






Thoracic Society of Australia and New Zealand (TSANZ) position statement on chronic suppurative lung disease and bronchiectasis in children, adolescents and adults in Australia and New Zealand

Anne B. Chang^{1,2,3}  | Scott C. Bell^{4,5,6}  | Catherine A. Byrnes^{7,8} |
 Paul Dawkins^{9,10} | Anne E. Holland^{11,12,13}  | Emma Kennedy^{14,15,16} |
 Paul T. King¹⁷  | Pamela Laird^{18,19,20}  | Sarah Mooney^{9,21} | Lucy Morgan²² |
 Marianne Parsons²³ | Betty Poot^{24,25} | Maree Toombs²⁶ | Paul J. Torzillo^{27,28} |
 Keith Grimwood^{29,30}

Correspondence

Anne B. Chang
 Email: anne.chang@cdu.edu.au

Funding information

National Health and Medical Research Council,
 Grant/Award Numbers: 1170958, 1154302;
 Children's Hospital Foundation

Handling Editor: Paul Reynolds

Abstract

This position statement, updated from the 2015 guidelines for managing Australian and New Zealand children/adolescents and adults with chronic suppurative lung disease (CSLD) and bronchiectasis, resulted from systematic literature searches by a multi-disciplinary team that included consumers. The main statements are:

- Diagnose CSLD and bronchiectasis early; this requires awareness of bronchiectasis symptoms and its co-existence with other respiratory diseases (e.g., asthma, chronic obstructive pulmonary disease).
- Confirm bronchiectasis with a chest computed-tomography scan, using age-appropriate protocols and criteria in children.
- Undertake a baseline panel of investigations.
- Assess baseline severity, and health impact, and develop individualized management plans that include a multi-disciplinary approach and coordinated care between healthcare providers.
- Employ intensive treatment to improve symptom control, reduce exacerbation frequency, preserve lung function, optimize quality-of-life and enhance survival. In children, treatment also aims to optimize lung growth and, when possible, reverse bronchiectasis.
- Individualize airway clearance techniques (ACTs) taught by respiratory physiotherapists, encourage regular exercise, optimize nutrition, avoid air pollutants and administer vaccines following national schedules.
- Treat exacerbations with 14-day antibiotic courses based upon lower airway culture results, local antibiotic susceptibility patterns, clinical severity and patient tolerance. Patients with severe exacerbations and/or not responding to outpatient therapy are hospitalized for further treatments, including intravenous antibiotics and intensive ACTs.
- Eradicate *Pseudomonas aeruginosa* when newly detected in lower airway cultures.

This document has been endorsed by the Thoracic Society of Australia and New Zealand on 15 February 2023.

For affiliations refer to page 347

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- Individualize therapy for long-term antibiotics, inhaled corticosteroids, bronchodilators and mucoactive agents.
- Ensure ongoing care with 6-monthly monitoring for complications and co-morbidities.
- Undertake optimal care of under-served peoples, and despite its challenges, delivering best-practice treatment remains the overriding aim.

KEYWORDS

adolescents, adults, bronchiectasis, children, evidence base practice, systematic review

INTRODUCTION

Chronic suppurative lung disease (CSLD) and bronchiectasis are characterized by chronic or recurrent wet/productive cough, and lower airway infections. Those with bronchiectasis are distinguished by radiographic evidence of bronchial dilatation secondary to structural airway injury. Other clinical and radiographic features may also exist as bronchiectasis is a heterogenous disease with many underlying aetiologies and co-morbidities.^{1–3}

Since our latest guideline,⁴ Australia has a diagnostic-related-group which recognizes that bronchiectasis is important. Global data have demonstrated increased prevalence, hospitalization and economic burden.^{5,6} A United Kingdom population-based study described an increase in point-prevalence of bronchiectasis between 2004 and 2013 (women = 350.5–566.1/100,000; men = 301.2–485.5/100,000).⁷ There are no updated Australian data (prevalence in Central Australian First Nations: children aged ≤15-years = 1470/100,000; adults = 103/100,000 derived from hospitalization data⁴), but in New Zealand (NZ) the incidence in children aged ≤14-years increased from 3.7/100,000 in 2001/2002 to 13.2/100,000 in 2017 and the population-prevalence was 180/100,000.⁸ Recently, a Brisbane-based paediatric study reported direct health service costs for bronchiectasis exacerbations in 2016 of 30,000 Australian dollars per hospitalization.⁹

Although the burden of CSLD/bronchiectasis is now recognized, diagnostic delays remain.^{10,11} Early diagnosis requires greater clinician awareness, whilst prompt investigations confirm the diagnosis and identify underlying aetiologies, treatable traits and co-morbidities ensuring prompt and appropriate care.

