

## BRONCHIECTASIS

Bronchiectasis is a disease in which there is permanent enlargement of parts of the airways of the lung due to chronic inflammation and/or infection. Mucus builds up in the small airways, resulting in airway dilatation and a chronic daily cough. Infection and chronic inflammation results in progressive lung damage. The condition is irreversible. Bronchiectasis is characterised by recurrent chest infections.

There is little information on the prevalence or incidence of bronchiectasis in Australia. In 2017 almost all deaths (96%) due to bronchiectasis as an underlying cause occurred in people aged 60 years and over.

### Causes

Many people with bronchiectasis will have no clearly identifiable cause for their disease. Bronchiectasis may be localised to one lobe or segment in the lungs or generalised in both lungs. The most common cause is infections such as pneumonia, whooping cough (pertussis) or tuberculosis (TB).

In addition, common lung diseases such as asthma and chronic obstructive pulmonary disease (COPD) can be associated with bronchiectasis in the presence of an allergy to the fungus *Aspergillus* (allergic bronchopulmonary aspergillosis or ABPA). Up to 50% of people with severe COPD will have co-existent bronchiectasis. Other chronic inflammatory diseases such as rheumatoid arthritis and inflammatory bowel disease can cause bronchiectasis.

### Symptoms

Symptoms typically include a chronic cough with mucus production. Other symptoms include shortness of breath, tiredness, coughing up blood, and chest pain. Sputum is usually purulent and may be bloodstained. A small proportion of people with bronchiectasis will have clubbing of the fingers. A high resolution computed tomography (HRCT) chest scan may show mucus plugging and thickened airway walls.

### Exacerbations

Many people with bronchiectasis experience exacerbations (flare-ups of symptoms) once or twice a year. Bacterial infections, respiratory viruses and air pollution can lead to flare-ups. The symptoms of an exacerbation include the deterioration of three or more symptoms including:

- Cough
- Sputum volume and/or consistency
- Sputum colour or presence of blood
- Breathlessness
- Fatigue or malaise

Severe exacerbations usually require hospital admission in the presence of:

- Worsening respiratory distress
- Worsening hypoxaemia from baseline state
- Acute-onset confusion
- Signs of sepsis or septic shock

### Self-management

Clearing mucus or sputum from the chest every day is very important to decrease the risk of flare-ups. Respiratory physiotherapists can support residents and carers in developing a daily clearance routine. This may include breathing exercises, using positive expiratory pressure devices, inhaling saline via a nebuliser, positioning to open the airways and a prescribed exercise program.

Pulmonary rehabilitation can be beneficial to improve exercise tolerance. Active Cycle of Breathing (ACBT) is a method used where cycles of controlled breathing are interspersed with deep breathing and huffing. Positive Expiratory Pressure (PEP) where breathing cycles are added to by breathing out against resistance can also help shift the distal mucus that can be difficult to cough up. It is important to maintain weight, muscle strength and mass through good nutrition and exercise. Smoking cessation is strongly recommended for all people with bronchiectasis.

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### Treatment

Early treatment with airway clearance and antibiotics is recommended to minimise lung damage. Nebulised saline can help break up sticky mucus. The choice of antibiotic should be based on the most recent sputum culture. Common infective organisms include *Haemophilus influenzae*, *Streptococcus pneumoniae*, *Moraxella catarrhalis*, *Pseudomonas aeruginosa* and *Staphylococcus aureus*.

The Australian Therapeutic Guidelines detail antibiotic therapy according to colonisation with *Pseudomonas aeruginosa* and whether the exacerbation is severe or non-severe. The presence of *P. aeruginosa* in the airways is associated with more frequent exacerbations, increased risk of hospitalisation and greater mortality.

For exacerbations of bronchiectasis in adults without longstanding *P. aeruginosa* colonisation, amoxicillin or doxycycline are recommended. Amoxicillin / clavulanate is recommended if infection with *Haemophilus influenzae* or *Moraxella catarrhalis* is suspected. Treatment for 14 days is usually required. Intravenous therapy with ceftriaxone, cefotaxime or amoxicillin / clavulanate may be required for severe bacterial exacerbations. Intravenous therapy is usually continued for at least 5 days, and then switched to oral therapy for a total duration of 10-14 days.

For bacterial exacerbations of bronchiectasis in adults with longstanding *P. aeruginosa* colonisation, amoxicillin or doxycycline are recommended. Treatment for 14 days is usually required. If improvement is not seen with initial antibiotic therapy and *P. aeruginosa* is identified in sputum, ciprofloxacin is recommended if the culture shows susceptibility. For severe exacerbations, intravenous therapy is recommended.

### Long-term antibiotics

Long-term antibiotic therapy for bronchiectasis is not considered appropriate as it promotes the development of antibiotic resistance. However, in patients who continue to have recurrent or severe exacerbations despite optimal airway clearance regimens, long-term antibiotics may be prescribed under specialist supervision.

Long-term use of macrolide antibiotics (e.g. azithromycin, erythromycin) has been shown to reduce mucus secretion and exacerbations in patients who still have exacerbations despite optimal clearance and exercise. The reduction in exacerbation frequency may be due in part to an anti-inflammatory effect of macrolides. Macrolide antibiotics

may cause clinically significant drug interactions, prolong the QT interval and increase adverse cardiac events, and cause hearing loss.

Use of nebulised antibiotics (tobramycin, colistin, aztreonam, ciprofloxacin) have been studied; however, most studies report no improvement in acute exacerbation frequency and may result in significant adverse effects such as bronchospasm and wheeze.

### Inhaler therapy

For people with co-existing asthma or COPD long-acting beta agonists (LABA), antimuscarinics (LAMA) and inhaled corticosteroids (ICS) may be beneficial. There is little evidence to support the use of these inhaled therapies in people without co-existent asthma or COPD. Short-acting bronchodilators are recommended before inhaled antibiotics to improve deposition in the lungs.

### Vaccination

Influenza and pneumococcal pneumonia vaccinations are recommended for people with bronchiectasis.

### Mental health

As with any chronic disease, anxiety and depression is common in bronchiectasis, and early detection and appropriate treatment can enhance quality of life.

### Summary

Bronchiectasis is a disease characterised by permanent dilatation of bronchi and bronchioles, resulting in a chronic cough with sputum production. Wheezing and shortness of breath are common during exacerbations of flare-ups. The basic aim in bronchiectasis management is to keep the airways as free of secretions as possible and minimise the number of infective exacerbations. Effective airway clearance is the cornerstone of bronchiectasis management.

### Further information and support

- Lung Foundation Australia <https://lungfoundation.com.au>
- Bronchiectasis toolbox <https://bronchiectasis.com.au>

### References

Australian Institute of Health and Welfare  
*Therapeutic Guidelines, 2019.*  
*Med J Aust 2018;209(4):177-83.*

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