



ISSN 2040-2023

July 2010

BTS Guideline for non-CF Bronchiectasis

A Quick Reference Guide

British Thoracic Society
www.brit-thoracic.org.uk

BTS GUIDELINE FOR NON-CF BRONCHIECTASIS – A QUICK REFERENCE GUIDE

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This quick reference guide should be read alongside the main guideline, and section numbers refer to the text of the main guideline.
Note that the references made in this document relate to the references available in the full guideline only.

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The full BTS Guideline is published in Thorax Vol 65 Supplement 1

Available online at: <http://www.brit-thoracic.org.uk/clinical-information/bronchiectasis.aspx>



SECTION 1: INTRODUCTION**Reason for BTS bronchiectasis guideline**

The diagnosis, investigation and particularly management of bronchiectasis has been largely empirical and the subject of relatively few controlled clinical trials. There are no clear guidelines, although an Australian position statement has been published concerning bronchiectasis in children.

The purposes of these guidelines were therefore threefold: (1) to identify relevant studies in non-cystic fibrosis (CF) bronchiectasis; (2) to provide guidelines on management based on published studies where possible or a consensus view; and (3) to identify gaps in our knowledge and identify areas for future study.

Guideline group members

The membership of the BTS Bronchiectasis (non-CF) Guideline Group is as follows: Dr Mark C Pasteur, Dr Diana Bilton, Dr Adam T Hill, Professor Andrew Bush, Dr Charles Cornford, Dr Steven Cunningham, Dr Xavier Emmanuel, Jane French, Dr Mike Greenstone, Professor David M Hansell, Alex Harvey, Dr Richard Herriot, Karen Heslop, Dr Pota Kalima, Frances Sinfield, Dr Samantha Sonnappa, Dr David A Spencer, Professor Robert A Stockley, Lorna Willcox, Dr Robert Wilson, Mr G Wyn Parry. The assistance of Julia Bott, Jennifer Pryor and Dr Colin Wallis is gratefully acknowledged.

How has the guideline been designed?

The guideline is divided into sections covering different aspects of the management of the condition. Guidance for children and adults is dovetailed together throughout to avoid repetition while acknowledging differences between these groups. Areas of particular or sole relevance to one or other of these groups are indicated. Sections 2 and 3 cover the background, clinical assessment and investigation of patients (including appropriate radiological and laboratory investigations). The principles and broad approach to management are discussed in Section 4 including recommendations for physiotherapy and non-antibiotic drug treatment. The use of antibiotics is covered in Section 5 and surgery and the management of advanced disease is covered in Section 6.

Definition

This guideline refers to the investigation and management of patients with symptoms of persistent or recurrent bronchial sepsis related to irreversibly damaged and dilated bronchi, namely, clinical bronchiectasis. It does not cover the management of cystic fibrosis (CF) and, for the purposes of the guideline, 'bronchiectasis' is synonymous with the term 'non-CF bronchiectasis'. Likewise, it does not focus on traction bronchiectasis secondary to other lung pathologies, particularly the interstitial lung diseases, which is commonly asymptomatic.

Methods

A literature search was performed using the following databases: Pubmed, Cinahl, Embase and AMED using the combined search terms 'bronchiectasis' and 'not CF' which resulted in 1803 references published in English. The abstract for each reference was retrieved and reviewed by each of the three members of the steering committee. The steering committee then decided, on the basis of the abstract, whether the full paper should be reviewed. Papers were assigned to one or more of the three working groups based on their relevance to each section and read by each member of the group. Those papers with information relevant to the guideline were included in the final document. The methods

for assessing the level of evidence for each paper and the grading of recommendations were those developed by the Scottish Intercollegiate Guidelines Network (SIGN) and as used in the British Thoracic Society (BTS)/SIGN British guideline on the management of asthma (see table 1). The evidence level for each paper is given at the end of each text section and the grade of recommendation follows each recommendation statement. This guideline is due for revision in 2011.

How common is bronchiectasis in adults and children in the 21st century?

The incidence varies widely between populations from 3.7/100 000 children in New Zealand to 52/100 000 adults in the USA. In the UK there are no recent studies, although mass chest x-ray features of bronchiectasis in the 1950s suggested a prevalence of 100/100 000. The prevalence increases with age. Some of these population studies, many of which were conducted years ago, have not included modern diagnostic techniques and specifically high-resolution CT scanning (HRCT). The importance of this issue is that the true incidence remains unknown in most populations.

What are the pathology and underlying causes?

