

# The burden of bronchiectasis in Pacific peoples

## Key concepts

- Rates of bronchiectasis among Pacific children are disproportionately high
- Bronchiectasis should be suspected as a diagnosis in Pacific adults and children, living in areas with a high prevalence of bronchiectasis, who have a chronic, wet cough
- Referral to secondary care is required for a formal diagnosis based on a CT scan
- Acute exacerbations of bronchiectasis should be treated with a two week course of antibiotics and more intensive chest physiotherapy
- Consider developing an action plan so that treatment can be initiated early in an exacerbation

In most developed countries the incidence of bronchiectasis has declined over the years. However, in New Zealand, rates remain higher than in other developed countries and are disproportionately high in Pacific and Māori children. The rate among Pacific children is reported as 12 times higher than the rate for European children in New Zealand.<sup>1</sup>

Clinicians, particularly those in the upper North Island, should take into consideration the higher prevalence of bronchiectasis in Pacific peoples when diagnosing the cause of cough.

## Characteristics of bronchiectasis

Bronchiectasis is defined as irreversible widening of the bronchi in the lungs. It is characterised by inflammation, destruction of bronchial walls and chronic bacterial infection. Patients with bronchiectasis usually have a chronic, wet cough. Severe or recurrent respiratory infections such as pneumonia, tuberculosis or pertussis can result in bronchiectasis, especially if access to care or treatment is delayed. N.B: This article refers only to non-cystic fibrosis bronchiectasis.

## Starship statistics on bronchiectasis

Starship Children's Health, Auckland run a specialised bronchiectasis clinic and since 1998 has maintained a computerised database of children with bronchiectasis. Approximately 170 children with bronchiectasis are currently under active review at this clinic.<sup>4</sup>

Data on 91 children who have been followed for five or more years have been reported.<sup>4</sup> The median age at diagnosis was seven years.

Some key findings from this study were:

- 59% of the children were of Pacific ethnicity
- 36% lived in areas with a New Zealand deprivation score of 10
- 67% lived in areas with New Zealand deprivation scores of 8–10
- 28% of clinic appointments were not attended
- 14% of children were “lost to follow-up”
- 45% of children had worsening lung function despite clinic follow-up
- 44% of children had co-existing asthma
- 7% (six children) died during the follow-up period

Pacific or Māori ethnicity, male gender or low socioeconomic status was predictive of poor lung function.

Early recognition of children (including infants) with a “chronic, wet cough”, especially those with recurrent respiratory infections, is critical in reducing the incidence of bronchiectasis in New Zealand. If bronchiectasis is untreated it becomes a progressive condition that reduces life expectancy.<sup>2</sup> Even with treatment, bronchiectasis may have a major impact on day-to-day life for the child, with reduced exercise tolerance and time off school. Parents and caregivers must deal with the increased demands of an unwell child and the need for time away from work. Repeated primary care visits are necessary along with outpatient appointments and hospital admissions.

An increased awareness of bronchiectasis appears to have resulted in less delay before diagnosis, earlier referral and a younger age at diagnosis than previously reported.<sup>1,3</sup> This has markedly increased the number of children with bronchiectasis who are under active review in specialist clinics.<sup>2,4</sup> However, recent analysis of data from Starship Children's Health, Auckland, shows that even with follow up in a specialist clinic, almost half the children continue to have deteriorating lung function.<sup>4</sup>

## Prevention of bronchiectasis

Strategies to help prevent bronchiectasis include:<sup>5</sup>

- Ensuring up to date immunisations, including flu vaccination if appropriate
- Consideration of the possibility of a foreign body and prompt referral and treatment if this is likely
- Avoidance of aspiration if possible, e.g. in a patient with neurologic dysfunction, and effective treatment if aspiration occurs
- Early identification and treatment of chest infections
- Provision of a smoke-free environment for children and smoking cessation programmes for adults
- Providing education about bronchiectasis
- Improving housing conditions, e.g. reducing overcrowding and improving insulation and heating
- Improving nutrition

