Chronic Cough Due to Bronchiectasis
ACCP Evidence-Based Clinical Practice Guidelines

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Background: Bronchiectasis is a condition that is characterized by the permanent dilation of bronchi with destruction of elastic and muscular components of their walls, usually due to acute or chronic infection. The cardinal symptom is a chronic productive cough.

Methods: Review of articles cited in the systematic literature search, along with others found in Ovid MEDLINE and the Cochrane Library (including the Cochrane Database of Systematic Reviews, the Cochrane Controlled Trial Register, and the Database of Abstracts of Reviews of Effectiveness) from 1966 through 2003.

Results/conclusions: High-resolution CT scanning of the chest is the preferred means of establishing the diagnosis of bronchiectasis. With the increasing use of antibiotics in the treatment of childhood infection in the last several decades, an increasing percentage of patients with bronchiectasis now have an underlying disorder that predisposes them to chronic or recurrent infection. These include cystic fibrosis, common variable immunodeficiency, HIV infection, primary ciliary dyskinesia, allergic bronchopulmonary aspergillosis, and chronic Mycobacterium avium complex infection. A variety of agents have been used to improve cough effectiveness and prevent infectious exacerbations in patients with bronchiectasis, with variable results. Chest physiotherapy offers a modest benefit in increasing sputum volume, but its long-term effectiveness is unknown. Selected patients with localized idiopathic bronchiectasis that causes intolerable symptoms despite maximal medical therapy should be offered treatment with surgery. Patients with exacerbations of bronchiectasis should be given antibiotics, with the choice of agents depending on the likely causative pathogens.

(CHEST 2006; 129:122S–131S)

Key words: allergic bronchopulmonary aspergillosis; antibiotics; bronchiectasis; bronchodilators; chest physiotherapy; cystic fibrosis; mucolytics; Mycobacterium avium complex; primary ciliary dyskinesia

Abbreviations: ABPA = allergic bronchopulmonary aspergillosis; CF = cystic fibrosis; GERD = gastroesophageal reflux disease; HRCT = high-resolution CT; MAC = Mycobacterium avium complex; PCD = primary ciliary dyskinesia; rhDNase = recombinant human DNase

Bronchiectasis is a condition characterized by the permanent dilation of bronchi with destruction of the elastic and muscular components of their walls, usually due to acute or chronic infection. Bronchiectasis due to recurrent or severe infection in patients without an apparent underlying cause has become less common with the increasing use of immunization and antibiotics in treating childhood infections, and with the declining incidence of pulmonary tuberculosis. In the last few decades, an increasing proportion of patients with bronchiectasis in developed countries have been found to have an underlying disorder (disorders are summarized in Table 1). Regardless of the etiology, the cardinal symptoms of bronchiectasis are chronic cough and sputum production, although some patients may have a nonproductive cough. Recurrent bacterial colonization and infection lead to progressive airway injury that is mediated by neutrophils, T lymphocytes, and monocyte-derived cytokines. The actions of inflammatory mediators, elastase, and...
collagenase lead in turn to inflammation and the destruction of the elastic and muscular components of bronchial walls, while the contractile force of the surrounding lung tissue exerts traction, expanding the diameter of the involved airways. Bronchiectasis is also associated with increased bronchial arterial proliferation and arteriovenous malformations, predisposing some patients to recurrent hemoptysis.

Although bronchiectasis can be classified radiographically and pathologically into cylindrical, varicose, and cystic varieties, these distinctions are not helpful in determining the etiology, treatment, and prognosis in individual patients. Bronchiectasis may have a focal or diffuse distribution. Focal bronchiectasis may follow a severe case of pneumonia, but with increasing use of antibiotics in recent years it is more often caused by bronchial obstruction by a foreign body, broncholith, a slowly growing tumor, anatomic distortion following lobectomy, or enlarged peribronchial lymph nodes. These predispose the patient to recurrent infection and bronchiectasis distal to the site of obstruction. Focal bronchiectasis due to bronchial obstruction is important to recognize because it may be amenable to local intervention with bronchoscopic procedures or surgery. Diffuse bronchiectasis is usually caused by an underlying disorder (Table 1).

