earlier diagnosis and intervention. It is not clear how the patient reported here inserted the needle into his heart. His story that he had been punched in the chest during a fight was not consistent with the evidence. The presence of numerous needles in his body, particularly in his abdomen and neck, suggested that they had been intentionally inserted and that the sewing needle also had been intentionally inserted into his heart.

There are few reports of self-inflicted injuries from needles in the heart in more recent medical literature. Eight cases of intracardiac injuries with needles have been reported in the past 20 years; the salient features of these reports are summarized in Table 1. Six of the eight cases were the result of self-injurious behavior. Seven of the eight cases were associated with serious complications including arterial embolization, mural thrombus formation, valvular dysfunction, cardiac tamponade, and constrictive pericarditis. In all of the cases, the needles were removed. In none of the cases was the needle completely intramyocardial in location or associated with the development of a pneumothorax.

A retrospective study on more than 200 intracardiac bullets and shrapnel included 23 cases in which the foreign bodies were fully intramyocardial; of these 23 patients, only 1 underwent surgery, an unsuccessful attempt at removal of the bullet. There were no deaths reported. The only complications included one case of pericarditis and the development of a fistula between the right coronary artery and the right ventricle in the patient in whom surgery was attempted. In a 1969 review, Schechter and Gilbert, however, emphasized that while needles which remain within the myocardium may remain clinically silent, their shape, in contrast to other foreign bodies such as bullets, also allows for rapid migration through tissues; there were three cases of the needles being extruded from the heart into the pleural space, but these cases were not associated with development of a pneumothorax.

A thorough discussion of the psychiatric aspect of this case is beyond the scope of this review, but it is of interest to note that self-mutilation does not appear in the fourth edition of the Diagnostic and Statistical Manual of Mental Disorders as a distinct entity. Self-mutilation can be observed in certain medical diseases, including the Lesch-Nyhan, de Lange, and Gilles de la Tourette’s syndromes and mental retardation, and can be associated with a variety of psychiatric conditions, such as personality and depersonalization disorders, schizophrenia, major depression, mania, and gender identity disorders. Self-mutilation is a diagnostic criterion in the borderline personality disorder and may be a component of factitious disorders. Alcohol and drug use increase the probability of such behavior. Self-mutilation can be thought of as an attempt on the part of the patient to relieve pathologic symptoms through multiple possible mechanisms. Such acts may reduce anxiety and tension especially when the patient is experiencing racing thoughts and varying emotions. These acts may also allow a cathartic release of anger, relieve depressive thoughts, remedy feelings of badness, and function as a means of drawing attention.

Although the patient may achieve the desired effects through self-mutilation, the underlying pathologic condition is not addressed, the relief provided is transient, and the resultant morbidity, if not mortality, is still a problem.

**Conclusion**

The case presented herein appears to be the only case of self-injury with a needle causing both a pneumothorax and cardiac penetration. This patient is most fortunate in that he did not suffer from any serious cardiac dysfunction on his initial presentation and in that he continues to do well from a medical standpoint despite apparent migration or embolization of the sewing needle.

**References**

3. Schechter DC, Gilbert L. Injuries of the heart and great vessels due to pins and needles. Thorax 1969; 24:246-53

**Lung Transplantation for Williams-Campbell Syndrome**

Scott M. Palmer, Jr., MD; Daniel T. Layish, MD; Peter S. Kassir, MD; Tim Oury, MD; Robert D. Davis, MD; and Victor F. Tapson, MD, FCCP

Williams-Campbell syndrome is a rare disorder characterized by a deficiency of cartilage in subsegmen-
tal bronchi leading to distal airway collapse and bronchiectasis. We report the first case of lung transplantation in a patient with end-stage lung disease secondary to Williams-Campbell syndrome. Although the patient did not have proximal airway collapse prior to transplantation, his posttransplant course was complicated by the development of bronchomalacia of the right and left mainstem bronchi. The patient experienced recurrent pulmonary infections and died of bacterial pneumonia 1 year after transplantation. Autopsy revealed cartilage deficiency in both right and left mainstem bronchi. A hypothesis may be made that a combination of proximal cartilage deficiency and posttransplant airway ischemia led to the development of bronchomalacia after lung transplantation. Thus, in contrast to previous reports, the cartilage deficiency in Williams-Campbell syndrome can involve both proximal and distal airways. Consequently, bilateral sequential lung transplantation may not be an effective therapeutic option in patients with this syndrome.

