Non CF Bronchiectasis

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Definition

Abnormal bronchial wall thickening and luminal dilatation of central and medium sized bronchi.

Epidemiology

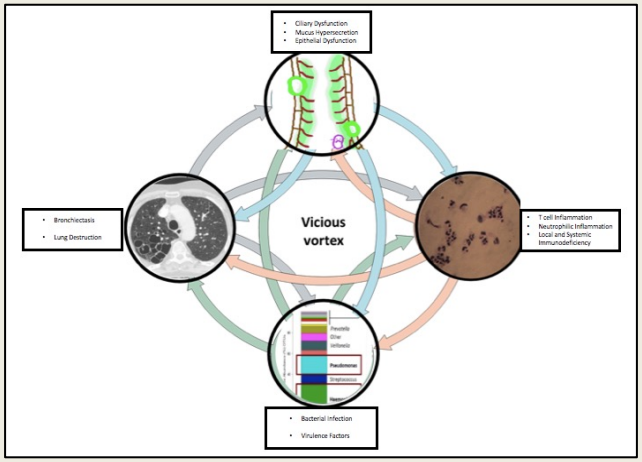
566 Female vs 486 Male cases/100,000. Increases in those over 60 years of age

Clinical Features

- **Cough:** >8 weeks with purulent sputum, bronchial infection and abnormal permanent dilation of bronchi, bronchial wall thickening, mucus plugging.
- **Chronic Airway Infection**
- **Exacerbations:**(50% will have 2 or more/year) associated with mortality, quality of life, lung function and further exacerbations.
- **Dyspnoea**
- **Haemoptysis**

Aetiology

Idiopathic	**50% idiopathic** (less likely if <50 at onset or rapidly progressive)
Associated Respiratory Diseases:	**Asthma** 40% of asthmatics with difficult to control asthma have radiological evidence of bronchiectasis. 27% of patients with bronchiectasis also have asthma. **COPD** (54.3% have both, more common in males with OR 1.62, higher if severe AFO and exacerbations, consider in COPD if 2 or more exacerbations per year or *Pseudomonas Aeruginosa* cultured) **ABPA** seen in 1-11% cases **Alpha 1 antitrypsin deficiency** cause in 1% of cases - suspect in basal emphysema **PCD** 0.9-10% see PCD Infection: Childhood measles, pneumonia, TB, whooping cough **Localised Disease**: Foreign Body, Tumour, Traction
Immunodeficiency	**5.8%** have immunodeficiency: isolated IgA deficiency or CVID (IgG,IgA and sometimes IgM), X linked Agammaglobulinemia (reduction in all classes)
Inflammatory Diseases	**Rheumatoid Arthritis** 2-5% of cases caused by RA (lung disease can preceded arthritis Other **CTD** 5% **Inflammatory Bowel Disease** 1-3%
Aspiration	1-11% have symptoms of aspiration (Hiatus Hernia, Tracheo-oesophageal Fistula, Neurological Disease)
Rare Causes	**Congenital:** Williams Campbell Syndrome (tracheobronchomalacia), Mounier Kuhn syndrome (bronchomegaly), Lung sequestration **Acquired:** Young syndrome – obstructive azoospermia, rhinosinusitis - mercury poisoning. Yellow Nail Syndrome: pleural effusions, lymphoedema



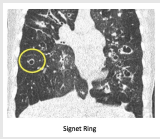
Laboratory Tests

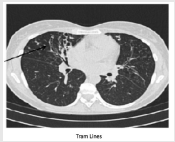
Bloods:	• FBC, Serum Protein Electrophoresis. Total IgE • Aspergillus specific IgE/Skin test • Pneumococcal (PPV23) anti-body response (4 fold at 4-6 weeks) * if low immunoglobulin levels/low normal • Connective Tissue Disease work up: * if features suggestive: Rheumatoid Factor, Anti CCP, ENA,ANA/ANCA • Alpha-1 Antitrypsin Levels • HIV test - depending on population prevalence
Sputum	• Minimum yearly, at least 1 for AFB pre long term macrolides.

Imaging

Chest Radiograph:
- Parallel line opacities (tram tracking), Tubular Opacities (mucus filled bronchi), Ring opacities (Dilated end-on bronchi).
- Abnormal in 87.8% but specificity 74%. Useful at baseline as comparator.

CT features
- Bronchus > Artery (broncho-arterial ratio >1.5) (signet ring)
- Lack of tapering (airway visible within 1cm of pleura)
- Features specific to aetiology: CF -upper lobe, PCD -middle lobe, Alpha 1 – emphysema, ABPA - infiltrates


Signet Ring


Tram Lines

Bronchoscopy :
- Focal disease to out rule foreign body/lesion.
- Not expectorating and clinically deteriorating

Pulmonary Function Tests
- Obstructive in over 50% but can be restrictive/mixed/normal

Treatment

Maintenance Therapy	• Physiotherapy: OD/BD >10 but <30 mins CT to tailor technique Review after 3 month Avoid head down if GORD NIV useful if early fatigue or exacerbation Modify if haemoptysis Hypertonic Saline • Pulmonary Rehabilitation • Vaccines: Flu, Pneumonia, COVID-19 • LABA /LAMA low grade evidence • ICS/PO steroids only with co-existing disease e.g. asthma/abpa (may have a role IBD associated bronchiectasis) • Treatment of cause (e.g. Immunodeficiency, GORD) • Monitor: BMI, Sputum Culture, Sats, Consider severity scoring (Bronchiectasis Severity Index/ FACED score)
Exacerbation Treatment	• Minimum 14 days up to 21days • Tailor to clinical response and microbiology (note in vitro and in vivo antibiotic sensitivities can differ) • Anti-Pseudomonal Beta-Lactam +/- aminoglycoside • Massive haemoptysis: bronchial artery embolization/surgery
Eradication Therapy	Pseudomonas Aeruginosa: 1. Oral (ciprofloxacin) or IV therapy x 2/52 2. 3 months inhaled agent (colomycin/gentamicin/tobramycin)
Long Term Antibiotics	• Consider if >3 exacerbations/yr • Pseudomonas Negative: azithromycin 250-500mg alt days/daily • Pseudomonas Positive: Inhaled Colomycin/Gentamicin +/- po azithromycin
Advanced Disease	• NIV • Oxygen • Surgery • Transplant

Specific Tests

Investigations:
- Sweat Test, Genetic Testing
- Features: Younger patients, upper lobe predominance, nasal polyps, chronic rhino sinusitis, recurrent pancreatitis, male infertility, malabsorption)

Primary Ciliary Dyskinesia Investigations (PCD):
- Nasal Nitric Oxide, nasal NO, brushings, genetic testing
- Features: productive cough since childhood, situs inversus, congenital heart disease, nasal polyps, chronic rhinosinusitis, middle ear disease, neonatal ARDS/ICU admission as neonate