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Thoracic Society of Australia and New Zealand guidelines

Chronic suppurative lung disease and bronchiectasis in children and adults in Australia and New Zealand

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Guidelines on managing chronic suppurative lung disease (CSLD) and bronchiectasis (unrelated to cystic fibrosis [CF]) in Australian Indigenous children initiated in 2002[1] were extended to include Indigenous adults in 2008[2] and children and adults living in urban areas of Australia and New Zealand in 2010.[3] Here, we present an updated guideline relevant for all sections of the community. The recommendations in this guideline are targeted principally to primary and secondary care, and are not intended for individualised specialist care. As with all guidelines, they are not a substitute for sound clinical judgement, particularly when investigating and treating such a phenotypically heterogeneous condition as bronchiectasis.[4]

Key updates

An increasing trend in the health burden of CSLD and bronchiectasis is recognised in both Indigenous and non-Indigenous settings in Australia, New Zealand and worldwide.[3,5-7] Some affluent countries report childhood fatalities,[7] and there is a growing appreciation of the economic cost.[5] Misdiagnosis or coexistence of bronchiectasis with other chronic respiratory diseases is also recognised increasingly. When these comorbidities are present, the prognosis is worse; for example, mortality increases in those with both chronic obstructive pulmonary disease (COPD) and bronchiectasis (hazard ratio, 2.54; 95% CI, 1.16–5.56).[6] As many as 9% of newly referred children with chronic cough in Australia have bronchiectasis,[8] and 40% of newly referred adults with difficult asthma have bronchiectasis.[9] As effective management influences prognosis and quality of life,[10] a heightened vigilance by health professionals is needed to ensure an early diagnosis is made and treatment is optimised.

There is still a paucity of data and clinical trials on bronchiectasis, but encouraging trends of better evidence have emerged. These include studies related to defining CSLD in children,[10,11] airway clearance, rehabilitation, and use of nebulised and long-term maintenance antibiotics to prevent exacerbations. These studies (obtained through systematic searches[12]) formed the basis for our updated recommendations. When evidence was lacking, Australian and New Zealand experts (the writing group) developed the recommendations, which were further informed by the voting group using a modified Delphi process and the GRADE (grading of recommendations assessment, development and evaluation) system.[13]

Of the 32 recommendations, eight are new (3, 8, 10, 14, 25, 30–32), seven unaltered (7, 17–18, 22–24, 29) and the remaining 17 were amended. We refer readers to the Thoracic Society of Australia and New Zealand website for the full guidelines (http://www.thoracic.org.au/professional-information/position-papers-guidelines/bronchiectasis) including information about our guidelines development process, details of the systematic searches, evidence for the recommendations, implications of the strength of recommendations, suggested antibiotic regimens for management and updated references.[12]

Number	Recommendation	GRADE category	Evidence level
Definitions			
1	a. Bronchiectasis is a clinical syndrome in a child or adult with the symptoms and/or signs outlined below as well as characteristic radiographic features on chest high-resolution computed tomography (c-HRCT). Symptoms and signs include recurrent wet or productive cough episodes (⩾3 per year), each lasting for > 4 weeks, with or without other features (for example, exertional dyspnoea, symptoms of airway hyperresponsiveness, recurrent chest infections, growth failure, clubbing, hyperinflation or chest wall deformity). In children, triggers for referral to a specialist include one or more of: ■ persistent wet cough not responding to 4 weeks of antibiotics; ■ ⩾3 episodes of chronic (> 4 weeks) wet cough per year responding to antibiotics; ■ a chest radiograph abnormality persisting > 6 weeks after appropriate therapy. b. Chronic suppurative lung disease is a clinical syndrome in children with the symptoms and/or signs outlined above, but who lack a radiographic diagnosis of bronchiectasis.	Strong	–

