Imaging of congenital disorders of esophagus in infancy and childhood

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Introduction

- From practical point of view the esophageal abnormalities have to be divided into congenital and acquired diseases
- Congenital diseases manifest themselves mostly in early life
- Acquired disorders are usually expected in later life
- Each group of abnormalities should be categorized etiologically
Congenital disorders of esophagus

1. Esophageal atresia (EA) and tracheo-esophageal fistula
2. Congenital esophageal stenosis
3. Esophageal duplication
4. Laryngo-tracheo-esophageal cleft (LTE)
5. Diverticula
6. Epidermolysis bullosa dystrophica
7. Extrinsic impression of esophagus by vascular ring or mediastinal masses
Esophageal atresia (EA)

- Frequency 1: 2500-3000 life births
- Exact etiology unknown, but it is a developmental disorder in formation and separation of the primitive foregut into trachea and esophagus
- Inability to swallow salivary secretions with overflow and aspiration
- Various types of esophageal atresia
VACTERL association is an acronym for a complex of anomalies affecting various systems:

- **V** = Vertebral
- **A** = Anorectal
- **C** = Cardiovascular
- **TE** = Tracheo-esophageal
- **R** = Renal
- **L** = Limbs
Preoperative imaging of EA

- Positioning of a radio-opaque feeding tube to the level of atretic pouch
- Cervicothoraco-abdominal plain film (AP), if necessary added with a lateral chest film
- Contrast pouchogram of atretic proximal esophagus is unnecessary because of risk of aspiration
- Use of non-ionic contrast medium is absolutely indicated in only suspected cases of TEF without atresia (H-type)
- Cardiac and abdominal sonography
- CT and MRI are only useful in exceptional cases
Radiological findings

- The radiological appearance varies with the type of lesion
- The proximal blind ending pouch is usually lucent and distended with air
- In cases with a distal TEF bowel gas is present
- A gasless abdomen is characteristic for EA without distal fistula (long gap esophageal atresia)
- Special attention should be paid in recognition of other GI tract obstruction, cardiopulmonary and skeletal abnormalities
Aspiration pneumonia after use of contrast medium by EA

Neonate with esophageal atresia and right-sided pulmonal agenesis.

Note the dilated proximal esophageal pouch with air.
Two cases of tracheo-esophageal fistula without atresia
Non-ionic contrast media has been used
Postoperative findings and complication of EA

- Gastro-esophageal reflux (70-100%)
- Esophageal stricture (up to 40%) specially following leakage
- Esophageal leak ± 18%
- Tracheomalacia
- Poor esophageal motility and lodging of large bolus of food above the anastomosis
- Recurrent fistula
- Additional lower congenital stenosis
- Incidental finding of a overlooked proximal fistula
Recurrent TEF

Second congenital stenosis by EA

Lodging of food

Proximal fistula years after primary correction
EA without distal fistula
Long gap (atresia)

A: primary correction
B: colonic interposition
C: jejunal interposition
Congenital esophageal stenosis

- Frequency 1 : 25,000-50,000 life birth
- One third of cases are associated with EA
- Located mostly in distal third of the esophagus
- Some stenoses are membranous web shaped such as diaphragm and can be multiple
- They may occur as an isolated narrowing due to ectopic cartilaginous tracheobronchial remnants; some other due to segmental hypertrophy of muscular layers or fibrosis of mucosal layers
- The stenosis should be differentiated from peptic stenosis related to gastro-oesophageal reflux disease
Note the different types of one or more congenital webs and stenoses
Esophageal duplication cysts

- Related to an abnormality in tubulation of esophagus
- Second most common duplication after ileal duplication (15-20%)
- Usually no communication with esophageal lumen
- Most common incidental finding on chest film or by contrast study of GI tract
- Additionally to contrast esophagogram, US, CT and MRI are all helpful to demonstrate the location, extension and its relation to neighbouring organs
- Duplication cysts should be differentiated from other cystic or solid mediastinal masses, especially from bronchogenic cysts
Two cases of duplication cyst of esophagus
Laryngotracheo-esophageal cleft (LTE)

- Very rare congenital abnormality
- Depending to its extension classified in four types
- Majority has associated congenital malformations such as esophageal atresia, congenital heart diseases and pulmonary abnormalities
- Clinical findings: cyanosis, choking on feeding and aspiration
- Contrast examination: free spillage of contrast from esophagus to trachea (should be avoided)
- CT is a possibility to demonstrate the LTE cleft
- Endoscopic evaluation is the first method of choice for diagnosis and classification of the LTE cleft
Two cases of laryngothracheoesophageal cleft
Diverticula

- Rarely to be observed in children
- Pressure diverticula are herniations of the mucosa and submucosa through congenitally weak sites of the esophagus wall
- Usually located above the clavicles
- Best visible in lateral or oblique projection
- Large diverticula can be observed following leak of repaired esophageal atresia and should be differentiated from congenital diverticula
Congenital diverticula located in upper part of the esophagus

Large diverticula following leakage of corrected esophageal atresia (acquired)
Epidermolysis bullosa dystrophica

- A congenital disease affecting squamous epithelium with involvement of esophagus
- Inherited in an autosomal recessive pattern
- Can produce strictures and dysmotility of esophagus
- Gastro-intestinal manifestation can occur in the absence of cutaneous changes
A case of epidermolysis bullosa dystrophica with esophageal stricture

Another case of epidermolysis bullosa dystrophica with atretic esophagus and pyloric canal
Vascular rings anomalies

- Extrinsic impressions on the esophagus
- The main complaints are related to the airways compression and rarely lead to dysphagia
- An anterior compression on the trachea with a posterior impression on the esophagus is a vascular ring
- A vascular ring is a result of either a double aortic arch or a right aortic arch with an aberrant left subclavicular artery and ductus arteriosus remnant
- Additionally to initial esophagram, CTA or MRA are absolutely indicated in preoperative deliniation of the anatomy of the vascular ring
Imaging of arcus aortae duplex with extrinsic esophageal compression