**CASE SERIES**

# Rare laryngotracheobronchial anomalies in children: a retrospective study of 249 bronchoscopic cases in a single center

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**Abstract**

**Background:** Laryngotracheobronchial anomalies in children are rare, and their clinical manifestations are diverse. In this study, we report the clinical aspects and prevalence rates of tracheal bronchus (TB) and congenital subglottic stenosis (CSS) in a select group of patients at our institution and briefly review and discuss the literature to draw attention to these rare anomalies.

**Case series:** We retrospectively reviewed the clinical records of 249 children for whom senior surgeons performed rigid bronchoscopy for suspected or confirmed food choking or foreign body aspiration in children between August 2013 and January 2020. Bronchoscopic findings and demographics of the patients with laryngotracheobronchial anomalies were documented. Four among the 249 patients (1.6 %) had right-sided TB (two males aged 24 and 42 months and two females aged 14 and 60 months), while three (1.2 %) had grade 1 CSS (one male aged eight months and two females aged 11 and 13 months). There was no previous history of endotracheal intubation for any subglottic stenosis (SS), though all three patients were admitted with sudden onset of respiratory difficulty. One of the TB cases had congenital cardiac anomalies, and two were symptomatic before their admission to the emergency department.

**Conclusions:** The prevalence of CSS and TB in the healthy population is low. Physicians dealing with the pediatric airways should consider such anomalies for prompt diagnosis, proper instrumentation, management, and follow-up of these cases. Our data also correlate with previous data, indicating that these anomalies' prevalence rates have not increased during the last few decades. HIPPOKRATIA 2023, 27 (2):59-63.

**Keywords:** Bronchoscopy, children, congenital, subglottic stenosis, tracheal bronchus

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## Introduction

Tracheobronchial anomalies, such as tracheal bronchus (TB) and congenital subglottic stenosis (CSS), are rare, and their clinical manifestations are diverse. They can be asymptomatic or cause difficulty in breathing, recurrent pulmonary infections, difficult intubation, or misplaced intubation tubes. Furthermore, malignancies have been reported in adults with TB, while the severe forms of CSS are life-threatening[1,2].

The reported incidence of TB ranges from 0.1 % to 5 %[3-10]. Although several scientific papers mention the clinical aspects, complications, and treatment of food choking (FC), foreign body aspiration (FBA), or other conditions that require rigid bronchoscopy (RB) or flexible bronchoscopy (FB) in children, only a few of these studies have focused on TB[6-10]. The lack of reports on TB cases may be due to several reasons. The attending surgeon's lack of anatomical knowledge or merely focusing on extracting the foreign body from the tracheobronchial tree may lead to overlooking the anomaly. Thus, existing data on its incidence or prevalence are limited. In addition, it can be asymptomatic for many years and evade detection[1,3].

The subglottic area is an anatomic region that extends just below the vocal folds to the lower border of the cricoid cartilage[11]. The incidence of subglottic stenosis (SS) ranges from 1 % to 8 %. However, most reported cases were acquired and reported together with the congenital cases[12]. Without a history of previous intubation, stenosis in this area is considered CSS. CSS is an element of the laryngeal anomalies spectrum resulting from failure to recanalize the laryngeal lumen completely. It may display various histopathological forms, such as membranous (typically circumferential) or cartilaginous[11,13]. The most common CSS form is the lateral elliptical shelf of the cartilage at the cricoid level[14]. Some forms of CSS regress as the children grow older. Therefore, its true prevalence or incidence remains unknown[11,13].

We retrospectively reviewed the charts of pediatric cases who underwent RB performed by senior surgeons at our institution for suspected or confirmed FC and FBA. We estimated the demographics, clinical aspects, and prevalence rates of TB and CSS in this selected patient group, reviewed relevant literature and discussed these rare anomalies to draw attention to them.

**Case Series**

The clinical records of 249 children who underwent bronchoscopy performed by senior surgeons at our institution for suspected or confirmed FC or FBA between August 2013 and January 2020 were retrospectively reviewed. The collected data included patient age and sex in 249 cases, previous symptoms attributed to the anomaly, location or side of the anomaly, and associated congenital anomalies in cases where TB or CSS were identified.

All bronchoscopies were performed under general anesthesia in the operating room with RB (StorzÒ rigid bronchoscopes No. 2.5, 3, 3.5, and 4, Karl Storz, Tuttlingen, Germany). All patients were routinely administered intravenously cefazoline sodium 50 mg/kg for prophylaxis and methylprednisolone 0.5 mg/kg to prevent additional airway edema. The Myer-Cotton scale was used to evaluate the airway size in patients with CSS.

Descriptive analysis results are presented as percentages or mean ± standard deviation. Informed consent was obtained from all patients regarding the operation type, possible complications, and possible publication of the results. Before the study, the Institutional Ethics Committee approved the study design (decision No. 09, date: 08/01/2020).