### Table 1—Disorders That Predispose the Patient to the Development of Bronchiectasis

<table>
<thead>
<tr>
<th>Disorders</th>
<th>Description</th>
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<tbody>
<tr>
<td>Focal distribution</td>
<td>Bronchial obstruction: Foreign body, Tumor, Broncholithiasis</td>
</tr>
<tr>
<td>Previous pneumonia</td>
<td>Compression by peribronchial lymph nodes</td>
</tr>
<tr>
<td>Diffuse distribution</td>
<td>CF, Reduced host immunity, Congenital and acquired hypogammaglobulinemia (especially IgG and/or IgG subclasses), HIV infection</td>
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<tr>
<td>Primary ciliary dyskinesia</td>
<td>α1-antitrypsin deficiency, Tracheobronchomegaly (Mounier-Kuhn syndrome)</td>
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<tr>
<td>Allergic bronchopulmonary mycoses</td>
<td>Cartilage deficiency (Williams-Campbell syndrome)</td>
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<tr>
<td>Chronic MAC infection</td>
<td>Young syndrome</td>
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<tr>
<td>Aspiration or toxic inhalation</td>
<td>Pulmonary sequestration</td>
</tr>
<tr>
<td>Rheumatoid arthritis</td>
<td>Marfan syndrome</td>
</tr>
<tr>
<td>Inflammatory bowel disease</td>
<td>Other congenital disorders</td>
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<td></td>
<td>Yellow nail syndrome</td>
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</table>

Although chronic productive cough is the cardinal feature of bronchiectasis, this symptom occurs far more commonly in patients with chronic bronchitis, asthma, upper airway cough syndrome due to a variety of rhinosinus diseases, previously referred to as *postnasal drip syndrome*, and gastroesophageal reflux disease (GERD). In a prospective, before-and-after intervention trial, a cohort of 71 immunocompetent adults who complained of expectoration of > 30 mL of sputum per day underwent a detailed protocol-driven diagnostic evaluation; upper airway cough syndrome was the cause in 40% of patients, asthma in 24% of patients, GERD in 15% of patients, bronchitis in 11% of patients, and bronchiectasis in only 4% of patients. Unusual causes of chronic cough with excessive sputum production may be clinically unsuspected initially; these include non-asthmatic eosinophilic bronchitis and bacterial infections.

### Materials and Methods

The articles produced by the Duke systematic literature search were reviewed (see section 2 of this guideline), along with others found in searches of Ovid MEDLINE and the Cochrane Library (including the Cochrane Database of Systematic Reviews, the Cochrane Controlled Trial Register, and the Database of Abstracts of Reviews of Effectiveness) from 1966 through 2003.
suppurative airway disease. In the latter disorder, patients do not have excessive sputum expectoration and may not have radiographic evidence of bronchiectasis, but have copious purulent secretions found on bronchoscopy.

Excessive sputum volume, purulence, and tenacity are nonspecific findings. The physical examination findings in patients with bronchiectasis may reveal variable degrees of rhonchi, crackles, and clubbing, or they may be normal. Also, the presence or absence of crackles on auscultation of the chest does not correlate with the presence of bronchiectasis as diagnosed by high-resolution CT (HRCT) scanning of the chest. Sputum cultures are often positive for Haemophilus influenzae, Staphylococcus aureus, Streptococcus pneumoniae, and Pseudomonas aeruginosa, but these pathogens may also be found in the airways of patients with chronic bronchitis. The isolation of mucoid strains of Pseudomonas suggests the diagnosis of cystic fibrosis (CF), isolation of Aspergillus suggests allergic bronchopulmonary aspergillosis (ABPA), and isolation of Mycobacterium avium complex (MAC) suggests chronic infection with that organism, but isolating these organisms is not specific for these disorders.

