BRONCHIECTASIS
BRONCHIECTASIS

• Age group 5-15 Yrs.

• Causes
  – Congenital / Hereditary
  – Acquired
DEFINITION

Abnormal permanent dilatation of bronchi or subsegmental bronchi with inflammatory destruction of peribronchial tissue [muscle, cartilage] with accumulation of secretions in the dependent bronchi
CAUSES

• In developing countries
  Frequently sequelae of acute infection.

• In developed world
  in association with underlying disorders such as
  cystic fibrosis
  immune deficiencies (including HIV)
  primary ciliary dyskinesia
  recurrent aspiration syndromes.
TYPES

Classic ‘Reid’ classification (gross histological appearance) - three different patterns

1. **Cylindrical bronchiectasis** - mildly uniform airway dilation
2. **Varicose bronchiectasis** - focally dilated areas between narrowed segments
3. **Saccular bronchiectasis** - balloon-like airway dilation with more disruption of lung parenchyma
PATHOGENESIS

- Chronic infection – recruitment of neutrophils, T-lymphocytes, monocyte-derived cytokines with release of inflammatory mediators - elastases & collagenases
- Loss of ciliated columnar epithelium
- Micro-abscess in bronchial wall with peribronchial inflammation
- Destruction - elastic & muscle tissue, cartilage of bronchus.
- Endarteritis of pulmonary vessels
- Bronchial arterial proliferation (predisposes to hemoptysis)
CONGENITAL CAUSES

• **Structural** – William Campbell Syndrome, Mounier Kuhn syndrome, airway malacia

• **Abnormal immune function** – agammaglobulinemia, combined immunodeficiency, neutrophil function abnormalities

• **Cystic fibrosis (CF)**

• **Ciliary abnormalities** - Kartagener's syndrome, Immotile cilia syndrome

• **Others** – yellow nail syndrome, alpha 1 anti-trypsin deficiency, ataxia telangiectasia
ACQUIRED CAUSES

Infection

Obstruction

Aspiration syndrome

BACTERIAL
Tb, Pertussis, Mycoplasma

VIRAL
Measles, Viral Pneumonia, HIV

Chronic lung allograft rejection

Foreign body

Foreign body

Foreign body

Lymph nodes

Lymph nodes

Lymph nodes

Tumors

Tumors

Tumors

Endobronchial TB

Endobronchial TB

Endobronchial TB

Inspissated mucus-

Inspissated mucus-

Inspissated mucus-

[cystic fibrosis,

[cystic fibrosis,

[cystic fibrosis,

Immotile cilia syndrome]

Immotile cilia syndrome]

Immotile cilia syndrome]
CLINICAL MANIFESTATIONS

• Most common symptom - persistent cough, typically "wet" or productive
• Episodic exacerbations of infection-
  • Increased cough and sputum production
  • Fever
  • Pleuritic chest pain
  • Dyspnea
• Absence of sputum production does not exclude bronchiectasis (younger children may not be able to expectorate
CLINICAL MANIFESTATIONS

- Hemoptysis - uncommon in children
- Occurs - erosion of inflamed airway tissue adjacent to pulmonary vessels.
- Bleeding
  - Mild, with blood streaked sputum
  - Profuse amounts of fresh bleeding if larger pulmonary vessels rupture.
CLINICAL MANIFESTATIONS

• Dyspnea and exercise intolerance

• Uncommon at presentation

• May develop as disease progresses

• May occur during acute exacerbation (intercurrent infection)
CLINICAL MANIFESTATIONS

• Cyanosis- severe bronchiectatic lung disease

• Severe hypoxemia due to mismatched pulmonary ventilation and perfusion.

• If hypoxemia - prolonged and profound- cause pulmonary hypertension & cor-pulmonale.
CLUES FOR AETIOLOGY

- **Failure to thrive** - cystic fibrosis (CF) and immunodeficiency disorders.

- **Chronic sinusitis** – cystic fibrosis, ciliary dysfunction disorders, immunodeficiencies

- **Chronic ear infection with or without otorrhea** - ciliary dysfunction.
CLUES FOR AETIOLOGY

- **Steatorrhea** — suggests cystic fibrosis.

- **Choking history** — foreign body aspiration or swallowing disorder with chronic aspiration of oropharyngeal contents.

