Airway Malformations and Bronchiectasis: A Pediatric Study

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Abstract
Congenital airway malformations are most often identified in early childhood. The development of bronchiectasis in association with malformations of the lower airway has been described, particularly among adult patients. The coexistence in a pediatric population of these conditions is not well described. This study was conducted to identify whether younger patients with airway malformations commonly develop bronchiectasis. International Classification of Diseases, Ninth revision (ICD-9 codes) were defined for airway anomalies and bronchiectasis. The electronic medical records system of a children’s hospital was then searched for the number of patients with upper airway anomalies with or without bronchiectasis. The airway database was then cross referenced with the ICD codes for bronchiectasis to identify patients with both conditions. There were 844 patients with airway anomalies and 117 with bronchiectasis in the electronic system during the time period of August 1, 2009 to September 30, 2014. There was only 3 patients identified with both bronchiectasis and airway anomalies. The coexistence of bronchiectasis is low among the pediatric population with upper airway anomalies studied. This would suggest that the children with airway anomalies have been treated with strategies that are effective in the prevention of recurrent lower respiratory tract infection. Further study may be done to define the effectiveness of various strategies in preventing aspiration and lower respiratory tract infection. In addition, this methodologic technique utilizing database integrative platforms is useful in the identification of patients for further study and to identify the coexistence of pediatric conditions.

Introduction
Congenital and acquired airway malformations may occur among infants and children. The defects can involve any anatomic region of the airway, from the nasal passages to the lower airway. There have been numerous reports identifying and describing an association between upper and lower airway disease. Among these conditions, bronchiectasis has been described among a subpopulation of patients with anatomic airway conditions.

Airway malacia results in localized airway collapse that can cause chronic cough. The consequence of airway malformation includes impaired local clearance mechanisms. This in turn may lead to severe localized infection, recurrent infections, and subsequent bronchiectasis. Bronchiectasis is a distortion of the airway resulting in dilation that can take the form of cysts, cylinders, or twisted distortion in the airway. These anatomic findings are termed cystic, cylindrical, and varicose forms of bronchiectasis. Recurrent infections of the airway can cause the development of bronchiectasis due to prolonged inflammation and chronic bacterial infection leading to destruction of the bronchial wall.

Williams-Campbell syndrome (WPS) is a rare syndrome, and cases of bronchiectasis are associated with the syndrome. Williams-Campbell syndrome is caused by defective or absent airway cartilage in subsegmental bronchi, which leads to distal airway collapse and bronchiectasis. Patients with WPS typically will have a normal trachea and proximal airway.

Although the coexistence of bronchiectasis and airway anomalies is better described in the adult population, the study of the pediatric association of these disorders is less well understood or described. The goal of this study was to describe the number of bronchiectasis cases among a pediatric population with airway anomalies. Airway anomalies of a wide variety were considered in studying a possible association given that these anomalies predispose to aspiration and infection of the lower airway.

Patients and Methods
Identification of Electronic Searchable Diagnostic Codes
The ICD diagnostic codes for upper airway anomalies and bronchial anomalies and bronchiectasis were identified. The ICD codes were then used to search the electronic medical records of a large urban children’s hospital.
Formulation of a Study Database

The electronic medical records database utilizes ICD coding to describe hospital and clinic outpatient visits. The center serves a population of more than 1 million patients.

The ICD code for anomalies of the larynx, trachea, and bronchi was 748.3. These included the airway anomalies of “atresia,” “dilation,” “diverticuli,” “anomalies,” and “agenesis” of the airway. The database was then searched for patients and patient visits with this ICD code. A subdatabase was formulated for use in this study. The diagnostic term “Bronchiectasis” ICD codes 494.0 and 494.1 were then cross referenced with the subdatabase. The patients who had both conditions were identified, and these were then reported as the number of cases with both conditions compared to total number with airway anomalies without bronchiectasis.

Additionally, the cohort with ICD codes 494.0 and 494.1 was further delineated to search for patients with multiple systemic anomalies (759.89). Similarly, the cohort with ICD code 748.3 was further cross referenced with the ICD code 759.89.

Statistical Methods

The method of description involves descriptive analysis of the number of visits and number of individual patients identified.

The calculations included (1) total number of patients with condition A and with condition B separately; (2) the total number of patients with both conditions A and B; (3) and the total number with A and B/total number with A alone.

Results

Subdatabase

The airway subdatabase was defined as patients with ICD codes 748.3 (congenital airway anomaly). Among the defects described were those patients with absence or agenesis, anomalies, atresia, dilation, stenosis, diverticuli, laryngoceles, and clefts of the airway. These 2 codes incorporated all of these listed anomalies. The airway subdatabase included patients seen between August 1, 2009, and September 30, 2014, at outpatient clinics and hospital facilities.

