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Review

Management of children and adolescents with bronchiectasis: summary of the ERS clinical practice guideline

Bronchiectasis, characterised by chronic wet/productive cough with recurrent respiratory exacerbations and abnormal bronchial dilatation on computed tomography scans, remains an increasingly recognised but often neglected chronic pulmonary disorder in children and adolescents. An early diagnosis combined with optimal management offers the prospect, at least in some patients, of curing a condition previously considered irreversible. However, unlike in adults, until now no international paediatric guidelines existed. The recently published European Respiratory Society clinical practice guidelines for the management of children and adolescents with bronchiectasis attempts to address this clinical information gap. The guidelines were formulated by panel members comprised of experts from several relevant health fields, the European Lung Foundation and parents of children with bronchiectasis. Systematic reviews and the GRADE (Grading of Recommendations, Assessment, Development and Evaluation) approach guided the nature and strength of recommendations. The recommendations are grouped into clinically relevant topics: diagnosis, evaluating for underlying causes, defining exacerbations, management, systematic care, monitoring, reversibility and prevention. The guidelines seek to achieve: 1) optimal lung growth, 2) preserved lung function, 3) enhanced quality of life, 4) minimal exacerbations, 5) few or no complications, and 6) if possible, reversal of lung injury for each child/adolescent with bronchiectasis. This review presents example cases that highlight the recommendations of the clinical practice guidelines.

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Educational aims

This article is intended for those involved in caring for children/adolescents with bronchiectasis. It aims to inform:

- Clinicians of the European Respiratory Society recommendations for the diagnosis and management of children/adolescents with bronchiectasis.
- Adolescents and parents of children/adolescents with bronchiectasis of these recommendations, so as to assist discussions with healthcare teams and help facilitate access to appropriate care.

@ERSpublications
An international guideline for managing children/adolescents with bronchiectasis is now available
https://bit.ly/3A3XYnN

What is bronchiectasis?

Bronchiectasis is a clinical syndrome of recurrent or persistent wet/productive cough, lower airway infection and/or inflammation, and abnormal bronchial dilatation on chest computed tomography (CT) scans. It is a heterogenous chronic pulmonary disorder with many risk factors and aetiologies (figure 1) [1, 2]. Moreover, wide regional variations in prevalence exist globally.

Interrupting the infection/inflammation cycle as early as possible with effective treatment is necessary to reverse and/or halt disease progression and structural lung injury [2, 3]. Without disrupting this cycle, ongoing impaired mucociliary clearance leads to repeated or persistent infection and inflammation and progressive airway injury, predisposing the damaged airway to further cycles of infection and inflammation (figure 1) [1].

Previously considered irreversible, current data show that paediatric bronchiectasis might be reversible over time [2]. Achieving this goal requires early disease recognition leading to appropriate investigations to both diagnose and manage bronchiectasis. Evaluation for treatable underlying causes and effective management are thus important [1, 2]. Clinicians now aim to achieve a cure in at least a subset of children [2, 4].

Although there are similarities between paediatric and adult bronchiectasis, such as wet/productive cough being the dominant symptom and having recurrent exacerbations, important differences also exist. These differences include the underlying aetiology and accompanying comorbidities [1], prognosis (when optimally treated) [2], lower airway microbiology (bacterial pathogens [5] and microbial communities [6]) and age-related immunological responses [7]. Children/adolescents also require developmentally appropriate care, support and supervision from their parents. Furthermore, some diagnostic [2, 4] and treatment methods differ, including airway clearance techniques (ACT), which are age and cognition dependent [4]. Compared with adult-onset bronchiectasis, those with untreated bronchiectasis symptoms from childhood have worse disease and a poorer prognosis [8]. Australian data indicate that >60% of adults with bronchiectasis have symptoms dating from childhood [8].

Children with bronchiectasis are still managed in some cystic fibrosis (CF) centres, where they may not receive optimal care for their needs. It is recognised that treatments effective in children with CF may not benefit, or may even be harmful for, those with bronchiectasis unrelated to CF. An example is inhaled recombinant human deoxyribonuclease (rhDNase), which in contrast with its actions in CF patients is associated with harm [4].

Bronchiectasis is under-diagnosed and often not managed optimally

Although no longer considered rare, bronchiectasis is still one of the most neglected lung disorders [9],

