

**Guideline for diagnosis and treatment of adult bronchiectasis**

This guideline is intended for use by both primary and secondary care.

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# 1. Definition

Bronchiectasis is defined as having inflamed, permanently and irreversibly damaged airways leading to symptoms of chronic cough, chronic sputum production and recurrent respiratory tract infections.

The prevalence is about 1:1000 = 0.1% and it is estimated in Lothian that about 800 people will be affected.

**Note: for detailed information, especially concerning secondary care, see the appendices.**

# 2. Aetiology (see also Appendix 1)

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| Infective | whooping cough  measles  tuberculosis  pneumonia |
| Bronchial obstruction | tumours  broncholithiasis  bronchial stenosis  lymph nodes  foreign body |
| Auto-immune | rheumatoid arthritis  systemic lupus erythematosus (SLE)  Sjogren’s syndrome |
| Inflammatory bowel disease | ulcerative colitis  Crohn’s |
| Vasculitis | Wegener’s |
| Cystic fibrosis |  |
| Ciliary defects |  |
| Immune deficiencies |  |
| Hyperimmune states | ABPA (allergic bronchopulmonary aspergillosis) |
| Congenital anatomic defects |  |
| Traction bronchiectasis associated  with pulmonary fibrosis |  |
| Alpha 1 antitrypsin deficiency |  |
| Chronic asthma |  |
| COPD |  |

**Note: in up to 50% of cases, no precipitating cause can be identified.**

# 3. Symptoms

* Chronic cough
* Chronic sputum production
* Recurrent respiratory tract infections.

# 4. How to diagnose bronchiectasis

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| **Non-smokers** | **Smokers** |
| Investigate if regular cough and sputum production | investigate if more than two respiratory tract infections/year |

* If you suspect bronchiectasis, send sputum for routine microbiology culture and arrange a chest x-ray
* If bronchiectasis is suspected, refer patients to consultant chest physician, Royal Infirmary of Edinburgh. Specialist bronchiectasis nurses are also based at the Royal Infirmary of Edinburgh.

# 5. Prognosis

* The disease is lifelong and the prognosis depends on the severity of the bronchiectasis. In severe cases, life expectancy will be reduced
* Patients chronically colonised with Pseudomonas aeruginosa tend to have poorer health related quality of life, more exacerbations and increased mortality.

# 6. Differential diagnoses

* COPD
* Asthma
* Post-nasal drip
* Tuberculosis
* Cancer
* Abscess
* Pneumonia.

# 7. Investigations (see also Appendix 2)

* + **PRIMARY CARE**
* **Chest x-ray** A normal chest x-ray does not exclude bronchiectasis - the gold standard is CT chest
* **Sputum** for routine microbiological culture.
* Approximately two thirds of patients with bronchiectasis have pathogenic bacteria isolated in the sputum. The most common organisms involved are:
  + *Haemophilus influenzae*
  + *Moraxella catarrhalis*
  + *Staphylococcus aureus*
  + *Streptococcus pneumoniae*
  + *Pseudomonas aeruginosa*
  + *Environmental mycobacteria* 
    - Check routine bloods - FBC, U+E, LFTs.

# 8. Definition of severity:

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| **Mild bronchiectasis** |
| * When stable, no sputum or mucoid/mucopurulent sputum * < 10 mls sputum produced per day * < 2 exacerbations per year * Normal lung function (FEV1, FVC and PEFR) * Normal chest x-ray * CT chest - tubular bronchiectasis ≤ 3 lobes * Not chronically colonised (chronically colonised = a sputum sample revealing a pathogen on two or more occasions in the past year when clinically stable). |

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| **Severe bronchiectasis** |
| * When stable, sputum mucopurulent or purulent * > 20 mls sputum produced in 24 hours * ≥ 3 exacerbations per year * Normal lung function or restrictive defect or obstructive defect * Chest x-ray: tram-lining, cystic changes, atelectasis * CT - varicose or cystic bronchiectasis affecting one or more lobesor multiple lobes affected with bronchiectasis * Chronically colonised. |

# 9. Treatment

The aim is to:

* Identify and treat underlying cause to prevent disease progression
* Maintain or improve pulmonary function
* Reduce exacerbations
* Improve quality of life by reducing daily symptoms and exacerbations.

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| **Advise the patient on the following:**   * Smoking cessation * Airway chest clearance procedures with physiotherapy * How to recognise and treat exacerbations promptly * How to recognise and access medical advice early in an exacerbation * The significance and importance of providing a sputum sample prior to treatment * Airway pharmacotherapy (see section 11 below) * Annual influenza vaccine * Pneumococcal immunisation (repeated every five years). |

**Note: treatment regimes with drugs and physiotherapy can be complex.**

Care must be taken to maximise the patient’s understanding of the disease. Most importantly patients from ethnic minorities and those with specific disabilities or mental health issues must be given appropriate information. Carers, caseworkers or advocates are often involved in helping with treatment and should also be included, to maximise their understanding.

