Management in primary care

The overall aim of therapy is to:
- improve symptoms
- prevent complications
- prevent decline in lung function

To support the above:
- identify and treat underlying cause to prevent disease progression
- maintain or improve pulmonary function
- reduce exacerbations
- implement any management advice from clinical immunologist if patient with primary or secondary immune deficiency
- use respiratory email advice line to support management plan – mailto:respconsultants.enh-tr@nhs.net
RED FLAGS for admission

Arrange hospital admission for adults who:

- are cyanosed or acutely confused
- have a respiratory rate of > 25 breaths per minute
- have signs of cardiopulmonary failure, e.g.:
  - marked breathlessness
  - rapid respiration
  - laboured breathing
  - worsening peripheral oedema
  - oxygen saturation < 93% on room air
- have a temperature of ≥ 38°C
- are unable to take or have failed to respond to oral therapy
- have pleuritic pain severe enough to inhibit coughing and the clearing of secretions

Where IV treatment can be provided at home some patients may remain at home with secondary care follow-up. Discuss with community respiratory nurse and acute consultant. Patients seen in ambulatory care and discharged home should be under the care of the community respiratory nurse.
**Annual Review**

All patients should be offered an annual review, which includes:

- smoking cessation advice
- review of the number of exacerbations in the last year
- assessment of breathlessness associated with activities of daily living
- review of sputum volume, character and undertake sputum culture and sensitivity test to assess for chronic bacterial colonization
- If patient is regularly productive of sputum, consider referral to pulmonary rehabilitation service to teach individualised chest clearance techniques
- if patient is short of breath on exertion MRC 2 or above consider referral to pulmonary rehab service for group exercise sessions

**Annual spirometry is not recommended for people who are stable, with minimal exercise limitation, and who have few exacerbations.**

Do not routinely repeat CXRs.
Consider specialist follow-up

Refer patient for specialist follow-up if:

- chronic colonization with *Pseudomonas aeruginosa*, opportunist mycobacteria, or meticillin-resistant *Staphylococcus aureus* (MRSA)
- deteriorating symptoms
- three or more infective exacerbations a year
- for consideration for long-term prophylactic antibiotics
- bronchiectasis associated with:
  - rheumatoid arthritis
  - immune deficiency
  - inflammatory bowel disease
  - primary ciliary dyskinesia (PCD)
  - allergic bronchopulmonary aspergillosis (ABPA)
  - advanced disease
Immunisation, treatment & review self-management plan

Immunisations
- annual influenza vaccination
- offer immunization against Streptococcus pneumoniae and seasonal influenza

Treatment and self-management
- treat co-morbid respiratory conditions, e.g. COPD
- if patient is regularly productive of sputum refer to pulmonary rehabilitation service to teach individualised chest clearance techniques
- if patient is short of breath on exertion MRC 2 or above pulmonary rehabilitation group exercise sessions
- ensure people who are able to self-manage medication have been advised to start antibiotics themselves on exacerbation.

They should:
- understand when it is appropriate to start treatment
- the importance of collecting sputum before starting treatment
- review and update self-management plan including how to recognize exacerbations and understand the condition. A patient information leaflet on bronchiectasis is available from the British Lung Foundation: [http://www.blf.org.uk/Conditions/Detail/Bronchiectasis](http://www.blf.org.uk/Conditions/Detail/Bronchiectasis)
Send sputum for culture and sensitivity

Commonly isolated pathogens are:
- Haemophilus influenzae
- Staphylococcus aureus
- Streptococcus pneumoniae
- Moraxella catarrhalis
- Klebsiella sp

The following pathogens require specialist involvement:
- Pseudomonas aeruginosa
- Aspergillus fumigatus
- Mycobacterium tuberculosis
- Mycobacterium avium complex
Infective exacerbations

- optimal management of infective exacerbation can only be achieved if there is knowledge of the microbiological cause:
  - sputum cultures are useful in the stable phase as they may inform treatment for a subsequent exacerbation
  - sputum sample should be sent for culture whenever possible at the start of an exacerbation and prior to starting antibiotic treatment
  - empirical therapy should be initiated whilst awaiting culture results
Indications of infective exacerbation

Diagnose an infective exacerbation requiring antibiotic therapy when there is:
- acute deterioration (usually over several days), with:
  - worsening cough
  - increased sputum volume, viscosity, or purulence
  - increasing wheeze
  - breathlessness
  - systemic upset
  - increased fatigue and malaise
  - fever
  - pleuritic chest pain