Because bronchiectasis is defined as abnormal dilation of airways, the diagnosis depends on visualizing the typical changes either radiographically or anatomically. Bronchiectasis is sometimes obvious on routine chest radiographs, but the diagnosis is usually established using HRCT scanning. This test compares favorably with bronchography, which had been considered a “gold standard,” but bronchography is now rarely performed because it is invasive, unpleasant, and risky, and because the HRCT scan is as accurate in diagnosis, with sensitivity and specificity exceeding 90%.

The key feature of bronchiectasis on HRCT scans is enlarged internal bronchial diameter, where the bronchi appear larger than the accompanying artery (called signet ring sign). Other HRCT scan findings in bronchiectasis include the failure of the larger airways to taper while progressing to the lung periphery, air-fluid levels in dilated airways, and the identification of airways in the extreme lung periphery. Indirect HRCT scan signs of bronchiectasis include bronchial wall thickening, mucoid impaction, and focal air-trapping.

**Recommendation**

1. In patients with suspected bronchiectasis without a characteristic chest radiograph finding, an HRCT scan should be ordered because it is the diagnostic procedure of choice to confirm the diagnosis. Level of evidence, low; benefit, substantial; grade of recommendation, B

**Specific Causes of Bronchiectasis**

Many cases of bronchiectasis result from permanent bronchial wall damage after a childhood respiratory infection, such as bacterial pneumonia, pertussis, or measles. In patients with healed tuberculosis or other fibrosing lung diseases (eg, fungal infection, sarcoidosis, or idiopathic pulmonary fibrosis), traction on major airways from surrounding fibrotic lung parenchyma may distort their architecture, leading to recurrent infection and traction bronchiectasis. A systematic search for an underlying cause of bronchiectasis often yields a diagnosis that is amenable to treatment. In a study of 150 adults with bronchiectasis, one or more causative factors were identified in 47% of cases, and the diagnosis had important prognostic or therapeutic significance in 15% of cases. In the latter category, patients with CF were referred to a specialty clinic, patients with common variable immunodeficiency were started on Ig replacement therapy, patients with ABPA were given a trial of corticosteroid treatment, and a patient with recurrent aspiration associated with GERD was treated with an intensive regimen of gastric acid suppression.

**Recommendation**

2. In patients for whom there is no obvious cause, a diagnostic evaluation for an underlying disorder causing bronchiectasis should be performed, because the results may lead to treatment that may slow or halt the progression of disease. Level of evidence, low; benefit, substantial; grade of recommendation, B

**CF**

Occurring in 1 per 2,000 to 3,000 live births, CF is probably the most common identifiable cause of bronchiectasis in the United States and Europe. A mutation of the gene that encodes for the CF transmembrane regulator protein disrupts the normal flow of fluid and electrolytes across cell membranes, altering the composition of secretions in the respiratory tract, pancreas, GI tract, and sweat glands. In the lung, this is manifested by excessive and tenacious mucus secretion, impaired mucus transport, dilation and hypertrophy of bronchial glands, and goblet cell hyperplasia, leading to proximal airway plugging and bronchiectasis. This diag-
nosis should be considered in children and younger adults with recurrent sinopulmonary infection and/or bronchiectasis, even in the absence of GI symptoms. The sweat test (called pilocarpine iontophoresis) is still the most accurate way to diagnose CF, as the result is positive in almost all cases when performed properly.23 Testing for the CF gene is less sensitive, as there are >700 mutations identified; commercial laboratories identify around 90% of CF mutations in the general population.

