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Perioperative management and outcomes of esophageal atresia and tracheoesophageal fistula

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ABSTRACT

Background/Purpose: Esophageal atresia/tracheoesophageal fistula (EA/TEF) is a rare congenital anomaly lacking contemporary data detailing patient demographics, medical/surgical management and outcomes. Substantial variation in the care of infants with EA/TEF may affect both short- and long-term outcomes. The purpose of this study was to characterize the demographics, management strategies and outcomes in a contemporary multi-institutional cohort of infants diagnosed with EA/TEF to identify potential areas for standardization of care.
Methods: A multi-institutional retrospective cohort study of infants with EA/TEF treated at 11 children's hospitals between 2009 and 2014 was performed. Over the 5 year period, 396 cases were identified in the 11 centers (7 ± 5 per center per year). All infants with a diagnosis of EA/TEF made within 30 days of life who had surgical repair of their defect defined as esophageal reconstruction with or without ligation of TEF within the first six months of life were included. Demographic, operative, and outcome data were collected and analyzed to detect associations between variables.
Results: Prenatal suspicion or diagnosis of EA/TEF was present in 53 (13%). The most common anatomy was proximal EA with distal TEF (n = 335; 85%) followed by pure EA (n = 27; 7%). Clinically significant congenital heart disease (CHD) was present in 137 (35%). Mortality was 7.5% and significantly associated with CHD ($p < 0.0001$). Postoperative morbidity occurred in 62% of the population, including 165 (42%) cases with anastomotic stricture requiring intervention, anastomotic leak in 89 (23%), vocal cord paresis/paralysis in 26 (7%), recurrent fistula in 19 (5%), and anastomotic dehiscence in 9 (2%). Substantial variation in practice across our institutions existed: bronchoscopy prior to repair was performed in 64% of cases (range: 0%–100%); proximal pouch contrast study in 21% (0%–69%); use of interposing material between the esophageal and tracheal suture lines in 38% (0%–69%); perioperative antibiotics ≥24 h in 69% (36%–97%); and transanastomotic tubes in 73% (21%–100%).
Conclusion: Contemporary treatment of EA/TEF is characterized by substantial variation in perioperative management and considerable postoperative morbidity and mortality. Future studies are planned to establish best practices and clinical care guidelines for infants with EA/TEF.
Level of Evidence: Type of study: Treatment study. Level IV.

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Esophageal atresia and/or tracheoesophageal fistula (EA/TEF) is an infrequently encountered congenital anomaly, wherein the esophagus fails to develop normally and blindly ends as a pouch in the neck or upper thorax. An abnormal communication from the trachea to the distal esophagus is commonly present. The worldwide incidence of EA/TEF is approximately one in 2500 to 4500 live births [1–4]. Without operative correction to establish esophageal continuity and divide the airway from the digestive tract, EA/TEF by itself is a life-threatening neonatal condition. While survival has dramatically improved over the last several decades, morbidity associated with EA/TEF and its associated anomalies remains high [1–3,5,6].

Historical data related to demographics, care and mortality of EA/TEF patients from single institutions need to be interpreted with caution because of small numbers and variations in practice with such a rare disorder. Recent reviews of EA/TEF management using large administrative databases, including the Pediatric Health Information System (PHIS) [5] and the Kid's Inpatient Database (KID) [6], are useful in describing epidemiology, survival, and variables associated with mortality. These studies are limited, however, by difficulty discerning essential clinical elements, including operative anatomic findings, long-term follow-up and the need for additional outpatient intervention.

We sought to provide a contemporary evaluation of the perioperative management of EA/TEF in the United States using the Midwest Pediatric Surgical Consortium, a collaboration of eleven academic children's hospitals. The study objective was to characterize the demographics, operative findings, management variation, and postoperative outcomes in infants with EA/TEF in order to identify potential areas for standardization of care.

1. Methods

1.1. Patients and study design

Following individual institutional review board approval, a retrospective cohort of infants diagnosed with esophageal atresia and/or tracheoesophageal fistula between 2009 and 2014 was identified across the eleven participating children's hospitals of the Midwest Pediatric Surgery Consortium (see Acknowledgments for list of centers). All patients with a diagnosis of EA/TEF (ICD-9750.3) made within 30 days of life who had surgical repair of their defect defined as esophageal reconstruction with or without ligation of TEF within the first six months of life (ICD-9 CM 31.73, 42.51, 42.85, 42.89, 43.19; CPT 43314 or 43,313, respectively) from January 1, 2009 to January 1, 2014 were included. Patients were identified from administrative hospital databases as well as practice databases. A minimum of one-year follow-up was obtained excluding those who died before the age of 1 year.