Bronchiectasis is a persistent or progressive condition characterised by dilated thick-walled bronchi. The symptoms vary from intermittent episodes of expectoration and infection localised to the region of the lung that is affected to persistent daily expectoration often of large volumes of purulent sputum. Bronchiectasis may be associated with other non-specific respiratory symptoms including dyspnoea, chest pain and haemoptysis, and may progress to respiratory failure and cor pulmonale.

The underlying pathological process is damage to the airways which results from an event or series of events where inflammation is central to the process. This is easy to understand as part of the 'vicious circle' hypothesis which has been applied to bronchiectasis and has been the major factor influencing current disease management.

The lung is continuously exposed to inhaled pathogens and (in many countries) environmental pollutants. The lung has a sophisticated primary and secondary defence system that maintains sterility of the normal lung. If this defence system is breached as in disorders of mucociliary clearance or specific antibody deficiencies, the lung becomes susceptible to infection, colonisation (the persistence of bacteria in the lower respiratory tract) occurs and the subsequent inflammation that causes airway damage further impairing host defences. Thus, once established, the defective defences can lead to a self-perpetuating cycle of events that facilitate bacterial colonisation and airway sterility becomes unlikely.

Primary defects in the lung defences are uncommon in patients investigated as adults, suggesting they are either subtle or do not influence the primary event. However, immunodeficiency may be more common when bronchiectasis presents in childhood.²³² Episodes causing clear lung damage such as previous pneumonias, gastric aspiration or viral illnesses in childhood would represent such initiating events, although recent evidence suggests these may be less common. The damage to the airway by such episodes would particularly impair the normal mucociliary function and hence clearance of inhaled pathogens initiating the inflammatory cycle.

However, despite many studies over the years using modern immunological techniques, not only have few primary deficiencies of host defences been found but up to 40% of patients do not even have a clear defining event that appears to initiate the process.

What is the outlook for these patients?

Most of the information on long term outcome is from historical data and

suggests that antibiotic therapy has had an effect. For instance, in the 1940s most patients diagnosed with bronchiectasis died before the age of 40 years but, by the 1960s, the average age of death had risen to 55 years. Nevertheless, this still indicates a significant reduction in life expectancy in patients with bronchiectasis. More recent data suggest a better prognosis,³⁷³ although it is recognised that the general health of patients with bronchiectasis can be poor²⁵¹ and certain subsets (particularly those colonised with *Pseudomonas aeruginosa*) are particularly affected, with continued ill health and progressive deterioration.

SUMMARY OF RECOMMENDATIONS

SECTION 2: BACKGROUND AND CAUSES

Congenital defects of large airways

- ▶ Congenital defects should be considered in all patients with bronchiectasis. [D]

Foreign bodies and aspiration

- ▶ Gastric aspiration should be considered as a cause in all patients. [D]

What is the current relevance of previous severe lower respiratory tract infections to patients with bronchiectasis?

- ▶ A history of previous severe lower respiratory tract infections due to bacterial and viral pneumonia, pertussis or tuberculosis should be sought in all patients with bronchiectasis. [C]
- ▶ Where possible, the temporal relationship of identified infections to the onset of chronic respiratory symptoms should be determined. [D]

Mycobacterium tuberculosis and opportunist mycobacteria

- ▶ All patients with repeated isolates of opportunist mycobacteria should have regular follow-up in secondary care. [D]

Immune deficiency and bronchiectasis

- ▶ The possibility of underlying immune deficiency, particularly antibody deficiency, should be considered in all children and adults with bronchiectasis. [A]
- ▶ Serious, persistent or recurrent infections, particularly involving multiple sites, or infections with opportunist organisms should raise the suspicion of immune deficiency. [D]
- ▶ The possibility of symptomatic or clinically silent bronchiectasis should be considered as a potential complication in all patients with immune deficiency, particularly primary antibody deficiency. [D]
- ▶ In patients with immune deficiency and patients with bronchiectasis, features in the history or clinical examination which may support the coexistence of both conditions should be considered and adequately assessed. [D]
- ▶ In patients with suspected or proven immune deficiency and bronchiectasis in combination, specialist aspects of diagnosis, monitoring and management should optimally be provided within a shared specialist care arrangement (joint working between chest physician and immunologist). [D]

What is the relationship of other airway diseases to bronchiectasis? What are the features of allergic bronchopulmonary aspergillosis (ABPA) as a cause of bronchiectasis?

- ▶ All patients with bronchiectasis should be assessed for evidence of

ABPA, which is a clinical diagnosis based on presentation and immunological tests (*Aspergillus*-specific IgE and IgG). [D]

Is asthma a cause of bronchiectasis?