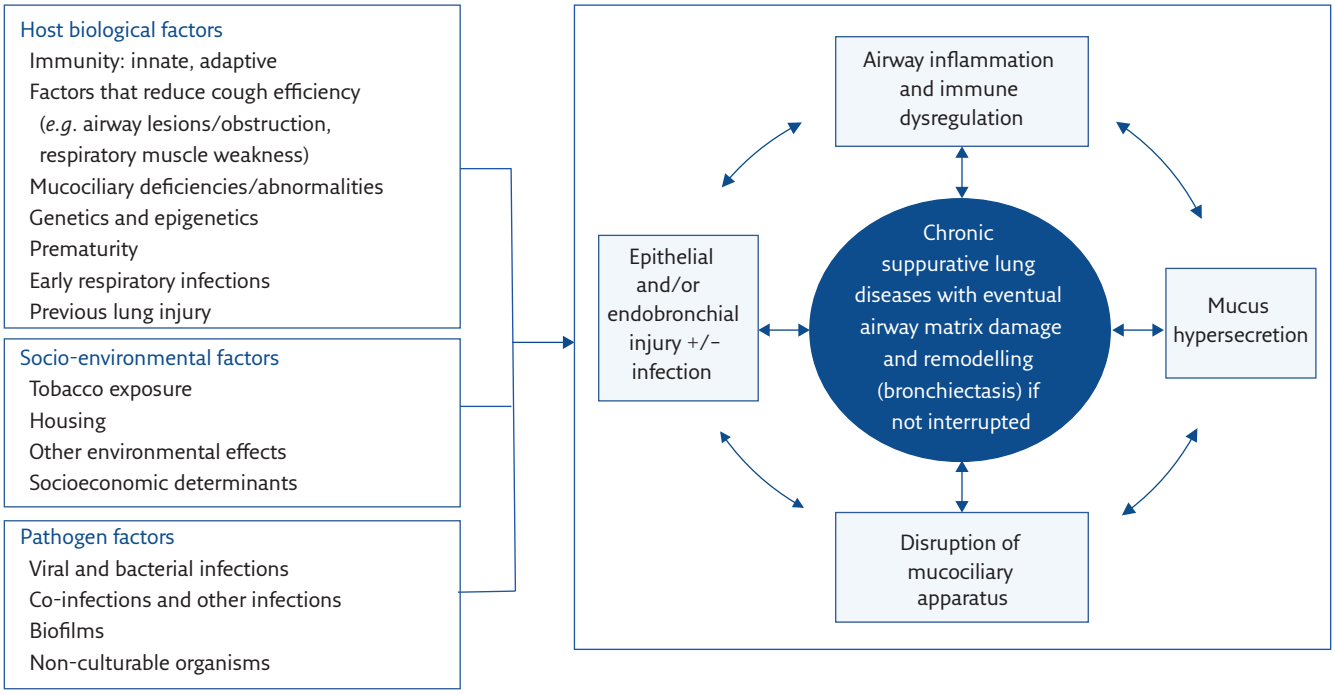

Figure 1 *Contributing factors (host, socio-environmental and pathogen) to the development of chronic suppurative lung disease with key features of the pathobiology of bronchiectasis. If the pathways are not interrupted, the resulting airway remodelling eventually becomes irreversible, leading to severe/irreversible bronchiectasis.*

with high individual disease burden, economic cost [10] and poor quality of life (QoL) for children/adolescents [11] and their parents [12]. Also, there are large disparities in the standards of care and outcomes between bronchiectasis and other chronic lung diseases [13], including those with bronchiectasis from within the same country [14].

In an international survey undertaken by the European Lung Foundation (ELF), the top clinical needs of parents of children with bronchiectasis included standard treatments afforded to other chronic lung diseases, such as having access to paediatric chest physiotherapy services and being taught ACT and how to use the equipment at home [15].

In this context there is a clear need for an international guideline for children/adolescents with bronchiectasis. The recent European Respiratory Society (ERS) clinical practice guideline (CPG) [4] serves this purpose. This article highlights several key points of the CPG and includes illustrative case studies.

The methodology of the guideline development process was outlined in the original guideline publication [4]. All references to the recommendations in this article refer to the summaries in table 2 of the CPG [4] (Patient, Intervention, Comparison, Outcome (PICO) questions) and table 3 of the CPG [4] (narrative questions (NQ)).

Diagnosis and evaluating causes

Case 1

This child (box 1) has the clinical syndrome of bronchiectasis and illustrates that a chest CT scan is required to confirm a diagnosis of bronchiectasis, using the paediatric criteria to define abnormal bronchial dilatation (PICO-1) [4]. Additionally, a minimum panel set of tests should be undertaken. Some children require additional assessment (*e.g.* for this child, a video fluoroscopic swallow study is

Box 1

Case 1, a girl aged 2 years and 3 months (corrected), was referred for recurrent wet cough. Her past history included being born preterm at 27 weeks' gestation and having had chronic neonatal lung disease. Her supplemental oxygen was ceased 20 months earlier.

Her recurrent wet cough began after she was hospitalised with "viral pneumonia" (although no viral pathogens were determined) >12 months ago. After 8 weeks of a persistent wet cough, she received a 4-week course of oral antibiotics, following which the cough improved, only to return 2 weeks later once the antibiotics were discontinued. Since then, in the last 9 months she has had repeated 4-week courses of antibiotics, but with the cough returning after 1–2 weeks of finishing the antibiotics. She was also noted to be "wheezy" on occasions, but does not get shortness of breath. In addition, she can sometimes cough and splutter with thin fluids and on occasions gags on purees and solids.

She attends childcare 1 day per week, is not exposed to tobacco smoke and is fully immunised in accordance with the national immunisation schedule. Her parents were not related to each other and there is no family history of primary immunodeficiency, bronchiectasis or CF.

Examination revealed a well-grown child (87th percentile for weight and 93rd for height). She had a spontaneous wet cough, a normal voice and a clear chest on auscultation, and her tympanic membranes were normal. She did not have chest wall deformity, digital clubbing or any respiratory distress. The remainder of her examination was also normal. Her chest radiograph showed nonspecific increased bronchovascular markings.

Bronchiectasis was suspected and the child underwent a chest CT scan under general anaesthesia. The multidetector CT scan with high resolution revealed several segments where the broncho–arterial ratio (BAR) was 0.9 in the lingula and left lower lobes.

