ESOPHAGEAL ATRESIA AND TRACHEESOPHAGEAL MALFORMATIONS

ES Boia[1], A Mittal[1]

[1]University of Medicine and Pharmacy "Victor Babes" Timisoara

Abstract

Tracheesophageal fistula (TEF) and esophageal atresia (EA) are surgical emergency, presenting during first moments after birth. They are usually associated with other congenital anomalies. They are usually complicated due to aspiration of gastric contents leading to pneumonia and respiratory distress. The anatomy end embryology of the esophagus, clinical manifestations, diagnosis and management of these conditions are presented in this paper work.

Key words: tracheesophageal fistula, esophageal atresia.

ANATOMY OF ESOPHAGUS

Layman calls it *food pipe*, an organ responsible for delivering food to stomach. It is a tabular structure with diameter of 2.5 cm and length 25 cm. Hence, it is important to note that whenever we want to instrumentise esophagus e.g. for nasogastric feeding or decompression, that its upper end lies at distance of 15 cm from incisors and lower end of esophagus lies at 40 cm.

Like rest of gut, esophagus also has the capacity to do peristalsis. Peristalsis is actually responsible for delivering food bolus to stomach.

According to the region, esophagus traverse is divided into:

- cervical esophagus
- thoracic esophagus
- abdominal esophagus

- Cervical esophagus starts from lower end of oropharynx, at the level of C_6.
- Thoracic esophagus lies in the mediastinum. At the level of T10, thoracic esophagus crosses diaphragm, a strong muscular layer separating thorax and abdomen.
- Abdominal esophagus at the level of T_{11} enters into stomach, forming a very sharp angle called cardiac angle.

Arterial supply of esophagus

It is very important to know the blood supply of esophagus, when we want to operate on esophagus. It has got complex blood supply according to anatomical division of esophagus from various arteries:

o *cervical esophagus:* Receives its blood supply from inferior thyroid artery

o *thoracic esophagus:* Receives its blood supply from branches of thoracic aorta and branchial artery

o *abdominal esophagus:* Receives its blood supply from left gastric artery and left inferior phrenic br. of abdominal aorta

Venous drainage of esophagus

Like arteries different veins drain esophagus:

o *cervical esophagus*: drained by inferior thyroid vein

o *thoracic esophagus*: drained by azygous vein, hemiazygous vein and accessory hemiazygous vein

o *abdominal esophagus*: drained by azygous vein and left gastric vein

Lymphatics of esophagus

o *cervical esophagus*: drained into deep cervical and paratracheal nodes

o *thoracic esophagus*: drained into mediastinal nodes

o *abdominal esophagus*: drained into nodes in relation with left gastric artery

Nervous supply of esophagus

Esophagus is supplied by autonomic nervous system both sympathetic and parasympathetic nerves.

o sympathetic nerves reaches it through splanchnic branches of sympathetic trunk

o parasympathetic nerves reaches it through vagus plexus around it.

EMBRYOLOGY OF ESOPHAGUS

Human fetus has been divided into *foregut, midgut* and *hindgut*. Esophagus develops from the part of foregut between pharynx and stomach. In early stages of development, it is short but elongates as formation of neck, descent of diaphragm and with enlargement of pleural cavities.

The muscles of esophagus arise from *mesenchyme* surrounding the foregut. Upper two third of esophagus is made up of striated muscles and lower one third is made up of smooth muscles.

Whole respiratory system develops from *median diverticulum* of foregut. That's why, its lining epithelium is derived from *endoderm* and connective tissue, cartilage and muscles are derived from *mesoderm*.

Free caudal end of diverticulum becomes *bifid*, each subdivision being called *lung bud*. Part of diverticulum cranial to this lung bud forms *larynx and trachea*, while lung buds form *bronchi and lungs*.

About 19th day of gestation, the foregut of human embryo is represented by a cell-layer tube, which extends from pharynx to stomach. Over several days, the ventral aspect of this foregut begins to thicken and form a groove lined by ciliated, stratified, columnar epithelium which becomes *respiratory mucosa*.

Separation of the dorsal foregut (esophagus) from tracheal ventral part occurs first at the carina and extends in a cephalad direction. By about 26th day of gestation, these two structures become up to level of larynx. Bronchi develop from posterolateral buds of trachea and grow to each side. This process of separation of dorsal and ventral foregut is called *epithelial 'ridge' ingrowth*.

HISTORY OF DISEASE

Thomas Gibson was the first, who in 1696 described an accurate clinical and pathological description of the most common anomaly, in which EA is associated with TEF. At that time this disease was considered as fatal condition, which is the not fatal anymore now days. Major breakthrough occurred in 1941, when American surgeon *Cameron Haight* achieved survival by successful anastomosing two ends of esophagus and thus overcoming obstruction in gastro-intestinal tract.

DEFINITIONS

Atresia: Means absence of a normal opening in hollow tract.

Fistula: Means an abnormal connection between two epitheliased structures in body.

Esophageal Atresia: This is a congenital disorder, when proximal and distal portions of Esophagus do not communicate. The proximal blind pouch has thick musculature and bigger diameter, while the distal portions have thin musculature. The proximal pouch usually lies at the level of T_2-T_4, but can be as short as up to C_7 and longer up to T_5.

Tracheesophageal Fistula: This is also a congenital disorder with abnormal disconnection between anterior esophagus to posterior membranous trachea. Occasionally, trachea trifurcates with fistula arising

o

between two primary bronchi. Rarely, fistula connects with bronchi.