NB: the presence of mucopurulent or purulent sputum alone, or the isolation of a pathogen alone without a deterioration in symptoms, is not necessarily an indication for antibiotic treatment

There is great variability in sputum appearance within and between patients:
- variation can occur with discoloured sputum produced within the same day as clear sputum
- many patients, whilst stable, will have sputum bacteriology that grows potentially pathogenic microorganisms
**Antibiotic therapy**

The decision to administer antibiotics as an outpatient or inpatient, orally or parenterally, should be based on the clinical scenario and likely pathogen:

- If previous cultures are available, treatment should be guided by known sensitivity patterns or according to NICE CKS and BTS guidelines: [https://www.brit-thoracic.org.uk/document-library/clinical-information/bronchiectasis/bts-guideline-for-non-cf-bronchiectasis/](https://www.brit-thoracic.org.uk/document-library/clinical-information/bronchiectasis/bts-guideline-for-non-cf-bronchiectasis/) (page iS)
- Send sputum for culture and sensitivity testing before starting antibiotics (even if the person is taking long-term antibiotics).

Collect expectorated sputum after deep coughing. Ensure prompt transport of specimens to the laboratory, as Haemophilus influenzae and Streptococcus pneumoniae may only be viable if the specimen is processed within 3 hours. Promptly prescribe an antibiotic for 10-14 days.

**Do not await the results of culture.**
First-line treatment, give for 10-14 days

- If there is no previous bacteriology, prescribe:
  - amoxicillin 500mg three times a day; or
  - clarithromycin 500mg two times a day (in patients who are penicillin-allergic); or
  - Doxycycline 200mg stat then 100mg OD
- High-dose oral regimens may be needed in patients with severe bronchiectasis chronically colonised with *Haemophilus influenzae*, e.g. amoxicillin 1g three times a day, or amoxicillin 3g two times a day
- Patients with *Pseudomonas* infection are at risk of developing chronic resistance and therefore should be referred to a specialist
- Failure to respond to an antibiotic course should prompt a repeat sputum culture
- Consider a short-acting inhaled beta2-agonist (e.g. salbutamol) if necessary for wheeze or breathlessness in the acute phase
- Ensure that a suitable airway clearance technique is used during the exacerbation
- Arrange an urgent appointment with a physiotherapist if the person has not already been taught this or if they cannot manage this alone
Follow-up of infective exacerbation

Review the following:
- response to empirical treatment when sputum culture and sensitivity results are available:
  - if the patient is responding well, continue with the prescribed antibiotic – do not change the treatment based on culture results
  - if the patient has not responded well to treatment, prescribe a different antibiotic
- effectiveness and acceptability of the chosen airway clearance technique within approximately 3 months of the initial exacerbation

Consider acute referral if the patient presents with:
- breathlessness with respiratory rate greater ≥ 25 breaths per minute
- intravenous therapy required in patients with clinical failure after oral antibiotics
- patients unable to take oral antibiotics
- patient is unable to cope at home
- patient develops cyanosis or is confused
- circulatory and/or respiratory failure
- temperature ≥ 38°C

If the patient deteriorates at any stage after starting treatment, reassess to see if hospital admission is indicated
Ongoing monitoring

Follow-up is divided between primary and secondary care, dependent on clinical need:

- long term prognostic data is limited to specialist centres, which skews the prognostic information towards more extreme cases
- the overall aim of follow-up is to monitor symptoms, prevent and/or manage complications, and prevent decline in lung function:
  - long-term follow-up should include monitoring of lung function, breathlessness associated with activities of daily living, sputum volume and character, frequency of exacerbations and antibiotic use, and microbiological status through sputum culture
  - review of smoking status, immunisation status, consideration for pulmonary rehab, how to identify and treat exacerbations (consideration for reserve course of antibiotics), treatment of comorbid conditions and appropriateness of self management plan
  - many patients will have positive sputum cultures for potentially pathogenic bacteria when clinically well; treatment should only be initiated on the basis of clinical status
  - certain potential pathogens should prompt repeat culture and/or specialist assessment even if the patient is well, e.g. *Pseudomonas* or *Mycobacterium* species
  - escalation of therapy may be prompted by:
    - increasing frequency of exacerbation
    - declining lung function
    - emergence of complications, including development of infection with particular organisms, e.g. *Pseudomonas* or *Mycobacterium* species

