The role of flexible bronchoscopy

IM Balfour-Lynn

A Adams

Abstract

Flexible bronchoscopy is an integral part of tertiary paediatric respiratory practice. It has a mainly diagnostic role, from direct observation (anatomy and structure, airway contents, dynamics), and sampling (lavage, biopsy and brushings). There are also some therapeutic roles, mainly re-inflation of an atelectatic segment or lobe. Clinical situations that may require referral for consideration of a bronchoscopy include chronic or recurrent stridor, severe wheeze, chronic cough, recurrent pneumonia, and pneumonia that fails to resolve. Additionally, bronchoscopy is often required in children with chronic suppurative lung disease, patients ventilated in a paediatric or cardiac intensive care unit, and children who have undergone transplantation. Finally its use in research is increasingly improving our understanding of the pathophysiology of asthma and cystic fibrosis.

Keywords bronchoscopy; children

Introduction

Flexible bronchoscopy is an invaluable investigation for respiratory paediatricians in tertiary centres. It is an obligatory part of their training, and in experienced hands, is a valuable and safe procedure to perform. It can also be enjoyable and interesting. However just because we can do it does not necessarily mean we should do it on all our patients, each case must be carefully considered, and contra-indications taken into account (Table 1). Broadly, the indication for a bronchoscopy is when the information required is best obtained (safely) by a bronchoscopy. Risk of complications (Table 2) must be set against potential benefit of the procedure. In many circumstances, a completely normal set of investigations can be as useful as a positive finding for the child, the parents and the clinician. The role of bronchoscopy can be divided broadly into diagnostic and therapeutic, with the former predominating (Table 3). It also has an important role in research, which will be mentioned briefly.

This review will outline various clinical scenarios in which flexible bronchoscopy is indicated, with the aim of helping general paediatricians decide who should be referred for further investigations that may well include a bronchoscopy. It would be unusual for the referring paediatrician to make the referral solely and specifically for a bronchoscopy. That might happen though, in the acute situation when an inhaled foreign body is suspected. When that is the case though, the child needs a rigid bronchoscopy, so should be referred to a paediatric Ear Nose & Throat, or Thoracic surgeon. There are also many indications for a bronchoscopy that will arise in patients who are already under the care of a respiratory paediatrician, and these will be considered later. First, though we discuss clinical scenarios in which referral to a respiratory paediatrician might well lead to a bronchoscopy.

IM Balfour-Lynn *BSc MD MBBS FRCPCH FRCP FRCS(Ed) DHMSA is a Consultant in the Department of Paediatric Respiratory Medicine at the Royal Brompton Hospital, London, UK. Conflicts of interest: none declared.*

A Adams *MBBS(Hons) BSc MRCPCH is a National Grid Trainee in Paediatric Respiratory Medicine in the Department of Paediatric Respiratory Medicine at the Royal Brompton Hospital, London, UK. Conflicts of interest: none declared.*

Contra-indications to flexible bronchoscopy

Absolute

- Airway too small
- Foreign body
- Massive haemoptysis

Relative

- Coagulopathy
- Severe airway obstruction
- Severe hypoxia
- Haemodynamic instability e.g. pulmonary hypertension, cardiac dysrrhythmias

Table 1

Clinical situations that may necessitate a bronchoscopy

Chronic or recurrent stridor

Acute stridor is usually due to viral laryngotracheobronchitis and there is certainly no indication for a bronchoscopy with this

Complications of flexible bronchoscopy

Physiological

- Hypoxia
- Hypercapnia
- Laryngospasm or bronchospasm
- Cardiac arrhythmia and bradycardia
- Cough post-procedure

Infection

- Minor fever in 24 h post-procedure (also due to cytokine release)
- Cross-infection from bronchoscope
- Transient bacteraemia
- Septicaemia if immunocompromised

Mechanical

- Epistaxis
- Laryngeal trauma
- Subglottic oedema
- Haemoptysis post-biopsy
- Pneumothorax (transbronchial biopsy)