A flexible bronchoscopy, undertaken under the same anaesthetic, showed mildly purulent secretions in several major bronchi. The bronchoalveolar lavage (BAL) cultured *Haemophilus influenzae* at 10^4 colony-forming units (cfu) per mL. The total white cell count in the BAL fluid was 1500×10^3 per mL, with 60% neutrophils (indicating airway neutrophilia) and an absence of airway eosinophilia (0.1%, which suggests that asthma medications would not be beneficial despite the history of wheezing).

Her immunoglobin (Ig)G level was mildly elevated. Her full blood count indices, IgM, IgA and IgE were within normal limits. Her vaccine responses to both tetanus and *H. influenzae b* vaccine antigens showed long-term protection levels. Her sweat chloride was not elevated (9 $mmol\cdot L^{-1}$). Testing for tuberculosis and HIV was not undertaken as there was no history of close contact with tuberculosis and the prevalence of both infections is very low in her community.

The speech pathologist review after her current cough settled was unremarkable. Her video fluoroscopic swallow study revealed no evidence of aspiration.

needed to exclude recurrent aspiration) when the child's history suggests other possible contributing factors (NQ-1).

Diagnosing paediatric bronchiectasis (PICO-1)

Diagnosing bronchiectasis at an early stage was a top priority for parents of children/adolescents with bronchiectasis and also for adults who had bronchiectasis as a child [15]. The positive impact of diagnosing bronchiectasis promptly, as described in the CPG's narrative evidence [4], included improving QoL [16] and lung function in many cohorts from various aetiologies (including primary immunodeficiency disorders [17]).

Abnormally dilated airways are the main CT scan characteristic of bronchiectasis. The old definition of abnormally dilated airways (broncho–arterial ratio (BAR; defined as the ratio of the inner diameter of the airway to the outer diameter of the adjacent artery) >1–1.5 as a single cut-off irrespective of age) was based on adult data. Although the data were limited to adult patients, the CPG's narrative evidence [4] documented the significantly higher sensitivity of multidetector CT scans combined with high-resolution CT (HRCT), compared to HRCT alone for detecting bronchiectasis.

The BAR correlates with age [18] and increases as bronchiectasis becomes more severe (from cylindrical to varicose to cystic [2]). Early diagnosis necessitates defining the BAR based upon paediatric rather than adult data. The summary of evidence includes two paediatric studies [19, 20], showing that the mean BAR is lower in children/adolescents than in adults.

Although the evidence for the type of CT scan and BAR cut-off was weak, the recommendation placed a relatively higher value on the more accurate and early detection of bronchiectasis and its importance for subsequent management, and a relatively lower value on the currently available evidence. However, as there are false-positive results with diagnosing bronchiectasis based purely on the BAR, the panel advocated that the BAR alone should not be used to diagnose bronchiectasis. Instead, it is best based on clinical features consistent with this diagnosis being present and then confirmed radiographically.

What standard tests that impact on clinical outcomes should be undertaken? (NQ-1)

In all children with bronchiectasis, the CPG determined that a standard set of investigations should be performed to screen for treatable causes of bronchiectasis that are either common or critical (*e.g.* immunodeficiency, infection or CF). Despite the very low-quality evidence, the severe consequences of missing treatable causes warrant investigations being undertaken.

The recommendation was conditional, based upon the large desirable effect and likely trivial undesirable effects of setting a standard set of investigations, and the risk and harm of not managing common or critical conditions related to bronchiectasis in children/adolescents. Lung function and respiratory cultures do not identify the cause, but help assess severity and guide antibiotic choices, respectively, and thus help optimise treatment. Implementation requires health services to increase accessibility to centres practising standard-of-care management for children/adolescents with bronchiectasis that includes undertaking the recommended minimum panel of tests.

What criteria should be used to define an exacerbation? (NQ-6)

Recognising respiratory exacerbations promptly is important, as exacerbations are associated with increased psychological stress and impaired QoL [21]. Hospitalised exacerbations are associated with accelerated lung function decline (forced expiratory volume in 1 s (FEV_1) −1.9% predicted per hospitalised exacerbation) and substantial healthcare costs [10, 22]. Also, as managing exacerbations is a key component of bronchiectasis care and one of the three top issues for parents, it is important to increase patient, parent/carer and health professional education in recognising exacerbations and commencing additional treatments.

The recommendation was based upon several prospective studies. We considered that ≥3 days of increased symptoms is required to fulfil the definition, except when either immunodeficiency (lower threshold is suggested) or hypoxia/dyspnoea (immediate treatment is mandated as this indicates the exacerbation is severe) are present. Children/adolescents with neurodevelopmental conditions may have subtle and/or individually recognised symptoms of an exacerbation, whereby earlier treatment may also be necessary.

Management

Airway clearance techniques (PICO-4)

ACT are universally undertaken in people with chronic suppurative lung diseases. Having access to physiotherapists with expertise in paediatric lung diseases and being taught the techniques and how to use the equipment at home were management priorities highlighted by the ELF survey [15].