# 10. Airway clearance

The aims of respiratory physiotherapy are to mobilise and aid expectoration of bronchopulmonary secretions, improve efficiency of ventilation, maintain or improve exercise tolerance, and reduce breathlessness and (thoracic) pain

* There are a wide variety of available airway clearance techniques and patients are encouraged to choose ones that they are comfortable with
* We recommend 20 to 30 minutes once or twice daily depending on patient’s severity
* All patients should see a respiratory physiotherapist.

# 11. Airway pharmacotherapy

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| **Proven therapies:** |
| * Bronchodilators - use of beta-2 agonist or anti-cholinergic as appropriate for symptom control if symptomatic breathlessness * Inhaled steroids can reduce sputum volume but there is no clear benefit in maintenance of FEV1 or reduction in exacerbation frequency. In severe bronchiectasis, a six-month inhaled corticosteroid trial is worthwhile (800µg daily inhaled corticosteroid). Only continue if there is clinical benefit. |

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| **No evidence of benefit:** |
| * Mucolytics - no role for recombinant human DNase; further trials are needed to establish efficacy of carbocysteine and inhaled mannitol * Corticosteroids - no evidence for a role for oral corticosteroids * Leukotriene receptor antagonist - no role. |

# 12. Exacerbations - when is an antibiotic needed?

Daily symptoms of cough and sputum production are frequent. More severe bronchiectasis patients often expectorate mucopurulent or purulent sputum and culture respiratory pathogens when apparently clinically stable. This is more common in adults. The presence of mucopurulent or purulent sputum alone, or the isolation of a pathogen alone is not necessarily an indication for antibiotic treatment.

Antibiotics are recommended for exacerbations that present with an acute deterioration (usually over several days) with worsening local symptoms (cough, increased sputum volume or change of viscosity, increased sputum purulence + increasing wheeze, breathlessness, haemoptysis) and/or systemic upset.

Managing patients with exacerbations requires an assessment of severity and decision about whether to treat the patient in the community or in hospital.

# 13. Which antibiotic to administer?

* Send sputum sample for culture prior to starting an antibiotic
* Start empirical antibiotics while waiting for culture results
* Previous sputum bacteriology can guide antibiotic use (see Appendix 3)
* If no previous bacteriology, first line treatment is:

Amoxicillin 500mg tds or clarithromycin 500mg bd for 7-14 days (if penicillin allergic) - 7 days for mild disease, 14 days if severe disease.

* High dose oral antibiotics eg Amoxicillin 1g tds or 3g bd may be required in severe bronchiectasis with chronic *Haemophilus influenzae* colonisation
* Failure to respond should prompt a repeat sputum culture
* Only change antibiotics if no clinical improvement (it can take several days before clinical improvement is seen) - be guided by antibiotic sensitivity results
* Criteria for IV antibiotics - refer them to the bronchiectasis service for assessment and possible admission:
  + unwell
  + resistant organisms not responsive to oral antibiotics
  + failed to respond to oral therapy (usually *Pseudomonas aeruginosa*).

# When is combination (dual) antibiotic therapy recommended?

* If 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
* Combination antibiotics are recommended for infections due to strains of *Pseudomonas aeruginosa* sometimesthat are resistant to one or more anti-pseudomonal antibiotics (including ciprofloxacin) or if the clinician suspects the patient will require many antibiotic courses, to reduce the development of drug resistance.
* MRSA should be treated with two oral antibiotics or a single intravenous agent. Intravenous vancomycin should only be used with appropriate and robust dosing and monitoring systems in place.

# When is combination (dual) antibiotic therapy NOT recommended?

* Combination antibiotics are **not** required in patients colonised with *Haemophilus influenzae*, *Moraxella catarrhalis*, *Staphylococcus aureus* (Methicillin sensitive) and *Streptococcus pneumoniae.*

# 14. Indicators for inpatient treatment

* Unable to cope at home
* Development of cyanosis or confusion
* Breathlessness with a respiratory rate ≥ 25 per minute
* Circulatory failure
* Respiratory failure with oxygen saturations <93% on room air
* Temperature ≥ 38°C
* Unable to take oral therapy.

# 15. Who should be followed up by secondary care? (if any of the following apply?

* Patients with recurrent exacerbations >3/year
* Deteriorating bronchiectasis with declining lung function
* Patients with moderate or severe bronchiectasis
* Patients receiving prophylactic antibiotic therapy (oral or nebulised)
* Patients who isolate *Pseudomonas aeruginosa* or enteric gram negative organisms, environmental mycobacteria or MRSA.