Table 2

Indications for flexible bronchoscopy

Clinical

Diagnostic

Look

- Anatomy (upper airways, major bronchi, subsegmental bronchi)
- Structure (mucosal quality, size of lumen)
- Dynamics (movement of glottis, change in luminal size, malacia, pulsatility)
- Contents (mucus plugs, secretions, blood, foreign body)

Sample

- Bronchoalveolar lavage: microbiology, cytology (including staining for lipid and haemosiderin-laden macrophages)
- Brushings: microbiology, cytology
- Biopsy: microbiology, histology

Therapeutic

- Suction/lavage: mucus plugs causing localized atelectasis
- Re-inflate atelectatic lobes/segments of lung
- Instillation of dornase alfa in cystic fibrosis
- Washout: alveolar proteinosis
- Assisted endotracheal intubation/check position of tube

Research

- Bronchoalveolar lavage (microbiology, inflammatory markers)
- Brushings (cell culture, microbiology, cytology)
- Biopsy (mucosal inflammation)

Table 3

diagnosis. However chronic or recurrent stridor may well need further investigation as it usually reflects an obstruction of the extrathoracic upper airways. Flexible bronchoscopy allows an examination of the adenoids, larynx, and hypopharynx in physiological conditions and often while stridor is audible. The child is anaesthetized via a facemask (rather than intubated) and breathing spontaneously, so this is a dynamic study with the laryngeal structure and function observed during inspiration and expiration. Bronchoscopy is not indicated in every infant with stridor, but should be performed in any child with symptoms starting at birth, severe or persistent symptoms, failure to thrive, or if it is associated with hoarseness or leads to oxygen desaturation or apnoeic episodes. A previous history of endotracheal intubation, even if brief such as during postnatal resuscitation, should lower the threshold for a bronchoscopy. Minor trauma of the delicate airway mucosa during an intubation can lead to fibrosis and scarring and the development of subglottic stenosis. If mild, the child may only have symptoms during intercurrent viral illnesses, but can then rapidly develop significant airway obstruction.

Laryngomalacia is the most common congenital laryngeal anomaly and the most frequent cause of persistent stridor in children. Other congenital anomalies of the larynx and extrathoracic trachea causing stridor include tracheomalacia, laryngocoeles, laryngeal webs, and congenital neoplasms such as haemangiomas. Paralysis of the vocal cords produces stridor in infants and children and is usually the result of congenital anomalies of the nervous system. Compression of the upper airway by a vascular ring, e.g. double aortic arch, can also cause stridor. Recurrent stridor in older children is less common but may also be an indication for airway endoscopy. In children with recurrent spasmodic croup, if the episodes are frequent or severe (leading to recurrent hospital admissions), a bronchoscopy is warranted to exclude other diagnoses.

Severe wheeze

The vast majority of infants with episodic viral wheeze or older children with asthma do not need complex investigations. However there are a small number of infants in whom the symptoms, when they occur, are severe and may require intubation and ventilation. A single admission to intensive care would not necessarily warrant a bronchoscopy, unless at the time associated pathology is strongly suspected, but if the child has frequent severe deteriorations then a bronchoscopy is indicated. This allows for the exclusion of associated pathology and congenital malformations, for example vascular rings, malacia of the bronchi or intrathoracic trachea, congenital cysts or webs. Analysis of the nature of the airway inflammation may possibly guide future treatment although therapeutic options are limited. Localized monophonic wheeze may indicate foreign body aspiration. A chest radiograph should have excluded significant hilar lymphadenopathy that can compress an airway and cause focal wheezing (e.g. from tuberculosis, lymphoma).

