Diagnosis and management of patients with bronchiectasis


Abstract
Diagnosis of bronchiectasis should be considered in individuals presenting with respiratory symptoms similar to asthma and chronic obstructive pulmonary disease that have not responded to usual treatment. This article provides an overview of the prevalence, diagnosis and management of bronchiectasis to inform nursing care and improve patient outcomes.

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Keywords
Airflow obstruction, bronchiectasis, lung infection, respiratory system and disorders

Introduction
The British Thoracic Society (BTS) (2010) defines bronchiectasis as a progressive condition characterised by dilated thick-walled bronchi and excess sputum production. Although bronchiectasis has been documented for some time (Wrong 2003, Roguin 2006), evidence relating to investigation, diagnosis and management of the condition is mainly empirical and guidance is limited (BTS 2010).

Bronchiectasis is typically classified as an obstructive lung disease because of related airflow obstruction and tendency of the bronchi to collapse. Problems with mucus clearance result from damaged mucosa, leading to frequent bacterial colonisation and infection in the lungs. In health, a small amount of mucus is produced within the air passages, swept along by the cilia and often swallowed.
without causing any problems (Payne 2007). In bronchiectasis, mucus clearance by the cilia is ineffective and mucus pools in the dilated airways, providing an ideal environment for infection (Payne 2007). This promotes an inflammatory response, leading to further bacterial colonisation and infection (BTS 2010) (Figure 1).

The symptoms of bronchiectasis vary between individuals, but often include a cough that produces sputum. Breathlessness may be a feature in the later stages of disease. Intermittent exacerbations of the condition may be misdiagnosed as chronic obstructive pulmonary disease (COPD) (BTS 2010). The disease can affect all age groups, although it appears to be more prevalent in women (BTS 2010). Bronchiectasis can also be relatively widespread throughout the lung or confined to one or more lobes of the lung.

Prevalence
Mass screening of the UK population for tuberculosis in the 1950s revealed incidental findings of bronchiectasis in an estimated 100 per 100,000 people (BTS 2010). However, there are no up-to-date estimates of prevalence in the UK so the true occurrence of the condition is unknown (BTS 2010). Severe bronchiectasis is less common in developed countries as a result of improvements in socioeconomic conditions, widespread immunisation programmes and effective antibiotic therapy (Sapey and Stockley 2004). Also, modern diagnostic techniques, particularly high-resolution computed tomography (HRCT), may mean that bronchiectatic changes in the lungs are identified more frequently and treated in a timely manner.

Causes
Bronchiectasis has a wide range of possible causes, commonly divided into congenital causes (Table 1) and acquired causes, which are more common (Table 2). It is important to note that 40% of cases of bronchiectasis are idiopathic (BTS 2010).

Signs and symptoms
The most common presenting symptoms of bronchiectasis are breathlessness and recurrent infections, often with accompanying cough (BTS 2010). People with bronchiectasis typically produce copious amounts of green or yellow sputum that can vary in amount, but may be up to 240mL (8oz) per day (Payne et al 2007). Copious sputum production may occur without any coloration, and it is also possible to have ‘dry bronchiectasis’ characterised by little or no sputum production. People with bronchiectasis may have bad breath suggestive of active infection (BTS 2010). Other symptoms include chest pain and haemoptysis, and progression of bronchiectasis may lead to respiratory failure and cor pulmonale (BTS 2010).

Complete time out activities 1 and 2

Diagnosis
Diagnosis of bronchiectasis is based on a comprehensive review of the patient’s clinical history, especially childhood and respiratory illnesses, and HRCT (BTS 2012). Bronchiectasis can be observed as having characteristic patterns on HRCT. These patterns include “tree-in-bud” abnormalities and cysts with definable borders (Gosset et al 2009). Tree-in-bud abnormalities indicate the presence of infection that has spread throughout the bronchi. The peripheral airways are full of mucus, pus or fluid. The bronchi are usually thick and dilated, making them more visible. Tree-in-bud abnormalities are associated with tuberculosis and bronchopneumonia, which can predispose a person to bronchiectasis. Cysts with definable borders show thickening of the bronchi indicative of bronchiectasis. HRCT is
recommended as the gold standard for diagnosing bronchiectasis (BTS 2010).