Study data were collected and managed using REDCap (Research Electronic Data Capture) software hosted at The Medical College of Wisconsin [7]. All study data entered were validated both centrally and at each individual institution for completeness of data entry and accuracy. Missing data were recollected and outliers were reconfirmed to be appropriate.

1.2. Methods

Collaborative consensus in identifying and defining relevant data elements for collection and analysis across the 11 institutions was obtained. Perioperative data collection included demographics, prenatal information when available, associated congenital anomalies, diagnostic imaging, physiologic status including American Society of Anesthesiology (ASA) Physical Status Classification at the time of operation, operative approach and findings, postoperative morbidity and mortality, and need for further intervention(s) up to one year following repair. Race/ethnicity was self-reported by patients during hospital registration. Congenital heart disease was defined as clinically identified cardiac defects other than patent ductus arteriosus and patent foramen ovale. Type of EA/TEF was categorized using the Gross five categories as determined at the time of surgical exploration for repair of the EA/TEF or ligation of the fistula (A–E classification) [8]. Long gap esophageal atresia was defined as an anatomic distance of three or more vertebral bodies between the proximal and distal esophageal segments determined on preoperative chest x-ray/esophageal pouch contrast studies or at the time of surgical exploration. Esophageal anastomotic stricture was defined as the need for dilatation of any type within one year of establishing esophageal continuity. Prenatal diagnosis or suspicion of EA/TEF was determined by reviewing the prenatal ultrasound reports for mention of polyhydramnios and/or absent/small stomach with mention of a possible diagnosis of EA within the report.

1.3. Statistical analysis

Statistical analysis was performed using SAS version 9.4 (SAS Institute Inc., Cary, NC, USA) and STATA (StataCorp. 2011. Stata Statistical Software: Release 12. College Station, TX: StataCorp LP). Continuous data were expressed as medians and interquartile ranges and discrete variables were expressed as percentages. For discrete variable comparisons, Pearson's Chi-Square Test was used. Fisher's Exact Test was used for associations of two binary variables which had small cell sizes. The Wilcoxon Rank-Sum test was used to compare medians. A p-value less than 0.05 was considered statistically significant.

2. Results

2.1. Study cohort

Over the five-year study period, 396 infants with EA/TEF were identified and analyzed from eleven children's hospitals. On average, 7 cases were identified per institution per year of the study; the cumulative number of patients enrolled per institution ranged from 16 to 56 total cases. The most common type of EA/TEF defect was proximal esophageal atresia with a fistula between the distal esophagus and trachea (type C) in 335 (84.7%) followed by pure EA (type A) in 27 (6.8%). The remaining distribution of EA/TEF types is shown in Table 1. Overall, EA/TEF was diagnosed or suspected prenatally in 53 patients (13.3%). Patients without a distal fistula (pure EA or esophageal atresia with proximal TEF) had a prenatal diagnosis established in 54% compared to only 10% in patients with a distal TEF (EA with a distal fistula or EA with proximal and distal fistula).

2.2. Patient characteristics

Of the 396 infants, 241 (61%) were male, and the race/ethnicity reflected the population of the Midwest United States with 308 (77.8%) Caucasian, 29 (7.3%) African American, 24 (6.1%) Hispanic, 9 (2.3%) Asian, 9 (2.3%) multiracial, and 17 (4.2%) other or not reported. One hundred ninety-nine patients (50%) had private insurance, 177 (45%) had public insurance including Medicaid, and 20 (5%) were self-pay or unknown. The median gestational age at birth was 37 weeks (interquartile range (IQR) 34–39) with a median birth weight of 2.6 kg (IQR 2.0–3.0). The median weight of infants at the time of surgical correction of their defect was lower in infants with a type C fistula (2.6 kg) as compared to those with type A, B, D and E defects ($p < 0.0001$).

Table 1
Distribution of anatomic findings using the Gross classification categories (n = 396).

	n (%)
Esophageal atresia with distal TEF (type C)	335 (84.7%)
Pure esophageal atresia without TEF (type A)	27 (6.8%)
TEF without esophageal atresia (type E or H-type)	18 (4.5%)
Esophageal atresia with proximal TEF (type B)	8 (2.0%)
Esophageal atresia with proximal and distal TEF (type D)	8 (2.0%)