The Child and Newborn

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Editorial

Tracheobronchial foreign body aspiration (FBA) is often a common cause of mortality and occasional morbidity in children. It is more common in children younger than two years of age. The peak incidence being between one and two years of age. The major reasons being that at this age, most children are able to move around, explore their world via the oral route and have the fine motor skills to put a small object into their mouths, but they do not yet have molars to chew food adequately. The confounding factors to FBA in this age group include access to improper foods or small objects and older siblings and casual caregivers (who may place food or objects into the mouths of infants or toddlers). Young children are also particularly vulnerable to foreign body obstruction because of the smaller diameter of their airway, which is vulnerable to obstruction.

Commonly aspirated objects in children include peanuts,, other nuts, popcorn, food particles, erasers, ear pins and pieces of toys. Common food pieces are the usual items aspirated by infants and toddlers, whereas nonfood items (e.g., coins, paper clips, pins, erasers, toy balloons, pen caps) are more commonly aspirated by older children. Balls, marbles, toy balloons and other toys are commonly involved in fatal FBA. Factors like roundness (round objects are most likely to cause complete airway obstruction and asphyxiation), failure to break apart easily, compressibility and smoothness, slippery surface make FBs more dangerous.

Most of the aspirated FBs in children are located in the bronchi. Laryngeal and tracheal FBs are less common. FB in right lung is more common (60 %) and most commonly in the in the right main bronchus.

FB may be lodged in the larynx or trachea in children younger than 1 year. Tracheal narrowing or weak respiratory effort may predispose to tracheal FB. Laryngotracheal FBs are associated with increased morbidity and mortality.

Clinical presentation of foreign body aspiration depends upon the degree of airway blockage and also on the location of the object. The age of the child, the type of object aspirated object and the elapsed time are other factors.

Children with suspected FBA who are stable should undergo a focused history and physical examination. A chest X ray is the baseline investigation. The caregivers should be asked about the history of choking episode. Physical examination may reveal wheezing, stridor and regional variation in breath sounds.

Bronchoscopy

The upper airway should be examined in all children with a moderate or high suspicion of FBA, typically using rigid bronchoscopy so that the object can be safely removed. Flexible bronchoscopy may be used for diagnostic purposes in cases in which the diagnosis is unclear, or if the FBA is known but the location of the object is doubtful.

Extraction of FB should be performed by an experienced operator to minimize the risk of complications. Repeated unsuccessful attempts to remove the FB may push it down to a distal position, making them more difficult to retrieve. The rare complications of FB extraction include pneumothorax, hemorrhage and respiratory arrest.

Dr Jaydeep Choudhury
Editor in Chief

Indian Academy of Pediatrics

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FOREWARD

Dear PEDIA SCOPE delegate,

Greetings from the Indian Academy of Pediatrics!!

Fiberoptic bronchoscopy (FOB) is an important entity in the armamentarium of procedures listed in diagnosis of respiratory problems. It is a universally accepted procedure both in the diagnosis and therapy of various pulmonary disorders. This procedure allows careful inspection of the bronchial tree for endobronchial lesion and foreign body and also helps in recovery of deep respiratory secretions, brushing and biopsy, which is useful in diagnosis of un- common infections, neoplasm and other non infectious causes. FOB not only helps in assessing the disease area but also provides better bacteriological and histological yield thus helping to reach a definite diagnosis.

It has been a long cherished dream for me to sensitize the pediatricians. in general, and intensivists and those interested in pulmonology, in particular, in this investigative modality. Hence this module PEDIA SCOPE was conceived to spread the message and to bring it closer to you. I am happy that with academic inputs of Dr. Pallab Chatterjee, Dr. Mahesh Mohite and other stalwarts, educational grant by M/S Cipla and technical support from M/S. Olympus we have been able to launch PEDIA SCOPE under IAP Action Plan 2019.

I sincerely hope that some of you will take up FOB in the years to come in your routine practice and thus contribute to improve the care of children in our country.

Happy learning.

Yours in academy service

Dr Digant D Shastri
IAP President, 2019

West Bengal Academy of Pedaitrics

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PREFACE

Dear members,

It is my proud privilege to present to you this module and manual of Pediascope, a Basic Bronchoscopy Course for Pediatricians. Conceived by our President, DrDigantShastri, this module comprises of around 10 workshops all over the country and aims to sensitize you to use this very useful, but underutilized, investigative procedure in your regular practice.

I am really grateful to DrVijayasekhar, who initiated me in this field of bronchoscopy and supported me unconditionally in my initial years, and Dr Robert Wood, for his training and guidance. I am thankful to DrGowrishankar, Dr Mahesh Mohite, Dr Shashank Kadam, Dr Ankit Parekh, Dr Ilin Kinimi, Dr Shilpa Hazare and all others who contributed in preparation of this module. I am also grateful to DrVijayasekhar, Dr Andrew Colin, DrSejalSaglani and others who lent their support to develop the module and the manual. I thankfully acknowledge the contribution of M/s Cipla Limited and M/s Olympus for all their help and support, without which this module would not have seen the light of the day.

I sincerely hope that this module will help you in the years to come.

Thanking you,
Yours in academy service,

Dr Pallab Chatterjee
DCH, MD(Ped), DNB(Ped)
European Diplomate in Pediatric Pulmonology
National Coordinator, Pediascope.

Chapter 1

Plan Your Scopy

Pallab Chatterjee
European Diplomate in Pediatric Respiratory Medicine

"Good judgement comes from experience; experience comes from poor judgement!"

Introduction

Bronchoscopy is the visual examination of the airways using either rigid or flexible instruments that are inserted into the airways. Gustav Killian, in 1897, was the pioneer in bronchoscopy, using a hollow metal tube. Until 1980s, only rigid scopes ("open tube") were available for pediatric use. The flexible scopes meant for adult use had a diameter of 5mm, which is the usual diameter of the infant trachea. Dr Robert Wood, in collaboration with Olympus Corporation, developed the flexible fiberoptic bronchoscope of outer diameter 3.5mm, with a suction channel of 1.2mm, that changed the entire scenario of Pediatric Pulmonology.

The Team

A bronchoscopy team consists of at least the following:

(i) Bronchoscopist (s) – physician (s)

(ii) Assistant(s) for the procedures – nurse, respiratory therapist

(iii) Anesthesiologist/sedation nurse – physician/ nurse

(iv) Clerical/billing staff

(v) Cleaning staff

The bronchoscopist is the team leader and should be personally competent to perform all the tasks of each of the team members, and be willing to do so when necessary. He should maintain close congenial relation with his surgical colleagues, whose help might be required in an emergency.

The assistant(s) need to be trained and skilled. His first responsibility is to the patient, followed by handling of the equipment, handling of the specimens and others, varying from institution to institution.

