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Bronchiectasis in primary care

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Non-cystic fibrosis (CF) (bronchiectasis) is a common chronic lung condition, which occurs due to damage to the airways leading to persistent cough, sputum production and recurrent chest infections (Hill et al, 2018). This article focuses on the adult patient and describes the pathophysiology, aetiology, investigation, and management of bronchiectasis in the primary care setting. The aim is to raise awareness of this disease, which is increasing in prevalence and to empower community nurses with information to support patients through the bronchiectasis disease trajectory.

KEYWORDS:

■ Bronchiectasis ■ Pathophysiology ■ Aetiology ■ Investigation ■ Management

The term bronchiectasis (brongcos, airways; ectasia, dilatation) was first used in 1819 by the French Physician Laennec to describe abnormal, irreversibly dilated and thick-walled bronchi, which he noted in lungs he examined during postmortems (Grippi et al, 2015). It is a chronic lung condition associated with a markedly increased mortality, defined by the British Thoracic Society (BTS, 2019) as the abnormal, irreversible dilatation of the bronchi. The majority of patients present with a chronic cough and sputum production (Rosen, 2006). A study conducted in the UK over nine years reported that bronchiectasis is surprisingly common, increasing in incidence and prevalence particularly in older age groups (Quint et al, 2016).

The dilation and thickening of the bronchi seen in bronchiectasis is demonstrated in *Figure 1*, and

is due to chronic inflammation. A persistent airway inflammatory response leads to the development of oedema in the bronchial wall and increased mucus gland hypersecretion, which compromises mucociliary clearance (Morrissey, 2007). The release of neutrophils,

lymphocytes and macrophages within the bronchial lumen, prompt the release of inflammatory cytokines and proteases which are responsible for destruction of the airways (Morrissey, 2007). Neutrophils alter the function of the cilia, hundreds of tiny hair-like structures in the respiratory tract, which beat rhythmically to sweep foreign particles trapped in mucus from the lungs. The impairment of cilia in bronchiectasis is complicated further in smokers, as the cilia become paralysed by cigarette smoke (Tortora and Derrickson, 2017). This vicious cycle of infection and inflammation eventually leads to colonisation of the airway with microorganisms, debilitating infections and progressive airway damage.

Bronchiectasis has many potential causes resulting from abnormalities of anatomy, immunity or function. However, in approximately 40% of

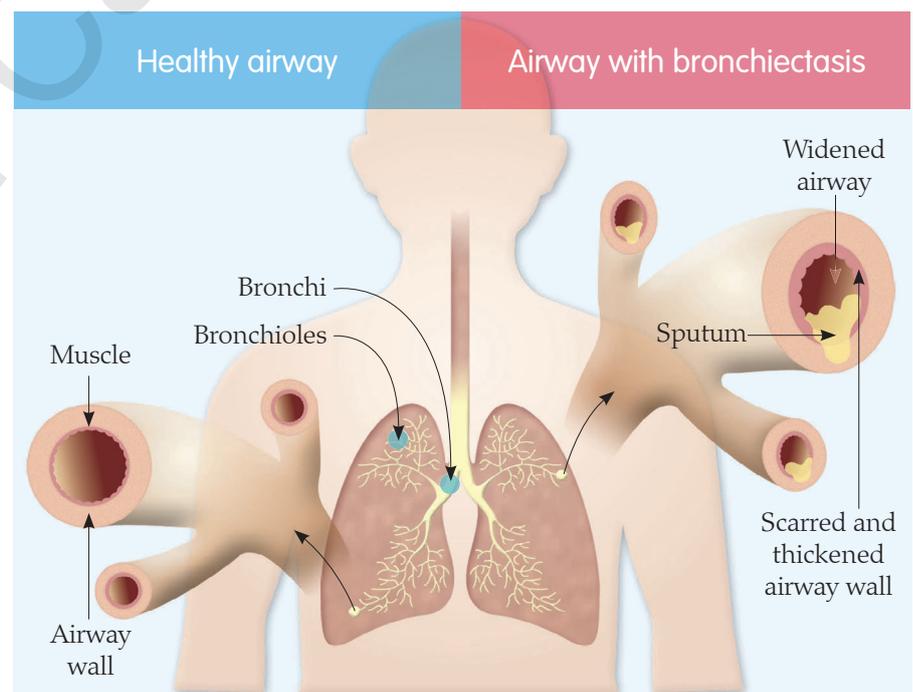


Figure 1.

