Bronchiectasis

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Background

- I have had a longstanding interest in bronchiectasis (Peter Holmes)
- Completed a PhD on this topic
- Have an ongoing research interest in this topic
- Initially regarded as being a boutique condition but now is very common (and has significant overlap with COPD)
Background

- What is bronchiectasis
- Clinical features
- Tests for diagnosis
- Treatment options
- Outcome in bronchiectasis
Definition of bronchiectasis

- From Greek
  - "Bronkhos" = the wind pipes of the lung
  - "Ektasis" = dilated, expanded
Normal Lung and Airways (Panel A) and the Lung of a Patient with Bronchiectasis (Panel B).
How does bronchiectasis occur

- Bronchiectasis is due to chronic inflammation in the bronchi that damages the airways and makes them wider
- This inflammation arises from bacterial infection in the lung
- The bacteria which cause this inflammation in the lung and found in the throat of all healthy adults
Bacteria in the upper and lower respiratory tract

**Upper airway microbiome**
- Bacteria present, no inflammation (commensal)
  - *H. influenzae*
  - *S. pneumoniae*
  - *M. catarrhalis*

**Lower airway microbiome**
- Colonization & chronic inflammation
The lung immune response

- The lung has an extensive immune response which helps stop bacteria from moving down from the throat into the lung.

- Sometimes the lung immune response does not control the bacteria and there may be:
  - Acute infection e.g. acute bronchitis or pneumonia
  - Chronic infection e.g. chronic bronchitis or bronchiectasis
How does bronchiectasis occur

- A defect in the lung immune response allows bacteria from the throat to move down into the lung airways where they cause inflammation
  - Number of causes of defects in lung immune response
    - Cystic fibrosis (very rare in adult)
    - Structural lung damage, especially COPD. Up to half of patients with COPD may develop bronchiectasis

- The bacteria become established in the lung and cannot be cleared

- There is progressive inflammation which results in lung damage
Airflow obstruction in bronchiectasis
Viral infections have a potentially important role

- Common viral infections such as the common cold and influenza acutely damage the lung immune response and may allow bacteria to proliferate.

- Viral infection may have 2 effects:
  - Lead to bacteria moving from the throat to the lung and set up chronic bronchitis “the cold that never went away”
  - May cause acute flare ups of chronic bronchitis
Bronchiectasis is characterised by abnormal and permanent airway (bronchial widened).

It is due to chronic bacterial infection of the lung that damages the airways and lung.

A defect in the lung immune response allows bacteria to move down from the throat and establish infection in the lung airways.
Clinical features of bronchiectasis

- The most common feature of bronchiectasis is a chronic cough which is generally productive of sputum.
- Chronic airway inflammation causes the productive cough.
- The sputum colour varies from clear/white to dark green.
Sputum characteristics

- Both the volume of the sputum and the colour of the sputum correlate with lung inflammation.

- The white cells in the lung have green granules and the more inflammation the greener the sputum.
Shortness of breath

- Shortness of breath (or dyspnoea) is also a prominent feature of bronchiectasis

- There are many different causes of this symptom

- In the context of bronchiectasis shortness of breath has 2 major causes
  - Lung inflammation
  - Airway obstruction
Haemoptysis

- Haemoptysis is the coughing up of blood
- Usually mixed in with sputum
- Inflamed airways bleed
- Bleeding is usually minor
Other symptoms

- Chronic fatigue is prominent especially in patients with more advanced disease.

- Patients with childhood onset disease often will have prominent sinus/upper respiratory tract disease.

- Reflux

- Urinary incontinence
Findings on medical examination

- Localised crackles can be heard in the lungs particularly at the bottom of the lungs

- Other signs are wheeze and if acutely unwell; temperature and elevated heart rate and respiratory rate
Chronic and acute symptoms

• Most patients will have mild to moderate symptoms particularly cough chronically

• Periodically there will be episodes of acute worsening e.g. exacerbations
  • Increased cough, more sputum and more purulent
  • Increased shortness of breath
  • Other symptoms; haemoptysis, constitutional upset

• These exacerbations appear to often be triggered by viral infections
Summary of symptoms

- Bronchiectasis is most often characterised by a chronic productive cough (with other symptoms) with periodic acute worsening

- There may be overlap with other respiratory diseases particularly airway diseases such as
  - Asthma
  - COPD
Tests for bronchiectasis

- Bronchiectasis is diagnosed now with a CAT scan of the chest

- The ready availability of CAT scans has markedly increased the awareness of bronchiectasis

- The requesting doctor needs to order a particular form of CT scan (a high resolution CT or HRCT) to best pick up bronchiectasis
CT scan and bronchiectasis
Sputum samples

- If possible we always like to have a sputum sample to determine which bacteria is causing the inflammation in the lungs

- Problems
  - Patients can’t produce a good quality sample
  - Recent use of antibiotics
  - The sputum itself is filled with chemicals which can kill bacteria
Blood tests

- Several blood tests are ordered and these are to assess if there is a detectable immune deficiency that may be treatable.

