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What Is Your Diagnosis?

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What's Wrong in the Airway?

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We present the case of a 5 year-old boy who was diagnosed, at birth, with Ivemark syndrome [1]. This syndrome includes multiple congenital heart defects, associating a double outlet right ventricle with left ventricle hypoplasia, a transposition of the great vessels, a large atrial septal defect with persistent left superior vena cava, an abdominal situs inversus with midline liver and asplenia, and rhizomelic chondrodysplasia with bilateral femoral shortening and growth deficiency.

Pulmonary artery banding was performed at 5 months. As haemodynamic degradation has progressively occurred, decision was made to proceed to surgical cavopulmonary bypass as a palliative procedure at the age of 5 years. At this time, the preoperative chest X-ray revealed right upper lobe atelectasis. Hence, bedside videobronchoscopy was performed. No mucus plugging and no external compression were observed, but a very unusual anatomy of the main carina was found (Fig. 1).

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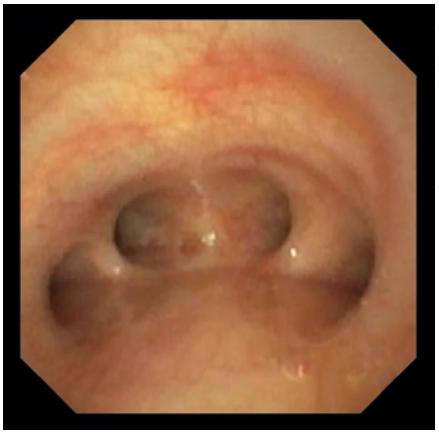

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Fig. 1. An unusual anatomy of the main carina.

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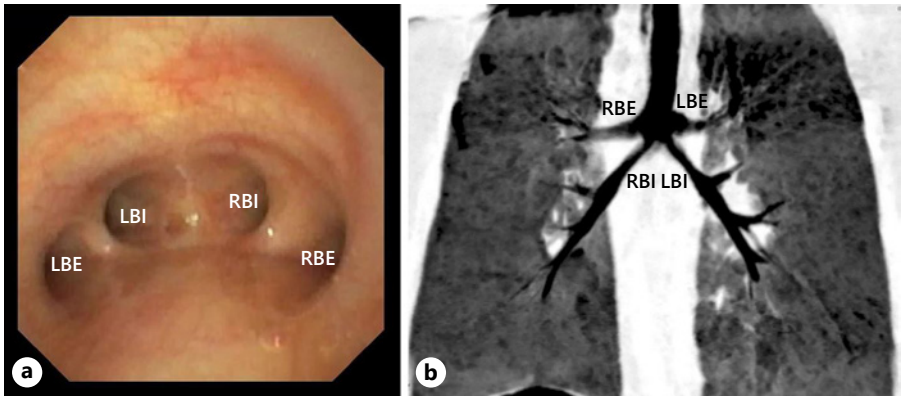

Fig. 2. Endoscopic (**a**) and CT images with labelled bronchi (**b**).

Indeed, 4 bronchial orifices are connected to the main carina. For convenience they are labelled as RBE, RBI, LBE, and LBI (R: right; L: left; E: external; I: internal; and B: bronchus) (Fig. 2a). Further bronchoscopic examination revealed the following findings:

- RBE branches off at 90°, has a regular size, is about 3–4 cm long, and divides into 3 segmental orifices typical of a right upper lobe.
- LBE has the same characteristics but only divides into 2 orifices, a posterior orifice and an intermediate segment, subsequently giving an apical orifice and an anterior orifice. This bifurcation evokes an anatomical variation frequently observed in the left upper lobe (LB1 & LB2–3).
- RBI is vertical, has a regular size, is about 4 cm long, and divides into a right middle lobe and a right lower lobe. It is obviously a right bronchus intermedius.
- LBI is about 20° vertical and exhibits the same characteristics as RBI, with an anterior division suggesting a left middle lobe.

The chest CT-scan shows 2 long-sized upper lobe bronchi connecting directly to the lower part of the trachea, just above the main carina (Fig. 2b). Tracheal bronchus, in which the right upper lobe directly originates from the trachea, is not a rare condition. Its prevalence has recently been estimated to 0.2–3% of children undergoing bronchoscopy [2]. In most cases, it is found incidentally and is thus considered as a variant of the normal tracheobronchial anatomy [3]. Nevertheless, in some patients it can be responsible for recurrent infections [4, 5]. As in the present case persistent right upper lobe atelectasis can be observed with no visible endobronchial obstruction, probably due to particular airflow limitations [6]. Haemoptysis and bronchiectasis have been described in older patients. Tracheal bronchus is reported with increased frequencies in children with various congenital heart defects, oesophageal atresia and foregut malformations, Down's syndrome, and can be associated with tracheomalacia and congenital tracheal stenosis [7, 8].

Conversely, bilateral tracheal bronchi seem rather exceptional, at least in adult patients [9]. However, in a series of 48 children with tracheal bronchus 3 of them were found to have a bilateral form [10]. Since bilateral tracheal bronchi are "usually" associated with right-sided pulmonary isomerism the extra orifice, labelled as LBE in the present report, should be regarded like a mirror-imaged right tracheal bronchus rather than a true left tracheal bronchus.

Right-sided bronchial isomerism or "bilateral right lung" is usually associated with Ivemark syndrome [7]. From an embryological point of view, this syndrome belongs to the heterotaxy group – that is, laterality disorders – where the situs ambiguous is somewhat confusing.

Conflict of Interest Statement

The authors declare no conflict of interest regarding this case report.

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Author Contributions

T.M.H.T. recorded the bronchoscopic and imaging data. L.D. and H.D. wrote the manuscript.

Keywords

Bronchoscopy · Tracheal bronchus · Child · Malformation · Heterotaxy

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