- ▶ In adults, asthma should be considered as the cause of bronchiectasis if no other cause is identified. [D]

Primary bronchiolar disorders

- ▶ The possibility of diffuse panbronchiolitis should be considered in patients of Far Eastern ethnic origin. [D]

What is the relationship of bronchiectasis to cystic fibrosis (CF)?

- ▶ For all patients with bronchiectasis, the possibility of underlying CF should be considered (see Section 3). [D]

Which connective tissue disorders are associated with bronchiectasis?

- ▶ A history of rheumatoid arthritis should be sought in all patients with bronchiectasis. [D]
- ▶ Closer follow-up of patients with rheumatoid arthritis-related bronchiectasis is warranted in view of a poorer prognosis. [C]

Inflammatory bowel diseases

- ▶ Bronchiectasis should be considered in patients with inflammatory bowel disease who develop a chronic productive cough. [D]

Disorders of ciliary function

- ▶ In all children with bronchiectasis, a detailed history of the neonatal period should be taken. [D]
- ▶ In children and adults with bronchiectasis, a history of chronic upper respiratory tract problems, particularly otitis media, should be sought. [D]
- ▶ Adults should be questioned about any history of infertility. [D]

Is α_1 -antitrypsin deficiency a cause of bronchiectasis?

- ▶ Routine screening for α_1 -antitrypsin deficiency is not required unless the radiological investigations suggest basal emphysema. [D]

Yellow nail syndrome

- ▶ The assessment of patients with bronchiectasis should include a search for features of yellow nail syndrome. [D]

The upper respiratory tract in bronchiectasis patients

- ▶ Every patient with bronchiectasis should have an assessment of upper respiratory tract symptoms. [D]

SECTION 3: CLINICAL ASSESSMENT AND INVESTIGATIONS

Who to investigate for bronchiectasis

Which children should be investigated for bronchiectasis?

Consideration should be given to evaluating a child for bronchiectasis who presents with: [D]

- ▶ Chronic moist/productive cough, especially between viral colds or with positive bacterial cultures.
- ▶ Asthma that does not respond to treatment.
- ▶ A single positive sputum culture, in the setting of chronic respiratory symptoms, for *Staphylococcus aureus*, *Haemophilus influenzae*, *Pseudomonas aeruginosa*, non-tuberculous mycobacteria or *Burkholderia cepacia* complex.

- ▶ An episode of severe pneumonia, particularly if there is incomplete resolution of symptoms, physical signs or radiological changes.
- ▶ Pertussis-like illness failing to resolve after 6 months.
- ▶ Recurrent pneumonia.
- ▶ Persistent and unexplained physical signs or chest radiographic abnormalities.
- ▶ Localised chronic bronchial obstruction.
- ▶ Respiratory symptoms in children with structural or functional disorders of the oesophagus and upper respiratory tract.
- ▶ Unexplained haemoptysis.
- ▶ Respiratory symptoms with any clinical features of CF, PCD or immunodeficiency.

Which adults should be investigated for bronchiectasis?

Bronchiectasis should be considered in all adults who have: [D]

- ▶ Persistent productive cough. Factors favouring further investigation are any one of the following :
 - young age at presentation;
 - history of symptoms over many years;
 - absence of smoking history;
 - daily expectoration of large volumes of very purulent sputum;
 - haemoptysis;
 - sputum colonisation with *P aeruginosa*.
- ▶ Unexplained haemoptysis or non-productive cough.
- ▶ Patients thought to have COPD may have bronchiectasis alone or in addition and referral for investigation is appropriate if:
 - management is not straightforward;
 - there is slow recovery from lower respiratory tract infections;
 - recurrent exacerbations;
 - there is no history of smoking.

Clinical presentation of bronchiectasis

What are the symptoms and signs of bronchiectasis in children?

- ▶ Respiratory symptoms, particularly cough and sputum production, should be assessed and recorded in all children with bronchiectasis. [D]
- ▶ There should be a high index of suspicion for diagnosing bronchiectasis in children with chronic respiratory symptoms. [D]
- ▶ The finding of persistent lung crackles on auscultation should alert the clinician to possible underlying bronchiectasis. [D]

What symptoms and signs should be assessed in an adult with bronchiectasis?

