

# Tracheal atresia associated with cor biloculare

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Atresia of the trachea is an extreme rarity (Willis, 1962), and cor biloculare is a very uncommon cardiac anomaly. These two abnormalities were found together in the case described, a combination which, we believe, has not been previously reported.

## CASE REPORT

The infant was the second child of healthy parents. There was no history of congenital abnormalities in the family. The mother's pregnancy had been normal up to the thirty-second week, when uterine enlargement was found to be equivalent to that of a 36-week pregnancy. Radiographic examination showed a single foetus. A diagnosis of moderate hydramnios was made. The mother's blood group was B Rhesus positive. She did not contract any infectious disease or take any drugs during her pregnancy.

Labour began spontaneously at 39 weeks. There was a precipitate vertex delivery of a female infant weighing 6 lb. 9 oz. On delivery the infant gave a few gasps and showed obvious respiratory difficulty. Tracheal intubation was unsuccessful. Tracheostomy was attempted but was abandoned when no trachea was found. Thirty minutes after delivery no heart beat could be detected.

**NECROPSY** There was marked cyanosis and complete tracheal atresia from immediately below the cricoid cartilage. A thin fibrous tissue condensation immediately anterior to the oesophagus suggested the usual course of the trachea. The two main stem bronchi were normal and joined each other smoothly in the mid-line, 3 cm. below the larynx (Fig. 1). A direct fistula, 1 mm. in diameter, from the oesophagus entered the bronchi at this point. The oesophagus was otherwise normal.

The route of the circulation is shown in Fig. 2. The heart was sited normally in the mediastinum. Blood entered the atrium from four sources—the pulmonary veins, the superior and inferior venae cavae, and the coronary sinus, which also drained a patent vein of Marshall. Blood passed through a single atrio-ventricular valve into an enlarged single ventricle with a wall 2 mm. thick. There was a normal pulmonary valve and proximal pulmonary artery. Distally, the pulmonary artery divided into three branches; the right and left pulmonary arteries, and

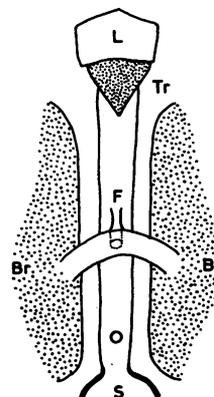


FIG. 1. Diagram of the tracheal atresia. L, larynx; Tr, tracheal remnant; F, tracheo-oesophageal fistula; Br, bronchi; O, oesophagus; S, stomach.

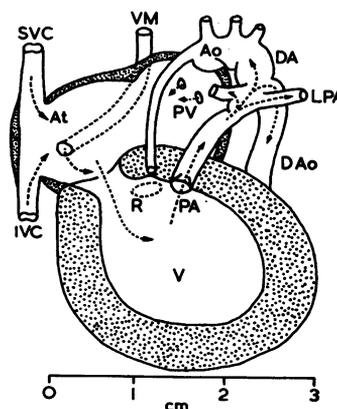


FIG. 2. Diagram of the heart showing the abnormal route of the circulation. SVC, IVC, superior and inferior venae cavae; VM, vein of Marshall; PV, pulmonary veins; At, single atrium; V, single ventricle; PA, pulmonary artery; R, rudimentary chamber; DA, ductus arteriosus; Ao, aorta; LPA, left pulmonary artery; DAo, descending aorta.

the direct origin of the descending aorta. A patent ductus arteriosus, 2 mm. in diameter, also arose at the site of trifurcation of the pulmonary artery and gave rise to the arch of the aorta and the great vessels. The ascending aorta was hypoplastic, measuring 0.5 mm. at its maximum diameter; the aortic valve was hypoplastic, tricuspid, and measured only 1 mm. across. A rudimentary left ventricle was found at the root of the aorta (Fig. 2). It was a small slit-like chamber 2 mm. across, with no communications, and was lined by hyperplastic endocardium up to 0.2 mm. thick. There was transposition of the aorta and pulmonary artery. The coronary arteries arose from the coronary sinuses and were of normal anatomical distribution.

Apart from the tracheal and cardiovascular anomalies described, no other congenital defects were found.

**HISTOLOGY** Sections of the condensed fibrous tissue from immediately below the cricoid cartilage did not show any cartilage, smooth muscle or respiratory epithelium. Only a few mucus glands were found in this area. No tracheal components were found anterior to the oesophagus. The sections of the lungs demonstrated extensive partial aeration of not only the bronchioles and alveolar ducts but many alveoli as well. All other organs were unremarkable on histological study.

#### DISCUSSION

**TRACHEAL ATRESIA WITH TRACHEO-OESOPHAGEAL FISTULA** The various types of tracheo-oesophageal fistulae are due to defective formation of the tracheo-oesophageal partition during the fourth and fifth weeks of embryonic life and a subsequent difference of growth rates between the trachea and the oesophagus (Gruenwald, 1940). The embryological basis of atresia of the trachea or oesophagus is unknown. Fluss and Poppen (1951) described three cases of tracheo-oesophageal fistula and atresia of the oesophagus associated with abnormalities of the great vessels but they were unable to relate the atresia in any way to the vascular anomalies.