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To ensure that the patients remain safe and comfortable during the entire procedure, someone other than the bronchoscopist must be responsible for effective sedation and monitoring of the patient. Bronchoscopy can be fun and very engrossing, and the "bronchoscopist's hypnosis" is something very real. The duty of the sedation nurse/anesthesiologist is to monitor the patient and maintain records. There should be very clear and effective communication with the bronchoscopist before, during and after the procedure.

The final product of bronchoscopy is information. Hence images, observations and interpretations must be prepared by the bronchoscopist in the formal report that is distributed to the referring physician. Hence the nursing and clerical staff play an important role in timely distribution of data.

Finally, it is important to for someone specially trained to be responsible for cleaning the instruments after each use and make it suitable for subsequent use.

The Venue

Bronchoscopy is usually performed in an appropriate venue that is safe and effective. There are three basic venues: operating theatre, bronchoscopy suites and intensive care units.

The Operating Theatre is usually the most appropriate venue. There is always an anesthesiologist to assist in sedation of the patient and to take care of any eventuality. However, there may be challenges in scheduling in a busy hospital. One also has to supervise that necessary equipments and supplies are available in the OT.

The Bronchoscopy Suite is an ideal venue for most bronchoscopists, where he is in his own 'territory'. The suite must be fully equipped for any emergency, like cardiac arrest, hemorrhage, pneumothorax, etc. There should be an alarm system in the hospital,

and a 'Code Blue' should be in place to get help in case the bronchoscopist is the only physician around. In most hospitals, the suite is usually shared with their adult colleagues (pulmonologists and gastroenterologists), and this results in more efficient utilization of resources.

It is possible to do a bronchoscopy at any bedside, though it should never be attempted outside an Intensive Care Unit. It may not be possible to shift the patient outside the ICU in many circumstances, and the bronchoscopist must evaluate the facilities available before embarking on the procedure. Availability of cleaning solutions and simple things like 'slip-tip' syringes (in contrast to the usual 'Luer-Lock' syringes) should be ensured.

The most dangerous time is immediately after the procedure, when the staff tend to relax their vigilance and the child can become suddenly apneic.As most bronchoscopies are done on an outpatient basis, there must be an appropriate and safe venue for the patient prior to the procedure and for post-procedure recovery and monitoring.

Pre-requisites

The pre-requisites before doing a fiberoptic bronchoscopy are as follows:

1. A clear indication for the procedure, where the risk/benefit is in the favor of the patient.
2. Appropriate instrument size, depending on the age and weight of the child, and whether intubated.
3. Available facilities for bronchoscopy and the skill of the bronchoscopist.
4. Available monitoring facilities before, during and after the procedure.
5. Skill and facilities for cardio-pulmonary resuscitation.
6. Reliable laboratory facilities for transfer of any specimen at the earliest.
7. Recording facilities during bronchoscopy and maintain a database.

The Instrument Trolley

The following equipments and drugs are to be readied in a systematic manner on the trolley:

1. 0.9% normal saline.
2. Appropriate size 'slip-tip' syringes filled with saline for BAL.
3. Appropriate size 'slip-tip' syringes filled with 1% lignocaine for local anesthesia (2% lignocaine diluted 1:1 with normal saline).
4. Romson's Mucus Extractor, for collecting BAL.
5. Lubricant jelly
6. Sterile gauze pieces, Alcoswabs, Gloves, Gown, mask
7. Resuscitation equipment: appropriate sized bag-and-mask, endotracheal tube, LMA, laryngoscope with spare battery.
8. ***Medications for sedation / analgesia:***

(a) Pre-medication: Chloral hydrate in the dose of 50-80 mg/kg few hours before procedure

(b) Local anesthesia: 2% Lignocaine jelly for the nose; 1% Lignocaine spray for airways

Intravenous sedation / analgesia:

(a) Midazolam (0.05 – 0.2mg/kg); Pethidine (1 – 2 mg/kg); Fentanyl (1-3 mcg/kg);

(b) Ketamine (1-3 mg/kg); Propofol (2.5 – 3.5 mg/kg)

Reversal agents:

(a) Naloxone (0.01 – 0.1 mg/kg; max 2mg); Flumazenil (0.01 mg/kg; max 0.2mg)

Data Handling

Often what is written is more important than what is done. Generation of a report with appropriate images, observations and their interpretation, and data generated from specimens obtained during the procedure is the ultimate goal of bronchoscopy.

All bronchoscopies should be video recorded and the bronchoscopist should go through the same at his leisure, as many findings may be missed during the procedure itself. As this entails generation of a large volume for images, there should be a system for recording and archiving of data for future use.

The procedure report should include the indications for the procedure, a brief history, a short description of the procedure, the findings and their implications, the complications, diagnosis, and a discussion with a plan for follow-up. There are various software packages that help generate a report, and most hospitals already have one in use by the adult scopists.

Specimen Handling

The most serious complication of bronchoscopy, other than death of the patient, is to get a wrong

answer. The commonest cause for this is mishandling of the specimens (BAL, biopsy, etc) Thebronchoscopist has to work in close liason with the laboratory and the pathologist/microbiologist needs to be intimated beforehand of the history and diagnostic possibilities. The BAL specimen must reach the laboratory within one hour of the procedure and promptly processed. Nothing can be more damaging than placing the biopsy specimen into the wrong preservative or if wrong tests are requested.

Communication

There should be effective and timely communication with all the team members and with the family prior to the bronchoscopy, and afterwards as well. Unfortunately, in this era of increasing medico-legal cases, a detailed explanation of the procedure and its complications should be given to the patient/ parents. An appropriate consent should be taken for the same. The referring physician should be communicated immediately so that proper care of the patient is taken and they remain satisfied, to ensure a steady flow of referrals, without which the program will not sustain.

Thus, to conclude, for a successful bronchoscopy program, one must build, train and nurture the team; ensure a proper venue; obtain and maintain proper equipment; handle data efficiently; maintain records; and communicate effectively.

Further reading :

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6. Payne D. Allergy. 2007 Jun;62(6):577-8.