CLASSIFICATION

These 2 abnormalities are congenital, that means they are present before birth. They can occur as separate entities but commonly occur together.
(see fig. 1)

o   *EA with distal TEF (84%)*: This is most common type of deformity among all. Here, a gap exists between two Esophageal segments. The length of this gap actually influences the simplicity and difficulty in correction. Occasionally, an overlap of segments and even muscular continuity exists. (fig. 1A)

o   *EA with proximal fistula (1%)*: This is a least common type among all. Here fistula usually arises at 1-4 cm proximal to the tip of upper esophageal pouch. (fig. 1B)

o   *EA with proximal and distal fistula (3%)*: Here in this abnormality there is usually no gap exists between two segments of Esophagus. So very easy to repair. (fig. 1C)

• *Type 3A EA with proximal and distal TEF with gap of >2cm*

• *Type 3B EA with proximal and distal TEF with gap of <2cm*

o   *Isolated EA (without TEF) (8%)*: Here in this abnormality usually a long gap exists between two segments of Esophagus. Stomach is small because no amniotic fluid reaches the stomach in uterus. (fig. 1D)

o   *Isolated TEF (no EA) (4%)*: Fistula is usually 2-4 mm in diameter occurs at any level from Cricoid to Carina but usually arises in lower cervical or upper thoracic area. Double or triple fistula can also occur. "N" type or "H" type indicates integrity of Esophagus. (fig. 1E)

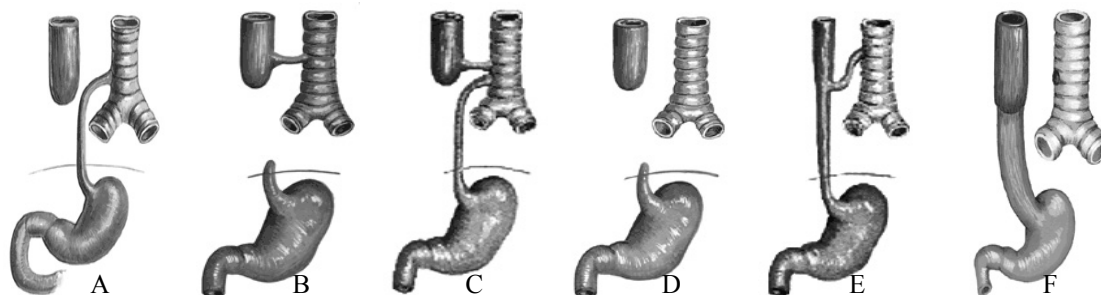o   *Repaired esophageal atresia with distal TEF*: (fig. 1F)

Fig. 1: Esophageal Atresia: A. with a distal tracheo-esophageal fistula, B. with a proximal fistula, C. with fistulae from both esophageal segments, D. isolated esophageal atresia without a fistula, E. TEF of the 'N' or 'H' type, F. repaired esophageal atresia.

Esophagus of all affected infants is deficient in neural tissue in *Auerbach's plexus*. Lower segment is more deficient than upper segment. This is the cause of a*bnormal peristalsis* demonstrated by Barium Swallow. *Manometric studies* have showed that this defect is congenital not result of operation. Discoordinated peristalses have been reported from level of fistula to the stomach in patient with isolated TEF.

ETIOLOGIC FACTORS AND PATHOGENESIS

Even today etiologies of all these congenital anomalies are not known completely but many theories have been postulated by different embryologists. But for parents of children with this anomaly, it is important to understand – it is not their fault and they could not have done anything to prevent it.

This defect may be result of interruption of process called *epithelial 'ridge' ingrowth*.

Smith's Theory of EA: This is the most widely accepted theory for formation of EA. He postulated that 'Lateral Esophageal Grooves' (naturally occurring areas of narrowing much like the ridges of epithelial proliferation that form the septum between Trachea and Esophagus) may turn dorsally and in formation of EA

There exists no convincing evidence of Mendelian Inheritance, however number of reports of multiple family members having EA and TEF exist. Several sets of identical twins having EA and TEF have occurred as well as mother and child and father and child have occurred.

The non random association of VACTERL may be evidence of generalized disturbance in embryogenesis. The exact nature of embryogenic insult that cause EA/TEF is not known but there is some evidence that vascular insufficiency; genetic factors; vitamins deficiency drug and alcohol exposure and viral, chemical and physical external events results EA/TEF.

EPIDEMIOLOGY

a. *Frequency*: 1 in 4000 live births has EA/TEF. Also incidence increases in 1st degree relatives.

b. *Race*: There is no association with any specific race, but in general white population have increased risk of having EA/TEF than non white population

c. *Sex*: Male to female ratio is 1,26.

d. *Age*: Recent studies have shown that EA/TEF increase in incidence with increase in maternal age (> 30 years)

PATHOPHYSIOLOGY

Normally Trachea and Esophagus are entirely separate lumen with no connection in between them. Hence, child can feed properly without any respiratory distress and feeding problems.

- But in case of isolated EA, there is accumulation of saliva and food in proximal pouch. Thus there is increased risk of aspiration of these collections resulting respiratory distress, atelectasis and pneumonia.

- When proximal pouch is associated with Proximal TEF, result definitely into an aspiration of various collections of proximal pouch result into various morbid conditions.

- EA with distal TEF is more fatal condition because this allows two way movements of gastric fluids and gases.

- When there is H/N type or isolated TEF, symptoms are less severe because some of Gases and Fluids went via esophagus to mouth.

