Bronchiectasis is defined pathologically as the abnormal, irreversible dilatation of diseased bronchi. First described as a clinical entity by Laennec in 1819, it is technically not a disease but rather the outcome of various insults to the lungs. The widening is usually most extensive in the medium-sized bronchi at the segmental or sub-segmental level, where the permanently dilated airways are inflamed and often obstructed with thick, purulent secretions. Pooling of secretions leads to intermittent infection and inflammation and results in progressive destruction of the bronchial walls. The small peripheral airways are often involved because of proximal obstruction. Patients with advanced disease have dilatation of the small distal airways.

In this review, we discuss causes of bronchiectasis, common signs and symptoms, diagnostic evaluation, and treatment with primary and adjunctive therapies. In addition, we present 3 case studies illustrating the management of different presentations of bronchiectasis: postinfection bronchial damage, postinhalation injury, and congenital airway obstruction.

ETIOLOGY

The cause of bronchiectasis is unknown in more than half of cases. Pulmonary conditions known to predispose patients to bronchiectasis include postinfection bronchial damage, postinhalation injury, hypersensitivity reactions, and congenital airway obstructive disorders (Table 1). Infection is the most common identifiable cause of bronchiectasis in middle-aged and elderly people. In developed countries, congenital disorders such as cystic fibrosis (CF) are the most likely cause of bronchiectasis in younger patients, whereas sequelae of pulmonary tuberculosis, pertussis, and severe measles are common causes in developing countries. Regardless of the cause, intense inflammation and inadequate host defense mechanisms are the 2 factors common to all cases of bronchiectasis.

SIGNS AND SYMPTOMS

The hallmark of bronchiectasis is sputum overproduction. Typical symptoms include fever, pleurisy, dyspnea, and chronic cough that produces pu-
rulent, often foul-smelling sputum with or without hemoptysis. The quantification of daily sputum production may be a useful indicator of disease severity. Less than 10 ml per day suggests mild bronchiectasis, whereas greater than 150 ml per day indicates severe bronchiectasis. Some patients, especially those with upper lobe involvement, may have cough with little or no sputum production, referred to as dry bronchiectasis.

Hemoptysis is a relatively common feature of bronchiectasis, occurring in approximately 50% of patients. It can vary from streaks in the sputum to massive bleeding; however, life-threatening hemoptysis is rare. Generally, small amounts of hemoptysis, which may occur with infective exacerbations of bronchiectasis, are not a cause for concern unless the bleeding persists. However, if massive amounts do occur, vigorous supportive therapy must be instituted. Urgent bronchoscopy should be performed to locate the site of bleeding, and, if conservative measures are ineffective, the patient can be treated by either bronchial artery embolization or resection of the involved lung segment.

Patients frequently have a persistent, year-round productive cough. The cough tends to worsen when patients are supine because secretions accumulate while the patient is in the dependent posture. Clubbing is frequently noted in moderate or advanced cases. Wheezes, rhonchi, crackles, and a pleural rub are often evident on auscultation of the lungs.

Exacerbations of bronchiectasis, considered any deviation from the patient’s baseline, can vary from mild to severe. A mild exacerbation involves a worsening of baseline cough and increased sputum production, with changes in sputum color and consistency. A moderate exacerbation is characterized by increased cough, increased sputum production with a change in color and consistency, increased fatigue, decreased appetite with or without weight loss, and a decrease in pulmonary function as measured by standard pulmonary function tests. A severe exacerbation is similar to a moderate one but is further characterized by a greater decrease in pulmonary function.

**DIAGNOSTIC EVALUATION**

The prevalence of bronchiectasis is probably underestimated. A common diagnostic challenge is to differentiate bronchiectasis from chronic bronchitis, which is similarly characterized by a daily productive cough and dyspnea. For many years bronchograms were used to differentiate the two diagnoses. Three patterns of bronchiectasis were classified by Reid in 1950: cylindrical or fusiform, varicose, and cystic or saccular. The clinical relevance of these 3 types is disputed, although the cystic type is generally thought to represent advanced disease.

This classification system was once very useful as a diagnostic aid when bronchography was the standard test to identify bronchiectasis. In recent years, however, computed tomography (CT) has replaced bronchograms as the definitive diagnostic test for bronchiectasis.