- ▶ Assessment of symptoms in patients with bronchiectasis should include a record of both sputum purulence and estimated or measured 24 h sputum volume when clinically stable. [D]
- ▶ The number of infective exacerbations per annum should be noted including frequency and nature of antibiotic usage. [D]

Investigations directed at underlying cause

Why should the underlying cause of bronchiectasis be established?

- ▶ Investigations should be performed to establish cause and severity of disease. [D]

What blood tests should be performed?

The following should be measured in all patients:

- ▶ serum immunoglobulins (IgG, IgA, IgM) and serum electrophoresis; [A]
- ▶ serum IgE, skin prick testing or serum IgE testing to *Aspergillus fumigatus* and *Aspergillus precipitins*. [C]

What immunological tests should be done on all patients?

- ▶ All patients with bronchiectasis should be screened at presentation for gross antibody deficiency by routine measurement of serum IgG, IgA and IgM levels and serum electrophoresis. [A]
- ▶ Respiratory and immunology units should develop additional local protocols for screening assessment of humoral responses to specific antigens; such screening may be universal (applied to all cases of bronchiectasis) or targeted (directed only at higher risk cases in whom common underlying causes of bronchiectasis have been excluded or who have other features of potential antibody deficiency) according to local preference or circumstances and should comprise [D]:
 - ▶ measurement of baseline specific antibody levels against tetanus toxoid and the capsular polysaccharides of both *S pneumoniae* and *H influenzae* type b (or suitable alternative peptide and polysaccharide antigens);
 - ▶ immunisation with appropriate vaccines followed by re-assay of individual specific antibody responses after 21 days where screening baseline levels are low.
- ▶ Where screening tests or clinical presentation indicate that further immunological investigation is warranted, this should be planned and undertaken within an agreed and integrated respiratory/immunology protocol. [D]

What are the second-line immunological investigations and when should they be performed?

Consideration of second-line assessment of immune competence is necessary in the following circumstances:

- ▶ Antibody screening investigations have demonstrated the presence of an antibody deficiency disorder (to refine diagnosis, detect immune complications and plan treatment). [D]
- ▶ In the presence of normal antibody screening test results where the following are present: [D]
 - clinical suspicion of immune deficiency (short stature, facial abnormality, cardiac lesions, hypocalcaemia, cleft palate, oculocutaneous telangiectasis, eczema, dermatitis, petechiae, manifestations of endocrinopathy, unexplained failure to thrive, enlargement of absence of lymphoid tissues, unexplained organomegaly, unexplained joint symptoms);
 - a family history of known or suspected immune deficiency;
 - infections which are serious, involving a threat to life, tissue destruction or which require/have required surgical intervention (eg, lobectomy, tonsillectomy, insertion of grommets, incision of boils), are persistent or recurrent despite multiple or prolonged courses of antibiotics, involve unusual/opportunistic microorganisms or involve multiple sites (eg, sinuses or middle ear in addition to the bronchial tree).

When should patients have gastrointestinal investigations?

- ▶ There should be a low threshold for gastrointestinal investigations in children. [D]
- ▶ Gastric aspiration should be considered in patients following lung transplantation. [D]

When should patients have investigations to exclude CF?

- ▶ All children and all adults up to the age of 40 presenting with bronchiectasis should have investigations for CF. [D]
- ▶ In adults, investigations should also be considered in those with: [D]
 - age at presentation >40 years and no other identified cause;
 - persistent isolation of *S aureus* in the sputum;
 - features of malabsorption;

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- male primary infertility;
 - upper lobe bronchiectasis;
 - a history of childhood steatorrhea.
- ▶ Screening investigations should include both: [D]
- two measurements of sweat chloride;
 - CFTR genetic mutation analysis.

When should patients have tests of ciliary function? What are the best tests to identify ciliary defects?

- ▶ Ciliary investigations should be considered in children with bronchiectasis when there is: [D]
- no other cause for bronchiectasis identified;
 - a history of continuous rhinitis since the neonatal period;
 - a history of neonatal respiratory distress;
 - dextrocardia.
- ▶ Ciliary investigations should be considered in adults only if there is a history of chronic upper respiratory tract problems or otitis media. Factors favouring investigation include: [D]
- problems since childhood;
 - childhood chronic otitis media;
 - predominantly middle lobe bronchiectasis;
 - infertility or dextrocardia.
- ▶ For adults, the saccharin test and/or exhaled nasal NO may be used to screen out those not requiring detailed ciliary function tests. [D]

What are the indications for bronchoscopy?