- **Dextrocardia** suggests primary ciliary dyskinesia.
1. General physical examination
2. Failure to thrive
3. Sinus and ear infections
4. Presence of congenital anomalies
5. Clubbing
   1. Schamroth sign- obliteration of the quadrangular space when both fingers kept in unison
   2. Distal phalangeal depth(DPD) to interphalangeal depth ratio(IPD) > 1
6. Coarse leathery crackle over areas of bronchiectasis
A  Schamroth sign
Normal
Clubbed

B  Phalangeal depth ratio
Normal
Clubbed

DPD  IPD
DIAGNOSIS

• Diagnosis depends on radiographically or anatomically visualizing abnormal dilatation of airways

• Diagnostic procedure of choice - High-Resolution Computed Tomography (HRCT) scan

• Other tests- diagnose underlying conditions.
LABORATORY INVESTIGATIONS

- Complete blood count with differential
- Sputum exam - volume, gram stain, C & S
- Immunodeficiency - Total IgM, IgA and IgG
- Tests for Tuberculosis –
  - Mantoux
  - Chest X-ray
  - Resting Gastric juice for AFB
- Test for cystic fibrosis (sweat chloride and/or DNA testing)
INVESTIGATIONS

- Flexible bronchoscopy
  - shows structural alteration of bronchial tree
  - bronchoalveolar lavage - remove mucus plugs/ to obtain lower airway cultures.
  - If an airway foreign body is discovered-rigid bronchoscopy -for removal

- HIV screening

- Ciliary biopsy
INVESTIGATIONS

• Test for GERD: 24 hour pH monitoring, upper gastrointestinal endoscopy and technetium milk scan scintigraphy.

• Test for aspiration due to inadequate airway protective mechanisms during swallowing or dysphagia-evaluated by video fluoroscopy.
INVESTIGATIONS

• Pulmonary function tests - severity of lung disease
• should be performed if the patient is able to do
• useful tool to evaluate long-term progression of lung disease.

• Most patients with bronchiectasis have features of obstructive lung disease (low FEV1 & FEV1/FVC ratio)
INVESTIGATIONS

• Test for allergic bronchopulmonary aspergillosis
  • Immunoglobulin E (IgE)

• Serum precipitins for *Aspergillus* species

• Sputum culture for fungus

• Aspergillus skin test
CHEST RADIOGRAPHY

- Dilated and thickened airways (tram-tracking or parallel lines)
- Irregular peripheral opacities that represent mucopurulent plugs
- Loss of lung volume and peribronchial fibrosis
Cystic fibrosis - upper lobes

Allergic bronchopulmonary aspergillosis - Centrally located bronchiectasis

Bronchopulmonary sequestration - lower lobe and usually unilateral
X-RAY AND CT CHEST – NOTE THE SEVERITY IN CT CHEST
IMAGING - CT

• Most sensitive to detect bronchiectasis - high resolution computed tomography (HRCT)

• Internal diameter of airway - larger than diameter of adjacent artery

• Airway wall thickening ("signet ring" shadows) with or without air fluid levels

• Volume loss, mucus plugging and air trapping
MEDICAL TREATMENT

- Control infection
- Physiotherapy
- Nutritional support
- Identify etiology and treat accordingly
MEDICAL MANAGEMENT

• Immediate
  – Medical treatment
  – Postural drainage
  – Relief of atelectasis
  – Treatment of associated problems

• Long term
  – Continuation of postural drainage
MANAGEMENT

Treatment – any identified underlying disorder

Therapy
  • reduce airway secretions
  • facilitating their removal - with chest physiotherapy & mucolytic agents

Pharmacotherapy
  • to improve mucociliary clearance.

Antibiotics
  • prevent & treat recurrent infections

Surgery – if localized disease - may be considered.
CHEST PHYSIOTHERAPY

• Facilitate mucous expectoration - manual and mechanical interventions
• Chest percussion
• Vibration
• Postural drainage
• Cough-assist devices & airway oscillation-serve as adjuncts to cough (most effective and efficient manner of clearing airway).
SURGICAL TREATMENT

- Only in unilateral disease
- Removal of affected segment/lobe/lung
- Surgery contraindicated if bilateral disease
- Unilateral - surgical resection - good prognosis
COMPLICATIONS

- Broncho-pneumonia
- Empyema
- Lung abscess
- Hemoptysis
- Metastatic abscess-brain
- Osteomyelitis
- Hemoptysis
- Cor pulmonale
- Amyloidosis
PREVENTION OF BRONCHIECTASIS

- Childhood immunization for measles and pertussis
- Screening for tuberculosis and treatment wherever needed
- Aggressive appropriate therapy of lower respiratory tract infections
- Therapy of child with chronic or recurrent respiratory problems due to recurrent aspiration and/or gastroesophageal reflux disease
PROGNOSIS

- Overall - prognosis – good
- In absence of an underlying condition-children with isolated bronchiectasis- good prognosis
- Progressive bronchiectasis from underlying disease or ongoing pulmonary insult –causes progressive obstructive defect & ultimately, respiratory compromise
Thank You