Respiratory Medicine
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Stephanie D. Davis
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Diagnostic Tests in Pediatric Pulmonology

Applications and Interpretation

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Preface

Over the past 20 years, diagnostic tests for pediatric pulmonologists have revolutionized care of children afflicted with respiratory disorders. These tests have been used to help not only in diagnosis but also in the management and treatment of these children. Bronchoscopic, imaging, and physiologic advances have improved clinical care and have also been used as outcome measures in research trials.

The aims of this book are to (1) describe the various diagnostic modalities (especially the newer ones) that are available for the evaluation of pediatric respiratory disorders; (2) understand the advantages and limitations of each test so that the clinician may choose the most appropriate modality; and (3) describe how best to interpret the key findings in a variety of tests as well as the possible pitfalls in interpretation.

The book focuses on the main diagnostic modalities used in the evaluation of pediatric patients with respiratory disorders and presents up-to-date information on a number of tests that are used for a variety of conditions encountered in the practice of pediatric pulmonology. The clinical applications of the tests are highlighted within each chapter.

The book contains 14 chapters written by 30 authors; the authors are both young pediatric pulmonologists who are emerging as leaders in our field as well as well-known international experts.

Target readers are practicing clinicians including pediatric pulmonologists, intensivists, pediatricians, and primary care practitioners. Other readers may include trainees, respiratory therapists, nurses, radiologists, and clinical researchers.

We would like to thank the staff at Springer, especially Maureen Alexander and Amanda Quinn, for endorsing and editing the book. We especially would like to thank our expert authors for writing such detailed and outstanding chapters. Finally, we would like to thank our families for their continual love, support, and encouragement during this endeavor.

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Chapter 1
The Evaluation of the Upper and Lower Airways in Infants and Children: Principles and Pearls from Four Decades in the Trenches

Robert E. Wood

Abstract Diagnostic bronchoscopy is an often underutilized technique in pediatric patients. However, with proper equipment, appropriate technical and cognitive skills, and effective and careful attention to safety and comfort, bronchoscopy can be a powerful tool for the pediatric pulmonologist. This review is a distillation of the author's four decades of experience.

Keywords Flexible bronchoscopy • Airway dynamics • Sedation/anesthesia for pediatric bronchoscopy • Airway management for pediatric flexible bronchoscopy • Indications for pediatric flexible bronchoscopy • Complications of pediatric flexible bronchoscopy • Techniques for pediatric flexible bronchoscopy • Clinical utility of pediatric flexible bronchoscopy

Bronchoscopy is a powerful diagnostic and therapeutic tool for the evaluation and management of children with pulmonary or airway issues. During the 1970s, dramatic progress was made in the development of instrumentation suitable for pediatric bronchoscopy, including the glass rod telescope for rigid instruments and a flexible bronchoscope small enough to be safely used in children. Over the ensuing nearly four decades, further progress has been made in instrumentation as well as experience in the utilization of these instruments.

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The discussion in this chapter is predicated on the assumption that the operator will be equipped with the proper equipment (which is properly cleaned and prepared for use in the patient), trained assistants, a proper venue, appropriate provision for sedation/anesthesia and monitoring of the patient's physiologic status, and a plan for safe recovery from the sedation, and that the parents/guardians have provided appropriate informed consent.

This chapter is primarily a distillation of my personal experience over the past four decades of spelunking in the pediatric airways. The views expressed are mine, and are based on more than 20,000 procedures. I have made (and learned from) many mistakes … my practices and perspective have evolved over this time.

Principles

There are four criteria for successful bronchoscopy: (1) safety, (2) safety, (3) comfort, and (4) achieving the correct diagnosis or result.

Other than death of the patient, the most serious complication of a bronchoscopy is to have done the procedure but obtained the wrong diagnostic or therapeutic result.

Match the instrument(s) to the patient and purpose of the procedure.

Be aware of the effect of sedation level and body position as well as the effect of the instrument itself and techniques utilized for airway management on the visualized anatomy and airway dynamics.

The airways begin at the nostril ….

Children often have more than one significant airway abnormality—examine the entire airway unless contraindicated.