**Radiologic studies**

In bronchiectasis, chest radiographic findings may or may not be abnormal. Abnormalities, when present, appear as nonspecific increased linear markings-parallel “tram lines” or “tram tracking”—indicating lack of bronchial tapering. Cystic spaces and honeycomb-like structures may be visible in the films of patients with more severe disease.

In recent years, high-resolution CT has become the gold standard for diagnosis of bronchiectasis. Thin-section CT has a sensitivity of 87% to 97% and a specificity of 93% to 100%. The abnormal signet ring sign is a common CT finding. In normal subjects, the bronchus and adjacent artery appear similar in size. In patients with bronchiectasis, however, the dilated bronchus appears larger than the artery, giving the appearance of a ring set with a stone—the “ring” being the wall of the dilated bronchus and the “stone” a cross-sectional image of the adjacent pulmonary artery.

**Pulmonary function testing**

Pulmonary function testing usually reveals an obstructive defect, with a reduced ratio of forced expiratory volume in 1 second to forced vital capacity (FEV/FVC). A restrictive defect may also be noted, particularly in patients with advanced disease and extensive parenchymal destruction.

**Bronchoscopy**

Bronchoscopic examination is useful primarily in evaluating the proximal airways for obstructing lesions and in assessing and localizing sources of hemoptysis. Bronchiectasis is a disease of distal airways; therefore, there is little additional indica-
tion for bronchoscopy in the diagnosis of bronchiectasis.

**TREATMENT**

Treatment of bronchiectasis is multifaceted. The goals of treatment are to reduce or eliminate the underlying host deficiency, improve clearance of secretions, control acute infections, relieve airway obstruction, reduce the load of the colonizing organism, and reduce inflammation. Inhaled bronchodilators help to relieve bronchoconstriction, and steroids reduce airway inflammation.

**Antibiotic therapy**

Antibiotic therapy is used to control acute infections, reduce organism load, and reduce inflammation. Therapy typically includes the use of oral, intravenous (IV), or aerosolized antibiotics, individually or in combination. The choice of antibiotics should be driven by sputum sensitivities. A list of common infecting organisms is given in Table I.

Oral antibiotics are routinely used for treatment of mild exacerbations. Most commonly preferred is an oral quinolone to which the organism is sensitive. Often, a second oral antibiotic is used in conjunction, usually a β-lactam or macrolide. Patients with bronchiectasis associated with CF typically require higher dosages or more frequent dosing intervals. The normal course of treatment in bronchiectasis is 14 days and is frequently accompanied by a tapering dose of prednisone to help decrease inflammation.

Treatment of moderate exacerbations may include the use of IV antibiotics (home or inpatient), a course of prednisone given for a longer period than for mild exacerbations, and aggressive bronchial hygiene. Antibiotics are administered for at least 2 weeks and are tapered in accordance with resolution of symptoms.

Treatment of severe exacerbations includes inpatient hospitalization, IV antibiotics, IV or oral steroids, and aggressive bronchial hygiene.

**Aerosolized antibiotics**

Aerosolized antibiotics are used for both acute and chronic therapy for bronchiectasis resulting from *Pseudomonas aeruginosa* infection. In both moderate and severe exacerbations, aerosolized antibiotics are used acutely in conjunction with IV antibiotics. Solutions most frequently aerosolized in *Pseudomonas* bronchiectasis are tobramycin solution for inhalation, Colistin, and β-lactam antibiotics. Tobramycin solution for inhalation (TOBI) has been shown to significantly improve lung function, lower the density of *P. aeruginosa* in sputum, and decrease the length of hospital stays in CF bronchiectasis. The drug is dosed at 300 mg and administered via nebulizer twice daily for a 28-day cycle, followed by a cycle of 28 days off therapy. Additional studies of the use of this drug in patients with bronchiectasis who do not have CF are under way.

**Bronchial hygiene**

Most of the morbidity and mortality in bronchiectasis is associated with pulmonary complications and respiratory failure. Evidence suggests that the progression of airway damage is the result of recurrent infection. It is postulated that the airway infections are accompanied by an inflammatory response, along with the release of neutrophilic proteases that damage the airways, leading to more severe bronchiectasis. Large amounts of secretions are produced in the diseased airways and are difficult for the patient to expectorate because of loss of mobility mechanisms. These secretions “pool” in the bronchi and become the media for the organisms responsible for these recurrent infections, allowing the cycle to continue. Clearing this sputum from the airways is a consistent problem in managing bronchiectasis. Adjunctive therapies, such as chest physiotherapy (CPT) and postural drainage (PD), have been shown to increase sputum clearance and help prevent acute episodes, but these techniques are labor intensive and costly in the acute setting. Newer devices have been developed that have been shown to be more effective in increasing sputum expectoration and easier for both patient and caregiver.