- ▶ In children, bronchoscopy is indicated when bronchiectasis affects a single lobe to exclude a foreign body. In some acutely ill patients it may achieve a useful microbiological result. [D]
- ▶ In adults with localised disease, bronchoscopy may be indicated to exclude proximal obstruction. [D]
- ▶ In adults, bronchoscopy and bronchoscopic sampling of the lower respiratory tract does not have a place in the routine investigation of patients with bronchiectasis. [D]
- ▶ For patients in whom serial testing of sputum does not yield microbiological information and who are not responding well to treatment, bronchoscopic sampling of lower respiratory tract secretions may be indicated. [D]
- ▶ Bronchoscopy is indicated if HRCT suggests atypical mycobacterial infection and sputum culture is negative. [D]
- ▶ Cytological examination of bronchoscopic specimens can provide evidence supporting gastric aspiration. [D]

Radiological investigations

What is the role of a chest x-ray?

- ▶ A baseline chest x-ray should be done in all patients. [D]
- ▶ Repeat chest x-rays need only be done if clinically indicated. [D]

What is the role of HRCT?

- ▶ HRCT is the radiological investigation of choice to establish the diagnosis of bronchiectasis. [D]

What is an optimum HRCT protocol for defining bronchiectasis?

- ▶ Standard HRCT protocol, single detector CT scanner: [D]
- patient position: supine, breath holding at full inspiration; optional ECG gating 120–140 kV; 100–180 mAs (dependent on patient habitus); acquisition time <1 s;
 - beam collimation 1 mm; 1 cm intervals;
 - reconstruction with ‘very sharp’ kernel.

- ▶ Volumetric HRCT protocol, 64-channel CT scanner: [D]
- patient position: supine, breath holding at full inspiration; 120–140 kV; 120 effective mAs; rotation time 0.5 s;
 - detector collimation 0.6 mm; section thickness 1 mm; pitch 0.9;
 - reconstruction with ‘very or ultra sharp’ kernel.

What are the HRCT features of bronchiectasis?

- ▶ Bronchial wall dilation (internal lumen diameter greater than accompanying pulmonary artery or lack of tapering) is the characteristic feature of bronchiectasis. [D]
- ▶ Bronchial wall thickening is often also present although harder to define. [D]

Can HRCT identify features of specific causes?

- ▶ HRCT features may be suggestive of certain underlying conditions but require correlation with clinical and laboratory assessments. [D]
- ▶ HRCT images should be examined for features suggesting ABPA, CF, immobile cilia, opportunist mycobacteria and tracheobronchomegaly. [D]

How are HRCT changes related to lung function?

- ▶ The severity of bronchiectasis on HRCT correlates with measures of airflow obstruction. [D]

How often should radiological investigations be repeated?

- ▶ Routine repeat chest x-ray or HRCT is not necessary; repeat imaging should be considered when there is clinical need. [D]
- ▶ In cases of humoral immune deficiency, repeat HRCT at intervals may be necessary to detect asymptomatic progression. This should be discussed with the patient’s clinical immunologist. [D]

What scoring systems should be used for research?

- ▶ Scoring systems based on studies of patients with cystic fibrosis are the best currently available and should be used until disease-specific scoring systems are available. [D]

Sputum microbiology

Which organisms are isolated from the lower respiratory tract in bronchiectasis?

- ▶ All children and adults with bronchiectasis should have an assessment of lower respiratory tract microbiology. [D]
- ▶ Persistent isolation of *S aureus* (and/or *P aeruginosa* in children) should lead to consideration of underlying ABPA or cystic fibrosis. [D]

How and when should standard microbiology be performed?

At what interval should it be repeated?

- ▶ Respiratory tract specimens should be obtained in all patients with bronchiectasis. [D]
- ▶ To maximise the chances of isolating *H influenzae* and *S pneumoniae*, specimens should reach the microbiology laboratory within 3 h. [D]

Lung function tests

Which lung function tests should be performed in children?

- ▶ In all children who are old enough (usually aged >5 years) forced expiratory volume in 1 s (FEV₁), forced vital capacity (FVC) and forced expiratory flow (FEF_{25–75}) should be measured at initial assessment. [D]

Which lung function tests should be performed in adults?

- ▶ All adults with bronchiectasis should have measures of FEV₁, FVC and PEF. [D]
- ▶ Repeat assessment of FEV₁, FVC and PEF should be made at least annually in those patients attending secondary care. [D]
- ▶ Patients with immune deficiency or PCD should have measurements of FEV₁, FVC at least four times each year. [D]
- ▶ Measurement of lung volumes and gas transfer coefficient may help in the identification of other causes of airflow obstruction such as COPD/emphysema. [D]
- ▶ Reversibility testing may identify improvement in lung function after bronchodilators and should always be considered if airflow obstruction is identified, especially in young people. [D]

Is there a role for exercise testing in bronchiectasis?