There are also a relatively small group of older children with problematic severe asthma. These children remain symptomatic (often with frequent admissions which may include periods of mechanical ventilation), despite remaining on high doses of inhaled corticosteroids and often regular oral corticosteroids. The initial consultation may immediately indicate the need for a bronchoscopy, for example if the child produces sputum, we have diagnosed bronchiectasis on a number of occasions, and even cystic fibrosis or primary ciliary dyskinesia in children referred for management of their 'asthma' (Figure 1). When an alternative diagnosis is not obvious, the children need to be categorized into difficult asthma, in which psychosocial and non-adherence issues predominate, and genuine severe therapy-resistant asthma. The Royal Brompton Hospital approach is a staged process; stage 1 includes assessment of allergy, lung function & reversibility, airway inflammation, the home environment, prescription uptake and psychosocial issues by respiratory nurse specialists. From that, around 50% cases go on to stages 2 & 3, which include a flexible bronchoscopy amongst other procedures. Alternative or concomitant airway pathology is excluded, and bronchoalveolar lavage and endobronchial biopsies are performed. The aim is to define the nature of the airway inflammation to enable phenotype-specific therapies to be considered.

Chronic cough

The British Thoracic Society 2008 guideline for the assessment and management of children with chronic cough, defined as cough lasting for more than 8 weeks, have identified situations needing to be excluded by a bronchoscopy. These include:

- suspicion of a foreign body which is best looked at with a rigid scope as the object can then be removed with grasping forceps

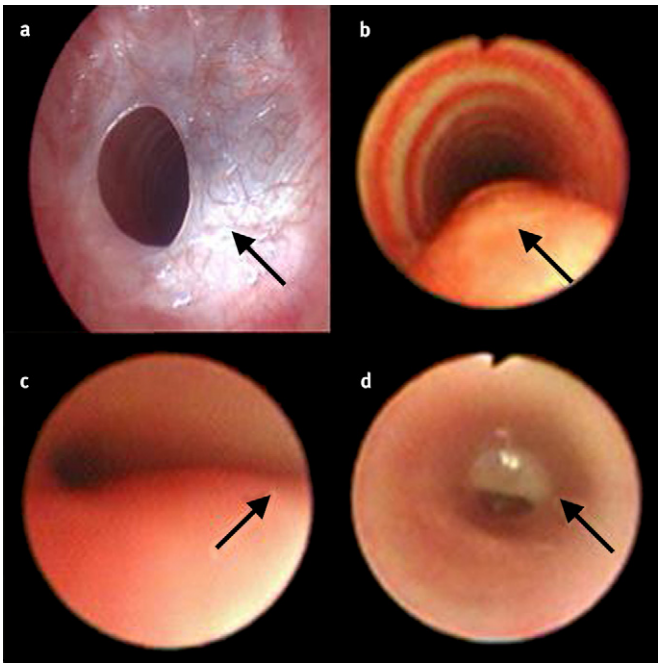

Figure 1 Four cases of problematic asthma with alternative diagnoses made after bronchoscopy — tracheal web blocking most of the tracheal lumen **a,** tracheomalacia **b,** double aortic arch compressing carina and obscuring right main bronchus **c,** primary ciliary dyskinesia with pus in the airway **d.**

- recurrent aspiration — presence of fat-laden macrophages in the lavage fluid may be an indication
- anatomical disorders, including bronchomalacia, tracheo-oesophageal fistula or congenital thoracic malformations.

We believe that if initial investigations are unhelpful in any child with a significant persistent chronic cough, a bronchoscopy is indicated. This is particularly the case if sputum is being regularly produced. We have had a number of cases in which bacteria are isolated from bronchoalveolar lavage fluid, usually *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Moraxella catarrhalis* in a situation where the child is not acutely unwell with pneumonia but has chronic symptoms. Sometimes these children fall into the definition of persistent bacterial bronchitis. Either way a month's course of oral antibiotics such as co-amoxiclav often resolves the symptoms.

Recurrent pneumonia

Every child is allowed to have pneumonia once without being extensively investigated as to why it happened. However a child who has had two or three separate episodes, especially when radiographically-confirmed, and especially if they required hospital admission deserves referral and investigation. These children are not usually the same as those frequently referred from general practice with the label of 'recurrent chest infections', who usually suffer from recurrent viral upper respiratory tract infection with recurrent cough and sometimes wheeze through the winter. Pneumonias recurring in the same lobe — especially the right middle lobe — may indicate an anatomical abnormality of the bronchus (narrowing, isolated bronchomalacia, extrinsic compression). When the infections are in different lobes, it may indicate aspiration, either from severe gastro-oesophageal reflux, primary aspiration from a swallowing abnormality, or an anatomical connection e.g. laryngeal cleft, tracheo-oesophageal fistula. Amongst the investigations, a bronchoscopy must be considered.