Studies have shown that the Flutter (Vario Raw Percutive, Aubonne, Switzerland), a mucus clearance device, significantly aids in expectoration of mucus. It is a small, hand-held device in the shape of a pipe. Within the Flutter is a high-density stainless steel ball resting in a plastic cone. As the patient exhales, the ball rolls out of place and back many times, resulting in variance of endobronchial pressure and expiratory airflow. The oscillations that are produced loosen mucus from the airway walls, and the acceleration of airflow moves the mucus up the airways. The oscillations also decrease collapsibility of the airways. As noted by Konstan et al., therapy with the Flutter results in an increased amount of sputum expectorated compared with voluntary coughing or postural drainage.

A newer way to administer CPT is through the use of high-frequency chest compression (HFCC). The
ThAIRapy vest (also known as The Vest Airway Clearance System, American Biosystems, Inc; St Paul, MN) is a portable HFCC system. It is an inflatable, tailored, fitted vest jacket attached to a large pump that generates high-frequency oscillations. The resulting pressures cause the vest to inflate and deflate against the chest wall. In this passive technique, the vibrations cause transient increases in airflow, resulting in improved mucus mobilization.

Studies have shown HFCC to be equally safe and effective when compared with CPT during acute pulmonary exacerbations, with the advantage of being easier to administer, less expensive, and less time-consuming. These studies present encouraging data on adjunctive therapies such as HFCC for bronchiectasis. In general, therapies that are portable and user friendly promote better compliance and improved bronchial hygiene.

Surgical therapy

In developing countries, bronchiectasis continues to be a major cause of morbidity and mortality. Higher rates of human immunodeficiency virus- and tuberculosis-related infections are associated with the increased incidence of bronchiectasis. Even with conservative medical therapy (antibiotics and PD), mortality rates remain high. In these severe situations surgical resection of the involved segments provides a possibility of cure as long as the disease is localized.

In a retrospective study conducted in Saudi Arabia, 85 patients with unilateral or bilateral bronchiectasis who had failed medical therapy and continued to suffer from a productive cough, hemoptysis, or failure to thrive, received surgical intervention. There were no surgical mortalities, and 63 patients had excellent results. The authors reinforced the need for careful CT and V/Q lung scanning before surgery not only to classify the disease, but to avoid recurrent postoperative symptoms.

In a 7-year review of patients in Germany, surgery was the intervention for those with recurrent pneumonias, significant hemoptysis, lung abscess, or empyema. It was found that the patients with cylindrical bronchiectasis were the best candidates. It was also noted that concomitant chronic sinusitis affected surgical prognosis adversely. In a review of 119 patients in Portugal who received surgical intervention for bronchiectasis, it was found that 68% of the patients were asymptomatic 4.5 years after surgery. Those who had complete resection of the disease had the best outcomes.

Although the rate of post surgical mortality is low, there remains a need for randomized studies comparing the benefits of surgical intervention with conservative therapy for bronchiectasis.

CASE STUDIES

Case study 1

The patient, an 82-year-old white woman, had a history of frequent episodes of bronchitis and daily sputum production, including a childhood history of frequent episodes of cough, bronchitis, and pneumonia. Sputum production had increased over the several days before she arrived and was yellow in color. She denied having any fever or chills. At 26 years of age, she was diagnosed with bronchiectasis after undergoing a bronchoscopic procedure. She did well until she reached her sixties when she noted increased dyspnea with activity, cough, and wheezing. In addition, she was hospitalized twice for pneumonia.

On physical examination, there were diffuse wheezes and crackles bilaterally and new lower-extremity edema. Her oxygen saturation was 90% on room air at rest. Chest radiograph showed a volume loss of the left lung and pleural changes over the left apex and base. On spirometry, her FEV1 was 0.82 L (46% of predicted). An echocardiogram revealed elevated pulmonary artery systolic pressures estimated at 70 to 75 mmHg.

