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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ISSN 2197-7372
ISBN 978-1-4939-1800-3
DOI 10.1007/978-1-4939-1801-0
Springer New York Heidelberg Dordrecht London

ISSN 2197-7380 (electronic)
ISBN 978-1-4939-1801-0 (eBook)

Library of Congress Control Number: 2014951680

Printed on acid-free paper

Humana Press is a brand of Springer
Springer is part of Springer Science+Business Media (www.springer.com)

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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S.D. Davis et al. (eds.), *Diagnostic Tests in Pediatric Pulmonology*,
Respiratory Medicine, DOI 10.1007/978-1-4939-1801-0_1

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

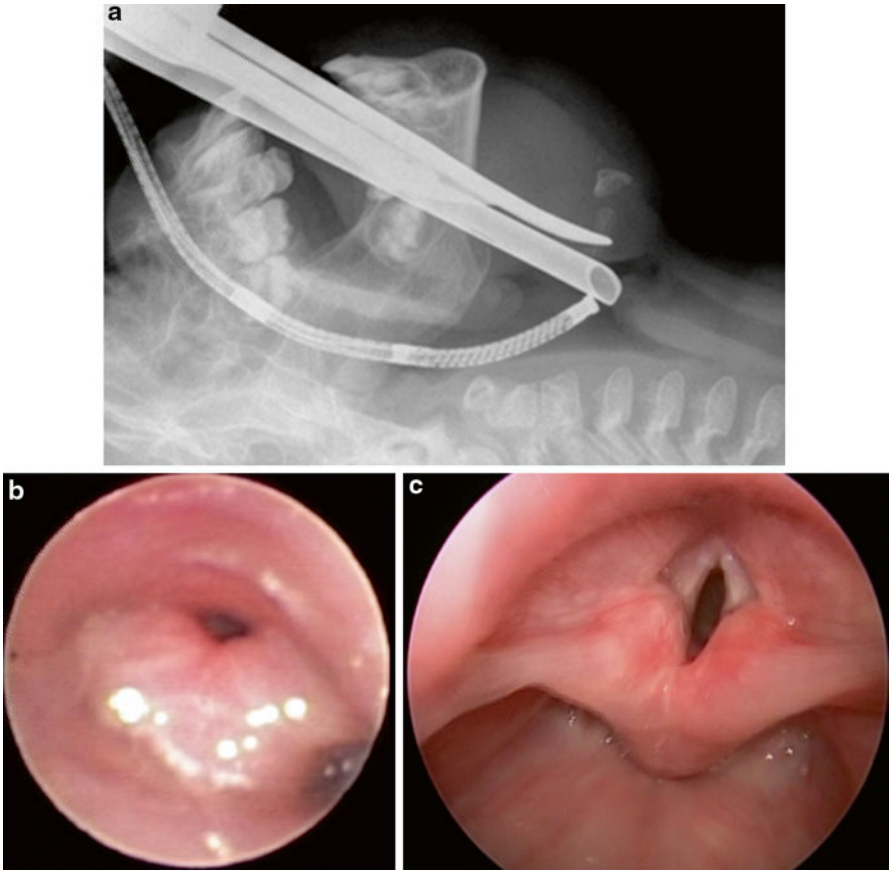

Fig. 1.1 Flexible and rigid instruments approach the larynx from very different perspectives. A rigid instrument necessarily elevates the hyoid and tongue base, lifting and distorting the larynx, while at the same time allowing more detailed anatomic evaluation as well as manipulation of the tissues 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;

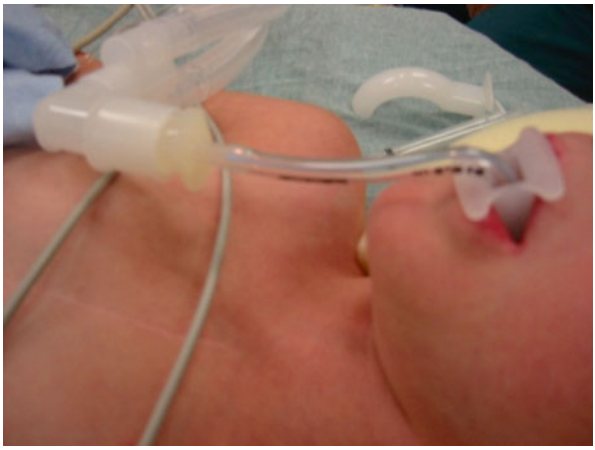

Fig. 1.2 Placement of a shortened RAE tube into the oral airway allows insufflation of oxygen and anesthetic gas and does not obstruct the space above the patient's face (which can interfere with manipulation of the flexible bronchoscope)