Pneumonia that fails to resolve

The majority of cases of community-acquired pneumonia resolve without complications. The British Thoracic Society 2002 guideline recommends a follow up chest radiograph in children with lobar collapse, an apparent round pneumonia, or for continuing symptoms. In the presence of persistent radiographic changes (especially atelectasis) and significant symptoms, a bronchoscopy may be warranted, and may be both diagnostic and therapeutic (Figure 2). If there is lobar collapse due to purulent secretions or a thick mucus plug, the suctioning performed during bronchoscopy can aid airway clearance; subsequent physiotherapy should lead to full re-expansion of the segment or lobe. Unfortunately if the lobe has been down for a prolonged time, it is not always possible to re-inflate it. Lavage of the lower airways may lead to isolation of the infecting organism, especially if atypical or resistant to the antibiotics the child has received, but more usually it is negative for bacteria if the child has had a prolonged antibiotic course. Occasionally we have found other reasons for failure of resolution, such as a narrowed bronchus, endobronchial tuberculosis, endobronchial polyps or tumour, or an unexpected foreign body. Persistent pulmonary infiltrates on radiographs are also indications for a bronchoscopy.

An indication for a bronchoscopy during the acute stages of pneumonia is in a child with particular underlying problems, where an urgent microbiological diagnosis is required to exclude atypical bacterial and fungal opportunistic infections. These include immunocompromised children, either from a primary immune problem or those undergoing chemotherapy. The other main group is those in an intensive care with suspected ventilator-associated pneumonia (see below).

Children already under the care of a respiratory paediatrician

Chronic suppurative lung disease

Children with CSLD principally include those with cystic fibrosis (CF), and non-CF bronchiectasis. Bronchoscopies are not used to clean out the lungs in children with CF, as it is pointless performing a prolonged procedure suctioning out thick viscid secretions that immediately reappear. However there is a role when there is focal lobar or segmental collapse as it may be

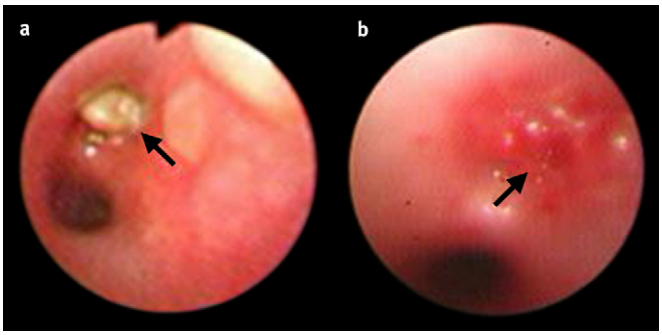

Figure 2 Two cases of persistent atelectasis following apparent community-acquired pneumonia. Bronchoscopy revealed the causes were due to airway blockage with distal collapse, from a plaque of endobronchial tuberculosis **a,** and a spindle cell tumour **b.**

possible to remove a mucus plug that is blocking a major bronchus causing distal collapse. After direct suctioning, dornase alfa can be instilled into the area and it is helpful if a physiotherapist is present during the procedure.

Another indication in CSLD (usually in CF patients) is to obtain microbiological diagnosis in young children, particularly looking for a first growth of *Pseudomonas aeruginosa*. Generally, patients do not expectorate until at least 6–7 years old and even then many CF children do not like coughing out sputum. Cough swabs are useful when positive, but a negative growth does not necessarily mean that there are no pathogenic organisms in the lower airways. If a young child is not as well as we would expect, we have a low threshold for performing a bronchoalveolar lavage. We also find it useful to perform a bronchoscopy in newly diagnosed CF patients, both after a symptomatic diagnosis and newborn screening. We also carry out a pH study at the same time to diagnose gastro-oesophageal reflux, which is common in CF infants. Finally, older CF children with intractable wheeze may benefit from a bronchoscopy to exclude tracheobronchomalacia.

