airway collapse on pretransplant pulmonary function testing or bronchoscopy, the reduced amount of cartilage in the proximal airways is not an adequate explanation for the development of bronchomalacia posttransplantation. A hypothesis may be made that the reduced amounts of cartilage in the proximal airways, however, made the patient herein reported particularly susceptible to bronchial ischemia after transplantation and led to the development of bronchomalacia. After lung transplantation, blood supply to the proximal airways often is compromised because of dependence upon collateral blood flow. Abnormalities consistent with reduced perfusion have been observed in proximal bronchial cartilage of asymptomatic lung transplant recipients.9

Bronchomalacia has been previously described in lung transplant recipients in association with obliterative bronchiolitis (OB).10 Impaired bronchial blood flow is thought to contribute to development of bronchomalacia in OB. Bronchial blood flow is reduced in animal models of acute rejection, and similar reductions may occur in patients with acute or chronic rejection.10 In addition, immunologic factors associated with OB may also contribute to the development of bronchomalacia with OB. However, autopsy studies in this patient revealed no evidence of any acute or chronic allograft rejection.

In conclusion, we describe the first case of lung transplantation for Williams-Campbell syndrome. Autopsy studies revealed cartilage deficiency which included proximal and distal airways, in contrast to previous reports of Williams-Campbell syndrome. Consequently, bilateral sequential lung transplantation is not recommended for this syndrome because of a high risk of postoperative airway complications. En bloc bilateral lung transplantation may offer a more viable therapeutic option for patients with Williams-Campbell syndrome although additional data are needed.

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Tracheal Bronchus*

A Cause of Prolonged Atelectasis in Intubated Children

Brian P. O'Sullivan, MD; Joseph J. Frassica, MD; and Shawn M. Rayder, MD

Tracheal bronchus is a common anomaly that occurs in approximately 2% of people. Two children with multiple medical problems which led to endotracheal intubation are described. The hospital course for each child was complicated by persistent right upper lobe atelectasis. The presence of a tracheal bronchus was not recognized in either case initially; identification of this anatomic variant allowed appropriate changes in airway management. The potential for tracheal bronchus to cause, or be associated with, localized pulmonary problems is reviewed. The diagnosis of tracheal bronchus should be considered early in the course of intubated patients with right upper lobe complications. (CHEST 1998; 113:537-40)

Key words: atelectasis; Down's syndrome; pediatrics; pig bronchus; right upper lobe; tracheal bronchus; tracheal stenosis; tracheoesophageal fistula; trisomy 21 syndrome

Abbreviations: ETT=endotracheal tube; PDA=patent ductus arteriosus

A right-sided bronchus arising from the trachea above the main carina, known as a tracheal bronchus or "pig bronchus," occurs in 0.1 to 5% of humans.1-4 This is often an incidental finding of no clinical significance; however, it can be associated with localized pulmonary problems including chronic atelectasis, recurrent infection, bronchiectasis, and cysts.5-10 Tracheal bronchus also may be seen in association with other congenital anomalies such as tracheoesophageal fistula, tracheal stenosis, and Down's (trisomy 21) syndrome.2,3,6,8,11

Although case reports of tracheal bronchus are common in the older medical literature, there has been little published about this entity recently. Two cases of tracheal bronchus presented here were unanticipated discoveries during the course of antegrade or retrograde endobronchial intubation. Dr. O'Sullivan identified the right bronchus at the primary intubation attempt and performed an immediate intubation of the left bronchus. The right bronchus was not intubated during the initial 24 hours of the hospital course and the right upper lobe atelectasis persisted. The possibility of a tracheal bronchus was considered when the patient was intubated 12 hours after arrival, and the bronchus was subsequently intubated, resulting in clearing of the right upper lobe. The second case was a 6-month-old infant who presented with recurrent episodes of bronchiolitis. Bronchoscopy identified pathologic right bronchus identified at birth which was not intubated at the initial hospitalization. The bronchus was successfully intubated during the second hospitalization.

