Bronchiectasis in Children

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Objectives

• Objective #1
discuss the specific etiologies and clinical presentation of bronchiectasis in childhood

• Objective #2
review approaches to diagnosis and management and current therapies for bronchiectasis in childhood
Bronchiectasis
Pathophysiology

chronic or repeated episodes of environmental insults + genetic vulnerability

bronchial injury and dilatation

Chronic infiltrates and atelectasis
Chronic productive or wet cough
Radiological imaging

Chest x-ray

HRCT scan
Pathology

Gross specimen
classic saccular enlargement
of the bronchi

Histology/pathology
dilated lumen of the bronchus and
chronic inflammation
in the bronchial wall
Causes of bronchiectasis

• Cystic fibrosis
• Impaired immune function
• Ciliary dyskinesia
• Clinical syndromes
• Congenital tracheobronchomegaly
• Aspiration syndromes
• Obstructive bronchiectasis
• Other pulmonary diseases
10-year-old boy with chronic infiltrates
12-year-old boy with cystic bronchiectasis since infancy
30-year-old male with recurrent pneumonia
25-year-old male diagnosed with asthma as a child
14-year-old male with situs inversus
Bronchiectasis in childhood

- cystic fibrosis (CF) is the most common cause of bronchiectasis in childhood

- non-CF bronchiectasis (systematic review)
  - specific etiologies on non-CF bronchiectasis in childhood (prevalence)
    - infectious (17%)
    - primary immunodeficiency (16%),
    - aspiration (10%)
    - ciliary dyskinesia (9%
    - congenital malformation (3%)
    - secondary immunodeficiency (3%)

Brower et al. BMC Pediatrics 2014, 14:299
Underestimate of cases

• Misdiagnosis
  • “difficult asthma”
  • Chronic obstructive pulmonary disease (COPD)
    • 29% of adults with COPD have underlying bronchiectasis
Genetics

• Interplay between genotype and environment = phenotypic expression of respiratory disease

• Frequency of CFTR genotypes

• Turkish study:
  • Consanguinity
    • Transporter associated with Antigen Presentation (TAP) gene polymorphisms in cohort of children with bronchiectasis
Innate pulmonary immune mechanisms

• Pro-inflammatory cytokine
• Adhesion molecule production and receptor expression
• Exaggerated neutrophilic response
• Metalloproteinases (MMP-2 and -9)
  • Isolated in sputum and BAL
  • Airway destruction by galtinases and collagenases
Reid’s subtypes
(bronchographic findings)

- Cylindrical
- Varicose
- Cystic

Cylindrical
Varicose and cystic changes
HRCT scoring systems
(more recent)

Markers of disease severity

• Cylindrical
• Saccular
saccular bronchiectasis
Increased bronchoarterial ratio

Signet ring sign
broncho-arterial ratio > 1

Diameter of the bronchial lumen divided by the diameter of accompanying artery
normal bronchoarterial ratio
< 5 years of age = 0.5
< 18 years of age = 0.8
fluid-filled dilated bronchi
bronchial wall thickening
Vascular changes

• Total bronchial arterial blood flow is increased

• Extensive precapillary anastomoses between the two arterial systems (shunt between pulmonary and systemic systems)

• Vascular remodeling of the pulmonary arteries and arterioles (pulmonary hypertension and cor pulmonale)
Airway mucosa abnormality (bronchoscopy)
Sputum markers

• Neutrophilic airway inflammation
  • CF sputum
    • Viscous
    • Elastic
    • Adhesive
Mucus-filled saccular airway changes
Respiratory pathogens

- *Streptococcus pneumoniae*
- *Haemophilus influenza non-type b*
- *Moraxella catarrhalis*
- *Pseudomonas aeruginosa*
Mechanisms of lung destruction

• Exaggerated or persistent pulmonary inflammation
• Balance between proteases and anti-proteases
• Collagenase activity (from neutrophils and bacteria)
• Metalloproteinases
• Impaired removal of apoptotic inflammatory cells
• Adhesion molecules
Management Principles

• Aggressive management of infections (antimicrobials)
• Airway clearance methods
• Attention to nutrition
• Vigilant monitoring of clinical trends
• Proactive care
Philosophy of antibiotic use

• Maintenance
• Intermittent
• Regular hospitalizations
Airway Clearance Techniques

• Mechanical “valve” devices (Flutter, Pep, Acapella, and others)
• Postural drainage and chest physiotherapy
• Therapeutic vest
Mucus alteration agents

- Inhaled $\beta_2$-agonists and/or anticholinergic bronchodilators
- Hypertonic saline or mannitol inhalation
- Dornase alfa (Pulmozyme)
- $N$-Acetylcysteine (Mucomyst)
New therapies

• Macrolides
• Anti-inflammatory
• Anti-oxidant
• Anti-secretagogue effects
• Statins
Other therapies/management

• Asthma therapy
• Environmental modification
• Prevention: Vaccines
Complications related to bronchiectasis

- Hemoptysis
- Lung abscess
- Pulmonary hypertension
- Sleep disorders
- Reactive airway disease
Lobectomy

• **Indications**
  - Poor control of symptoms
  - Poor growth inspite of optimal medical therapy
  - Severe and recurrent hemoptysis uncontrolled by bronchial artery embolization

• **Relative indication**
  - Localized disease with moderate persistent symptoms

• **Contraindications**
  - Widespread bronchiectasis
  - Young child (<6 years of age)
  - Minimally symptomatic disease
Bronchiectasis and health care

• Identifying children for appropriate referral
• Confirm diagnosis and investigate etiology
• Assess severity
• Develop management plan
• Multidisciplinary team approach to chronic care
• Public health issues