Chapter 2

Know Your Bronchoscope : Types of Bronchoscopes and Recording Systems, Cleaning and Disinfection

Shashank Kadam

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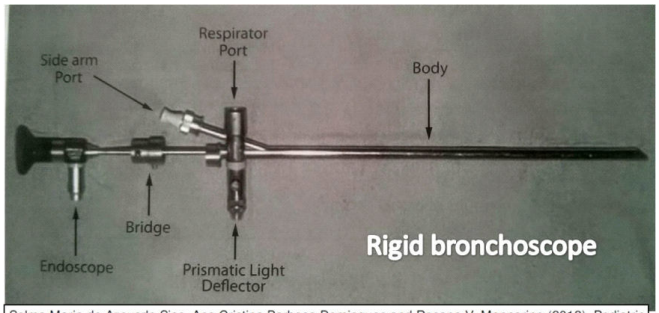
Bronchoscopy the visual examination of the airways is usually performed for diagnostic purposes, but it is also useful for certain therapeutic maneuvers. Bronchoscopy may be performed with either rigid or flexible (video/fiber-optic) instruments, depending on the particular needs of the patient and the skills and instrumentation available to the bronchoscopist.

The rigid ("open tube") bronchoscope consists of a metal tube of appropriate diameter and length, which is passed into the trachea, and through which the operator may look and the patient may breathe. The instrument is not a simple metal tube; it is equipped to deliver anesthetic gases, and light to the distal tip.



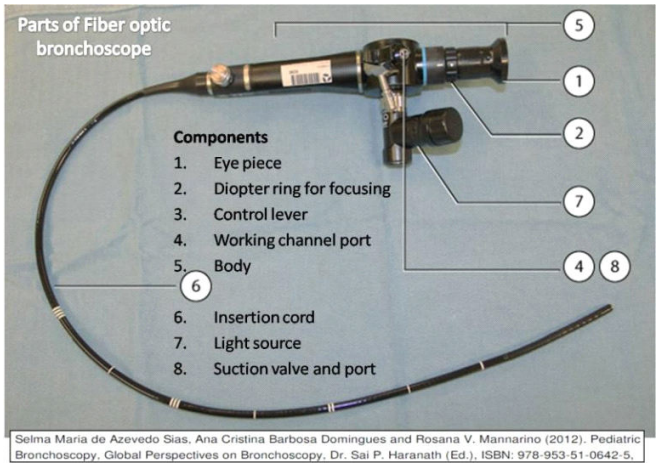
Selma Maria de Azevedo Sias, Ana Cristina Barbosa Domingues and Rosana V. Mannarino (2012). Pediatric Bronchoscopy, Global Perspectives on Bronchoscopy, Dr. Sai P. Haranath (Ed.), ISBN: 978-953-51-0642-5,

The nomenclature of bronchoscope sizes can be confusing. In general, rigid instruments are defined by the diameter of the largest instrument that will pass through the bronchoscope, while flexible bronchoscopes are defined by their outer diameter. For example, a 3.5-mm flexible bronchoscope will easily pass through a "3.5-mm" rigid bronchoscope.

The flexible bronchoscope is essentially a solid instrument that is composed of thousands of glass fibers that carry the image and the light for illumination. The tip of the instrument can be deflected to guide it into the desired path or location. Most flexible bronchoscopes have a small suction channel through which secretions may be aspirated, fluids may be delivered to the airways, or small flexible instruments may be passed.

The smallest scope is 2.2mm with no channel for instrumentation. The common used scopes range from 2.8mm to 4.9mm in diameter and they possess an instrumentation channel from 1.2mm to 2.2mm in diameter. Adult scopes are even larger.

More information about bronchoscopes, their different sizes are available on manufacturer's website.



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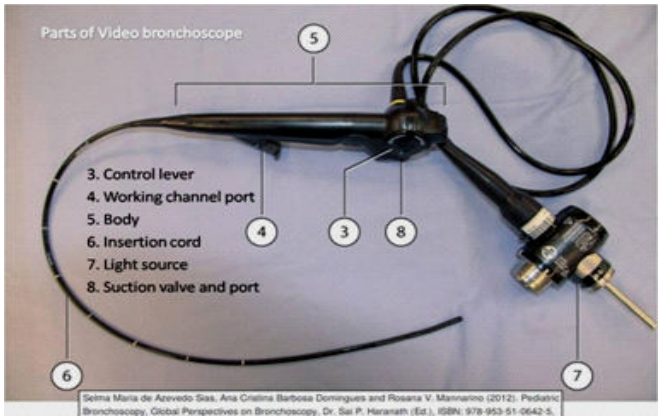

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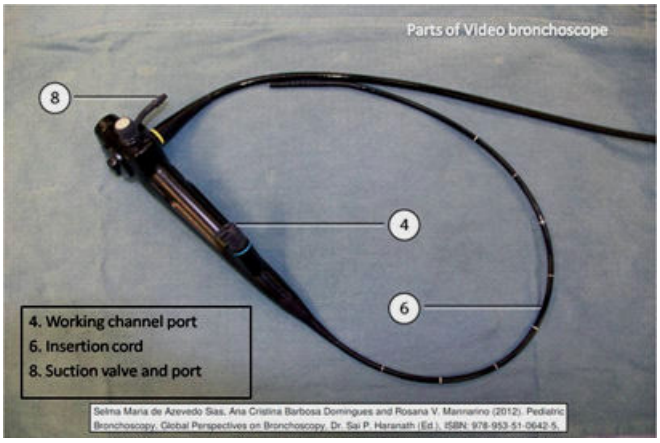
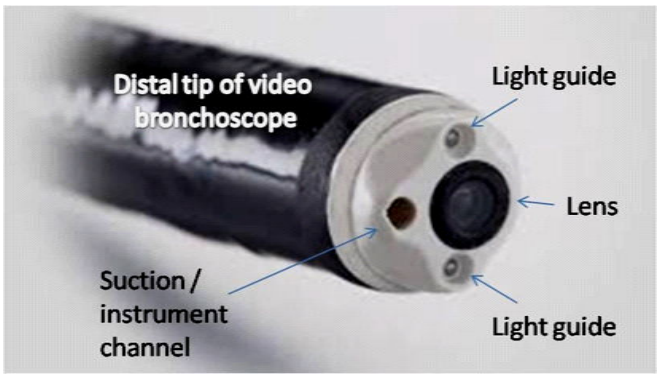




Flexible fiber-optic bronchoscopes are limited in their optical performance by the number of glass fibers that compose the image. Newer pediatric instruments now mostly utilize a video chip at the working tip (and thus generate an image with greater resolution).

Video Bronchoscope has a small ccd chip in the head which converts images into current and is transmitted through a wire to a processor which reconverts it into image as seen on monitor. The tip has the camera so is the most delicate part of the video-scope.

Recording your procedure

Video recording the procedure allows later review of the observations and sometimes leads to a revision of the diagnosis. To augment teaching, consultative reports, and even research data acquisition, serious consideration should be given to recording all procedures.