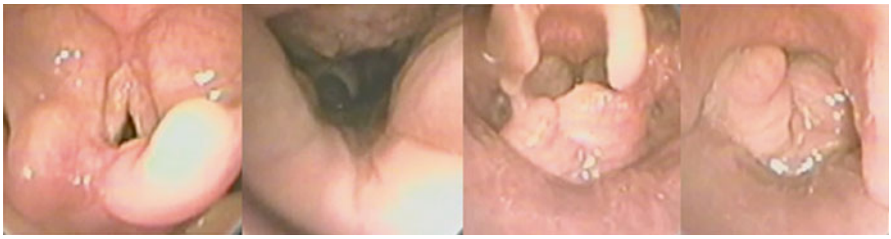

Fig. 1.3 How an LMA can lead to erroneous diagnoses. The *first panel* shows the larynx of a child with MPS II with an LMA in place. The patient has a history of significant stridor, but through the LMA, the larynx does not look too abnormal (and no stridor could be heard). The LMA was removed; the *second 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

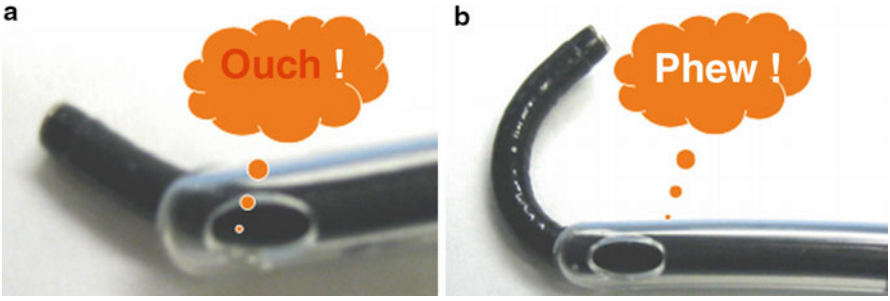

Fig. 1.5 Care must be taken when passing a flexible bronchoscope through an artificial airway (endotracheal or tracheostomy tube) to not flex the tip of the instrument until the bending segment has passed beyond the end of the tube. Otherwise, the bronchoscope can be damaged

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

tube, a very high level of airway pressure ("inadvertent PEEP") can develop, even leading to a tension pneumothorax. Conversely, excessive suctioning when the instrument is passed through a relatively small tube can result in a dramatic decrease in the patient's functional residual capacity and rather impressive oxygen desaturation can result. This is generally easily managed, however, by removing the bronchoscope and applying positive pressure ventilation through the endotracheal or tracheostomy tube (an alveolar recruitment maneuver is often most beneficial).

Techniques for Flexible Bronchoscopy

In the majority of diagnostic flexible bronchoscopies, it is desirable to obtain a specimen (bronchoalveolar lavage "BAL") for cytologic and microbiologic analysis. This specimen should be representative of the state of the lungs prior to the procedure—it is therefore important to minimize the risk of aspiration of oral secretions before the specimen can be obtained. The nose and hypopharynx should be gently suctioned prior to inserting the bronchoscope. Continuous insufflation of oxygen (~2 L/min) through the suction channel of the bronchoscope during passage through the nose and to the larynx can minimize contamination of the suction channel. Application of topical lidocaine to the larynx, while essential, also immediately leads to the risk of aspiration. Employing a small volume (0.5 mL) can help minimize this. However, when a patient is lying supine, the carina is at an approximately 30° downhill position from the larynx, and it is very common to visualize secretions draining from the mouth towards the carina as the bronchoscope is initially inserted. Suctioning of these secretions will of course contaminate the instrument and therefore the subsequent specimen, as will delay in obtaining the BAL specimen (Fig. 1.6)

