

Airway Anomalies in Neonates with Complex Congenital Heart Disease

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Abstract

The presence of severe airway anomalies in infants with congenital heart disease is linked to poor prognosis. Our short report describes the outcomes of seven infants with complex congenital heart disease and tracheoesophageal fistula with esophageal or tracheal atresia. We reviewed the prenatal and postnatal course of the mothers and infants to identify prenatal markers and overall outcomes. There was prenatal suspicion for tracheal anomaly in only two of the patients. Morbidity and mortality was high among our patients: four neonates died within the first days of life, and one of the three living infants required tracheostomy. Our report indicates a need for heightened awareness of potential airway anomalies in patients with complex congenital heart disease in order to improve prenatal diagnosis and counseling.

Keywords: Congenital heart disease; Tracheoesophageal fistula; Tracheal atresia; Esophageal atresia; Airway anomaly; Prenatal diagnosis

Abbreviations: CHD: Congenital Heart Disease

Introduction

The presence of airway anomalies such as tracheoesophageal fistula and tracheal agenesis greatly affects the morbidity and mortality of patients with congenital heart disease (CHD). These neonates typically have emergent symptoms of airway obstruction immediately after birth resulting in a difficult postnatal course. The mortality among patients with CHD and concurrent severe airway anomaly is significantly increased when compared to CHD patients without airway abnormalities [1-3]. Morbidity of surviving patients is high; complications may include need for tracheostomy, pulmonary hypertension, frequent hospitalizations, feeding difficulties, severe reflux and recurrent respiratory illnesses resulting in significant long term disability [3].

Prenatal diagnosis of tracheal and esophageal anomalies is difficult [4]. Despite advances in ultrasound technology, most patients continue to be diagnosed postnatally when neonates become symptomatic with excessive secretions, respiratory distress, cyanosis, difficulty feeding or inability to pass a nasogastric tube [5].

We describe a series of patients with prenatally diagnosed complex CHD who were diagnosed either prenatally or postnatally with tracheoesophageal fistula and esophageal or tracheal atresia. We aim to report the outcomes as well as the prevalence of prenatal markers of airway anomalies within this cohort in order to improve prenatal diagnosis of airway anomalies among all patients with CHD.

Methods

Approval was obtained through the Institutional Review Board at our institution. The Fetal Cardiac Database, which tracks all fetuses prenatally diagnosed with cardiac disease at our institution, was searched for all patients with prenatal or postnatal diagnosis of upper airway anomaly. The medical charts for each infant and mother were reviewed for prenatal findings on fetal echo and ultrasound, delivery course, and postnatal course including outcomes of death or need for tracheostomy. In addition, available fetal ultrasound images were reviewed by a maternal-fetal-medicine specialist for additional findings that may have been undetected at the time of the initial study.

Results

Seven infants born between 2001-2015 had both prenatally diagnosed complex CHD and prenatally or postnatally diagnosed tracheoesophageal fistula with tracheal agenesis and/or esophageal atresia. The findings in these patients are summarized in Table 1.

Patient	GA	Birth Weight	Heart Defect	Airway Anomaly	Other Prenatal Findings	Other Postnatal Findings	Outcome
A	37	2.38	TCA, PA	TEF/EA	None	Malrotation	Alive
B	36 1/7	2.74	DORV, HLH	TEF/EA, TA	Polyhydramnios, 2VC	Hemivertebrae, polydactyly	Died at 3 days
C	34 6/7	2.49	DORV, TGA	TEF, TA*	Polyhydramnios, 2VC, renal and skeletal abnormalities	Forearm malformation, imperforate anus, hydronephrosis	Died at 2 days
D	39 3/7	2.85	DORV, TOF	TEF/EA*	Absent stomach bubble, polyhydramnios, vascular, CNS and skeletal abnormalities	Rib fusion, dysgenesis of corpus callosum	Alive, tracheostomy
E	31	1.78	TOF	TEF/EA	Hydrops, 2VC, imperforate anus, genitourinary and skeletal abnormalities	Vertebral/rib anomalies, camptodactyly, ambiguous genitalia, imperforate anus, cloacal malformation, pelvic kidney	Died at 11 days
F	39 3/7	3.07	DORV, HLH, TAPVR	TEF/EA	None	Vertebral/rib anomalies	Died at 1 day
G	39 4/7	3.34	TA/IAA	TEF/EA	None	None	Alive

*: Diagnosed prenatally

Abbreviations: 2VC: 2 Vessel Umbilical Cord; CNS: Central Nervous System; DORV: Double-Outlet Right Ventricle; EA: Esophageal Atresia; GA: Gestational Age; HLH: Hypoplastic Left Heart; PA: Pulmonary Atresia;

TA: Tracheal Atresia; TA/IAA: Truncus Arteriosus with Interrupted Aortic Arch; TCA: Tricuspid Atresia; TAPVR: Total Anomalous Pulmonary Venous Return; TEF: Tracheoesophageal Fistula; TGA: Transposition of the Great Arteries; TOF: Tetralogy of Fallot

The cardiac diagnoses were variable but mainly involved conotruncal anomalies. Four patients had double outlet right ventricle variants: two with associated hypoplastic left heart, one with transposition of the great arteries and one with tetralogy of Fallot physiology. The remaining three patients had isolated tetralogy of Fallot, tricuspid and pulmonary atresia, and truncus arteriosus with interrupted aortic arch.