During the seven-year study period, 249 RBs were performed by senior surgeons for suspected or confirmed FC or FBA in children. Among the 249 cases (139 male, 110 female), four (1.6 %) right-sided TB and three (1.2 %) CSS (grade I) cases were identified. The mean age of the 249 cases was 22.6 ± 22.22 months.

Two of the four TB cases (24-month-old and 42-month-old males) had no previous respiratory complaints or symptoms. One patient (a 14-month-old female) had a history of recurrent bronchitis and lower respiratory tract infections, and one patient (a 60-month-old female) had a history of croup. A foreign body was found in the two cases, in the left main bronchus of the 14-month-old female and the right main bronchus of the 24-month-old male. Food particles did not obstruct TB in any case. One male patient had a history of patent ductus arteriosus (PDA) ligation and underwent surgery for an atrioventricular septal defect.

Three CSSs (an eight-month-old male, an 11-month-old female, and a 13-month-old female) were asymptomatic before emergency admission. There was no history of endotracheal intubation in either case. One female patient was admitted with suspected food choking during pear eating. The other female and male patients were admitted for suspected FBA following sudden onset respiratory difficulty while lying in their cradles without parental supervision. CSS caused difficult intubation during rigid bronchoscopy with No. 3.5 rigid bronchoscope. Therefore, No. 3 and 2.5 rigid bronchoscopes were used in these cases. No food or foreign bodies were found in either patient. No other anomalies, such as laryngomalacia or tracheomalacia, were detected or noticed during RB.

**Discussion**

The true incidence of laryngotracheobronchial anomalies, such as CSS and TB, among healthy individuals is difficult to determine. Most TB patients remain asymptomatic for several years. By contrast, CSS is easy to diagnose because it becomes symptomatic in the first year of life. However, some mild forms of CSS improve as the child grows and remain silent[1,3,11,13].

*Tracheal bronchus*

TB is one of the two major bronchial anomalies of the tracheobronchial tree, with the second being an accessory cardiac bronchus. TB includes a broad spectrum of bronchial anomalies originating from the main bronchus or trachea and directed to the upper lobe[1]. TB has different types and can be classified as follows: type 1, a bronchus originating from the junction of the lower and middle one-third of the trachea; type 2, a bronchus that arises from the lower one-third of the trachea; and type 3, just proximal to the typical origin of the right main bronchus (carinal trifurcation)[6,15]. The term supernumerary TB, which can be confused with TB, refers to a bronchus that supplies an associated tracheal lobe other than the upper lobe. Without computed tomography or magnetic resonance imaging, it is challenging to distinguish these anomalies by RB or FB[3,6].

Dave et al[6] reported eleven TB cases in 1,021 RB procedures performed during elective surgery in children aged 0-6 years, with TB prevalence being 1.06 % in their series. One year later, Ruchonnet-Metrailler et al reported 57 TB cases in a cohort of 5,970 FB pediatric cases undergoing respiratory symptoms investigation, a reported prevalence of 0.9 %[7]. In 2018, Perez Ruiz et al[9] reported 26 (1.9 %) TB in 1,337 cases. The reported rate of TB ranges from 0.1 to 5 % in the literature[3-10]. We found four TB in 249 cases in our study, thus a prevalence rate of 1.6 % for TB, which is within the range of the previous reports. All TB in our series were in the right lateral wall of the trachea (Figure 1), as highlighted in previous literature[1,4,6,7,16]. The male-to-female ratio for TB in the present study was 1:1 (Table 1), in contrast to that in other studies[6,7].

Literature indicates that TB is often complicated by other congenital anomalies such as Down syndrome, congenital heart disease, and other congenital airway malformations such as laryngomalacia or tracheal stenosis[4,6]. Dave et al[6] reported that all eleven TB cases had at least one congenital anomaly in their series. Ruchonnet-Metrailler et al[7] reported that 61.5 % of the 57 TB cases had associated anomalies. A recent Spanish multicenter study by Moreno et al[10] states that tracheomalacia and laryngomalacia were found in 33 % and 9 % of TB cases, respectively. Regarding other congenital anomalies, 32 % had congenital cardiovascular malformations, 28 % had gastroesophageal reflux, 22.5 % had congenital tracheal stenosis, and 8.3 % had Down syndrome. We found only one (25 %) TB case with a congenital cardiac anomaly, which is attributed to our study's limited number of cas-