(CHEST 1998; 113:534-37)

Key words: bronchiectasis; congenital; lung transplantation

Abbreviations: OB=obliterative bronchiolitis

In 1960, Williams and Campbell described five children with symptoms of cough, wheezing, and recurrent pulmonary infections.1 Bronchography in these patients revealed thin-walled bronchiectasis with associated airway dilation on inspiration and collapse with expiration. Autopsy studies performed in one child revealed cartilage deficiency in the subsegmental bronchial tree. After Williams described an additional 11 children with similar features, the name Williams-Campbell syndrome was given to the constellation of airway collapse, bronchiectasis, and bronchial cartilage deficiency.2,3 Although the syndrome has been best described in children, there have been recent descriptions in adults as well.4 Recently, an adult with end-stage lung disease secondary to Williams-Campbell syndrome underwent bilateral lung transplantation.

Case Report

A 28-year-old white man was referred to Duke University Medical Center for consideration of lung transplantation for Williams-Campbell syndrome. The patient's childhood was remarkable for several episodes of pneumonia and the development of dyspnea on exertion by the age of 14. There was no family history of lung disease. The patient had smoked one pack of cigarettes per day from age 14 until 21 years. Despite significant exercise limitations, he declined medical evaluation until the age of 23 years. At the time of his initial presentation in 1990, the patient's arterial blood gas values, while breathing room air, revealed a Pao2 of 66 mm Hg and a PCO2 of 50 mm Hg. Pulmonary function tests revealed an FEV1 of 1.10 L (27% predicted), and FVC of 2.50 L (40% predicted), and a diffusion of carbon monoxide of 21% predicted. The flow-volume curve revealed no intrathoracic or extrathoracic airway obstruction. A CT scan of the chest revealed extensive thin-walled cystic bronchiectasis. Bronchoscopy confirmed expiratory collapse of the distal airways. Results of the α-1-antitrypsin level and sweat chloride tests were normal. A presumptive diagnosis of Williams-Campbell syndrome was made.4

The patient suffered a progressive decline in pulmonary function and was seen at Duke University Medical Center for consideration of lung transplantation. By that time, the patient had developed profound hypoxemia and hypercarbia. While breathing room air, arterial blood gas values were Pao2 of 31 mm Hg, PCO2 of 74 mm Hg, and a hemoglobin oxygen saturation of 57%. He required continuous therapy with 6 L of oxygen by nasal cannula. Echocardiography revealed normal left ventricular function with moderate right ventricular dysfunction and a negative saline microcavitation study. Catheterization of the right side of the heart revealed pulmonary artery pressures of 48/32 mm Hg. Eventually the patient developed respiratory failure requiring tracheostomy and nocturnal mechanical ventilation for several months prior to transplantation.

In August 1995, the patient underwent bilateral sequential lung transplantation. The cold ischemic time was 345 min for the left lung and 615 min for the right lung. The patient did well after the surgery and was discharged home on the 19th postoperative day. Maintenance immunosuppression therapy with cyclosporine, azathioprine, and prednisone was continued; he also received ganciclovir and cytomegalovirus immunoglobulin because of his cytomegalovirus mismatch status (donor positive-recipient negative). The patient also received trimethoprim-sulfamethoxazole for Pneumocystis carinii prophylaxis.

Pathologic examination of the patient's native lungs confirmed the diagnosis of Williams-Campbell syndrome. Cut sections of both lungs revealed numerous large cysts which measured up to 6 cm in maximum dimension and grossly had the appearance of sacular bronchiectasis. On histologic examination, sections from all lobes of both lungs contained numerous cyst-like spaces lined by ciliated respiratory epithelium. In addition, there were focal areas of paracartilaginous emphysema. A striking feature was the absence of cartilage in the walls of the medium to small airways (Fig 1). The sacular bronchiectasis, paracartilaginous emphysema, and diminished cartilage are all characteristic of Williams-Campbell syndrome.

After transplantation, the patient's pulmonary function significantly improved. While breathing room air, the patient's arterial blood gas values revealed a Pao2 of 72 mm Hg, a PCO2 of 44 mm Hg, and a 94% hemoglobin oxygen saturation. Spirometry revealed an FEV1 of 2.04 L with an FVC of 2.80 L. A surveillance bronchoscopy performed 1 month after transplantation revealed well-healed bronchial anastomoses with no evidence of necrosis or infection. However, bronchomalacia was noted with collapse of the native trachea and mainstem bronchi. Multiple surveillance transbronchial biopsies were performed after transplantation and revealed no evidence of infection or rejection. Several months after transplantation, the patient experienced recurrent bacterial pulmonary infections. Because the recurrent infections were believed to be related to proximal airway collapse, Palmaz stents (Johnson and Johnson Interventional Systems; Warren, NJ) were placed in the trachea and in the right and left mainstem bronchi. Continuous positive airway pressure was also used to maintain airway patency. Despite stent placement and continuous positive airway pressure, the patient required frequent