Dilation and thickening of the bronchi seen in bronchiectasis (adapted from BLF, 2019).

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adults, it is idiopathic with no clear initiating event or underlying cause (BTS, 2019). Bronchiectasis may affect one lobe of the lung, or multiple lobes in one or both lungs (Bourke and Burns, 2015).

Patients can develop bronchiectasis as a complication of a bacterial or viral infection, for example pneumonia caused by pathogens such as *Streptococcus pneumoniae*, *Staphylococcus aureus* or a *Klebsiella*. A history of prior childhood infections due to viruses such as measles, influenza or pertussis may be significant. A non-tuberculosis mycobacteria (NTM) infection is a potential opportunistic cause. In developing countries, tuberculosis remains a common cause (BTS, 2019). An exaggerated response to fungal infections, such as *aspergillus fumigatus*, may be a precipitant (BTS, 2019).

People with immunodeficiencies, such as human immunodeficiency virus (HIV), lymphoma or myeloma, or those with immunoglobulin disorders, are predisposed to infection which may facilitate bacterial or fungal colonisation of the lower respiratory tract and the subsequent development of bronchiectasis (O'Donnell, 2018). There is evidence that recurrent aspiration pneumonia may also lead to bronchiectasis, and recurrent small volume aspirations, as seen with gastro-oesophageal reflux disease (GORD), may be an exacerbating cause in some patients (McDonnell et al, 2018). Bronchial obstruction can be a factor. For example, the inhalation of a foreign body, a bronchial carcinoma, or enlarged lymph node can lead to bronchiectasis distal from the obstruction (Bourke and Burns, 2015).

Bronchiectasis can also result from rare congenital defects such as primary ciliary dyskinesia (PCD), connective tissue disorders and alpha 1 antitrypsin deficiency, an enzyme which protects the lungs from inflammation caused by infection and inhaled irritants (Chalmers and Sethi, 2017).

There is an established coexistence that occurs with conditions such as inflammatory

bowel disease, rheumatoid arthritis and pulmonary fibrosis, and bronchiectasis is also commonly found in patient with a diagnosis of asthma and chronic obstructive pulmonary disease (COPD) (National Institute for Health and Care Excellence [NICE], 2018). Although this article relates to non-cystic fibrosis (CF) bronchiectasis, it is important to consider that the diagnosis of cystic fibrosis is being made with increasing frequency in adults. These patients may previously have received diagnoses of asthma, chronic bronchitis, or emphysema (Nick et al, 2005). The literature suggests that CF should be suspected in adults under the age of 50 with a *Pseudomonas* or *Staphylococcus* infection, male infertility, or other extra pulmonary features, i.e. pancreatic symptoms and malabsorption (Chalmers and Sethi, 2017).

It is important for clinicians to have an awareness of the possible causes and disease overlap in order to identify and treat patients with bronchiectasis effectively, with comprehensive history-taking being paramount. The majority of respiratory tract infections in primary care are self limiting, however patients presenting with persistent cough and daily production of a large volume of sputum with recurrent infections should alert clinicians to the possibility of bronchiectasis. Patients may also report breathlessness, chronic rhinosinusitis, haemoptysis, fatigue, pleuritic chest pain, and, in more severe cases, weight loss (NICE, 2018). On clinical examination, there may be coarse crackles, wheeze, large airway rhonchi (low pitched snore-like sounds) on auscultation of the chest, and finger clubbing may be present (Bourke and Burns, 2015).

