

## Short reports

### Congenital broncho-oesophageal fistula associated with tracheal agenesis

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Congenital malformations affecting the trachea and oesophagus are not infrequent, but certain forms are quite rare. The complicated mode of development of the tracheobronchial tree from the floor of the foregut leads to various combinations of atresia and fistula formation. Some of these occur with sufficient frequency to allow classification; the rare variations pose puzzling problems of morphogenesis. The rarity of the malformation presented here prompted this report.

#### Case report

A male infant was born after 37 weeks of an uncomplicated fourth pregnancy to a 21-year-old woman whose previous obstetric history included one abortion and one stillbirth. The infant was delivered by spontaneous breech and did not breathe or cry at birth. He was "intubated" and intermittent positive-pressure respiration was given, but spontaneous respiration was never established. The initial heart rate of 50 beats/min gradually declined and the infant was declared dead 40 minutes after birth. A tracheo-oesophageal fistula was suspected clinically.

At necropsy the infant weighed 2120 g and had features of borderline prematurity. The ears were low set and the nose beaked and the eyes were set widely apart. The scrotum was bifid and the anus imperforate. Only a single umbilical artery was found.

The trachea could not be identified. The larynx could not be examined as permission for a restricted necropsy only was obtained and the oesophagus was transected below the level of the larynx. When the oesophagus was opened the right and left main bronchi were seen opening on to the anterior wall of the organ, but they were widely patent (fig 1). Below this the oesophagus continued on to the stomach. There was a shallow, linear groove on the luminal aspect of the anterior wall of the oesophagus, above the entry of the bronchi. At the upper end of this groove a nubbin of firm tissue, 1 mm in diameter, was present. The lungs were atelectatic and hypoplastic by weight (combined weight 30 g).

Other relevant features noted were an infracristal ven-

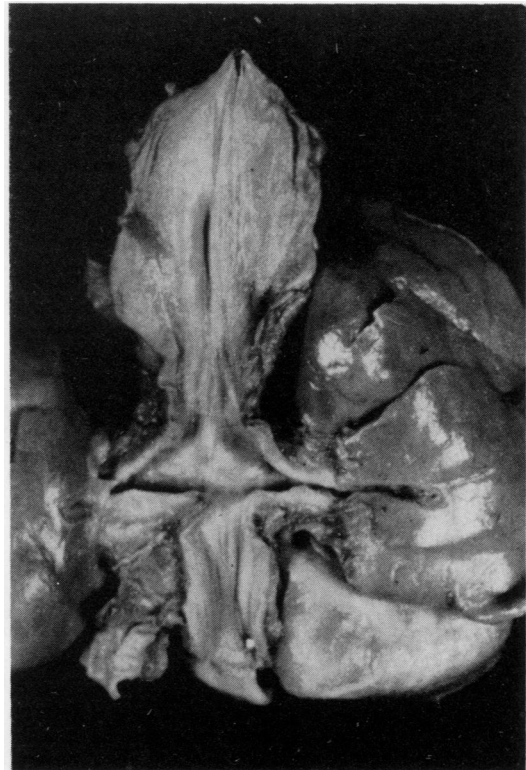


Fig 1 Necropsy specimen showing the opened-out oesophagus with bronchi arising directly from it.

tricular septal defect, a secundum type of atrial septal defect, anal canal agenesis with a common vesicorectal chamber (cloaca), complete absence of the right kidney and ureter, and a dysplastic left kidney.

Histological examination of transverse sections from the oesophagus showed a shallow central fissure on the luminal aspect lined by cuboidal-to-columnar epithelium, which was continuous with the oesophageal epithelium on the lateral aspect. In the connective tissue surrounding the fissure a zone of mucous glands bordered by islands of cartilage was present (fig 2).

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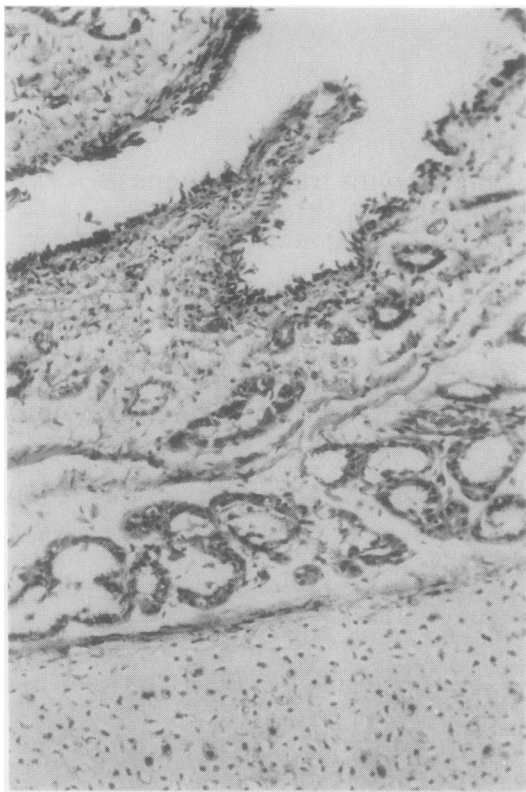


Fig 2 Histological section from the region of the oesophageal fissure showing mucosa with mucous glands and cartilage (below). (Haematoxylin and eosin  $\times 160$ .)

### Discussion

Agenesis of the trachea is a rare malformation. Since the original description by Payne in 1900,<sup>1</sup> Warfel and Schulz<sup>2</sup> found only 26 cases, including one of their own, in published reports up to 1976. An additional five were reported

by Effmann *et al* in 1975.<sup>3</sup> Floyd *et al* classified tracheal agenesis into three types, type III—which was the rarest variant—consisting of complete absence of the trachea with both bronchi arising directly from the oesophagus.<sup>4</sup> Type I comprises agenesis of the upper trachea with a normal distal trachea; some workers do not accept this as true tracheal agenesis.

Coexistent congenital anomalies of other viscera are frequently associated,<sup>2</sup> as in the present case. The presence of only one umbilical artery has also been reported.<sup>3</sup> Histological studies have apparently been neglected, however, and in the present instance microscopic examination showed a groove with surrounding mucous glands and cartilage. Associated laryngeal malformation, including absence of the larynx, has been reported in the past<sup>2</sup>; unfortunately the larynx could not be examined in this case.

The trachea develops by the formation of a tracheal groove on the floor of the foregut followed by septation, which is effected by convergence of the lateral aspects of the foregut. Hence the development of fistulas, clefts, and atresias can easily be envisaged. The existence of the microtracheal groove seen in this case suggests that when septation of the foregut does not occur the tracheal epithelium and surrounding mesenchyme do not undergo normal development to reach their normal size. The relatively normal bronchial development continues independently of septation of the foregut.

### References

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- <sup>2</sup> Warfel KA, Schulz DM. Agenesis of the trachea. Report of a case and review of the literature. *Arch Pathol Lab Med* 1976;**100**:357–9.
- <sup>3</sup> Effmann EL, Spackman TJ, Berdon WE, Kuhn JP, Leonidas JC. Tracheal agenesis. *Am J Roentgenol Radium Ther. Nucl Med* 1975;**125**:767–81.
- <sup>4</sup> Floyd J, Campbell DC jun, Dominy DE. Agenesis of the trachea. *Am Rev Respir Dis* 1962;**86**:557–60.