- ▶ Exercise tests have a role in investigating children in whom symptoms are out of keeping with lung function or HRCT measurements. [D]
- ▶ In adults, exercise testing should be part of a pulmonary rehabilitation programme. [D]

Can lung function tests be used to assess response to antibiotic treatment?

- ▶ Routine measurement of lung function is not necessary in the assessment of response to short-term antibiotic therapy but, if performed, may offer objective evidence of improvement. [D]
- ▶ FEV₁ and FVC should be measured before and after intravenous antibiotic therapy as this may give objective evidence of improvement. [D]
- ▶ Spirometry and lung volumes should be measured in all patients before and after commencing long-term oral or nebulised antibiotic therapy. [D]

SECTION 4: MANAGEMENT – PRINCIPLES AND GENERAL APPROACH

General approach and treatment of the specific underlying cause

- ▶ Identify and treat underlying cause to prevent disease progression. [D]
- ▶ Maintain or improve pulmonary function. [D]
- ▶ Reduce exacerbations. [D]
- ▶ Improve quality of life by reducing daily symptoms and exacerbations. [D]
- ▶ In children, achieve normal growth and development. [D]
- ▶ Patients with primary or secondary immune deficiency should be under joint care with a clinical immunologist. [D]
- ▶ Patients with CF should be referred to a CF specialist centre. [D]

Role of primary care

What is the interface between primary and secondary care?

Patients who should have regular follow-up in secondary care include: [D unless stated]

- ▶ all children with bronchiectasis;
- ▶ patients with chronic *P aeruginosa*, opportunist mycobacteria or methicillin-resistant *S aureus* colonisation;
- ▶ deteriorating bronchiectasis with declining lung function;
- ▶ recurrent exacerbations (≥3 per year);
- ▶ patients receiving prophylactic antibiotic therapy (oral or nebulised);
- ▶ patients with bronchiectasis and associated rheumatoid arthritis, [C] immune deficiency inflammatory bowel disease and PCD;

- ▶ patients with ABPA;
- ▶ patients with advanced disease and those considering transplantation.

Role of nurses

What role do nurses play in the management of bronchiectasis?

- ▶ Primary and secondary care nurses should receive training in the management of bronchiectasis. [B]

Physiotherapy: airway clearance techniques and exercise

Which airway clearance technique(s) should be taught?

- ▶ Patients should be made aware of the airway clearance techniques available. [D]
- ▶ HRCT images should be reviewed to complement the physiotherapy assessment and assist planning appropriate clearance techniques. [D]
- ▶ Patients should, where possible, be encouraged to be independent with their chosen airway clearance technique. [D]
- ▶ Patient preference and adherence to treatment must be taken into account. [D]
- ▶ The active cycle of breathing techniques (plus postural drainage) and oscillating positive expiratory devices (plus postural drainage and the forced expiration technique) should be considered when offering individuals with non-CF bronchiectasis effective airway clearance techniques. [A]
- ▶ The inclusion of postural drainage should be considered for all airway clearance techniques. [B]
- ▶ The inclusion of the forced expiration technique should be considered for all airway clearance techniques. [B]
- ▶ Autogenic drainage and PEP may be offered to patients as an alternative airway clearance technique in non-CF bronchiectasis if other techniques are not effective or acceptable to the patient. [D]
- ▶ Where postural drainage is essential for clearing secretion in a breathless patient, consider offsetting the increased load by the use of non-invasive ventilatory support such as NIV or intermittent positive pressure breathing. [D]
- ▶ Modified gravity-assisted positions (no head-down tilt) should be offered where the conventional tipped position is contraindicated or unacceptable to the patient. [D]
- ▶ During an acute exacerbation or when the patient is more fatigued than usual, manual techniques may be offered as a part of an airway clearance technique regimen. [D]

Are adjuncts to airway clearance techniques useful?