Reduced Host Immunity

Patients with congenital and acquired hypogammaglobulinemia (especially involving IgG and/or IgG subclasses) have an increased frequency of recurrent rhinosinusitis, respiratory infections, and bronchiectasis.24 Hypogammaglobulinemia is diagnosed by measuring the serum levels of IgG and IgG subclasses, along with those of IgM and IgA. In primary hypogammaglobulinemias, the IgG level is typically <5 g/L, and the IgA level is <0.1 g/L; antibody deficiency can be confirmed by documenting decreased antibody production to two or more vaccines, including tetanus, diphtheria, measles, mumps, and pneumococcal vaccine. Patients with IgG deficiency may benefit from IV Ig replacement, which is associated with a reduced frequency of infection and a slowing of the progression of bronchiectasis.25,26 Even though this therapy is considered to be the standard of care for primary immunodeficiency diseases with hypogammaglobulinemia, there are no double-blind placebo-controlled trials evaluating threshold Ig levels for starting treatment, the effect of the standard dose of IV Ig (400 mg/kg every 28 days) on outcomes, and whether higher doses are more effective.26 Those with isolated IgG subclass deficiency and recurrent infection may benefit from therapy with IV Ig, but this has not been investigated in controlled clinical trials.26

Patients with HIV infection are also predisposed to developing recurrent sinopulmonary infections and bronchiectasis, probably because of abnormal B-lymphocyte function.27,28 The treatment of patients with HIV infection and bronchiectasis with IV Ig has not been studied systematically. The impact of combination antiretroviral therapy on the severity and course of airway disease is also unknown, although the restoration of immune function would be expected to be beneficial.

Primary Ciliary Dyskinesia

This refers to a group of autosomal-recessive disorders associated with defective ciliary structure (almost always in the dynein arms) and function.29 Absent or discoordinated movement of cilia in the respiratory tract leads to recurrent sinus, ear, and lung infections, which in turn leads to bronchiectasis. The name Kartagener syndrome refers to the triad of situs inversus totalis, bronchiectasis, and either nasal polyps or recurrent sinusitis, occurring in approximately half of patients with primary ciliary dyskinesia (PCD).30 Male patients with PCD are almost always infertile due to immotile sperm.29 The disorder is diagnosed by assessing ciliary motility in semen samples or in bronchial wall or nasal biopsy specimens, or by electron microscopy that shows abnormal cilia morphology. In a study29 of 78 patients with PCD diagnosed using a combination of compatible clinical features and together with tests of ciliary ultrastructure and function, chronic rhinitis/sinusitis occurred in all patients, recurrent otitis media occurred in 95% of patients, neonatal respiratory disease occurred in 73% of patients, and situs inversus occurred in 55% of patients. All patients had defects in the ciliary structure, two thirds of which occurred in the outer dynein arm. Nasal nitric oxide production was very low compared with that in healthy subjects and may be of diagnostic value. H influenzae was the most common bacterial isolate found in sputum, but Pseudomonas isolates were also common. Although the isolation of mucoid strains of Pseudomonas in sputum is considered to be a marker of CF, it was found in 15% of subjects with PCD.

ABPA

Immune-mediated inflammation in response to inhaled Aspergillus fumigatus antigen causes this disorder in some patients with chronic asthma or CF.31 When a similar disorder is caused by other species of Aspergillus or other fungi, the term allergic bronchopulmonary mycosis may be preferable.32 The essential diagnostic criteria for the diagnosis of ABPA in patients with bronchiectasis are the central distribution of disease (inner two thirds of chest on CT scan), asthma, immediate cutaneous reactivity to Aspergillus antigen, elevated total serum IgE level (>417 IU/L or 1,000 ng/mL), and elevated serum levels of IgE-A fumigatus (Aspergillus precipitins) or IgE-A fumigatus in comparison to those seen in sera from skin-test positive patients who did not have ABPA.33 Patients may have pulmonary infiltrates and eosinophilia, but these findings are not needed to establish the diagnosis. Mucoid impaction of the bronchi and bronchocentric granulomatosis are also associated with ABPA.

