Pediatr Radiol (2015) 45:1230–1235
DOI 10.1007/s00247-015-3276-1

HISTORICAL PERSPECTIVE

From Vogt to Haight and Holt to now: the history of esophageal atresia over the last century

Oliver J. Muensterer · Walter E. Berdon

Received: 14 April 2014 / Revised: 20 August 2014 / Accepted: 2 January 2015 / Published online: 11 February 2015
© Springer-Verlag Berlin Heidelberg 2015

Abstract Esophageal atresia was first classified by the Boston Children's Hospital radiologist Edward Vogt in 1929 and has been a major challenge in its characterization and management ever since. It defied all attempts at repair until University of Michigan thoracic surgeon Cameron Haight's first successful fistula ligation and primary esophageal anastomosis in 1941. Haight worked with the pediatric radiologist John Holt. This historical review describes advances in pre- and postnatal diagnosis.

Keywords Esophageal atresia · Tracheoesophageal fistula · Infant · History · Vogt · Haight · Holt

Early history

Esophageal atresia (EA) and tracheoesophageal fistula (TEF) defied primary surgical repair well into the 20th Century. Over time, several attempts were made at palliating the condition by placing a gastrostomy, ligating the lower esophagus at the cardia, and/or performing a cervical esophagostomy. All of the patients invariably died of their disease.

O. J. Muensterer (✉)
Department of Pediatric Surgery, University Medicine Mainz, Langenbeckstrasse 1, 55131 Mainz, Germany
e-mail: oliver.muensterer@att.net

W. E. Berdon
Department of Radiology, Morgan Stanley Children's Hospital, Columbia University, Bronx, NY, USA

Edward Vogt and the classification of esophageal atresia

The American radiologist Edward Vogt (Fig. 1) proposed the first classification of the spectrum of EA and TEF anomalies in 1929 [1]. He trained in radiology in Boston and New Haven, before being appointed as the first chief of radiology at the Children's Hospital in Boston, MA [2].

Interestingly, the Vogt classification is still widely cited in Europe, while it has been mostly replaced in North America by the Gross classification (Table 1). Most clinicians agree that the best way to describe the malformation is by its true anatomical description, avoiding any misunderstanding. Whatever classification or description used, it must be remembered that the vast majority, at least three-quarters of cases, present with a blind-ending proximal pouch and distal tracheoesophageal fistula. The second-most-common anomaly is pure esophageal atresia without fistulas.

Initial postnatal imaging and management

As Vogt already described in his publication, most of the patients have a blind-ending upper pouch with a distal tracheoesophageal connection. After birth, failure to pass a nasogastric tube is usually the pathognomonic sign of esophageal atresia. If encountered, the tube should be left in place to mark the length of the pouch. Alternatively, air can be instilled into the nasoesophageal tube to demonstrate a negative contrast contour of the pouch (Fig. 2). While some advocate instilling a small amount of water-soluble contrast, care must be taken to avoid aspiration into the lungs (Fig. 3).

The tell-tale indication of a distal tracheoesophageal fistula is generally regarded to be air within the stomach on the first radiograph. However, in rare cases, a gasless abdomen may be the result of a mucous plug in the TEF that resolves and