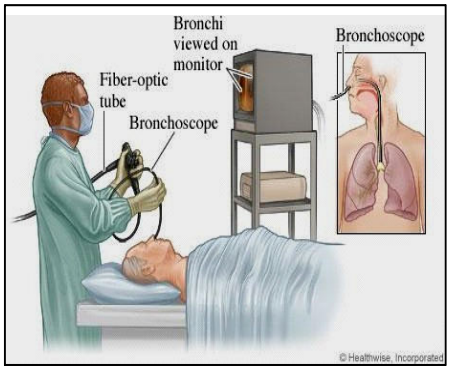
The duration of bronchoscopy examinations varies considerably depending on the diagnostic and therapeutic procedures used. It can last more than 20 minutes if a complex diagnostic work-up is included. With wide access to videobronchoscopy-, the whole procedure can be recorded as a video sequence. It is important to record the full bronchoscopy procedure as documentation when liability issues are at stake. Furthermore, an automatic recording of the whole procedure enables the bronchoscopist to focus solely on the performed procedures.

Software is available for recording the procedure on laptop/desktop. The output from the video processor goes to the laptop/desktop along with the monitor. The recorded video and images can be reviewed, and if need be, edited before giving a copy to the patient.

The reporting format is available in the software and images can be added in the report.

The report should mention patient details, date, time, bronchoscopist's name, indication for bronchoscopy, all areas viewed, any abnormality in structure, procedures performed and samples collected along with details of anesthesia used, any events during procedure.


Bronchoscopy tower



Care and Maintenance of Bronchoscopes

Bronchoscopy is not a sterile procedure, since the instruments through a non-sterile area (the nose and/or mouth). However, bronchoscopes and associated instruments must be cleaned and sterilized before use. Transmission of infectious agents from patient to patient due to inadequate cleaning or sterilization procedures has been well documented.

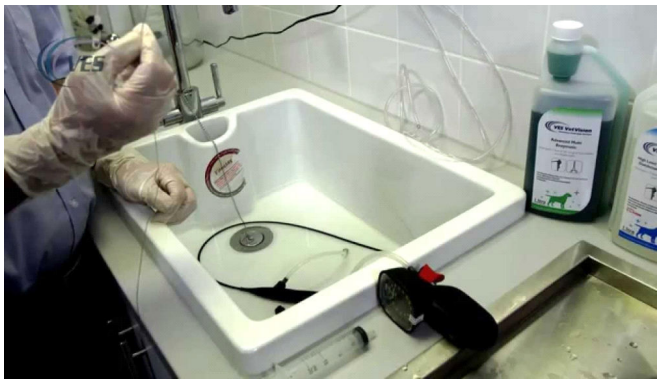
In general, bronchoscopic equipment should be cleaned as soon as possible after use because dried blood and mucus are much more difficult to remove and will prevent adequate sterilization by any method. At a minimum, the instruments should be flushed

with water immediately after use, and, if possible, soaked in an enzymatic detergent until formal cleaning can be done.

Rigid bronchoscopes are cleaned by vigorous brushing with detergent, followed by rinsing; they may be sterilized by steam autoclaving. Glass rod telescopes and other associated components may not be exposed to steam, however, and must be sterilized with ethylene oxide or with liquid agents such as glutaraldehyde or peracetic acid/ o-pthalaldehyde (OPA).

Flexible bronchoscopes are cleaned by careful scrubbing of the exterior with a soft cloth and enzymatic detergent. The suction channel must be cleaned by multiple passes of an appropriate cleaning brush. Thorough rinsing is followed by high-level disinfection (with glutaraldehyde or peracetic acid) or sterilization (with ethylene oxide).

The lenses of rigid telescopes and flexible



bronchoscopes must be carefully scrubbed and polished with a soft cloth during cleaning. Otherwise, small amounts of protein left on the lens will accumulate over time, making the image progressively less satisfactory. Flexible bronchoscopes and glass rod telescopes are made of glass and are fragile (not to mention expensive). They must never be dropped or subjected to forces that will cause breakage.

Flexible bronchoscopes should never be passed through a patient's mouth unless protected by a rigid bite block; an endotracheal tube will not protect the bronchoscope from severe damage by teeth. The care with which instruments are cleaned and handled must match the care with which they are utilized in the patient's airway.

Individuals who are responsible for cleaning and preparing the instruments must be well trained and supervised. The bronchoscopist must assume full responsibility for the care and cleaning of the instrument.

Further Reading :

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2. de Azevedo Sias, Selma Maria, Ana Cristina Barbosa Domingues, and Rosana V. Mannarino. Pediatric Bronchoscopy. INTECH Open Access Publisher, 2012.

Chapter 3

Normal Anatomy In Bronchoscopy

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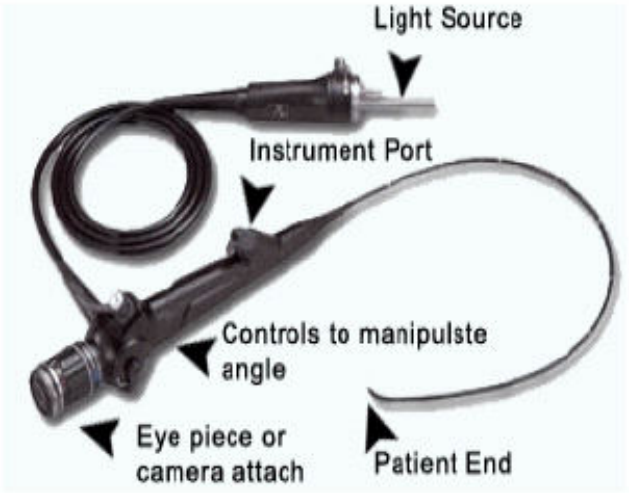