Currently in Australia and NZ, there are large equity gaps within,^{12,13} and between,^{14,15} settings. Australian First Nations with bronchiectasis die >20-years earlier than other Australians with bronchiectasis.¹⁵ In addition, children/adolescents with bronchiectasis unrelated to cystic fibrosis (CF) have substantially poorer lung function, receive lesser care (as judged by numbers of clinic visits, physiotherapist reviews and lung function tests) than age-matched children/adolescents with CF.¹² Optimal care for people with bronchiectasis is crucial as poor treatment risks premature and accelerated pulmonary decline.^{1,2,16} Moreover, in children/adolescents, it is acknowledged that mild cylindrical bronchiectasis is potentially reversible, indicating an achievable cure for some patients.^{1,3,16}

This Thoracic Society of Australia and NZ (TSANZ) position statement builds upon previous guidelines^{4,17–20} for

managing CSLD/bronchiectasis in children/adolescents and adults in Australia and NZ, including urban and rural-remote First Nations people. It seeks to increase awareness of CSLD/bronchiectasis amongst clinicians and health service managers, and to encourage consumers to advocate for best-practice care. However, it does not substitute for sound clinical judgement when diagnosing and managing these heterogenous disorders.^{1,2,16}

The objectives are to:

1. Increase awareness of the disease burden associated with CSLD/bronchiectasis;
2. Encourage earlier and improved diagnosis and best-practice management of CSLD/bronchiectasis, including important co-morbidities;
3. Present an updated multi-disciplinary position statement on managing children/adolescents and adults with CSLD/bronchiectasis living in Australia and NZ.

The accompanying technical report for this position statement is presented in Appendix S1 (in the Supporting Information). We reviewed the latest guideline,⁴ and undertook an updated search (Appendix S2 in the Supporting Information); of the 33 statements, one is new, 28 were modified and four unchanged.

AWARENESS AND DEFINITIONS

Statement-1

- 1a. Bronchiectasis is a clinical syndrome in children/adolescents or adults with the symptoms and/or signs in Box 1 and presence of characteristic radiographic features on chest computed-tomography (CT) scans.
- 1b. CSLD is a clinical syndrome in children/adolescents with the symptoms and/or signs outlined in Box 1, but where radiographic signs of bronchiectasis are lacking.

Statement-2

- Patients with symptoms and/or signs suggestive of bronchiectasis require a chest CT-scan to confirm the diagnosis and to assess severity and extent of bronchiectasis.

BOX 1 Definition (related to Statement-1)

Presentation in children/adolescents is with recurrent wet/productive cough for >4-weeks, with or without other features (faltering growth, exertional dyspnoea, recurrent chest infections, digital clubbing, hyperinflation and chest wall deformity). While in adults, characteristic clinical features are chronic cough, sputum production and recurrent exacerbations.

In children/adolescents, triggers for referral to a specialist include: >3 episodes of chronic (>4-weeks) wet/productive cough per year, chronic wet/productive cough not responding to 4-weeks of antibiotics, recurrent (>2) episodes of pneumonia, severe asthma (with chronic wet/productive cough) and persistent (>6-weeks) lung parenchymal abnormalities on chest radiographs.

- In children/adolescents, seek specialist advice before ordering a chest CT-scan and paediatric-specific diagnostic radiographic criteria should be used.
- In all age groups, multi-detector CT-scans with high-resolution CT reconstruction is the preferred technique to diagnose bronchiectasis.

Statement-3

Consider a chest CT-scan in adult patients with other airway diseases (e.g., asthma and chronic obstructive pulmonary disease [COPD]), but who also have the clinical phenotype of bronchiectasis (chronic cough, sputum production and recurrent exacerbations), especially in those with more severe airway disease.

INVESTIGATING A PATIENT WITH CSLD/ BRONCHIECTASIS**Statement-4**

Obtaining further history/clinical examination for specific underlying causes may determine subsequent investigation and management.

In children/adolescents, these should include:

- CF (faltering growth, chronic diarrhoea, heat prostration with hyponatraemia, positive family history, *Pseudomonas aeruginosa* in sputum);
- Underlying immune deficiency (recurrent severe, protracted infections, infections with unusual pathogens, chronic diarrhoea, severe dermatitis, faltering growth);
- Primary ciliary dyskinesia (neonatal respiratory distress in a term infant, recurrent otitis and sinopulmonary infections from the first-year of life, heterotaxy);

- Recurrent aspiration (cough and/or choking with feeds, 'wet' sounding voice, cyanotic episodes); and
- Inhaled foreign body (history of sudden choking, cough, wheeze/stridor, cyanosis, focal decreased breath sounds or wheezing, refractory pneumonia or persistent atelectasis).

In adults, these should include:

- Parameters suggestive of CF (early-onset symptoms, family history, pancreatitis, chronic gastrointestinal symptoms, co-existent liver disease, male infertility);
- Features of primary ciliary dyskinesia (recurrent upper and lower respiratory tract infections from early life, heterotaxy, infertility);
- Evidence of underlying immune deficiency (including recurrent and/or severe sinopulmonary infections, unusual pathogens, severe dermatitis, chronic diarrhoea);
- Features of connective tissue diseases (e.g., rheumatoid arthritis);
- Features suggestive of COPD (also consider alpha-1-antitrypsin deficiency in this group) and asthma;
- Features of inflammatory bowel disease;
- Recurrent aspiration episodes (these can occur post-bariatric surgery or with a hiatus hernia, and cause symptoms of cough and/or choking with meals, or may even be occult);
- Features suggestive of an inhaled foreign body.

Statement-5

Perform or refer for baseline investigations (Box 2).

