Introduction

Esophageal atresia and tracheoesophageal fistula (EA/TEF) are congenital defects in which the esophagus (the tube that carries food and liquid from the mouth to the stomach) and trachea (windpipe) fail to develop properly. This defect occurs during the first 4 weeks of fetal life when the esophagus and trachea are forming. There is currently no known specific cause for EA/TEF, and it is not believed to be inherited.

There are at least 5 types of esophageal and tracheal abnormalities that can occur. All of these prevent your baby from feeding safely at birth and can lead to additional gastrointestinal and respiratory complications.

Esophageal Atresia

In most cases of esophageal atresia, the formation of the esophageal tube is interrupted creating two separate tubes: an upper (proximal) tube connected to the mouth and a lower (distal) tube connected to the stomach. These separated tubes are sealed off creating a blind pouch on either side; the gap between these pouches can be short or very long. Saliva can accumulate in the upper pouch as it cannot drain into the stomach.

Tracheoesophageal Fistula

Newborns with esophageal atresia often also have a tracheoesophageal fistula which is a direct connection between the esophagus and the trachea (windpipe). Usually this fistula connection forms off of the lower pouch; however, it can also form off of the upper pouch or a connection can be formed with both pouches, and in some cases, a fistula will form without atresia. The tracheoesophageal fistula can lead to breathing problems (aspiration pneumonia) due to saliva draining from the upper pouch into the trachea, or stomach contents may enter the trachea from the lower pouch.
Common Types of EA/TEF

Diagnosis

Esophageal atresia and tracheoesophageal fistula is difficult to detect before birth. The most common ultrasound findings are polyhydramnios (an increased volume of amniotic fluid) and a small or absent stomach; however these signs may also be present for reasons other than esophageal atresia.

If no diagnosis is made before birth, EA/TEF is soon recognized after birth as most newborns have feeding problems right away. Often these newborns will have coughing and choking with feedings, and sometimes with accumulation of saliva. Any attempt to use a feeding tube is stopped by the atresia of the esophagus.

Typically, the final diagnosis is then made by X-ray. The type of esophageal atresia and the distance between the esophageal ends will be evaluated and explained to you by your baby's surgeon.
Also, TOFS: Tracheo-Oesophageal Fistula Support is another organization created to support families of children with EA/TEF

• http://www.tofs.org.uk
• info@tofs.org.uk

Common medications

Ranitidine/Zantac
• This medicine is used to treat gastroesophageal reflux disease, heartburn, acid indigestion, and sour stomach.
• Ranitidine prevents symptoms and damage to the gastrointestinal tract caused by stomach acid or infection.

Lansoprazole/Prevacid
• This medicine is used to prevent or treat gastrointestinal ulcers or ulcers of the esophagus.
• This medicine is used to treat gastroesophageal reflux disease and heartburn.
• Lansoprazole prevents symptoms and damage to the gastrointestinal tract caused by stomach acid or infection.

Omeprazole/Prilosec
• This medicine is used to prevent or treat gastrointestinal ulcers or ulcers of the esophagus.
• This medicine is used to treat gastroesophageal reflux disease and heartburn.
• Omeprazole prevents symptoms and damage to the gastrointestinal tract caused by stomach acid or infection.

Metoclopramide/Reglan
• This medicine is used to prevent or treat nausea and vomiting.
• This medicine is used to treat gastroesophageal reflux disease.
• This medicine may improve feeding problems and spitting up.
• Metoclopramide increases gastric emptying of food and decreases symptoms of nausea, vomiting, heartburn, and fullness.

Associated birth defects
In some cases, newborns with EA/TEF may have one or more associated congenital abnormalities including:
• Gastrointestinal defects (i.e., malrotation, imperforate anus, etc.)
• Cardiac defects
• Urinary system defects
• Limb development abnormalities
• Chest wall and spine abnormalities
Some of these defects may be diagnosed at birth and others may not become apparent until later. Chest wall and spine abnormalities may develop as the child grows. We will follow your child in our outpatient office to monitor for these changes and make recommendations, as needed.

How is EA/TEF repaired?
Your baby will be cared for in an Intensive Care Nursery by specially trained doctors and nurses who will explain the type of defect your infant has and the type of surgical repair required. If a tracheoesophageal fistula is present, it must be surgically closed soon after birth. During fistula closure, if the gap between the two ends of the esophagus is small, they will be sewn together and the esophageal atresia will be repaired. This is called a primary repair and can be often done in a single operation; however, complicated defects may need to be completed in stages during two operations. For example, if the upper and lower pouches are too far apart to bring together, only the fistula will be closed and the repair of the esophagus may be delayed. If the gap is very wide, an alternative method of surgical repair of esophageal atresia may be offered.

At the time of surgery, some newborns will have a gastrostomy (or feeding tube) placed through the abdomen into the stomach or a feeding tube placed from the mouth or nose to the stomach while the esophagus heals.