Initial tests carried out in primary care might include a sputum culture to identify pathogens and enable targeted antibiotic treatment. The discovery of a pathogen such as *Pseudomonas* is associated with severe disease and poorer quality of life. The isolation of *Pseudomonas* in the sputum for the first time in a patient should prompt an attempt

at eradication therapy and this may involve admission to hospital for a 14-day course of intravenous (IV) antibiotics (BTS, 2019). A chest X-ray, although often normal in appearance, will help to exclude other pathology, such as infection, cancer or signs of chronic lung conditions, e.g. COPD. A high resolution computer tomography scan (HRCT) of the chest is the radiological investigation of choice in the diagnosis of bronchiectasis (BTS, 2019). Spirometry is routinely carried out in primary care and those with bronchiectasis may exhibit airflow obstruction, although this does not help diagnosis of the cause of obstruction, but is a marker of disease severity. A full blood count (FBC) will aid in determining the presence of a superimposed infection and, if the eosinophil count is high, an underlying allergic bronchopulmonary aspergillosis may be suspected (O'Donnell, 2019).

It is recommended by the BTS (2019) that referral to secondary care is considered where there is diagnostic uncertainty in order to investigate and confirm underlying causes, as well as to share management of patients with severe and complex disease. Respiratory specialist teams may have access to tests not available in primary care, such as HRCT scans. Other tests that are requested during work up are immunoglobulin, IgE specific to fungal precipitins, functional antibodies (response to vaccinations), and myeloma (a blood cancer arising from the plasma cells) (Chalmers and Sethi, 2017; BTS, 2019).

Once a diagnosis has been reached, the main treatment goals are to:

- ▶ Reduce exacerbations
- ▶ Preserve lung function
- ▶ Improve quality of life (Chalmers and Sethi, 2017).

Treatment of any underlying disease or comorbidity is paramount. A cornerstone of treatment is effective airway clearance to keep sputum moving (BTS, 2019). This can be hard work for patients, and physiotherapists are vitally important in teaching techniques such as the

active cycle of breathing. These are breathing exercises that help loosen and mobilise sputum in the airways. This involves breathing control and deep breathing performed in cycles until the chest is cleared. Sputum clearance techniques should be carried out daily, and can be increased during exacerbations. Patients are encouraged to maintain oral hydration. There are devices to help, such as the acapella/flutter using a form of vibrating oscillation to help loosen and mobilise sputum. Mucolytic drugs, such as carbosisteine (for maintenance therapy) and erdosteine (acute treatment for 10-day periods), help patients to breakdown sputum and easier to cough up (BTS, 2019). Nebulised and hypertonic saline humidify secretions, reducing tenacious secretions and promoting chest clearance. However, hypertonic saline is usually trialled with a healthcare professional, as in some patients this can provoke a bronchospasm, which can be reversed by administering bronchodilators such as inhaled or nebulised salbutamol (BTS, 2019).

The prompt and effective management of exacerbations, defined as the worsening of symptoms, such as an increase in cough, breathlessness, sputum volume and purulence occurring over several days, is essential. Patients also often report fatigue as a symptom during an exacerbation (BTS, 2019). A sputum sample should be taken and antibiotics chosen based upon current and previous pathogens, sensitivities, and local microbiology guidance. A standardised 14 days of treatment is recommended (BTS, 2019).

NICE (2018) recommends that all adults with bronchiectasis should be offered an annual review in primary care. This review should

Red Flag

Those patients with resistant bacteria and/or those with more severe signs and symptoms, such as sepsis red flags or haemoptysis, should be managed in hospital (Chalmers and Sethi, 2017).

include advice on smoking cessation, discussion around the number of exacerbations in the past year — three or more may trigger referral to secondary care. An assessment of breathlessness using, for example, the Medical Research Council (MRC) dyspnoea score, can identify patients disabled by breathlessness who may benefit from pulmonary rehabilitation and inspiratory muscle training (an exercise and educational programme for people with chronic lung disease). Advice on diet and exercise is recommended for all patients, exercise itself is a form of airway clearance. It is also important to ask about the impact of bronchiectasis on each individual, as it is known that these patients have increased anxiety and depression scores, experience fatigue, and have a poorer quality of life. Handheld self-management plans are recommended, as these can empower patients to recognise and manage exacerbations and obtain suitable antibiotics for effective treatment. Influenza and pneumococcal vaccinations should be offered to patients (NICE, 2018; BTS, 2019).

