Approach to Bronchiectasis

1. Diagnosis

2. Management
Case 1

- Mr B, 76yo male, non smoker
- Has pulmonary tuberculosis 20 years ago, treated
- Since then daily productive cough
- Sputum are mucopurulent in nature
- He also has breathlessness, worse with exertion
Case 1

- On examination, he has finger clubbing
- Left lower zone crackles were heard, intensity slightly altered with coughing
- No other additional breath sounds
Case 1

- Chest X Ray
### Case 1

--- SPIROMETRY ---

<table>
<thead>
<tr>
<th></th>
<th>Pred</th>
<th>LLN</th>
<th>Actual</th>
<th>%Pred</th>
</tr>
</thead>
<tbody>
<tr>
<td>FEV1 (L)</td>
<td>2.66</td>
<td>1.75</td>
<td>1.94</td>
<td>73</td>
</tr>
<tr>
<td>FVC (L)</td>
<td>3.74</td>
<td>2.52</td>
<td>2.94</td>
<td>79</td>
</tr>
<tr>
<td>FEV1/FVC (%)</td>
<td>80</td>
<td>72</td>
<td>66</td>
<td>82</td>
</tr>
</tbody>
</table>

[Graph depicting spirometry data]
What is bronchiectasis?

• Bronchiectasis is a condition with chronic, irreversible dilatation of one or more bronchi
FORMS OF BRONCHIAL DILATATION

Dilatations of the air sacs occur due to bronchiectasis, as depicted below.

- Saccular
- Cylindrical
- Varicose

- Mucus and pus


REDUCTION IN BRONCHIAL SUBDIVISION IN BRONCHIECTASIS

BY

LYNNE McA. REID*

*From the Royal Melbourne Hospital*
Diagnosis

• High resolution computed tomography (HRCT) of chest
Pathophysiology

Neutrophil Inflammation (Proteases) → Airway Destruction and Distortion (Bronchiectasis) → Abnormal Mucus Clearance → Bacterial Colonization

Figure 1. Vicious cycle hypothesis. Host-mediated inflammatory response to foreign material and bacteria in the airway causes tissue damage resulting in bronchiectasis, which contributes to abnormal mucus clearance and further bacterial colonization.
What causes Bronchiectasis?

- Post infection
  - Bacterial, TB, adenovirus, measles
- Congenital
  - Primary ciliary dyskinesia, cystic fibrosis
- Immunodeficiency
  - hypogamaglobulinemia
- Aspiration or foreign body
- Rheumatoid arthritis
- Idiopathic (up to 50% of patients)\(^1-4\)

### TABLE 1. ETIOLOGIES OF NON–CYSTIC FIBROSIS BRONCHIECTASIS

<table>
<thead>
<tr>
<th>Cause</th>
<th>n ( % of study population)</th>
</tr>
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<tbody>
<tr>
<td>Autoimmune disease</td>
<td></td>
</tr>
<tr>
<td>Rheumatoid arthritis</td>
<td></td>
</tr>
<tr>
<td>Sjögren's syndrome</td>
<td></td>
</tr>
<tr>
<td>Cilia abnormalities</td>
<td></td>
</tr>
<tr>
<td>Primary ciliary dyskinesia</td>
<td></td>
</tr>
<tr>
<td>Connective tissue disease</td>
<td></td>
</tr>
<tr>
<td>Tracheobronchomegaly (Mounier-Kuhn syndrome)</td>
<td></td>
</tr>
<tr>
<td>Marfan's disease</td>
<td></td>
</tr>
<tr>
<td>Cartilage deficiency (Williams-Campbell syndrome)</td>
<td></td>
</tr>
<tr>
<td>Hypersensitivity</td>
<td></td>
</tr>
<tr>
<td>Allergic bronchopulmonary aspergillosis (ABPA)</td>
<td></td>
</tr>
<tr>
<td>Immune deficiency</td>
<td></td>
</tr>
<tr>
<td>Immunoglobulin deficiency</td>
<td></td>
</tr>
<tr>
<td>HIV infection</td>
<td></td>
</tr>
<tr>
<td>Job’s syndrome</td>
<td></td>
</tr>
<tr>
<td>Inflammatory bowel disease</td>
<td></td>
</tr>
<tr>
<td>Ulcerative colitis</td>
<td></td>
</tr>
<tr>
<td>Crohn's disease</td>
<td></td>
</tr>
<tr>
<td>Injury</td>
<td></td>
</tr>
<tr>
<td>Pneumonia/childhood infections</td>
<td></td>
</tr>
<tr>
<td>Aspiration</td>
<td></td>
</tr>
<tr>
<td>Smoke inhalation</td>
<td></td>
</tr>
<tr>
<td>Malignancy</td>
<td></td>
</tr>
<tr>
<td>Chronic lymphocytic lymphoma</td>
<td></td>
</tr>
<tr>
<td>Stem cell transplantation; graft-versus-host disease</td>
<td></td>
</tr>
<tr>
<td>Obstruction</td>
<td></td>
</tr>
<tr>
<td>Tumor</td>
<td></td>
</tr>
<tr>
<td>Foreign body</td>
<td></td>
</tr>
<tr>
<td>Lymphadenopathy</td>
<td></td>
</tr>
<tr>
<td>Other</td>
<td></td>
</tr>
<tr>
<td>α₁-Antitrypsin deficiency</td>
<td></td>
</tr>
<tr>
<td>Yellow nail syndrome</td>
<td></td>
</tr>
<tr>
<td>Young's syndrome</td>
<td></td>
</tr>
<tr>
<td>Post infection</td>
<td>52 (32)</td>
</tr>
<tr>
<td>Idiopathic</td>
<td>43 (26)</td>
</tr>
<tr>
<td>PCD</td>
<td>17 (10)</td>
</tr>
<tr>
<td>ABPA</td>
<td>13 (8)</td>
</tr>
<tr>
<td>Immune deficiency</td>
<td>11 (7)</td>
</tr>
<tr>
<td>Ulcerative colitis</td>
<td>5 (3)</td>
</tr>
<tr>
<td>Young's syndrome</td>
<td>5 (3)</td>
</tr>
<tr>
<td>Pan bronchiolitis</td>
<td>4 (2)</td>
</tr>
<tr>
<td>Yellow nail syndrome</td>
<td>4 (2)</td>
</tr>
<tr>
<td>Mycobacterium infection</td>
<td>4 (2)</td>
</tr>
<tr>
<td>Rheumatoid arthritis</td>
<td>3 (2)</td>
</tr>
<tr>
<td>Aspiration</td>
<td>2 (1)</td>
</tr>
<tr>
<td>Cystic fibrosis</td>
<td>2 (1)</td>
</tr>
<tr>
<td>Total</td>
<td>165</td>
</tr>
</tbody>
</table>

