



Esophageal atresia and tracheo-esophageal fistula

David C. van der Zee, MD, PhD^{a,*}, Stefaan H.A. Tytgat, MD, PhD^b,
Maud Y.A. van Herwaarden^b

^a Professor of Pediatric Surgery, Dept. Pediatric Surgery, University Medical Center Utrecht, The Netherlands

^b Pediatric Surgeon, Dept. Pediatric Surgery, University Medical Center Utrecht, The Netherlands

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ABSTRACT

Management of esophageal atresia has merged from correction of the anomaly to the complete spectrum of management of esophageal atresia and all its sequelae. It is the purpose of this article to give an overview of all aspects involved in taking care of patients with esophageal atresia between January 2011 and June 2016, as well as the patients who were referred from other centers. Esophageal atresia is a complex anomaly that has many aspects that have to be dealt with and complications to be solved. By centralizing these patients in centers of expertise it is believed that the best care can be given.

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Introduction

Esophageal atresia has always been the hallmark of pediatric surgery. In the past, focus used to be on the correction of the malformation. In more recent years emphasis has shifted toward the complete spectrum of management of esophageal atresia and all its sequelae.^{1–6} INoEA is an International Network of all pediatric specialisms as well as patient organizations involved in dealing with all aspects of esophageal atresia and follow-up. It becomes more and more clear that the pediatric surgeon is merely a link in this chain. It is up to the pediatric surgeon to determine whether he or she wishes to be the spider in the web and serve as the coordinator for the management of children born with esophageal atresia.

Esophageal atresia has been the focus point in our department for many years, not only on treating the malformation,⁷ but also on managing sequelae like gastro-esophageal reflux disease,⁸ esophageal stenosis, and tracheomalacia.⁹ The department developed the jejunal interposition technique for long-gap esophageal atresia¹⁰ and was one of the forerunners starting up the thoracoscopic approach for managing esophageal atresia repair.¹¹ In 2007, the first thoracoscopic traction technique for long-gap esophageal atresia was described.¹² The department is a recognized center of expertise in the Netherlands today.

In 2012, the department published its 10-year outcome of thoracoscopic esophageal atresia repair,⁷ showing a learning curve over the first five years, and an improved outcome over the last five years, in spite of the fact that at that time most procedures were carried out by the junior staff members. This is a follow-up

that gives an overview of the department activities over the last six years since the last publication.

Patients

There is a standardized protocol for all patients admitted with esophageal atresia, including preoperative rigid tracheo-bronchoscopy and genetic counseling, and postoperative follow-up until transition into adulthood (Table 1). The pediatric surgeon coordinates all investigations.

Between January 2011 and June 2016, a total of 64 patients were admitted in the department with esophageal and/or tracheal anomalies. This included 15 referrals and/or second opinions from elsewhere. There were 47 children with type C esophageal atresia. Of the patients with long-gap esophageal atresia 5/10 also had a proximal fistula (type B). Five children had a pure type A atresia (Table 2). Nine of 16 patients with tracheomalacia were referred from elsewhere, 14 had esophageal atresia. Twenty-one children had concomitant anomalies (33%).

Principally, all children are operated thoracoscopically. The technique has been described earlier and will only be summarized here.^{7,13} All patients undergo a preoperative rigid tracheo-bronchoscopy to determine the presence and location of a fistula and occurrence of tracheomalacia, either by collapse of the tracheal rings and/or pars membranacea.

Thereafter the patient is turned into a left 3/4 prone position at the left side of the operating table to allow easy access into the thorax. The port position for type C and long-gap esophageal atresia is the same. Depending on age and weight a first 5 mm trocar is placed approximately 1 cm below and anterior to the scapula tip. The thorax is insufflated with 3–5 mm Hg CO₂ at 1–2 L/min. In a triangle around, two 3 mm trocars are placed under direct vision. After sufficient desufflation of the right lung

* Corresponding author.

E-mail address: d.c.vanderzee@umcutrecht.nl (D.C. van der Zee).

Table 1
Protocol esophageal atresia.

