Esophageal Atresia and Upper Airway Pathology

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KEYWORDS

• Esophageal atresia • Long gap esophageal atresia • Tracheoesophageal fistula
• Esophageal stenosis • Tracheomalacia • Recurrent fistula
• Gastroesophageal reflux disease

KEY POINTS

- Esophageal atresia is an anomaly with frequently occurring sequelae requiring lifelong management and follow-up.
- There is an increasing awareness of the anesthetic and surgical implications in the care of neonates with esophageal atresia.
- Because of the complex issues encountered, patients with esophageal atresia preferably should be managed in centers of expertise.

INTRODUCTION

With improved neonatal care focus in esophageal atresia, the current focus in outcomes has shifted from mortality to morbidity. Esophageal atresia is not merely a congenital malformation that warrants surgical treatment but is an anomaly with frequently occurring sequelae requiring lifelong management and follow-up.[1] The pediatric surgeon may play a key role in the management of children born with esophageal atresia, which implies not only using advanced techniques to restore continuity but also providing optimal perinatal care and follow-up into adulthood. In a recent publication from the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition–North American Society for Pediatric Gastroenterology, a joint article from the European and North American pediatric gastroenterology societies, guidelines were presented for the evaluation and treatment of gastrointestinal and nutritional complications in children with esophageal atresia.[1]

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In addition to paying more attention to follow-up, there is an increasing awareness of the anesthetic and surgical implications in the care of these newborns. Disquieting reports have recently been published on the possible negative effects on neurodevelopment in children born with noncardiac major congenital anomalies.[2,3] Perinatal brain monitoring has become a major issue in managing this cohort of patients.[4–7]

The Department of Pediatric Surgery at the University Medical Center in Utrecht is a center recognized by the government for the authors' expertise in esophageal atresia and upper airway management. Patients are referred from throughout the country and abroad for specific management or reiterative surgery and airway problems. Results were recently published.[8] This article describes the management of minimally invasive surgery in esophageal atresia and upper airway pathology.

PRENATAL DIAGNOSIS

Prenatal symptoms may be polyhydramnios detected on physical examination or ultrasound.[9,10] Routinely performed ultrasound during pregnancy may show regurgitation of amniotic fluid during swallowing or the absence of gastric contents, in particular in patients with type A atresia with no fistula.[9–12] Antenatal MRI has a high sensitivity for the confirmation of esophageal atresia.[10,12,13] The sensitivity is significantly higher in high-level fetal centers.[10,13]

POSTNATAL DIAGNOSIS AND WORKUP

There are principally 6 types of esophageal anomalies (**Fig. 1**). The most common sign of esophageal atresia is the inability to advance a nasogastric tube during postnatal care. Usually a plain thoracic and abdominal radiogram will show the curling of the nasogastric tube in the proximal esophagus. Additionally, air in the stomach and intestines indicates the presence of a distal fistula, whereas the absence of air indicates isolated esophageal atresia. On establishing the diagnosis, patients are admitted into the neonatal intensive care unit (NICU) and are given an intravenous (IV) drip, an arterial line, and a orogastric tube to empty the proximal esophagus (**Box 1**). Nowadays, all neonates have near-infrared spectrometry and α-electroencephalogram (EEG) to monitor brain oxygenation. Further immediate workup consists of consultation of the pediatric cardiologist to determine cardiac malformations and locate the side of the descending aorta as well as the performance of an ultrasound of kidneys and brain. Consultation of a geneticist, an ophthalmologist, and other pediatric specialists depending on concomitant anomalies are performed as necessary.

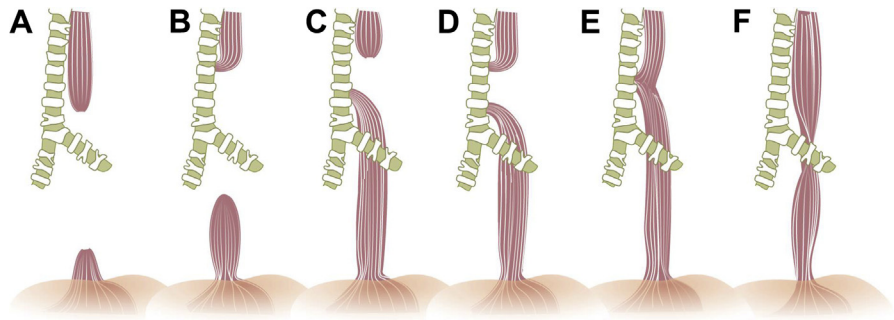

Fig. 1. Six types of esophageal atresia: type A, no fistula; type B, proximal fistula; type C, distal fistula; type D, both proximal and distal fistula; type E, only fistula; type F, esophageal stenosis.