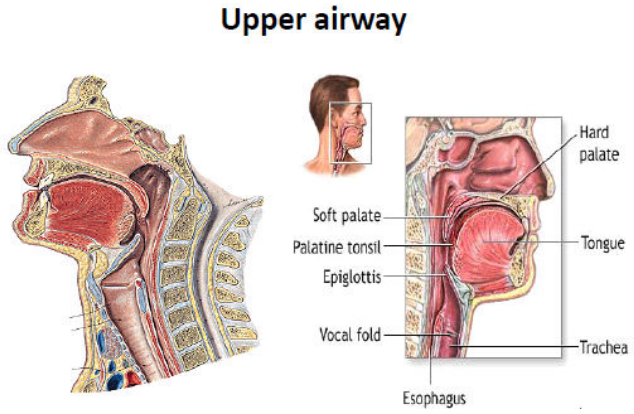

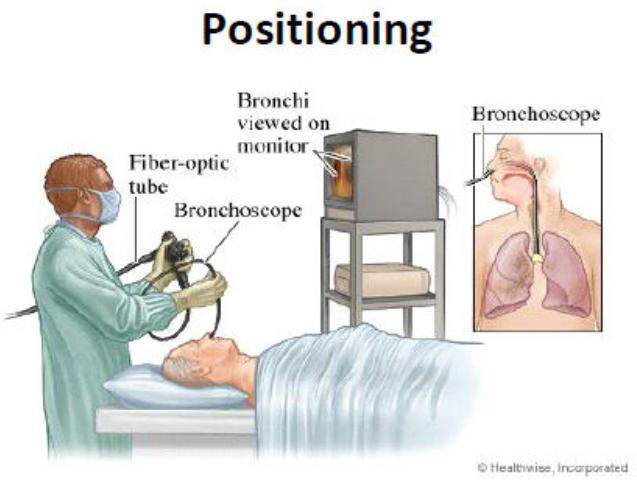

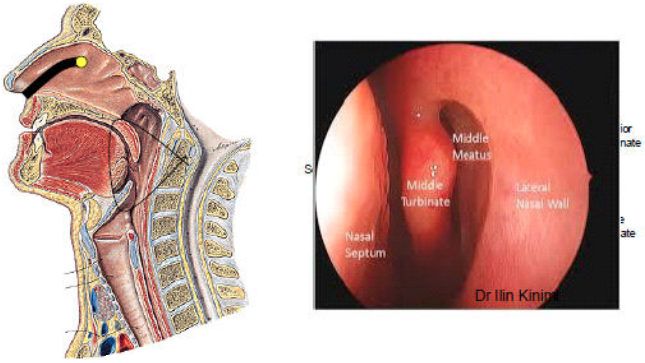

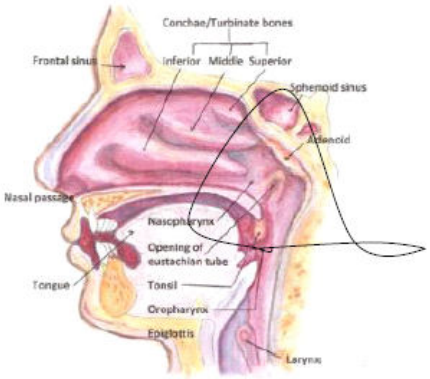

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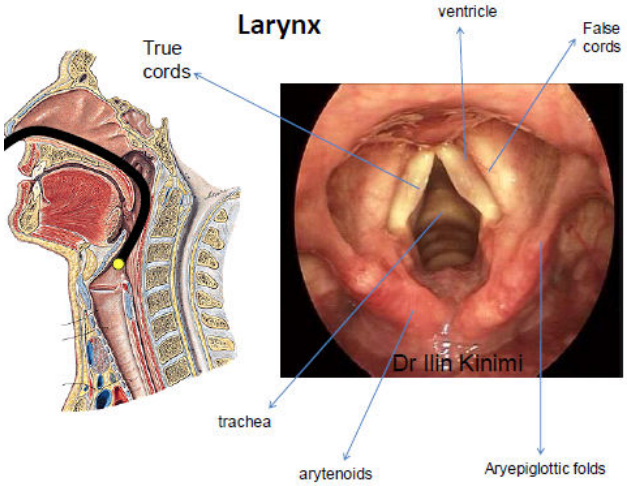
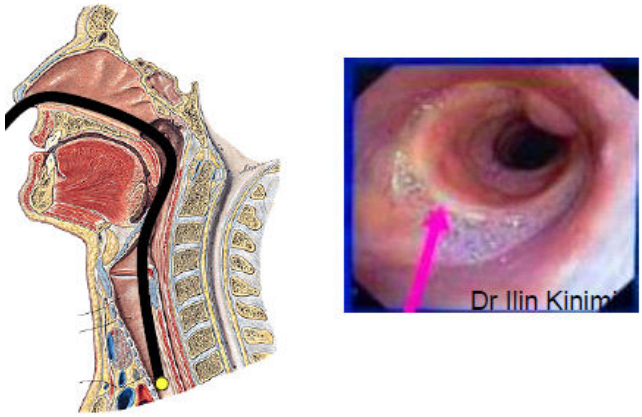
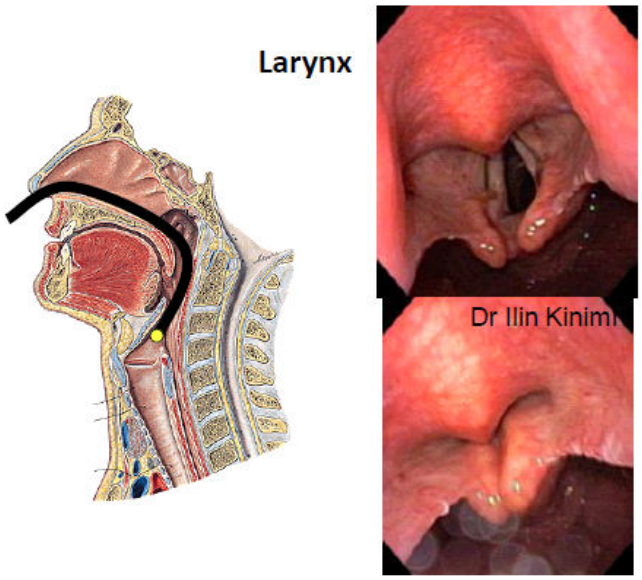
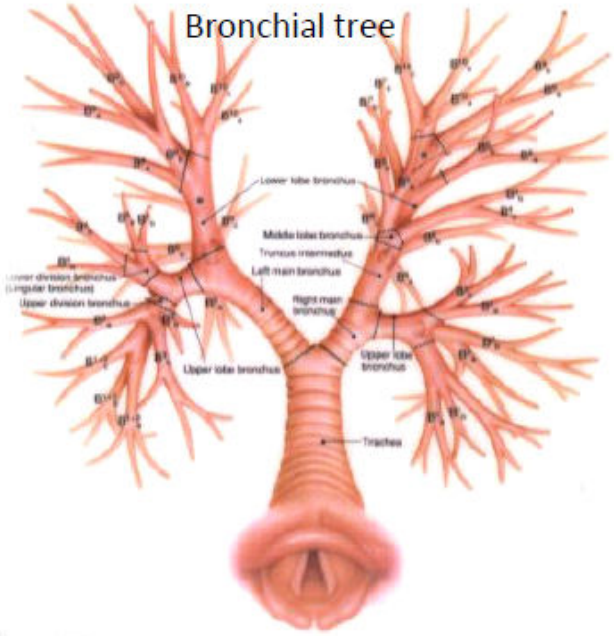
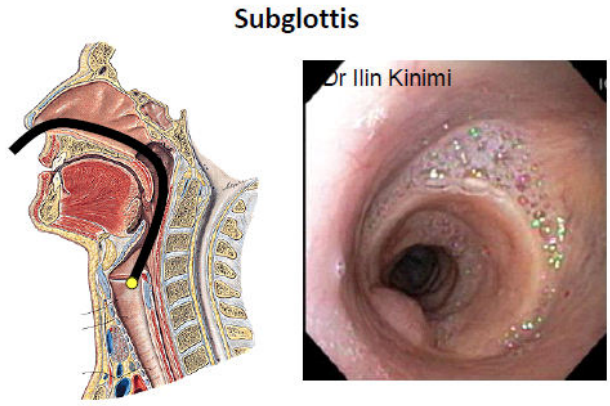
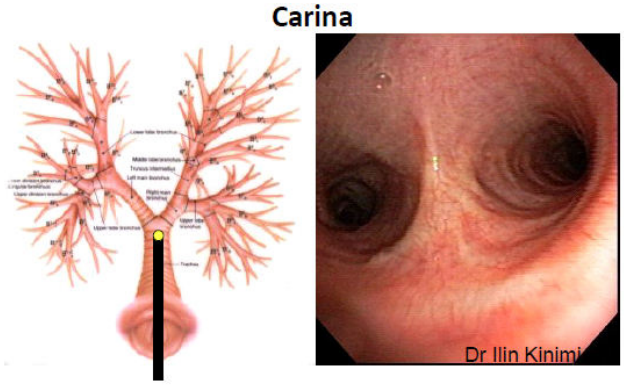