As the disease is a manifestation of a hypersensi-
tivity reaction rather than an infection, treatment is aimed at immune modulation. The administration of oral prednisone to patients with ABPA is associated with the improvement of asthma, and the presence of pulmonary infiltrates and eosinophilia, with reduction in serum levels of IgE, and probably reduced progression of bronchiectasis. The optimal dose is unknown, but 0.5 mg/kg/d is recommended, followed by a gradual taper and adjustment based on the patient’s condition. The long-term use of corticosteroids is often necessary, but carries risk, including the development of invasive Aspergillus infection. Therefore, the antifungal agent itraconazole was tested in patients with ABPA as an adjunctive, steroid-sparing agent in two randomized placebo-controlled studies. Both studies spanned 16 weeks of treatment and showed reduced levels of markers of systemic immune activation (serum IgE level and eosinophil count). The study by Wark et al also showed reduced levels of markers of airway inflammation in induced sputum. Neither study showed significant changes in lung function, although the study by Wark et al showed that subjects receiving itraconazole experienced fewer exacerbations of disease requiring increased doses of corticosteroids. Therefore, while itraconazole appears to be promising as adjunctive treatment for patients with ABPA, long-term trials are needed to assess the clinical efficacy and safety in patients in different disease severity strata. There have been no randomized controlled trials to evaluate the use of antifungal therapies in patients with ABPA complicating CF.

MAC Infection

A clinicopathologic subtype of chronic pulmonary infection with this pathogen occurs predominantly in patients who are white women in their seventh or eighth decade of life with no underlying immune compromise (called the Lady Windemere syndrome). It is unknown why this group is at increased risk, but an undefined inherited or acquired immune deficiency state is presumed to exist. These patients typically present with chronic cough, which is often associated with fever and weight loss. The symptoms are insidious, and there is often an interval of several years between the onset of illness and the diagnosis. Radiographs and CT scans show bronchiectasis and nodular densities, especially in the middle lobe and lingula. The nodules are typically distributed around peripheral vessels and airways, frequently with a tree-in-bud configuration. In the past, these patients were thought to have preexisting bronchiectasis and “colonization” with MAC, but the presence of granulomas in the airways and the response to antmycobacterial therapy indicates that MAC infection is the primary disorder that leads to progressive airway damage and bronchiectasis. The diagnostic criteria for pulmonary infection with MAC have been reviewed elsewhere and are based on expert opinion. Sputum cultures are not sufficiently sensitive to establish the diagnosis in many patients. In patients with typical clinical and radiographic features of MAC infection, it is diagnosed in some patients with bronchoscopic biopsies, and other patients are treated empirically. There have been no large randomized trials of the treatment of MAC-associated lung disease, but based on in vitro data, there is expert consensus that treatment with a macrolide (eg, clarithromycin or azithromycin) with ethambutol and a rifamycin (eg, rifabutin or rifampin) constitute first-line therapy for patients with severe or progressive symptoms. However, these regimens are prolonged and often poorly tolerated, and patients often relapse.

Other Disorders

Up to 3% of patients with rheumatoid arthritis have symptomatic bronchiectasis, and up to 30% have HRCT-diagnosed disease. Some have recommended testing for serum rheumatoid factor in patients with bronchiectasis with no defined etiology. Inflammatory bowel disease, and especially chronic ulcerative colitis, is associated with recurrent respiratory tract infection and bronchiectasis. Mechanisms may include airway infiltration by immune effector cells, enhanced autoimmunity, and complications of immune-modulating therapy. A variety of congenital disorders listed in Table 1 predispose the patient to chronic respiratory infection and bronchiectasis; these have been discussed in detail elsewhere.