Statement-6

Obtain further history/clinical examination to determine markers of severity, impact of illness, co-morbidities and modifiable risk factors. History should include: frequency and impact of exacerbations and hospitalisations, degree of effort limitation, exposure to tobacco smoke, vaping/e-cigarettes and other aerotoxicants, immunization and early childhood medical and social history, including housing. Clinicians should be aware that some ethnicities (e.g., Australian First Nations, Māori and Pacific Islanders) have more severe disease and experience worse outcomes than other ethnic groups.

In children/adolescents, growth and developmental assessment should be undertaken.

In adults, for severity and impact assessment, consider using clinical severity (e.g., Bronchiectasis severity index, FACED) and quality-of-life scores to guide management at baseline and follow-up. Assess for modifiable risk factors and treatable co-morbidities that are more common in those with bronchiectasis (COPD, asthma, atopy, rhinosinusitis, cardiovascular disease, anxiety and depression, stress urinary incontinence and low body-mass index). In severe cases and those with significant co-morbidities (e.g., rheumatoid

BOX 2 Panel of tests (related to Statement-5)

The minimum baseline investigations in children/adolescents should include:

- Full blood count;
- Major immunoglobulin classes G, A, M, E;
- Sweat test for chloride testing;
- Culturing airway secretions, including specialized cultures for mycobacterial species in sputum-producing patients (consider referring for induced sputum or bronchoscopy in those unable to expectorate sputum);
- Spirometry when age appropriate, usually in those aged >6-years.

In addition, consider the following after discussion with a specialist

- Exhaled fractional nasal nitric oxide, nasal ciliary brushings and/or genetic testing for primary ciliary dyskinesia;
- Cystic fibrosis transmembrane conductance regulator gene variant/mutation testing when there is a high index of suspicion for cystic fibrosis, but where sweat testing (for chloride) is equivocal or negative;
- Bronchoscopy for foreign body, airway abnormality and specimens for culture of respiratory pathogens, including mycobacteria;
- Assessment for aspiration (primary or secondary), for example, modified barium swallow for primary aspiration;
- Additional immunological tests (neutrophil function tests and lymphocyte subsets, IgG subclasses, antibody responses to vaccine antigens);
- Human immunodeficiency virus testing;
- Echocardiogram (when concerned about pulmonary hypertension).

The minimum baseline investigations in adults should include:

- Full blood count;
- Major immunoglobulin classes (G, A, M, E);
- Culturing airway secretions in patients who can expectorate sputum—including for fungi and mycobacterial species—consider referring for induced sputum or bronchoscopy in those unable to expectorate sputum;
- Spirometry;
- *Aspergillus* serology (IgG precipitins, specific IgE to aspergillus or skin prick tests to *Aspergillus*).

In addition, consider the following:

- Alpha-1-antitrypsin levels if there is evidence of chronic obstructive pulmonary disease/emphysema;
- Autoimmune serology if clinically indicated (e.g., anti-CCP, RF, ANA, ANCA, ENAs);
- Vitamin D levels if at risk of deficiency;
- Exhaled fractional nasal nitric oxide, nasal ciliary brushings and/or genetic testing for primary ciliary dyskinesia;
- Sweat test (for chloride) and/or extended cystic fibrosis transmembrane conductance regulator gene variant/mutation testing (in those with early onset of symptoms, family history, pancreatitis, chronic gastrointestinal symptoms, co-existent liver disease, male infertility);
- Bronchoscopy for investigation of suspected foreign body, airway abnormality or in order to obtain specimens for culture of respiratory pathogens (including mycobacteria) in those who are unable to expectorate sputum;
- Assessment for aspiration (oesophageal pH monitoring, multi-channel intraluminal impedance, endoscopic or radiographic evaluation);
- Additional immunological tests (IgG subclasses, functional antibody responses to vaccine antigens (including to 23-valent pneumococcal polysaccharide vaccine antigens), neutrophil function tests, lymphocyte subsets);
- Human immunodeficiency virus or human T-lymphotrophic virus type 1 testing if clinically suspected;
- Echocardiogram, especially in adults (if there is suspicion of secondary pulmonary hypertension);
- Detailed lung function testing, including body plethysmography, gas transfers, measures of small airway function and walk test (e.g., 6-minute walk test, if there are symptoms of dyspnoea).

arthritis, other autoimmune conditions, inflammatory bowel disease or those taking corticosteroids, immunosuppressive agents or biologic drugs), referral to specialist bronchiectasis services/clinicians is recommended.

MANAGEMENT AND MONITORING

Box 3 outlines the aims of optimal treatment of people with bronchiectasis.

Statement-7

Aim to optimize general well-being, symptom control, lung function, quality-of-life and reduce exacerbation frequency and prevent excessive decline in lung function. This may require intensive medical therapy.

Statement-8

Develop treatment plans for exacerbations for each patient, linking them to primary healthcare and specialist or hospital facilities. When appropriate, this includes individualized and self-initiated management action plans.

Statement-9

Base antibiotic selection (Table 1) on lower airway culture results (sputum, bronchoscopy washings [adults and older children/adolescents] or bronchoalveolar lavage [young non-expectorating children]) when these are available, local antibiotic susceptibility patterns, clinical severity and patient tolerance, including allergy.