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bronchus seen in children with other severe illnesses are reported to remind clinicians of the potential complications of this relatively common airway anomaly.

**REPORT OF CASES**

**CASE 1**

A male infant at 31 weeks' gestation was born with esophageal atresia and tracheoesophageal fistula. He underwent multiple surgical procedures and his hospital course was complicated by sepsis and the need for long-term ventilatory support. Roentgenograms of the chest during this time showed recurrent atelectasis. The endotracheal tube (ETT) was believed to be embedded in the right lateral wall of the trachea as evidenced on several x-ray films.

During induction for end-to-end anastomosis of the esophagus, the anesthesiologist was unable to selectively intubate the right main bronchus as planned. Rigid bronchoscopy revealed what was thought to be a narrowed right main bronchus, and the procedure was aborted. A subsequent bronchoscopy revealed a pouch arising from the right tracheal wall approximately 1 cm above the carina. This had been mistaken as a stenotic right main bronchus at the previous surgery. In addition, there was marked tracheomalacia of the midtrachea involving the anterior and right lateral walls. It was thought that the pouch was either a congenital diverticulum of the trachea or a traumatic dilatation secondary to chronic endotracheal intubation.

Subsequently, a barium swallow was performed during which the patient aspirated barium into the airway. The resultant bronchogram revealed that the pouch was, in fact, the opening of a tracheal bronchus with no upper lobe bronchi arising from the right main bronchus (Fig 1). This bronchus was severely kinked 1 cm from its takeoff from the trachea. In retrospect, the x-ray films of the chest which showed the ETT apparently embedded in the right lateral wall of the trachea were actually depicting intubation of the tracheal bronchus.

Knowledge of the presence of the tracheal bronchus led to careful positioning of the ETT and monitoring of its position throughout the remainder of the hospitalization. In turn, this led to fewer ventilatory problems and decreased right upper lobe atelectasis. It also allowed the surgeons and anesthesiologists to prepare appropriately for selective airway intubation at the time of esophageal reanastomosis. Despite this, the child had a stormy course due to multiple surgical complications and sepsis and died at 16 months of age.

**CASE 2**

A 16-month-old girl with Down's syndrome was admitted to the pediatric ICU with respiratory failure secondary to respiratory syncytial virus infection. Early in this patient's life, a diagnosis of patent ductus arteriosus (PDA) had been made. The family had not pursued medical care for the PDA.

The child was intubated in the emergency department. The attending physician had difficulty passing a 4.0-mm ETT more than a centimeter or two below the vocal cords; however, in the pediatric ICU the next day the ETT was advanced farther into the trachea. The child was treated with high-frequency oscillatory ventilation and then, after 8 days, was supported with conventional ventilation. An echocardiogram showed the presence of a PDA with marked pulmonary hypertension. Persistent right upper lobe atelectasis was noted throughout this time. On conventional ventilation, the patient had transient periods of hypoxia or hypercarbia, or both, which seemed to be related to the position of the ETT and of the patient's head and neck. Review of the plain radiographs suggested a right tracheal bronchus (Fig 2). A helical CT scan of the chest was performed.

![Figure 1](image1.png) **FIGURE 1.** Inadvertent bronchogram of patient 1 obtained during barium esophagogram. Note the severely narrowed right upper lobe bronchus arising from the trachea above the main carina.

This demonstrated both tracheal stenosis at the level of the thoracic inlet and a right upper lobe bronchus coming off the trachea just above the main carina (Fig 3). No right upper lobe bronchus was seen emanating from the right main bronchus. The patient's airway was managed with a 4.5-mm ETT placed in the upper trachea (proximal to the stenosis). Right upper lobe atelectasis resolved slowly, and the patient was extubated successfully. When the child was in stable condition, the PDA was ligated. Two weeks following PDA ligation, she had a sudden hypoxic event associated with marked systemic hypotension. The

![Figure 2](image2.png) **FIGURE 2.** Roentgenogram obtained in the pediatric ICU demonstrating right upper lobe atelectasis. The thin arrow indicates the main carina. The curved arrow indicates the origin of the tracheal bronchus.
child died despite reintubation and full resuscitation efforts. The family denied permission for an autopsy.

