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Managing concomitant congenital diaphragmatic hernia, esophageal atresia, and tracheoesophageal fistula: A case report of a premature infant that achieved survival☆

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ABSTRACT

Newborns with the constellation of congenital diaphragmatic hernia (CDH), esophageal atresia (EA), and tracheoesophageal fistula (TEF) present a unique clinical situation that requires well-coordinated multidisciplinary management as it is most commonly fatal. The authors describe successful management of a premature infant diagnosed with left CDH, EA, and TEF in the United States, the first such case to be reported in this country.

Few cases of coexisting congenital diaphragmatic hernia (CDH), esophageal atresia (EA), and tracheoesophageal fistula (TEF) are described in the literature, with most failing to survive the perinatal period. When associated with a congenital cardiac anomaly, the mortality is even greater. The rarity of this constellation of abnormalities is demonstrated by epidemiological studies based on the California Birth Defects Monitoring Program in 2004 demonstrating only 17 cases of patients with CDH, EA, and TEF, of which 16 were stillborn or died [1]. The following case report describes a premature infant who was successfully managed at the Children's Hospital of the University of Virginia Health System (Charlottesville, Virginia, USA) with left CDH, EA, TEF, and initially presumed coarctation of the aorta.

1. Case report

A premature male infant was born via precipitous spontaneous vaginal delivery at 34 weeks gestational age weighing 1.7 kg, with known left-sided CDH, diagnosed prenatally at 20 weeks gestation, and suspected congenital cardiac disease on prenatal echocardiography. Apgar scores were 3, 5, and 8 at 1, 5, and 10 min respectively. Immediately after birth the patient was intubated, started on high-frequency oscillatory ventilation, and admitted to the neonatal intensive care unit (NICU). An enteric tube was inserted by mouth but would not pass more than 11 cm, raising the suspicion for esophageal atresia. Chest x-ray (CXR) confirmed the presence of a large left congenital diaphragmatic hernia with multiple air-filled loops of bowel, as well as the enteric tube terminating in the mid-thoracic esophagus (Fig. 1). Esophageal atresia was confirmed on follow up x-ray with contrast injection (Fig. 2).

Further evaluation revealed normal appearing kidneys, no hydrocephalus or cerebral hemorrhage, but possible coarctation of the aorta with a large patent ductus arteriosus, bidirectional shunting, and a patent foramen ovale. The patient was administered prostaglandins. An attempt was made to prevent or minimize air passage through the TEF by advancing the endotracheal tube past the fistula. However, the stomach and intestines continued to distend, making ventilation increasingly difficult (Fig. 2). A decision was made to take the patient emergently to the operating room for surgical management. Via a transverse left upper quadrant laparotomy incision, the CDH was addressed first. The intestines were reduced out of the left chest. Similar to the approach described in the Journal of Pediatric Surgery in 1996 by Sapin E et al., the TEF was controlled with a Silastic (Dow Corning, Midland, MI) vessel loop as a Rommel tourniquet at the

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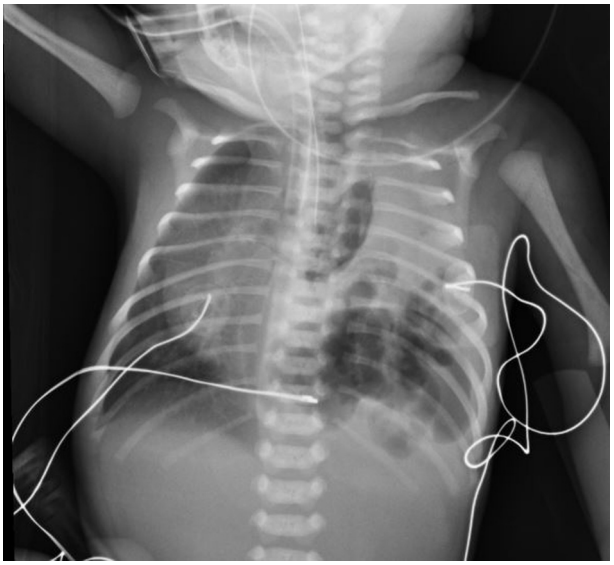

Fig. 1. Initial CXR showing herniated bowel contents in left hemithorax, as well as oral-gastric tube terminating in mid-esophagus.