Statement-10

When *P. aeruginosa* is newly detected, eradication therapy should be offered (Figure 1).

Statement-11

In patients not requiring parenteral antibiotics for an acute exacerbation, oral antibiotics are prescribed for at least 14-days based upon available airway microbiology results. Close follow-up to assess treatment response is necessary.

Statement-12

Inadequate response should prompt repeating lower airway cultures, including for potential pathogens not detected on routine culture (e.g., non-tuberculous mycobacteria; NTM)

BOX 3 Main aims of optimal management of children/adolescents and adults with CSLD and bronchiectasis

Main aims are to:¹

1. Preserve lung function and halt disease progression.
2. Optimize well-being and quality-of-life for the patient and family.
3. Minimize the frequency and severity of respiratory exacerbations.
4. Prevent complications.

Additional aims in children/adolescents are to:

5. Optimize lung growth.
6. If possible, reverse structural lung injury.

and assessment of whether a change in antibiotic therapy, including parenteral antibiotics, and hospitalization are required.

Statement-13

Patients failing oral antibiotic therapy for an acute exacerbation should receive intensive airway clearance strategies and parenteral antibiotics based upon the latest lower airway culture results. Close follow-up is required.

- a. In children/adolescents, this requires supervised treatment for at least 14-days;
- b. In adults, intravenous antibiotics should be for at least 5-days and only transitioned to an oral regimen following clear evidence of clinical improvement. A total of at least 14-days antibiotic therapy should be prescribed. The transition from intravenous to oral antibiotics should depend upon appropriate oral alternatives (based upon sputum culture results) and, if effective, adjunct therapies, such as airway clearance strategies, can be maintained in an ambulatory care setting and where there is ongoing outpatient review.

Statement-14

Long-term oral macrolides should be considered for patients with the frequent exacerbator phenotype (≥ 3 exacerbations requiring antibiotics in the preceding 12-months) and without contraindications to these agents. Such courses of macrolides should be for at least 6-months with regular

TABLE 1 Antibiotic selection guide (related to Statement-9).

| | Mild-to-moderate exacerbations (Oral therapy) | Moderate-to-severe exacerbations (Intravenous therapy) |
|--------------------------------------|---|---|
| Initial empiric therapy ^a | <i>Children:</i> amoxicillin-clavulanate <i>Adults/adolescents:</i> amoxicillin-clavulanate or doxycycline ^b <i>All ages:</i> ciprofloxacin if <i>P. aeruginosa</i> in recent cultures | <i>All ages:</i> amoxicillin-clavulanate, cefotaxime or ceftriaxone (amoxicillin, amoxicillin-clavulanate or cefuroxime in New Zealand) <i>All ages:</i> piperacillin-tazobactam or ceftazidime ± tobramycin ^c if severe or <i>P. aeruginosa</i> in recent cultures |
| <i>Specific pathogens</i> | | |
| <i>H. influenzae</i> ^d | | |
| β-lactamase –ve | Amoxicillin | Ampicillin (amoxicillin in New Zealand) |
| β-lactamase +ve | Amoxicillin-clavulanate or doxycycline ^b | Amoxicillin-clavulanate, cefotaxime or ceftriaxone (amoxicillin-clavulanate or cefuroxime in New Zealand) |
| <i>S. pneumoniae</i> | Amoxicillin | Benzylpenicillin G, ampicillin (amoxicillin in New Zealand) |
| <i>M. catarrhalis</i> | Amoxicillin-clavulanate | Amoxicillin-clavulanate, cefotaxime or ceftriaxone (amoxicillin-clavulanate or cefuroxime in New Zealand) |
| <i>S. aureus</i> MRSA | Di-/flucloxacillin seek specialist advice ^e | Flucloxacillin seek specialist advice ^e |
| <i>P. aeruginosa</i> | Ciprofloxacin (max 14-days) | Piperacillin-tazobactam or ceftazidime ± tobramycin ^c |
| <i>Aspergillus</i> or NTM | Seek specialist advice | Seek specialist advice |

Abbreviations: MRSA, methicillin-resistant *S. aureus*; NTM, non-tuberculous mycobacteria species.

^aIn addition to clinical severity, initial empiric therapy is also guided by previous lower airway culture results (sputum, bronchoscopy washings or bronchoalveolar lavage), local antibiotic susceptibility patterns, patient tolerance and hypersensitivity to antibiotics and prior responses to antibiotic treatments. In children too young to expectorate sputum, and in older patients when no previous lower airway culture results are available, prescribed empiric antibiotic therapy should be active against *Haemophilus influenzae*, *Streptococcus pneumoniae* and *Moraxella catarrhalis*.¹⁶

^bDoxycycline is used only in adults and adolescents.

