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Radiology

IMAGING IN TRACHEAL AGENESIS - A CASE REPORT

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ABSTRACT Tracheal agenesis, a rare congenital anomaly with an unknown etiology1, involves partial or incomplete underdevelopment of trachea. It is often associated with maternal polyhydramnios, premature delivery, trachea-oesophageal or broncho-oesophageal fistula. Type I tracheal agenesis is characterized by agenesis of the proximal trachea and by the presence of a distal tracheoesophageal fistula, whereas type II is defined by a complete absence of the trachea and by the presence of normal bifurcating bronchi. In type III, the two main bronchi arise independently from the esophagus2.We report a case of a newborn baby with severe respiratory distress, with no audible crying immediately after birth. CT was done due to challenges encountered during intubation.

KEYWORDS :

INTRODUCTION:

Complete tracheal agenesis (TA) is a rare and often lethal congenital anomaly with only 200 cases reported in literature. Its incidence is approximately 1in 50,000 births[3].

It is characterized by complete or partial absence of trachea below the larynx with or without tracheoesophageal fistula. Clinical presentation is characterized by rapid onset of severe respiratory distress at birth, absence of audible crying, and impossibility of endotracheal intubation.

According to recent theories lower respiratory tract develops as a respiratory diverticulum from the ventral aspect of the foregut which then elongates caudally to form the trachea. Arrested elongation of the trachea results in type 1 TA while arrested elongation with fusion to form carina results in type 2 TA and without fusion to type 3 TA. The exact etiology is not known but genetic factors have been implicated.

CASE DETAILS:

A newborn male baby of 1820 g was born at 34 weeks 6 days of gestation to a primigravida by C-section. She immediately showed severe respiratory distress and no audible crying. Intubation was difficult despite good visualization of the glottis. Polyhydramnios was present in 30 weeks and 34 weeks of antenatal screening Chest radiograph for initial screening and HRCT chest was advised in view of difficult intubation.

IMAGING FINDINGS:

The chest radiograph shows normal aerated lungs. The endotracheal tube is slightly low but otherwise is unremarkable. The nasogastric tube also is present and its distal end is in the stomach (Figure 1)

CT showed no tracheal air from the level of the glottis to the carina with normal main bronchi fused in the midline and communication between the esophagus and carina representing a Floyd type 2 tracheal agenesis. The endotracheal tube and Nasogastric tube are seen in one common lumen (dilated esophagus) beyond the glottis (figures2,3,4,5).

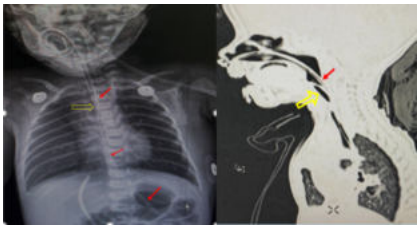


Fig1: Chest radiograph shows normal aerated lungs with endotracheal tube(yellow arrow),nasogastric tube(red arrow) with distal end in the stomach and chest leads.

Fig2: CT saggital section showing nasogastric tube (red arrow) and the endotracheal tube (yellow arrow) in larynx.

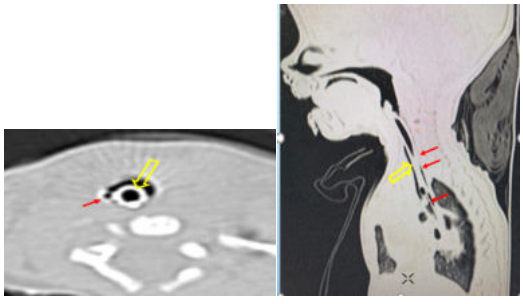


Fig3 :CT axial showing the nasogastric tube(red arrow) and the endotracheal tube(yellow arrow) side by side in the proximal dilated esophagus.The trachea is absent.

Fig4 : CT saggital section showing the nasogastric tube(red arrow) and the endotracheal tube(yellow arrow) side by side in one common lumen (i.e dilated esophagus). The trachea is absent.

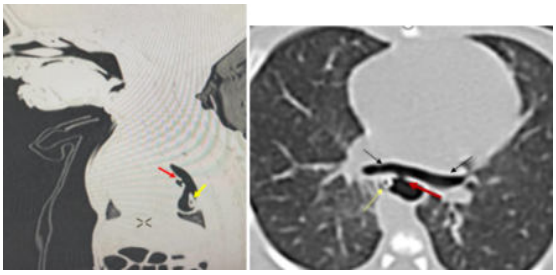
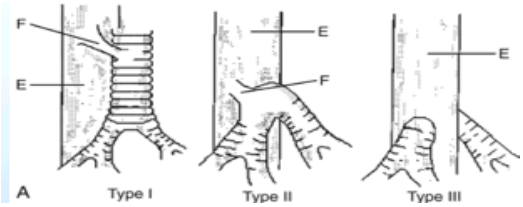


Fig.5 : CT saggital(a) and axial(b) sections at the level of the carina shows a markedly dilated distal esophagusand the nasogastric tube (yellow arrow).The trachea is completely absent. Two main bronchi (black arrows) join the midline at the carina and communicate with esophagus(red arrow) with a length of 1.4mm and caliber of 1.2mm-FLOYD'S TYPE 2 TRACHEAL AGENESIS.

DISCUSSION:

Tracheal agenesis is an uncommon congenital condition marked by

incomplete tracheal development, often occurring partially or entirely. It's frequently associated with maternal polyhydramnios and may coincide with additional anomalies such as tracheoesophageal or bronchoesophageal fistulas. This condition presents in three primary types, with the second type being the most prevalent.



FLOYD'S CLASSIFICATION OF TRACHEAL AGENESIS

Type I - Agenesis of the proximal trachea with a normal distal trachea and a TEF (Tracheoesophageal fistula)

Type II: Total TA with normal main bronchi fusing in the midline at the carina (TEF may or may not be present)

Type III: Complete TA with a separate origin of the main bronchi from the esophagus (E)

Newborns affected by tracheal agenesis typically encounter severe breathing difficulties, lack an audible cry, and cannot be intubated below the larynx at birth. Diagnosis is considered if there's an observed improvement in lung function after attempting esophageal intubation following unsuccessful standard intubation.

In approximately 90% of cases, associated congenital malformations affect various body systems, commonly involving the cardiovascular, gastrointestinal, and genitourinary systems. Prenatal imaging may reveal indicators of Congenital High Airway Obstruction Syndrome (CHAOS), such as enlarged lungs, altered diaphragm shape, fetal hydrops, and ascites, with the possibility of polyhydramnios being present.

Moreover, the presence of a tracheoesophageal fistula might alleviate pressure on the fetal lungs, potentially masking signs of airway obstruction or CHAOS in prenatal imaging. Postnatally, affected infants show signs such as aphonia, respiratory effort 4without audible breath sounds, and difficulty placing an endotracheal tube. Temporary oxygenation in cases involving tracheoesophageal fistula might be achieved through esophageal intubation.

CONCLUSION:

Tracheal agenesis, a rare congenital anomaly with an unknown etiology, involves partial or incomplete underdevelopment of trachea. Floyd classified tracheal agenesis into three types, with type II being the most common. CT imaging is a simple, quick and accurate imaging method to show in detail the anatomical situation of the airway and the precise length and degree of tracheal malformation.

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