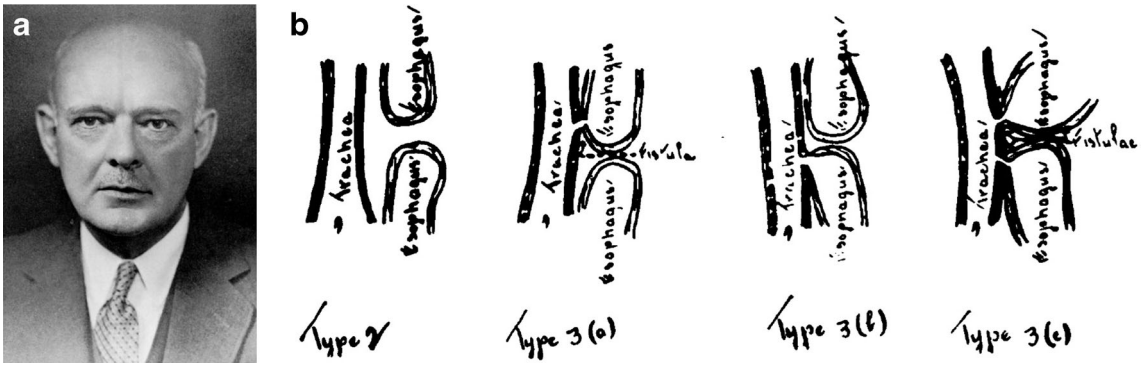

Fig. 1 The American pediatric radiologist Edward C. Vogt (**a**, 1891–1980) provided the first classification (**b**) in 1929 for the spectrum of esophageal atresia and tracheoesophageal fistula, which is still widely used in Europe [1, 2]

Table 1 Classifications of esophageal atresia and/or tracheoesophageal fistula

Vogt [1]	2	3 a	3 b	3 c
Gross [3]	A	B	C	D
Ladd [4]	I	II	III, IV	V
Spitz [5]	b	e	a	c
Anatomical description	Pure esophageal atresia	Esophageal atresia with proximal fistula	Esophageal atresia with distal fistula	Esophageal with double fistula (proximal and distal)
Incidence (%)	18–20	low, but may be more common than appreciated	75–80	low

permits air to pass distally only after some elapsed time (Fig. 4). Repeat radiographs, or preoperative bronchoscopy and esophagoscopy, are helpful in these rare instances.

Recent series have reported a higher incidence of proximal tracheoesophageal fistula than previously cited (Fig. 5), perhaps up to 5% of all cases [6]. Preoperative knowledge of a proximal fistula is extremely important for the pediatric surgeon so that it can be addressed adequately during the repair. If missed, chronic aspiration with pneumonia can result.

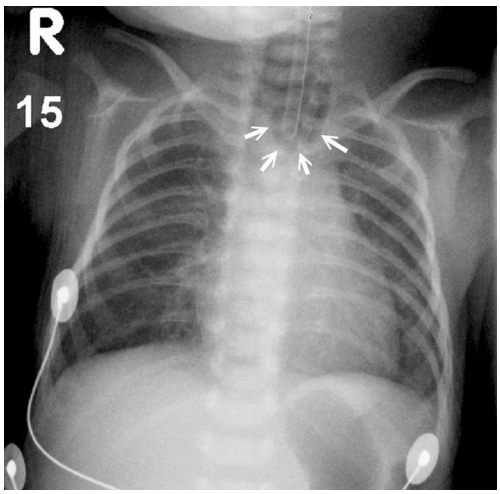

Fig. 2 Air contrast radiograph of the upper pouch (*arrows*). Also note air in stomach, indicating the presence of a distal tracheoesophageal fistula

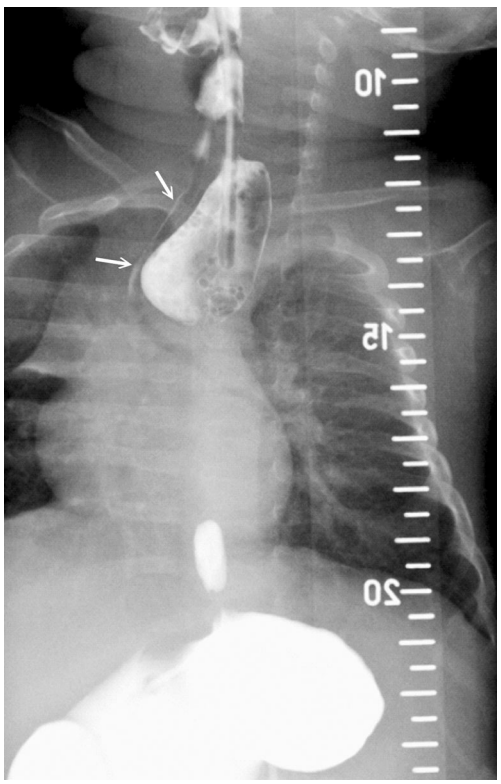

Fig. 3 Upper pouch contrast study and "gapogram" in a 2-month-old with pure esophageal atresia. Too much volume was used in this case, leading to aspiration of some contrast into the trachea (*arrows*). The other interesting finding is the degree of tracheal compression that can result from a distended upper pouch, potentially leading to respiratory distress. Contrast in the stomach is instilled via the gastrostomy