*Patients with recurrent infections and chronic Pseudomonas spp. who require nebulised antibiotics should receive on-going monitoring. Secondary care will initiate nebulised antibiotics. This may be continued by primary care where agreements for standard practice exist. This should be supported by a community respiratory nurse.*
**Referral Criteria**

Consider referral to secondary care for patients with the following features:

- clinical diagnosis of bronchiectasis who require a high resolution CT (HRCT) for confirmation of diagnosis
- haemoptysis
- deteriorating bronchiectasis with declining lung function
- colonised with:
  - chronic *Pseudomonas aeruginosa*
  - opportunist mycobacteria
  - meticillin-resistant *Staphylococcus aureus* (MRSA)
- recurrent LRTI - > 3 per annum
- bronchiectasis and associated rheumatoid arthritis, immune deficiency, inflammatory bowel disease, and primary ciliary dyskinesia (PCD)
- allergic bronchopulmonary aspergillosis (ABPA)
- advanced disease and those considering transplantation
- family history
- abnormal spirometry, e.g. FEV1 < 70% predicted
Information for Patients

'Bronchiectasis' from Patient UK: https://patient.info/health/bronchiectasis-leaflet
'Coughing up blood (haemoptysis)' from Patient UK: https://patient.info/health/coughing-up-blood-haemoptysis

'Wheeze' from Patient UK: https://patient.info/health/wheeze

'Bronchiectasis' from the British Lung Foundation: www.blf.org.uk
Nebuliser Support from the British Lung Foundation: https://www.blf.org.uk/support-for-you/nebulisers

'Breathlessness' from the British Lung Foundation: www.blf.org.uk

'Living with a lung condition' from the British Lung Foundation: www.blf.org.uk
Bronchopulmonary hygiene

The cycle of recurrent infection, bronchial damage, and impaired ciliary function leads to mucus accumulation:

- patients with hypersecretion of mucus should be referred to specialist pulmonary rehab team to encourage expectoration and enhance clearance:
  - it is essential for all patients with daily production of purulent sputum to be taught airway clearance by a suitably qualified physiotherapist
  - patients with a non-productive cough should be taught an appropriate technique to use during exacerbations
Manage complications

Haemoptysis is a common complication in patients with bronchiectasis:
• due to proliferation, dilatation, and malformation of the bronchial arteries surrounding the damaged airways
• episodes are typically related to an infective exacerbation
• small volume haemoptysis is usually managed with antibiotic therapy only if with an infective exacerbation and/or procoagulant agent, e.g. tranexamic acid, initiation would be on the advice of a specialist only
Management of underlying causes - additional specialist input

- Immunodeficiency should be jointly managed by an immunologist and a respiratory physician
- Connective tissue disease should be jointly managed by a rheumatologist and a respiratory physician
- Inflammatory bowel disease should be jointly managed by a gastroenterologist and a respiratory physician
- Cystic fibrosis should be managed under the care of a specialist cystic fibrosis centre
Contact secondary care respiratory physician if considering adjuvant pharmacological therapy

- bronchodilators - should only be prescribed after a trial of therapy has demonstrated improvement of symptoms of lung function
- long-term antibiotic therapy
- nebulised saline - may be considered to increase sputum yield, reduce sputum viscosity and improve ease of expectoration
- corticosteroids (inhaled or oral), not recommended for use unless there is coexistent asthma
Referral criteria for secondary care respiratory consultant

- a clinical diagnosis of bronchiectasis requires a HRCT for confirmation of diagnosis
- haemoptysis
- deteriorating bronchiectasis with declining lung function
- patients colonised with:
  - chronic *Pseudomonas aeruginosa*
  - opportunistic mycobacteria
  - meticillin-resistant *Staphylococcus aureus* (MRSA)
- recurrent LRTI - more than three per annum
- patients being considered for long-term prophylactic antibiotic therapy (oral or nebulised)
- patients with bronchiectasis and associated rheumatoid arthritis, immune deficiency, inflammatory bowel disease, and primary ciliary dyskinesia (PCD)
- patients with allergic bronchopulmonary aspergillosis (ABPA)
- patients with advanced disease and those considering transplantation
- family history
- abnormal spirometry, e.g. FEV1 < 70% predicted