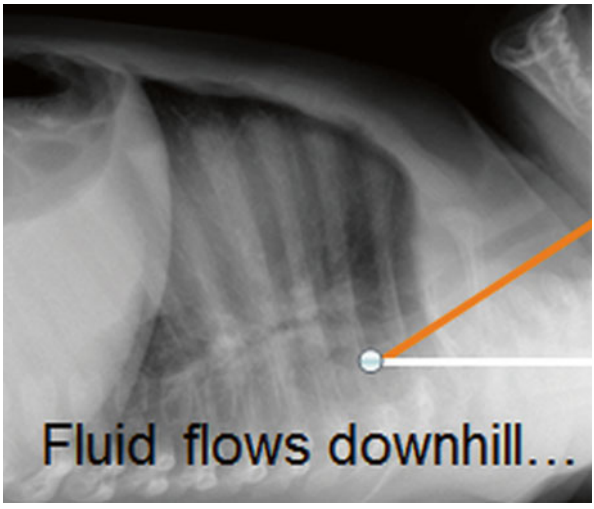

Fig. 1.6 Secretions readily drain from the larynx to the carina when the patient is supine. This often results in contamination of BAL specimens by oral secretions

If the proposed site for BAL specimen collection can be determined beforehand, it can be very helpful to immediately go to this site and perform the BAL; after the specimen has been obtained, one can aspirate secretions, either endogenous or aspirated, in order to evaluate the remaining airway anatomy, etc. Sometimes, however, there will be no clue in advance, and a very quick inspection of the bronchial anatomy (which should take no more than ~10 s once the tip of the bronchoscope reaches the carina, assuming that there are minimal secretions present, so that the anatomy can be clearly seen) can inform the site selection.

When it is important to obtain a BAL specimen with absolutely minimal risk of contamination by upper airway secretions, the most effective technique is to electively intubate the patient without placing any topical anesthetic on the larynx and then pass the bronchoscope through the clean endotracheal tube. After obtaining the BAL specimen, the endotracheal tube can be removed (if desired) and the anatomic (and dynamic) evaluation can then be completed. When I utilize this technique, in patients in whom I expect a sterile BAL specimen, the culture is indeed sterile more often than not.

When there is diffuse lung disease, the precise location from which a BAL specimen is obtained may be relatively unimportant. However, site selection can be a serious concern. For example, if there is a peripheral lesion seen on chest radiography, one may wish to sample that specific area. It is easy to sample a different area than the one intended, by simply passing the bronchoscope too far distally, and missing the bronchus leading to the intended target. This is an especially insidious problem when one is utilizing a smaller diameter instrument in a larger patient. Even one bronchial generation, which can be only 2–3 mm, can make a difference (Fig. 1.7).

There have been attempts to "standardize" BAL technique, with the goal of achieving a consistent dilution of alveolar lining fluid (ALF) components in the specimen obtained. However, it makes no rational sense to specify the aliquot volume (for example, xx mL/kg or xx ml/100 mL estimated FRC) unless the size of the bronchoscope and the bronchial generation number into which the tip of the instrument will be gently wedged are also specified (see Fig. 1.7). Almost by definition, each bronchial generation reduces the volume of lung being sampled beyond the tip of the bronchoscope by half, thereby potentially doubling the concentration of ALF constituents in the resulting specimen.

When airway dynamics are an important part of the evaluation, it may be necessary to lighten the level of sedation (this may be most effectively done after the anatomic evaluation has been completed). Bronchoscopy performed under deep anesthesia or with neuromuscular paralysis is almost guaranteed to prevent the accurate diagnosis of dynamic airway problems. It is not at all uncommon for the anatomy to look perfectly normal until the patient coughs, at which time surprisingly dramatic bronchomalacia or tracheomalacia may become apparent (Fig. 1.8). For this reason, it may also be desirable not to routinely apply topical anesthetic agents to the distal airway until the airway dynamics have been adequately evaluated. When a more involved or prolonged procedure is needed, the sedation can be deepened (or the procedure may be temporarily interrupted while an endotracheal tube or LMA is placed to provide for positive pressure ventilation during the remainder of the procedure).