Number	Recommendation	GRADE category	Evidence level
Investigations of a patient with CSLD or bronchiectasis			
2	a. Patients with symptoms and/or signs suggestive of bronchiectasis require a c-HRCT scan to confirm the diagnosis and to assess the severity and extent of bronchiectasis. b. In children, seek specialist advice before ordering a c-HRCT scan; child-specific criteria should be used. c. In both adults and children, a multidetector CT scan with HRCT reconstruction is the preferred technique to diagnose bronchiectasis.	Strong	Moderate
3	Consider a c-HRCT scan in adults with COPD and either ⩾3 exacerbations per year, very severe disease (forced expiratory volume in 1 second [FEV_1] < 30% predicted or requiring domiciliary oxygen) or whose sputum contains organisms atypical for COPD (ie, *Aspergillus* species, *Pseudomonas aeruginosa* or non-tuberculous mycobacteria).	Low	Low
4	Obtaining further history for specific underlying causes may determine subsequent investigation and management. This includes history of, or suggestive of: ▪ cystic fibrosis (family history, pancreatitis, chronic gastrointestinal symptoms, male infertility); ▪ underlying immune deficiency or ciliary dyskinesia (recurrent sinusitis, extrapulmonary infections, including discharging ears and severe dermatitis, and male infertility); ▪ recurrent aspiration (cough and/or choking with feeds or meals; after bariatric surgery; may be occult); ▪ an inhaled foreign body.	Strong	Moderate
5	Perform or refer for baseline investigations. Minimum investigations are: ▪ full blood count and major immunoglobulin classes G, A, M, E; ▪ sweat test in all children and selected adults (see full guidelines[12]); ▪ culture of airway secretions, including specialised cultures for mycobacteria, particularly non-tuberculous mycobacteria in sputum-producing patients; ▪ spirometry and lung volumes (patients aged > 6 years); and serological tests for *Aspergillus* species. In selected patients, other investigations should be considered (see full guidelines[12]).	Strong	Moderate
6	Obtain further history to determine markers of severity, impact of illness, comorbidities and modifiable risk factors. History should include frequency of exacerbations and hospitalisations, degree of effort limitation, exposure to tobacco smoke and other pollutants, childhood history, and housing.	Strong	Low
Management			
7	Aim to optimise general wellbeing, symptom control, lung function and quality of life, and to reduce exacerbation frequency and prevent excessive decline in lung function. This may require intensive medical therapy.	Strong	High
8	Develop treatment plans for exacerbations for each patient, linking them to primary health care and specialist or hospital facilities. When appropriate, this includes individualised and self-initiated management action plans.	Strong	Low
9	Base antibiotic selection on lower airway culture results (sputum, bronchoscopy washings [adults and older children] or bronchoalveolar lavage [young non-expectorating children]) when available, local antibiotic susceptibility patterns, clinical severity and patient tolerance, including allergy (Appendix).	Strong	Moderate
10	When *P. aeruginosa* is first detected, consider discussion with a specialist in this field regarding suitability for eradication treatment.	Weak	Low
11	In patients not requiring parenteral antibiotics for an acute exacerbation, oral antibiotics are prescribed for at least 10 days based on available airway microbiology results. Close follow-up to assess treatment response is necessary.	Strong	Low
12	Inadequate response should prompt repeat of lower airway cultures and assessment of whether parenteral antibiotic therapy and hospitalisation are needed.	Strong	Moderate
13	Patients in whom oral antibiotic therapy for an acute exacerbation fails should receive intensive airway clearance strategies and parenteral antibiotics based on the latest lower airway culture results. Close follow-up is required. a. In children, this requires supervised treatment for at least 10–14 days. b. In adults, intravenous antibiotics should be administered for at least 5 days and often need to be followed by oral antibiotics. Conversion from intravenous to oral antibiotics depends on appropriate oral alternatives and whether effective adjunct therapies, such as airway clearance strategies, can be maintained in an ambulatory care setting and with ongoing outpatient review.	Strong	Moderate
14	Long-term oral antibiotics should not be prescribed routinely. Macrolides (or other antibiotics) can be considered for a therapeutic trial over a limited period (eg, up to 12–24 months) in selected patients (eg, those with frequent exacerbations [⩾3 exacerbations and/or ⩾2 hospitalisations in the previous 12 months]). Before commencing macrolide antibiotics: ▪ seek respiratory/infectious diseases specialist advice; ▪ ensure non-tuberculous mycobacteria infection is excluded in patients capable of providing a sputum specimen; ▪ perform electrocardiography in adults for assessment of QT interval corrected for heart rate.	Strong	Moderate
15	Long-term nebulised antibiotics should not be prescribed routinely. Consider a therapeutic trial in children and adults with frequent exacerbations and/or *P. aeruginosa* infection.	Strong	Moderate

