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## Journal of Pediatric Surgery CASE REPORTS

journal homepage: [www.jpascasereports.com](http://www.jpascasereports.com)Congenital diaphragmatic hernia and complete tracheal rings:  
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## ARTICLE INFO

## Article history:

Received 6 September 2015

Accepted 12 October 2015

## Key words:

Congenital diaphragmatic hernia

CDH

ECMO

Complete tracheal rings

Slide tracheoplasty

## ABSTRACT

A term newborn female presented with prenatally diagnosed congenital diaphragmatic hernia and postnatally diagnosed complete tracheal rings and tracheal stenosis. Initially, the spells associated with tracheal stenosis were misdiagnosed as pulmonary hypertension. Bronchoscopy showed a critically narrowed airway, and veno-arterial ECMO stabilized the baby for further workup. The endotracheal tube was removed while on ECMO to avoid further injury to the airway. Staged CDH repair followed by slide tracheoplasty were performed on ECMO. The multidisciplinary approach included neonatology, general surgery, otolaryngology, cardiac surgery and anesthesiology.

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Congenital diaphragmatic hernia (CDH) associated with complete tracheal rings is a very rare combination with only one other case reported in the literature [1]. Management of CDH frequently requires extracorporeal membrane oxygenation (ECMO) in patients with severe pulmonary hypoplasia with associated pulmonary hypertension for pre-operative stabilization [2,3]. Neonatal repair of complete tracheal rings is commonly done utilizing cardiopulmonary bypass [4]. We present a unique case involving the repair of a CDH and complete tracheal rings on ECMO.

## 1. Case report

A 26-year-old G6P2133 pregnant woman underwent evaluation at 27 4/7 weeks gestation at the Center for Fetal Diagnosis and Treatment. High-resolution prenatal ultrasound showed left CDH

with stomach and bowel within the left chest, heart displaced to the right, and liver within the abdomen. Right lung area to head circumference ratio (LHR) measured 2.9 and an observed to expected LHR was 1.09. The left kidney was absent and there was evidence of interdigital webbing on both hands. Fetal echocardiography demonstrated normal cardiac structures and function. Ultrafast fetal body MRI observed to expected total lung volume (O/E TLV) measured 0.79. There were no signs of tracheal obstruction or stenosis. Microarray was consistent with a normal female.

At 38 2/7 weeks the mother had cervical ripening and induction of labor in the Special Delivery Unit (SDU) at our institution. The newborn was intubated by 3.5 min of life. APGARS were 5 at 1 min and 9 at 5 min. An anterior anus was found in addition to the known prenatal findings of a left CDH. The patient was weaned to minimal ventilatory support in the first several days of life. Prior to CDH repair, she started to have desaturation spells, marked by severe cyanosis requiring positive pressure ventilation with high pressures and sedation. ECHO and physical examination were not compatible with pulmonary hypertension (PHT) crises. During an acute event on day of life (DOL) 12 the patient had complete loss of chest wall movement despite paralysis, and high frequency oscillatory ventilation (HFOV) resulting in profound hypercarbia and respiratory

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acidosis. Hyperextension of the neck in preparation for ECMO cannulation resulted in marked improvement of chest wall movement and ventilation. Thus, cannulation was deferred.

Rigid bronchoscopy showed distal tracheal stenosis with complete tracheal rings and small main stem bronchi. Narrowing was initially considered non-critical. However, a repeat rigid bronchoscopy one week later (DOL 19) just prior to planned CDH repair showed progression with edema, thick secretions and near complete stenosis (Fig. 1). Given the precarious nature of the airway and impaired ventilation, the decision was made to place the patient on ECMO and subsequently repair the CDH followed by repair of the trachea. Veno-venous ECMO cannulation was technically not possible due to the vessel size; therefore veno-arterial (VA) cannulation was performed with 10 Fr. cannulae. Given the concern for mechanical irritation from the endotracheal tube (ETT), the patient was extubated to a RAM Nasal Cannula® (Neotech™ Products Inc., Valencia, CA, USA) on ECMO day 1 (Fig. 2).