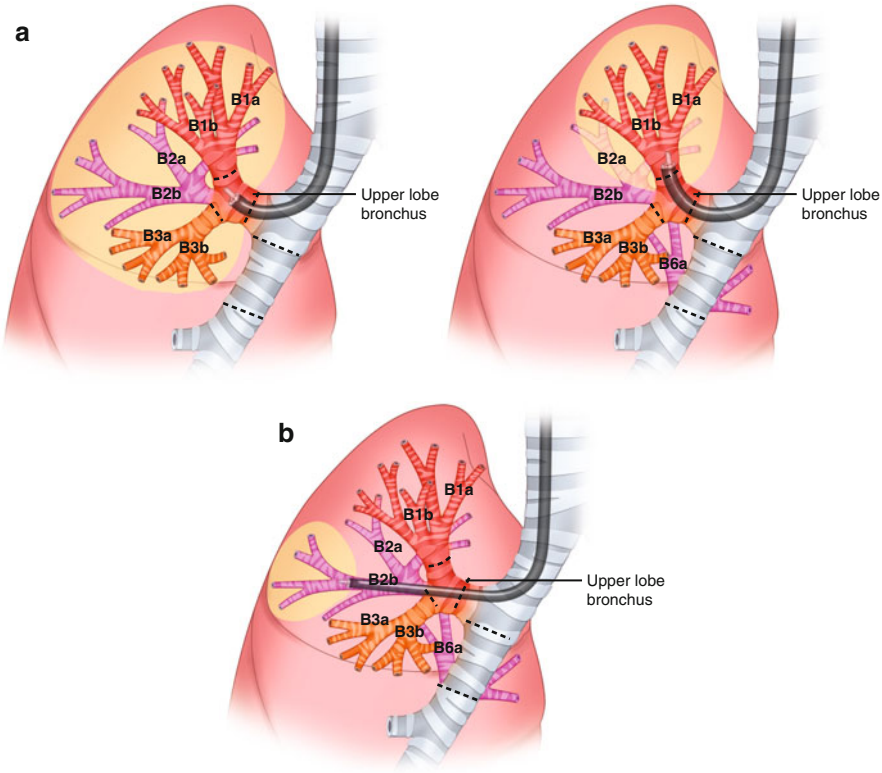

Fig. 1.7 The effect of scope size and position on BAL: Advancing the tip of the bronchoscope to a more peripheral position (which may be especially easy to do when a smaller diameter instrument is used) can result in sampling of a much smaller lung volume than may be intended (or recognized). This can, in some circumstances, produce erroneous results

This is often very useful when there is extensive mucus plugging or some other indication for a more prolonged procedure, especially a procedure that will require extensive suctioning.

The bronchoscopist should systematically evaluate the anatomy of the entire airway, beginning at the nostril. Generally, the easiest pathway through the nasal airway is through the middle meatus, and it is a smart idea to evaluate both sides of the nose, as unilateral obstruction is not rare. The presence of an oral airway can push the soft palate down, and make the adenoids appear to occupy much more of the airway than is the case under natural conditions. An oral airway can also push the tongue base down over the larynx, giving the appearance of glossoptosis. It is important to remove the oral airway, at least long enough to adequately evaluate the anatomy and dynamics of the upper airway. As the bronchoscope is advanced beyond the choana, the operator must also be alert to the changes that are produced by relatively small changes in the position of the head and neck. This is particularly

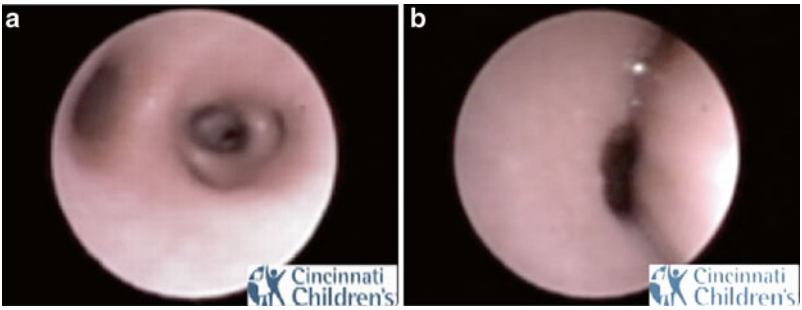

Fig. 1.8 (**a** and **b**) Two views of the bronchus intermedius, taken with the tip of the bronchoscope in the same position, approximately 0.5 s apart. The image on the *right* demonstrates significant bronchomalacia that was only apparent when the patient coughed