- Also because of congenital deficiency of neural tissue in Auerbach's plexus there is in coordination in peristalsis in esophagus of infants with EA/TEF. These are demonstrated by manometric and gastrostomy. They even persist after surgical repair of defect.

- Tracheal and pulmonary development have been hampered in fetus with EA/TEF because:
  o Swallowing of Amniotic Fluid results Dilation of Proximal Esophageal Pouch, this cause increased pressure on Trachea. These results into mal development of cartilage rings called *Tracheomalacia*.
  o Also TEF causes flow of Pulmonary Amniotic Fluid to pass into stomach, this results in decreased branching of Bronchi and Alveoli because of decreased Intra Pulmonary pressure.

CLINICAL PRESENTATION

o excessive salivation
o drooling
o poor feeding
o coughing, choking and gagging associated with attempted feedings
o bluish coloration of skin associated with attempted feeding.
o excessive amniotic fluid during pregnancy (polyhydroamnios)

The first sign of EA in fetus is Maternal Polyhydroamnios which has got broad differential diagnosis:

1. agnathia, microstomia, synotia
2. DiGeorge velocardiofacial syndrome
3. placental insufficiency
4. maternal dm
5. chromosomal disorders, iso-immunologic diseases, congenital abnormality
6. epignathus
7. hydrolethalus
8. tetra logy of fallot, treacher collins syndrome
9. myasthenia gravis, pseudohypoaldosteronism

Also polyhydroamnios is associated with *premature birth* because of increased amniotic pressure. About one-third of infants with EA weigh less than 2500 gm. If there is associated TEF then there is decrease in amniotic fluid amount.

Classically, the neonate with EA presents with copious, fine, white frothy bubble in mouth and sometimes in nose. These secretions may clear with aggressive suction but return. There are episodes of cough, choking and cyanosis, typically exaggerated with attempted feeding.

Cyanosis is a result of laryngospasm (a protective mechanism that body has to prevent aspiration into trachea). Over the time respiratory distress develops.

Abdomen is distended if there is associated TEF and scaphoid if there is isolated EA

Hence, in newborn prematurity, polyhydroamnios and any demonstrable element of VACTERL warrants a more thorough search for EA.

DIAGNOSIS

EA with or without TEF, is a fairly common congenital disorder of a neonate who develops feeding difficulties and respiratory distress in the first few days of life. Early diagnosis is necessary to minimize pulmonary complications.

A. Prenatal diagnosis

o *Prenatal maternal sonography*: Although sonography postnatally has no significance in evaluation of EA with/without TEF, but prenatal maternal sonography is suggestive of these possible conditions.

- *Pouch sign*: This is usually seen as aechoic shadow in middle of fetus during 26th week of gestation. This is confirmatory for EA. But this requires experience.

- *Presence or absence of gastric air bubble* suggests EA with TEF or isolated EA respectively.

- *Polyhydroamnios* is also present.

The prenatal diagnosis of EA is low, unless there is *pouch sign* and *polyhydroamnios* together.

Polyhydroamnios alone is a poor indicator of EA. Only 1 out of 12 patients with polyhydroamnios has EA. Similarly, a small or absent fetal stomach gas bubble has multiple association in addition to EA. Prenatal sonography can also cause many cases of TEF to be missed because amniotic fluid from lungs may pass to stomach and result is normal appearing fluid filled stomach shadow.

o *Prenatal MRI of fetus*: Today in the time of modern diagnostic tools, MRI is important confirmatory diagnostic of EA/TEF prenatal. This allows visualization of entire lesion and anatomic relationships unlike sonography. In modern medicine this is a method of choice. This also allows recognition of other associated congenital anomalies. However fetal MRI is difficult in polyhydroamnios because of poor image quality, also fetal movements cause poor imaging.

This is 100% confirmatory. However doubtful cases should be screened for EA/TEF postnatal.

B. Postnatal diagnosis

o *Nasogastric Intubation of Neonate*: Here we prefer to use 8F in premature and 10-12F intern infants. We perform this technique when we suspect EA either by prenatal sonography or by clinical picture of neonate.

Normally gastric cardia of infant's lies at 17 cm from gums of infant, but in case of EA, tube typically stops at 10-12 cm. This technique is not confirmatory as external compression of esophagus can lead to false positive test. In this case repeat the test. If we are using soft tube, then it may coil inside the pouch and can lead to false negative test. Hence, nasogastric intubation should always be followed by whole chest and abdomen simple radiography.

This is very low for this technique as lots of false positive and false negative test results associated. Also tear of oropharynx or esophagus should be considered especially in patient, who underwent various attempts at feeding tube placement following delivery. Also with this technique we can not differentiate between types of EA/TEF.

o *Radiography*: This is very important diagnostic tool and should always follow nasogastric intubation. Here both PA and lateral views should be taken. Both chest and abdomen should be included in single radiograph for correct diagnosis of EA/TEF with type. With this we can also confirm location of aortic arch, which is important regarding surgical repair of defect. Also we can detect congenital anomalies like cardiac silhouette: boot shape cardiac shadow in tetralogy of Fallot, vertebral anomalies, aspiration pneumonia especially of right upper lobe and patchy atelectasis are frequently present. Localization of aortic arch is important because thoracotomy is always done on the side opposite to aortic arch.

Radiographic signs of right sided aortic arch

A. Right ascending Aorta, indicated by an opaque shadow on right side of mediastinum.

B. Right sided tracheal indentation and/or deviation.

We can also know the position of aorta be passing umbilical artery catheter, by seeing on which side of vertebral column its shadow lays.