# Normal secondary care follow-up:

* Patients with moderate and severe disease should be followed up twice yearly
* Mild cases will be discharged from the clinic, but should be re-referred if symptoms worsen.

# 16. Structured secondary care letters

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| Secondary care clinic letters, from outpatient settings are structured and will have the following information:   * Diagnosis * CT scan result * BMI * Spirometry * Smoking history * In addition to medical staff, whether the patient has been seen by the specialist nurse and physiotherapist * Number of antibiotic courses since last assessment * Number of IV antibiotic courses since last assessment * Number of hospital admissions * Sputum production and sputum colour * Whether chronically colonised * Usual pathogen * Investigations * Recommended antibiotic for exacerbation * Medication summary * Follow-up/review date. |

# 18. Bronchiectasis service pathway

Assess patient for exacerbation

Send sputum for culture and sensitivity   
and start patient on antibiotics   
(do not wait for test results)

When culture and sensitivity results come back, check to ensure appropriate antibiotic.

**Only change antibiotic if no clinical improvement**

* previous sputum results can guide   
  antibiotic use
* if no prior sputum result, treat with   
  amoxicillin 500mg tds
* 7 days (mild disease)
* 14 days (severe disease)
* (clarithromycin 500mg bd for 7 to14 days   
  if penicillin allergic).

Patient has own supply at home?

Use this supply   
and advise to contact GP within 24 hours   
of starting to   
update record

**Indications for IV antibiotics and hospital admission**

**Indications for IVs:**

* unwell
* resistant organism not responsive to oral antibiotics
* failed response to oral antibiotics.

Discuss with bronchiectasis team

Use normal hospital admission policy

**Indications for  
hospital admission:**

* unable to cope at home
* cyanosis or confusion
* breathlessness with  
  RR ≥ 25 per minute
* circulatory failure
* respiratory failure with sats <93% on room air
* temperature ≥ 38°C
* unable to take oral therapy.

# Appendix 1 - Aetiology (expanded)

|  |  |
| --- | --- |
| Infective | * whooping cough * measles * tuberculosis * pneumonia |
| Bronchial obstruction | * endobronchial tumours * broncholithiasis * bronchial stenosis resulting from infections * encroachment of hilar lymph nodes * foreign body aspiration |
| Auto-immune | * rheumatoid arthritis * systemic lupus erythematosus (SLE) * Sjogren’s syndrome |
| Inflammatory bowel disease | * ulcerative colitis * Crohn’s |
| Vasculitis | * Wegener’s |
| Cystic fibrosis |  |
| Ciliary defects |  |
| Immune deficiencies |  |
| Hyperimmune states | * ABPA (allergic bronchopulmonary aspergillosis) |
| Congenital anatomic defects | * [Bronchopulmonary sequestration](http://emedicine.medscape.com/article/1005815-overview) * Williams-Campbell syndrome (congenital cartilage deficiency) * Mounier-Kuhn syndrome (tracheobronchomegaly) * [Swyer-James syndrome](http://emedicine.medscape.com/article/361906-overview) (unilateral hyperlucent lung) * Yellow nail syndrome |
| Traction bronchiectasis associated with pulmonary fibrosis |  |
| Alpha 1 antitrypsin deficiency |  |
| Chronic asthma |  |
| COPD |  |

**Note: in up to 50% of cases, no precipitating cause can be identified.**

# Appendix 2 - Secondary care investigation

The following blood tests should be performed in all patients:

* Full blood count and white blood cell differential
* Erythrocyte sedimentation rate and C-reactive protein
* Urea and electrolytes and liver function tests
* Serum immunoglobulins (G, A, M, E), protein electrophoresis and IgG subclasses
* IgE and IgE to Aspergillus
* Citrullinated peptide (replaced Rheumatoid Factor)
* ANA, and anti-double stranded DNA, if ANA positive
* ANCA
* Alpha 1 antitrypsin level and phenotype.

Sputum

* Routine microbiological culture
* 24-hour sputum for fungal culture if allergic broncholpulmonary aspergillosis suspected
* Three sputum samples for TB culture should be sent if there is upper lobe, middle lobe, lingular or nodular bronchiectasis

Lung function

* FEV1, FVC and PEF
* FEV1, FVC, lung volumes and gas transfer should be measured at baseline including reversibility to beta-agonist and anti-cholinergic if baseline spirometry shows a restrictive or obstructive picture.

Radiology

* HRCT for all patients.

Exclude cystic fibrosis (measurements of sweat chloride and CFTR genetic mutation analysis)

* All patients aged <40 presenting with bronchiectasis should have investigations for cystic fibrosis
* Consider in aged >40 if persistent isolation of Staphylococcus aureus in the sputum, features of malabsorption, male primary infertility, upper lobe bronchiectasis, a history of childhood steatorrhoea.