"WNL" too often really means, "We Never Looked."

The endoscopic findings must be interpreted *in the context of the patient's history*—some things that look bad may not be physiologically important and may be the result of the sedation or conditions under which the examination is performed. Or vice versa ….

Stridor is always visible.

Every bronchoscopic procedure performed in children should be recorded so that the video record can be examined again when necessary.

Indications for Procedures

There are only two indications for bronchoscopy in children, diagnostic and therapeutic. Diagnostic bronchoscopy is indicated when there is information in the lungs or airways of the child, necessary for the care of the child, that is best obtained with a bronchoscope. Similarly, therapeutic bronchoscopy is indicated when it is the best way to achieve the necessary therapeutic goals. The specific indications for bronchoscopy will vary considerably among different institutions, as there will inevitably be wide variation in the patient populations.

A Basic Philosophy of Bronchoscopy

No one knows what lurks in the airways of a child, and surprises abound. The bronchoscopist must be careful to examine the entire airway in each patient, unless there is a very good reason not to do so. For example, an intubated immunosuppressed patient who is thrombocytopenic does not need to have a scope passed through the nose unless in search of specific pathology, as there is more risk than benefit involved.

The bronchoscopist must adopt a surgical mentality—you are sent to drain the swamp, not merely to survey and report back—i.e., take care of things that can be taken care of …. Discovering and then simply reporting the finding of a mucus plug is not enough—remove the mucus plug if it is possible/reasonable to do so. Every diagnostic bronchoscopy has the potential to also be a therapeutic procedure. Likewise, every therapeutic bronchoscopy includes a diagnostic component. When a flexible bronchoscope is employed to facilitate a difficult intubation, for example, the operator should recognize and report the abnormal anatomy or other factors that make the intubation difficult; otherwise, a golden opportunity may be missed, and the patient may be forced to undergo yet another procedure.

The goals of bronchoscopy are to evaluate the airway anatomy, dynamics, and contents, to obtain appropriate specimens for further analysis (as indicated), to relate the findings to the patient's history and clinical context, and to improve the patient's clinical status when feasible. When contemplating a bronchoscopy, the assessment of risk must also include the risk of *not* doing the bronchoscopy.

Bronchoscopy is a visual procedure—the work product is primarily *images*. Every procedure should be recorded for review at some later point in time—this will improve the quality of patient care, facilitate teaching (parents, patients, and medical trainees), and reduce the potential for medicolegal liability. Written documentation is also important, and should include enough descriptive language to enable the reader to develop a reasonably accurate picture of what was actually seen and done.

Instruments

Diagnostic and/or therapeutic bronchoscopy may be done with either rigid or flexible instruments, and in many cases, either instrument will suffice for the patient's immediate need. However, there are clearly indications for which a rigid instrument is much more suitable, and some for which a flexible instrument is more suitable. Additionally, for the adequate evaluation of some pediatric patients, utilization of *both* rigid and flexible instruments may be necessary.

A bronchoscope must be small enough to safely traverse the airway of the patient. The most common flexible instrument utilized in pediatric patients today has an outer diameter of 2.8 mm, and this instrument can be safely used in children as small as approximately 600 g (although in children smaller than about 1,200 g, great care must be taken to ensure adequate ventilation or very rapid completion of the procedure). This instrument (and its predecessor, which is approximately 3.7 mm)

has a 1.2 mm suction channel; this limits the devices that can be passed through the channel. Instruments with a larger suction channel can be used in older children, and may be necessary when airway secretions are extremely thick or instrumentation is necessary.

Rigid instruments utilize a glass rod telescope, which produces an image with extremely high resolution. Rigid bronchoscopes and telescopes are available in a variety of sizes. A major limitation of rigid instrumentation is that it is necessary to pass the instrument through the mouth, extending the neck and elevating the mandible. This may not be possible in all patients, and, in any event, will distort the anatomy and airway dynamics.