Although barium studies are rarely indicated because of high risk of aspiration chemical tracheobronchitis but sometimes to measure the length of gap between proximal and distal esophageal pouch. Here we put contrast material in proximal pouch through mouth and in distal pouch via gastrostomy, and measure length of gap and also helps in visualizing other gastrointestinal abnormalities.

Radiographic findings on children with EA/TEF according to type of anomaly present

a) *Isolated EA*

▪ Dilated air filled blind Proximal Pouch, which often displaces trachea anteriorly

▪ A gasless abdomen may be depicted. Air is normally present in stomach even after 15 min of birth

▪ Lower pouch can be seen by gapogram or air

b) *EA with TEF Distal*

▪ Proximal pouch and gaseous stomach and small bowel (because air passes through fistula)

▪ Images may be airless if TEF is occluded

▪ Excessive air may be present in esophagus, although some air in esophagus is

normal in neonates and children because of aerophagy common in bottle fed infants

c)   *EA with TEF Proximal*
▪   Similar to isolated EA on plain X-Ray
▪   Barium swallow examination may fail to demonstrate this anomaly
▪   Fistula visualization requires rapid sequence or videoflouroscopic studies during cautious filling of proximal pouch

d)   *Isolated TEF*
▪   Recurrent pneumonia may be present with a widespread pneumonia pattern
▪   Fistula delineation is difficult
▪   Excessive air may be present in esophagus

If H-Fistula is strongly suspected on clinical basis, a prone video-esophagogram should be obtained to determine fistula's size and location.

Stringer technique: Patient lying prone on waist height footstep with fluoroscopic table erect and by using cross table flouroscopy, serial injection of isotonic, non ionic contrast agent or dilute barium is administered through a naso-esophageal catheter. As the catheter is withdrawn proximally, tracheal filling is noted at situ of fistula.

Finding of a coiled nasogastric tube at radiographic examination confirms EA. Occasionally; the tube may coil because of external esophageal compression and here repeat placement should be attempted. If the tube passes to stomach, the possibility of the tube passing through a TEF must be investigated. The infant's cry should be profoundly affected if the tube passes through vocal cords on the way through a distal TEF and into stomach.

Special Concerns in Contrast Radiography
A.   Contrast radiography has no special role in diagnosis of EA/TEF, except for localizing TEF.
B.   Barium is best contrast material but can cause aspiration pneumonitis and pulmonary edema.
C.   Extra luminal barium causes granulomatous and fibrotic reaction. Hence fibrous mediastinum.
D.   Aqueous low osmolality agents such as Visipaque and Optiray are preferred. They are expensive but have no deleterious effect on gastro intestinal tract. It is preferred in premature children and neonates with suspected esophageal perforation. They also remain in gastro intestinal tract for long time because of low absorbability (because they are hyperosmolar and hypertonic)
E.   Hypo-osmolarity aqueous agents get quickly diluted and are less fluoroscopically visible because of they have low coating ability.
F.   They also cause hypovolemia, severe dehydration and pneumonitis by significant irritation of trachea and bronchi.
G.   Hence, contrast radiography of neonates with EA/TEF should be done under experienced hands.

o   *Tracheoscopy and esophagoscopy*

They have no diagnostic value but can be used to confirm the position of TEF.

DIFFERENTIAL DIAGNOSIS
o   Esophageal cancer
o   Esophageal diverticula
o   Esophageal rupture
o   Esophageal stricture
o   Esophagitis
o   Gastresophageal reflux disease
o   Pneumonia, aspiration
o   Respiratory failure
o   Tracheal tumors
o   Tracheomalacia
o   Zenker diverticulum

ASSOCIATED ANOMALIES
o   Congenital heart diseases, intestinal atresia, imperforate anus, skeletal anomalies and renal anomalies suggest VACTERL association: **v**ertebral, **a**nal, **c**ardiac, **t**racheal, **e**sophagus, **r**enal, **l**imb
o   Esophageal Atresia may also be seen in the CHARGE association: **c**oloboma, congenital **h**eart disease, choanal **a**tresia, **g**rowth and mental **r**etardaion, **g**enital hypoplasia, **e**ar anomalies

Approximately half of the patients with EA with or without TEF have associated anomalies. Many of these are major and adversely affect the patient's immediate and long term outlook. Associated anomalies, in fact, cause substantially more deaths than malformations. Smaller birth weight infants have more anomalies than the larger birth weight one. Associated anomalies are found 3 times more often in infants weighing less than 2000 gm than those weighing 2500 gm. Isolated EA have more incidence of associated anomalies with any type of anomalies. Infants with VACTERL association tend to have a higher proximal esophageal pouch and more complications with higher rate of mortality. VACTERL association is not a syndrome but a non-random association. If an infant has one of these anomalies another one should be suspected.

Cardiac anomalies are most common and most lethal. Gastrointestinal anomalies are frequently seen but correctable. A variety of anomalies both trivial and serious occur. Some are more immediate risk to life and require treatment before EA and TEF repair. The majority of the anomalies, however, do not interfere with immediate care of infant.

MANAGEMENT

I. Pre-operative management
o   confirming the diagnosis and type of anomaly
o   evaluating the pulmonary status, treating the pulmonary problem and preventing tracheal contamination
o   searching for and if necessary treating other major associated problems

Diagnostic procedures described earlier should be done as soon as after initial assessment and support is provided.