It is possible to diagnose bronchiectasis in the absence of HRCT using only clinical history of frequent respiratory infections, sputum culture and inflammatory markers in the blood. However, this is not recommended as diagnosis is not definitive (BTS 2010). The BTS (2010) provides criteria for diagnosing bronchiectasis in children (Box 1) and adults (Box 2).

Bronchiectasis may occur in isolation or coexist with other more common respiratory disorders such as COPD (O’Brien et al 2000, Patel et al 2004) and asthma (Bisaccioni et al 2009). It is important that healthcare professionals are able to differentiate between these conditions to ensure that the patient’s treatment is targeted and effective.

**Management**

Initial management of bronchiectasis involves control of chronic day-to-day symptoms and acute exacerbations of the condition. If damage is extensive and the patient develops respiratory failure, palliative care may be required. The BTS (2010, 2012) recommends multidisciplinary involvement in secondary care, whether as an inpatient or an outpatient, led by a respiratory physician specialising in severe disease. Patients with mild disease can be appropriately cared for in primary care. Irrespective of the care setting, education should be provided for all healthcare professionals who may come into contact with patients who have bronchiectasis (BTS 2010).

**Primary prevention**

To prevent bronchiectasis, immunisation of children against measles and pertussis should be encouraged because severe lung infections can damage the lungs at the time of infection, and acute respiratory infections should be actively and appropriately treated (BTS 2010). Although smoking and passive smoking have not been definitively linked to bronchiectasis, they are lung and airway irritants that people should be encouraged to avoid (Sellick and Widdicombe 1971).

**Underlying conditions and coexisting disease**

Any coexisting problems such as asthma and COPD should be treated. This is important because coexisting diseases may delay the suspicion and diagnosis of bronchiectasis (BTS 2010).

**Physiotherapy and chest clearance**

Since people with bronchiectasis generally produce excess sputum, techniques to assist with chest clearance are an important component of care. The use of the Active Cycle of Breathing Technique (ACBT) can be useful in the clearance of sputum. This technique

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**TABLE 1**

<table>
<thead>
<tr>
<th>Congenital causes of bronchiectasis</th>
<th>Effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary ciliary dyskinesia, also known as Kartagener’s syndrome</td>
<td>Defective cilia action means mucus secretions are not cleared effectively, resulting in an inflammatory response and infection.</td>
</tr>
<tr>
<td>Cystic fibrosis</td>
<td>Ineffective secretion clearance and repeated infection.</td>
</tr>
<tr>
<td>Young’s syndrome</td>
<td>Viscous sputum causes an inflammatory response and increased risk of infection.</td>
</tr>
<tr>
<td>Alpha-1-antitrypsin deficiency</td>
<td>Lung damage.</td>
</tr>
<tr>
<td>Immunodeficiency</td>
<td>Poor immune response to infection because of immunosuppression.</td>
</tr>
<tr>
<td>Marfan’s syndrome</td>
<td>Relatively rare pulmonary complications of bronchial reactivity and pneumothorax.</td>
</tr>
<tr>
<td>Williams-Campbell syndrome</td>
<td>Defective cartilage in the bronchi makes them more prone to collapse.</td>
</tr>
</tbody>
</table>

(Adapted from British Thoracic Society 2010)

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**TABLE 2**

<table>
<thead>
<tr>
<th>Acquired causes of bronchiectasis</th>
<th>Effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acquired immunodeficiency syndrome</td>
<td>Poor immune response to infection because of immunosuppression.</td>
</tr>
<tr>
<td>Tuberculosis</td>
<td>Bronchial stenosis or fibrotic changes in the lung cause secondary traction in the bronchi, resulting in lung damage and increased risk of infection.</td>
</tr>
<tr>
<td>Hiatus hernia</td>
<td>Aspiration of gastric acid into the lungs causes tissue damage.</td>
</tr>
<tr>
<td>Rheumatoid arthritis</td>
<td>Link stronger in patients with rheumatoid arthritis who smoke, reflecting poor immune response to infection because of immunosuppression.</td>
</tr>
<tr>
<td>Environmental exposure</td>
<td>Infection, inhalation, aspiration, alcoholism, drug use, allergy and allergic bronchopulmonary aspergillosis predispose to increased risk of infection.</td>
</tr>
</tbody>
</table>

(Adapted from British Thoracic Society 2010)
encourages the person to undertake a relaxed, diaphragmatic style of breathing, allowing increased expansion of otherwise consolidated areas of the lungs, therefore aiding mucociliary clearance (Thompson et al 2002).