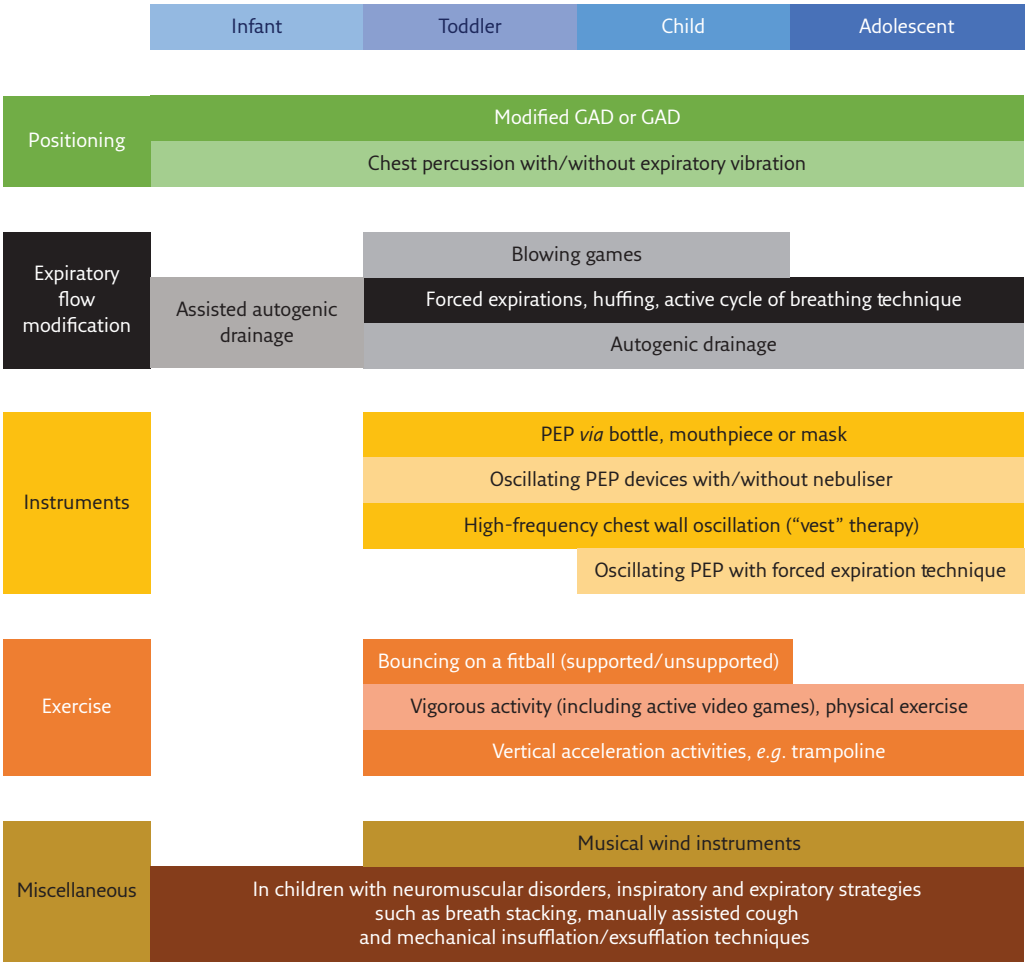

Figure 2 *There are many different ACT. In children/adolescents, these are age-specific and best taught by physiotherapists experienced in managing children/adolescents with bronchiectasis. GAD: gravity-assisted drainage; PEP: positive expiratory pressure. Reproduced from the ERS CPG [4] with permission.*

Many ACT exist and the cognitive ability of children/adolescents varies widely (figure 2). Thus, the panel recommended that individualised therapy be taught, and reviewed at least biannually, by paediatric-trained chest physiotherapists. Increased airway secretions occur during exacerbations and more intensive ACT would be beneficial but may require adjustment to fit the circumstances (*e.g.* exercises may be impractical).

Case 2

This case (box 2) illustrates the CPG's approach to the use of empirical antibiotics for exacerbations and, when the exacerbation is unresolved, the need for intravenous antibiotics (PICO-5) and more intensive ACT (PICO-4). When possible, sputum should be re-cultured during exacerbations and, if *Pseudomonas aeruginosa* is newly isolated, PICO-6 should be applied. As the child had severe recurrent exacerbations, long-term azithromycin was commenced (PICO-7). Lastly, this case emphasises that bronchiectasis can be stable (or even reversed) in children who are managed optimally, making lung surgery of bronchiectatic lobes unnecessary in almost all cases (NQ-7) [2].

Antibiotics for exacerbations (PICO-5)

Treating acute respiratory exacerbations with antibiotics is considered standard care in children and adults with bronchiectasis [4, 23, 24]. While no placebo randomised controlled trials (RCTs) exist in adults, we found one high-quality RCT that showed amoxicillin-clavulanate was superior to placebo at resolving symptoms after 14 days of treatment in children with mild (non-hospitalised) exacerbations [25]. Amoxicillin-clavulanate also significantly reduced the duration of the exacerbation, while in contrast this was similar between azithromycin and placebo amongst those whose symptoms resolved by day 14 [25]. Although we recommended 14 days of antibiotics, the optimal treatment duration is yet to be studied. Nevertheless, patients should

Box 2

Case 2 is a boy who was first referred at age 5 years. His past history included repair of his tetralogy of Fallot at 6 months, which was uneventful. He has had a chronic wet cough for at least 3 years, but his growth and development are normal. He did not have a history of persistent nasal congestion/discharge, recurrent otitis media or otorrhoea and there were no symptoms and signs suggestive of an underlying immunodeficiency. Clinical examination was remarkable only for localised crackles on auscultation over the right lower lobe; digital clubbing was not present and both tympanic membranes and his nasal passages were normal in appearance. His chest radiograph demonstrated situs solitus and atelectasis of the right lower lobe.

His panel of minimum tests were unremarkable and his vaccine response to polysaccharide pneumococcal 23-valent was robust. As he had a past history of congenital heart disease, nasal cilia biopsy was undertaken. The nasal cilia motility and electron microscopy results were both inconclusive and later gene analyses showed he had primary ciliary dyskinesia.