Pulmonary status is evaluated by respiratory rate, degree of respiratory distress, cyanosis, rales or rhonchi and chest radiograph. Occasionally, oxygen saturation and blood gases valve is obtained. To present further aspiration, pharynx should be suctioned continuously. Patient should always be cared in Fowler's position, with head elevated at approx 45^0. This position helps in minimizing the amount of gastric fluid that refluxes back through the distal TEF into trachea by placing the gastresophageal junction above the level of gastric fluid, so that regurgitation is more likely to evacuate gas than liquid. All the patients should be put on combination of Ampicillin and Gentamicin, to cover broad spectrum. If there is clinical or radiographic evidence of significant atelectasis or pneumonia, a decompression Stamm gastrostomy should be done, in order to prevent further gastric reflux through distal TEF and its consequences. Significant improvement of atelectasis or pneumonia prior to major thoracic procedure results in smoother operative and postoperative course. Patients usually respond well within 24 to 72 hrs, at which time the anomaly is repaired.

As we all know major cause of death in these pediatric patients is concomitant disease, so systematic evaluation is of great value. Some conditions need specific therapy and responds well to therapy are congenital heart disease, idiopathic respiratory distress syndrome (IRDS), intestinal and anal atresia etc.

IRDS is a difficult problem to manage. Infants having this require mechanical ventilatory assistance, often with significantly increased intratracheal pressure. Presence of TEF results distension of stomach and further respiratory distress because of diaphragm elevation. This stomach distension frequently leads to gastric rupture, if not vented by gastrostomy. Gastrostomy causes decompression of trachea via fistula to a degree that sufficient intratracheal pressure can't be maintained to provide adequate ventilatory support. This problem can be overcome by temporary occlusion of TEF with Fogarty balloon passed bronchoscopically. A more permanent and secure occlusion is division and suture of TEF as an urgent procedure.

Laryngotracheoesophageal cleft or congenital subglottic stenosis associated with EA or TEF usually requires a prompt tracheostomy.

Some premature infants, those who are quite ill when the cause is not readily apparent, and those who are septic, benefit from a period of support, evaluation and therapy prior to esophageal repair. These patients plus those with major anomalies are probably best treated by gastrostomy to prevent gastroesophageal reflux and its consequences, by central venous catheter for parenteral nutrition and by antibiotics and other appropriate therapy – surgical or medical. When patient stabilizes and begins to improve, EA/TEF repair should be done. Timing is obviously important.

II. Operative management

Repair is performed under general anesthesia. During repair of EA/TEF needs serious involvement of anaesthesiologist in surgical field also, other than anesthetic management. It is he who guides the surgeon in locating and identifying fistula and proximal end of esophagus.

After making a posterolateral incision just below the tip of the scapula, the latissimus dorsi and the serratus anterior muscles are retracted anteriorly. The fourth rib is resected subperiosteally, an incision is made through the deep periosteum. The cut edges are grasped with small mosquito clamps. The plane of endothoracic fascia is developed with blunt dissection. The pleura are gently dissected from chest wall posteriorly to the apex of the chest and two to three interspaces below the incision. If a small opening is made in the pleura, it is closed with small ligature.

The dissection follows the curve of the ribs to the mediastinum. The azygos vein is retracted anteriorly with pleura after dividing the two highest intercoastal veins, as they enter the azygos. The vagus nerve is identified, as it runs along the right side of both the proximal and distal esophagus.

The distal esophagus is identified, looped and freed up to its junction with the post trachea. Traction sutures are placed on the tracheal end of the fistula, which is divided close to the trachea. The trachea must not be narrowed. The tracheal end of the fistula is closed with interrupted sutures. Adjacent tissue, if available is tacked over the closure.

Traction sutures are placed in the end of the upper pouch. The proximal esophagus is freed well into the neck. A proximal pouch fistula to the trachea is possible. The dissection stays close to the pouch. The vagus nerves are avoided. The distal esophagus is freed only as much as necessary to approximate the ends of the Esophagus.

The end of the upper pouch is excised. The posterior row of sutures are placed through all layers of the esophagus and tied inside the lumen. The anterior row is tied on the outside. A 10F catheter is left in the reteropleural space near the anastomosis. The wound is closed in anatomic layers.

Summary of management

- Feeding is withheld and suction applied to esophageal pouch
- Nursed in head elevated position
- Associated congenital abnormalities are identified
- Surgery required with in first 24 hrs of life
- Operation involves
  1. right thoracotomy and extrapleural approach
  2. azygos vein is divided
  3. TEF is divided
  4. esophagus mobilized and primary anastomosis is usually achieved
  5. if anastomosis impossible, a staged procedure is required
  6. gastrostomy performed and fistula divided at initial operation

7.   esophagus replaced by colon or stomach after a few months

Situation

For infants with long gap EA, however, the operation may be much more difficult and the results not always good. The two ends of the esophagus may be thought to be too far apart, or the tissues too thin, raising concern that the repair would be under too much tension and not hold up. Whether the gap is merely long or too long depends on these factors and the viewpoint of the surgeon. For gaps 2-3 cm long or 2-4 vertebral bodies apart, a primary repair will usually be carried out. Surgeons are realizing that some tension will be tolerated by a well constructed anastomosis. As the distance increases, however, so do the likelihood of complications with an attempted primary repair. To avoid this possibility, many surgeons will use an esophageal substitute such as stomach or colon.

a. *The True Primary Repair*

Despite the difficulties imposed when a long gap is present, we believe a true primary repair using the child's own esophagus will be best for the long term. A true primary repair can be defined simply as joining the two esophageal ends together and leaving the stomach entirely below the diaphragm. The stomach must remain in the abdomen where it belongs. Furthermore, no circular incision is made through the esophageal muscles. A circular cut through the muscle wall will allow the remaining tissue to stretch; a circular myotomy. Circular myotomies are not used because of the potential for complications from the weakened esophageal wall. The area of myotomy is unsupported by muscle and may balloon up to a serious degree.