The traditional techniques for flexible bronchoscopy involve transnasal passage, thus enabling examination of the entire airway, and placing minimal traction on airway structures, giving the most effective visualization of airway dynamics. However, transnasal passage means that the tip of the instrument must be flexed forward to view and enter the larynx (Fig. 1.1), making evaluation of the posterior aspects of the larynx much more difficult. It can be virtually impossible to diagnose a laryngoesophageal cleft, for example, with a flexible bronchoscope. A rigid bronchoscope, on the other hand, approaches the larynx from a very different angle (Fig. 1.1), and is the instrument of choice for evaluation of the anatomic details of the larynx, and especially the posterior commissure. Children suspected of aspiration should in most cases be evaluated with both rigid and flexible instruments in order to definitively ensure that there is no laryngoesophageal cleft or "H-type" TE fistula.

Sedation, Anesthesia, and Airway Management for Flexible Bronchoscopy in Children

It is possible to examine a child's airway without sedation. The most common setting for this approach is a simple evaluation of the nasopharyngeal airway and larynx in an office setting, including the endoscopic assessment of swallowing. Most children do not like this, and it may be difficult for the operator as well. However, full assessment of vocal cord function may require this approach. When the bronchoscope needs to be passed beyond the glottis, it is much wiser and safer to provide sedation for pediatric patients.

In the early days of pediatric flexible bronchoscopy, most procedures were done with sedation provided by the bronchoscopist. Today, most procedures are performed with the aid of an anesthesiologist, and this is very appropriate, in order to enhance safety; it also enables the use of agents that are generally restricted to use by anesthesiologists and can provide a more smooth and comfortable evaluation. However, choice of the wrong technique for sedation is one of the easiest ways to achieve the wrong diagnosis. Sedation that is too deep may mask dynamic pathology, and sedation that is not sufficiently deep may increase the risk of complications and possibly lead to termination of the procedure before the answer has been obtained. It is vitally important that the bronchoscopist and the person responsible