With respect to airway anomalies, all infants had tracheoesophageal fistula. Five patients had associated esophageal atresia, one patient had associated tracheal agenesis, and one patient had both esophageal atresia and tracheal agenesis. There was prenatal suspicion for tracheal anomaly in only two patients; one had an absent stomach bubble and severe polyhydramnios, and the other had severe polyhydramnios in the setting of renal and skeletal anomalies. In the remaining five infants with postnatal airway diagnoses, severe polyhydramnios was prenatally identified in one, and hydrops fetalis with genitourinary abnormalities, suspected imperforate anus and thumb abnormalities was diagnosed prenatally in another. Three infants had no extracardiac abnormalities identified on fetal ultrasound.

Fetal ultrasound images were available for retrospective review in five of the infants. In the infant with hydrops and multiple other anomalies (patient E), a small and poorly visualized stomach bubble was identified on retrospective review. Based on initial image reports, the patient's findings were thought to be secondary to a gastrointestinal malformation

and there was no mention of prenatal suspicion of an airway anomaly. For the remaining four patients with available fetal ultrasound images, the retrospective review was consistent with initial image reports; however, none of the patients had dedicated airway views available for review.

Postnatally, four of the infants had findings consistent with a VACTERL association. Two of the remaining three patients had additional extracardiac anomalies. All patients had normal chromosome karyotypes; patient E had a nonspecific chromosomal abnormality on microarray.

Mortality was high among this group of patients. Four of the seven infants died within the first few days of life. Two infants with tracheal agenesis had no surgical options for airway repair and support was withdrawn. The other two infants died from complications of necrotizing enterocolitis and respiratory failure. Of the three living infants, one required tracheostomy.

Discussion

Our patient series reinforces the previously described high morbidity and mortality of patients with complex CHD and severe airway anomalies. Patients with fetal diagnosis of conotruncal cardiac anomalies appear at higher risk. Based on retrospective review of the fetal ultrasound images in our series, the presence of severe polyhydramnios, especially with additional defects suggestive of an airway syndrome (such as VACTERL association) should raise suspicion for the presence of a severe airway

anomaly and prompt further airway evaluation. Thorough evaluation for airway abnormalities is ideally performed at a Maternal-Fetal-Medicine center experienced in the diagnosis of these anomalies utilizing dedicated airway views on ultrasound. Fetal MRI may also be useful in cases where airway abnormality is suspected on ultrasound but difficult to define.

Heightened awareness of potential airway anomalies in CHD patients can improve prenatal detection and lead to more specific and appropriate prenatal counseling and delivery planning for this high risk group of patients. Counseling regarding the high mortality and potential for immediate distress is important as this markedly differs from what is expected for infants born with CHD alone. Planning for delivery at a tertiary care pediatric center with immediate resuscitation and airway evaluation by appropriate specialists is warranted and may improve outcomes.

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References

1. Diaz LK, Akpek EA, Dinavahi R, Andropoulos DB (2005) Tracheoesophageal fistula and associated congenital heart disease: implications for anesthetic management and survival. *Paediatr Anaesth* 15: 862-869.
2. Güçer S, Ince T, Kale G, Akçören Z, Özkutlu S, et al. (2005) Noncardiac malformations in congenital heart disease: a retrospective analysis of 305 pediatric autopsies. *Turk J Pediatr* 47: 159-166.
3. Hayashi T, Inuzuka R, Shiozawa Y, Shindo T, Shimizu N, et al. (2013) Treatment strategy and long-term prognosis for patients with esophageal atresia and congenital heart diseases. *Pediatr Cardiol* 34: 64-69.
4. Pedersen RN, Calzolari E, Husby S, Garne E, EUROCAT Working group (2012) Oesophageal atresia: prevalence, prenatal diagnosis and associated anomalies in 23 European regions. *Arch Dis Child* 97: 227-232.
5. Lim FY, Crombleholme TM, Hedrick HL, Flake AW, Johnson MP, et al. (2003) Congenital high airway obstruction syndrome: natural history and management. *J Pediatr Surg* 38: 940-945.

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