true for the tongue base. Dynamic abnormalities in the supraglottic region are also dependent on inspiratory effort (and the pressure gradient generated) as well as muscle tone, and this will vary significantly, depending on the level of sedation. Some children who have no history of stridor or upper airway obstruction can appear to have laryngomalacia or glossoptosis, and vice versa. The bronchoscopist must correlate the endoscopic findings within the context of the clinical history of the patient. If there is a history of stridor, but no abnormalities are apparent during the examination, the level of sedation should be changed until the symptoms are reproduced, so that an accurate diagnosis can be achieved. If there is audible stridor, the vibrating structures producing the sound must always be visualized; if not, the only explanation is that one is not looking in the right place. If there appears to be a significant dynamic abnormality but there is no history of upper airway obstruction or noisy breathing, this may be due to the effects of sedation, and not clinically relevant. In other cases, however, the history is incomplete or even wrong…

Because the tip of the flexible bronchoscope must be flexed anteriorly to view and then enter the larynx, it is much easier to obtain a view of the anterior commissure than of the posterior commissure. The posterior aspects of the larynx can often be more easily evaluated as the bronchoscope is being withdrawn than during insertion.

Complications of Bronchoscopy

All human activity involves risk. If the potential risk outweighs the potential benefit, then the activity should not be performed. This is also true of bronchoscopy. A complication may be defined as the occurrence of an event that is unexpected, and either causes harm to the patient or results in a significant change in the performance of the procedure. The most common "complication" listed in literature reviews is transient oxygen desaturation. Low oxygen saturation is often noted as a relative

contraindication to doing a bronchoscopy. However, in many patients, the need for the procedure outweighs the potential risk of producing some hypoxemia (indeed, the low oxygen saturation is often the very *indication* for the procedure, with the anticipation that the result of the procedure will be an improvement in the hypoxemia). There is little evidence to suggest that transient oxygen desaturations result in harm to the patient, and I do not feel that they should be considered a true complication. The operator and anesthesiologist can do much to minimize the potential, and to respond appropriately when a desaturation event does occur, but procedures should not be terminated simply because there are some desaturation events that resolve promptly and with reasonable effort. If a patient is unable to ventilate adequately to maintain oxygenation, then the procedure can be continued after providing an artificial airway (endotracheal tube or LMA).

Mechanical complications can include mucosal edema or hemorrhage, and pneumothorax. Microbiological complications include introducing pathogens into a previously non-infected lung or spread from an infected to a non-infected portion of the lungs; this is most likely to occur when there has been failure to adequately clean and disinfect/sterilize the instrument between patients. The most serious complication, other than death of the patient, is cognitive: failure to obtain the correct diagnosis or therapeutic outcome. There are many paths that can lead to this unhappy state of affairs.

One subtle but potentially very serious complication is failure to do the procedure when it is truly needed. I have seen a number of children who have undergone major thoracic surgical procedures (repair of VSD, repair of pulmonary artery sling, as two examples) and despite difficult intubations, only after the chest was closed did someone think to do a bronchoscopy … which revealed potentially life-threatening airway anomalies (complete tracheal rings). The incidence of complete tracheal rings is on the order of 60 % in children with pulmonary artery slings ….

When discussing the risk of a proposed procedure with parents/guardians, it is important to place the potential risks in proper context. It is also important to recognize, and to point out to parents, that sometimes, the most valuable finding is the definitive exclusion of serious pathology that had been suspected (and worried/agonized about) prior to the procedure.