^cAlthough treating serious *Pseudomonas aeruginosa* infections in adults with combined beta-lactam and aminoglycoside antibiotic therapy provides no additional clinical benefit than when using a single beta-lactam agent,²¹ the role of single beta-lactam antibiotics compared with combination antibiotic therapy for *P. aeruginosa* associated exacerbations in bronchiectasis remains unproven.²² Thus, in the absence of evidence, the current standard of care of combination parenteral antibiotic therapy for moderate to-severe exacerbations in children/adolescents should continue, as it should in adults when multi-resistant *P. aeruginosa* strains are suspected. Otherwise, in elderly patients and those with significant medical co-morbidities, beta-lactam monotherapy is recommended. Whenever systemic aminoglycoside antibiotics are prescribed, careful monitoring for toxicity and measurement of serum drug levels are required. Seek specialist advice when treating multi-resistant organisms, including *P. aeruginosa*.

^dRoutine beta-lactam susceptibility testing for *H. influenzae* is unreliable and no longer performed by many microbiology laboratories for respiratory isolates. Amoxicillin/ampicillin remains standard therapy for beta-lactamase negative strains, while amoxicillin-clavulanate, cefotaxime/ceftriaxone or cefuroxime (New Zealand) are appropriate for beta-lactamase positive strains. Seek specialist advice if further susceptibility testing is required.

^eSpecialist advice should be sought for treating MRSA strains in accordance with local susceptibility patterns and infection control policies.

assessments to determine if there is a clinical benefit. Those receiving longer courses should be reviewed regularly for adherence, development of adverse events (especially gastrointestinal and auditory) and emergent macrolide-resistant bacteria, and to determine risk versus benefit.

Before commencing long-term macrolides; (a) seek specialist advice, (b) ensure NTM infection is excluded in patients capable of providing a sputum specimen, (c) perform an electrocardiogram in adults to determine their QTc interval (an electrocardiogram should also be ordered in children/adolescents when there is a history of acute cardiac events, if there is a family history of prolonged QT syndrome and/or arrhythmias, or if already receiving QT prolonging medication) and (d) warn adults that macrolides may adversely affect their hearing, and if confirmed the antibiotic will be discontinued.

Statement-15

Long-term (>3-months) inhaled antibiotics (e.g., aminoglycosides, colistin) should not be prescribed routinely for

patients with clinically stable bronchiectasis. They should be considered in adults with bronchiectasis who have *P. aeruginosa* infection and frequent exacerbations (≥3 episodes/year). Similarly, long-term inhaled antibiotics should be considered in adult patients if oral macrolides are either contraindicated or poorly tolerated or if they have not reduced the frequency of pulmonary exacerbations (irrespective of *P. aeruginosa* status). Seek specialist advice for children/adolescents.

Those receiving long-term inhaled antibiotics should be reviewed regularly to monitor their clinical progress, and to assess therapy adherence. Also, discuss (with the patient) if the antibiotic is still being tolerated, and determine the overall risk versus benefit of this treatment.

Statement-16

Inhaled and oral corticosteroids should not be prescribed routinely in either the short or long-term, unless there is an established diagnosis of co-existing asthma and/or eosinophilic airway inflammation. In

