Bronchiectasis

Definition of bronchiectasis

- Abnormal and permanent dilatation of airways
  - Bronchial walls become inflamed, thickened and irreversibly damaged
  - The mucociliary elevator is impaired
  - Mucus accumulates leading to increased susceptibility to infection

Epidemiology of bronchiectasis

- Prevalence in the UK estimated as 100/100,000
  - Prevalence increases with age

Causes of bronchiectasis

- Congenital
  - Cystic Fibrosis
  - Primary Ciliary dyskinesia (sinusitis, bronchiectasis and azospermia)
  - Kartagener's (primary ciliary dyskinesia with dextrocardia and situs inversus)
  - Young's syndrome (azospermia and sinusitis)
  - Pulmonary sequestration
- Mechanical obstruction
  - Foreign body
  - Bronchial carcinoma
  - Post-TB Stenosis
  - Lymph node
- Post-Infective
  - Measles
  - TB
  - Pertussis
  - Bacterial and viral pneumonia
- Granulomatous Disease
  - TB
  - Sarcoidosis
- Usual interstitial pneumonia (cryptogenic fibrosing alvelolitis)
- Immune over-activity
  - Allergic broncho-pulmonary aspergillosis (ABPA)
  - Inflammatory bowel disease
  - Rheumatoid arthritis
  - Sjorgrens
  - Post lung transplant
- Immune deficiency
  - Hypogammaglobulinaemia
  - Selective immunoglobulin deficiencies (IgA and IgG_2)
  - Secondary
    - HIV, malignancy
- Aspiration
  - Chronic alcoholics
  - GORD

Presentation of bronchiectasis

- Symptoms
  - Cough
  - Shortness of breath
  - Excessive sputum production
  - Recurrent chest infections
  - Haemoptysis
- Signs
  - Cachexia and lymph nodes
  - Clubbing
  - Hyperinflation
  - On Auscultation
    - Coarse crackles in affected areas: mixed character, alter with coughing
    - Squeaks and Wheeze
    - Inspiratory clicks

Differential diagnosis of bronchiectasis

- Pulmonary fibrosis
- Bronchial carcinoma
- Chronic lung abscess
- Asbestosis

Investigation of bronchiectasis

- Sputum culture and cytology
- CXR
  - Tramlines and ring shadows. Bullae.
- HRCT
  - "Signet ring" sign: thickened, dilated bronchi larger than the adjacent vascular bundle
- Sinus x-rays
  - 30% have concomitant sinusistis
- Spirometry
  - Normal/ restrictive picture
- For a specific cause:
  - Bronchoscopy
  - Immunoglobulins
  - Aspergillus RAST and skin prick testing
  - Sweat electrolyte test
  - Mucocilliary clearance
    - Nasal saccharine taste test: 1mm cube of saccharine placed on inferior turbinate should be tasted within 30mins)

Management of bronchiectasis

- Non-pharmacological
  - MDT
  - Physiotherapy
    - Postural drainage
    - Active cycle breathing
  - Smoking cessation
  - Immunisations

- Medical
  - Antibiotics
    - To treat exacerbations refer to local guidelines but examples include:
      - Amoxicillin 500mg tds or clarithromycin 500mg bd for 2 weeks as 1st line
      - Ciprofloxacin in pseudomonas colonisation
      - High dose maybe needed in severe bronchiectasis with Haemophilus infuenzae B colonisation e.g. amoxicillin 1g tds
    - Long term antibiotics
      - Consider in patients having ≥ 3 exacerbations per year or patients with fewer exacerbations causing significant morbidity e.g. low dose azithromycin three times per week
      - Inhaled antibiotics can also be used
  - Bronchodilators/ inhaled corticosteroids if there is any evidence of airflow obstruction
  - Inhaled Saline
  - NIV/ Intermittent positive pressure may be used to augment tidal volume and reduce work of breathing
- Surgical
  - Resection in localised disease
  - Lung transplant (heart/lung transplant)
  - Bronchial artery emobolisation or surgery for management of haemoptysis

Complications of bronchiectasis

- Progressive respiratory failure
- Cor pulmonale
- Pneumonia
- Pneumothorax
- Empyema
- Life-threatening haemoptysis: Mycotic aneurysm (esp. in patients with CF)
- Secondary amyloidosis

Prognosis of bronchiectasis

- Vastly improved with antibiotic therapy, but most still eventually progress to respiratory failure due to chronic damage.

Common questions concerning bronchiectasis

- What organisms are commonly associated with bronchiectasis?
  - Staph aureus
  - Haemophilus influenza
  - Pseudomonas
  - Rarer:
    - Pneumococcus
    - Klebsiella
- What is Yellow Nail Syndrome?
  - Bronchiectasis + yellow nails + Lymphoedema
- What do you know about Cystic fibrosis?
  - Autosomal recessive disorder of the cystic fibrosis transmembrane conductance regulator (CFTR) – chromosome 7 (Delta F508). 1:2500.
    - Induces low salt and chloride excretion into airways leading to increased viscosity of secretions

- Presentation
    - Lung: recurrent chest infections, bronchiectasis
    - ENT: nasal polyps and sinusitis
    - GI: Meconium ileus, malabsorption, intestinal obstruction, steattorrhoea (pancreatic insufficiency)
    - Other: Fertility, arthropathy
- Investigations
    - Sweat electrolyte test
        - Chloride >60 is diagnostic
    - CXR
        - Peribronchial thickening (but can be normal)
    - CT (HR)
    - DNA analysis for genotype
- Management
    - Antibiotics
    - Pancreatic/nutritional supplements
    - Inhaled antibiotics, steroids and recombinant human DNase
    - CFTR gene therapy
    - Lung transplant
- Complications
    - DM (10%) – treat with insulin
    - ABPA (15%)
        - High dose steroids
    - Infections, osteoporosis, liver disease
- Prognosis
    - Median survival 40 years