The role of CT in the diagnosis of tracheal agenesis: a case report
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Section: Paediatric radiology
Area of Interest: Paediatric Anatomy Oesophagus
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: Experimental
Special Focus: Pathology Fistula Congenital Case Type: Clinical Cases
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Patient: 1 days, male

Clinical History:
A premature male baby was born to a 28 year old Caucasian woman at 29+2 weeks gestation. The boy was pale, hypotonic, and without audible cry. He made no breathing movements. Bag and mask ventilation was difficult and multiple attempts to intubate the infant were unsuccessful.

Imaging Findings:
Fiberlaryngoscopy was performed which showed no tracheal opening. Oesophagoscopy was performed and an endotracheal tube (ETT) was placed in the oesophagus. Chest X-ray showed deep placement of the ETT. CT scanning was performed as tracheal agenesis (TA) was suspected. CT scan revealed absence of trachea up to a few millimetres over the level of carina with a fistula between oesophagus and the short distal tracheal segment. ETT was passing through the fistula and was placed in the right main bronchus (Fig. 1, 2). The main bronchi were normal [1]. The changes were compatible with tracheal agenesis Floyd type I. CT showed no other associated anomalies. Surgical reconstruction for the TA was considered impossible and ventilatory support was withdrawn. The boy died within 12 hours of birth. Autopsy (Fig. 3) confirmed a Floyd type I TA. No other congenital malformations were found. Extended chromosome study showed normal karyotype 46XY.

Discussion:
Tracheal agenesis (TA) refers to a very rare and fatal congenital anomaly characterized by complete or incomplete failure of tracheal development with or without tracheoesophageal fistula [1]. Prevalence of TA is about 1 per 50,000 newborns [1] with a male to female ratio of 2:1 [3].

Classification of TA by Floyd et al [2] is widely used and divides it into three types (Fig. 4). [4] In type I the trachea is absent except for a short distal segment with a normal carina. In this type, there is a tracheoesophageal fistula connecting the distal part of the trachea to the oesophagus. In type II the trachea is completely absent; the two main bronchi join at the carina and in almost all cases there is a carino-oesophageal fistula. In type III, the trachea and carina are absent and each of the main bronchi join the oesophagus from either side [2].

Type I is the rarest type and accounts for about 13% of all TA. The incidence of type II and III is 65% and 22% respectively [3]. Most of the cases of TA are associated with other congenital anomalies of the heart, limbs, and
gastrointestinal and genitourinary tracts [4].

As in our case, TA typically presents at birth with respiratory distress, no audible cry and failure to intubate. Moreover, it should be suspected in cases of polyhydramnios and prematurity [3].

CT scanning plays a significant role to clearly outline the anatomy of the airways, in diagnosing associated anomalies, and deciding the feasibility of any surgical intervention [5]. CT imaging should therefore be considered in newborns with suspected TA.

**Differential Diagnosis List:** Tracheal agenesis Floyd type I, Laryngeal atresia, Severe congenital tracheal stenosis

**Final Diagnosis:** Tracheal agenesis Floyd type I

**References:**

Figure 1

Description: Shows a short distal tracheal segment, oesophagus and two main bronchi. Origin: Department of radiology Rigshospitalet Copenhagen Denmark.
Description: Endotracheal tube in the right main bronchus after passing through the fistula between short distal tracheal segment and oesophagus. Origin: Department of radiology Rigshospitalet Copenhagen Denmark.
Description: CT demonstrates placement of endotracheal tube in the right main bronchus after passing through the fistula between distal tracheal segment and oesophagus. Normal main bronchi and a dilated distal oesophagus (E) are demonstrated. Origin: Department of radiology Rigshospitalet Copenhagen Denmark.
Description: CT demonstrates the endotracheal tube in the proximal esophagus. The trachea is absent.

Origin: Department of radiology Rigshospitalet Copenhagen Denmark.
Figure 5

Description: CT demonstrates the endotracheal tube in the proximal esophagus. The trachea is absent.

Origin: Department of radiology Rigshospitalet Copenhagen Denmark.
Figure 6

Description: Floyd classification of tracheal agenesis [2] Origin: Department of radiology
Rigshospitalet Copenhagen Denmark