Primary Care Respiratory Society UK - Opinion No.41

Opinion

Bronchiectasis

Overview: what is bronchiectasis?

First described by Laënnec in 1819,[1] bronchiectasis is a debilitating long-term disease of children and adults caused by chronically inflamed and damaged airways. The diagnosis of bronchiectasis is based on both clinical and radiological features. Chronic cough and sputum production and recurrent respiratory tract infections are characteristic of bronchiectasis. High-resolution computed tomography (HRCT) is the diagnostic modality of choice in bronchiectasis.

In the 1950s (i.e. pre-HRCT) in the UK, about one person in 1,000 was estimated to have bronchiectasis.[2] The current prevalence is unknown. This article will focus on non-cystic fibrosis bronchiectasis in adults.

Case finding in primary care

The BTS guidelines[3] suggest that bronchiectasis should be considered in adults with:

- a persistent productive cough;
- a young age of presentation;
- a history of symptoms over many years;
- absence of smoking history;
- daily expectoration of large volumes of very purulent sputum;
- haemoptysis;
- sputum colonisation with Pseudomonas aeruginosa;
- coarse crackles in the lung on auscultation.

Bronchiectasis may be misdiagnosed as chronic obstructive pulmonary disease (COPD) or the two conditions may co-exist and should be considered if:

- management of the COPD is not straightforward;
- recovery from lower respiratory tract infections is slow;
- the patient has recurrent infections;
- there is <15 pack year smoking history.[3]

Patients suspected of having bronchiectasis require an HRCT scan. In primary care this may necessitate referral to secondary care for further screening and investigations.

Causes of bronchiectasis

Common causes of bronchiectasis and relevant investigations are outlined in Table 1,[4,5] but in up to 53% of cases no cause is identified. The commonest cause is post infection in up to 42% (e.g. following pneumonia, whooping cough, measles and tuberculosis).[4,5]

Table 1. Underlying causes of bronchiectasis

Aetiology	Incidence	History/signs	Investigations
Idiopathic	Up to 53%		Diagnosis of exclusion
Post infection	Up to 42%	History of pneumonia, pertusis, measles, TB	CXR or CT scan evidence of previous infection
Immune defect	8%		Decreased immunoglobulin levels, or functional antibody deficiency
Allergic bronchopulmonary aspergillosis	7%	History of asthma.	Eosinophilia, raised IgE (total and *Aspergillus* specific); fleeting infiltrates or proximal bronchiectasis on CXR and/or HRCT
Aspiration/gastro-oesophageal reflux	4%	History of aspiration or reflux	Foreign body/mucus plugging on bronchoscopy
Rheumatoid arthritis (RA)	3%	History of RA	Positive autoimmune screen
Cystic fibrosis (CF)	3%	Age <40 years; malabsorption; male infertility; diabetes	CFTR mutations; positive sweat test
Ciliary dysfunction	1.5%	Situs inversus; productive cough; deafness; infertility	Abnormal ciliary beat pattern in two separate sites on the respiratory tract
Ulcerative colitis	<1%	Malabsorption; diarrhoea; weight loss; joint pain	Colonoscopy and biopsies suggestive of inflammatory bowel disease
Congenital	<1%		N/A

HRCT=high resolution computed tomography; CFTR=cystic fibrosis transmembrane conductance regulator, CXR = chest X-ray

X-ray findings – what to look for?

The chest X-ray is usually normal unless patients have severe bronchiectasis.[6,7] Characteristic chest X-ray appearances include crowding of bronchi, parallel line opacities (tram lines) caused by thickened dilated bronchi, ring opacities or cystic spaces as large as 2 cm in diameter resulting from cystic bronchiectasis (Figure 1), sometimes with air-fluid levels, and oligaemia as a result of a reduction in pulmonary artery perfusion.[8]

Recommendation:
BTS guidelines recommend a baseline chest X-ray be done in all patients with bronchiectasis and a repeat chest X-ray if clinically indicated.[3]

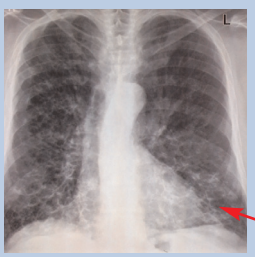
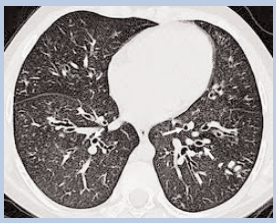


Figure 1: Chest X-ray in a patient with extensive bronchiectasis

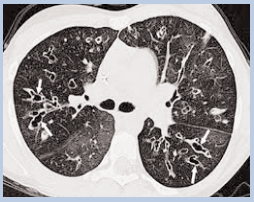
HRCT is the gold standard investigation. Bronchiectasis is diagnosed when there is bronchial wall dilatation, defined as the internal lumen of the bronchus being greater than that of the accompanying pulmonary artery.

Recommendation:
HRCT is the imaging modality of choice to diagnose bronchiectasis.