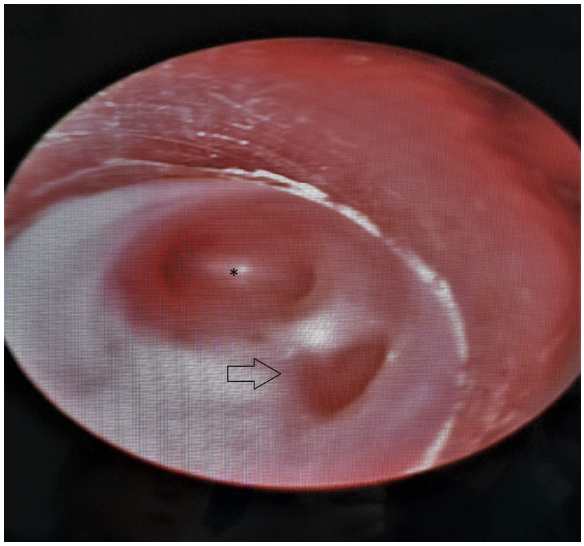

**Figure 1:** Endoscopic view of right-sided type-1 tracheal bronchus (black asterisk: tracheal carina, black arrow: tracheal bronchus).

es. If the number of cases were higher, other anomalies would be possible to detect. On the other hand, we may presume that more asymptomatic TB cases could be present among the otherwise normal population since no syndromic associations or other congenital airway anomalies were detected in our series.

The main difference between the current study and previous large cohorts was that we performed RB for suspected or confirmed FC or FBA[6-10]. Thus, none of the cases we included was symptomatic for lower respiratory infection or atelectasis. However, one of the four TBs had been previously treated for recurrent bronchitis and lower respiratory tract infections. Another patient had a history of croup. These findings may correlate with previous reports stating that TB can present with wheezing, stridor, or recurrent lower respiratory tract infections, atelectasis, asthma, or bronchitis. However, in large cohorts, no clear association was found between TB and those clinical conditions[6,7]. Thus, we cannot undoubtedly state that TB caused previous respiratory problems in our patients.

We did not observe any complications during intubation, bronchoscopy, or endotracheal tube insertion in the reported TB cases. However, in cases with difficult intubation or difficult ventilation, unexplained intraoperative hypoxemia, or persistent atelectasis, TB should be suspected as one of the causes[4,17,18]. Although there is no specific treatment for asymptomatic TB, lobectomy is the preferred treatment for the affected lobe with persistent respiratory problems[4].

*Congenital subglottic stenosis*

Among the congenital anomalies of the airway, CSS is the third most common anomaly after laryngomalacia and vocal cord paralysis and is a result of recanalization failure of the larynx[11-13]. It is a common cause of stridor in neonates, infants, and children. Other symptoms of this condition include dyspnea, brassy cough, recurrent croup, and recurrent upper respiratory tract infection[2,5,12,13].

In our series, CSS cases had no previous symptoms such as stridor, dyspnea, barking, or croup but sudden onset respiratory difficulty before emergency admission. There was no history of hospitalization for respiratory symptoms or endotracheal intubation. One female patient was admitted with sudden difficulty in breathing while eating a pear. Therefore, the presumed diagnosis was food choking (Table 1). Two patients were admitted with sudden difficulty in breathing while they were lying in their cradles without parental supervision. However, the 13-month-old female patient also had inspiratory stridor at admission, which was mistakenly considered a symptom of tracheal occlusion by a foreign body. CSS was identified during RB instead of foreign body or food.

The symptoms of CSS are usually evident in the first few weeks or months after labor, depending specifically on the severity of the pathology. More severe cases become evident at the time of labor[2,13]. The late onset of symptoms in the reported cases was related to the lower grade of the CSS. Recent literature indicates mild to mod-

**Table 1:** Demographics and clinical characteristics of the four children with tracheal bronchus and three children with congenital subglottic stenosis, diagnosed during rigid bronchoscopy for suspected or confirmed food choking or foreign body aspiration.

| Anomaly type | Sex | Age (month) | Previous symptom | Location/side | Associated Anomalies |
|---|---|---|---|---|---|
| TB | Female | 14 | Recurrent bronchitis and lower RTI | Right | None |
| TB | Male | 24 | None | Right | None |
| TB | Male | 42 | None | Right | PDA and AV septal defect |
| TB | Female | 60 | Croup | Right | None |
| CSS | Female | 11 | Suspected FC | Subglottic area | None |
| CSS | Male | 8 | Sudden onset respiratory distress | Subglottic area | None |
| CSS | Female | 13 | Sudden onset respiratory distress | Subglottic area | None |

TB: tracheal bronchus, CSS: congenital subglottic stenosis, RTI: Respiratory tract infection, PDA: patent ductus arteriosus, AV: Atrioventricular, FC: food choking.