Treatment

In addition to the treatment of an identified underlying disease, pharmacotherapy may be used to enhance bronchodilation and improve mucociliary clearance, antibiotics may be used to prevent and treat recurrent infection, maneuvers may be designed to mobilize secretions (eg, chest physiotherapy), and mucolytic agents and occasionally surgery may be used to treat localized disease. The goal of treatment is generally to improve the symptoms of cough, sputum production, and dyspnea, and to prevent the progression of airway damage. Evaluating the effects of treatment in
patients with bronchiectasis is limited by study methodology. Randomized trials of these treatments generally lack proper control groups, and the surrogate end point of “effectiveness” is usually the volume of sputum production or the clearance of radiolabeled aerosol from the lung. More meaningful studies should focus on the number of exacerbations over time and on measures of health-related quality of life.

TREATMENT OF STABLE BRONCHIECTASIS

Bronchodilators

Bronchodilators, including short-acting and long-acting β-agonists, anticholinergics, leukotriene antagonists, and theophylline are useful in the management of COPD and/or asthma. Because many patients with bronchiectasis also have COPD, and many show signs of airflow obstruction and bronchial hyperreactivity, patients commonly receive bronchodilator therapy. However, there have been no randomized studies that have validated their usefulness in the management of cough, sputum production, dyspnea, or other end points in patients with bronchiectasis.45–48

Recommendation

3. In patients with bronchiectasis with airflow obstruction and/or bronchial hyperreactivity, therapy with bronchodilators may be of benefit. Level of evidence, expert opinion; benefit, small; grade of recommendation, E/C.

Mucolytics

The purpose of mucolytic drugs is to assist tracheobronchial clearance by altering the properties of sputum. Of this class of agents, only recombinant human DNase (rhDNase) was studied in randomized controlled trials in stable patients with bronchiectasis not caused by CF.49–51 DNA release by neutrophils in airways increases sputum viscosity, and rhDNase administered by aerosol digests DNA, thereby decreasing sputum viscosity and hopefully mucus plugging, infection, and inflammation. This agent was not associated with significant benefit in these two trials, and cannot be recommended for patients with idiopathic bronchiectasis. However, patients with CF treated with rhDNase enjoyed spirometric improvement for up to 2 years compared with placebo, with a nonsignificant reduction of risk of infectious exacerbations.52

Recommendation

4. In patients with bronchiectasis caused by CF, rhDNase should be used to improve spirometry. Level of evidence, low; benefit, small; grade of recommendation, C.

Antiinflammatory Agents

Inhaled corticosteroids are used commonly to treat bronchial inflammation and to prevent exacerbations of asthma and COPD. Two randomized short-time (4 and 6 weeks, respectively) placebo-controlled trials53,54 of inhaled corticosteroids in patients with idiopathic bronchiectasis showed nonsignificant trends toward improved FEV1, FVC, diffusing capacity of the lung for carbon monoxide, and residual volume, but no effect on total lung capacity, sputum production, cough, wheeze, or dyspnea. Oral corticosteroids may be more effective in treating bronchial inflammation, but there have been no randomized controlled trials addressing clinical end points in patients with bronchiectasis, other than when it is associated with known corticosteroid-responsive conditions like asthma, COPD, rheumatoid arthritis, and ulcerative colitis.55 However, the administration of oral corticosteroids at a prednisolone-equivalent dose of 1 mg/kg on alternate days appears to slow the progression of lung disease in CF patients, but is associated with significant side effects, especially in children.56–59 Therefore, systemic corticosteroid administration may be considered in patients with CF, but concerns about side effects preclude a general recommendation for its use.

Recommendation

5. In patients with CF, prolonged treatment with systemic corticosteroids should not be offered to most patients because of significant side effects. Level of evidence, low; benefit, conflicting; grade of recommendation, I.

