CONGENITAL TRACHEOBRONCHIAL BRANCING ANOMALIES – TYPES AND CLINICAL SIGNIFICANCE

Kamil Syga¹,B,D, Julia Lewtak¹,CD, Kaja Iwaniuk¹,B,D,F, Paweł Stanicki¹,B,E, Zuzanna Białkowska¹,2,AE-F, Aleksander Daniluk¹,AE-F, Jakub Buczek¹,2CD, Maciej Samczuk¹,2,E, Hubert Stachowicz¹,B,CD, Jan Ostański,J,C,E

¹ Human Anatomy Research Group, Medical University, Lublin, Poland
² 1st Military Clinical Hospital with the Outpatient Clinic, Lublin, Poland
³ Department of Correct, Clinical and Imaging Anatomy, Medical University, Lublin, Poland

Introduction and Objective: The tracheobronchial tree consists of the pathways through which air reaches the pulmonary alveoli. Anomalies in the tracheobronchial tree can be both congenital and acquired. Congenital anomalies include tracheal bronchus, accessory cardiac bronchus, lung aplasia, hypoplasia, or agenesis. The aim of this review is to summarise knowledge about the most common congenital anomalies in the tracheobronchial tree, and associated symptoms.


Brief description of the state of knowledge. Accessory cardiac bronchus (ACB) and tracheal bronchus (TB) are the 2 most common congenital anomalies of the tracheobronchial tree. The incidence of ACB ranges from 0.07% – 0.5%, and TB between 0.9% – 3%. There are 3 types of ACB, with type I being the most common. TB is most frequently detected on the right side. Both anomalies are often asymptomatic; however, diagnosing them may be crucial in cases of recurrent symptoms of unknown origin, such as cough, haemoptysis or recurrent pneumonia. They may also contribute to complications in surgical procedures conducted on the respiratory tract or within the chest cavity.

Summary. Understanding the occurrence of anatomical anomalies in the branching of the tracheobronchial tree can facilitate the diagnosis of atypical respiratory symptoms and clarify the cause of procedural failures, such as bronchoscopy, intubation or thoracic surgery.

Key words
bronchi, abnormalities, hemoptysis

INTRODUCTION

The tracheobronchial tree consists of the trachea, main bronchi – right and left, and branches derived from their divisions which, upon entering the lungs, further divide into smaller bronchi. Understanding the occurrence of tracheobronchial tree anomalies is crucial in differentiating the causes of respiratory system-related symptoms, such as cough, haemoptysis, recurrent pneumonia, or even a serious condition like ARDS [1, 2]. The aim of the study is to outline the most common congenital anomalies of the tracheobronchial tree, taking into account the resulting symptoms and technical challenges during examinations, such as bronchoscopy or intubation [3], as well as in chest surgery, particularly in cases of lung resection or transplantation [2].

Anatomy. The trachea begins at the level of the C6 vertebra and ends at the level of the Th4 vertebra, dividing into 2 main bronchi. The right main bronchus is wider and shorter than the left, measuring approximately 2 cm in length. The left main bronchus is narrower and longer, with a length of 4.5 cm [4]. The right main bronchus divides – based on its topography relative to the right pulmonary artery – into the eparterial bronchus, which enters the upper lobe of the right lung, and the hyparterial bronchus, with one entering the middle lobe and the other entering the lower lobe of the right lung. From the left main bronchus, 2 hyparterial bronchi arise, supplying the upper and lower lobes of the left lung. Upon entering the lung lobes, all branches of the main bronchi become intrapulmonary bronchi, named after the corresponding lung lobe. The lobar bronchi further divide into segmental bronchi, 10 in each lung, which in turn branch into subsegmental bronchi [5]. The nomenclature proposed by Boyd in 1955 is commonly used to describe the normal anatomy of the bronchi [6].

MATERIALS AND METHOD

This literature review is based on articles published in the PubMed database using the following terms: ‘tracheobronchial branches abnormality’, ‘tracheal bronchus’, and ‘accessory cardiac bronchus’. Nineteen articles published between 2001–2023 were utilized, which focused on the occurrence of anomalies in the tracheobronchial tree, and statistical data on the frequency of ACB from 3 studies conducted using computed tomography between 1999–2022. Similarly, statistical data on the frequency of TrB came from 4 studies...
conducted using bronchoscopy between 2014–2021. In both cases, the structure of the tracheobronchial tree in patients was analyzed for the presence of either of the mentioned anomalies. The results were collected and presented numerically and as a percentage (Tab. 1 and 3).

