Radiological findings of esophageal atresia, tracheoesophageal fistula, the association VATER / VACTERL and postoperative complications

Poster No.: C-0158
Congress: ECR 2013
Type: Educational Exhibit
Authors: R. Sanchez Oro¹, R. Pastor Toledo¹, L. Ariño Montaner¹, J. Joudanin Seijo², A. Meseguer Carrascosa¹, J. Palmero da Cruz¹;
¹Valencia/ES, ²Valencia/ES
Keywords: Pediatric, Thorax
DOI: 10.1594/ecr2013/C-0158

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR's endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method is strictly prohibited.

You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.

Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

- Describe the different types of esophageal atresia and radiological findings.
- Review their associations with other congenital anomalies (VATER / VACTERL association).
- Know the postoperative complications of esophageal atresia repair and tracheoesophageal fistulas.
- Show illustrative examples.

Background

Esophageal atresia (EA), with or without tracheoesophageal fistula (TEF), is the most important congenital malformation of the esophagus. Its incidence is approximately 1 in 3000-4500 live births.

The trachea and esophagus start as a single structure in fetal life. It is generally accepted that the etiology of this condition is a faulty separation of the two. This often leads to a segmental atresia which most frequently is associated with TEF.

The most common types are presented in figure 1 and are the following: A. EA without fistula (5-10%).
B. EA with proximal fistula (1%).
C. EA with distal fistula (80-90%).
D. EA with proximal and distal fistula (1-3%).
E. H-type fistula without atresia (5-8%).

Descriptive terms are better than numbers or letters when communicating with pediatricians and surgeons. Most children with EA begin with drooling that can trigger coughing and choking early after birth. This, in turn, may cause aspiration into the tracheobronchial tree.

Images for this section:
Fig. 1: Figure 1: Diagram representing the different types of esophageal atresia and tracheoesophageal fistula and their relative incidence: A, esophageal atresia without fistula; B, esophageal atresia with proximal fistula; C, esophageal atresia with distal fistula; D, esophageal atresia with proximal and distal fistula; E, tracheoesophageal fistula without atresia; e, esophagus; s, stomach.
Imaging findings OR Procedure details

The radiologist should examine the infant with EA after failing to introduce a nasogastric tube. Initial chest and abdominal x-rays, will characteristically show the nasogastric tube bent in the upper chest, in the proximal reservoir.

In patients with a clinical suspicion of EA, the chest x-ray should show the nasogastric tube looped in the proximal esophageal pocket. Discarded, therefore, would be the H-type fistula without atresia. The next step would be to observe whether or not there is gastrointestinal aeration, which if affirmative would indicate an EA with only a distal fistula (Fig. 3) or an EA with distal and proximal fistulas (Fig. 4 and 5). If there is no gastrointestinal aeration, this would rule out a distal fistula. In this case it might be an EA with a proximal fistula (Fig 6 and 7) or an EA without fistula. When there is a distal fistula, the abdomen is distended, if not, the abdomen is scaphoid.

The proximal fistula is shown by performing an upper intestinal transit (Fig 7). It is recommended to administer non-ionic, soluble contrast under a fluoroscope. Only a small amount should be introduced through a nasogastric tube. This should be done with the infant in the lateral position and then switching to prone position to facilitate viewing the fistula. The infant should not lie supine to avoid the risk of aspiration. The fistula will extend from the anterior wall of the esophagus at an angle towards the rear wall of the trachea or, less frequently, the left bronchus.

The TEF without EA, H- or N-type fistula, can be difficult to diagnose. Any child with a history of choking on liquid food, coughing during feeding and recurrent episodes of unexplained pneumonia should be suspect for this type of pathology. Usually there is a single fistula, although occasionally there may be more located on several levels. Most frequently they are congenital, although there are cases where the fistulas are acquired secondary to trauma, infection, esophageal diverticula, foreign bodies and necrotizing vasculitis.

About half of the children with esophageal atresia have other congenital anomalies. The association of these anomalies is given the acronym VATER:

- Vertebral abnormalities: at any level of the spine and which may consist of aplasia or hypoplasia of the pedicles or vertebral bodies (Fig 8).
- Anorectal anomalies: the most common is upper rectal atresia with a fistula to the bladder or vagina (Fig 9).
- TEF.
- EA.
Renal dysplasia (Fig 10, 11 and 12)

The acronym VATER has been expanded to VACTERL in order to include cardiac and limb alterations (Fig 13).

This is a casual association of alterations due to an unknown error or errors in embryogenesis during the 5th week of gestation. Thus, while many children with EA have one or more VACTERL or VATER alterations, few have all of them. Overall, the most frequent anomalies in children with EA are those of the cardiovascular system and gastrointestinal tract.

The most common cardiovascular disorders are ventricular communications, persistent ductus arteriosus and tetralogy of Fallot. Other less frequent ones are single ventricle, transposition of the great arteries and a single umbilical artery. Approximately 5% of children have a right aortic arch. Surgery can cause problems if a right thoracotomy is performed. The surgeons must be informed of the position of the aortic arch since the posterolateral thoracotomy must be done on the side opposite the aortic arch.

Of the associated gastrointestinal disorders, other atresias, especially anal and duodenal, are the most frequent. One of the most curious is that which affects the vertebrae and ribs. In about 25% of cases, there are 13 or more thoracic vertebrae and rib bodies (Fig 14) or 6 or more lumbar vertebral bodies.