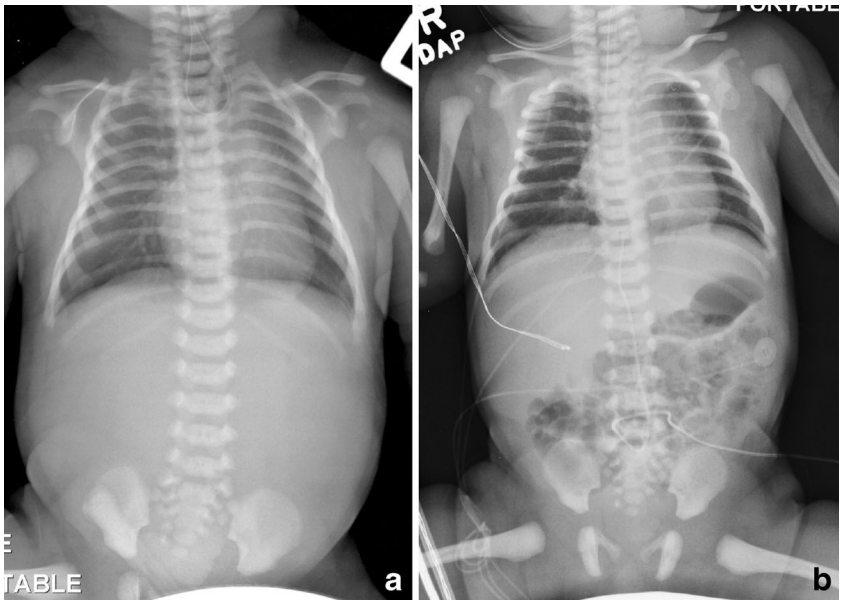

Fig. 4 Type 3b with initially gasless abdomen in a newborn with esophageal atresia and distal tracheoesophageal fistula. A mucous plug precluded air to pass below the diaphragm (**a**). A repeat film 12 h later demonstrates the typical gas pattern of the lesion (**b**)

Cameron Haight, John Holt and the first primary successful repair

Cameron Haight (Fig. 6), a thoracic surgeon trained in adult procedures at the University of Michigan, was the first to perform the first primary repair of esophageal atresia with distal tracheoesophageal fistula, as published in 1941 [7]. He had tried the operation before on several patients without success.

The initial radiograph showed a blind-ending upper pouch and a markedly distended stomach (Fig. 7). The patient survived postoperative bouts of cyanosis, as well as a large anastomotic leak and stricture that required dilatation. After being hospitalized for 20 months, she was discharged on an oral diet. This little girl who first survived primary anastomosis for EA with distal TEF was also the last patient Haight saw before his death in 1970 [8].

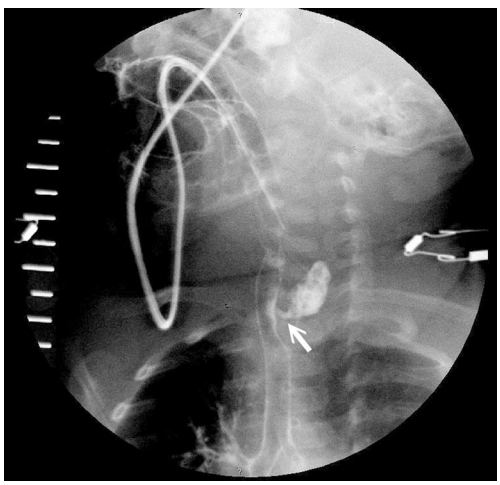

Fig. 5 An unsuspected proximal fistula (Vogt type 3a) is seen on this upper pouch contrast study (*arrow*)

The pediatric radiologist working with Haight at the University of Michigan was John F. (Jack) Holt (Fig. 6). As a young attending, he was picked by his chief of radiology to dedicate time to this new clinical problem. Together, they published their landmark paper on 46 patients, employing the Vogt classification [9].