Number	Recommendation	GRADE category	Evidence level
16	Inhaled and oral corticosteroids should not be prescribed routinely unless there is an established diagnosis of coexisting asthma or COPD.	Strong	Low*/ moderate†
17	Inhaled bronchodilators should not be prescribed routinely but used only on an individual basis.	Strong	Low
18	Recombinant human deoxyribonuclease is contraindicated in CSLD and bronchiectasis.	Strong	High
19	Mucoactive agents, including hypertonic saline and mannitol, are currently not recommended for routine use. Consider a therapeutic trial in children and adults with frequent exacerbations.	Weak	Moderate
20	Airway clearance techniques are recommended and a respiratory physiotherapist's advice should be sought. Individualise airway clearance therapy.	Strong	Moderate
21	Adults with bronchiectasis and exercise limitation should receive pulmonary rehabilitation.	Strong	Moderate
22	Regular physical activity is recommended for children and adults with CSLD or bronchiectasis.	Strong	Low
23	Assess and optimise nutritional status.	Strong	Moderate
24	Promote elimination of smoking, including second-hand smoke exposure.	Strong	High
25	Promote avoidance of environmental airborne pollutants.	Strong	Low
26	Regularly monitor and manage complications and comorbidities. When present, manage following standard guidelines. Regular review consists of at least an annual review in adults and 6-monthly review in children. A multidisciplinary team is preferable, especially at the initial evaluation. Review includes: ▪ assessment of severity, which includes oximetry and spirometry; ▪ sputum culture (when available) for routine bacterial and annual mycobacterial culture; ▪ management of possible complications and comorbidities, particularly gastro-oesophageal reflux disease or aspiration, reactive airway disease or asthma, COPD, otorhinolaryngeal disorders, urinary incontinence, mental health and dental disease. Less commonly, patients require assessments for sleep disordered breathing and cardiac complications; ▪ checking adherence to therapies and knowledge of disease processes and treatments.	Strong	Moderate
27	Although surgery is not indicated normally, assessment by a multidisciplinary team expert in CSLD and bronchiectasis care may be required in some circumstances.	Strong	Moderate
Public health issues, prevention and appropriate health care delivery			
28	Vaccinate according to the National Immunisation Program Schedule. Ensure timely annual influenza vaccination and that pneumococcal vaccines are administered following national guidelines.	Strong	Moderate
29	Coordinated care by health care providers is necessary. If bronchiectasis is suspected, specialist evaluation is recommended to confirm the diagnosis, investigate the aetiology, assess severity and develop a management plan. Patients with moderate or severe disease are best managed using a multidisciplinary approach to chronic care with individualised case management. Clinical deterioration should prompt early referral to services with CSLD and bronchiectasis expertise.	Strong	Low
30	Specialist review should be undertaken for patients with moderate disability or progressive lung disease. This includes consideration for lung transplantation.	Strong	Low
31	Providing health care for Indigenous people in rural and remote regions requires flexible and adaptive arrangements. However, this should not alter the objective of delivering best-practice treatment to this population.	Strong	Low
32	Given the high prevalence of CSLD and bronchiectasis in Indigenous Australians, Maori and Pacific Islander children and adults, a high index of suspicion with early diagnostic investigation should be established, as well as best-practice treatment. Interpreters and local health workers should be available for educating patients about the disease and its management.	Strong	Moderate

* For oral corticosteroids. † For inhaled corticosteroids. ◆

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