Upper Airway Anomalies in Congenital Tracheoesophageal Fistula and Esophageal Atresia Patients

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ABSTRACT

Objective: To examine the prevalence of upper airway anomalies in patients diagnosed with congenital tracheoesophageal fistula and esophageal atresia (TEF/EA).

Methods: A retrospective review was conducted of all TEF/EA patients seen at a tertiary pediatric hospital between January 2008 and December 2013. Inclusion criteria included evaluation by the otolaryngology service. Exclusion criteria included age > 18 years, acquired TEF/EA, subsequent rule out of TEF/EA, and otolaryngology evaluation for reasons not pertaining to the airway. Data including demographics, co-morbidities, presenting symptoms, surgical interventions, laryngoscopic and bronchoscopic examinations, and subsequent medical and surgical management were collected and analyzed.

Results: 139 children were included in the analysis. 56.1% (n=78) male, 43.9% (n=61) female. All patients underwent either flexible laryngoscopy or direct laryngoscopy and bronchoscopy. 4.3% patients (n=6) were diagnosed with laryngomalacia. 21.6% (n=30) were found to have vocal fold paresis or immobility. Laryngeal cleft was diagnosed in 25.9% (n = 36). 18 patients or 12.9% were diagnosed with subglottic stenosis. Tracheomalacia was the most common airway finding, diagnosed in 37.4% (n=52) patients.

Conclusion: Patients diagnosed with congenital TEF/EA have a high rate of secondary upper airway anomalies. Consideration should be given to perform a complete airway evaluation in all of these patients.

INTRODUCTION

Congenital tracheoesophageal fistula (TEF) is an abnormal communication between the esophagus and trachea that can be associated with esophageal atresia (EA). The overall incidence of TEF/EA ranges from 1 in every 2500 to 4000 live births.[1]

There is a high frequency of anomalies associated with TEF/EA which can be of great clinical importance. The most commonly associated malformations are cardiovascular, including Tetralogy of Fallot and ventricular septal defects, in 34% of cases.[2] TEF/EA can also be related to other congenital abnormalities, as part of the VACTERL association (vertebral, anal, cardiac, trachea-esophageal fistula, renal, limb defects).[3]

To our knowledge, there are no published reports that describe complete upper airway findings in TEF/EA patients. This study therefore examines the prevalence of upper airway anomalies in pediatric patients diagnosed with congenital TEF/EA. The diagnosis and management of these head and neck issues are discussed.

METHODS AND MATERIALS

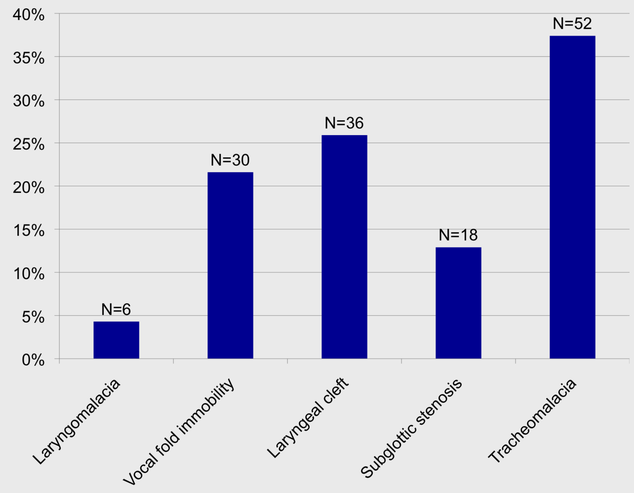
The Pediatric Health Information System (PHIS) database was queried for TEF/EA patients seen at Boston Children's Hospital (BCH) from January 2008 to December 2013. All patients with an ICD-9 code of 750.3 (tracheoesophageal fistula, esophageal atresia and stenosis) were searched.