With the inclusion of the present case, 14 cases of tracheal atresia have been described and they are summarized in Table I. Three types have been described: (1) the upper trachea is atretic, but the lower portion, the bifurcation, and the main bronchi are normal; (2) complete absence of the trachea, with the main bronchi joining each other in the mid-line; (3) the bronchi arising directly from the oesophagus (Floyd *et al.*, 1962). Two of the cases (Payne, 1900; Milles and Dorsey, 1950) were without tracheo-oesophageal communication, and two (Hempel, 1956; Payne, 1900) had associated laryngeal atresia.

TABLE I

Case No.	Authors	Year	Type (Floyd <i>et al.</i> )	Associated Abnormalities
1	Payne	1900	II <sup>1</sup>	Laryngeal atresia
2	Beneke	1905	II	—
3	Walcher	1928	III	—
4	Fritz	1933	I	None
5	Benešová and Peter	1934	III	None
6	Marek	1940	III	—
7	Potter and Bohlender	1941	II <sup>1</sup>	None
8	Milles and Dorsey	1950	II	Patent foramen ovale, aortic arch stenosis, bilateral cryptorchidism
9	Kessel and Smith	1953	II	None
10	Sandison	1955	I	Hypoplasia of the right kidney
11	Hempel	1956	II	Laryngeal atresia
12	Devenis and Otis	1957	II	Meckel's diverticulum
13	Floyd, Campbell, and Dominy	1962	III	Imperforate anus, patent ductus arteriosus, atrial and ventricular septal defects, aberrant portal vein, single aplastic kidney
14	Fonkalsrud, Martelle and Maloney	1963	II	Ventricular septal defect, right polycystic kidney
15	Present case	1964	II	Cor biloculare, transposition of great vessels

<sup>1</sup> Without associated tracheo-oesophageal fistula.

Almost all the infants were stillborn, but a few survived for a very short time after birth (Sandison, 1955; Floyd *et al.*, 1962), and reconstructive surgery was attempted in the latter case.

The case reported by Fonkalsrud *et al.* (1963) was unique. Radiographic diagnosis of tracheal atresia with bronchi arising from the oesophagus was made shortly after birth. Surgery was done with respiration controlled through an oropharyngeal tube. The procedure was a gastrostomy with isolation of the oesophagus and exteriorization of the proximal oesophagus. The airway was maintained by a tracheotomy tube through the orifice of the proximal lumen of the isolated oesophagus. Despite other multiple abnormalities, the infant survived to seven weeks after the operation when he died of bronchopneumonia. The condition still presents a challenging problem to paediatric surgery.

It is interesting to note that histologically the lungs showed partial aeration owing to misplacement of the endotracheal tube into the oesophagus, with passage of air through the tracheo-oesophageal fistula into the lungs, an identical occurrence to that reported by Floyd *et al.* (1962).

In these rare cases of tracheo-oesophageal fistula and tracheal atresia, theoretically at least it may be possible to inflate the lungs through an oesophageal tube as an emergency measure.

**COR BILOCULARE** Cor biloculare is probably the rarest of all cardiac anomalies (Wood, 1956). Abbott (1936) found only 14 cases of cor biloculare in 1,000 cases of congenitally abnormal hearts.

Three varieties of this condition are described (Brown, 1950): (1) Cor biloculare with an undivided truncus arteriosus; (2) with division into aorta and pulmonary artery; (3) with incomplete septal formation but with a persistent single atrio-ventricular valve.

The case described appears to belong to the third group except that it contains a non-functioning rudimentary chamber. This rudimentary chamber, because of its close relationship to the origin of the aorta, probably represents the remnant of the bulbus cordis. This has been demonstrated in those rare cases of cor triloculare biatriatum with functioning rudimentary chamber (Holmes, 1824; Abbott, 1901; Sharp, 1957; Rosenquist, Olney, and Roe, 1963).

Despite the severity of the abnormality, survival in the presence of cor biloculare is not unknown. Abbott (1936) gave the mean age at death as  $3\frac{1}{4}$  years. Carr, Goodale, and Rockwell (1935) and Nelson and Wells (1948) reported cases surviving to 36 and 27 years, respectively.

The extreme difficulty in the diagnosis of cor biloculare has been pointed out by Campbell, Gardner, and Reynolds (1952), who were unable to make the diagnosis even after full investigation. The physical signs are very variable, and even cyanosis may be absent. McCracken (1962) reported a patient who survived to 15 weeks in whom there was a complete absence of physical signs.

Cor biloculare is almost always associated with other visceral, skeletal or cardiovascular anomalies (Brown, 1950). In the present case these were confined to aortic hypoplasia, transposition of the great vessels, persistence of the vein of Marshall, and tracheal atresia.

#### SUMMARY

An infant with complete tracheal atresia and tracheo-oesophageal fistula associated with cor biloculare and transposition of the great vessels is described. This combination of rare congenital anomalies has not previously been reported.

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The views expressed herein do not necessarily coincide with those of the Surgeon-General, United States Air Force.

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