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hospitalization for recurrent bacterial pneumonia. Thirteen months after transplantation, the patient died of *Pseudomonas aeruginosa* pneumonia. Autopsy examination of the lungs confirmed bilateral acute and chronic bronchopneumonia. Extensive bronchiectasis was also found in the transplanted lungs. There was no acute or chronic allograft rejection, and viral immunostains were negative. A decreased amount of cartilage was observed in the patient’s native right and left mainstem bronchi.

**DISCUSSION**

Williams and Campbell1 and Williams and colleagues8 described a total of 16 children with an unusual type of bronchiectasis. All patients presented in early childhood with symptoms of cough, wheezing, and recurrent pulmonary infections. Physical examination often revealed a barrel-shaped chest, inspiratory and expiratory wheezes, and clubbing. Pulmonary function tests in most patients revealed moderate to severe obstruction with increased residual volumes and reduced diffusion of carbon monoxide. The chest radiograph and bronchography demonstrated thin-walled cystic bronchiectasis with associated ballooning of the smaller bronchi on inspiration and collapse with expiration. Pathologic studies performed in one child revealed absence of cartilage in subsegmental bronchi, leading Williams and Campbell to postulate a congenital cause for the syndrome.

Since these initial reports, there have been several other clinical and pathologic descriptions of Williams-Campbell syndrome. Mitchell and Bury3 provided a second detailed autopsy study in a child with suspected Williams-Campbell syndrome and noted absence of cartilage in smaller bronchi consistent with Williams’ initial report. Familial occurrence of Williams-Campbell syndrome has now been reported in two separate groups of patients and provides further support for a congenital cause in this syndrome.5,6 Several cases of adults with Williams-Campbell syndrome have been recently described, although without pathologic confirmation.4,7,8

The diagnosis of Williams-Campbell syndrome requires an appropriate clinical history, characteristic inspiratory airway collapse, and exclusion of other causes of congenital or acquired bronchiectasis,5,6 In Williams’ initial report, bronchography was used to identify airway collapse, although more recently, inspiratory and expiratory CT images have proven useful in the diagnosis of this syndrome.3,5 Other acquired and congenital conditions associated with bronchiectasis, including ciliary dyskinesia, cystic fibrosis, allergic bronchopulmonary aspergillosis, and immunoglobulin deficiencies, must be excluded.

The clinical course of Williams-Campbell syndrome is variable. Among the 21 children described in the English-language literature, five progressed rapidly to respiratory failure and death while the remainder survived into adulthood with variable degrees of recurrent infections and respiratory limitations.5 Long-term follow-up in these patients, however, is limited, and patients with Williams-Campbell syndrome who survive to adulthood may progress to respiratory failure, as was the case with the patient reported here. Although there is no specific therapy for Williams-Campbell syndrome, antibiotics and chest percussion are employed to treat recurrent infections associated with bronchiectasis.2 Surgical resection of severely bronchiectatic lobes has been described in this condition without significant improvement noted.5

Since there is no known therapy for Williams-Campbell syndrome, lung transplantation was considered in the patient reported here. Although lung transplantation has not been previously described in a patient with Williams-Campbell syndrome, bilateral sequential lung transplantation was considered a viable therapeutic option. Previous pathologic descriptions of patients with Williams-Campbell syndrome demonstrated cartilage deficiency and bronchiectasis confined to the distal airways, suggesting that bilateral sequential lung transplantation would be curative.

Although the patient initially did well after transplantation, his posttransplant course was complicated by the development of bronchomalacia of the mainstem bronchi contributing to recurrent pulmonary infections and death 1 year after transplantation. Autopsy studies performed in our patient revealed cartilage deficiency in the native right and left mainstem bronchi, in contrast to previous pathologic descriptions of Williams-Campbell syndrome. The proximal cartilage deficiency, however, was not as extensive as in the distal airways where a complete absence of cartilage was observed.

Because the patient did not have evidence of proximal...
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.

Bronchomalacia has been previously described in lung transplant recipients in association with obliterative bronchiolitis (OB).

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. 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. In 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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REFERENCES


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.

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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=palnt 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. 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.

Tracheal bronchus also may be seen in association with other congenital anomalies such as tracheoesophageal fistula, tracheal stenosis, and Down's (trisomy 21) syndrome.

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...