His first chest multidetector CT scan with HRCT showed bronchiectasis with localised collapse of the right lower lobe. Flexible bronchoscopy revealed mild airway bronchomalacia of the main right lower lobe bronchus. He had purulent secretions in several major bronchi and BAL cultured *H. influenzae* (10^6 cfu·mL^{-1}) and *Moraxella catarrhalis* (10^5 cfu·mL^{-1}). The total white cell count was 5100×10^3 per mL of BAL fluid with airway neutrophilia of 85%.

Case 2 was hospitalised and managed with 14 days of intravenous ceftriaxone and intensive twice daily airway clearance by a paediatric physiotherapist. The family also received bronchiectasis-specific education and at discharge the child's cough was dry and chest crackles had resolved. Although the bronchiectatic changes were localised, his family was counselled against lung surgery.

Over the next 3 months, his wet cough recurred and chest crackles returned. Failing to respond to 3 weeks of oral amoxicillin-clavulanate, he was re-hospitalised for empiric intravenous ceftriaxone and intensive twice daily airway clearance. A sputum culture did not isolate any respiratory bacterial pathogens. On discharge he was commenced on regular azithromycin (doses of 10 mg·kg^{-1}, three times a week), which he received for the next 2 years.

When aged 6 years, his spirometry technique was good and revealed that his FEV_1 was 82% predicted and forced vital capacity (FVC) was 81% predicted. He was reviewed every 3–4 months in a specialist outpatient clinic where he was seen by a respiratory physiotherapist, nurse and specialist respiratory physician. When sputum was available, it was cultured routinely every 6–12 months. In total, case 2 had five hospitalisations over 13 years for exacerbations. When he transferred recently to the adult respiratory service, his spirometry values were FEV_1 80% and FVC 80% predicted.

have access to appropriate antibiotics for the recommended duration of treatment.

Antibiotics to reduce recurrent pulmonary exacerbations (PICO-7)

Some children/adolescents have frequent exacerbations. In the CPG examination of the question of whether these children should receive long-term macrolide antibiotics, three RCTs were found. Data showed that macrolides reduced the number of children/adolescents experiencing any exacerbations during the trial period (relative risk (RR) 0.86, 95% CI 0.75–0.99). The largest of the RCTs found that using long-term azithromycin halved the frequency of exacerbations (incidence rate ratio 0.5, 95% CI 0.35–0.70) and possibly reduced hospitalisation (odds ratio 0.25, 95% CI 0.06–1.07) [26]. For reducing exacerbations, the quality of evidence was high. A recently published 6-month-duration RCT involving children/adolescents and adults with primary ciliary dyskinesia also described significantly reduced exacerbation rates by a similar effect size [27].

There are, however, some contraindications to using macrolides. These include an abnormal ECG, liver function abnormalities and azithromycin hypersensitivity. The CPG considered that an ECG is not necessary before commencing macrolides, except when there is a family history of prolonged QT syndrome, arrhythmias and acute cardiac events.

Serious adverse events were numerically lower in the azithromycin group (RR 0.57, 95% CI 0.31–1.05), compared to placebo [26]. However, long-term use of azithromycin is associated with significant increases in macrolide-resistant bacteria in the upper airways. Thus, those receiving longer treatment courses (>24 months) should continue to be evaluated for risk *versus* benefit, particularly to monitor for nontuberculous mycobacteria (NTM). Moreover, as adherence (≥70%) to the macrolide regimen improved efficacy [26] and reduced antibiotic resistance [28], strategies to promote adherence are advocated.

Antibiotics for eradication treatment (PICO-6)

Lower airway infection with pathogenic microorganisms is associated with deteriorating clinical status and lung function in both the bronchiectasis [29] and CF [30] literature. While there is currently no evidence for early eradication from well-conducted trials in children/adolescents with bronchiectasis, the CPG panel considered that virtually all physicians with specific expertise in paediatric bronchiectasis would undertake interventions to eradicate initial or new isolates of *P. aeruginosa*. In this recommendation, a higher value is placed upon both the theoretical benefits of eradication and patient/carer values than upon possible treatment-related adverse effects.

The evidence from three before-and-after trials in adults showed improved QoL and reduced exacerbation rates and hospitalisation when compared to the pre-eradication period. Once *P. aeruginosa* infection is confirmed (*e.g.* not a transient coloniser in upper airway samples from a clinically stable patient), eradication treatment should be administered promptly. Without clear evidence for one regimen over another, figure 3 illustrates commonly used approaches in children/adolescents by experts in the field.

Mucoactive agents and asthma medications (PICO-2 and PICO-3)

The CPG [4] found RCTs in adults involving rhDNase, hypertonic saline, mannitol and bromhexine. Considering the overall evidence, we recommend against the use of bromhexine because of adverse events, and rhDNase in particular is contraindicated as there is risk of substantial harm (increased risk of exacerbations and accelerated lung function decline). The data for hypertonic saline and mannitol are equivocal, and the balance probably favours administering hypertonic saline and mannitol in some, but not all patients, as there is an added treatment burden and intolerance associated with these therapies. However, when compared with controls, mannitol was found to be beneficial (significantly fewer exacerbations, prolonged time to