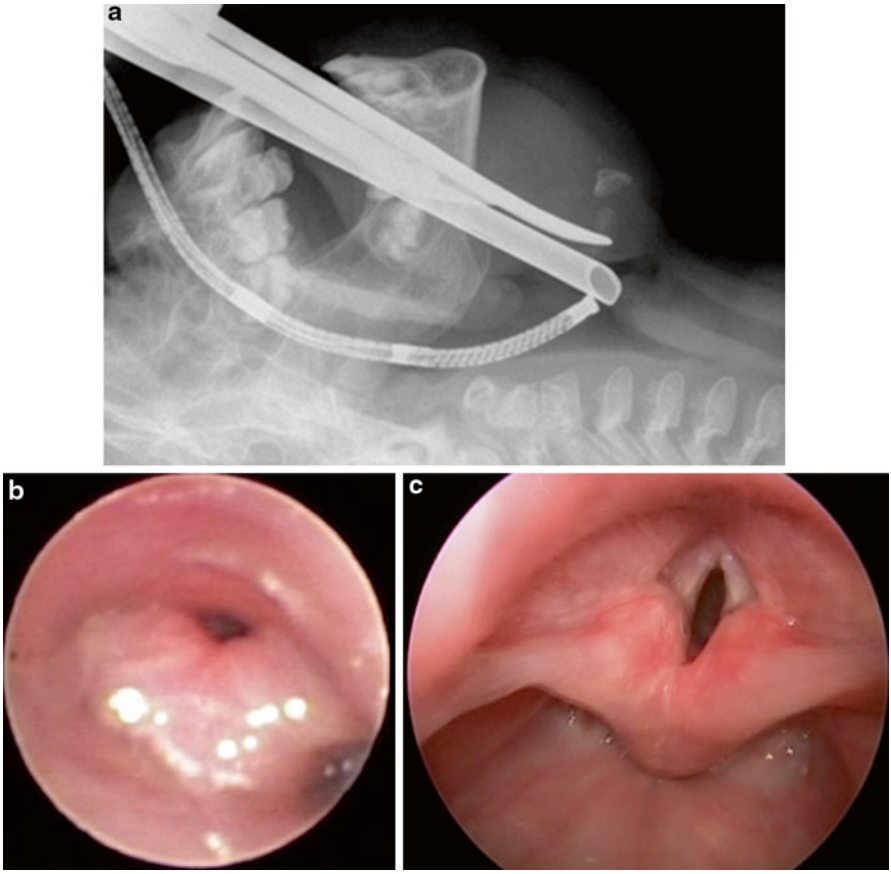

Fig. 1.1 Flexible and rigid instruments approach the larynx from very different perspectives. A rigid instrument necessarily elevates the hyoid and tongue base, lifting and distorting the larynx, while at the same time allowing more detailed anatomic evaluation as well as manipulation of the tissues under direct vision. The flexible instrument, on the other hand, approaches from behind, and is much more suitable for evaluation of laryngeal dynamics. When there is any suspicion of posterior laryngeal pathology (laryngoesophageal cleft, for example) both instruments may need to be employed in order to obtain a full understanding of the laryngeal anatomy and dynamics. (**a**) Lateral radiograph showing the path taken by rigid and flexible instruments. Note the elevation of the hyoid and tongue base by the rigid instrument and the angle of approach to the larynx by both instruments (this is not the same patient as in **b** and **c**). (**b**) The larynx of a child with a history of inspiratory stridor, seen by a flexible instrument. There is no traction on the larynx, and in this view, the mucosa overlying the arytenoid cartilages completely obscures the view of the glottis, and produces significant inspiratory obstruction. (**c**) The same larynx as seen by a rigid instrument. The larynx is being elevated by a rigid laryngoscope. The mucosa, which through the flexible instrument looked redundant and possibly edematous, now looks anatomically normal, and there is no obvious obstruction

for the sedation and monitoring of the child both have an adequate understanding of the purpose of the procedure and that they communicate effectively before, during, and after the procedure. It is often useful to change the level of sedation during the course of an examination. For example, a deeper level of sedation at the beginning may facilitate the anatomic evaluation and collection of specimens, while lightening the sedation near the end of the procedure may facilitate documentation of abnormal airway dynamics.

The precise techniques utilized for sedation of children for bronchoscopic procedures is as much a matter of personal preference as anything, as long as the patient is safe and the goals of the procedure are adequately met. Mask induction followed by establishment of intravenous access and maintenance with a short-acting parenteral drug can be a very effective technique.

Pediatric bronchoscopy is certainly among the most challenging tasks an anesthesiologist is called upon to perform. As bronchoscopists, we violate virtually every principle that anesthesiologists hold near and dear: we want control of the airway, we want to see the airway obstruct (at least, long enough for us to be able to understand why the child's airway is obstructing), and we often want to see the child cough (so that we can see lower airway dynamics). Modern bronchoscopes employ digital display, and it is very helpful for the anesthesiologist to be able to visualize what the bronchoscopist is seeing. This does not in itself suffice for effective communication between the bronchoscopist and the anesthesiologist.

Airway management can be one of the most contentious issues between the anesthesiologist and the bronchoscopist. Typically, the child is placed under light anesthesia so that spontaneous ventilation is maintained, and an oral airway is placed. The bronchoscope is then inserted through one nostril. However, the presence of the oral airway distorts the anatomy, and it often needs to be removed, at least temporarily, while the upper airway anatomy and dynamics are assessed. Once this is accomplished, the oral airway can be reinserted and the bronchoscope directed into the lower airways. The bronchoscopist should evaluate the position of the oral airway (in many cases, the oral airway may actually push the posterior tongue over the larynx, obstructing, rather than opening, the airway). It is quite effective (so long as the patient is breathing spontaneously) to insert an endotracheal tube into the oral airway to provide for delivery of oxygen and anesthetic gas directly to the larynx (Fig. 1.2).