Michael Harrison, the side of the arch and implications for repair

In 1977, Michael Harrison, a pediatric surgeon in Los Angeles, reviewed 130 infants with esophageal atresia and found the presence of a right aortic arch in 5% of the patients [10]. He recommended operating on the contralateral side of the arch. This pediatric surgical dogma is still adhered to in most centers [11]. Recently, however, a number of publications have questioned this proposition and have described equally good outcomes whether the repair is performed from the left or the right side in case of right aortic arch [12]. Harrison's publication includes very useful diagrams on the anatomical relationships of the structures as seen by the surgeon at thoracotomy.

The rare EA/TEF patients who present with heterotaxia and polysplenia should have abdominal US with Doppler studies performed, looking for continuation of the inferior vena cava into the azygos. In these rare cases, ligating the azygos during the TEF repair may be lethal [13].

Treatment options for pure esophageal atresia

In pure esophageal atresia, the proximal and distal esophageal pouches are commonly too far away from each other to allow

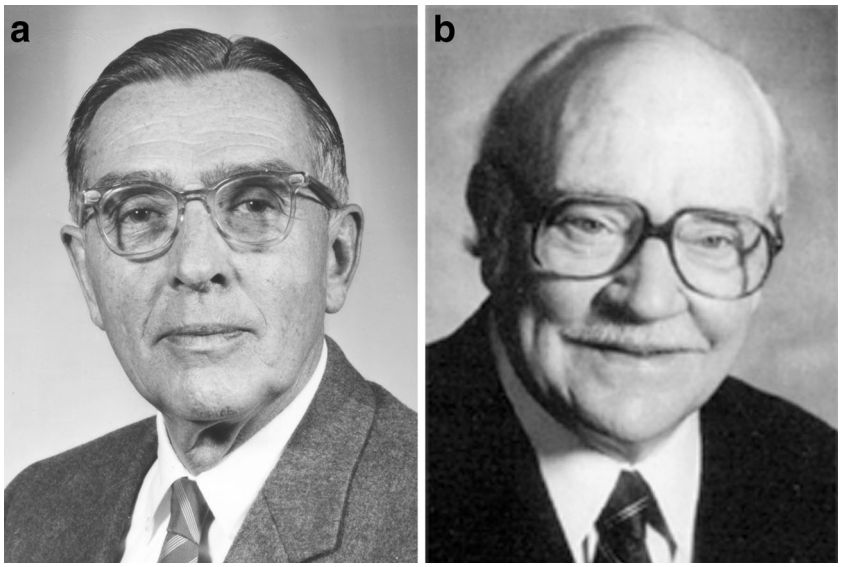

Fig. 6 Cameron Haight (**a**) performed the first successful primary esophageal atresia repair in 1941 (from public domain). He worked together with University of Michigan's pediatric radiologist John Holt (**b**, courtesy University of Michigan archives)

for initial primary repair. Therefore, several therapeutic options have been devised, all of them with caveats. Interposition of the transverse colon was first described for lye strictures and later adapted for treatment of esophageal atresia by German and Waterston [14]. Spitz at Great Ormond Street Hospital in London [15], and Coran at Haight and Holt's alma mater in Ann Arbor [16] propagated gastric transposition with cervical anastomosis of the fundus to the upper pouch. At the University of Iowa, Kimura [17] described a staged extrathoracic advancement of the cutaneous esophagostomy in set intervals until the upper segment was long enough for anastomosis. Most recently, Foker at the University of Minnesota [18] published a method in which external traction is placed on the proximal and distal ends until these overlap (Fig. 8), at which time a primary anastomosis can be performed.

Unfortunately, all these techniques are fraught with complications, some of them common, and others unique to the method employed. It is important that the pediatric radiologist be familiar with these methods to correctly interpret the obtained imaging studies and detect potential problems early on.

