



Bronchiectasis:

Rates still increasing among Pacific peoples

What is bronchiectasis?

Bronchiectasis is a lung disease characterised by irreversible bronchial dilation and chronic inflammation, resulting in chronic wet cough. Recurrent cycles of infection and inflammation impair mucociliary clearance, leading to progressive remodelling and scarring of the bronchial walls. Symptoms of bronchiectasis are often present for many years before the diagnosis is made.¹

Bronchiectasis occurs in both children and adults. Although a relatively uncommon condition, bronchiectasis disproportionately affects Māori and Pacific peoples and people from lower socioeconomic communities. Clinicians need to consider this when diagnosing and treating chronic, wet cough in these patients. Empiric treatment of recurrent chest infections with antibiotics may be considered in some patients with risk factors to help prevent the development of bronchiectasis.

Although bronchiectasis is most often found in children with cystic fibrosis (in developed countries), this article focuses on non-cystic fibrosis bronchiectasis.

What causes bronchiectasis?

Bronchiectasis is commonly caused by recurrent or severe respiratory infections such as pneumonia (both bacterial

and viral), tuberculosis, adenovirus, measles and pertussis.² It may also be associated with; congenital syndromes and abnormalities, chronic obstructive pulmonary disease (COPD), gastro-oesophageal reflux, smoking and passive smoking, mucociliary dysfunction, immune deficiency, pulmonary fibrosis, post-obstruction (e.g. with an unrecognised foreign body), recurrent aspiration and systemic inflammatory diseases (e.g. rheumatoid arthritis, sarcoidosis). Overcrowding and socioeconomic deprivation are also important contributing factors.^{1,2}

The rate of bronchiectasis in New Zealand adults is not known, however, prevalence increases with age. In the United States, the estimated prevalence of bronchiectasis ranges from 4.2 per 100 000 in people aged 18 – 34 years to 272 per 100 000 in people aged over 75 years.³

Pacific children are at increased risk of developing bronchiectasis

In New Zealand, Pacific children aged under 15 years have an estimated incidence rate of bronchiectasis of 17.8 per 100 000.⁴ This compares to 4.8 and 1.5 per 100 000 for Māori and New Zealand European children respectively, of the same age.⁴ Capital & Coast DHB reported that from 2002 – 2006, the risk of being hospitalised due to bronchiectasis was almost 11 times higher for a Pacific child aged under 14 years and four times higher for a Māori child, than for a New Zealand European child.⁵

Socioeconomic deprivation is associated with an increased risk of developing bronchiectasis. The relative risk of a child living in a decile nine or ten community (most deprived) developing bronchiectasis is over 15 times higher than for a child living in a decile one area (least deprived).⁵ Issues such as housing and financial support may need to be considered in the management of the condition.

When to suspect bronchiectasis

The symptoms and signs of bronchiectasis are not diagnostic and may be mild in the early stages of the illness.

Consider bronchiectasis in a child with:^{1,2,6}

- A chronic wet/productive cough lasting longer than six weeks, especially between viral infections (N.B. if there is suspicion of an inhaled foreign body, the child should have a chest x-ray after two weeks)
- Wheeze that does not respond to treatment
- Partial resolution of severe pneumonia or recurrent pneumonia
- Persistent lung crackles
- Persistent x-ray changes
- Respiratory symptoms with structural or functional disorders of the oesophagus and upper respiratory tract

Other clinical features of bronchiectasis include dyspnoea, chest pain, clubbing, hyperinflation or chest wall deformity and failure to thrive.¹

Consider bronchiectasis in an adult with a chronic productive cough and:²

- A long history of respiratory symptoms
- Large volumes of purulent sputum on a daily basis
- Haemoptysis
- No history of smoking*

* N.B. Although smoking can be a factor in the development of bronchiectasis, it is not a major cause. COPD is a more common respiratory disease in adults, especially among people who smoke.

The prevalence of bronchiectasis increases with age, however, younger age at presentation should increase the clinical suspicion of bronchiectasis, because a diagnosis such as COPD is less likely in a young adult.

Bronchiectasis can co-exist with, or be misdiagnosed as, other chronic respiratory diseases that are more commonly seen in primary care, e.g. asthma, COPD, rhinosinusitis and tracheobronchial infection.⁷ Bronchiectasis should be considered in patients being treated for COPD when; management is complicated, there is slow recovery from lower respiratory tract infections, there are frequent exacerbations, there is no history of smoking or sputum is colonised with *Pseudomonas aeruginosa*.²

Investigating suspected bronchiectasis

All patients with suspected bronchiectasis should be referred to secondary care for confirmation of the diagnosis, exploration of underlying aetiology and ongoing management and support. High-resolution computed tomography is the current diagnostic gold standard.¹ Several investigations can be performed in primary care before a referral is made.

A sputum sample should be collected for microbiological analysis.² This information is important for selecting antibiotics to treat future exacerbations. Ideally, this should be done when the patient is stable and not currently taking antibiotics.