Larynx
ventricle
True cords
False cords
Dr Ilin Kinimi
trachea
arytenoids
Aryepiglottic folds


Trachea
Dr Ilin Kinimi

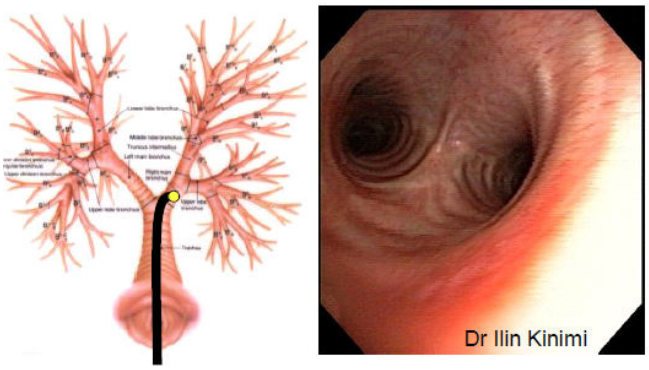

Larynx
Dr Ilin Kinimi


Bronchial tree
Trachea

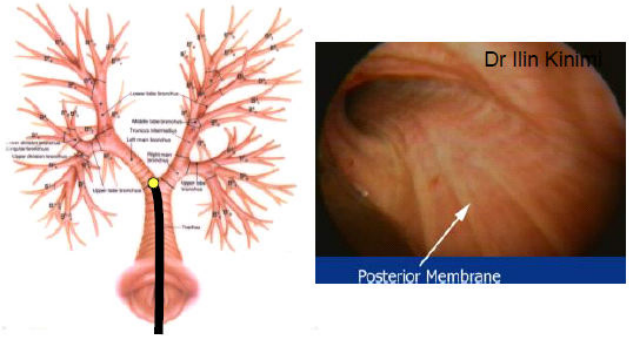

Subglottis
Dr Ilin Kinimi


Carina
Dr Ilin Kinimi

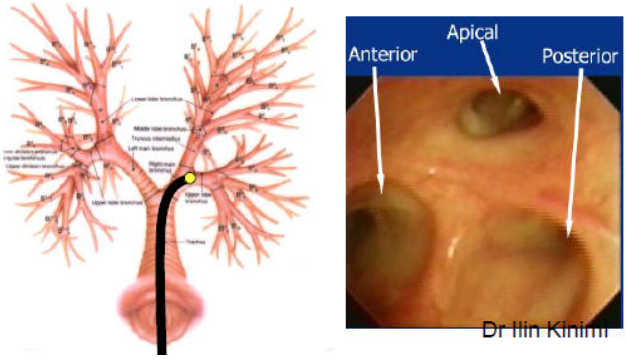
Right main bronchus



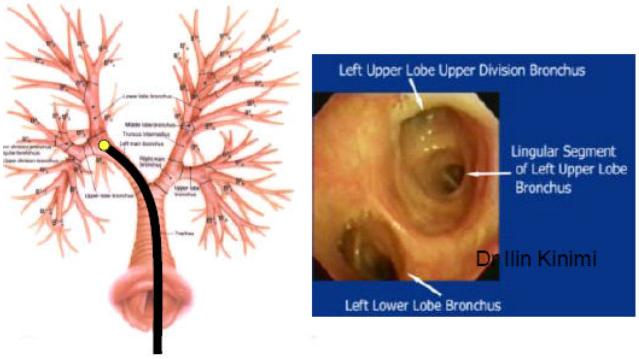
Left main bronchus



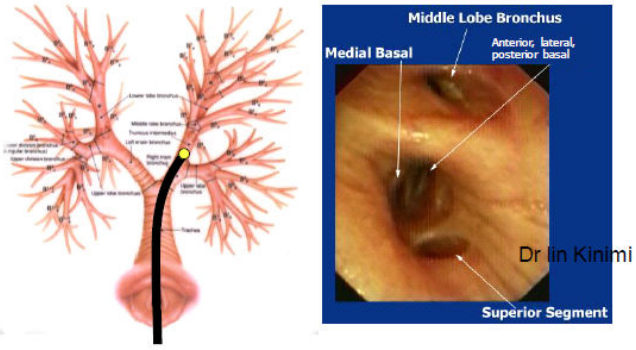
Right upper lobe bronchus



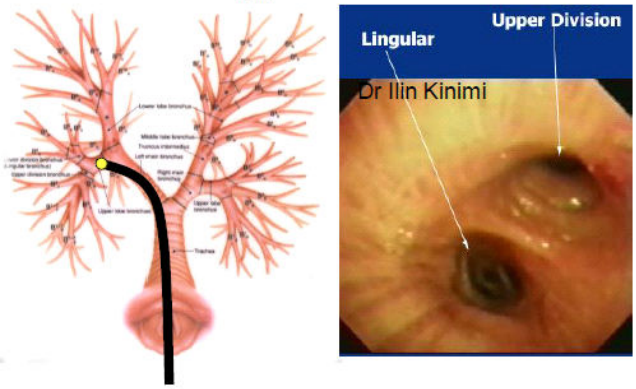
Left main bronchus



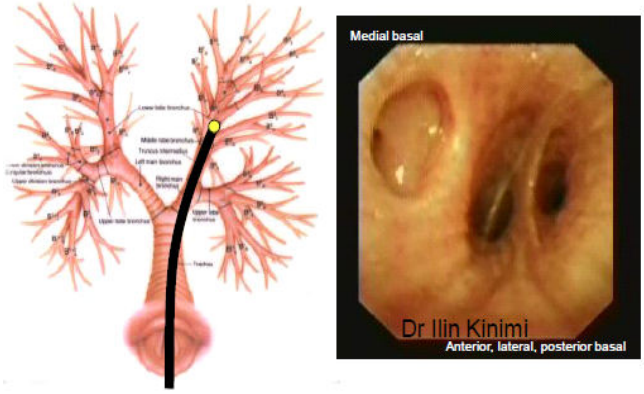
Right bronchus intermedius



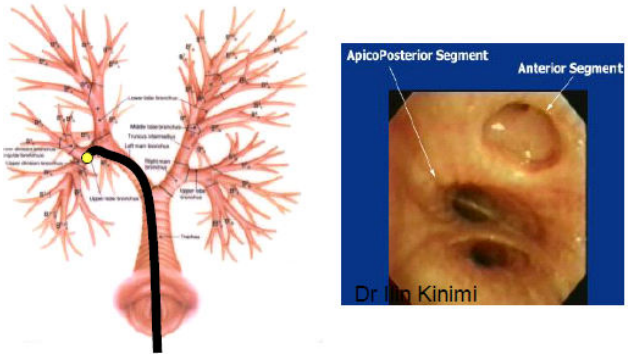
Left upper lobe bronchus



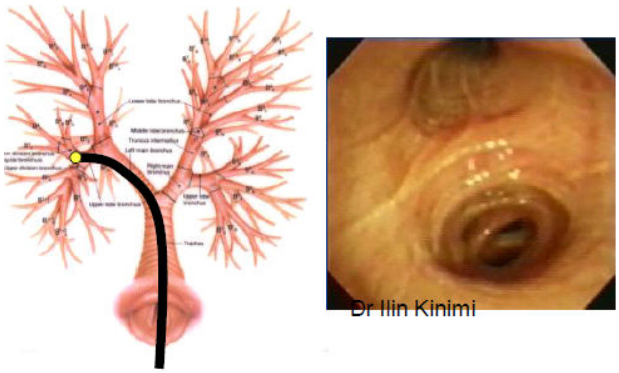
Distal right bronchus intermedius



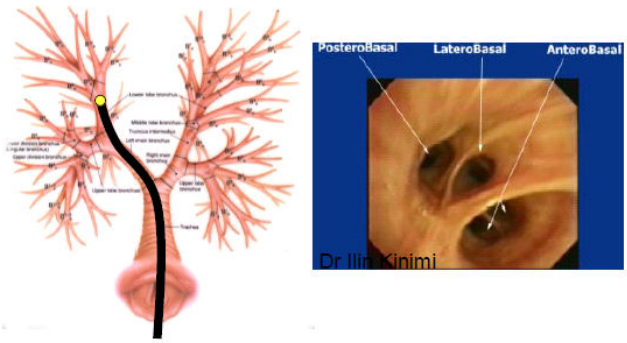
Left upper lobe upper division bronchus



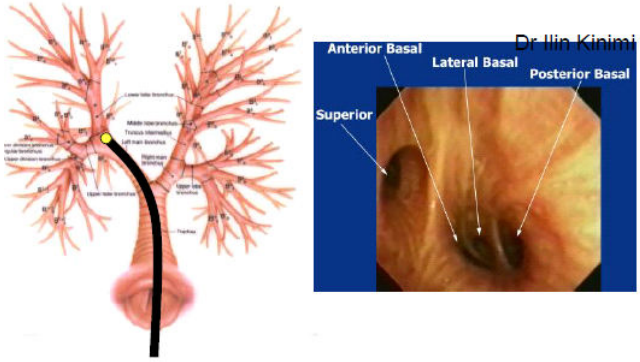
Left lingular lobe bronchus



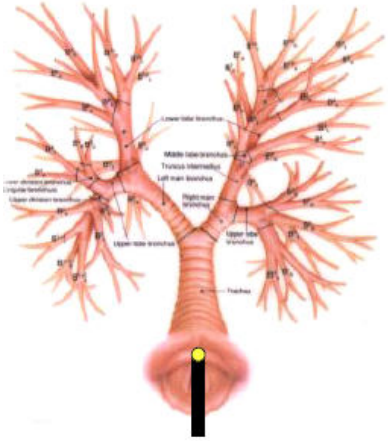
Distal left lower lobe bronchus



Left lower lobe bronchus



Subglottis



Chapter 4

Diagnostic Bronchoscopy: Indications, Techniques And Complications

Sejal Saglani
Professor of Respiratory Paediatrics
National Heart and Lung Institute, Imperial College, London

Indications

Fibreoptic, or flexible bronchoscopy is undertaken in children for several reasons. The indications can be broadly divided split into:

Diagnostic bronchoscopy

1. Unexplained / unusual symptoms and a suspected upper airway abnormality – such as such as stridor since birth, worsening stridor or development of recurrent croup or stridor in older school-age children.
2. Persistent or very symptoms of an otherwise common condition – such as severe asthma or preschool wheeze
3. Recurrent lower respiratory symptoms without an underlying diagnosis – such as recurrent pneumonia or recurrent chest infections