Prenatal detection and imaging

Since the 1980s, the principle modality for prenatal screening and imaging diagnosis has been ultrasonography [19]. The typical findings raising the suspicion for EA/TEF are

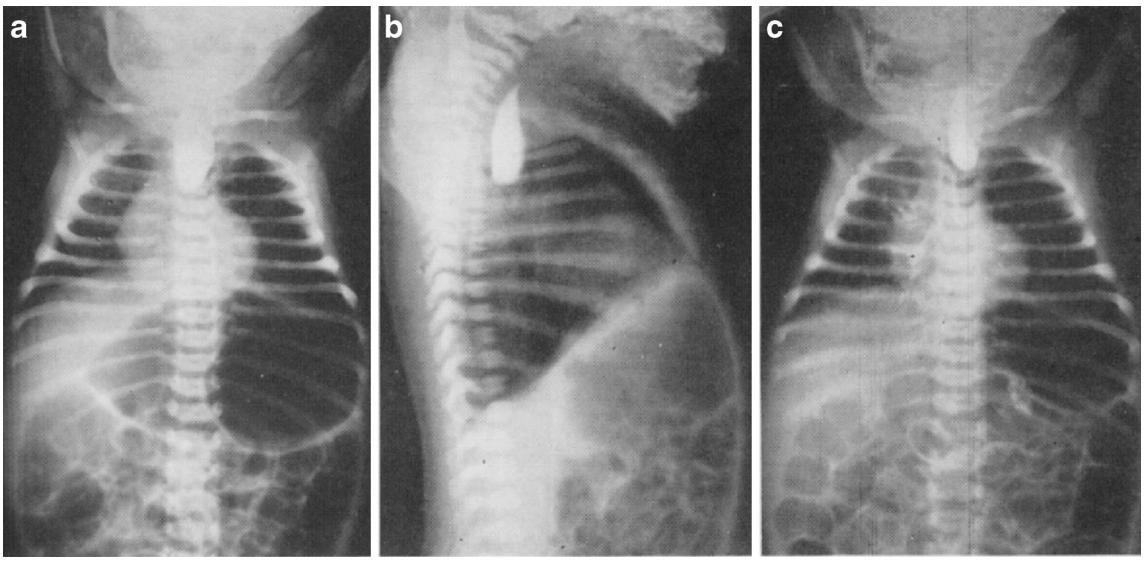

Fig. 7 Initial radiographs with upper pouch contrast study (Vogt type 3b) of Haight's patient [12]. In the initial posteroanterior and lateral views (**a** and **b**, respectively), the upper pouch contains contrast and the stomach is markedly distended, while a later posteroanterior film shows contrast aspirated into the bronchial tree (**c**) (reprinted with permission from the Journal of the American College of Surgeons, formerly Surgery Gynecology & Obstetrics)

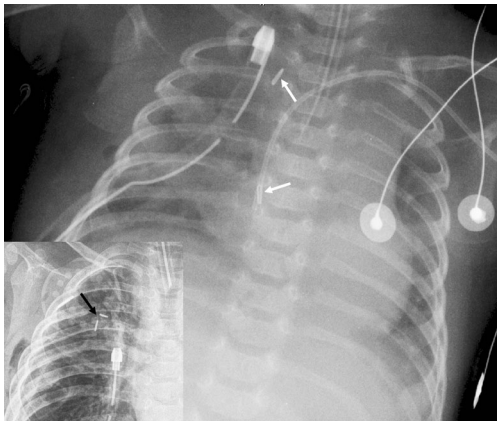

Fig. 8 Chest radiographs of the Foker external traction procedure for long-gap esophageal atresia. Immediately after the initial operation, the tips of the upper and lower esophageal pouches are marked by clips (large image, *white arrows*). Daily increasing traction is applied, until the tips approximate 5 days later (left lower inset, *black arrow*), indicating that a low-tension anastomosis can be performed

polyhydramnios and a small fetal stomach. Intraluminal pressures in utero are not sufficient for the stomach to become distended prenatally. Follow-up magnetic resonance imaging (MRI), as demonstrated in Fig. 9, may be better to diagnose