Patients were included in the study if they were evaluated by the BCH otolaryngology service at any point in time. Exclusion criteria included age > 18 years or acquired TEF. Patients were also excluded if the diagnosis of EA and/or TEF was subsequently ruled out, or if patients were evaluated by the otolaryngology service for reasons not pertaining to the airway.

Data including demographics, co-morbidities, presenting otolaryngologic symptoms, TEF/EA surgical interventions, flexible laryngoscopic examinations, direct laryngoscopic / bronchoscopic examinations, airway findings, and subsequent medical and surgical management were collected and analyzed. The Boston Children's Hospital Institutional Review Board approved this study and its guidelines were followed.

RESULTS

Percentage of Secondary Airway Anomalies



- 430 patients identified with a diagnosis of TEF/EA between 1/2008 and 12/2013
- 139 patients who underwent complete otolaryngology diagnostic evaluation were included in analysis
- 85 patients had type C or type D TEF; 38.8% (n=54) had pure long gap EA
- All patients evaluated by the otolaryngology service underwent either bedside flexible laryngoscopy or direct laryngoscopy and bronchoscopy in the OR.
- In 13.9% (n = 19) patients, a tracheotomy was already in place at the time of their first otolaryngology evaluation. Inclusive of all the above airway abnormalities diagnosed, 6 patients, (4.41%) eventually required subsequent tracheotomy for airway management.

Patient Demographics and Comorbidities

	N	%
Demographics		
Gender		
Male	78	56.1%
Female	61	43.9%
Comorbidities		
Cardiac	92	66.2%
ASD/VSD	59	42.4%
Tetrology of Fallot	10	7.2%
Dextrocardia	5	25.2%
Right aortic arch	3	2.2%
Respiratory	88	63.1%
Asthma	40	28.8%
Bronchiectasis	4	2.9%
Gastrointestinal	97	69.8%
Imperforate Anus	8	5.8%
Reflux	93	66.9%
Intestinal atresia	2	1.4%
Diaphragmatic hernia	1	0.7%
Genitourinary	41	29.5%
Hypospadia	7	5.0%
Horseshoe kidney	5	3.6%
Polycystic kidney	1	0.7%
Renal agenesis	2	1.4%
Syndromes	82	59.0%
VATER/VACTERL	68	48.9%
CHARGE	7	5.0%
Chromosomal Abnormalities	9	6.5%
Trisomy 21	8	5.8%
Trisomy 18	1	0.7%

DISCUSSION

Despite the association of TEF/EA with anomalies of the respiratory tree, controversy exists regarding the need for a pre-operative bronchoscopy before TEF/EA repair.[4-10]

In this study, 139 patients were evaluated by the otolaryngology service for symptoms of aspiration, respiratory distress, and hoarseness.

The rates of secondary upper airway lesions were high with 87 patients, or 62.6% of the 139 patients, being diagnosed with an additional upper airway problem.

It is important to note that the prevalence of upper airway anomalies in this study is reported in TEF/EA children that were referred to the otolaryngology service. This was 139 patients out of a total of 430 patients at our institution diagnosed with TEF/EA. We accepted this selection bias as we know that airway examinations completed by different services focus on different findings.

Based on the findings of this study, we recommend complete airway examination in patients with congenital TEF/EA preoperatively. Given the rates of secondary airway anomalies, treatment and management decisions may be altered.

Additionally, we suggest collaboration between pediatric surgery and otolaryngology services to optimize diagnosis and treatment of symptomatic anomalies in the upper respiratory tract. We also suggest routine re-evaluations to assess the evolution of the upper airway conditions over time.

CONCLUSIONS

Patients diagnosed with congenital TEF/EA have a high rate of associated and secondary upper airway anomalies, including tracheomalacia, laryngeal cleft, and vocal fold immobility. Consideration should be taken to perform a complete airway evaluation in all of these patients pre-operatively and post-operatively. We also suggest further collaboration with otolaryngology and pediatric surgery services to optimize care for these medically complex children.

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