**BOX 1**

**Criteria for diagnosing bronchiectasis in children**

Bronchiectasis should be considered in a child presenting with:
- Persistent productive cough.
- Asthma that is unresponsive to treatment.
- Single positive sputum culture of one of the following: *Staphylococcus aureus, Haemophilus influenzae, Pseudomonas aeruginosa,* non-tuberculosis mycobacteria, and *Burkholderia cepacia*.
- An episode of severe pneumonia or recurrent pneumonia.
- Pertussis-like illness persisting over six months.
- Persistent and unexplained physical signs, such as failure to thrive and weight loss.
- Chest abnormalities on X-ray.
- Localised bronchial obstruction.
- Structural or functional disorders of the oesophagus and upper respiratory tract.
- Unexplained haemoptysis.
- Respiratory symptoms associated with cystic fibrosis, primary ciliary dyskinesia or immunodeficiency.

(Adapted from British Thoracic Society 2010)

**BOX 2**

**Criteria for diagnosing bronchiectasis in adults**

Bronchiectasis should be considered in an adult presenting with:
- Persistent productive cough, particularly if any one of the following factors is present:
  - Age of less than 50 years.
  - History of cough, sputum production and chest infections.
  - No history of smoking.
  - Expectorating large amounts of purulent sputum daily.
  - Haemoptysis.
  - Sputum colonisation with *Pseudomonas aeruginosa*.
- Unexplained haemoptysis or non-productive cough.
- Patients diagnosed with chronic obstructive pulmonary disease (COPD) in whom:
  - Management is not straightforward.
  - There is slow recovery from lower respiratory tract infection.
  - There are recurrent exacerbations of COPD.
- There is no history of smoking.

(Adapted from British Thoracic Society 2010)

Useful adjuncts to the ACBT technique are percussion, vibration, and shaking to dislodge sputum from the chest walls, enabling the patient to expectorate more easily (Thompson et al 2002). Another intervention to aid chest clearance is use of the Flutter device (Thompson et al 2002). The Flutter device can also be used to aid mucus clearance by vibrating and loosening mucus from the airway walls; intermittently increasing endobronchial pressure, which helps to maintain the patency of the airways during exhalation so that mucus does not become trapped as it moves up the airways; and accelerating expiratory airflow, which assists the upward movement of mucus through the airways so that it can be cleared easily (Konstan et al 1994, Thompson et al 2002). Healthcare professionals can also advise the patient on techniques such as postural drainage, although this is not frequently used as it is felt other methods are more effective and better tolerated by patients. Patients with bronchiectasis should be referred to a respiratory physiotherapist for advice about the best method of chest clearance (BTS 2012).

**Pharmacotherapy**

Antibiotics are the main treatment for recurrent lung infections caused by bronchiectasis (BTS 2010). A two-week course of oral antibiotics, depending on local antibiotic prescribing guidance, is used most commonly to treat these infections. It is recommended that a sputum sample is sent to the laboratory before each course of antibiotics is prescribed (BTS 2010). For recurrent or difficult to treat infections, it may be necessary for the person to receive a course of intravenous antibiotics depending on the bacterial infection. Intravenous antibiotics can be used to target specific bacteria and can be administered in higher doses (BTS 2010). Some areas will provide a hospital-at-home support service for administration of intravenous antibiotics or may teach the person to self-administer medicines (BTS 2010). Treating bronchial infections with antibiotics will help to prevent the destructive cycle of infection, damage to the bronchi and recurrent infection (BTS 2010).