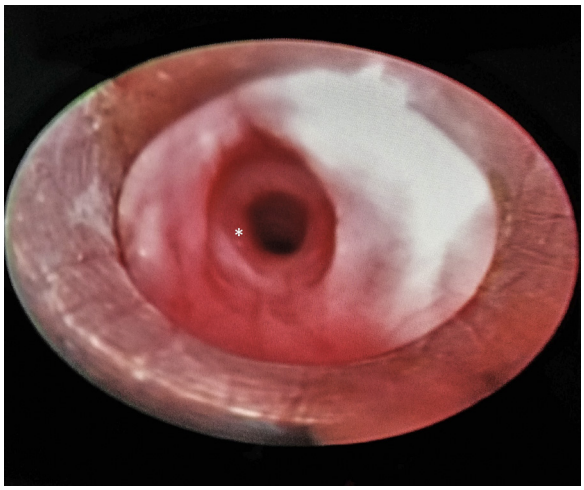

**Figure 2:** Endoscopic view of Myer-Cotton grade 1 congenital subglottic stenosis (white asterisk: stenosis).

erate (namely Myer-Cotton grade I-II) CSS may resolve within years and not become evident during the patient's lifetime[11-13]. In our series, none of the patients had a previous history of airway intubation, and both patients had membranous, circumferential CSS (Figure 2).

The clinical measurement of subglottic diameter has changed tremendously. Early studies suggested that the normal subglottic lumen diameter should be six mm in size in term infants. However, autopsy studies have revealed that the effective or intramucosal diameter of the subglottic region is approximately 4.5 mm in diameter in neonates[2]. Currently, the Myer-Cotton grading system is commonly used for grading subglottic stenosis cases, and in full-term neonates, subglottic diameters of four mm or less are considered stenotic[11,12,19]. According to the Myer-Cotton grading system, grade I stenosis is a subglottic area stenosis of up to 50 % of its diameter. Grade II refers to 51-70 % stenosis; grade III refers to 71-99 % stenosis; and grade IV refers to no visible lumen[19].

Roberts et al[20] reported that No 3.5 RB (external diameter: 5.7 mm, corresponding to No 4 to No 4.5 tracheal tube) is preferred in children older than six months. No 3 RB is suitable for term infants up to six months of age, and No 2.5 RB is preferred in premature babies[20]. From this point of view, the subglottic area should have passed with at least No 3.5 RB in our two cases as they were older than six months. Also, the 13-month-old female patient should have been intubated with No. 4 RB. However, we failed with No. 4 and 3.5 RB and utilized No. 3 and 2.5 RB in our cases. These findings correlate with the literature in terms of subglottic stenosis definition[11,19].

CSS has been reported to be associated with certain syndromic conditions such as Down syndrome, CHARGE, 22q11 deletion syndrome, or other congenital head and neck lesions[11,13]. In our series, CSS cases had no syndromic condition or other congenital anomalies. Thus, we may speculate that most grade I CSS cases remain silent, not just because they improve with time[2], but also because they are not associated with any congenital condition requiring further investigation.

The treatment and follow-up of CSS cases depend on the severity of the lesion. Nonoperative conservative management is usually sufficient in grade I and some grade II lesions. Some patients require gentle dilatation with dilators. Grade III and IV lesions usually require surgical intervention[2,11-13]. In our series, all RBs were followed by endotracheal intubation, and we utilized No 3.5 endotracheal tubes in the three CSS cases. They did not experience any problems during intubation, extubation, or the postoperative period. Two cases were followed for two years, and one case was followed for one-and-a-half years, and none experienced any problems to date.

RB is the gold standard treatment for FBA in children[21]. However, recent reports state that FB is superior in extracting foreign bodies as it is less traumatic and uncomplicated and can access distal airways in children[22,23]. Nevertheless, it should be noted that FB is not free from complications, especially in children younger than two years of age. In addition, if FB fails, RB is necessary, especially in long-standing and lodged foreign bodies[23]. In a recent literature review, RB was the mainstay for foreign body removal in children and adolescents[21]. Despite this, FB is ahead of RB in treating FBA in children in new publications, and it will replace RB in the future[22,23].

The major limitations of the current study are as follows: a) we performed a retrospective chart review of a specific group of patients. Ethical concerns make it impossible to perform bronchoscopy (rigid or flexible) in a healthy population to determine the exact prevalence or incidence of TB or CSS. Thus, the prevalence rates reported in this study do not reflect the actual prevalence of asymptomatic TB or CSS in the population. However, it correlates with previous data, which may indicate that the prevalence rates of these anomalies have remained the same during the preceding few decades. b) Another limitation is that we performed RB instead of FB, as we do not perform FB in our clinic.

## Conclusion

Laryngotracheobronchial anomalies can be found incidentally during RB or FB performed in children for unrelated reasons. Although the prevalence of asymptomatic TB and CSS is low, there may be more asymptomatic cases in healthy populations. Thus, physicians dealing with pediatric airway diseases should consider such anomalies for proper and prompt diagnosis and proper management before and/or during bronchoscopy in these cases.

## Conflict of interest

The authors have nothing to disclose.

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