RESULTS

Anomalies of the tracheobronchial tree can be both congenital and acquired. Congenital anomalies include tracheal bronchus, additional cardiac bronchus, as well as lung aplasia, hypoplasia, or agenesis. Acquired changes involve thickening of bronchial walls, bronchial dilation, and bronchial constriction. Thickening of the bronchial walls is a respiratory tract response to irritants, accompanied by bronchial oedema and inflammation. Bronchial dilation is caused by infections, lung fibrosis, aspirations, and cystic fibrosis. Causes of stenosis include foreign body aspiration, acute aspirations, excessive dynamic collapse of the airways, tumours, granulomatous disease, and asthma [7].

The best non-invasive method for evaluating changes in the tracheobronchial tree is computed tomography (CT). The sensitivity of this method in detecting trachea and main bronchial diseases is 97% [8].

The prevalence of tracheobronchial tree anomalies is estimated to be 1–12%. Diagnosis is challenging due to the often asymptomatic nature of these pathologies. However, among patients, cough, haemoptysis, and recurrent episodes of chest infections may occur more frequently. Diagnosis of these disorders is crucial, especially when planning lung surgery or attempting endotracheal intubation. This not only helps avoid perioperative complications, but also allows for further diagnostic evaluation as tracheobronchial anomalies often co-exist with other anomalies from this group, congenital heart defects, or developmental anomalies of the anterior intestine [9].

Accessory cardiac bronchus. This is an anomaly first described by Brock in 1944. According to him, the mentioned anomaly is a supernumerary bronchus observed on the inner wall of the right main bronchus or intermediate bronchus, opposite the beginning of the upper lobe bronchus of the right lung. In most cases, the accessory cardiac bronchus occurs asymptptomatically. Occasionally, there may be intermittent haemoptysis, infection, or malignant changes [10]. Typically, haemoptysis is preceded by long-term smoking, beginning with a cough which, over time transforms into the expectoration of whitish secretion and later haemoptysis [11]. The estimated frequency of occurrence of this anomaly is between 0.07% – 0.5%, and it has been proven that this anomaly is more common in men than in women (2.8:1). The accessory cardiac bronchus develops around the 29th – 30th day of foetal life [10], and is considered to be a remnant of the primordial cardiac bronchi that did not undergo regression during embryogenesis [12].

There are three types of accessory cardiac bronchus (ACB):
1) Iorticvicular – with a blind-ended, short, additional tracheal bronchus without adjacent lung tissue;
2) ductal – which supplies a small lung lobe through small branches;
3) intermediate – with long, blind-ended projections or supplying a lung lobe [10].

Accessory cardiac bronchus can also have an atypical location. Previously, it was believed to arise only from the intermediate bronchus and the right main bronchus. In 2018, Ghaye et al. described the first case of ACB originating from the left main bronchus. Two years later, Trisolini et al. reported the first case of this bronchus originating from carina. Recent studies by Liqiang et al. demonstrated a new origin of the accessory cardiac bronchus. They examined 11 patients with ACB and showed that the accessory cardiac bronchus originates from the basal bronchus of the lower lobe. In 6 cases it was the bronchus of the right lung, and in 5 cases – the bronchus of the left lung. However, it is essential to consider a differential diagnosis, as the bronchus identified by them as an accessory cardiac bronchus may turn out to be a physiological subsegmental medial basal bronchus (B7). This type of bronchus arises from the medial wall of the lower lobe bronchus, and is located next to the accessory cardiac bronchus diagnosed among the studied 11 patients [13].

The influence of ACB on airflow depends primarily on...
the diameter and the presence of a blind ending. However, it has been proven that individuals with an accessory cardiac bronchus in the right lung may have reduced lung flow, regardless of the diameter or depth of this supernumerary bronchus. This suggests that a better post-operative diagnostic method for forced expiratory volume in the first second (FEV1) in patients with ACB undergoing lung resection, is quantitative lung ventilation scintigraphy, rather than an anatomy-based standard [14].