Nebulised antibiotics may be used prophylactically and are recommended in patients with three or more exacerbations of bronchiectasis per year (BTS 2010). Patients who are prescribed nebulised antibiotics will need advice on looking after their nebuliser and correct administration of antibiotics. Bronchodilators may be
prescribed to help with breathlessness or may have already been prescribed for coexisting disease, such as asthma or COPD. Inhaled corticosteroid therapy used daily has been found to reduce sputum production and decrease airway constriction over a period of time, while helping to prevent progression of bronchiectasis (BTS 2010). However, this is not recommended by the BTS (2010) unless the patient has asthma. Patients who are prescribed inhalers should be encouraged to bring these with them to their healthcare appointment, and patients’ technique should be checked regularly.

**Complete time out activity**

Expectorants or mucolytics are often prescribed to help reduce the viscosity of sputum and assist with expectoration, yet there is a lack of evidence for their use (Crockett et al 2010). If prescribed, these medications are usually taken on a regular basis to help maintain thinning of sputum (Crockett et al 2010).

**Self-management**

Bronchiectasis can be self-managed, which involves daily chest clearance and use of antibiotics and bronchodilators. In a study by Lavery et al (2007), patients with bronchiectasis demonstrated the potential to self-manage medication and airway clearance. Perceived obstacles to self-management included lack of information and confidence (Lavery et al 2007). Healthcare professionals should encourage and promote self-management by providing disease-specific information and advising on appropriate chest clearance techniques.

**Pulmonary rehabilitation**

A formal regular programme of exercise and education known as pulmonary rehabilitation is recommended for people with bronchiectasis, if breathlessness affects activities of daily living (BTS 2012). People with bronchiectasis will benefit from the exercise and educational components of pulmonary rehabilitation. Exercise will help to control the symptoms of breathlessness and improve exercise tolerance, while education about nutrition and relaxation techniques, for example, will improve the patients’ overall outcomes.

**Surgery**

When other treatments have been tried and failed surgery may be necessary. Surgery may involve removal of a damaged lobe of the lung (BTS 2010). However, not all patients will be suitable candidates for surgery, and treatment of this nature is generally considered in those with well-defined lobular bronchiectasis (BTS 2010). Patients who are considered for surgery will need to receive adequate support and information.

**Acute exacerbations**

People with bronchiectasis are more likely to experience chest infections (O’Donnell et al 1998). O’Donnell et al (1998) identified several factors linked to exacerbations of bronchiectasis, including four or more of the following:

- Change in sputum production.
- Increased dyspnoea.
- Increased cough.
- Temperature >38°C.
- Increased wheeze.
- Lethargy, fatigue, general malaise or reduced activity levels.
- Reduced lung function.
- Radiological changes, such as bronchiectatic changes on chest X-ray or evidence of infection on HRCT.
- Increased chest sounds or changes in chest sounds.

Oral antibiotics can be used in the management of patients with mild exacerbations. Moderate-to-severe exacerbations require intravenous antibiotic therapy, and treatment is usually for 14 days (BTS 2010). If an exacerbation is identified, the BTS (2012) advises sending sputum samples to the laboratory and starting antibiotic therapy, rather than waiting for the results. Common bacteria found in sputum in people with bronchiectasis are listed in Box 3.

The guidance advises testing for *Aspergillus fumigatus* in all people diagnosed with bronchiectasis and for an annual sputum culture to be sent to the laboratory as part of a regular review (BTS 2012). Annual sputum cultures identifying chronic colonisation, for example with *Haemophilus influenzae*, *Staphylococcus aureus*, *Streptococcus pneumoniae*, *Moraxella catarrhalis*, *Pseudomonas aeruginosa*, *Haemophilus influenzae*, *Aspergillus fumigatus*.

(British Thoracic Society 2010)

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may need more aggressive treatment with intravenous antibiotics (BTS 2010). The BTS (2012) has developed quality statements for the management of clinically significant bronchiectasis in adults, which can be used by healthcare professionals to ensure that patients receive optimum care (Box 4).

Complete time out activity

Hydration and good oral hygiene
Encouraging a good level of hydration, around 1.2-3L of water per day, will help to reduce sputum viscosity (NHS Choices 2013). Adequate hydration may also help to reduce halitosis associated with bronchiectasis (Scully and Porter 2008).