Similarly, the administration of ibuprofen to patients with CF with the goal of reducing inflammation showed modest benefits in patients with mild disease, but its use also cannot be recommended at this time because of the potential of side effects with prolonged use.60

Recommendation

6. In patients with CF, prolonged courses of ibuprofen should not be used. Level of evidence, low; benefit, conflicting; grade of recommendation, I.
Antibiotics

The systemic and inhaled administrations of antibiotics were investigated as ways to prevent exacerbations of bronchiectasis by breaking the putative vicious cycle of bacterial colonization leading to inflammation and further airway injury. A literature review by the Cochrane group revealed six randomized controlled trials in patients with idiopathic bronchiectasis, but these studies had differences in design that limited the ability to generalize the outcomes. In general, they suggested that long-term therapy with antibiotics is effective for reducing sputum volume and purulence, but has limited impact on the frequency of exacerbations and the natural history of the condition, and may facilitate the emergence of resistant organisms.

Recommendation

7. In patients with idiopathic bronchiectasis, the prolonged systemic administration of antibiotics may produce small benefits in reducing sputum volume and purulence, but may also be associated with intolerable side effects. Level of evidence, low; benefit, conflicting; grade of recommendation, I

A randomized placebo-controlled trial of inhaled tobramycin delivered by jet nebulizer in patients with CF showed that it was well tolerated, and was associated with improved pulmonary function, decreased density of P. aeruginosa in sputum, and decreased risk of hospitalization.

Recommendation

8. In patients with CF, therapy with aerosolized antipseudomonal antibiotics are recommended. Level of evidence, low; benefit, intermediate; grade of recommendation, C

However, a similar trial in patients with bronchiectasis not caused by CF was less encouraging; although the treatment group had decreased density of P. aeruginosa and was judged more likely to have an “improved medical condition,” they were also more likely to report increased cough, dyspnea, wheezing, and noncardiac chest pain.

Recommendation

9. In patients with idiopathic bronchiectasis, aerosolized antibiotics should not be used. Level of evidence, low; benefit, negative; recommendation, D

Bronchopulmonary Hygiene

A variety of manual and mechanical interventions, including chest percussion, vibration, postural drainage, mechanically assisted cough, and airway oscillation, are used commonly to facilitate the expectoration of mucus. These maneuvers are also referred to as chest physiotherapy, and formerly as pulmonary toilet. Their primary role is to facilitate the clearance of lower airway secretions in conditions associated with the hypersecretion of mucus and the inability to expectorate effectively, as in patients with bronchiectasis. The role of bronchopulmonary hygiene in treating bronchiectasis and chronic bronchitis is discussed in a later section of this guideline. Although these techniques are considered to be mainstays in the treatment of bronchiectasis, they have modest effects on increasing sputum volume, and the long-term effectiveness of these interventions is unknown.

Recommendation

10. In patients with conditions associated with the hypersecretion of mucus and the inability to expectorate effectively, chest physiotherapy should be used and patients should be monitored for symptom improvement. Level of evidence, expert opinion; benefit, small/weak; grade of recommendation, E/C

Surgery

There have been no randomized controlled studies comparing surgery with more conservative management, and the literature consists of retrospective reviews of case series. With proper patient selection, surgery is associated with excellent outcomes and low perioperative mortality rates. Surgery to resect the bronchiectatic lung should be limited to patients with local disease who have not responded to medical therapy. Double-lung transplantation is commonly offered to patients with CF.

Recommendation

11. In selected patients with localized bronchiectasis that causes intolerable symptoms despite maximal medical therapy, surgery should be offered. Level of evidence, low; benefit, substantial; grade of recommendation, B

Treatment of Exacerbations

As bacterial exacerbations of bronchiectasis account for substantial morbidity, antibiotics are a mainstay of
treatment. Antibiotics are probably effective for exacerbations characterized by change in sputum production, increased dyspnea, increased cough, fever, increased wheezing, malaise, fatigue, lethargy, reduced exercise tolerance, reduced pulmonary function, new pulmonary opacities (although this may represent pneumonia), and changes in chest sounds.51

**Recommendation**

12. In patients with exacerbations of bronchiectasis, antibiotics should be used, with the selection of agents depending on the likely pathogens. Level of evidence, low; benefit, substantial; grade of recommendation, B