4. To obtain airway samples to identify infection in children with a known diagnosis such as cystic fibrosis

Therapeutic bronchoscopy

1. To treat areas of lobar or sub-segmental collapse
2. To remove a foreign body

The respiratory paediatrician most commonly undertakes flexible bronchoscopy predominantly focussing on lower airway samples / treatments, whilst ENT or thoracic surgeons perform rigid bronchoscopy and micro-laryngo bronchoscopy to treat upper airway and laryngeal lesions and for removal of foreign bodies.

Fibreoptic bronchoscopy is usually undertaken electively in patients when they are clinically stable to investigate symptoms or for diagnostic purposes. They may also require to be undertaken on the intensive care unit. These emergency procedures carry significantly more risk and must be undertaken after carefully considering the benefits against the risks. PICU bronchoscopy indications include assessment of airway narrowing / external compression following cardiac surgery, obtaining samples for culture usually in the immunocompromised child, persistent lobar / segmental collapse, or difficult intubations.

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Techniques

Visualisation of the airway

Most centres in Europe perform paediatric bronchoscopy under general anaesthetic for optimal safety. A skilled paediatric anaesthetist is essential to ensure procedural success. Use of inhaled anaesthesia and a face-mask id ideal is it allows the upper airway to be visualised. The bronchoscope is introduced via the nostril through a facemask. The larynx can vocal cords can be visualised, and it is helpful if the anaesthetist allows spontaneous ventilation without PEEP (Payne Arch Dis Child 2009, Saglani Thorax 2013).