gastroesophageal junction [2]. This was brought out of the right side of the abdomen through a stab incision. The posterolateral diaphragm defect was then closed with use of a Gore® Dualmesh (W.L. Gore & Associates, Inc, Newark, DE). A Stamm gastrostomy using a Malecot catheter was placed and brought out through a separate abdominal stab incision and connected to a one-way water seal valve. This allowed additional control of the trache-oesophageal fistula, as well as continued access to the stomach in case primary esophageal repair was not possible at the subsequent operation. Placement of the gastrostomy allowed for decompression of intra-luminal air from the distended intestine, which facilitated abdominal closure. The patient tolerated the procedure well and was maintained on mechanical ventilation in the NICU pending definitive TEF repair. Immediate post-operative CXR showed resolution of the left CDH with successful reduction of the herniated abdominal contents (Fig. 3).

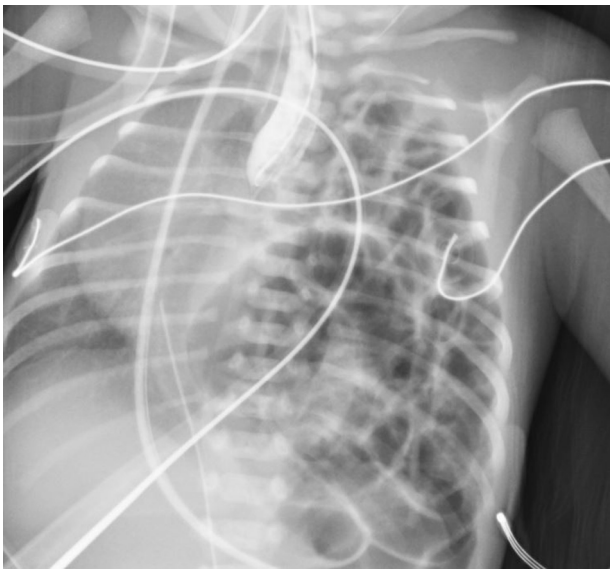

Fig. 2. CXR with contrast injection via oral-gastric tube demonstrating blind-ending esophageal pouch and worsening gaseous distension of small bowel.

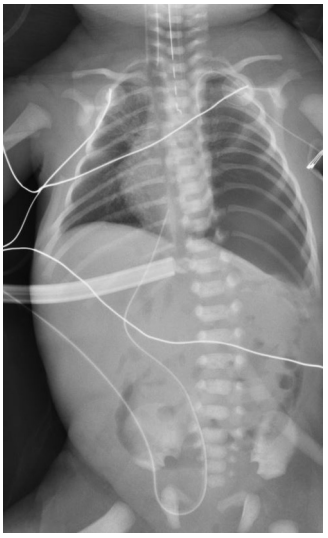

Fig. 3. Post-operative CXR showing repair of left-sided CDH.

After a trial of right-side up oscillatory ventilation, and a trial of conventional ventilation, that both demonstrated adequate left lung functionality, the patient was taken back to the operating room on day-of-life (DOL) 12. A left-sided chest tube was placed to drain a pleural effusion that had failed to resolve. Then, via a right thoracotomy, the tracheoesophageal fistula was ligated and a tension-free, primary end-to-end esophageal anastomosis was accomplished. A right-sided chest tube was placed and the patient returned to the NICU in stable, but critical condition.

He was transitioned to conventional mode ventilation on DOL 15. Trophic tube feeds were started via the gastrostomy tube on DOL 19. Prostaglandins were stopped and the left chest tube was removed on DOL 20. Cardiac echo on DOL 21 showed no aortic coarctation, a small PDA with left to right shunting, and a persistent left superior vena cava draining into a dilated coronary sinus. Upper GI contrast study showed no obstruction or leak at the esophageal anastomosis on DOL 24 (Fig. 4). The patient was extubated the same day. The right chest tube and NG tube were removed on DOL 25. Patient's respiratory status continued to improve and he was weaned to high-flow nasal cannula (HFNC) oxygen on DOL 28.

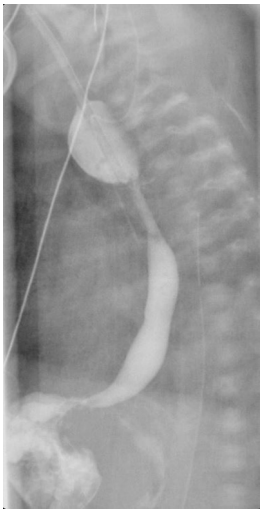

Fig. 4. Upper GI contrast study showing patent esophageal anastomosis without obstruction or leak.