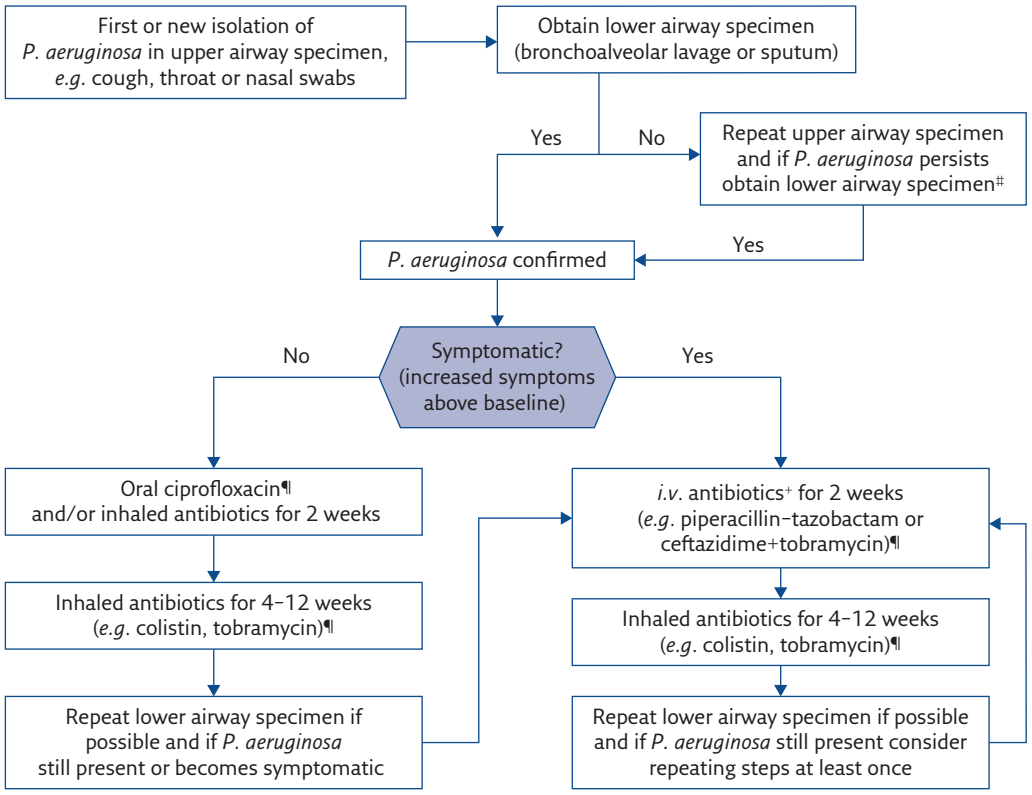

Figure 3 *Suggested management approach used by the panel when Pseudomonas aeruginosa is first or newly isolated in a child with bronchiectasis. The suggested approach depends upon: 1) the specimen type, and 2) whether the child is symptomatic. However, panel members acknowledged the approach to initiating eradication treatment is controversial. Some physicians may still feel it is appropriate to initiate eradication therapy based only on a single upper airway specimen, even when symptoms and evidence of benefit in such circumstances are absent. ‡: if no lower airway specimen available, no treatment if asymptomatic; treat with intravenous anti-pseudomonal antibiotics for 2 weeks if symptomatic. ¶: antibiotic choices are dependent upon patient factors (e.g. adherence, tolerance and preference), availability of antibiotics and P. aeruginosa susceptibility profile. +: although there is no trial evidence, many paediatricians use a combination of two intravenous antibiotics; the recommendation for administering two antibiotics when employing short (2-week) intravenous antibiotic courses is made to align with the studies included in the systematic review and the ERS adult guidelines [23]. Reproduced from the ERS CPG [4] with permission.*

next exacerbation, and symptomatic improvement) in a subgroup of adults with a high symptom burden [31]. When they are prescribed, education on using these medications and their associated equipment care is needed.

Although inhaled corticosteroids (ICS) are commonly used in some settings for children/adolescents with bronchiectasis, direct evidence is lacking for any benefit when using ICS either alone or in combination with long-acting β_2-agonists (LABA). Data for this recommendation were based largely on adult studies. Furthermore, there are increased adverse events when ICS are used, and the risk increases with higher ICS doses (*e.g.* increased risk of NTM infection, pneumonia and tuberculosis in adults with bronchiectasis and other chronic pulmonary disorders who received ICS [32, 33]).

Nevertheless, there is a subgroup with overlapping bronchiectasis and asthma symptoms. If treatment with ICS or ICS/LABA is contemplated, documenting benefit should be undertaken (if possible with objective tests, *e.g.* spirometric response following short-acting β_2-agonists).

Surgery (NQ-7)

While surgery for bronchiectasis is now rarely undertaken in high-income countries, it remains relatively common in low- and middle-income settings. Panel members rarely advocate surgery to control bronchiectasis. Surgery is only considered after maximal medical therapies (*e.g.* ACT, long-term antibiotics, *etc.*) have failed and the child/adolescent's QoL remains significantly impaired. When contemplated, a multidisciplinary approach is essential, and the decision should be based upon the individual's clinical state and local surgical expertise.

If surgery is undertaken, it should take place in specialised centres. A series of tests (*e.g.* ventilation–perfusion scan, bronchoscopy, chest CT scan) and optimising the patient's lung function pre-surgery are required.

Case 3

This case (box 3) highlights the approach to education when a child is first diagnosed with bronchiectasis, including recognition of exacerbations and education on various ACT. Also, the CPG's recommendations of systematic care and regular monitoring are emphasised by this case study.