Because of her history of bronchiectasis and increasing sputum production, she was given 500 mg of oral azithromycin as a single dose on day 1 and 250 mg on days 2 to 21 (prescribed for a total of 21 days), and she was given 40 mg of oral prednisone once a day with a tapering dose. Before she could return to clinic, she was admitted to the hospital with acute dyspnea and wheezing. Her sputum culture was positive for P aeruginosa. The antibiotic was changed to 500 mg of levofloxacin. In addition, it was noted that with activity, her oxygen saturation dropped to 86%, therefore, home-based oxygen therapy at 2 L by nasal cannula was prescribed.

Because of her frequent hospitalizations, inhaled TOBI (twice a day via nebulizer) was added to her therapy on a 28-days-on/28-days-off schedule on a limited basis. She subsequently returned to the clinic on an as-needed basis for HFCC with the ThAIRapy vest, continued to use her home oxygen, and joined a pulmonary rehabilitation program that she continues to attend.

Case study 2

The patient was a 42-year-old white man who had been in generally good health until 3 years previously, when he was injured in an apartment fire. He
suffered carbon monoxide poisoning and smoke inhalation that caused a chemical pneumonitis that resulted in bronchiectasis and bronchiolitis. He was also left with vocal cord paralysis from emergent intubation.

His symptoms included shortness of breath on moderate exertion and a persistent cough productive of a large amount of yellow-green sputum. The productive cough was worse in the morning but persisted throughout the day, and it became problematic for the patient in his occupation as a waiter. On physical examination, coarse crackles were heard bilaterally, louder at the right base. Possible early clubbing of the fingers was also noted.

Chest radiograph showed cystic changes consistent with bronchiectasis at the lung bases in a reticular nodular pattern (Fig 1). High-resolution CT of the chest revealed cystic bronchiectasis involving both lower lobes, as well as varicose bronchiectasis involving the upper lobes (Fig 2). Spirometry revealed a mild obstructive pattern, with an FVC of 2.63 L (72% of predicted), an FEV₁ of 1.79 L (60% of predicted), and an FEV₁/FVC ratio of 68%.

He was treated with a triamcinolone acetonide steroid inhaler, an albuterol bronchodilator inhaler, and theophylline. Intermittent pulmonary infections were treated empirically with various antibiotics, and during these exacerbations the dosage of the steroid inhaler was temporarily increased until the infection resolved. Although coughing spasms and purulent sputum have continued to be a problem for him, he has not been hospitalized and has remained stable.

Case study 3

The patient was a 29-year-old white woman with CF diagnosed at birth. She became colonized with _P. aeruginosa_ in her early twenties and has had an exacerbation once or twice a year since then. During each exacerbation, she has responded well to standard IV antibiotic therapy while hospitalized.

Four years previously she was infected with a resistant strain of _P. aeruginosa_ that was treated successfully with nebulized standard tobramycin. During the next 2 years, she required 5 courses of IV antibiotics and 2 admissions to the hospital. During this time, her spirometry readings showed a drop in FEV₁ from 2.16 to 1.60 L.

Two years earlier, after completing a 2-week course of IV antibiotics, she reported that the antibiotics had failed to resolve her CF exacerbation. At this time, she was given 300 mg of TOBI twice a day via jet nebulizer on a 28-days-on/28-days-off schedule. After two cycles of TOBI and a 3-week course of IV antibiotics, she noted that she was feeling better than she had at baseline. Her FEV₁ had improved...
CONCLUSION

The case studies illustrate 3 different presentations of bronchiectasis: postinfection bronchial damage, postinhalation injury, and congenital airway obstruction. The 3 patients discussed share a common need for consistent bronchial hygiene and pharmacologic intervention.

For patients with bronchiectasis, the constant cough, sputum production, and dyspnea from obstruction of airways by purulent secretions often result in physical inactivity. This pulmonary deconditioning is similar to that seen in patients with other chronic lung diseases. Rehabilitation including education and exercise may increase muscle strength, optimize medication usage, and decrease the work of breathing. The social contact and psychosocial support of the group setting may help patients improve their quality of life and become more independent. Although it would seem intuitive that a program of pulmonary rehabilitation may be useful for patients with bronchiectasis, no confirming studies have been performed to date. Future research is needed to support our hypothesis that pulmonary rehabilitation should be considered as adjunctive therapy in the plan of care.

REFERENCES