The Clinical Utility of Bronchoscopy

The specific indications for bronchoscopy, as well as the findings, will vary enormously from institution to institution, and depend on many factors, the most important of which is the referral practice in that institution. At Cincinnati Children's Hospital, we have a referral pattern that encompasses virtually the entire USA, and a very high number of children who are being referred for consideration for airway reconstructive surgery. This is a very different patient population from that which might be seen, for example, in a hospital that focuses on children's oncology. Because of the high percentage of our patients with structural airway problems, we

perform ~50 % of our flexible bronchoscopies in conjunction with our otolaryngology colleagues, who perform rigid endoscopy. One might question the practice of doing both flexible and rigid bronchoscopy in the same patient by two different physicians, and that would be a legitimate question, if the only issue were the technical performance of the procedure. However, there is a world of difference between a flexible bronchoscopy performed by a pulmonologist and rigid bronchoscopy performed by a surgeon; we have very different perspectives, different instruments, and different procedural goals, we look at different aspects of the airway, and our follow-up is different. We believe that 1 + 1 =>2. We also frequently engage our colleagues in gastroenterology and do a triple endoscopy; we attempt to make the most effective and efficient use of anesthesia events as possible.

When we perform multidisciplinary evaluations, we sequence the procedures so that the flexible bronchoscopy/BAL is performed first, the rigid laryngoscopy/bronchoscopy is performed second, and then the patient is electively intubated for the GI endoscopy. This maximizes the potential to obtain a valid BAL specimen without contamination from prior laryngeal anesthesia and manipulation.

The clinical utility of a bronchoscopy will depend on the indication for the procedure, the care and skill with which it is performed, and other factors. No listing of "diagnostic yield" will be applicable to other institutions. Surprise diagnoses are common; I found a clinically unsuspected foreign body in 1 % of the first 1,000 flexible bronchoscopies I did (excluding patients in whom the presence of a foreign body *was* suspected). We often evaluate patients prior to bone marrow ablation for a bone marrow transplant; it is not uncommon to find previously unsuspected problems, including occult infection, anatomic problems, evidence of ongoing aspiration, or significant amounts of retained secretions. In those patients in whom the findings are normal, we may, in retrospect, question why we did the procedure. However, in those patients in whom there are significant findings, their entire management may be changed. Since it can be extremely difficult to define in advance who may have an abnormality, an attitude of "guilty until proven innocent" is defensible, *within reason* (we are not excused from using common sense and careful clinical judgment). Our approach to patients referred for airway reconstruction is based on the recognition that lung disease (due to aspiration or anatomic abnormalities, for example) is a major risk factor in the potential success of the surgical procedures, and we aggressively manage such patients before clearing them for repair.

Simple factors can dramatically reduce the clinical value of a bronchoscopy. The value of a culture of a BAL specimen can be neutralized, for example, by antibiotic therapy prior to the bronchoscopy. Failure to obtain a specimen from the proper location or failure to perform the appropriate analyses on the specimen can also lead to erroneous diagnosis. It is not safe to assume that lung disease is uniformly distributed throughout the lungs; I have seen many patients in whom one part of the lung was heavily inflamed and infected, while a BAL from another part of the lung was sterile, and yielded no evidence of inflammation on cytologic examination. The operator must review all available information prior to performing the bronchoscopy, and must also examine all parts of the lung, unless there is a truly compelling reason not to do so.

The decision to perform a bronchoscopy is predicated on the clinical situation, the experience and skill of the bronchoscopist, and an (admittedly, subjective) assessment of the risk/benefit ratio. For what specific indications is flexible bronchoscopy most likely to be useful? As noted, this will depend in large degree on the patient population, so let us look at some principles …. The most profound statement of relevance here is, "statistics do not apply to individuals," and one truly never knows what may be found.

In the following discussion, I will focus on generalities rather than specifics; this is not an attempt to provide a comprehensive review of the literature … this is a distillation of my own four decades of experience with pediatric flexible bronchoscopy ….

