Bronchiectasis, Chronic Suppurative Lung Disease and Protracted Bacterial Bronchitis

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Bronchiectasis is a structural airway disease characterized by dilated bronchi and bronchioles due to severe or recurrent lower airways inflammation. Bronchiectasis can occur as a result of chronic pulmonary aspiration. Bronchiectasis may also be associated with a wide variety of systemic diseases, which should be considered in the differential diagnosis. Children with bronchiectasis typically have a chronic or recurrent productive cough and carry a significant burden of disease with a considerable impact on quality of life. The diagnosis of bronchiectasis is made by high-resolution chest computerized tomography. Aggressive management of bronchiectasis is necessary to reduce the daily symptom burden and frequency of exacerbations. Chronic suppurative lung disease may be a precursor to bronchiectasis, only lacking the defining radiographic features of bronchiectasis. Children with chronic suppurative lung disease may have the same symptoms as children with bronchiectasis and should be treated similarly. Protracted bacterial bronchitis is defined as a cough lasting at least four weeks that responds to antibiotic therapy. Protracted bacterial bronchitis may occur following a viral respiratory tract infection. Protracted bacterial bronchitis can be treated with a prolonged course of empiric antibiotics. Further evaluation is necessary if a child with suspected protracted bacterial bronchitis does not adequately respond to antibiotics as chronic suppurative lung disease or bronchiectasis must be considered.

1. Bronchiectasis is a structural airway disease caused by chronic or recurrent lower airways inflammation that results in bronchial dilation.
2. Children with bronchiectasis typically have a chronic or recurrent cough that carries a significant disease burden and negative impact on quality of life.
3. Patients with chronic suppurative lung disease may have the same symptoms as those with bronchiectasis, lacking only the distinguishing radiographic features of bronchial dilation seen with bronchiectasis.
4. Protracted bacterial bronchitis is defined as a wet cough lasting at least four weeks that responds to a course of antibiotics without a clear alternative etiology.

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Bronchiectasis

Bronchiectasis is a structural disease of the airways characterized by chronically dilated bronchi and bronchioles.[1] Bronchiectasis occurs when the lungs and lower airways are subjected to either an initial severe or ongoing insult. This insult results in mucosal inflammation and thickening, impaired mucus clearance and eventual damage to the bronchial wall and cartilage with resultant bronchial dilation. Bronchiectasis may be localized or diffuse. In advanced stages, the airways become collapsible and patients suffer from chronic obstructive lung disease. When bronchiectasis is associated with bronchial vascular proliferation, the patient may develop hemoptysis. Severe disease may also be associated with hypoxemia or pulmonary hypertension. Traditionally, bronchiectasis has been defined as irreversible dilation; however, early disease seen on high-resolution computed tomography (HRCT) scans has been noted to resolve in some cases.[2]

From an aerodigestive perspective, bronchiectasis can be associated with chronic pulmonary aspiration and is more likely to be found in children with neurologic impairment or a reported history of gastroesophageal reflux.[3] Other diagnoses must be considered in a patient presenting with bronchiectasis, including: cystic fibrosis (CF), primary ciliary dyskinesia (PCD), immunodeficiency syndromes and HIV infection, allergic bronchopulmonary aspergillosis (ABPA), severe or recurrent lower respiratory tract infections and post-obstructive pneumonia due to

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retained foreign bodies and congenital or developed obstructive airway lesions (Table 1).[4] Bronchiectasis may also be associated with bronchiolitis obliterans, connective tissue disorders, autoimmune disease, inflammatory bowel disease, or rare congenital airway malformations such as Williams-Campbell syndrome[5] or Mounier-Kuhn syndrome. Causative infectious agents include *Bordetella pertussis*, *Mycobacterium tuberculosis*, atypical *Mycobacterium species* and viral respiratory tract infections such as Influenza and Adenovirus. Many cases of bronchiectasis in children have no identifiable cause.[6,7]

Patients with bronchiectasis typically have a chronic or recurrent cough. The cough is usually loose and may be productive or associated with chest pain or dyspnea. The child may be treated for recurrent pneumonia or bronchitis. Focal or diffuse rales or rhonchi may be noted on physical examination; wheezing is a less common finding. Digital clubbing and chest wall deformities are more commonly found with advanced disease.[7] Failure to thrive may be present in some cases. There is significant reported burden of disease and impact on quality of life, especially during symptom exacerbations.[8]

TABLE 1. Diseases associated with bronchiectasis.