On ECMO day 7, primary left CDH repair was performed. Intraoperative findings included a thick hernia sac, containing small bowel, colon, stomach and spleen. The airway was managed with a laryngeal mask airway. On ECMO day 12, the patient was reintubated with a 2.0 uncuffed ETT (placed through the stenotic section of trachea) to remove thick mucous and facilitate lung recruitment for a computed tomography angiogram (CTA) of the chest to evaluate the airway anatomy prior to slide tracheoplasty. CTA showed anomalous takeoff of right upper lobe bronchus from the trachea and no pulmonary artery sling. Slide tracheoplasty was performed through a median sternotomy the following day. Repair involved a 2 cm long tracheal stenosis, 3 mm at its narrowest, starting 4 rings inferior to the cricoid cartilage and extended distally to 1.5 cm above the carina (Fig. 3). The patient returned to the NICU post-operatively on ECMO with a 3.5 ETT and patent airway. On ECMO day 16 the patient was decannulated and she was extubated to high flow nasal cannula 15 days later. On postoperative day 29 from tracheal surgery, rigid bronchoscopy showed reconstructed, widely patent airway (Fig. 4). The patient was discharged home on room air and nasogastric tube feedings.

## 2. Discussion

The combined diagnosis of CDH and complete tracheal rings is exceedingly rare with only one other reported case in the literature



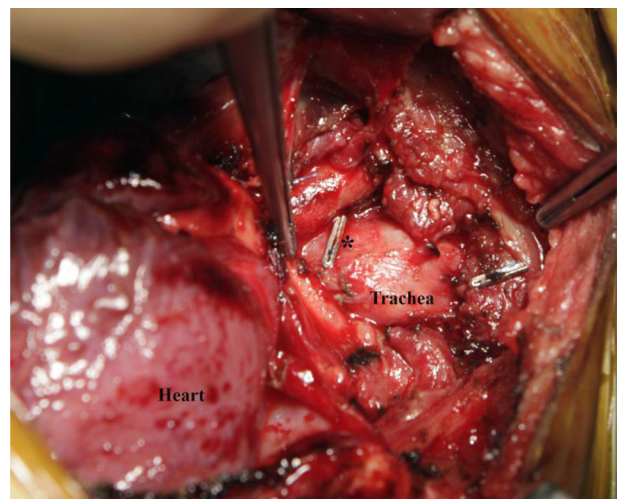
**Fig. 1.** Bronchoscopy on day of life 19 showed complete tracheal rings, edema and near complete stenosis.



**Fig. 2.** The patient was extubated to a RAM Cannula® while on veno-arterial ECMO support.

[1]. Longaker and colleagues described a patient with initial CDH patch repair and subsequent 2/3 tracheal resection to repair the complete tracheal rings. Both repairs were performed without ECMO. The patient was ventilated across the operative field for the tracheal repair. Tsang et al. described the first tracheoplasty in 1989 without the use of ECMO [5]. Patients with complete tracheal rings usually do not necessitate preoperative stabilization with ECMO, unless they present with near complete stenosis and life threatening airway obstruction [6,7]. Although preoperative ECMO is not required, recent reports suggest that slide tracheoplasty should be performed on cardiopulmonary bypass if possible [4].

Our patient had progressive narrowing of the airway over the first days of life. The appearance of stenosis on initial bronchoscopy was considered noncritical, but became increasingly edematous and erythematous. The presence of the CDH necessitated mechanical ventilation and likely potentiated the airway problems. Extubation following ECMO cannulation helped to decrease inflammation prior to repair. CDH repair was performed first in order to avoid further manipulation of the airway and mediastinum once tracheal repair was performed. Stabilization with ECMO also allowed careful study of the airway prior to surgical repair. The



**Fig. 3.** Intraoperative image after completed slide tracheoplasty on veno-arterial ECMO support. The clip marked with \* is the distal end of the tracheal anastomosis.



**Fig. 4.** Bronchoscopy on postoperative day 29 after slide tracheoplasty shows a widely patent airway.

patient was reintubated for the CTA of the chest to optimize lung recruitment and visualization of mediastinal structures. The CTA showed an anomalous right upper lobe bronchus originating directly from the trachea; a finding described in 20% of patients with tracheal stenosis [8].

In pediatric and adult ECMO, patients are sometimes maintained extubated on cardiopulmonary bypass [9,10]. Early extubation after slide tracheoplasty predicts a smoother recovery period [11]. Our patient was extubated on postoperative day 18 following tracheoplasty requiring prolonged mechanical ventilation due to the associated CDH. The timing of surgical repairs was deliberate. The multidisciplinary approach to this patient with close collaboration and co-ordination between Neonatology, Anesthesiology, General Surgery, Otolaryngology and Cardiac Surgery was key to successful short- and mid-term outcome for this critically ill infant [12].

### 3. Conclusion

The combined management of CDH and complete tracheal rings was successful with a staged multidisciplinary approach and the

use of ECMO. Consider this rare combined diagnosis in any patient with a CDH in whom there is also a clinical picture of airway obstruction.

### Conflicts of interest

None.

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