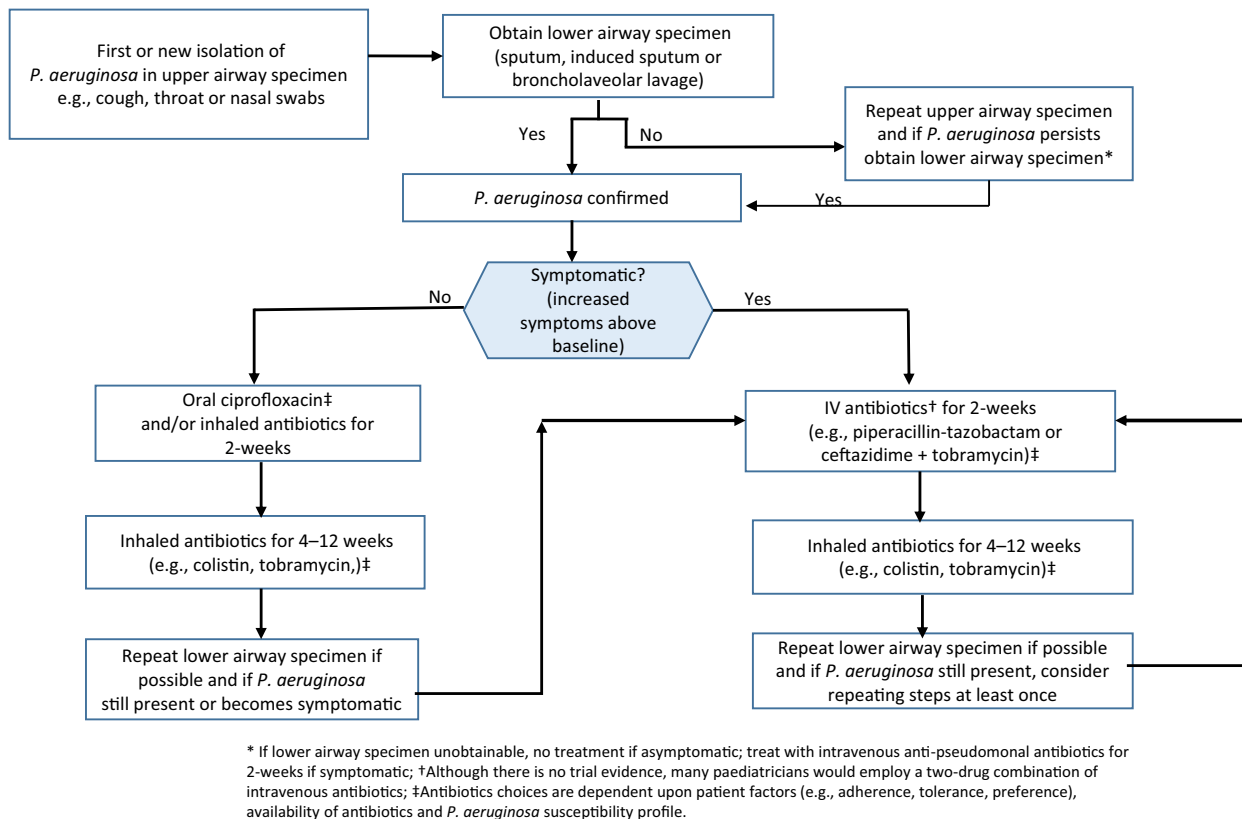


FIGURE 1 Suggested pathway for treatment when *Pseudomonas aeruginosa* is newly detected. Figure adapted with permission from the European Respiratory Society paediatric guideline for managing children/adolescents with bronchiectasis.¹⁶

adults with bronchiectasis, inhaled corticosteroids may be beneficial in those with peripheral blood eosinophilia (>3%).

Statement-17

Inhaled bronchodilators should not be prescribed routinely and instead used only on an individual basis (e.g., before using inhaled mucoactive agents or inhaled antibiotics or before undertaking ACTs).

Statement-18

Recombinant human deoxyribonuclease is contraindicated in CSLD/bronchiectasis.

Statement-19

Mucoactive agents, including inhaled isotonic saline, hypertonic saline and mannitol, are currently not recommended routinely. Consider a therapeutic trial in children/adolescents and adults with continuing frequent exacerbations despite other interventions (e.g., long-term antibiotics). If bronchial hyper-responsiveness is present, a rapid-onset

bronchodilator should be given pre-administration of hypertonic saline or mannitol.

Statement-20

ACTs are recommended. These should be individualized, and their methods and frequency reviewed, preferably biannually by a respiratory physiotherapist.

Statement-21

Offer pulmonary rehabilitation (including exercise training) to adults with bronchiectasis, especially those who have reduced exercise tolerance and/or frequent exacerbations.

Statement-22

Regular physical activity is recommended for children/adolescents and adults with CSLD/bronchiectasis.

Statement-23

Assess and optimize nutritional status.

Statement-24

Promote elimination of smoking, including vaping/e-cigarettes, cannabis and second-hand smoke exposure.

Statement-25

Promote avoidance of airborne pollutants, including traffic emissions, biomass smoke and other settings with poor air quality.

Statement-26

Regularly monitor and manage complications and comorbidities (described below). When present, these are managed following standard guidelines. Review at least annually in adults and 6-monthly in children/adolescents, ideally using a multi-disciplinary team approach. The review includes assessment of:

- a. Severity, which includes oximetry and spirometry;
- b. Sputum culture (when available) for routine bacterial and annual mycobacterial culture;
- c. Management of treatable traits and possible complications and/or co-morbidities, particularly for nutritional deficiencies, gastro-oesophageal reflux disease/aspiration, asthma, COPD (in adults), allergic bronchopulmonary aspergillosis and specific pathogens (e.g., *P. aeruginosa* or NTM), odontogenic and otorhinolaryngeal disorders, urinary incontinence, anxiety and mental health disorders. Less commonly, patients require assessments for sleep-disordered breathing and cardiac complications;
- d. General well-being and bronchiectasis-specific health education, including that related to exacerbations.

Statement-27

Although surgery is not indicated normally, there may be circumstances requiring assessment by a multi-disciplinary team expert in bronchiectasis. Factors to address are firstly ensuring it is focal disease, then optimizing medical management, and next considering the patient's age, symptoms, underlying aetiology (likelihood of recurrence), and the facility where surgery can be undertaken (expertise and availability of pre- and post-surgical care).

Statement-28

Vaccinate according to National Immunization Program Schedules, which include pneumococcal and influenza vaccine recommendations for high-risk patients with chronic lung disorders. Ensure timely annual seasonal influenza

vaccination. COVID-19 vaccination should follow national public health policies. Finally, consider vaccinating household contacts annually with the seasonal influenza vaccine.

Statement-29

A multidisciplinary approach to healthcare is recommended. Key considerations should focus upon tailoring care to the individual, reducing barriers to care, improving adherence to treatment and providing culturally responsive healthcare. Evaluation by clinicians with expertise in CSLD/bronchiectasis is recommended to confirm the diagnosis, investigate the aetiology, assess severity, provide education and develop individualized treatment and self-management plans. Clinical deterioration should prompt early referral to specialist CSLD/bronchiectasis services.