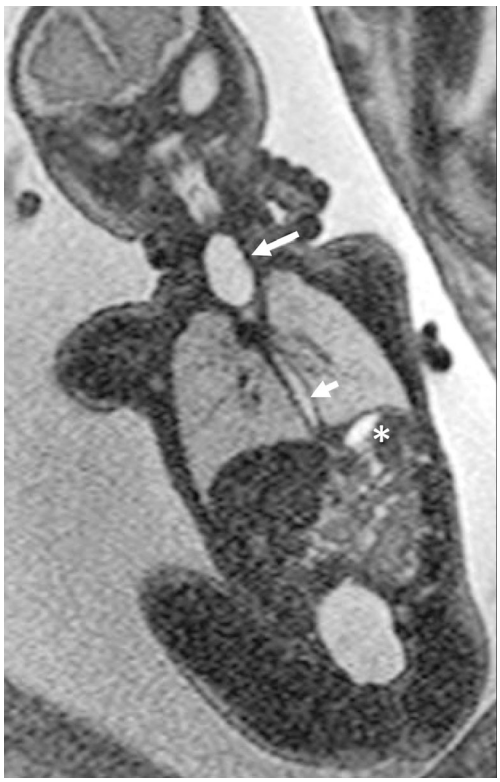

Fig. 9 T2-weighted MRI of esophageal atresia in a 28-week fetus (Vogt, type 3b). Note the distended upper pouch (*long arrow*) and slender distal esophagus (*short arrow*) as well as the small stomach (*), which is typically not distended in utero due to the lack of intraluminal pressure (Victoria T et al. [2011] Fetal MRI of common non-CNS abnormalities: A review. Appl Radiol 40:8–17, reprinted with kind permission from Anderson Publishing Ltd.)

EA/TEF in the fetus as well as screen for concomitant anomalies [20].

Despite advances in prenatal US and MRI, however, many cases are still not diagnosed until after delivery.

Prenatally or postnatally, associated anomalies should be investigated. The VATER association described by Smith in 1973 [21] is the most common type, including vertebral, anorectal, tracheoesophageal and renal malformations. Other common additional anomalies are those affecting the heart (cardiac) and limbs (particularly radial), incorporated into the term VACTERL.

Conclusion and outlook

It is important to note that esophageal atresia is a lifelong disease and that surgery restores the anatomy but not the peristaltic functionality of this complex organ. The long-term impact on esophageal motility was, in fact, noted soon after the first successful surgeries by the pediatric radiologist John Kirkpatrick at St. Christopher's Hospital for Children in Philadelphia [22]. Consequently, pediatric radiologists may be involved in investigating a multitude of different primary and secondary complications. Early after the repair, these are most commonly anastomotic strictures or leaks. Later, the patients have a high risk of presenting with gastroesophageal reflux disease, food bolus impaction or, less commonly, recurrent tracheoesophageal fistula. Management requires close cooperation between the pediatric radiologist, the pediatric surgeon and the gastroenterologist.

While the typical problems of strictures and leaks have been observed since Haight repaired his first patient, the new approaches pose additional challenges. Gastric transposition may lead to cervical strictures and leaks, pulmonary aspiration and metaplasia at the pharyngeogastric anastomosis, as well as dumping syndrome [23]. Frequent complications have been noted after the Foker procedure as well [24], including the tearing out of traction sutures or anastomotic breakdown due to poor blood supply.

Despite the described advances, making the correct diagnosis of esophageal atresia and tracheoesophageal fistula is still challenging, particularly in patients with unusual anatomical variations. Prenatal imaging requires a team approach beyond obstetric US, including pediatric diagnostic radiology. Although improved anesthesia and intensive care have increased overall survival in these patients, the major problems identified by Haight and Holt half a century ago, prematurity and cardiac comorbidities, still influence outcome.

Recently, in utero diagnosis has improved with careful sonographic surveillance. Fetal MRI may add additional information for the exact diagnosis of EA/TEF, along with any associated anomalies.

In the future, more precise prenatal diagnosis may be possible and advances in tissue engineering may permit the construction of a new esophagus using a patient's own stem cells. Recently, a tracheal interposition has already been tissue engineered and successfully implanted in a child with satisfactory outcome [25]. For the involved scientists, growing a new esophagus is the declared next frontier to be conquered [26].