Systematic care (NQ-3)

Paediatric systematic care

Routine immunisation, exercise (aerobic and non-aerobic) and good nutrition are undeniably beneficial, and additional vaccinations for children/adolescents with bronchiectasis likely beneficial, although the quality of the evidence is very low and the magnitude of effect specific for bronchiectasis is unknown. The positive effects of psychological support and teaching appropriate equipment use and care for children/adolescents with chronic illness are standard practice in specialised units caring for children/adolescents with bronchiectasis.

Box 3

Case 3, aged 4 years, is known to have common variable immunodeficiency (CVID) and was referred by an immunologist colleague. He had moved recently from another major capital city. When first seen, despite his known CVID and chronic wet cough (for as long as the parents can recall), he had never been diagnosed as having bronchiectasis. A chest multidetector CT scan with HRCT was undertaken, which confirmed multilobar bronchiectasis.

After bronchoscopy, which showed purulent secretions and BAL cultured *H. influenzae* (10^8 cfu·mL^{-1}), he was hospitalised for intravenous antibiotics. During the hospitalisation, he received intensive airway clearance and his family was given a personalised home ACT programme (PICO-4).

His parents were particularly concerned about whether his bronchiectasis would deteriorate and specifically asked about recognising exacerbations. The family received bronchiectasis-specific education, which consisted of recognition of exacerbations (NQ-6) and all the aspects of systematic care (NQ-3).

Prior to discharge, they received a bronchiectasis action management plan [34] and a plan on how bronchiectasis is monitored (NQ-4). They were specifically told to avoid close contact with children with CF to prevent cross-infection.

When he was reviewed in clinic 2 weeks after discharge, his wet cough had fully resolved. His parents commented how his energy levels had returned and both child and family's QoL had improved. The respiratory specialist reiterated all aspects of monitoring and systematic care. A letter communicating the above with the child's family doctor was also undertaken. Ideally, if modifiable risk factors are recognised and addressed promptly, bronchiectasis may be prevented, while its optimal treatment may avert disease progression, and if bronchiectasis is diagnosed early it may also be reversible in some children/adolescents (NQ-2).

There is low-quality evidence that exercise training leads to fewer pulmonary exacerbations and longer time to first exacerbation. Support for exercise training must be ongoing, to achieve and maintain the best effect upon respiratory health. In children/adolescents, there are no agreed formal pulmonary rehabilitation programmes and no data on what exercise interventions are most important. Whether a formal exercise programme is superior to encouragement of an active lifestyle is unclear.

Cross-infection

There are limited data on infection control policies for patients with bronchiectasis. The CPG [4] advocates avoiding direct contact with CF patients and following local policies if managed within a CF centre.

Education of families of a child/adolescent with bronchiectasis needs to include standard infection control procedures, including cough and hand hygiene measures. After writing the CPG, the coronavirus disease 2019 (COVID-19) pandemic led local health authorities to introduce additional non-pharmacological public health measures to interrupt virus transmission.

Monitoring (NQ-4)

Frequency of clinic attendance, airway microbiology testing and tests to detect complications

The Task Force panel and parent/patient advisory group (PAG) advocated for regular clinical care and monitoring by specialists. Data from the CPG's systematic review support outpatient clinic reviews every 3–6 months. Outpatient sputum culture surveillance every 6–12 months, when such specimens are available, is derived from expert opinion [24]. However, the desirable frequency of outpatient clinic attendance and airway microbiology surveillance is dependent upon various factors. These relate to the patient (*e.g.* age, underlying aetiology, illness severity, comorbidities and ability to reliably expectorate spontaneous or induced sputum) and setting (*e.g.* travelling long distances for clinic attendance).

There is a need to support access for children/adolescents to centres practising the standard of care, especially in low/middle-income countries. It is also important to educate clinicians, families and patients on the role of surveillance sputum cultures for informing future treatments, even in those with clinically stable bronchiectasis.

What investigations should be undertaken in children who are gradually deteriorating?

Specialists with expertise in bronchiectasis at tertiary paediatric hospitals currently use a model of care that includes standardised approaches for assessing stability and detecting deterioration. These include the monitoring of clinical symptoms, the frequency and severity of respiratory exacerbations, and lung function indices. When deterioration occurs, the evidence presented in the CPG supports assessing and investigating for treatable traits (*e.g.* new infection, possible comorbidities (*e.g.* asthma, gastro-oesophageal reflux disease, nutritional deficiencies, dental or sleep disorders), treatment adherence (including ACT), and exposure to tobacco smoke and other aero-toxicants (*e.g.* taking up vaping)). The recommendation was based upon indirect evidence that the current standard of care in specialist settings leads to improved lung function, reduced exacerbations and better QoL after diagnosis.

Should repeat chest CT scans be undertaken?

Evidence from the CPG's narrative summary found several studies where CT scans were repeated, and these were based largely upon individuals undergoing this investigation for clinical reasons. The panel considered that indications to repeat CT scans include documenting reversal of bronchiectasis (*e.g.* for medical insurance or reducing care burden for parents) or when there is an acute or gradual deterioration (*e.g.* to assess for new treatable disease or justify more intensive treatments). Obtaining additional CT scans needs to be balanced against the reported increased lifetime cancer risk, which is both age and dose dependent.