Many bronchoscopists and anesthesiologists routinely perform their procedures through a laryngeal mask airway (LMA). While this is easy, and allows positive pressure ventilation from start to finish, there are many reasons to condemn this as a routine practice. An LMA completely bypasses the nasopharyngeal airway, and many diagnoses will be missed. The LMA also presses against the post-cricoid region of the larynx, and can interfere with vocal cord motion; it also can put downward traction on the post-cricoid mucosa, making it impossible to adequately diagnose some forms of laryngomalacia (see Figs. 1.3 and 1.4). An LMA does not prevent laryngospasm or even, necessarily, aspiration of oral secretions. When positive pressure support is given through the LMA, it can be impossible to adequately evaluate tracheomalacia or bronchomalacia. On the other hand, there are clearly some circumstances where the use of an LMA may be appropriate and effective;

Fig. 1.2 Placement of a shortened RAE tube into the oral airway allows insufflation of oxygen and anesthetic gas and does not obstruct the space above the patient's face (which can interfere with manipulation of the flexible bronchoscope)

Fig. 1.3 How an LMA can lead to erroneous diagnoses. The *first panel* shows the larynx of a child with MPS II with an LMA in place. The patient has a history of significant stridor, but through the LMA, the larynx does not look too abnormal (and no stridor could be heard). The LMA was removed; the *second panel* shows hypopharyngeal collapse (this photo does not show the full extent of the collapse, which was complete). The *third panel* shows the larynx with mandibular lift; the mucosa overlying the post-cricoid area is redundant, and the arytenoids are large. The *final panel* shows the dramatic inspiratory prolapse of the arytenoid mucosa when mandibular lift was relaxed. The LMA did not allow evaluation of the supraglottic airway, and the traction on the post-cricoid mucosa created by the tip of the LMA in the upper esophagus made it impossible to appreciate the laryngomalacia

these primarily involve situations in which there is no clinical concern about the upper airway anatomy or dynamics, and the child may be too small to utilize an endotracheal tube with the flexible bronchoscope. However, as the 2.8 mm flexible bronchoscope can readily and safely be used through a 3.5 mm endotracheal tube, there are relatively few situations in which this may be the technique of choice. If the bronchoscopist feels strongly that an LMA is essential to safe and effective

Fig. 1.4 The laryngeal mask airway can be useful, but it is inappropriate to employ the device for every procedure as the primary technique for airway management

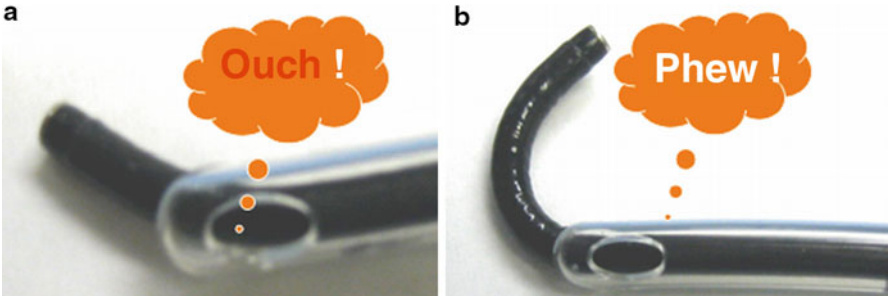

Fig. 1.5 Care must be taken when passing a flexible bronchoscope through an artificial airway (endotracheal or tracheostomy tube) to not flex the tip of the instrument until the bending segment has passed beyond the end of the tube. Otherwise, the bronchoscope can be damaged

evaluation of the *lower* airways, then very serious consideration should be given to an evaluation of the *upper* airways without the presence of the LMA. If this is done as the last step in the global procedure, then there will be less chance for contamination of the BAL specimens with upper airway secretions, and can be done as the patient recovers from the sedation.

It is often necessary or desirable to perform a flexible bronchoscopy through an endotracheal tube. Care must be taken to ensure that the tube is adequately lubricated (otherwise, manipulation of the bronchoscope may be difficult, or the bronchoscope may be physically damaged). Care must also be taken to ensure that the tip of the flexible bronchoscope extends far enough beyond the end of the endotracheal (or tracheostomy) tube before the tip is flexed; attempting to flex the tip of the scope while the bending segment of the instrument is still within the confines of the tube can result in breaking the control wires (Fig. 1.5).

A 2.8 mm bronchoscope can be safely utilized through a tube that is only 3.5 mm in diameter. However, this will result in a high level of obstruction to airflow through the tube. It is much easier to force air through the tube and into the lung than for the air to passively escape, and if there is not a sufficient leak around the outside of the