## When to suspect bronchiectasis

Bronchiectasis should be suspected in an adult who presents with a chronic productive cough, breathlessness and a history of a severe lower respiratory tract infection as a child or recurrent lower respiratory infections.<sup>6</sup> A classic presentation is an adult who has a history from a young age of repeated chest infections, daily sputum production and no history of smoking. Other clinical features that may be present in adults include haemoptysis (in up to 50% of cases) and non-pleuretic chest pain that occurs between exacerbations (approximately 30% of cases).<sup>6</sup>

Bronchiectasis should be suspected in a child who has a chronic wet cough (children aged less than five years usually swallow sputum). They may have shortness of breath, particularly with exercise. A child with wheeze, which may have been thought to be due to asthma, but has responded poorly to treatment, should raise suspicion of the possibility of bronchiectasis.<sup>6</sup> Haemoptysis is only rarely reported in children presenting with bronchiectasis.<sup>2</sup>

### Examination is not diagnostic

Clinical examination findings in patients with bronchiectasis are variable and not diagnostic.<sup>2, 6</sup>

During exacerbations:

- Fever may or may not be present
- Oxygen saturation may be borderline or reduced
- Respiratory rate and effort may be increased
- Crackles, usually coarse, may be localised or widespread. If present they are usually in the lower lung fields.
- Rhonchi may be present in about one-third of cases. Asthma may co-exist with bronchiectasis, however, rhonchi may be due to increased bronchial secretions rather than true bronchospasm.

Clinical signs of chronic lung disease, such as digital clubbing and chest deformities, e.g. Harrison's sulci,

pectus carinatum and hyperinflation, may be present. Studies have reported clubbing in up to 50% of children with bronchiectasis and chest wall abnormality in 40–60%.<sup>2, 4, 7</sup>

### Investigation and diagnosis of bronchiectasis

A good quality sputum sample should be obtained, although in younger children this may not be possible. In a secondary care setting, nasopharyngeal aspirate may be useful.<sup>2</sup>

Indicators of inflammation in the serum (increased white cell count and CRP) may be present even between exacerbations, but this does not alter management.<sup>2</sup>

A chest x-ray, although abnormal in the majority of cases, may not be diagnostic but should be arranged mainly to exclude other causes of chronic cough.<sup>6</sup>

Referral to secondary care is usually required for a formal diagnosis of bronchiectasis as CT scanning rather than bronchoscopy is now regarded as the test of choice.<sup>6, 7, 8</sup> Other investigations in secondary care may be performed to help determine the underlying cause of bronchiectasis and include:

- Gene testing to exclude cystic fibrosis
- Serum immunoglobulins (IgG, IgA and IgM) to screen for primary immunodeficiency (approximately 9% of children<sup>4</sup>)
- Bronchoscopy, e.g. in a child with suspected foreign body aspiration

In nearly 50% of cases of bronchiectasis, despite extensive investigation, no underlying aetiology is found.<sup>4, 8</sup>

### Treatment of bronchiectasis in primary care


In an acute exacerbation of bronchiectasis that can be managed in the community, a two week course of a suitable antibiotic should be prescribed.<sup>2, 6</sup> The choice of antibiotic should reflect the results of the sputum culture and sensitivities if possible. Appropriate initial antibiotic

choices include amoxicillin, erythromycin, cefaclor or co-trimoxazole.<sup>2, 6</sup> If there is wheeze or breathlessness, a short acting inhaled beta<sub>2</sub> agonist may be required. Inhaled corticosteroids are not indicated unless there is coexisting asthma.<sup>6,7</sup>

Regular chest physiotherapy in patients with known bronchiectasis can help with the drainage of secretions and help to prevent disease progression. Ideally physiotherapy should be increased during an exacerbation of bronchiectasis. Teaching a family member to assist with chest physiotherapy exercises can improve adherence to treatment and secretion drainage.

Consider developing an action plan<sup>2</sup> and providing a prescription for antibiotics so that treatment can be started early in an exacerbation.

A review visit is recommended to assess the response to the antibiotic and to check the sputum results if the antibiotic was started empirically. If there is deterioration or the patient has not improved with initial antibiotics, hospital admission may be required for intravenous antibiotics and more intensive physiotherapy.<sup>2, 6, 7</sup>

 For further information about bronchiectasis see: “Asthma and chronic cough in Māori children” BPJ 13 (May, 2008) and “Cough in children” BPJ 29 (July, 2010)

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