Statement-30

All people with CSLD/bronchiectasis should receive care by a specialist respiratory service with expertise in bronchiectasis. More frequent review for those with bronchiectasis should be undertaken in patients with any of the following: moderate disability, deteriorating lung disease, haemoptysis, pulmonary hypertension, frequent exacerbations (≥ 3 /year), lower airway *P. aeruginosa* or NTM infection, severe asthma symptoms, presence of underlying cause (e.g., primary ciliary dyskinesia, immunodeficiency, autoimmune diseases), advanced disease and if they are being considered for lung transplantation.

Statement-31

Providing healthcare for Australian First Nations, Māori and Pacific Islanders and under-serviced communities in rural-remote regions requires flexible and adaptive arrangements. However, it should not alter the objective of delivering best-practice treatment to these populations.

Statement-32

Given the high prevalence of CSLD/bronchiectasis in Australian First Nations, Maori and Pacific Islanders, a high index of suspicion with early diagnostic investigation, and institution of best-practice treatment should be established. Interpreters and local health-workers should be available for education regarding the disease and its management.

Statement-33

Health services should provide transitional care that supports the requirements of adolescents with

bronchiectasis. A purposeful, planned and developmentally appropriate transition, which includes engagement of the paediatric and adult multi-disciplinary teams, utilization of transition guidelines, and clear documentation of transfer processes are key goals as they transition from child to adult health services.

CONCLUSION

Although CSLD/bronchiectasis are described as amongst the most neglected pulmonary disorders,²³ since the latest TSANZ guideline in 2015⁴ our knowledge and evidence base^{1,3,24,25} for managing these conditions has expanded considerably. An important advance was recognizing that with optimal care, early cylindrical bronchiectasis in children/adolescents is potentially reversible.^{1,16} Another was identifying the importance of 'treatable traits', which seek to address the heterogeneity of CSLD/bronchiectasis by individualizing the pulmonary, aetiological, extra-pulmonary and environmental/lifestyle impacts of each disorder.^{1,3,26} Accordingly, and with the assistance of consumers, this TSANZ position statement is designed to help clinicians, health service managers, patients and families achieve the patient-oriented goals of optimizing lung growth in children, preserving lung function, minimizing exacerbations, optimizing quality-of-life, preventing complications and disease progression and if possible, reversing early bronchiectasis. Finally, the inequity of resources available for people with bronchiectasis living in Australia and NZ¹²⁻¹⁵ also needs addressing.

AUTHOR CONTRIBUTION

Anne B Chang: Conceptualization (lead); data curation (lead); methodology (lead); project administration (lead); writing – original draft (lead). **Scott Cameron Bell:** Data curation (supporting); writing – review and editing (supporting). **Catherine A Byrnes:** Data curation (supporting); writing – review and editing (supporting). **Anne Holland:** Data curation (supporting); writing – review and editing (supporting). **Emma Kennedy:** Data curation (supporting); writing – review and editing (supporting). **Paul King:** Data curation (supporting); writing – review and editing (supporting). **Pamela Laird:** Data curation (supporting); writing – review and editing (supporting). **Sarah Mooney:** Data curation (supporting); writing – review and editing (supporting). **Lucy Morgan:** Data curation (supporting); writing – review and editing (supporting). **Marianne Parsons:** Data curation (supporting); writing – review and editing (supporting). **Betty Poot:** Data curation (supporting); writing – review and editing (supporting). **Maree Toombs:** Data curation (supporting); writing – review and editing (supporting). **Paul J Torzillo:** Data curation (supporting); writing – review and editing (supporting). **Keith Grimwood:** Data curation (supporting); writing – review and editing (equal).

AFFILIATIONS

- ¹Australian Centre for Health Services Innovation, Queensland University of Technology, Brisbane, Queensland, Australia
- ²Department of Respiratory & Sleep Medicine, Queensland Children's Hospital, Brisbane, Queensland, Australia
- ³NHMRC Centre for Research Excellence in Paediatric Bronchiectasis (AusBREATHE), Child Health Division, Menzies School of Health Research, Charles Darwin University, Darwin, Northern Territory, Australia
- ⁴Thoracic Medicine, The Prince Charles Hospital, Brisbane, Queensland, Australia
- ⁵Faculty of Medicine, University of Queensland, Brisbane, Queensland, Australia
- ⁶Translational Research Institute, Brisbane, Queensland, Australia
- ⁷Department of Paediatrics, University of Auckland, Auckland, New Zealand
- ⁸Starship Children's Hospital, Auckland, New Zealand
- ⁹Department of Respiratory Medicine, Middlemore Hospital, Auckland, New Zealand
- ¹⁰University of Auckland, Auckland, New Zealand
- ¹¹Department of Physiotherapy, Alfred Health, Melbourne, Victoria, Australia
- ¹²Central Clinical School, Monash University, Melbourne, Victoria, Australia
- ¹³Institute for Breathing and Sleep, Heidelberg, Victoria, Australia
- ¹⁴College of Medicine and Public Health, Flinders University, Darwin, Northern Territory, Australia
- ¹⁵Board of Northern Territory General Practice Education Ltd, Darwin, Northern Territory, Australia
- ¹⁶Pandanus Medical NT, Millner, Northern Territory, Australia
- ¹⁷Departments of Respiratory and Sleep Medicine and Medicine, Monash Medical Centre, Monash University, Melbourne, Victoria, Australia
- ¹⁸Department Physiotherapy, Perth Children's Hospital, Perth, Western Australia, Australia
- ¹⁹Wal-yan Respiratory Research Centre, Telethon Kids Institute, Perth, Western Australia, Australia
- ²⁰Department of Paediatrics, School of Medicine, University of WA, Perth, Western Australia, Australia
- ²¹School of Clinical Sciences, AUT University, Auckland, New Zealand
- ²²Department of Respiratory Medicine at Concord and Nepean Hospitals, School of Medicine, Faculty of Medicine and Health, University of Sydney, Sydney, New South Wales, Australia
- ²³Representative of Parent Advisory Group, NHMRC Centre of Research Excellence in Paediatric Bronchiectasis, Cough and Airways Research Group, Australian Centre for Health Services Innovation, Queensland University of Technology, Brisbane, Queensland, Australia
- ²⁴Respiratory Department, Hutt Hospital, Te Whatu Ora Capital, Coast and Hutt Valley, Lower Hutt, New Zealand

