



British Paediatric Orphan Lung Diseases (BPOLD)

Name of Disease:

Post-infectious Obliterative Bronchiolitis (OB)

Clinician:

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Clinical details:

Definition

Clinical Presentation

Obliterative bronchiolitis (OB) is a typically irreversible lung disease that occurs in children after infection of the lower respiratory tract, resulting in bronchial injury. There is concentric narrowing and distortion of the bronchiole walls caused by inflammation and fibrosis. The alveoli are not involved. OB should not be confused with BOOP (Bronchiolitis Obliterans with Organising Pneumonia) from which it differs clinically, radiologically and histologically, and in response to steroids (Ryu 2003).

OB can be mistaken for asthma as some of the presenting symptoms such as cough, wheezing and dyspnoea are similar. OB should be suspected if symptoms persist, exercise intolerance is prolonged and respiratory symptoms are disproportionately severe to chest X-ray findings.

- Evidence gap: There is no standard clinical definition for post infectious bronchiolitis obliterans.
- Suggested definition: In a previously well child, an identified, presumed or confirmed viral respiratory tract infection, is followed by continued increased work of breathing and auscultated crepitations at 28 days following onset with evidence of hypoxaemia (possibly only during sleep). Chest CT demonstrates evidence of patchy gas trapping and bronchial wall injury (thickening or ectasis).

Causes

Adenovirus is the most commonly associated infective aetiology, but other infective agents may also cause the condition.

Diagnosis and investigations

An insidious failure of clinical improvement following a respiratory viral infection is the most common presentation. Work of breathing does not resolve as expected and respiratory crackles (crepitations) and low oxygen saturation persists or are made unduly worse by subsequent minor respiratory infection.

Chest xrays vary significantly but may demonstrate hyperinflation and widespread atelectasis. Tests rule out cystic fibrosis, immune function deficits and gastro-oesophageal reflux. CT chest may be

diagnostic (CT is best performed under general anaesthetic with low dose volumetric acquisition of inspiratory views and then HRCT views at the end of passive expiration – which may be unduly prolonged). CT may reveal "mosaic" shadowing emphasised in expiration, bronchial wall thickening, hyperlucent lung and bronchiectasis (Lynch 1999).

➤ Evidence gap:

- There is no study assessing minimum diagnostic criteria or the consistency of CT findings in bronchiolitis obliterans.

Lung biopsy does not always confirm the diagnosis of OB because of the patchy distribution of disease and diagnosis is usually based on a combination of history, physical examination, chest X-ray, pulmonary function tests and HRCT.

Course

The injury is OB is generally non-progressive (though progression of infective injury may occur). Whilst death may occur early in very severe cases, many children with significant OB continue with stable oxygen requirement and activity levels for many years. In some cases resolution occurs, most often when there is not widespread involvement and associated bronchiectasis is minimal.

Treatment

There are no large scale randomised controlled trials of treatment in OB. Treatment of OB is supportive, and includes antibiotics and physiotherapy plus oxygen therapy, if the patient is hypoxic. Many patients are treated empirically with bronchodilators and corticosteroids and responses may be variable. There have been no published randomised controlled trials of any treatment. Surgery may be an option for a small number of patients with severe localised disease and lung transplantation is an option in end-stage disease. The clinical course is very variable and the prognosis for any individual is often difficult to predict.

- Evidence gap: there is a need for standardised protocols to gather clinical data

Useful references:

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- Colom AJ, Teper AM, Vollmer WM, Diette GB. Risk factors for the development of bronchiolitis obliterans in children with bronchiolitis. *Thorax* 2006; 61:503-506.
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- Yates B, Murphy DM, Forrest IA, et al. Azithromycin reverses airflow obstruction in established bronchiolitis obliterans syndrome. *Am J Respir Crit Care Med* 2005; 172:772-775.

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- Smith KJ, Fan LL. Insights into postinfectious bronchiolitis obliterans in children. *Thorax* 2006; 61:462-463.
- Zhang L, Irion K, Kozakewich H, et al. Clinical course of postinfectious bronchiolitis obliterans. *Pediatr Pulmonol* 2000; 29:341-350.

Web links:

UK Website with information for patients and parents and fundraising

<http://www.breathtakers.co.uk>