Reversibility and prevention (NQ-2)

Mild bronchiectasis is potentially reversible and/or preventable in some patients, although the quality of evidence was low to very low. The resolution/improvement rates may be as much as 64% after appropriate treatment in children/adolescents [35]. The CPG's systematic review found several modifiable factors that may promote reversibility and/or prevent bronchiectasis. These include early identification and treatment of inhaled foreign bodies (<14 days [36]), preventing early and severe pneumonia, preventing recurrent protracted bacterial bronchitis, treating primary and acquired immunodeficiency disorders causing bronchiectasis, promoting breastfeeding and immunisation, and avoiding tobacco smoke, vaping and other air pollutants.

Conclusions and future perspectives

The recently published ERS CPG [4] aims to assist health professionals in providing the best care management of children/adolescents with

bronchiectasis. By doing so, it is possible to optimise postnatal lung growth, preserve lung function, enhance QoL, minimise exacerbations, prevent complications and, if possible when diagnosed early, reverse the radiographic changes on CT scans. For each recommendation, the CPG [4] was guided by parents' views and sought to balance benefits and risks associated with each of the treatment approaches.

Although we used the best available evidence at the time of writing the CPG, there are large knowledge gaps for managing bronchiectasis, a neglected chronic disorder with known inequities. The research priorities identified by ELF and the PAG are now established [15] and need addressing to improve the lives of children/adolescents with bronchiectasis and their families. Precision medicine approaches defining endotypes and phenotypes should also be identified, evaluated and advanced into practice.

We are hopeful that the newly established ERS Clinical Research Collaboration for paediatric bronchiectasis (Children's Bronchiectasis Education, Advocacy and Research Network (Child-BEAR-Net); www.ersnet.org/science-and-research/clinical-research-collaboration-application-programme/child-bear-net/) will progress research and collaboration in paediatric bronchiectasis and advance this neglected field.

Self-evaluation questions

1. Regarding the radiographic diagnosis of bronchiectasis in children/adolescents, which of the following is false?
   a) The diagnosis of bronchiectasis requires radiographic evidence in addition to presence of a clinical syndrome
   b) A chest CT scan is required to diagnose bronchiectasis as chest radiographic findings are insensitive
   c) Multidetector CT chest scans with HRCT reconstructs are the current gold standard for diagnosing bronchiectasis
   d) The adult criterion of BAR >1–1.5 should be used to diagnose bronchiectasis in children/adolescents
2. In addition to a chest CT scan, what other tests should be undertaken in all children/adolescents with bronchiectasis (*i.e.* minimal panel of tests)? Which of the following is false?
   a) Standard tests are required as they may influence treatment
   b) All children should have a full blood examination, immunological tests (total IgG, IgA, IgM, IgE, specific antibodies to vaccine antigens) and a sweat test
   c) Investigations for primary ciliary dyskinesia are also required in all patients
   d) Lower airway bacteriology and lung function testing (when age appropriate) should be undertaken in all patients
3. Regarding radiographic bronchiectasis reversibility in children/adolescents, which of the following is true?
   a) Yes, reversibility is always possible
   b) No, reversibility is never possible
   c) Reversibility is sometimes possible if bronchiectasis is diagnosed early and optimally treated
   d) Reversibility is possible only if there is no underlying cause
4. Regarding ACT in children/adolescents with bronchiectasis, which of the following is true?
   a) ACT should be used every single day, irrespective of severity and/or condition of bronchiectasis
   b) The types of ACT for children/adolescents usually change as they mature and are best taught by an expert chest physiotherapist
   c) During an exacerbation, ACT should remain the same
   d) Not all children/adolescents with bronchiectasis require ACT education
5. Regarding the new isolation of *P. aeruginosa*, which of the following is true?
   a) If the child/adolescent does not have CF, treatment is unnecessary
   b) *P. aeruginosa* cannot be eradicated in patients with bronchiectasis
   c) *P. aeruginosa* is confined to children/adolescents with mild bronchiectasis
   d) Eradication of *P. aeruginosa* should be undertaken once the infection is confirmed

Key points

- Bronchiectasis in children and adolescents is defined as a clinical syndrome of recurrent or persistent wet/productive cough, lower airway infection and/or inflammation, and abnormal bronchial dilatation on chest CT scans.
- In 2021, the ERS Task Force published a clinical practice guideline for the management of children and adolescents with bronchiectasis, using the ERS model and GRADE methods.
- 14 questions were posed, leading to 26 recommendations that addressed the diagnosis, evaluation of causes, definition of exacerbations, management, monitoring, and reversibility and prevention of paediatric bronchiectasis.
- ELF undertook an international survey on the clinical needs and research priorities of parents of children with bronchiectasis and adults who had bronchiectasis as a child, which informed the guideline.

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Conflict of interest

A.B. Chang reports grants from the National Health and Medical Research Council (NHMRC), Australia, for various project grants and a Centre of Research Excellence relating to various aspects of bronchiectasis in children, as well as an NHMRC Practitioner fellowship (1058213), during the conduct of the study; and fees paid to her institution from GSK (Independent Data Monitoring Committee (IDMC) member for an unlicensed vaccine), Merck (advisory member of study design for an unlicensed molecule for chronic cough), and Moderna (IDMC member for a COVID-19 vaccine for children), outside the submitted work. K. Grimwood reports grants from the National Health and Medical Research Council (NHMRC), Australia, for various project grants and a Centre of Research Excellence relating to various aspects of bronchiectasis in children, during the conduct of the study. J. Boyd is an employee of the European Lung Foundation. R. Fortescue has nothing to disclose. Z. Powell has nothing to disclose. A. Kantar has nothing to disclose.

Suggested answers

1. d.
2. c.
3. c.
4. b.
5. d.

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