²⁵School of Nursing Midwifery, and Health Practice, Victoria University of Wellington, Wellington, New Zealand

²⁶School of Public Health, University of Sydney, Sydney, New South Wales, Australia

²⁷Nganampa Health Council, Alice Springs, Northwest Territories, Australia

²⁸Royal Prince Alfred Hospital, University of Sydney, Sydney, New South Wales, Australia

²⁹School of Medicine and Dentistry, Menzies Health Institute Queensland, Griffith University, Southport, Queensland, Australia

³⁰Departments of Infectious Diseases and Paediatrics, Gold Coast Health, Southport, Queensland, Australia

ACKNOWLEDGEMENTS

Health professionals who contributed to the Delphi statement: *Nurses and Physiotherapists*: Kathleen Hall, Christine Wilson, Annemarie Lee, Donna Mason, Gabrielle McCallum, Anna Middleton, Lesley Versteegh, Kathy Lawton. *Adult-based Medical Practitioners*: Lucy Burr, Bart Currie, John Kolbe, Lata Jayaram, Chen Li Holmes-Liew, Peter Middleton, Jonathan Rutland, Frank Thien, Grant Waterer, Conroy Wong, Tim Baird, Daniel Smith, James Geake, Rachel Thompson. *Paediatric-based Medical Practitioners*: Vikas Goyal, Peter Morris, Anna Mulholland, Brent Masters, Andre Schultz, Gurmeet Singh, Dhanusya Sivananthan, Rahul Thomas, Danielle Wurzel, Julie Marchant, Bernadette Prentice, Hiran Selvadurai. *General Practice*: Kylie Vuong.

Research funding: Open access fees were provided by the TSANZ. Supported by the National Health and Medical Research Council (NHMRC) Centre for Research Excellence in bronchiectasis for children (grant 1170958). Anne B. Chang is funded by a NHMRC practitioner fellowship (grant 1154302) and the Queensland Children's Hospital Foundation. Open access publishing facilitated by Charles Darwin University, as part of the Wiley - Charles Darwin University agreement via the Council of Australian University Librarians.

CONFLICTS OF INTEREST STATEMENT

Catherine A. Byrnes: Trustee, Bronchiectasis Foundation; Editor, New Zealand Formulary for Children at the Starship Children's Hospital. Has received funds from Health Research Council of New Zealand research, CureKids research and FluLab International research on the subject matter. Anne B. Chang: European Respiratory Society (ERS) Guidelines Committee Member, ERS Long Strategy Member, APSR Strategic Planning Member and Australian Bronchiectasis Consortium and Registry. Has written for UpToDate and has received multiple NHMRC and MRFF grants on the subject matter. Sarah Mooney has received funds from Asthma NZ to present at GP and annual conference. Lucy Morgan: Past Chair of Australian Bronchiectasis Consortium and Registry. Has received personal funds from Astra Zeneca, Zambon, Insmad and Glaxo; and administrative support from the Lung Foundation of Australia. Received payment as a PI in commercial clinical trials. Betty

Poot: Currently involved in recruiting patients to the NZ bronchiectasis registry where data has been used for conference presentations and for writing up manuscript for publication. Scott C. Bell, Paul Dawkins, Keith Grimwood, Anne E. Holland, Emma Kennedy, Paul T. King, Pamela Laird, Marianne Parsons and Paul J. Torzillo have nothing to disclose.

DATA AVAILABILITY STATEMENT

Not applicable.

ORCID

Anne B. Chang  <https://orcid.org/0000-0002-1331-3706>

Scott C. Bell  <https://orcid.org/0000-0001-8651-7139>

Anne E. Holland  <https://orcid.org/0000-0003-2061-845X>

Paul T. King  <https://orcid.org/0000-0001-5197-1163>

Pamela Laird  <https://orcid.org/0000-0002-9799-0471>

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

How to cite this article: Chang AB, Bell SC, Byrnes CA, Dawkins P, Holland AE, Kennedy E, et al. Thoracic Society of Australia and New Zealand (TSANZ) position statement on chronic suppurative lung disease and bronchiectasis in children, adolescents and adults in Australia and New Zealand. *Respirology*. 2023;28(4):339–49. <https://doi.org/10.1111/resp.14479>