# Appendix 3 - Common organisms associated with an exacerbation with the recommended antimicrobial agents

|  |  |  |  |
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| **Organism** | **Recommended first line treatment** | **Recommended second line treatment** | **Length of treatment** |
| ***Streptococcus pneumoniae*** | Amoxicillin  500mg tds (po) | Clarithromycin 500mg bd (po)\* | 7 to14 days |
| ***Haemophilus influenzae*,**  **β-lactamase negative** | Amoxicillin  500mg tds (po)  Amoxicillin 1G tds (po)  Amoxicillin 3G bd (po) | Doxycycline 100mg bd (po)  or  Clarithromycin 500mg bd\* (po)  or  Ciprofloxacin  500 mg\* bd (po)  or  Ceftriaxone 2G od\* (IV) or 1g bd if administering as iv bolus (outpatient). | 7 to 14 days  In BTS guidelines this is detailed under haemophillus influenzae |
| ***Haemophilus influenzae*,**  **β-lactamase positive** | Co-amoxiclav  625mg TDS (po) | Doxycycline 100mg bd (po)  or  Clarithromycin 500mg bd\* (po)  or  Ciprofloxacin 500mg bd\* (po)  or  Ceftriaxone 2G od\* (IV) or 1g bd if administering as iv bolus (outpatient). | 7 to 14 days |
| ***Moraxella catarrhalis*** | Co-amoxiclav  625mg TDS (po) | Doxycycline 100mg bd (po)  or  Clarithromycin 500mg bd\* (po)  or  Ciprofloxacin  500mg bd\* (po)  or  Ceftriaxone 2G od\* (IV) or 1g bd if administering as iv bolus (outpatient). | 7 to 14 days |
| ***Staphylococcus aureus* (MSSA)** | Flucloxacillin  500mg QDS (po) | Clarithromycin  500mg BD (po)\* | 7 to 14 days |
| ***Staphylococcus aureus* (MRSA)**  **Oral preparations** | Rifampicin  300mg bd (po)  +  Doxycycline  200mg od (po) | Rifampicin 300mg bd (po) + Trimethoprim 200mg bd (po)  **Third line**  Linezolid 600mg bd (po) -requires microbiology approval | 14 days  First and second line reverse to this in BTS  14 days |
| ***Staphylococcus aureus* (MRSA)**  **Intravenous preparations** | Vancomycin\*  **see link 1 below**  or Teicoplanin\* 400mg od (iv)  **see link 2 below** | Linezolid 600mg bd (po)  - requires microbiology approval | 14 days |
| **Coliforms e.g. Klebsiella, Enterobacter** | Oral Ciprofloxacin  500mg bd\* (po) | Intravenous Ceftriaxone 2G od or  1g bd if administering as iv bolus (outpatient). | 7 to 14 days |

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| **Organism** | **Recommended first line treatment** | **Recommended second line treatment** | **Length of treatment** |
| ***Pseudomonas aeruginosa*** | Oral Ciprofloxacin  500mg bd\* (po)  (750mg bd in more severe infections) | Monotherapy Intravenous  Ceftazidime 2G tds\*  or  Tazocin 4.5G tds\*  or  Meropenum 2G tds\*  or  Aztreonam 2G tds\*  **Combination therapy:**  The above can be combined with gentamicin\* (refer to respiratory pharmacist) or colistin 1-2 MU tds\* (under 60 kg, 50 000-75 000 units/kg daily separated into 3 divided doses) OR oral ciprofloxacin 500mg BD\*  **See link 3 below, section 1.3 for gentamicin dosing** | 14 days  Tazocin is a brand which we no longer use. Refer to as tazobactam/  piperacillin |

Note: caution with aminoglycosides in pregnancy, renal failure, elderly or on multiple other drugs.

\* dose needs adjusted for impaired renal function

**Useful links**

**1. Guide to Vancomycin dosing**

<http://intranet.lothian.scot.nhs.uk/NHSLothian/Healthcare/A-Z/amt/AntimicrobialGuidelines/Pages/vancomycin_dosing.aspx>

2. Electronic Medicines Compendium - Targocid 200mg & 400mg

<http://emc.medicines.org.uk/document.aspx?documentId=9110>

* 1. Guide to Gentamicin dosing

[**http://intranet.lothian.scot.nhs.uk/NHSLothian/Healthcare/A-Z/amt/AntimicrobialGuidelines/Pages/gentamicin\_dosing.aspx**](http://intranet.lothian.scot.nhs.uk/NHSLothian/Healthcare/A-Z/amt/AntimicrobialGuidelines/Pages/gentamicin_dosing.aspx)