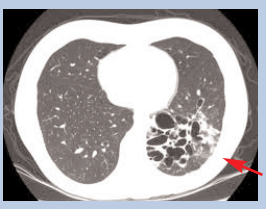
Figure 2: Reid classification of bronchiectasis according to the degree of bronchial dilatation.[9]



Mild: Tubular bronchiectasis
Minimally dilated bronchi



Moderate: Varicose bronchiectasis
Dilated bronchi resemble varicose veins



Severe: Cystic bronchiectasis

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When to refer to a specialist clinic?

In addition to confirming a diagnosis, the BTS guideline[3] suggests referring patients with:

- recurrent exacerbations (≥3 per year);
- deteriorating bronchiectasis with declining lung function;
- chronic *Pseudomonas aeruginosa*, opportunist mycobacteria or methicillin-resistant *Staphylococcus* aureus colonisation;
- consideration or already on long term antibiotic therapy;
- bronchiectasis associated with rheumatoid arthritis, immune deiciency, inflammatory bowel disease and ciliary dyskinesia;
- allergic bronchopulmonary aspergillosis;
- advanced disease and those considering transplantation.

Role of primary care in caring for people with bronchiectasis

General education

Primary care can support the provision of information for patients and/or carers including:

- Advice and help to stop smoking.
- Annual flu vaccination and pneumococcal vaccination every 5 years.
- Importance of airway pharmacotherapy.
- When and how to seek medical attention so that exacerbations can be treated promptly.

Airway clearance

Respiratory physiotherapy mobilises and aids expectoration of bronchopulmonary secretions with the aim of reducing symptoms, exacerbations and improving quality of life. There are a variety of techniques available – the choice depending on patient preference in conjunction with the physiotherapist.

Recommendation:
All patients with bronchiectasis should see a specialist chest physiotherapist. Chest physiotherapy is advised twice daily for 20-30 mins in patients with more advanced bronchiectasis.

Sputum microbiology

Almost two thirds of patients with bronchiectasis are chronically colonised with organisms.[10] Common pathogens are *Haemophilus influenzae, Moraxella catarrhalis, Staphylococcus aureus, Streptococcus pneumoniae, Pseudomonas aeruginosa* and occasionally environmental mycobacteria. In adults, *Haemophilus* influenzae is the most common organism isolated and is found in about 35% of patients.[10] Patients chronically colonised with *Pseudomonas aeruginosa* have increased hospital admissions, worse quality of life and may have an accelerated decline in forced expiratory volume in 1 second.[11]

Recommendation:
At each visit, all patients should be encouraged to provide a sputum sample for routine microbiology culture. This can guide antibiotic therapy for exacerbations and long-term management strategies.

Airway pharmacotherapy

Spirometry should be performed in adults with bronchiectasis and patients with airflow obstruction should be assessed for reversibility to β_2-agonist and anticholinergic bronchodilators. Short-acting bronchodilators (β_2-agonists or anticholinergics) are used in patients who are breathless on exertion, stepping up to long-acting bronchodilators if patients are still breathless.

Inhaled corticosteroids are only indicated to treat co-existent asthma or COPD.

Recommendation:
Bronchodilators are recommended if there is dyspnoea. Continue as maintenance treatment if lung function or symptoms improve with treatment.

Management of an exacerbation

Sputum should be sent for microbiology culture but dont wait for the culture results and start immediate empirical antibiotic therapy. Antibiotics are recommended for exacerbations that present with an several days of worsening local symptoms (cough, increased sputum volume, increasing sputum purulence, increasing wheeze, breathlessness, haemoptysis) and/or systemic upset.[3]

Antibiotic recommendations

If preious sputum bacteriology is known, initial treatment is guided by reported sensitivity. Otherwise, treatment should be started empirically with amoxicillin 500mg three times a day or clarithromycin 500mg twice a day (in penicillin allergy) for 14 days. If there is no clinical improvement, antibiotics can be modified once the pathogen is isolated and antibiotic sensitivities are available. Failure to respond to an antibiotic course should prompt a repeat sputum culture.

Recommendation:
If no previous sputum microbiology is available, start amoxicillin empirically 500mg three times a day or clarithromycin 500mg twice a day for 14 days if penicillin-allergic.

When should patients be admitted to hospital?

Inpatient treatment is recommended if the patient is unable to cope at home. Other indications include development of cyanosis or confusion; breathlessness with a respiratory rate ≥25/min; circulatory or respiratory failure; temperature ≥38°C; unable to take oral therapy; or if intravenous therapy is required (e.g. because of resistance of organisms to oral antibiotics or failure to respond to oral therapy – this is most likely to apply to patients with *Pseudomonas aeruginosa*).[3]

Good practice points[3]

1. All patients should be assessed for underlying cause(s).
2. Patients with an infective exacerbation of bronchiectasis should be assessed for inpatient or outpatient treatment.
3. Sputum microbiology should be checked prior to commencing antibiotics.
4. If no sensitivities are available treat empirically with amoxicillin or clarithromycin.
5. Exacerbations should be treated with antibiotics for 14 days.
6. All patients with bronchiectasis should see a chest physiotherapist.
7. Primary and secondary care nurses should receive training in management of bronchiectasis.

Conclusion

Bronchiectasis is a chronic debilitating progressive disease. Patient education and prompt treatment of exacerbations with antibiotics reduces symptoms and improves health-related quality of life. Successful management of bronchiectasis is dependent on appropriate treatment, both in primary and secondary care.

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