With the esophageal ends joined together and the stomach below the diaphragm, the child has by far the best chance of eating normally. Later problems are also much less likely to occur.

The result of a true primary repair is always the same, the esophageal ends are joined together and the stomach kept below the diaphragm. For most of these infants it can be done at one operation. This has proven true, even if there is a long gap between the esophageal ends. The two esophageal ends can be brought together, even sometimes under a great deal of tension and the repair will still hold together. Therefore, even babies whose gaps are rather long can have an initial true primary repair.

b. *Stimulating the esophagus to grow*

It is not always possible, however, to do a true primary repair initially. If the child has been born with most of the esophagus missing or the first operation has failed, or the upper pouch has been brought out the neck (a spit fistula), the gap will be too great for an immediate (one step) true primary repair. For these children, the esophagus must be made to quickly grow so the repair can be accomplished. We have found that the growth will be rapid and may take only a few days or, at most, 12-14 days. Over this relatively short period of time, the ends of the esophagus will grow significantly and allow a true primary repair to be carried out. The rapid growth of the esophagus is the most important discovery we have made and allows these operations to be carried out.

At the first operation, the two esophageal ends are put on traction towards each other. Occasional, when the gap is not overly long, the traction sutures will rapidly stimulate enough esophageal growth relatively rapidly. When this appears to be the case, the traction sutures are placed internally. After 2-3 days time, the incision is reopened and the esophageal ends sewn together.

For the very longest gap infants, however, more time will be needed. The traction sutures are placed in the esophageal ends and brought through the skin to the outside of the chest wall. This allows the traction to be increased daily and maximizes the growth stimulus. These children are kept on the ventilator and heavily sedated so they do not tear the traction sutures loose. Even the very longest gaps rapidly respond to this growth stimulus. When the ends are virtually together, the infant is returned to the operating room and the esophagus joined.

c. *Esophageal Substitutions (Interposition Grafts)*

Usually at other hospitals, if the gap is very long a true primary repair is not recommended or attempted. If the esophageal ends can not be brought together then another tubular organ must be used to bridge the gap and provide continuity. The most commonly used esophageal substitutions; include colon interpositions, the creation of a stomach tube or a pull-up of the stomach (gastric transposition).

The interposition grafts, with the exception of the jejunum, cause increasing problems and severe consequences with time. Pulling part of the stomach up into the chest so that the two esophageal ends can be joined together is not a true primary repair. Any partial division or elongation or an upward pull-up of the stomach will lead to significant long term consequences and would not meet the definition of a true primary repair.

The most commonly used esophageal substitutions include colon interpositions, the creation of a stomach tube or a pull-up of the stomach (gastric transposition). The consequences of these will be discussed under early and long-term results but suffice it to say, the likelihood of a difficult early course is high.

III. Post-operative management

The intubated patient is transported to neonatal intensive care unit. Antibiotics are continued until the chest drain is removed and endotracheal tube is suctioned necessary. Oral suctioning to a depth of no more than 7 cm from the lips is performed every half an hour on the first day, then every hour or more frequently as necessary on second day. The chest draining tube is placed in 2 cm of water, only to seal it (under water seal), it is not connected to a suction pump as this encourage an anastomotic leak. Morphine is infused as necessary for patient's pain relief and peripheral parenteral nutrition should be commenced. Endotracheal tube should remain in place to ensure ventilation.   Premature   extubation   and   subsequent

reintubation in the setting of a freshly closed tracheal fistula invites reopening of the fistula.

Watch for saliva exiting out the chest drain, this is a signal of anastomotic leakage. Often, it is accompanied by visible distress. Signs of sepsis may or may not be present. A chest radiograph should be obtained. Provided that the baby is stable, a contrast enhanced study of the esophagus with soluble isotonic medium may be performed on day 6 or 7 to assess for leaks and to view the repair. If the esophagus is patent and reasonably sized, the baby may be feed starting with expressed breast milk is ideal. Then the chest drain is removed. As soon as the baby feeding well, the intravenous line is discontinued and the baby can be discharged. Oral ranitidine prescribed for 6 months because of propensity for gastresophageal reflux in this group of patients and because of the risk of strictures as a secondary effect.

FOLLOW-UP CARE

If all is well with the patient and if the parents have been briefed on what to look for, reasonable follow up regimen may include the following steps:

- Make contact with the community physician who is responsible for general medical condition of child and ensure that he or she is briefed on the baby's history, condition and expected outcome
- The nurse in the surgical team should follow up by telephone in one week
- The surgeon should follow up in one month to interview the parents and generally assess the condition, growth and heavily at the surgical site
- The patient should return at 3 months for a similar assessment
- At a 1-year follow up and general assessment, a swallowing function, respiratory issues and gastresophageal reflux should be addressed

Radiologic assessment of the esophagus is required only if a significant history of choking, cyanosis, regurgitation, dysphagia, growth failure, coughing or wheezing exists. Subsequent endoscopic evaluation can be performed as indicated.

Follow-up care when the child is older can be performed as needed. Specific reassessment of patient which is aged approx. 12 yrs is advised; at that time, endoscopic assessment of the patient should be planned. Barrett esophagus and even subsequent malignant change is possible in condition because of the propensity for gastresophageal reflux. Follow-up with periodic endoscopy every few years until the patient is an adult can be justified for this reason.

