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A nationwide analysis of clinical outcomes among newborns with esophageal atresia and tracheoesophageal fistulas in the United States

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ABSTRACT

Background: The aim of this study was to examine national outcomes in newborn patients with esophageal atresia and tracheoesophageal fistula (EA/TEF) in the United Sates.

Methods: Kid's Inpatient Database (KID) is designed to identify, track, and analyze national outcomes for hospitalized children in the United States. Inpatient admissions for pediatric patients with EA/TEF for kid's Inpatient Database years 2000, 2003, 2006, and 2009 were analyzed. Patient demographics, socioeconomic measures, disposition, survival and surgical procedures performed were analyzed using standard statistical methods.

Results: A total of 4168 cases were identified with diagnosis of EA/TEF. The overall in-hospital mortality was 9%. Univariate analysis revealed lower survival in patients with associated acute respiratory distress syndrome, ventricular septal defect (VSD), birth weight (BW) < 1500 g, gestational age (GA), time of operation within 24 h of admission, coexisting renal anomaly, imperforate anus, African American race, and lowest economic status. Multivariate logistic regression identified BW < 1500 g (odds ratio [OR] $= 4.5$, $P < 0.001$), operation within 24 h (OR $= 6.9$, $P < 0.001$), GA <28 wk (OR $= 2.2$, $P < 0.030$), and presence of VSD (OR $= 3.8$, $P < 0.001$) as independent predictors of in-hospital mortality. Children's general hospital and children's unit in a general hospital were found to have a lower mortality rate compared with not identified as a children's hospital after excluding immediate transfers ($P = 0.008$).

Conclusions: BW < 1500 g, operation within 24 h, GA < 28 wk, and presence of VSD are the factors that predict higher mortality in EA/TEF population. Despite dealing with more complicated cases, children's general hospital and children's unit in a general hospital were able to achieve a lower mortality rate than not identified as a children's hospital.

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1. Introduction

Esophageal atresia (EA) is a rare congenital anomaly characterized by an anatomical discontinuity of the esophagus. It may occur with or without tracheoesophageal fistula (TEF), which is characterized by an abnormal connection between the lumen of the esophagus and trachea [1]. The two disease entities are thought to have the same embryonic developmental origin, and they are found to accompany one another in most of the affected pediatric patients. The frequency of EA with or without TEF appears to be 1.27–4.55 per 10,000 live births [2–5]. Overall, 93%–94% of these patients present with a combination of both EA and TEF, whereas approximately 6%–7% of newborns manifest with solely EA [1–6]. Differentiation of these anatomical anomalies is essential to determine the correct medical management and surgical approach in this patient population [1,6–8].

Patients with EA/TEF oftentimes exhibit a syndromic presentation associated with other congenital and chromosomal abnormalities. Most commonly, patients may simultaneously present with VACTERL (vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb) syndrome, CHARGE (coloboma, heart, choanal atresia, retarded growth, genital hypoplasia, and ear abnormality) syndrome, Trisomy 18 (Edward syndrome), Trisomy 21 (Down syndrome), or a cardiac defect [1,3,5,9].

Currently, the only curative option for EA with or without TEF is surgical correction. However, since the initial operative management of a newborn with EA and TEF in 1939, there has been a tremendous improvement in neonatal critical care, surgical techniques, and anesthesia techniques including mechanical ventilation, which ensure the successful repair and decrease the rate of complications [10–14]. Notably, all these factors have attributed to the increase in survival rate of patients presenting with this condition [14–16]. In most of the current reports, a survival rate of more than 90% can be expected among neonates with EA and TEF when excluding neonates with lethal congenital anomalies. Furthermore, while low birth weight (LBW) and the presence of a cardiac anomaly yields a lower survival rate among affected newborns, full-term infants with no associated congenital anomaly may have a survival rate approaching 100% [3,4,15,17–20].

As the only all-payer inpatient care database for children in the United States, the Kid's Inpatient Database (KID) was specifically designed to identify, track, and analyze national outcomes for hospitalized children [21,22]. By examining the information provided in KID, we were able to identify high-risk patients based on socioeconomic status and ethnicity, along with determining national outcomes and factors predictive of survival in newborn patients with EA and TEF.

2. Methods

A retrospective analysis of KID 2000, 2003, 2006, and 2009 was performed to identify all incident cases of pediatric EA and TEF [22]. KID is one of a family of databases in the Healthcare Cost and Utilization Project, which was created by the Agency for Healthcare Research and Quality. The database contains information that is released every 3 y on more than 100 clinical and nonclinical variables for approximately 2–3 million hospital admissions per year in more than 2500–4000 community hospitals encompassing a total of 44 states. To compensate for missing states, all cases were weighted appropriately to project nationally representative statistical estimates. According to the American Hospital Association, the definition of a community hospital includes public hospitals, academic medical centers, and pediatric hospitals.

In addition to patient demographics, admission and discharge status, length of stay (LOS), total hospital charges, and hospital characteristics, KID includes up to 25 diagnostic and 15 procedural codes per discharge based on the International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM). Hospitals are classified according to the National Association of Children's Hospitals and Related Institutions types: not identified as a children's hospital (NIACH), children's general hospital (CGH), and children's unit in a general hospital (CUGH). KID uses systematic random sampling to select 10% of uncomplicated in-hospital births and 80% of complicated in-hospital births along with other pediatric cases from each participating hospital. In multiple instances, the KID registry has been used to report outcomes for a variety of congenital and acquired illnesses in children, as well as socioeconomic factors and resource utilization [23–28].

In this study, discharge records of patients aged <8 d at admission with a diagnosis of EA/TEF from the KID database were extracted using the ICD-9 code 750.3. An 8-d cutoff was selected to optimally target all interhospital transfers while excluding readmissions and reoperations. Because current ICD-9 code does not differentiate different types of EA/TEF, we attempted to group patients based on the surgical procedures they received (Table 4). Because different types of EA/TEF require distinct management plans from one another, this type of grouping may roughly represent different types of EA/TEF [1,6–8]. Furthermore, four principal surgical procedures including tracheal fistula repair, esophageal reconstruction (includes all surgical procedures performed to reconnect the esophageal gap), esophagostomy, and surgical feeding tube placement were used to further screen the patient population. We suggest that at least one of these four procedures would be required for the immediate survival of the patient. Those patients who did not undergo any of the four principal surgical procedures ($n = 1675$) were further analyzed. This group of patients can be attributed to either immediate transfer to other hospitals (84%) or death without surgical intervention (14%). A small subgroup of patients could not be accounted by the four principal surgical procedures, immediate transfer or death were identified ($n = 162$). All 162 patients' discharge statuses were labeled as "routine." Because of the confliction among their diagnosis, treatment received and discharge status, these patients were excluded from the analysis.

Patient demographics, clinical characteristics, socioeconomic measures, hospital type, and survival rate for this patient cohort was assessed. The primary outcome measure was survival at discharge. Of note, only percentages based on available data for each individual variable are provided. Missing data points were excluded from each respective univariate and multivariate analysis.