Post-infectious
Adenovirus
Influenza
Haemophilus influenzae
Pseudomonas aeruginosa
Bordetella pertussis
Mycobacterium tuberculosis
Atypical *Mycobacterium species*
Aspergillus
Immunodeficiency states
Primary immunodeficiency
Acquired immunodeficiency (post-chemotherapy, post-transplant)
HIV/AIDS
Cystic fibrosis
Primary ciliary dyskinesia
Chronic pulmonary aspiration
Allergic bronchopulmonary Aspergillosis (ABPA)
Bronchiolitis obliterans
Post-obstruction
Foreign body
Airway mass
Lymphadenopathy
Right middle lobe syndrome
Connective tissue disorders
Williams-Campbell syndrome
Mounier-Kuhn syndrome
Autoimmune disease
Inflammatory bowel disease
Yellow nail syndrome

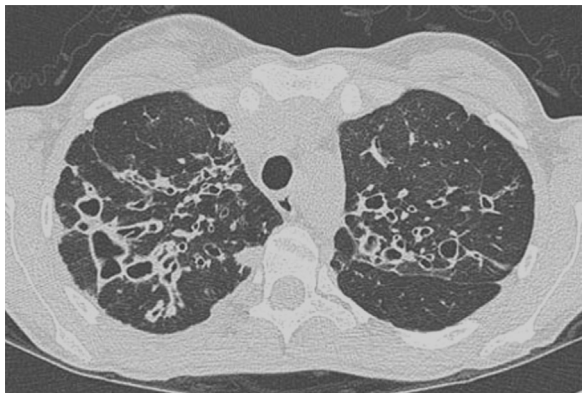

Figure 1. Bilateral bronchiectasis. Note thickened bronchial walls and varying degrees of bronchial dilation with bilateral cystic bronchiectasis.

Diagnosis of bronchiectasis is made via imaging. In cases of severe bronchiectasis, dilated airways may be identified by distinct parallel markings, or tram-tracking, on chest radiographs. A normal radiograph does not rule-out the presence of bronchiectasis; rather, imaging with HRCT should be used to make the diagnosis. The performance of magnetic resonance imaging (MRI) has steadily improved in assessing chronic lung disease[9,10]; however, HRCT remains the gold standard. Bronchiectasis is characterized by dilated peripheral bronchi, bronchial wall thickening, presence of bronchi within 1 centimeter of the pleura and bronchi with an internal diameter greater than that of the adjacent pulmonary artery, otherwise known as the "signet ring" sign.[11] The Reid classification defines bronchiectasis as (1) cylindrical, (2) varicose, and (3) cystic, the most severe form (Figure 1).[12] Flexible bronchoscopy may be part of the initial evaluation and bronchoalveolar lavage may be used to obtain lower airway specimen to help guide therapy.[13] There may be visual signs of severe bronchiectasis on flexible bronchoscopy. Severely dilated airways may not taper as expected; however, the presence of normal-appearing airways on bronchoscopy cannot rule-out bronchiectasis as a diagnosis. Bronchoalveolar lavage will reveal a neutrophilic predominance and offending organisms may be present. The most common organisms found in patients with bronchiectasis are *Haemophilus influenzae*, *Streptococcus pneumoniae*, *Moraxella catarrhalis*, *Staphlococcus aureus* and *Pseudomonas aeruginosa*.[14]

Determining the underlying etiology of the bronchiectasis is critical. A detailed history is performed to help guide testing. Additional evaluation may include a