- ▶ Sterile water inhalation may be used before airway clearance to facilitate clearance. [B]
- ▶ The use of nebulised normal saline prior to airway clearance could be considered to increase sputum yield, reduce sputum viscosity and improve ease of expectoration. [B]
- ▶ The use of nebulised hypertonic saline prior to airway clearance could be considered to increase sputum yield, reduce sputum viscosity and improve ease of expectoration. [B]
- ▶ When nebulised hypertonic saline is first administered, FEV₁ or PEF readings should be done before and 5 min after treatment to assess for possible bronchoconstriction. [D]
- ▶ When nebulising hypertonic saline, pretreat with a bronchodilator in those with bronchial hyper-reactivity. [D]
- ▶ Consider using nebulised β₂ agonists prior to treatment to enhance sputum clearance. [B]
- ▶ NIV/intermittent positive pressure breathing may be used to augment

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tidal volume and reduce the work of breathing in those patients who are becoming fatigued and finding their standard airway clearance difficult. [D]

How soon should the patient be reviewed after the initial assessment?

- ▶ Effectiveness and acceptability to the patient of the airway clearance technique should be reviewed within approximately 3 months of the initial visit. [D]

What is the role of exercise?

- ▶ Pulmonary rehabilitation should be offered to individuals who have breathlessness affecting their activities of daily living. [B]
- ▶ Inspiratory muscle training can be used in conjunction with conventional pulmonary rehabilitation to enhance the maintenance of the training effect. [B]

Airway pharmacotherapy

Are mucolytics and hyperosmolar agents of benefit in the long term to patients with bronchiectasis?

- ▶ Recombinant human DNase should not be used in adults with bronchiectasis. [A]
- ▶ Recombinant human DNase should not be used in children with bronchiectasis. [D]

Are bronchodilators of use in bronchiectasis?

- ▶ It seems appropriate to assess patients with airflow obstruction for reversibility to β_2 agonist and anticholinergic bronchodilators and to institute therapy where lung function or symptoms improve on therapy. [D]
- ▶ Methylxanthines have no routine role in bronchiectasis. [D]

Are inhaled corticosteroids a useful treatment for bronchiectasis?

- ▶ Inhaled steroids should not be used routinely in children with bronchiectasis (outside of use for those patients with additional asthma). [D]
- ▶ In adults, current evidence does not support routine use of inhaled corticosteroids in bronchiectasis (outside of use for those patients with additional asthma). [B]

Leukotriene receptor antagonists and other anti-inflammatory agents

- ▶ There is no evidence for a role for leukotriene receptor antagonists or other anti-inflammatory drugs in bronchiectasis. [D]

SECTION 5: MANAGEMENT: ANTIBIOTIC THERAPY

Defining and managing exacerbations

Which antibiotic regimen is recommended for exacerbations in adults?

- ▶ Before starting antibiotics, a sputum sample should be sent off for culture. [D]
- ▶ Empirical antibiotics should be started while awaiting sputum microbiology. [D]
- ▶ If there is no previous bacteriology, first-line treatment is amoxicillin 500 mg three times a day [B] or clarithromycin 500 mg twice daily (in patients who are penicillin-allergic) for 14 days. [C]
- ▶ High-dose oral regimens (eg, amoxicillin 1 g three times a day or amoxicillin 3 g twice daily) may be needed in patients with severe bronchiectasis chronically colonised with *Haemophilus influenzae*. [B]
- ▶ Ciprofloxacin should be used in patients colonised with *Pseudomonas aeruginosa* with cautious use in the elderly. [B]

- ▶ Previous sputum bacteriology results can be useful in deciding which antibiotic to use. Table A1 highlights the recommended first-line and alternative treatments for the common bacterial pathogens implicated in exacerbations of bronchiectasis. [C]
- ▶ Antibiotics can be modified subsequently once the pathogen is isolated only if there is no clinical improvement and the treatment should then be guided by antibiotic sensitivity results. [D]
- ▶ Failure to respond to an antibiotic course should prompt a repeat sputum culture. [D]
- ▶ Intravenous antibiotics should be considered when patients are particularly unwell, have resistant organisms or have failed to respond to oral therapy (this is most likely to apply to patients with *Pseudomonas aeruginosa*). [C]
- ▶ There is no evidence to support the routine use of antiviral drugs in exacerbations. [D]

When are combination (dual) antibiotic regimes required?