Treatment of the additional cardiac bronchus is introduced when there are significant life-threatening complications for the patient. Surgical treatment is primarily applied. The main reasons for performing this operations include severe abscesses not responding to antibacterial treatment, severe haemoptysis, and lung cancer, which etiologically may originate from the accessory cardiac bronchus [15].

Tracheal bronchus (TrB). The term tracheal bronchus has been known since 1785 when it was first described by Sandifort [6]. It is a rare congenital anomaly, encompassing a set of bronchial abnormalities branching-off from the trachea towards the upper lobes of the lungs. They typically originate about 2 cm from the carina, but they can occur as high as the level of the cricoid cartilage. Literature reports the frequency of occurrence ranging from 0.9% – 3% in the population [16]. Tracheal bronchus is almost always located on the right side, with a frequency of 0.1%-1.3% in adults and 1.5%-2.0% in children. It may be responsible for ventilating the entire upper lobe or only 1–2 segments. Occasionally, the tracheal bronchus may develop as very small, narrowed, or blind-ended, referred to as congenital right tracheal diverticulum [6].

There are 3 embryologic developmental theories regarding the tracheal bronchus: reduction, migration, and selection. The reduction theory suggests that it originated from the regression of a previously developed bronchus. The migration theory posits that it is the result of elongation or displacement of a branch of the bronchial tree located under the pulmonary artery. The selection theory, on the other hand, argues that the tracheal bronchus arises from local disruptions in morphogenesis [3,16]. Several classifications categorizing the tracheal bronchus have been identified [7,16,17].

The main division of TrB categorizes the anomaly into displaced tracheal bronchus, when it accompanies the right upper lobar bronchus which is properly constructed, and supernumerary tracheal bronchus, when it accompanies the normal right upper lobar bronchus, but lacks one branch. In cases where the entire right upper lobar bronchus is displaced, this type is referred to as pig bronchus, as it corresponds to the normal anatomical structure of the bronchial tree of a pig [16,17].

Table 1. Comparison of frequency of ACB occurrence in CT studies

<table>
<thead>
<tr>
<th>Imaging technique</th>
<th>Author</th>
<th>Total No. of patients</th>
<th>No. of patients with ACB</th>
<th>Gender</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Computed Tomography</td>
<td>B Ghaye et al. (1999)</td>
<td>11,159</td>
<td>9 (0.08%)</td>
<td>M: 5 (55.6%); F: 4 (44.4%)</td>
<td>100% asymptomatic</td>
</tr>
<tr>
<td>Computed Tomography</td>
<td>Elf Nisa Unlu et al. (2016)</td>
<td>5,790</td>
<td>12 (0.2%)</td>
<td>M: 7 (58.3%); F: 5 (41.7%)</td>
<td>100% asymptomatic</td>
</tr>
<tr>
<td>Computed Tomography</td>
<td>Liqiang Sun et al. (2022)</td>
<td>10,287</td>
<td>40 (0.39%)</td>
<td>M: 23 (57.5%); F: 17 (42.5%)</td>
<td>Mostly asymptomatic</td>
</tr>
</tbody>
</table>

Table 2. Anatomic classification

Anatomic classification

- **I** TrB located more than 2 cm from the carina, with distal tracheal stenosis
- **II** TrB is located more than 2 cm from the carina, without distal tracheal stenosis
- **III** TrB is located less than 2 cm above the level of the carina

Figure 3. Types of tracheal bronchus. Source: Diagram created by Dr. med. Magdalena Chmiel