PROGNOSIS AND COMPLICATIONS

The majority of patients will experience few problems after initial period. However, for others, problems may persist for many years:

- swallowing problems
- stricture
- gastresophageal Reflux

- long gap esophageal atresia
- tube feeding
- respiratory problems

Traditionally, the prognosis for children with EA/TEF was centered on the Waterston Risk Classification, which is based on the birth weight and presence of pneumonia and associated congenital abnormalities. Because of advancements in Neonatal Care, however, this risk classification is no longer prognostic. Pneumonia may be successfully treated, except in some infants with severely low birth weight. Currently, cardiac and chromosomal abnormalities are the most significant causes of death. Regardless of classification scheme, infants with a birth weight less than 1500 gm, major cardiac abnormalities, severe associated anomalies, preoperative ventilator dependence, and/or a long gap are at increased risk.

Preoperatively, the greatest risk to the child with EA/TEF is aspiration gastric rupture has been reported in patients with TEF who are receiving ventilator support. Air is forced through the fistula into the distal esophagus and then into the stomach.

The severity of complications after EA/TEF repair is often dictated by the extent of repair required. Primary anastomosis and fistula closure has fewer complications than esophageal replacement. The most common complications include anastomotic leakage, recurrent fistula, stricture and gastresophageal reflux.

Anastomotic leakage into mediastinum occurs in 14-21% of children that have undergone a surgical repair of EA/TEF. Leaks usually results from small friable lower segment, ischemia of esophageal ends, excess anastomotic tension, sepsis, technically poor suturing techniques and inaccurate mucosal apposition. Most leaks are small, occur later (after first 48 hrs of repair) and require only conservative management with cessation of oral intake. Total Parenteral Nutrition (TPN) and antibiotics are needed. Spontaneous healing occurs in 95% of leaks when chest drain is present. More significant leaks occur early – within the first few days and should be explored immediately. Major anastomotic disruptions occur in only 3-5% of leaks, but large leaks can be total and require surgical repair. Mediastinal leaks can lead to TEF recurrence; therefore, they should be monitored carefully.

Fistula Recurrence between the esophagus and trachea is observed in 3-14% of the patients treated for EA/TEF. Fistula usually recurs within a few months, but they may occur as late as 2 years after surgery. An anastomotic leak with local inflammation and erosion at the previous repair site, ischemia and surgical dissection too near the trachea may cause a recurrent fistula. This condition should be suspected when choking episodes occur during feeding and/or when recurrent pneumonia is observed. The best methods of diagnosis are bronchoscopy and oesophagography under videofluroscopic guidance with the patient in prone position and with bolus injections of contrast agent into a nasesophageal tube. Fistula do not close spontaneously and require surgical division and ligation. About 10-20% of cases recur after the first TEF recurrence.

Esophageal Strictures occur in 40% of cases after surgical EA/TEF repair. Strictures result from natural healing as a result of fibrosis, a difference in the sizes of the 2-anastomosed segments, tension and GER. Leaks as well as the use of 2-layer anastomosis and/or silk sutures, enhance stricture formation. Strictures may be diagnosed with barium swallow examination or oesophagoscopy. Although barium swallow study aids stricture reduction by dilatating the anastomotic site, decreasing the size discrepancy between 2 segments and loosening the fibrosis of healing, it is not completely effective and dilatations are required for resolution. Dilatation is 90% effective but strictures that do not respond to dilatation must be surgically resected.

Gastresophageal Reflux is a common complication, occurring in 40-70%of patients after EA repair. Symptoms of GER include coughing, apnea, recurrent pneumonia, failure to thrive and stricture formation. A barium swallow examinations may demonstrate GER, which is caused by tension, dysmotility of the lower esophagus and an altered angle of Hiss due to distal esophageal mobilization. GER may be medically treated by keeping the patient in a prone head-up position after feeding; by thickening the food; by giving smaller and more frequent meals. If problem persists, acid reduction agents such as histamine H2-receptors blockers and prokinetic agents may be administered. If medical therapy is unsuccessfuly, fundoplication may be considered. Fundoplications are required in about half of the patients with GER. GER tends to diminish with time, but long term GER leads to mucosal changes such as Oesophagitis and Barrett Esophagus.

Swallowing Problems due to altered esophageal peritalsis is seen approx. in all patients after EA/TEF repair. Normally, synchronized waves of contraction of the esophagus walls carry food down to the stomach. These are far less organized after EA/TEF repair. This can mean that food is brought back up, making eating enough to keep weight on-alone enough for a child to grow difficult.

Long Gap Esophageal Atresia is finding in small group of children with EA have too large gap in the esophagus to repair straight away. These children have to be tube-fed for some weeks or months until either the gap has decreased enough to permit the repair; or the surgeon decided that the gap will never close sufficiently and alternative part of Gastrointestinal tract must substitute for Esophagus (the stomach, jejunum or colon). In the latter case, sometimes the upper esophagus is brought out into the neck so that the child learns to eat during the time before such an 'Esophageal Substitution Procedure' can be performed. Even though the food will not reach the stomach, this practice *Sham Feeding* makes learning to eat far easier after a functional 'pipe' between the mouth and stomach has been accomplished. A tube is also placed through the skin and body wall into the stomach so that food can be given at the same time. In this way the child learns to associate eating and swallowing with the feeding of a full stomach. The arrangement is called a "cervical esophagostomy and gastrostomy".