**DISCUSSION**

Tracheal bronchus is a term applied to any airway which arises from the trachea above the level of the main carina. This is almost always a right upper lobe bronchus; however, left tracheal bronchi have been described rarely. There are many possible configurations of right-sided tracheal bronchus and right upper lobe bronchus. The right tracheal bronchus may be a displaced bronchus with all three segmental branches arising from it with no right upper lobe connection to the right main bronchus (as in both of the cases reported herein), or the tracheal bronchus may consist of only a right upper lobe apical bronchus with anterior and posterior upper lobe branches arising from the right main bronchus. Supernumerary bronchi leading to the right upper lobe with a normal, trifurcated right upper lobe bronchus coming off the right main bronchus also have been reported.

The true incidence of tracheal bronchus has been debated, but it is likely that the occurrence approximates 2%. Le Roux reported 30 anomalous right upper lobe airways in a review of 1,000 consecutive bronchograms; however, only 19 of these aberrant airways arose from the trachea (1.9% of all bronchograms). McLaughlin et al reported an incidence of tracheal bronchus of 2% in a series of over 400 pediatric bronchoscopies. Although lower and higher incidences have been reported, it is obvious that this is not a rare anatomic variant and that all physicians see patients with tracheal bronchus at some point.

It has been stated that “once identified, the tracheal bronchus has no particular significance.” In many cases, this is true. Unfortunately, the two patients described in this report had severe underlying problems and complicated hospital courses which were made worse by the presence of a tracheal bronchus that was not recognized during initial treatment. Tracheal bronchus was not the primary cause of illness in either of these children and was overlooked in part due to the fact that attention was focused on their other medical problems. Pooling of secretions and obstruction of the tracheal orifice of the tracheal bronchus by the ETT contributed to the problem of persistent atelectasis. Inadvertent intubation of the tracheal bronchus led to regional variations in ventilation which complicated care. Similar problems have been reported with tracheal bronchus in patients undergoing intubation for anesthesia.

The incidence of tracheal bronchus in association with other anomalies is unknown. Le Roux, Atwell, and Ritsema reported the incidence of tracheal bronchus in large series of bronchograms. In more than 40 cases of anomalous right upper lobe bronchi, no mention is made of associated abnormalities other than Down’s syndrome. Although this supports the belief that tracheal bronchus is generally an isolated, clinically irrelevant finding, case reports of tracheal bronchus have focused on its association with other airway, lung, and foregut malformations. McLaughlin et al reported associated anomalies in 78% (14 of 18) of children with tracheal bronchus. Other findings in their patients included rib abnormalities, tracheoesophageal fistula, Down’s syndrome, and VATER (vertebral defects, anal atresia, tracheoesophageal fistula, esophageal atresia, renal defects, and radial dysplasia) syndrome. Cantrell and Guild noted 3 cases of aberrant right upper lobe bronchi reported in 13 patients with segmental tracheal stenosis.

Both patients reported here had associated findings. The first patient had tracheoesophageal fistula and tracheomalacia in addition to tracheal bronchus. Although not one of the common anomalies seen with tracheoesophageal fistula, the diagnosis of a foregut malformation should make the clinician suspect the possibility of an accompanying tracheal bronchus. The second had Down’s syndrome and tracheal stenosis. Stenosis was suspected at the time of intubation and, in retrospect, was a clue to the presence of tracheal bronchus.

Of note is the apparent association of tracheal bronchus with Down’s syndrome. Although the specific incidence of tracheal bronchus in children with Down’s syndrome is not known, 2 of 30 patients with anomalous right upper lobe bronchi reported by McLaughlin et al were stated to be “mongols” and 2 of 18 patients in the report by McLaughlin et al had Down’s syndrome. In addition to the children reported here, two other patients at this center have been identified over a period of the last 6 years with asymptomatic tracheal bronchus without Down’s syndrome or other anomalies. Thus, one of the four patients we have seen with tracheal bronchus also had Down’s syndrome. Given the incidence of Down’s syndrome in the general population (1 in 600 to 800 births), people with this chromosomal abnormality appear to be overrepresented in the reported cases of tracheal bronchus.