**ABPA** = allergic bronchopulmonary aspergillosis.  
**PCD** = primary ciliary dyskinesia.

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**Aetiology in adult patients with bronchiectasis**

A. Shoemark, L. Ozerovitch, R. Wilson

Evaluation

• After the diagnosis of bronchiectasis, we need to identify
  – potentially treatable causes,
  – microbiologic pathogens, and
  – functional assessment
Evaluation

• Personal history: childhood symptoms, infertility, previous pneumonia/viral illness, gastric aspiration, asthma, joints pain or deformity

• Family history: primary ciliary dyskinesia (PCD) (Kartagener's syndrome – sinusitis, situs inversus and bronchiectasis), cystic fibrosis (CF)
Evaluation

• Investigations:
  – Full blood count with differential
  – Immunoglobulin levels (IgG, IgM and IgA), consider IgE when eosinophilia
  – Rheumatoid factor
  – Sputum smear and culture for bacteria, mycobacteria (NTM) and fungi (mainly *Aspergillus*)
Additional Tests

• Testing for cystic fibrosis
  – Sweat chloride (minimum 2 requirements) and/or mutation analysis of the cystic fibrosis transmembrane conductance regulator (CFTR) gene (if clinical suspicion high)
• Nasal nitric oxide test
• Alpha-1 antitrypsin level and genotype
• HIV antibody test
• Swallow study/pH monitoring
• Bronchoscopy
**Table 3. Diagnostic Testing for Bronchiectasis.**

<table>
<thead>
<tr>
<th>Level of Testing</th>
<th>Appropriate Tests</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Blood</td>
</tr>
<tr>
<td>Primary</td>
<td>Complete and differential blood count, IgG, IgA, IgM</td>
</tr>
<tr>
<td>Secondary</td>
<td>Rheumatoid factor; IgE, aspergillus precipitins (ABPA); IgG subclasses; alpha(_1)-antitrypsin level</td>
</tr>
</tbody>
</table>

*The causes for suspicion of bronchiectasis are chronic cough, daily production of mucopurulent sputum, and persistent focal infiltrate on radiography. CT denotes computed tomography, and ABPA allergic bronchopulmonary aspergillosis.*

**Bronchiectasis**

Alan F. Barker, M.D.

Case 2

- Mr P, 40yo male, non smoker
- Known bronchiectasis due to primary ciliary dyskinesia
- Having fever and more productive cough for past 3 days
- Sputum became more purulent
- Has minimal haemoptysis, not worsening
Case 2

- On examination, he was febrile (38.3°C) but appeared not in distress
- Bilateral crackles heard on lung examination, more at lower zones
- No additional breath sounds heard
- Chest radiograph did not show any consolidations/pleural effusion
Exacerbations

Figure 2  Definition of an exacerbation needing antibiotic therapy.