A chest x-ray should be arranged.² The presence of dilated and thickened airways (tramlines) is highly suggestive of bronchiectasis. A normal chest x-ray does not exclude bronchiectasis, however, it is useful for excluding other causes of persistent cough. A child with a wet cough lasting longer than two weeks should also have a chest x-ray to exclude the possibility of an inhaled foreign body.

A full blood count and CRP may show non-specific changes, e.g. infection, inflammation, anaemia or polycythaemia.

Immunoglobulins (IgA, IgG and IgM) should be requested to assess basic immune function.

A Mantoux or interferon gamma release assay (IGRA) should be arranged to exclude tuberculosis. IGRA testing, e.g. Quantiferon-TB Gold assay, should be first-line in patients who have previously received a Bacille Calmette-Guérin (BCG) vaccination, as 3 – 5% of people who receive a BCG vaccination as an infant and approximately one-third of those who receive it as an adult will return a false-positive Mantoux result.⁸



Management of bronchiectasis

The goals of management are the prevention and treatment of exacerbations and the prevention of lung function decline, through improved airway secretion clearance and antibiotics. Treatment plans are an important component of this strategy. Children with bronchiectasis require long-term follow-up in secondary care and should be under the care of a paediatrician as the monitoring of nutrition, growth and development are essential. Management of bronchiectasis is similar to cystic fibrosis care. Chest physiotherapy for airway secretion clearance and antibiotics are the mainstays of treatment. It is important that clear information is given so that patients and their families understand the rationale behind the treatment plan.

Smoking should be avoided and homes should be **smoke-free**. Patients can be provided with a “back-pocket” **prescription for antibiotics**, to allow for the prompt treatment of exacerbations.⁹ All patients with bronchiectasis should receive an **annual influenza vaccination**. **Pneumococcal vaccination** is also recommended, however, this is unfunded for adults (unless pre/post splenectomy).¹⁰ Pneumococcal vaccination is part of the New Zealand Childhood Immunisation Schedule.

Chest physiotherapy should be performed at home once or twice daily when the patient is well, and should be intensified if cough increases or exacerbation occurs. A physiotherapist should create an individualised airway secretion clearance programme for patients and their families. Family and patient comprehension of this programme is likely to influence compliance. Regular exercise should also be encouraged.¹ There is little evidence to support the routine use of mucolytics in patients with bronchiectasis.¹¹

Spirometry can be used at each review (if available) to record any deterioration in lung function.¹ N.B. Spirometry usually cannot be performed in children aged under five years.

Bronchodilators and corticosteroids should not be routinely prescribed for patients with bronchiectasis (particularly children), but may be considered in some cases for adults. Patients with bronchiectasis and co-existing asthma should continue to use beta-2 agonists as prescribed.¹ Inhaled corticosteroids provide only modest benefit to patients with bronchiectasis. Inhaled and oral corticosteroids should only be prescribed where there is an established diagnosis of co-existing asthma.¹

Managing exacerbations

Prompt use of broad-spectrum antibiotics and increased physiotherapy are essential in treating bronchiectasis

exacerbations.⁹ The choice of antibiotic should always be guided by sputum culture. If no sputum culture information is available, then a sample should be taken and empiric treatment initiated. *Haemophilus influenzae* should be assumed to be present and amoxicillin administered first-line.^{2, 12} Cefaclor, erythromycin or doxycycline in adults are alternatives if the patient is allergic to penicillin.⁹ High-dose amoxicillin, e.g. 1 g three times a day or even up to 3 g twice daily, may be considered for adults with severe bronchiectasis who are chronically colonised with *H. influenzae*.² Antibiotics are required for at least two weeks.

Follow-up is recommended to assess the effectiveness of any antibiotic treatment. If the patient has not improved, hospital admission for intravenous antibiotics may be required. When sputum culture indicates the presence of *Pseudomonas aeruginosa*, treatment with ciprofloxacin for a maximum of 14 days is indicated.¹

For children, eradication of *P. aeruginosa* is more aggressive and may require hospital admission, intravenous and nebulised antibiotics. A paediatrician should be consulted following all new *P. aeruginosa* positive cultures.


Hospital admission is indicated for all patients with bronchiectasis who display any of the following clinical features:²

- Breathlessness with an elevated respiratory rate and increased effort to breathe in children or respiratory rate > 25 breaths/minute in adults
- Circulatory or respiratory failure or cyanosis
- Temperature $\geq 38^{\circ}\text{C}$
- Unable to take oral treatment
- Intravenous treatment required

Socioeconomic circumstances and ability for patients, or parents/caregivers in the case of children, to cope at home may lower the threshold for referral.

Acute haemoptysis may occur during an exacerbation, more often in adults than children. This can be a life-threatening situation and patients who have expelled a volume of blood greater than 25 mL require urgent hospital assessment.²

Occasionally patients require surgical resection for symptoms resistant to standard treatments. This is unusual and would be determined in secondary care.

 For further information see: “The burden of bronchiectasis in Pacific peoples”, BPJ 32 (Nov, 2010).

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