Paediatric intensive care unit (PICU)

Bronchoscopy in a PICU is more hazardous due to haemodynamic instability or coagulopathies, but has an important role in a number of circumstances.

- **Ventilator-associated pneumonia** can be difficult to diagnose. Clinical and chest radiograph signs may be non-specific, and there are many other causes of infiltrates: for example, pulmonary oedema, aspiration and segmental mucus plugging. The trachea is usually colonized by gram-negative rods shortly after ventilation so lower airway cultures are often contaminated. Using a bronchoscope for bronchoalveolar lavage or a protected specimen brushing is worthwhile, and a non-bronchoscopic bronchoalveolar lavage also has a role to play.
- **Assessment of airway patency**, particularly in children with congenital heart disease is frequently necessary due to the potential for extrinsic compression of the airway from vascular structures and stents. It is also often necessary after cardiac surgery, as localized tracheomalacia or bronchomalacia may be apparent once a vascular structure has been moved off the airway.
- **Assessing patency of an endotracheal or tracheostomy tube** is critical in a ventilated child with a sudden deterioration. Even when a suction catheter passes easily, the tube may be blocked. This can be seen at bronchoscopy and remedied. If there is any doubt, it is usually simpler to change the tube, but there may be reluctance to do this if re-intubation is likely to be difficult.
- **Difficult intubations** are not too common, but there are certain patients in whom it can be problematic. These include an unstable cervical spine, midfacial disease (for example craniofacial syndrome), or mandibular hypoplasia (Treacher Collins syndrome or Pierre Robin sequence). Intubation can then be performed by 'railroading' the endotracheal tube over the bronchoscope, so that intubation takes place under direct vision.
- **Selective endotracheal intubation** may be indicated if there is unilateral pathology necessitating different ventilatory strategies: for example, a unilateral uncontrolled air leak caused by a bronchopulmonary fistula or by barotrauma in a ventilated neonate. This procedure is technically much easier if the longer left main bronchus is to be intubated; selective right-sided intubation risks occluding the right upper lobe bronchus or leaving an unstable tube position.
- **Airway stent assessment** is an infrequent indication as stents are not commonly used in children. Nevertheless in selected cases a stent may offer a good alternative to prolonged ventilation. Stent placement is a surgical procedure usually done with a rigid bronchoscope, but after it is inserted, the position and the state of any distal malacia can be checked using flexible bronchoscopy. Post-stent endobronchial washout may need to be performed repeatedly due to impairment of mucociliary clearance.
- **Post-extubation stridor** is not uncommon, especially after prolonged ventilation. Bronchoscopy is indicated if there has been at least one failed extubation apparently caused by upper airway obstruction, especially if corticosteroids have been given. The differential diagnosis includes airway oedema, scarring, subglottic stenosis, and malacia (either acquired as a complication of prolonged ventilation, or resulting from pre-existing disease).

Transplantation

In post-transplant patients, pulmonary symptoms are common and may be due to sepsis or graft versus host disease (GVHD). The CT chest scan may not differentiate between GVHD and infection due to fungi or atypical mycobacteria. Bronchoalveolar lavage is mandatory, and mucosal biopsies should be taken as this may aid cytomegalovirus and fungal diagnoses. Transbronchial biopsy is the method of choice for diagnosing GVHD. In children who have had a lung transplant, bronchoscopies are routinely performed to assess airway patency, obtain a microbiological diagnosis, and assess rejection with a transbronchial biopsy.