- Increased cough + wheeze + breathlessness + systemic upset
- Increased sputum volume or change in viscosity
- Increased sputum purulence

British Thoracic Society guideline for non-CF bronchiectasis

M C Pasteur, D Bilton, A T Hill, on behalf of the British Thoracic Society Bronchiectasis (non-CF) Guideline Group

Thorax 2010;65:1—58. doi:10.1136/thx.2010.136119
Exacerbations

• Before starting antibiotics, a sputum sample should be sent off for culture

• If there is no previous bacteriology, first-line treatment is co-amoxiclav 625mg twice daily for **14 days**¹,²

• Ciprofloxacin 500-750mg twice daily should be used in patients colonised with *Pseudomonas aeruginosa*

• Cautious use in elderly subjects *Clostridium difficile* colitis

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Exacerbations

• 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 *P aeruginosa*)
Management

• Goals of treatment
  – Identify and treat underlying cause to prevent disease progression
  – Maintain or improve pulmonary function
  – Reduce exacerbations
  – Reduce complications and mortality
Airway Hygiene

• Physiotherapy
  – Active cycle of breathing techniques, huff coughs
  – Postural drainage
  – Oscillating positive expiratory devices
  – High-frequency chest wall oscillation
Airway Hygiene

- **Adjuncts**
  - Nebulised saline\(^1\): increase sputum yield, reduce viscosity, improve ease of expectoration
  - Hypertonic saline\(^2\) (3-7%) may induce bronchoconstriction (consider pretreatment with nebulised bronchodilator)
  - Nebulised terbutaline (5mg)\(^1,2\): direct hydration and/or \(\beta_2\) adrenergic stimulation

Recombinant human DNase should **NOT** be used in non-CF bronchiectasis => worse outcome\(^3\)

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Bronchodilator

• $\beta_2$ agonists$^{1,2,4}$
  – Can be considered in those have reversibility of airway obstruction by $\beta_2$ adrenergic stimulants

• Anticholinergic agents$^3$
  – Some adults may gain a useful response$^1$

Anti-inflammatory

• Corticosteroids
  – systemic corticosteroids are not used for patients with non-CF bronchiectasis (except patients with allergic bronchopulmonary aspergillosis)
  – not enough evidence to support routine use of inhaled corticosteroids in bronchiectasis (outside of use for those patients with additional asthma)
Anti-inflammatory

• Macrolides

Azithromycin for prevention of exacerbations in non-cystic fibrosis bronchiectasis (EMBRACE): a randomised, double-blind, placebo-controlled trial

Conroy Wong, Lata Jayaram, Noel Karalus, Tom Eaton, Cecilia Tong, Hans Hockey, David Milne, Wendy Fergusson, Christine Tuffery, Paul Sexton, Louanne Storey, Toni Ashton

Lancet 2012; 380: 660-67

Effect of Azithromycin Maintenance Treatment on Infectious Exacerbations Among Patients With Non-Cystic Fibrosis Bronchiectasis
The BAT Randomized Controlled Trial

JAMA. 2013;309(12):1251-1259

Effect of Long-term, Low-Dose Erythromycin on Pulmonary Exacerbations Among Patients With Non-Cystic Fibrosis Bronchiectasis
The BLESS Randomized Controlled Trial

JAMA. 2013;309(12):1260-1267

1. Potential resistant strains of bacteria and NTM
2. QT interval lead to fatal arrhythmias
Antibiotics

1. Attempt to eradicate Pseudomonas and/or MRSA,

2. To suppress the burden of chronic bacterial colonization, or

3. To treat exacerbations
Exercise

• Pulmonary rehabilitation should be offered to individuals who have breathlessness affecting their activities of daily living

• A retrospective study\(^1\) showed significant improvement in 6-minute walk distance and health-related quality of life scores after participated supervised exercise sessions

Others

• **Mucolytic**
  – Bromhexine is shown to be beneficial in the treatment of bronchiectasis exacerbations\(^1\)

• **Surgery**
  – Lung resection
  – Lung transplantation

Figure 4. Overview of a comprehensive approach to bronchiectasis management. AAT = α₁-antitrypsin; ATS/IDSA = American Thoracic Society/Infectious Diseases Society of America; IgG = immunoglobulin G; HRCT = high-resolution computed tomography; NTM = nontuberculous mycobacteria. *A 2-week course is suggested.
Complications

• Recurrent pneumonia
• Progressive respiratory failure/cor pulmonale
• Pneumothorax
• Massive haemoptysis
Prognosis

• The prognosis of bronchiectasis is undefined
• A number of factors contribute to poorer outcome
• “FACED” bronchiectasis prognostic score
• “Bronchiectasis Severity Index” (BSI)
THANK YOU
MCQ 1

Which of the following statement is **NOT** true?

A. Cystic bronchiectasis is the most severe form

B. Age is one of the prognostic factor for bronchiectasis

C. Chest x ray can confidently exclude bronchiectasis

D. Bronchiectasis can be presented with obstructive defect on spirometry
During infective exacerbation of bronchiectasis, which of the following is true

A. Duration of antibiotic should be 5 to 7 days
B. Systemic steroid should be given for all patients
C. Sputum culture should be sent prior to administration of antibiotic
D. All patients need to be hospitalised
MCQ 3

Which of the following is **NOT** the cause of bronchiectasis with upper respiratory symptoms/sinusitis

A. Cystic fibrosis
B. Primary ciliary dyskinesia
C. Young syndrome
D. Rheumatoid arthritis
**Figure 4** Eradication algorithm for *Pseudomonas aeruginosa* in adults. Attempt to eradicate with a 2-week course of oral ciprofloxacin (step 1). If step 1 fails, further regimens may be considered (step 2).