Post operative radiology studies (contrast esophagram)
Several days after surgery a special x-ray, called a contrast esophagram, will be performed to determine if the esophageal repair has healed. The x-ray is used to determine if there are any leaks around the site of the repair; these leaks indicate that the repair still needs time to heal. If leaks are visible, X-ray studies will be repeated every several days and if a chest tube was place by the surgeon at the time if the operation, it will remain in place until the esophageal repair has completely healed.

Feeding and nutrition
The team in the Intensive Care Nursery will help design a feeding program to meet your baby's needs. These feeding guidelines will be reviewed with you before you take your baby home.

Feeding tubes
If your baby has a feeding tube at the time of the operation it will be used to deliver feedings directly
into the stomach while the esophagus heals. Some children, with more complicated defects, will require a tube placed into the stomach through the abdomen. This gastrostomy tube or G-tube allows feedings to be delivered directly into the stomach. Gastrostomy tubes are usually removed once your child is gaining weight on oral feeding only. The length of time the gastrostomy tube needs to stay in place varies from child to child and can be needed for weeks, months or rarely years.

**Gastrostomy home care & supplies**
If your baby has a gastrostomy tube for feedings, you will be instructed how to give feedings before you take your child home. The discharge planner will order tube feeding supplies, formula and a feeding pump (if needed) from a home care company. Supplies will be sent to your home each month, as long as your child needs tube feedings. Home nursing visits will be ordered if your child has a gastrostomy tube in order to ease the transition from hospital to home.

**Difficulty with swallowing & feeding**
Children with EA/TEF often have difficulty swallowing (dysphagia) due to narrowing or poor motility of the repaired esophagus muscle and acid reflux. As a result, they eat and drink slower than other children. New food textures must be introduced slowly, and solid food should be cut into small pieces or strips to make swallowing easier. While rare, it is possible for food or foreign bodies to become stuck in the esophagus; these may pass on their own or in extreme cases require removal by the surgeon. It is best to allow your child to eat and drink at a pace that is comfortable for them – by observing your child, you will quickly become an expert on your child’s feeding patterns.
Some children experience slow growth if eating is difficult. During your follow up appointments we will weigh and measure your child and you will see our nutritionist for recommendations.

**Esophageal stricture and dilation**
Children may develop a stricture (or narrowing) of the esophagus at the repair area which makes it difficult for children to feed. Strictures are typically noticed due to symptoms that occur after discharge including:
- Appetite reduces more and more at each feeding
- Choking or coughing with feeding
- Gaining weight more slowly than expected
- Taking more time to complete a feeding
If a stricture is suspected, we will schedule your child for an esophagram to look at the contour of the esophagus and evaluate how well the esophagus empties into the stomach. Upon diagnosis of a stricture, the surgeon may recommend a dilation (or stretching) of the esophagus in the operating room. The need for dilation varies, some children never require dilation and some children need several.

**Gastroesophageal reflux disease (GERD)**
Children with esophageal atresia commonly have gastroesophageal reflux disease (GERD), a back flow of contents from the stomach into the esophagus. To prevent this irritation, your baby will be started on an antacid medication and a motility medication to enhance stomach emptying. These medications are continued after hospital discharge for as long as needed, with dosages adjusted by your pediatrician as your child gains weight. Please do not stop these medications without consulting with your child’s surgeon.

**Respiratory complications**
Children with tracheoesophageal fistula commonly have respiratory symptoms including wheezing, cough, bronchitis and less commonly pneumonia. The severity of these respiratory problems vary; children with more severe problems may benefit from pediatric pulmonary care.

**Tracheomalacia**
The trachea may also be affected by a condition known as tracheomalacia, where the trachea is softer and less rigid than normal. This results in breathing problems and a barking cough which gets worse with upper respiratory infection or exertion. The severity of tracheomalacia varies from child to child and usually improves as the child grows older, but may require additional treatment.

**Respiratory syncytial virus (RSV)**
Respiratory syncytial virus is a common cold virus that can make newborns very ill. Children born with EA/TEF may be more sensitive to upper respiratory infections such as respiratory syncytial virus (a common cold virus). An RSV infection can lead to bronchitis or pneumonia and sometimes require hospitalization. In order to prevent this, we recommend that children with EA/TEF receive a monthly vaccine called Synagis® during the first winter of your child’s life.

**Follow up appointments**
Your baby will be seen in the surgery clinic several times the first year and at least every year thereafter or more frequently as needed.

**California Children’s Services (CCS)**
Children with EA/TEF may be eligible to receive insurance coverage through the California Children’s Services, as long as other criteria are met. After delivery, you will be referred to your local CCS county office where you can fill out an application. If your child is enrolled in CCS, regular visits to our CCS paneled surgeon will help maintain your coverage.

**Support resources for your family**
EA/TEF Family Support Connection has a web site to help you learn as much as possible about Esophageal Atresia/Tracheoesophageal Fistula.
- http://www.eatef.org/
- info@eatef.org