The number of patient visits for upper airway anomalies and conditions was 1863. The number of patients with upper airway anomalies was 844. There were 117 patients with bronchiectasis and 571 visits for this diagnosis during this time period. There were only 3 pediatric patients with both an airway anomaly and bronchiectasis. This suggests a 3 of 844 or 0.35%, occurrence of both conditions in a pediatric population. One possible explanation of this finding is that there is early recognition and intervention which is effective in preventing lower respiratory tract disease.

Among the patients with upper airway anomalies, 33 had multiple systemic anomalies. Among the patients with bronchiectasis, 4 had multiple systemic anomalies. The co-occurrence was 33 of 844 (3.9%) and 4 of 117 (3.4%), respectively.

Discussion and Conclusions

The development of dilation and distortion of the airway termed Bronchiectasis has been described in association with airway anomalies. The condition of congenital malacia or acquired narrowing of the airway may predispose to the development of Bronchiectasis. Aspiration and subsequent risk for lower respiratory tract infection is one possible mechanism.

The development of bronchiectasis may take years to develop following recurrent infections, or it can be defined as an anatomic dilation, for example, WPS. The occurrence of bronchiectasis among a pediatric population with malacia, airway anatomic defects with a variety of anomalies of the airway has not been extensively studied using a large database.

This study represents the first to the author’s knowledge to utilize a large pediatric database to identify the coexistence of both conditions and to describe the incidence of bronchiectasis among a pediatric population of patients with airway anomalies. While the study is a single-center study, the database is based on a large patient population in an urban county.

One congenital condition that has been associated with recurrent infections and bronchiectasis is Williams-Campbell syndrome (WCS). In one report of imaging in WCS, extensive cystic bronchiectasis was reported. The patient demonstrated airway ballooning during inspiration and collapse during the expiratory phase of respiration. These findings were described as an absence of cartilaginous rings from the main stem bronchi to subsegmental bronchi.

In the characterization of WCS, spiral dynamic computerized tomography (CT) can assist in establishing the diagnosis of bronchiectasis. The congenital malformation of the airway in WCS involves absence or diminished cartilaginous rings around the bronchi resulting in dilation and distal collapse. There has been at least 1 report of the familial occurrence of WCS among 2 related infants within 1 year of life. The developmental genetics though is not well defined.

There are other rare congenital conditions of the airway that have been linked to the development of lower airway recurrent infections and subsequently bronchiectasis. One such condition, Mounier-Kuhn syndrome (MKS) with less than 200 cases described since 1932, can be associated with tracheal and bronchial diverticuli. In MKS, there is abnormal dilatation of segments of the tracheal bronchial airway or tracheobronchomegaly. This rare disorder can result in pooled infected secretions and recurrent respiratory tract infections. This can in turn distort the airway and lead to bronchiectasis. In one study of the association of tracheomegaly and bronchiectasis among 75 adult patients referred for assessment of possible bronchiectasis, 17% were found to have both findings.

Among a pediatric population, chronic cough may be caused by airway malformations leading to decreased clearance and recurrent infections. The association between pediatric airway anomalies and chronic cough is established but not one of the more common causes. More common causes include asthma...
and bronchitis. The pathophysiologic connection between congenital anomalies to pediatric bronchiectasis is less clear. The findings would suggest that among a large database of pediatric patients (\(n = 844\)), there is a low incidence of associated bronchiectasis related to any cause, including immune defects.

While the study has drawbacks including the possibility of miscoding and missed cases during the electronic search, these are deemed to be unlikely to have a major impact on the findings, since the database included 844 patients with upper airway anomalies. The hospital system uses medical review and quality control to confirm ICD coding by the provider. The ICD code for airway anomalies was broad based and included those patients with multiple different airway anomalies.

The study suggests that the association of bronchiectasis and childhood airway anomalies warrants further study and that early detection and control of infection among these patients may prevent recurrent lung injury. As imaging and bronchoscopic techniques in viewing the dynamics of the airway improve, earlier detection is anticipated. Airway growth continues throughout development and parallels the somatic growth of children born with anomalies of the airway.

This study adds to the current base of clinical knowledge in this area and highlights that congenital malformations of the airway in children may not be highly associated with bronchiectasis. Prevention strategies include vaccination, surgical repair, antireflux and aspiration measures, speech therapy, prophylactic antibiotics, and maximization of somatic and lung growth.

Further investigation of early detection and treatment strategies may lead to a better understanding of the mechanisms of lower airway disease among patients with airway anomalies.

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