Diet
Adequate nutrition is important for people with respiratory disease, as they tend to have reduced appetite and may be prone to weight loss. Patients with chronic infection tend to lose their appetite or their sense of taste because of mouth breathing or because they can taste the infection. Small frequent meals are often tolerated, and advice from a dietitian may be useful (BTS 2010).

Monitoring
Lung function with beta agonist reversibility (assessing the patient’s response to a bronchodilator medication) is recommended as a baseline to assess for an asthmatic response as an underlying disease process (BTS 2010). Pulse oximetry is useful in identifying early signs of declining oxygen levels and potential development of respiratory failure, and a gas transfer coefficient may be undertaken (BTS 2010). This gas transfer coefficient is the value of the transfer factor divided by the alveolar volume. This value is an expression of the gas transfer ability per unit volume of lung. If the patient has a disease that causes a decrease in lung surface area, or has had a lung removed, there is a decrease in transfer factor. Annual lung function measurements are recommended, including before and after administration of intravenous antibiotics, and before commencing prophylactic nebulised antibiotics (BTS 2010).

Complications
Haemoptysis is a relatively common complication of bronchiectasis occurring in around 50% of patients (Silverman et al 2003). Small blood vessels can fracture as a result of prolonged coughing, and sputum may be streaked with blood. Although not life threatening, this can be worrying for patients. A rarer, but more serious, complication of bronchiectasis is coughing up 100mL of blood in a 24-hour period (Silverman et al 2003). This occurs when one or more of the blood vessels that supply the lungs split. This is often a medical emergency and may necessitate surgery to stop the bleeding.

BOX 4
Quality statements for the management of clinically significant bronchiectasis in adults

- People with a clinical diagnosis of bronchiectasis have the diagnosis confirmed by a computed tomography scan of the chest (using 1mm slices).
- People with bronchiectasis are taught appropriate airway clearance techniques by a specialist respiratory physiotherapist, and advised of the frequency and duration with which these should be carried out.
- People with clinically stable bronchiectasis have sputum bacteriology culture recorded at least once each year.
- Sputum is sent for bacterial culture at the start of an exacerbation before starting antibiotics. Empirical antibiotic therapy should commence as soon as feasible. It is not necessary to wait for the sputum culture results.
- People with bronchiectasis to attend pulmonary rehabilitation if they have breathlessness affecting their activities of daily living.
- People with bronchiectasis receiving intravenous antibiotic therapy to have an objective evaluation of the efficacy of their treatment and the result recorded.
- Services for people with bronchiectasis to include provision of nebulised prophylactic antibiotics for suitable patients supervised by a respiratory specialist.
- People with bronchiectasis to be investigated for allergic bronchopulmonary aspergillosis, common variable immunodeficiency and cystic fibrosis (if indicated) as these are specific treatable causes.
- People with bronchiectasis to have an individualised written self-management plan.
- People with bronchiectasis who meet the criteria for continuing secondary care to be managed by a multidisciplinary team led by a respiratory physician.
- Services for people with bronchiectasis to include provision of home intravenous antibiotic therapy for exacerbations in selected patients.
(British Thoracic Society 2012)
As bronchiectatic changes in the lungs progress, respiratory failure can occur and damage to the lungs may result in heart failure (BTS 2010). Pulse oximetry, clinical examination and careful history taking should be part of the patient review to identify falling oxygen levels, worsening or new symptoms and swelling of the ankles, suggesting respiratory failure (BTS 2010).

**Prognosis**

Morbidity associated with bronchiectasis is significant, and although prognosis has improved, some patients will have reduced life expectancy (BTS 2010). The outlook for patients with bronchiectasis is variable and depends on the underlying cause, degree of damage and success of management. Therefore, patients should be diagnosed and treated in a timely manner to improve their outcomes and quality of life.

**Conclusion**

Although bronchiectasis may result in significant morbidity, prognosis has improved with earlier diagnosis leading to timely and more effective interventions. It is vital that healthcare professionals consider a diagnosis of bronchiectasis in adults and children presenting with respiratory symptoms and disorders that have not responded to treatment. This will enable appropriate treatment to be commenced, thus improving outcomes for patients.

References