Research

Research into the basic mechanisms of respiratory disease in children is critical to our understanding of disease processes, which may lead to new therapeutic options. For ethical reasons, a bronchoscopy cannot be performed in children solely for research purposes (unlike in adults). Opportunities must therefore be taken to use clinically indicated procedures as a means of obtaining material for research. Such procedures must, however, constitute minimal risk to the child, and in our experience, procedures such as endobronchial biopsy or mucosal brushings have not proven to be a problem, as procedures are performed by senior, experienced bronchoscopists. Using some surplus bronchoalveolar lavage fluid for research purposes is also perfectly acceptable. In addition, bronchoscopy can be useful in airway surface measurements, in particular airway surface liquid composition and potential difference. The latter may also be helpful when there are difficulties diagnosing CF in a child too young to tolerate nasal potential difference measurements. ◆

FURTHER READING

Balfour-Lynn IM, Spencer H. Bronchoscopy — how and when? *Paed Resp Rev* 2002; **3:** 255–64.

Bracken M, Fleming L, Hall P, et al. The Importance of nurse-led home visits in the assessment of children with problematic asthma. *Arch Dis Child* 2009; **94:** 780–4.

Bush A. Bronchoscopy in paediatric intensive care. *Paediatr Respir Rev* 2003; **4:** 67–73.

Bush A. Primum non nocere: how to cause chaos with a bronchoscope in the ICU. *Chest* 2009; **135:** 2–4.

De Blic J, Midulla F, Barbato A, et al. Bronchoalveolar lavage in children. ERS Task force on bronchoalveolar lavage in children. European Respiratory Society. *Eur Respir J* 2000; **15:** 217–31.

Hilliard TN, Sukhani S, Francis J, et al. Bronchoscopy following the diagnosis of cystic fibrosis. *Arch Dis Child* 2007; **92:** 898–9.

Jaggar SI, Haxby E. Sedation, anaesthesia and monitoring for bronchoscopy. *Paediatr Respir Rev* 2002; **3:** 321–7.

Midulla F, de Blic J, Barbato A, et al. Flexible endoscopy of paediatric airways. *Eur Respir J* 2003; **22:** 698–708.

Molina-Teran A, Hilliard TN, Saglani S, et al. Safety of endobronchial biopsy in children with cystic fibrosis. *Pediatr Pulmonol* 2006; **41:** 1021–4.

Payne D, McKenzie SA, Stacey S, et al. Safety and ethics of bronchoscopy and endobronchial biopsy in difficult asthma. *Arch Dis Child* 2001; **84:** 423–6.

Regamey N, Balfour-Lynn I, Rosenthal M, et al. Time required to obtain endobronchial biopsies in children during fiberoptic bronchoscopy. *Pediatr Pulmonol* 2009; **44:** 76–9.

Regamey N, Hilliard TN, Saglani S, et al. Quality, size, and composition of pediatric endobronchial biopsies in cystic fibrosis. *Chest* 2007; **131:** 1710–7.

Saglani S, Nicholson AG, Scallan M, et al. Investigation of young children with severe recurrent wheeze: any clinical benefit? *Eur Respir J* 2006; **27:** 29–35.

Suri R, Balfour-Lynn IM. When to do a flexible bronchoscopy. *Curr Paediatr* 2004; **14:** 306–12.

Practice points

Infants with chronic stridor – when to consider a bronchoscopy

- Symptoms from birth
- Severe or persistent symptoms
- Episodes of oxygen desaturation
- Apnoeic episodes
- Failure to thrive
- Hoarseness
- Previous endotracheal intubation

Indication for rigid bronchoscopy (rather than flexible)

- Foreign body removal
- Severe haemoptysis
- Hypoxia (allows ventilation through the scope)
- Small airway (allows ventilation down the 2.5 mm internal diameter rigid scope)
- Viewing the posterior aspect of the larynx or upper trachea (for H-type tracheo-oesophageal fistula, laryngeal cleft, bilateral abductor vocal cord paralysis)
- Interventional bronchology (lasers, stents)

Therapeutic flexible bronchoscopy

- Re-inflation of lobar atelectasis by suctioning (& use of dornase alfa in CF)
- Difficult intubation – railroading endotracheal tube over bronchoscope
- Selective endotracheal intubation of left or right main bronchus
- Alveolar filling disorders e.g. alveolar proteinosis or lipid aspiration – large volume lavage