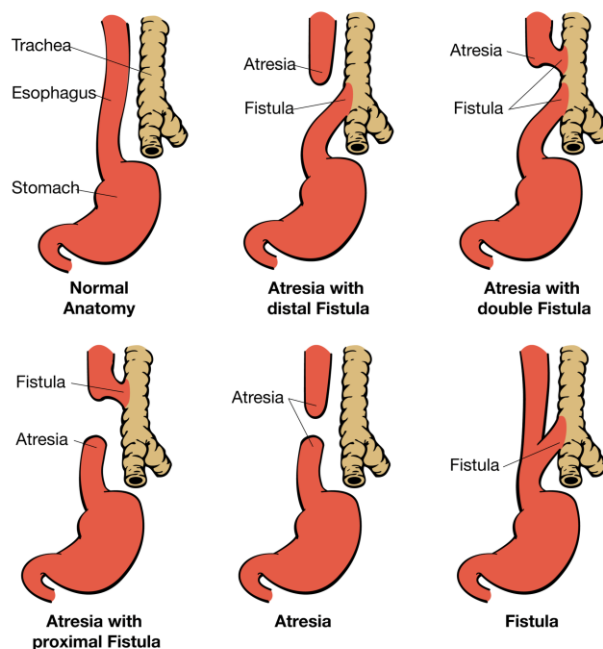


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Are there associated conditions/defects?

TEF/EA is not thought to be passed down within the family.

There are other birth defects that can be seen in babies who have TEF/EA.

- Chromosomal defects such as Trisomy 13, 18 or 21 (Down syndrome)
- **VACTERL** association is a group of 1 or more abnormalities that can be seen together in up to 50% of children with EA/TEF.
 - **V** – vertebrae (spine) defects
 - **A** – Anal defects (imperforate anus most common)
 - **C** – Cardiac defects (ventricular septal defect most common)
 - **T** – Tracheoesophageal fistula
 - **E** – Esophageal atresia
 - **R** – Renal (kidney) defects (horseshoe, missing, polycystic)
 - **L** – Limb defects [missing bone(s) in the legs or arms]
- **VACTERL** defects are often diagnosed with a full physical examination, x-rays, and/or ultrasound. They do not require painful testing.

How is TEF/EA treated?

- The treatment for TEF/EA is surgery.

What happens before surgery?

- Your baby will be in the Neonatal Intensive Care Unit (NICU) before and after surgery.
- If your baby is premature, they will be given all the support needed for their prematurity.
- Your baby will be given breathing support if needed.
- Intravenous fluids will be given for nutrition and hydration.
- A tube will be placed through your baby's nose or mouth to help suction saliva from the upper part of the esophagus.
- Some or all of the tests for the VACTERL association will be done at your baby's bedside.
- Blood will be drawn for blood type, electrolytes and for signs of infection.
- Depending on how your baby is doing, antibiotics may be given.

What happens during surgery?

- First, your baby will be given a breathing tube (if they don't already have one) and they will be put to sleep by the Anesthesiologist.
- All surgery is done through an opening in the side of your baby's chest. Depending on the type of surgery, this will either be one long opening or several small openings. The type of incision is decided by your surgeon.
- A feeding tube or gastrostomy tube (g-tube) may be placed in your baby's stomach to use to feed the baby after surgery.
- A chest tube (drain) will stay in place until the drainage has stopped. Before removing this tube, a special x-ray study will be done to make sure there is not a leak between the two ends that were sewn together.



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When can my baby eat again after surgery?

- Your baby may not be able to eat for the first week after surgery.
- When feedings begin, the surgeon may only allow feeds through a feeding tube. The tube will either be placed in the nose during the operation (nasogastric or NG tube) or directly into the stomach (g-tube)
- When your baby is allowed to eat by mouth, they may have difficulty because they have not eaten by mouth before.
- During this time, feeding can be given through the feeding tube and s/he can practice with smaller volume feedings by mouth.
- Eating everything by mouth by bottle or breastfeeding can take a while in the baby with TEF/EA. Teaching them to feed by mouth takes patience and practice.

Can I breast feed after surgery?

- Breastmilk is best for your baby so we will work with you to help you be able to breastfeed once it is safe for your baby.
- Until you can breastfeed your baby, please pump your breast milk and store it in the freezer. It will be used when your baby starts receiving milk through the feeding tube.

What are the complications after surgery?

- A leak of saliva where the two sections were sewn together can occur. A chest tube may need to be placed to collect the drainage. It will take time for the leak to stop and the area to fully heal.
- Gastroesophageal reflux disease (GERD) is very common after TEF/EA surgery. GERD is a condition where feedings may back up from the stomach into the esophagus. This is treated with medicine.
- The newly connected esophagus will not function perfectly after it is reconnected. The passage of liquid and food may be slower compared to a baby that didn't have this surgery. As your baby grows and starts to eat table food, your surgeon will tell you which foods your baby may have difficulty swallowing.
- As the connection heals, scar tissue will form. If the scar tissue gets too tight, it can cause a narrowing (stricture). This narrowing may make it more difficult for your baby to swallow food. This stricture can be treated by widening or dilating it.
- Tracheomalacia is a common complication after surgery. Tracheomalacia is a softening of the cartilage of the windpipe or a "soft spot" on the windpipe that can occur especially at the location where the TE fistula was located.
 - Due to tracheomalacia, your baby's cry or cough may sound like a "barking" sound similar to a seal.

When can we go home?

- This is a difficult question to answer and each baby with TEF/EA is different.
- The answer depends on many things including: if your baby was premature or not, if the esophagus could be connected right away, the breathing status of the baby, and also if there are other medical problems identified.
- If the baby was born early, you can expect them to be in the hospital for several weeks or months
- If the baby was closer to full term, the esophagus may be connected right away and if there wasn't a leak, you can expect to be in the hospital for a few weeks.



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How do I care for my child at home?

- You will be taught prior to discharge how to
 - Feed your baby if s/he has a feeding tube.
 - Give the baby medications, if necessary.
 - Use the proper recipe to mix formula or breast milk if your baby needs more calories.
 - Care for the incision and dressing, if still applicable.
 - Learn signs and symptoms of infection.
 - Learn signs and symptoms of narrowing where the esophagus was put back together.
 - When to call your pediatrician with concerns or problems.
 - When to call your surgeon with concerns or problems.
- You may also have home nursing visits, early intervention, speech therapy, physical therapy, and/or occupational therapy once you are discharged to home.
- Ask when the site of the surgery can get wet and when you can give your baby a tub bath.

When should I call the office?

If your baby experiences any of the following, please call our office:

- Temperature greater than 100.3°F (38.0°C).
- Frequent vomiting or diarrhea, especially if the vomit is bright yellow or green.
- Difficulty breathing, breathing that is harder or faster than normal for your baby.
- If the lips or skin around the mouth turns blue in color.
- If the “barking” cough increases or gets worse.
- If there is increased drooling. This could be a sign that the esophagus is becoming narrow.
- If there is choking or vomiting after feeding when feeding by mouth – may need a dilation.
- If you had to call 911 or take your child to the emergency room for a choking episode.
- If you need a prescription refill for the reflux medication(s).

Your child will need to follow up with the surgeon.

You will receive specific instructions for follow up when your child is discharged home.

Please don't hesitate to call our office if you have any problems or concerns.

Surgical provider: _____

Office Number: _____

After hours number, if applicable: _____

Thank you for allowing us to care for your child.