- Most of the time these blood tests will not change management.
Lung function testing

- Patients with bronchiectasis over time tend to develop a drop in lung function particularly the development of airflow obstruction.

- Lung function testing may be important in the diagnosis of other lung conditions such as asthma and COPD.
Other tests

- Sometimes other tests are used but these are really for the specialist respiratory physician

- Other tests include
  - Cardiac assessment; echocardiogram
  - Oxygen assessment
  - Ear, nose and throat (ENT) review
  - Clinical nutrition review
Summary of tests

- The diagnosis of bronchiectasis is made by CT scan
- Important other tests include, sputum sample and lung function testing
- Testing helps differentiate bronchiectasis from other conditions
- Patients will generally have symptoms for at least a year before a diagnosis is made (in our study 20 years!)
Treatment for bronchiectasis

- Bronchiectasis is a very variable condition

- It may be difficult to distinguish from other lung conditions

- Patients will generally always have some symptoms no matter how successful treatment is

- Management is directed at both chronic symptoms and the episodes of acute worsening (exacerbations)
Treatment of bronchiectasis

- Treatment (particularly with antibiotics) has greatly improved outcome but treatment strategies are still not that well defined

- Markedly increased interest and newly available guidelines for therapy
  - Medical Journal of Australia (MJA) guidelines on the management of bronchiectasis
  - These guidelines are widely available for both GPs and specialists
Antibiotics

- Remain the cornerstone of treatment
- Should be guided (if available) by results from sputum
- Generally require a double course
- Some antibiotics are fast acting (e.g. penicillin) whilst others are slow acting but are good at penetrating into the lung tissue where the reservoir of infection is present (e.g. rulide or doxycycline)
Antibiotics and exacerbations

- Initially in patients with bronchiectasis use antibiotics only for exacerbations (particularly if increased sputum volume or purulence)

- Double course

- Early initiation of treatment (often patient initiated)

- If not improving in a few days, need escalation of therapy (stronger oral antibiotics ➔ hospital in the home ➔ hospitalisation)
Other uses of antibiotics

- We sometimes use maintenance antibiotics for patients with severe symptoms (e.g. doxycycline or clarithromycin)

- Inhaled antibiotics are sometimes used but are not widely available and are expensive
Physiotherapy

- An important feature of lung disease is that airway secretions (from the inflammation) need to be cleared and failure to clear these secretions results in more infection/inflammation and more secretions.

- Physiotherapy particularly in acute exacerbations facilitates clearance of sputum and chronically in patients with more severe disease.
Physiotherapy

- The a variety of methods but most commonly used are specific breathing exercises (dynamic cycle of breathing) and devices
- Need to be shown this by a phyiotherapist with a specialist interest
- An active lifestyle probably improves sputum clearance
Bronchodilators

- At this stage there is little evidence that bronchodilators such as salbutamol (ventolin) have any effect in bronchiectasis (in contrast to asthma) and are not recommended as standard therapy for bronchiectasis.
Corticosteroids

- Corticosteroids which contain cortisol (e.g. prednisolone) are antiinflammatory are first line therapy for asthma.

- However as they suppress the immune response (which is partially controlling the infection in bronchiectasis) they can make the infection worse.

- The use of cortisol both as a tablet and as a puffer is not recommended for the treatment of bronchiectasis unless there is associated asthma.
Vaccination and other treatments

- Regular vaccines are recommended
  - Annual influenza vaccine
  - 5 yearly pneumococcal vaccine (new Prevenar 13 vaccine)

- Other therapy
  - ENT review
  - Oxygen
  - Transplant
Patient driven treatment

- As bronchiectasis is variable and patients respond differently an individualized regime should ideally be developed for each patient in conjunction with a respiratory physician.

- Patients drive a lot of the management
  - Recognizing when they are becoming unwell
  - Self initiation of antibiotics, physiotherapy etc.
  - Avoid exposure to viral infections if possible
Summary of management

- Antibiotics and physiotherapy are the main treatment options

- Early initiation of therapy for exacerbations and escalation as required

- Individualized regime for each patient (and distinguish from other respiratory conditions such as asthma)
Outcome in bronchiectasis

- Bronchiectasis is a heterogeneous condition.
- Patients will generally always have symptoms.
- A small proportion of patients will have rapidly progressive disease and a small proportion will have almost no symptoms.
- The clinical picture is affected by age.
Outcome in bronchiectasis

- Two common presentations of bronchiectasis
  - CHILDHOOD ONSET DISEASE: starts in early childhood with significant symptoms and often sinus involvement, improves as an adolescent then comes back from mid 50s
  - ADULT ONSET DISEASE: typically starts from mid 50s, often as a seeming viral infection (the cold that never went away), prominent in COPD

- Bronchiectasis tends to be very slowly progressive and the severity of illness is related to the duration of the productive cough
Progress in bronchiectasis

- Bronchiectasis is developing a much higher profile in recent years
- Progress is being made in the understanding of this condition
- As part of this the ALF is developing a national registry of bronchiectasis patients and one of the sites is through Monash
Acknowledgements

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