A carefully performed flexible bronchoscopy with appropriate BAL can usually yield a definitive etiologic diagnosis of pneumonia. In the vast majority of patients with pneumonia, however, this is not an enormous diagnostic challenge, and bronchoscopy is not likely to be cost effective, nor is the benefit likely to exceed the risk (although minimal) and cost. However, in a patient who is at risk for unusual organisms, who is immunosuppressed, who has an unusual clinical presentation, or who does not respond to treatment with empirically chosen antibiotics, bronchoscopy becomes much more reasonable. One situation which frequently arises in busy pediatric hospitals is pneumonia in the immunocompromised host. There is often a clinical urgency to initiate therapy ASAP, and not wait for the patient to achieve a satisfactory NPO status and for a procedural time slot to become available. Such patients are often immediately begun on Thundercillin, Megastompamycin, Amphoterrible, and assorted other agents in a desperate attempt to get control of the putative infection before it gets out of hand, and this is often a life-saving maneuver. A subsequent bronchoscopy and BAL are therefore much less likely to yield a definitive diagnosis, and even when it does, many practitioners will not alter the antimicrobial treatment plan, fearing that the BAL might still have missed something. The pulmonologist usually hears about such patients after a week or two of unsuccessful therapy, and then it is challenging to decide whether a bronchoscopy can be justified, since the yield can be fully expected to be low. It has been my personal experience in many of these patients that we find something not infectious—pulmonary hemorrhage, for example, or a foreign body or an endobronchial lesion—that explains the clinical history. It is impossible to cite any meaningful statistics to help decide which patient in this situation can be anticipated to benefit from bronchoscopy ….

Recurrent or persistent pneumonia is a very valid indication for bronchoscopy. While the vast majority of pneumonias in children are viral, pneumonia that is recurrent in the same area of the lungs is often due to a specific anatomic problem (including occult foreign body aspiration). Some such patients are discovered to have recurrent pulmonary hemorrhage, which can occur without overt hemoptysis or other clinical manifestations.

Noisy breathing is another common indication for diagnostic bronchoscopy. Most patients with a history of recurrent wheezing have a form of asthma. These patients do not need bronchoscopy. However, I have seen many patients with "severe" asthma, not responsive or only very poorly responsive to conventional asthma treatment, who do not have asthma. Rather, they have anatomic abnormali-

ties, occult foreign bodies, bronchomalacia, bronchial compression, etc. Some of these children have become Cushingoid due to escalating dosing with systemic steroids in an ill-fated attempt to gain control of their severe asthma symptoms …. Many physicians believe that poorly controlled asthma is a *contraindication* to bronchoscopy—in fact, it can be a highly productive *indication* for bronchoscopy.

Persistent stridor in an infant is a common cause of much anxiety on the part of parents, grandparents, and pediatricians. Most such children have laryngomalacia, and can be expected to "grow out of it"—it could be argued that bronchoscopy is unnecessary. However, any child with persistent stridor who is also failing to thrive, or who requires supplemental oxygen without an identifiable pulmonary cause, or whose parents cannot sleep at night due to the anxiety produced by their child's noisy breathing can be greatly benefitted by a diagnostic bronchoscopy. Knowledge is power, and if the parents can be assured, definitively, that there is no other lesion, they will sleep better, be more confident in their care of the child, and will be much less likely to go searching from doctor to doctor for CT scans, etc. There is great power in the definitive knowledge that your child's noisy breathing is truly benign. On the other hand, 15–20 % of patients whose airways I have examined for stridor, and in whom I find laryngomalacia, also have other significant airway abnormalities such as tracheomalacia, tracheal or bronchial compression, and complete tracheal rings. Other published reports have noted similar results. The great Godfather of bronchoscopy, Chevalier Jackson, said (in 1915) "If in doubt as to whether bronchoscopy should be performed, bronchoscopy should always be performed." I agree.

Children with obstructive sleep apnea most commonly have adenoidal or tonsillar hypertrophy, and can be treated with simple measures (T&A). However, if these measures do not relieve the obstruction, examination with a flexible instrument can be very helpful. Glossoptosis, laryngomalacia, and other problems can be identified. As discussed earlier, the bronchoscopist must pay close attention to the position of the head and neck, and to the level of sedation. If, during the examination, there is no noise, and no dynamic collapse is seen, the level of sedation should be altered so that the obstruction will occur and can be documented.

