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A Case Series On Tracheal Atresia

Dr Apoorva R M, Dr S.G. Matti, Dr Girish Hiremath
(Junior Resident, Department Of Paediatrics, Koppal Institute Of Medical Sciences, Koppal, India)
(Professor And Hod, Department Of Paediatrics, Koppal Institute Of Medical Sciences, Koppal, India)
(Associate Professor And Hod, Department Of Paediatrics, Koppal Institute Of Medical Sciences, Koppal, India)

Abstract:
Background*: Agenesis, aplasia or atresia of the trachea is a rare congenital anomaly that to date seems to be incompatible with life. Tracheal atresia is a congenital absence of a normal passage that does not imply a particular length of involvement. Tracheal atresia encompasses cases of agenesis (total absence of trachea) as well as varying lengths of tracheal mal-development[1]. We describe here 3 cases of tracheal agenesis.*
Materials and Methods*: It is a descriptive case series done with three neonates in tertiary care teaching hospital in Department of Paediatrics.*
Conclusion: *tracheal atresia is a rare anomaly and clinicians need to be aware of this condition to provide adequate supportive management at the time of delivery and subsequently.*
Keyword: *congenital anomalies; intubation; tracheal agenesis.*

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I. Introduction

Tracheal atresia (TA) is a rare and lethal congenital anomaly, in which there is a complete interruption or absence of the trachea. It was initially described in 1900 and since then only a few cases have been published worldwide[1]. The prevalence of tracheal atresia is less than 1:50,000 with a male to female ratio of 2:1. In general, 52% of cases are associated with premature delivery and approximately half of the cases are associated with polyhydramnios[2]. Tracheal atresia should be included in the diagnosis when the following clinical signs are manifested: neonate with history of polyhydramnios, absence of audible breath sound at birth, failure to intubate beyond vocal cords and respiratory distress. Till date only one patient has survived beyond neonatal age.

II. Material And Methods

This is a descriptive study which was carried out in Department of Paediatrics at Koppal Institute of Medical Sciences done over duration of three months from October 2022 to December 2022.

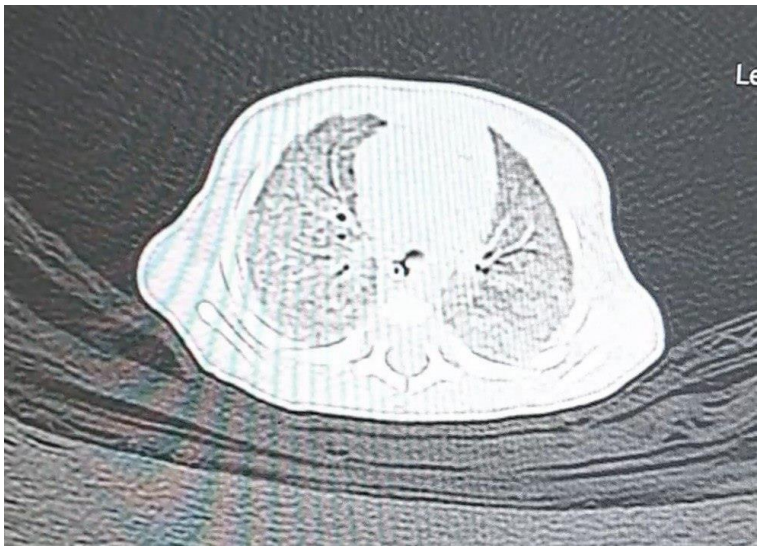
Study Design: A descriptive study

Study Location: This was a tertiary care teaching hospital-based study done in Department of Paediatrics at Koppal Institute of Medical Sciences, Koppal, Karnataka.

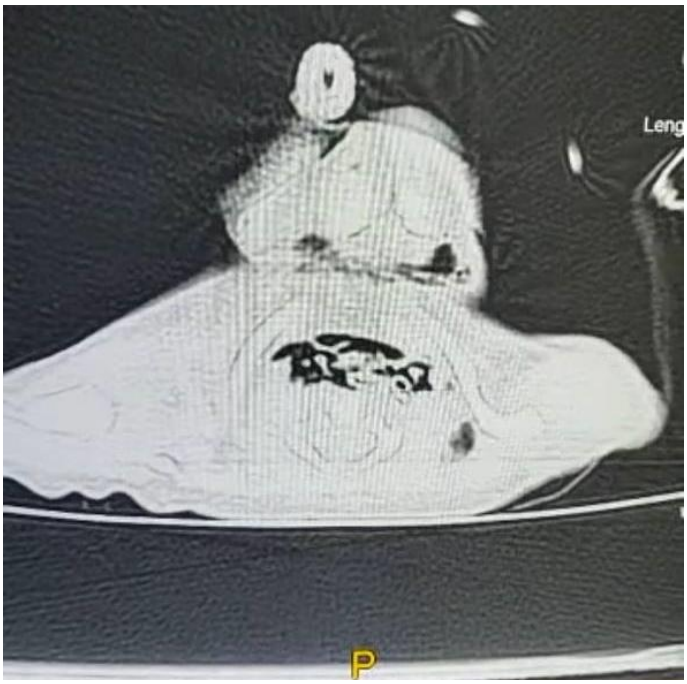
Study Duration: October 2022 to December 2022.