Immediate postoperative complications after repair of EA and TEF are anastomotic dehiscence and recurrent TEF (Fig 15). Recurrent TEF, appearing in 10% of cases, may be difficult to detect. In the case of isolated TEF, for example, it may require several examinations.

The most common long-term complications are recurring respiratory and gastrointestinal problems such as coughing, choking, dysphagia, failure to thrive, apnea, cyanosis and stridor. Probably the most common cause of dysphagia is stenosis of the anastomosis (Fig 16). Food impaction proximal to the stenotic anastomosis is very frequent. GER also occurs (Fig 17) in the majority of children after repair of EA and can cause esophagitis and stenosis. Subsequently, after repair of the EA, there may be anastomotic stenosis and peptic stricture, and much less frequently, congenital stenosis.

Tracheomalacia is somewhere present in almost all patients with atresia repair and may be symptomatic or asymptomatic. In some cases, it may be severe enough to cause apnea that compromises the patient's life and requires aortopexy.
Years after the EA repair, there can be severe progressive scoliosis, concave toward the side of the thoracotomy. It is more common if there has been severe mediastinitis and secondary empyema to the esophageal anastomotic dehiscence.

Images for this section:

Fig. 2: Figure 3: The nasogastric tube is looped in the proximal esophageal pocket, passage to the stomach is not possible. Distal intestinal aeration is observed. These are findings that correspond to esophageal atresia with a distal tracheoesophageal fistula. We may also see an increase in bilateral granular endoplasmic density with a loss of volume, related to surfactant deficiency disease.
**Fig. 3:** Figure 4: The same patient as in figure 5. The nasogastric tube is positioned in the proximal esophageal reservoir, distal intestinal aeration is observed. Therefore, there exists an esophageal atresia with a distal fistula. There is also an increase in paraumbilical density in relation to omphalocele and 11 pairs of ribs.
**Fig. 4:** The same patient as in figure 54. An upper GI transit is performed by injecting isosmolar water-soluble contrast via the nasal tube. There is a dilated proximal esophageal atresia that ends at the level of the T7. The fistular passage to the tracheobronchial tree is identified by the contrast. Therefore, this patient had atresia with both distal and proximal fistulas.
**Fig. 5:** Figure 6: The same patient as in figure 7. Nasogastric probe ends at the level of the D5, in the proximal esophageal pocket because of the esophageal atresia. Intestinal aeration is not observed, which indicates the absence of a distal fistula.

**Fig. 6:** Figure 7: The same patient as in Figure 6. Esophageal transit: isosmolar water soluble contrast is introduced by NGP, located in the proximal third of the esophagus, esophageal pocket is filled, and the contrast passes into the tracheobronchial tree via the proximal fistula. Intestinal transit: soluble contrast is introduced through a gastrostomy tube, filling the stomach and small intestine.
Fig. 7: Figure 8: D6 and D10 hemivertebrae
**Fig. 8:** Figure 9: Patient with anal atresia, after the introduction of water-soluble contrast by colostomy. The extravasation of the vaginal fistula can be seen.
Fig. 9: Figure 10: Figures 10 and 11. Nephrostomy tube in right hemiabdomen. Bilateral ureterohydronephrosis delay in eliminating the contrast and dilation of pyelocaliceal systems and ureters up to the vesicoureteral junction. Ureters dyed with contrast and without a visible movement of contrast to the bladder until viewed on an x-ray made later (Figure 11); therefore, it is a case of a stenosis in the bilateral vesicoureteral junction.
Fig. 10: Figure 11: Figures 10 and 11. Nephrostomy tube in right hemiabdomen. Bilateral ureterohydronephrosis delay in eliminating the contrast and dilation of pyelocaliceal systems and ureters up to the vesicoureteral junction. Ureters dyed with contrast, without a visible movement of contrast into the bladder until viewed on an x-ray made later (Figure 11); therefore, it is a case of a stenosis in the bilateral vesicoureteral junction.
**Fig. 11:** Figure 12: Left flank ultrasound where one can see akidney in profile: black arrow, left kidney; red star, aorta; blue star, inferior vena cava; yellow arrow, renal isthmus; white arrow, right kidney.
Fig. 12: Figure 13: Lack of first metacarpal of the right hand, with three rudimentary phalanges in the first finger of the hand. Eleven pairs of ribs can also been seen.
**Fig. 13:** Figure 14: 13 pairs of ribs and hemivertebrae in S1

**Fig. 14:** Figure 15: Soluble isosmolar contrast is administered orally. An esophagus of normal caliber is seen, with the passage of contrast to the tracheobronchial tree by previous reperfusion of the tracheal esophageal fistula located in the cervical esophagus.
Fig. 15: Figure 16: The area of smallest diameter is displayed in the upper third of the esophagus, a result of the stenosis of the anastomosis.
**Fig. 16:** Upper intestinal transit showing a stenosis in the middle esophagus, related to corrective surgery for atresia. The passage of contrast to stomach through wide, incompetent hiatus, results in gastroesophageal reflux.
Conclusion

It is important to be able to recognize in images the characteristic traits of isolated esophageal atresia or of those associated with tracheoesophageal fistulas. Just as important is the knowledge of the associated malformations in order to be able to detect them early, since the mortality of esophageal atresia is largely a result of the associated anomalies.

References


Personal Information