Adults

- ▶ Combination antibiotics are not required in patients colonised with *Haemophilus influenzae*, *Moraxella catarrhalis*, *Staphylococcus aureus* (methicillin-sensitive) and *Streptococcus pneumoniae*. [D]
- ▶ If there is more than one pathogen, select an antibiotic that will cover both pathogens. If this is not feasible due to resistance patterns, combination antibiotics may be required. [D]
- ▶ In patients who culture *Pseudomonas aeruginosa* that is sensitive to ciprofloxacin, monotherapy with oral ciprofloxacin can be used as first-line treatment (table A1). [B]
- ▶ In patients who have not responded to oral ciprofloxacin, monotherapy with an antipseudomonal intravenous antibiotic should be considered (table A1). [D]
- ▶ Combination antibiotics should be used for infections due to strains of *Pseudomonas aeruginosa* that are resistant to one or more antipseudomonal antibiotics (including ciprofloxacin) or if the clinician suspects the patient will require many subsequent antibiotic courses to reduce the development of drug resistance. [D]
- ▶ MRSA should be treated with two oral antibiotics or a single intravenous agent (see table A1). [D]
- ▶ Intravenous aminoglycosides should only be used with appropriate and robust dosing and monitoring systems in place that have been agreed with local microbiologists and pharmacists (Appendix 1). [D]

Children

- ▶ In children who culture *Pseudomonas aeruginosa* that is sensitive to ciprofloxacin, monotherapy with oral ciprofloxacin can be used (table A1). [B]
- ▶ Those children whose sputum cultures yield pathogens with multiple resistant patterns should be considered for combination antibiotic therapy (in particular for *Pseudomonas aeruginosa*) (table A1). [D]
- ▶ Identification of MRSA infection should prompt a dedicated eradication programme that in children may include a course of intravenous antibiotics, should oral antibiotics be unsuccessful (table A1). [D]

Do long-term oral antibiotics influence long-term outcome in adults?

- ▶ Patients having ≥ 3 exacerbations per year requiring antibiotic therapy or patients with fewer exacerbations that are causing significant morbidity should be considered for long-term antibiotics. [C]
- ▶ In the first instance, high doses should not be used to minimise side effects. [C]
- ▶ The antibiotic regimen should be determined by sputum microbiology

when clinically stable (table All). [D]

- ▶ Long-term quinolones should not be used until further studies are available. [C]
- ▶ Macrolides may have disease-modifying activity and preliminary data suggest the need for a large randomised controlled trial. [C]

Do long-term nebulised antibiotics influence long-term outcome in adults?

- ▶ Patients having ≥ 3 exacerbations per year requiring antibiotic therapy or patients with fewer exacerbations that are causing significant morbidity should be considered for long-term nebulised antibiotics. [C]
- ▶ In such patients, long-term nebulised antibiotics should be considered if chronically colonised with *Pseudomonas aeruginosa* (table All). The choice of antibiotic should be guided by the antibiotic sensitivity results. Further studies are needed to address the optimal antibiotic choice and doses required. [C]

Antibiotic resistance

What is the impact of long-term antibiotics on antibiotic resistance in adults?

- ▶ Long-term antibiotics may result in antibiotic resistance in individual patients and alternative antibiotics should be chosen depending on sensitivity results. [D]
- ▶ Long-term ciprofloxacin should not be used. [D]

Is there clinical relevance of in vitro antibiotic resistance patterns in adults and children?

- ▶ Treatment should be guided by antibiotic sensitivity results but is often empirical based on previous sputum bacteriology. [D]
- ▶ Some patients may respond to antibiotic treatment despite resistance to that drug in vitro. Antibiotics should only be changed if there is no clinical response. [D]

SECTION 6: SURGERY, COMPLICATIONS OF BRONCHIECTASIS AND MANAGEMENT OF ADVANCED DISEASE

Surgery for bronchiectasis

Is there a role for surgery in the management of patients with bronchiectasis?

- ▶ Lung resection surgery may be considered in patients with localised disease in whom symptoms are not controlled by medical treatment. [D]
- ▶ Patients undergoing surgery should have a review by a chest physician before referral. [D]

Massive haemoptysis

- ▶ Bronchial artery embolisation and/or surgery is first-line therapy for the management of massive haemoptysis. [D]

Non-invasive ventilation

- ▶ Non-invasive ventilation can improve quality of life in some patients with chronic respiratory failure due to bronchiectasis. [D]
- ▶ Evidence for survival benefit is lacking, although some patients are successfully treated with non-invasive ventilation for significant lengths of time which may reduce hospitalisations. [D]

This Quick Reference Guide contains the synopsis of recommendations taken from the main Guideline document. For the purposes of consistency, the numbering of Sections, Tables and Figures in this Guide is taken from the main document. Details of the methodology and grading system are also given in the full document. Readers are advised to refer to the main Guidelines document for full details:

<http://www.brit-thoracic.org.uk/clinical-information/bronchiectasis.aspx>



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British Thoracic Society Reports, Vol 2, No 2, 2010
ISSN 2040-2023

www.brit-thoracic.org.uk