It is recommended that a sample of sputum for culture and sensitivity should be sent at annual review, or if sputum has become persistently purulent between exacerbations (BTS, 2019). This is important, as specialist follow-up is required for people with chronic colonisation of pathogens, such as *Pseudomonas aeruginosa*, opportunist *Mycobacteria*, or methicillin-resistant *Staphylococcus aureus* (MRSA). In secondary care, nebulised antibiotics, such as colistin and gentamicin, are initiated both for treatment and as a means of prevention of exacerbations in colonisation (persistent bacterial growth that cannot be eradicated). Some patients benefit from continuous antibiotic treatment with macrolide antibiotics, such as azithromycin, which is started on a three times per week dose, i.e. monday, wednesday and friday. These preventative strategies have been shown to increase the time between exacerbations and improve quality of life (BTS, 2019). Although careful consideration is taken beforehand in terms of sensitivities, comorbidities

and the side-effect profiles, and an electrocardiogram (ECG) should exclude prolonged QTc interval. Patients should be counselled to report any potential hearing loss before starting azithromycin (BTS, 2019).

Use of inhaled bronchodilators is most effective in bronchiectatic patients with co-existing asthma/COPD. However, BTS (2019) does suggest a trial in patients troubled by significant breathlessness. However, there is no role for inhaled or oral steroids in patients without co-existing disease (BTS, 2019).

A review of oxygen saturation with pulse oximetry will identify patients (those with SpO₂ <92%) who would benefit from long-term oxygen assessment. Pulse oximeters are now widely available in the community and are a cost-effective, non-invasive means of measuring SpO₂. They work by the principles of spectrophotometry, comparing how much red light and infrared light is absorbed by the blood, measuring amounts of oxygenated and deoxygenated haemoglobin and providing a percentage reading of how much of the haemoglobin is carrying oxygen (Hinkelbein et al, 2007). The BTS guidelines for oxygen assessment should support practitioners with background information (NICE, 2018; BTS, 2019).

A small number of patients may benefit from thoracic surgery, when optimal medical management has been achieved and there is focal (in one lobe of the lung) and resectable disease. The criteria for referral for lung transplantation for bronchiectasis patients is:

- ▶ Age 65 years or less
- ▶ Severe airway obstruction as measured by lung function testing with rapid progressive respiratory deterioration (BTS, 2019).

A predictive tool, the bronchiectasis severity index, has been developed, which suggests mortality and exacerbation risk based upon a clinical scoring system — available at: www.bronchiectasisseverity.com/

(Chalmers and Sethi, 2017). This can be used alongside the Gold Standards Framework (GSF) Prognostic Indicator Guidance (Royal College of General Practitioners [RCGP], 2011) 'surprise question' to identify patients who may be approaching end of life and therefore would benefit from a coordinated palliative care support framework, aiming to achieve the best quality of life for patients with bronchiectasis and their families.

CONCLUSION

Bronchiectasis is a complex condition, it may present in patients with existing respiratory diagnoses and, untreated, can significantly impair quality of life. Community nurses are well placed to recognise signs and symptoms and to support patients with exacerbations, prompting referrals as appropriate to secondary care and advocating for patients and their families through the disease trajectory, and effectively

coordinating management of this chronic lung disease. **JCN**

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KEY POINTS

- The dilation and thickening of the bronchi seen in bronchiectasis is due to chronic inflammation.
- Bronchiectasis has many potential causes resulting from abnormalities of anatomy, immunity or function. However, in approximately 40% of adults, it is idiopathic with no clear initiating event or underlying cause.
- Prompt and effective management of exacerbations, such as an increase in cough, breathlessness, sputum volume and purulence occurring over several days, is essential.
- It is important to ask about the impact of bronchiectasis on each individual, as it is known that these patients have increased anxiety and depression scores, experience fatigue, and have a poorer quality of life.



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