Summary work-up on admission in NICU
– Preoperative work-up
o IV drip, arterial line, and lab
o Replogle tube drainage
o Near-infrared spectrometry (NIRS), a-EEG
o Consultation pediatric cardiologist + echocardiogram
o X-Thorax/abdomen and ultrasound kidneys
o Consultation genetics
o Consultation ophthalmologist
o Other investigations depending on concomitant anomalies
– Preoperative multidisciplinary consultation on intended procedure
– After induction preoperative rigid tracheo-bronchoscopy
– Operative thoroscopic correction esophageal atresia (EA) under NIRS and a-EEG control
– Postoperative ventilation for 24–48 h on indication
– Start ranitidine
– Start feeding through nasogastric tube after 24 h
– Start oral feeding when no saliva and/or infectious symptoms
– Transfer to ward when stable and spontaneous breathing
– MRI brain after 1 week
– Life support course parents
– Discharge from hospital when on full enteral feeds and no other sequelae
Follow-up
– Follow-up after 2, 4, and 8 weeks or earlier when indicated
o Consultation with dietician (by telephone)
o Consultation with pediatric pulmonologist (yearly, antibiotic prophylaxis during winter if necessary, lung function test at 6 years)
o Consultation neonatologist (3, 6, 9, 12, 18, and 24 months when Bailey test)
o Consultation pediatric gastroenterologist (esophagoscopy in case of stenosis or reflux; standard Barrett investigation at 12 and 17 years)
o Consultation pediatrician
o Consultation other specialists on indication
o Regular follow-up at 3, 6, 9, and 12 months, then every 2 years thereafter

dissection is started from the distal fistula, which can be found by tracing the vagal nerve below the level of the azygos vein. If the azygos vein is in the way it is taken down with diathermia or the 3 mm sealer. After sufficient mobilization, a transfixing suture Vicryl® 4 × 0 (Medtronic, Eindhoven, Netherlands) is placed in the fistula at the level of where it enters the tracheal wall and tied.

The esophagus is transected distally. In case of a long-gap esophageal atresia the distal esophagus is usually withdrawn more distally and sometimes has to be dissected out of the hiatus, but again can be usually traced by following the vagal nerve downwards. Thereafter, the proximal esophagus can be dissected up to the aperture. By carefully pushing on the Replogle tube dissection can be facilitated. If a proximal fistula is present this is dissected and closed first, depending on the level, either through the neck or thoracoscopically. In type C the anastomosis can be made with Vicryl® 5 × 0. A first 10 cm suture is used for the posterior side close to the vertebral column. The proximal esophagus is hitched with the lumen still closed so that some pressure can be exerted while approximating the two ends. If necessary, a second suture can be placed on the opposite side to approximate the two ends with a sliding technique. After tying the first suture, this can be led outside to stabilize the anastomosis line while running the back wall with the second suture. This suture can be tied with the short end of the first suture. A transanastomotic tube can be advanced into the stomach. If there is enough length the same suture can run back over the front wall and tied of on the opposite side. Otherwise a new suture can be used. No drain is left behind. In case of type A or B atresia a primary anastomosis is not possible and traction sutures are applied on the proximal and distal esophagus with the use of an Endoclose® (Covidien, Eindhoven, Netherlands) device. The sutures are introduced and retrieved crosswise to exert traction on both the ends. At the level of the pouches a clip is applied to the sutures to follow the approximation radiologically. On the outside the sutures are pierced through a piece of silicone tubing with the Endoclose® and brought under tension with mini-Mosquitos. After finishing the thoroscopic procedure a laparoscopy is performed to fix the stomach against the ventral abdominal wall to prevent migration of the stomach into the thorax, what otherwise would be prevented by the gastrostomy, which nowadays is not given anymore, because the procedure is carried in the first week of life. In the following days only the tension on the sutures is checked, but no additional tension is applied to prevent the sutures from tearing out. Usually after 2–3 days there is no more approximation due to adhesion of the esophageal ends to the surrounding lung tissue. In that case the patient is taken back to theater and the adhesions are carefully taken down. In most cases after 1–2 days the esophageal ends have approximated sufficiently for making a delayed primary anastomosis. Depending on the ease of the anastomosis a drain may be left behind. Only in case of long-gap esophageal atresia a contrast study is made after five days, before starting oral feeds.

Follow-up is according to the protocol (Table 1).

Results

Type C esophageal atresia

There were 47 patients with type C esophageal atresia. In one child with psychomotor retardation postoperative leakage occurred requiring drainage for three days. In another extremely dysmature and premature child (32 4/7 weeks, 1470 g) born after fetal distress (APGAR 4, 7, and 9) through cesarean section with Vacterl association (ARM, VSD, polycystic kidney, and vertebral anomalies) the distal esophagus ended in the right main bronchus and could only be anastomosed with difficulty. She developed postoperative leakage requiring drainage. Seventeen patients required one or more dilatations postoperatively (36%). Four children ultimately underwent a laparoscopic antireflux procedure. In one child a persisting stenosis was ultimately resected thoracoscopically. In two children a recurrent fistula occurred after dilatation that was closed thoracoscopically. In the postoperative

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