Sample size: 3 patients

Case 1: A 2.1kg male neonate was born at 36 weeks of gestation via caesarean section to a primigravida mother, with complication of polyhydramnios. At birth neonate was apnoeic. Hence resuscitation steps were taken. Intubation was attempted, larynx visualised, but on repeated attempts with 3.5, 3 and 2.5 et tubes, it was unsuccessful. Hence bag and mask ventilation continued. Nasogastric tube was passed successfully. The neonate expired due to worsening respiratory insufficiency and bradycardia unresponsive to cardiopulmonary resuscitation. Ct scan was performed to ascertain the cause of intubation failure which revealed complete absence of trachea below larynx, normal carina and right and left main bronchi with small fistulous communication (diameter 1mm) between anterior wall of Oesophagus and carina suggesting type 2 TA (Floyd's classification).



Case 2: A 2.25kg female neonate was born at 37weeks 5 days of gestation via caesarean section to a gravida 2 mother. There was no risk factor in the mother like polyhydramnios. At birth neonate did not have spontaneous respiration. Neonatal resuscitation steps were carried. As per neonatal resuscitation protocols intubation was attempted, under vision vocal cords were seen and multiple attempts to pass endotracheal tube were done, but it was unsuccessful. Nasogastric tube was passed. Bag and mask ventilation continued. The neonate could not be survived even after all cardiopulmonary resuscitation measures. Ct scan report – trachea not visualised separately with normally developed carina and right and left main bronchi s/o tracheal agenesis and a small fistulous communication between anterior wall of Oesophagus and carina suggesting type 2 TA (Floyd's classification) with sub cutaneous emphysema.



Case 3: A 2.05kg female neonate was born at 35weeks 4 days of gestation via caesarean section to a primigravida mother who had polyhydramnios. The neonate did not cry on birth and neonatal resuscitation measures were started which were futile. Nasogastric tube could be successfully passed. Under vision with vocal cords visualised intubation was attempted. As there was intubation failure with possible sizes of endotracheal tube, emergency tracheostomy was done, which was again futile. Tracheal rings were not present and no hollow structure found. The neonate expired due to worsening respiratory failure. Ct scan done – trachea and carina not visualised. Main bronchi is seen arising from anterior wall of oesophagus, suggests type 3 TA according to Floyd's classification.

III. Discussion

Tracheal Atresia is rare congenital upper-airway malformation[3]. Embryologically an aberrant (ventral or dorsal) septation is thought to be responsible for tracheal atresia/agenesis[4]. Prenatal US screening is the first exam that can possibly alert the clinician to a diagnosis. The classical signs could be enlarged hyperechogenic lungs, fluid-filled dilated trachea, and bronchi with absent flow in the trachea during breathing with or without cardiac dysfunction, diaphragmatic flattering, and massive ascites[5]

Postnatal diagnosis can be made by recognition of a combination of clinical signs: respiratory distress with breathing movements without appropriate air entry and absence of audible cry. Preterm labor, low birth weight (<2500 g), and polyhydramnios are often noticed. The newborn desaturates quickly, becoming bradycardic, while clinicians face the unanticipated impossibility of endotracheal intubation. Bag-valve-mask ventilation allows for temporary oxygenation, but still ventilation remains unsatisfactory as airflow resistance is high. However, bag-valve-mask ventilation leads to progressive gastric distention, which in turn lowers pulmonary compliance, further impairing ventilation and ultimately leading to the need for repeated gastric decompressions. Once other difficult airway causes have been ruled out and more advanced intubation techniques fail, if a TA is suspected and the patient improves with bag mask ventilation, the best option to secure the airway, pending further evaluation, is esophageal intubation. To secure the airways for a longer period insertion of a tracheal tube through the esophagus into the fistula or positioning a temporary clip at the gastroesophageal junction by a laparotomy and a preemptive gastrostomy tube can be performed. Next, an emergency cervical exploration should be attempted to perform a tracheotomy. Nonetheless, treatment of TA is always challenging as there is no clear recommendation on the best approach, and there is little time for airway assessment and intervention before severe hypoxic brain damage occurs[6].

TA was first described by Payne in 1900 and later classified into three types by Floyd. Floyd classification: (**Type 1**) The proximal trachea is atretic, the carina is normal, and the distal tracheal segment is connected to the esophagus through a fistula; (**Type 2**) the trachea is absent, and the main bronchi join at the carina. A carino-esophageal fistula is almost inevitably present; (**Type 3**) the trachea and the carina are missing, and the main bronchi directly join the esophagus[7]. The relative frequency of these types I, II, and III is 13%, 62%, and 25% respectively[8].

In our case series first two cases had type 2 anomaly where as third case had type 3 anomaly. They were either preterms or early terms. Two cases were associated with polyhydramnios. None of the patient could be survived. Certainly, if a fetal MRI is performed after ruling out other causes of polyhydramnios and if a prenatal diagnosis is made, pregnancy counselling is mandatory on whether the fetus will be compatible with life or no. Also a tertiary care management can be advised[9]. However, the surgical management of neonates with TA is difficult. A systematic surgical approach does not exist but limited success of surgical management is reported and despite progress of surgical developments, the prognosis is poor. Limited reports are available on long term survival in patient with TA[10].

IV. Conclusion

Till date only one patient of type II Floyd classification has survived beyond neonatal age. Currently this anomaly is incompatible with life. Clinicians need to be aware of such congenital airway malformations, as prenatal diagnosis will aid termination or plan for a multidisciplinary management of delivery. Whenever prenatal diagnosis is not possible, difficulty during intubation and other clinical features should immediately alert the clinician of a possible airway malformation as immediate resuscitation and palliative surgery can improve the longevity of the neonate. Wide research on the surgical management is still to be done as current methods are not favourable for sustaining life.

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