Preterm Newborn with Isolated Congenital Tracheal Stenosis: A Very Rare and Fatal Condition

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Abstract: Congenital tracheal stenosis is a lethal and very rare congenital malformation with an estimated incidence of two per 100,000 newborns. It leads to functional atresia of the trachea. It usually leads to infant death if there is no accessory connections like tracheo-oesophageal fistula to allow for spontaneous breathing. We report a male infant born at 32 weeks of gestation presented with respiratory distress immediately after delivery due to congenital tracheal stenosis. The infant was discharged successfully and now in regular follow up with adequate post natal growth.

Keywords: Preterm infant, Congenital tracheal stenosis.

INTRODUCTION
Tracheal stenosis leading to functional atresia of the trachea is a very rare and lethal congenital tracheal malformation. The associated malformations usually seen with it are anomalies of the vertebral anomalies, anal atresia, cardiovascular anomalies, tracheo-oesophageal fistula, esophageal atresia, renal/radial anomalies, and limb defects (VACTERL associations)—and tracheal agenesis, cardiac, renal and duodenum malformations (TACRD association) [1]. Affected infant presents with severe respiratory distress just after birth. Without associated tracheo-oesophageal fistula (TEF), severe tracheal stenosis results in functional atresia of the trachea which is lethal to life and usually results in infant death within first minutes of life despite all efforts for neonatal resuscitation.

We report on a preterm neonate with severe congenital tracheal stenosis resulting in functional tracheal atresia. The child developed respiratory distress immediately after birth. It is novel case with infant having absence of TEF at birth, in spite of that we were able to save the neonate and discharge successfully.

CASE REPORT
A male infant was born at 32 weeks of gestation to a 20 year-old primi gravida, by cesarean section because of preterm labor pain and fetal distress. Pregnancy was uncomplicated and the Apgar scores were 6, 7 and 8 at 1, 5 and 10 minutes, respectively. The infant developed respiratory distress and high oxygen demand immediately after birth and was admitted to the neonatal intensive care unit with nasopharyngeal continuous positive airway pressure support. On admission, the patient presented with respiratory acidosis with hypercapnia with a pCO₂ of 70 mmHg. Because of persisting respiratory distress and progression of CO₂ retention, the decision was made to intubate and ventilate the infant. Direct laryngoscopy revealed a clearly visible glottis plane with unaffected vocal cord mobility. But it was impossible to advance tracheal tubes of various sizes beyond the vocal cords. Hence baby was intubated with endotracheal tube no 2.5 and fixed just above vocal cords. Thereafter baby started maintaining saturation hence urgent com scan was done which revealed tracheal stenosis just below vocal cords till just above carina but internal diameter of 5mm both above and below stenosis. Hence again intubation was tried with endotracheal tube no 3 with stylet, which after manipulation was able to pass through stenotic area.

After stabilisation bronchoscopy was done which was suggestive of stenosis of complete tracheal ring from 2 to 5 ring and revealed congenital high-grade stenosis of the trachea directly distal to the vocal cords over a distance of more than 1 cm. There was no additional malformation except that the normal architecture of the proximal trachea was replaced by fibrous connective tissue the severely stenotic lumen of the distal part of trachea was surrounded by compact cartilage. Child was managed symptomatically and weaned off ventilator and discharged on breast feed, nebulization with steroid and advised followup. One month follow up revealed child to be stable and gaining weight normally with intermittent stridor.
DISCUSSION

Tracheal malformations including severe congenital stenosis and atresia are very rare malformations and have estimated incidence of two per 100,000 live births [2]. In the medical literature less than 100 cases have been reported till yet and the first case description was done by Payne in 1900 [3]. In the first case report tracheal malformation was associated with esophageal atresia proximal to the TEF.

The aetiology of tracheal atresia and tracheal agenesis is still unclear, but it is postulated that abnormal epithelial–mesenchymal interactions may be responsible for it as embryo logically respiratory tract develops from the ventral wall of the foregut in the third embryonic week [4]. Hence, tracheal atresia can be thought as a result of defective foregut differentiation, whereas tracheal atresia results from faulty development of an already differentiated anatomical structure.

Due to associated malformations usually seen in infants of tracheal agenesis, some authors refer to tracheal atresia as part of the VACTERL association spectrum [5]. More recently in literature, few cases of tracheal atresia have been reported with other associated malformations described as TACRD association [4, 5]. In most of the cases of tracheal atresia, usually an associated TEF is present; less cases without TEF have been published [1].

In the infant, in this case report, neither fit into Floyd’s classification of tracheal atresia nor any associated TEF/VACTERL/TACRO anomalies were found. Thus it is one of the rare cases, where an isolated tracheal atresia occurred. In contrast to our patient, in none of the reported cases without TEF, a residual lumen of the trachea was described in literature. Reviewing the published literature on congenital tracheal malformations, the reported case is unique for two reasons: firstly no other malformations were found on examination and investigation and secondly, a small residual lumen of the patient’s trachea was evident.

Tracheal atresia is one of the rarest tracheal anomaly seen. It should be suspected in pregnancies with a history of polyhydramnios and infant with severe respiratory distress without an audible cry and difficult intubation. There is a sexual predilection with male to female ratio 2:1 and an association with prematurity. In the reported case, the bronchoscopic examination revealed a severely stenotic trachea with an endoluminar width that extended from the subglottic region to the bifurcation.

If the diagnosis of tracheal atresia is suspected prenatally, fetal ultrasound and fetal MRI can be used to confirm the diagnosis antenatally. The foetus who are diagnosed antenatally with tracheal agenesis should be delivered in tertiary care hospital where all specialist care is available as the infant is in severe emergency condition after birth. From the results of the MRI, a decision should be made to employ an ex utero intrapartum tracheotomy (EXIT) to ventilate the lungs of the neonate. EXIT tracheotomy will be only successful in cases with a patent distal trachea [6, 7].

CONCLUSION

Tracheal atresia is a rare congenital malformation and the management of neonates with tracheal atresia pose a challenge to neonatologist. The resuscitative management could be improved with the diagnosis of tracheal atresia being made prenatally. There is no definitive treatment at present but tracheal tissue generation by tissue engineering gives ray for hope for future.

REFERENCES