Conflicts of interest None

References

1. Vogt EC (1929) Congenital esophageal atresia. AJR Am J Roentgenol 22:463–465
2. Lewis EO (1981) In memoriam: Edward C. Vogt, M.D. Radiology 140:244
3. Gross RE (1953) The surgery of infancy and childhood. WB Saunders, Philadelphia
4. Ladd WE (1944) The surgical treatment of esophageal atresia and tracheoesophageal fistulas. N Engl J Med 230:625–637
5. Spitz L (2007) Oesophageal atresia. Orphanet J Rare Dis 2:24. Available online at http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1884133/#B76. Accessed 27 November 2013
6. Parolini F, Morandi A, Macchini F et al (2013) Esophageal atresia with proximal tracheoesophageal fistula: a missed diagnosis. J Pediatr Surg 48:E13–E17
7. Haight C, Towsley H (1943) Congenital atresia of the esophagus with tracheoesophageal fistula: extrapleural ligation of fistula and end-to-end anastomosis of esophageal segments. Surg Gynecol Obstet 76:672–688
8. Langston HT (1984) The first successful total repair of congenital atresia of the esophagus with tracheoesophageal fistula. Ann Thorac Surg 38:72–74
9. Holt JF, Haight C, Hodges F (1946) Congenital atresia of the esophagus and tracheo-esophageal fistula. Radiology 47:457–470
10. Harrison MR, Hanson BA, Mahour GH et al (1977) The significance of right aortic arch in repair of esophageal atresia and tracheoesophageal fistula. J Pediatr Surg 12:861–869
11. Allen SR, Ignacio R, Falcone RA et al (2006) The effect of a right-sided aortic arch on outcome in children with esophageal atresia and tracheoesophageal fistula. J Pediatr Surg 41:479–483
12. Wood JA, Carachi R (2012) The right-sided aortic arch in children with oesophageal atresia and tracheo-oesophageal fistula. Eur J Pediatr Surg 22:3–7
13. Evans WN, Kogut K, Acherman RJ (2013) Preserving the azygos vein when repairing esophageal atresia and tracheoesophageal fistula accompanied by interrupted inferior vena cava. Pediatr Surg Int 30:345–347
14. German JC, Waterston DJ (1976) Colon interposition for the replacement of the esophagus in children. J Pediatr Surg 11:227–234
15. Spitz L (1984) Gastric transposition via the mediastinal route for infants with long-gap esophageal atresia. J Pediatr Surg 19:149–154
16. Macksood DJ, Blane CE, Drongowski RA et al (1997) Complications after gastric transposition in children. Can Assoc Radiol J 48:259–264
17. Kimura K, Soper RT (1994) Multistaged extrathoracic esophageal elongation for long gap esophageal atresia. J Pediatr Surg 29:566–568
18. Foker JE, Linden BC, Boyle EM Jr et al (1997) Development of a true primary repair for the full spectrum of esophageal atresia. Ann Surg 226:533–541
19. Farrant P (1980) The antenatal diagnosis of oesophageal atresia by ultrasound. Br J Radiol 53:1202–1203
20. Victoria T, Johnson AM, Chauvin NA et al (2011) Fetal MRI of common non-CNS abnormalities: a review. Appl Radiol 40:8–17
21. Quan L, Smith DW (1973) The VATER association. Vertebral defects, anal atresia, T-E fistula with esophageal atresia, radial and renal dysplasia: a spectrum of associated defects. J Pediatr 82:104–107
22. Kirkpatrick JA, Cresson SL, Pilling GP 4th (1961) The motor activity of the esophagus in association with esophageal atresia and tracheoesophageal fistula. Am J Roentgenol Radium Ther Nucl Med 86:884–887
23. Spitz L (2014) Esophageal replacement: overcoming the need. J Pediatr Surg 49:849–852
24. Liszewski MC, Bairdain S, Buonomo C et al (2014) Imaging of long gap esophageal atresia and the Foker process: expected findings and complications. Pediatr Radiol 44:467–475
25. Elliott MJ, De Coppi P, Speggiorin S et al (2012) Stem-cell-based, tissue engineered tracheal replacement in a child: a 2-year follow-up study. Lancet 380:994–1000
26. Maghsoudlou P, Eaton S, De Coppi P (2014) Tissue engineering of the esophagus. Semin Pediatr Surg 23:127–134