In summary, tracheal bronchus can interfere with routine care of children in the operating room and in the pediatric ICU. Undoubtedly, this applies to adult patients in these settings, too. It is important for clinicians to keep this airway anomaly in mind when caring for patients who have unusual sensitivity to ETT position or who have

**Figure 3.** Helical CT scan of patient 2 demonstrates tracheal bronchus arising from the trachea just above the main carina. There is some narrowing seen at the level of the thoracic inlet (arrow). A slightly anterior image (not shown) clearly demonstrated tracheal stenosis at this level.
chronic right upper lobe atelectasis, especially in those patients who have Down’s syndrome, tracheal stenosis, or foregut malformations.

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Vocal Cord Dysfunction Associated With Exercise in Adolescent Girls*

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Vocal cord dysfunction (VCD) has been reported in adolescents only rarely. Two patients are described whose initial diagnosis was exercise-induced bronchospasm (EIB). However, evaluation revealed inspiratory stridor and flattening of the inspiratory limb of the flow-volume curve. Flexible fiberoptic rhinolaryngoscopy revealed adduction of vocal cords during inspiration. Patients were treated with speech therapy and have remained free of symp-

toms. VCD should be considered in adolescents who are unresponsive to treatment for EIB.

(CHEST 1998; 113:540-42)

Key words: asthma; exercise-induced bronchospasm; laryngoscopy; stridor; vocal cord dysfunction

Abbreviations: EIB=exercise-induced bronchospasm; PFT=pulmonary function test; VCD=vocal cord dysfunction

Vocal cord dysfunction (VCD), described as paradoxical adduction of vocal cords during inspiration, has been reported in adolescents only rarely. Most of the cases reported have been in adults; VCD is not a well-recognized condition in children. VCD can be induced by exercise, and it can, therefore, resemble asthma: patients with misdiagnosed VCD have had frequent emergency department visits, prolonged hospitalizations, therapy with high-dose corticosteroids, intubation, and even tracheostomy.1-5 Two children with VCD are reported here in order to enhance the awareness of the physician about this condition.

CASE REPORTS

Case 1

A previously healthy 11-year-old girl was referred to the Pulmonary Clinic for evaluation of exercise-induced bronchospasm (EIB). She had a 2-week history of wheezing after exercise; one such episode of wheezing led to syncope while she was in the gymnasium. She had a history of prolonged viral infections as a child and “wheezing” in the past. The family medical history did not disclose asthma or atopy. Environmental history was unrevealing. Results of the physical examination were normal. Pulmonary function tests (PFTs) before and after exercise showed a greater than 15% decrease in FEV1 and more than 15% improvement in expiratory flow rates after bronchodilators. Albuterol by metered-dose inhaler was recommended prior to exercise but failed to bring complete relief. At 13 years of age, results of a physical examination and PFTs before and after exercise were within normal limits. Flow-volume loops after exercise showed flattening of the inspiratory limb. The expiratory-inspiratory flow ratio at 50% of vital capacity was 6.27 (normal, <1.00). In the following 3 weeks, she had 5 episodes of dyspnea; 4 of them were self-limiting but the fifth episode, following a fight with a friend, prompted her to visit the emergency department. She complained of episodes of tightness of the throat and chest, dyspnea, and wheezing. She looked very anxious, and an examination revealed inspiratory stridor mainly in the anterior area of the upper portion of the chest. No improvement was seen following use of bronchodilators. Flexible fiberoptic rhinolaryngoscopy showed adduction of the vocal cords during inspiration. She was referred for speech therapy and has been asymptomatic for the past 2 years.

Case 2

A previously healthy 12-year-old girl was referred to the Pulmonary Clinic for the evaluation of EIB. A few weeks before,