There have been no published randomized placebo-controlled trials on the antibiotic treatment of exacerbations of bronchiectasis since the last American College of Chest Physicians cough guideline,66 and no new recommendations can be offered. Treatment should initially include an agent that is active against *H influenzae* and *S aureus*; patients with recent antibiotic exposure or CF should be considered to be at high risk of infection with Pseudomonas and treated accordingly. Patients should receive at least 2 weeks of antimicrobial therapy; sputum culture and sensitivity testing should be performed in patients who do not have an adequate response, and they should guide the choice of a longer course of an oral agent, or even prolonged IV antibiotic therapy.2

**Conclusions**

Bronchiectasis is diagnosed in approximately 4% of patients with chronic cough. HRCT scanning of the chest is the best test to establish this diagnosis. Most cases of bronchiectasis in adults are idiopathic, but in the absence of an obvious cause, a diagnostic evaluation for an underlying disorder should be performed, as treatment may slow or halt the progression of airway disease. Bronchodilator agents, anticholinergic agents, and oral and inhaled steroids are often used in stable patients with bronchiectasis as well as during acute exacerbations despite the lack of objective evidence of benefit. Antibiotics and bronchopulmonary hygiene are mainstays in the treatment of exacerbations, but the prolonged use of systemic and inhaled antibiotics cannot be recommended to prevent these episodes except in patients with bronchiectasis caused by CF. Surgery is beneficial in a carefully selected subgroup of patients with focal disease and intractable symptoms.

**Summary of Recommendations**

1. In patients with suspected bronchiectasis without a characteristic chest radiograph finding, an HRCT scan should be ordered because it is the diagnostic procedure of choice to confirm the diagnosis. Level of evidence, low; benefit, substantial; grade of recommendation, B

2. In patients for whom there is no obvious cause, a diagnostic evaluation for an underlying disorder causing bronchiectasis should be performed, because the results may lead to treatment that may slow or halt the progression of disease. Level of evidence, low; benefit, substantial; grade of recommendation, B

3. In patients with bronchiectasis with airflow obstruction and/or bronchial hyperreactivity, therapy with bronchodilators may be of benefit. Level of evidence, expert opinion; benefit, small; grade of recommendation, E/C

4. In patients with bronchiectasis caused by CF, rhDNase should be used to improve spirometry. Level of evidence, low; benefit, small; grade of recommendation, C

5. In patients with CF, prolonged treatment with systemic corticosteroids should not be offered to most patients because of significant side effects. Level of evidence, low; benefit, conflicting; grade of recommendation, I

6. In patients with CF, prolonged courses of ibuprofen should not be used. Level of evidence, low; benefit, conflicting; grade of recommendation, I

7. In patients with idiopathic bronchiectasis, the prolonged systemic administration of antibiotics may produce small benefits in reducing sputum volume and purulence, but may also be associated with intolerable side effects. Level of evidence, low; benefit, conflicting; grade of recommendation, I

8. In patients with CF, therapy with aerosolized antipseudomonal antibiotics are recommended. Level of evidence, low; benefit, intermediate; grade of recommendation, C

9. In patients with idiopathic bronchiectasis, aerosolized antibiotics should not be used. Level of evidence, low; benefit, negative; recommendation, D

10. In patients with conditions associated with the hypersecretion of mucus and the inability to expectorate effectively, chest physiotherapy should be used and patients...
should be monitored for symptom improvement. Level of evidence, expert opinion; benefit, small/weak; grade of recommendation, E/C.

11. In selected patients with localized bronchiectasis that causes intolerable symptoms despite maximal medical therapy, surgery should be offered. Level of evidence, low; benefit, substantial; grade of recommendation, B.

12. In patients with exacerbations of bronchiectasis, antibiotics should be used, with the selection of agents depending on the likely pathogens. Level of evidence, low; benefit, substantial; grade of recommendation, B.

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