Tube Feeding is done in children who have problems with eating (often due to reflux or strictures, as described above) will need to receive some form of tube feeding to supplement their nutrition intake. Here we use either nasogastric tube or gastrostomy to provide food.

Respiratory Problems, because in TEF there is also an abnormality affecting the trachea. Children often suffer respiratory problems – commonly Asthma and repeated infects, also accompanied by the so called 'TEF Cough' – a harsh, barking cough caused by a softness or floppiness (tracheomalacia) of the normally rigid tracheal wall near the site of repair. The floppy trachea can, in some babies, contribute to so-called 'near death' attacks, when the baby goes blue and may pass out. This occurs when the less rigid airway collapses usually at a time when the child makes heavy breathing efforts, for example when crying or coughing. Although it is frightening to observe. Once the child relaxes, the airway opens up and the problems resolve itself. Occasionally surgery may be required in severe cases though this is unusual.

An esophageal substitution causes additional complications. Esophageal replacement has been associated with an increased surgical morbidity rate and a 68% complication rate. These conditions include the following:
- colon: cervical leaks, pulmonary problems, graft necrosis, redundancy ger, anastomotic stenoses, strictures.

In addition colon graft is thin walled and has poor function: subject to pathology (polyps, villous adenomas); slow transit that leads to dilation over patient's life time (this leads to anaemia, poor weight gain, recurrent pulmonary infections and redundancy); intestinal obstruction in 20% of patients; colon graft ulceration in 10% of cases, especially in reterosternal placements (this may lead to barret's epithelium in the lower esophagus if the lower segment is unused); limited mucosal acid resistance (the colon tends to dilate and form loops).
- gastric tube: frequent leaks and strictures; high incidences of fistulas, stenoses and peptic ulcerations; peptic esophagitis; extensive GER in cervical esophagus, which may lead to peptic ulceration and Barrett epithelium.
- gastric transposition: leakage in 6% cases; stricture at the anastomotic site in 12% of cases; microvasculature easily disturbed with handling; late dialations; long term effects of intrathoracic stomach ulcerations; aspiration.
- jejunum: infarction common; high incidence of peptic ulceration; high free graft failure rate.

Here are the few questions often put by worried parents, so a doctor should always be ready to answer these

Q1. What is a tracheesophageal fistula?

A fistula is a connection between 2 tubes. The breathing tube that connects the nose and mouth with the lungs is called the trachea. The swallowing tube is the esophagus. The breathing tube and the swallowing tube aren't supposed to be connected. But when a child has a tracheesophageal fistula, the fistula connects the 2 tubes. This means that food or milk in the stomach can get into the lungs. This can cause breathing problems and even pneumonia.

Q2. What causes atresia and fistula?

We don't really know what causes these problems. When the esophagus and the trachea grow in the embryo, they start from the same bit of tissue. Sometimes the tubes don't develop right. We don't think these problems are inherited.

Q3. Are these problems common?

About 1 baby out of 4,000 babies has one or both of these problems. They usually occur together. But sometimes a baby has atresia with no fistula.

Q4. How does the doctor know this is what's wrong with my baby?

Most babies with this condition have feeding problems right away. They may spit up a lot or have lots of bubbly mucus in their mouth. If your baby has a fistula, breathing may be hard. If your doctor thinks your baby has one of these conditions, an x-ray can help make the diagnosis.

Q5. How is this problem fixed?

Your baby will need surgery to fix the problem. First, the swallowing tube must be connected to the stomach. Then, if a fistula is connecting the esophagus to the trachea, it must be closed. Your child's doctor will decide when to do the surgery. If the baby isn't premature and doesn't have any other problems (like pneumonia or birth defects), the surgery can usually be done when the baby is just a few days old.

Q6. How long will my baby be sick?

In uncomplicated cases, your baby may be eating by one week after surgery. Meanwhile, until your baby can swallow milk or formula, your baby will be fed through a vein (this is called an "IV") or through a stomach tube.

Before regular feeding starts, an x-ray can check for holes at the place the surgeon fixed. However, if your baby was premature, the recovery time might be a little longer. Another factor is how complicated the operation is. If the surgery is harder, it takes a few days longer for your baby to recover. Your baby will stay in the hospital during this time.

Q7. Does my baby have any other problems?

Some babies with esophageal atresia have heart problems, kidney problems, stomach and bowel problems or muscle and bone problems. A physical exam by your doctor, maybe with some other x-ray or ultrasound pictures, will usually show if your baby has other problems. If your baby has other problems, the surgery to fix the swallowing tube might have to wait.

Q8. Will my baby have other problems in the future?

Babies born with esophageal atresia sometimes have long-term problems. Probably the most common problem is gastresophageal reflux disease, what doctors call heartburn. Heartburn is a burning feeling caused by acid that comes up from the stomach into the swallowing tube. It can usually be treated with medicine. Another problem is scar tissue. Sometimes scar tissue grows where the esophagus connects to the stomach. This scar tissue can make swallowing hard or painful because the food can't get past the scar tissue easily. Sometimes another surgery is needed to open the scar tissue. Your child may need more x-rays or endoscopy later. Endoscopy is a way of taking a picture. A narrow tube holding a tiny camera is put into the swallowing tube. The picture helps your doctor see inside the esophagus and stomach.

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Correspondence to:
Eugen Sorin Boia,
Gospodarilor Street, No. 42,
Timisoara 300778,
Romania
E-mail: eemboia@rdslink.ro