Chronic cough is a common indication for bronchoscopy. In this case, airway dynamics are often as important as the findings on BAL (microbiology, cytology). Patients with tracheomalacia or bronchomalacia may develop an intractable cough due to mechanical trauma to the mucosa produced by the cough. I have seen a number of patients previously diagnosed with "psychogenic cough" who actually had tracheomalacia or (central) bronchomalacia—understanding the nature of the problem can lead to resolution. I teach these patients to cough against pursed lips to maintain some back pressure and thus avoid making the "barking" sound with their cough, and this reduces the risk of mechanical irritation produced by the cough itself (which can then perpetuate the cough).

Not every child with atelectasis requires bronchoscopy, although the procedure can be both diagnostic and therapeutic. Most children with atelectasis resolve quickly on their own. However, if the atelectasis is functionally significant, or is recurrent, or is persistent, then bronchoscopy can be valuable. Mucus plugging, foreign bodies, bronchial compression or stenosis, and other diagnoses lurk, waiting

to be discovered. In a significant percentage of patients with atelectasis, no anatomic abnormality will be discovered; in these patients, the cytology and microbiology of BAL specimens will be important, and the BAL may in itself be helpful to speed resolution of the atelectasis (by loosening mucus plugging beyond the visual range of the bronchoscope).

One area of confusion is the bronchoscopic diagnosis of aspiration. Ideally, aspiration could be defined by the discovery of a marker that can only get into the lungs by aspiration. Many surrogate markers have been evaluated, including lipid-laden macrophages (LLM) and gastric enzymes. Unfortunately, there is no marker that is both specific and sensitive, and the clinician must place everything into the proper context for interpretation. For example, while it is clear that aspiration of lipid-containing liquids or even solids can produce an elevated number of LLM in subsequent BAL specimens, a patient who is NPO and aspirates only oral secretions cannot be expected to produce lipid laden macrophages. Even a patient who is clearly aspirating may have a highly variable number of LLM, depending on the amount of material aspirated, the lipid content of the material, the physical state of the material (i.e., liquid vs. solid), and most importantly, the time between the aspiration event and the sampling. Sampling immediately after an aspiration event cannot be expected to yield LLM—the process takes time. It may also take weeks to clear LLM after a single aspiration event, but the factors influencing the rate of clearance are totally unknown and almost surely variable. Finally, processes other than aspiration can produce LLM, including bone marrow infarction (as in sickle cell anemia) and hemophagocytic lymphohistiocytosis. The presence of large numbers of dead or dying neutrophils could be expected to result in LLM, since alveolar macrophages readily ingest dead cells, but, perhaps surprisingly, many patients with chronic purulence (i.e., cystic fibrosis) do not have elevated LLM in their BAL specimens.

In the evaluation of a patient with suspected aspiration, I do look at the percentage of lipid laden macrophages. I also look for large numbers of squamous epithelial cells (assuming that the BAL specimen has not been contaminated with saliva during the procedure—see previous discussion), and large numbers of "oral flora" on culture. I place these findings in the context of the child's history and known anatomic/functional defects, and will report "findings consistent with aspiration" but almost never am willing to state that the BAL findings are "diagnostic of" aspiration.

Pediatric radiologists often tease their pulmonary colleagues that multi-detector CT techniques have made bronchoscopy obsolete. While it is true that imaging techniques can yield much important information about the lungs and airways, the truth is that bronchoscopy and radiologic techniques are complementary. Neither can give the entire picture, and both are often necessary for accurate and complete evaluation of patients. For example, while a CT scan can demonstrate extrinsic compression of central airways, and identify the offending structure, radiographic studies are often confused by the accumulation of airway secretions, and in any case, the radiographic studies do not provide microbiologic information nor therapeutic benefit. On the other hand, bronchoscopy can easily miss sampling a lesion that is beyond the visual range unless the bronchoscopy is guided by radiologic imaging.

Bronchoscopy can be a very important adjunct to surgical manipulation of the pediatric airway. Transillumination of the bronchi can assist the surgeon in the identification of specific regions of the lung. Direct observation of the trachea during an aortopexy, for example, can improve the likelihood that the surgical procedure will be effective.

The potential value of diagnostic bronchoscopy in children is perhaps best embodied in the statement of the indications for bronchoscopy: information in the lungs or airways of the child, necessary for the care of the child, and best obtained with the bronchoscope.

"Seek, and ye shall find." Matt 7:7