



British Paediatric Orphan Lung Diseases (BPOLD)

Bronchiectasis of Unknown Cause - [Dr Adam Jaffe](#)

Consultant and Honorary Senior Lecturer in Respiratory Research Great Ormond Street Hospital for Children and Institute of Child Health. London.

Definition

Bronchiectasis is a pathological description of a disease process that has many possible causes. The characteristic features are abnormally dilated thick-walled bronchi that are inflamed and chronically infected by bacteria. It was previously thought to be irreversible but reports are emerging which challenge this dogma (Eastham 2004).

Causes

The causes of bronchiectasis are multifactorial and include: cystic fibrosis; primary ciliary dyskinesia; gastro-oesophageal reflux; repeat chest infections; inhaled foreign bodies and immunodeficiencies. In some series, no cause has been found in up to 48% of children (Edwards 2003).

Clinical Presentations

Children usually present with sputum production, wheeze or shortness of breath. They may be clubbed. Occasionally radiological changes are detected early in at-risk groups in the absence of symptoms.

Investigations

Computerised tomography of the chest is the gold standard for the diagnosis where dilatation of an airway greater than the accompanying vessel fulfills the radiological criteria for bronchiectasis. Changes may be evident on a chest X-ray but it may not be sensitive enough to detect mild bronchiectasis. Other investigations are aimed at excluding known causes e.g.: sweat test, nasal biopsy, nasal nitric oxide, reflux and aspiration studies and investigations for immunodeficiencies.

Treatment

When no specific cause is found then the treatment approach is similar to the management of pulmonary involvement in cystic fibrosis. Patients should be taught appropriate physiotherapy techniques and exercise encouraged. There should be a low threshold for oral antibiotic use in infective exacerbations. Children with severe bronchiectasis may require regular intravenous antibiotics. Some children will be treated with prophylactic antibiotics. The use of macrolides, such as azithromycin (Jaffe 2001) is increasing due to their potential anti-inflammatory properties. Occasionally, surgical resection is an option if the disease is localised.

Useful references:

The need to redefine non-cystic fibrosis bronchiectasis in childhood

**Eastham KM, Fall AJ,
Mitchell L, Spencer DA**

**Thorax
2004;59:324-327**



British Paediatric Orphan Lung Diseases (BPOLD)

Retrospective review of children presenting with non cystic fibrosis bronchiectasis: HRCT features and clinical relationships	Edwards EA, Metcalfe R, Milne DG, Thompson J, Byrnes CA	Pediatr Pulmonol 2003;36:87-93
Anti-inflammatory effects of macrolides in lung disease	Jaffe A, Bush A	Pediatr Pulmonol. 2001;31:464-73

Web links:

[NELH - Bronchiectasis](#)

[Lung UK](#)