In 78% of children with tracheobronchial tree anomalies – right tracheal bronchus – other disorders can exist, such as Down syndrome and other chromosomal aberrations, chest malformations, laryngotracheal stenosis [6], VATER syndrome, tracheoesophageal fistula, oesophageal atresia, congenital heart defects, lung hypoplasia, and spondyloesis [16,17]. According to a study by Al-Naimi from July 2007 – November 2020, which examined the frequency of TrB in patients who had bronchoscopy and imaging studies, such as X-rays, congenital anomalies were associated with as many as 80% of patients with TrB, of which 40% were congenital heart defects [17]. In a study by Pérez Ruiz, 24 of 26 patients diagnosed with TB [18] had other significant congenital defects. TrB in children can be associated with recurrent pneumonia, cough, audible stridor, wheezing, or respiratory failure [6, 17]. There are also cases where TrB is present in healthy children without any symptoms or accompanying diseases. In adults it often presents without symptoms and is diagnosed incidentally, most commonly during bronchoscopy [6, 17]. However, adults can also manifest symptoms of recurrent respiratory infections, cough, or haemoptysis [17], as described in Kumar’s case. The presence of a tracheal bronchus becomes significant when intubation is planned. Intubation of the tracheal bronchus can lead to respiratory acidosis due to the cutoff of the oxygen supply to the remaining parts of the lungs. On the other hand, skipping this bronchus during intubation can lead to lung collapse [19].
Accessory cardiac bronchus (ACB) and tracheal bronchus (TB) are the 2 most common congenital anomalies of the tracheobronchial tree. Classical literature often reports the incidence of ACB ranging from 0.07% – 0.5%, while in the current review of studies, this frequency ranged from 0.08% – 0.39%. This anomaly was detected using computer tomography. In the literature reviewed, there is a noticeable prevalence of ACB in males, as noted in some cited scientific works. Classically, 3 types of accessory cardiac bronchus are mentioned: I – diverticular, II – tubular, and III – intermediate, with type I being the most common. The majority of ACB cases are asymptomatic. On the other hand, the frequency of TB occurrence is reported to be between 0.9% – 3% in the population, most commonly on the right side.

The primary method for detecting anomalies is bronchoscopy. TrB is classified into 3 main divisions, with the most commonly used division categorizing them into supernumerary and displaced bronchi. Although the frequency of TB is low, and its presence often asymptomatic, diagnosing this anomaly can be crucial in the case of recurring symptoms of unknown origin, such as pneumonia, cough, or haemoptysis. Thorough diagnosis of these anomalies allows for ruling out suspicions of other potential, often dangerous, causes of symptoms presented by patients. Additionally, awareness of the occurrence of these anomalies, or knowledge of possible atypical branches of the tracheobronchial tree, can impact the success of surgical procedures performed on the respiratory pathways or within the chest. Therefore, after analyzing the data presented in this review it is easy to conclude that further research and diagnosis of patients born with ACB, TrB, or other abnormalities in the respiratory pathways, are necessary.

**Acknowledgements**
The authors express their gratitude to Magdalena Chmiel, MD, for creating Figure 3.

**Disclosures**
All authors have read and agreed with the published version of the manuscript.

**Conflict of interest.** The authors declare no conflict of interest.

**Funding statement.** No external funding was received for the performance of this review.

**Statement of informed consent.** Permission was obtained to use images from the computer tomography study conducted at the Department of Medical Radiology I, Medical University, Lublin, Poland.

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</tr>
</thead>
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<td>Bronchoscopy</td>
<td>Mital H Dave et al. (2014)</td>
<td>1,021</td>
<td>11 (1.06%)</td>
<td>M: 3 (27%); F: 8 (72%)</td>
<td>100% (11/11)</td>
</tr>
<tr>
<td>Bronchoscopy</td>
<td>Isabelle Ruchonnet-Mertrailler et al. (2015)</td>
<td>5,790</td>
<td>57 (0.9%)</td>
<td>M: 31 (54.3%); F: 26 (45.7%)</td>
<td>61.5% (35/57)</td>
</tr>
<tr>
<td>Bronchoscopy</td>
<td>Estela Perez-Ruiz et al. (2018)</td>
<td>1,317</td>
<td>26 (1.9%)</td>
<td>K:11 (42.3%); M:15 (57.6%); K: 11 (42.3%)</td>
<td>92% (24/26)</td>
</tr>
<tr>
<td>Bronchoscopy</td>
<td>Anal Al-Naimi et. al. (2021)</td>
<td>1,786</td>
<td>20 (1.12%)</td>
<td>M: 13 (65%); F: 7 (35%)</td>
<td>80% (16/20)</td>
</tr>
</tbody>
</table>

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5. Adam Bochenek, Michał Reicher, Anatomia człowieka tom 2, PZWL wydanie XIII 2016
9. Sabri YY, Yamamah HGAEN, Taymour TA. Role of multislice computed tomography in evaluation of congenital tracheobronchial branching

**Figure 4.** Upper lobe tracheal bronchus. Source: *Collection* of the Department of Medical Radiology I, Medical University, Lublin