Bronchoscopic procedures

Once the airway anatomy has been assessed, a broncho-alveolar lavage is routinely undertaken in all patients as a diagnostic procedure. BAL samples are processed for cytology, bacterial culture and viral PCR. Endobronchial biopsy is also performed in all patients as a diagnostic procedure, but especially in children with severe recurrent wheezing and asthma – to allow assessments of tissue inflammation and remodelling.

Additional procedures may be performed for clinical or research indications. These include endobronchial brushings to obtain epithelial cells

which can be cultured to assess epithelial function, or may be used for bacteriology or an assessment of the microbiome.

Complications

The most critical step to avoid complications is to ensure all procedures are undertaken under direct vision. (Video of endobronchial biopsy will be shown). Transbronchial biopsy is ONLY performed in patients that have undergone lung transplanatation in whom rejection or bronchiolitis obliterans syndrome is suspected. This procedure is only undertaken for clinical reasons in children, never for research.

The main complications that may occur during the procedure include desaturation or cough, both of which are significantly reduced when the procedure is undertaken under general anaesthesia and are also minimised with an experienced paediatric anaesthetist. Children with upper airway abnormalities may have worse stridor after the procedure, and there is a risk of a fever post procedure, caused mainly by broncho-alveolar lavage. However, none of these complications are prolonged and should not necessitate a longer hospital stay. It should be possible to discharge the patient 4-6 hours after the procedure once they have recovered from the anaesthetic and had some oral intake.

Bronchoscopic samples for research

We have ethics and currently use all bronchoscopic samples that are surplus to clinical requirements for research studies (with parental informed consent). This includes use of BAL and endobronchial biopsies.

Certain procedures are only undertaken for research purposes – with parental consent. This includes endobronchial brushings. Newer research techniques are also being introduced that allow collection of concentrated airway fluid samples. The dilution of BAL often makes detection of inflammatory mediators difficult / unreliable. We are therefore using newer techniques to overcome this including collection of mucosal lining fluid from the airway wall onto an absorptive matrix (leukosorption) which can then be spun down and the fluid can be assessed for inflammatory mediators.

A summary of the techniques used, including videos of their performance, indications and findings from interesting cases and recent paediatric bronchoscopic research will be discussed during the workshop.

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Chapter 5

Flexible Bronchoscopy and Airway Anomalies

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Introduction

Among the airway anomalies, most commonly encountered problems are laryngomalacia, vocal cord paralysis, laryngeal web,laryngeal cleft, congenital subglottic stenosis, tracheoesophageal fistulas and laryngotracheal stenosis.A child with airwayanomaly can have a wide range of symptoms like coughing,noisy breathing (stridor), hoarseness, wheezing, shortness of breath,reflex apnea, choking with feeds and aspiration.Early diagnosis of anomaly is essential to prevent pulmonary damage and associated morbidity.

The diagnostic evaluation of a child with suspected airway malformation should begin with detailed antenatal history. Detailed clinical history and thorough physical examination play an important role in the diagnosis of congenital malformations of the respiratory tract. Persistent wheeze and persistent stridor are the common respiratory sounds that warrant investigation. In small group of patients these malformation goes unrecognized for a long time. Congenital malformations of nose, nasopharynx, larynx and upper trachea pose a medical emergency as they invariably compromise the respiratory function.

In addition imaging studies, CT scan, laryngoscopy, flexible bronchoscopy and barium swallow play an important role in the diagnosis of congenital malformations of the respiratory tract.

Flexible bronchoscopy under local anesthesia has the ability to directly observe airway anatomy / function and make an accurate diagnosis.Flexible bronchoscopy is considered as gold standard tool in the diagnosis of anomalies with dynamic movements like laryngomalacia or tracheomalacia and vocal cord pathology.

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Bronchoscopy is done transnasally after 4 % lidocaine was applied locally to the nasopharynx. During the procedure 2% lignocaine in the dose of 5 mg/kg diluted with equal volume of normal saline is instilled by "Spray and proceed technique" through the working channel. Supplemental humidified oxygen is administered by keeping the oxygen catheter closer to the other nostril and saturation is monitored by pulse oximetry. Common airway anomalies and the contribution of flexible bronchoscopy in the diagnosis are discussed below.

Laryngomalacia

Laryngomalacia is the most common congenital laryngeal anomaly which causes stridor in infants. It is considered as a benign cause of inspiratory stridor. Immaturity of cartilage results in collapse of supraglottic structures (arytenoids, epiglottis, and ary-epiglottic folds) inwards during inspiration and results in low-pitched inspiratory stridor which worsens with agitation, crying and feeding. The noisy breathing in infants with laryngomalacia improve with sleep or prone position(Fig.1).

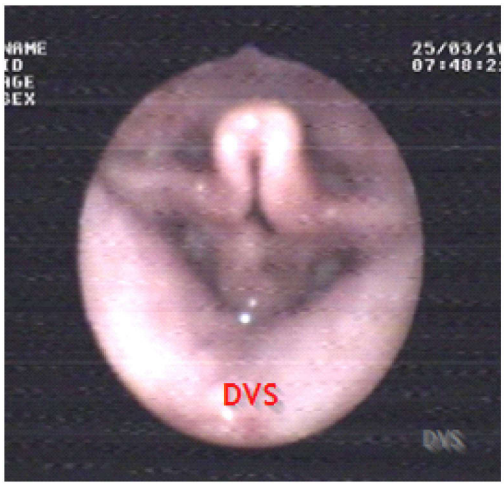

Fig.1 Bronchoscopic view of laryngomalacia

The symptoms usually become apparent by 2 weeks of age, worsen during first few months and then generally resolve by 12 to 18 months of age. If stridor presents in the first week of life, anomalies other than laryngomalacia should be considered. The diagnosis is confirmed by outpatient flexible laryngoscopy or bronchoscopy. Epiglottis is omega shaped due to shortened aryepiglottic folds or it may beds over the glottis. Children with mild laryngomalacia (majority) will only have stridor without any other serious symptoms. In severe cases the entire supraglottic structures may sink into the glottic opening with apparent life-threatening events.

Subglottic stenosis

The subglottis extends from the lower surface of the true vocal cords to the lower surface of the cricoid cartilage. Subglottic stenosis may be classified as either acquired or congenital. At times it becomes difficult in differentiating a congenital subglottic stenosis from acquired one when the neonate is subjected to emergency intubation. Majority of subglottic stenosis are acquired (95%) and the most common cause is difficult endotracheal intubation (iatrogenic due to intubation injury), associated with inflammatory-type response.

Congenital subglottic stenosis is uncommon.

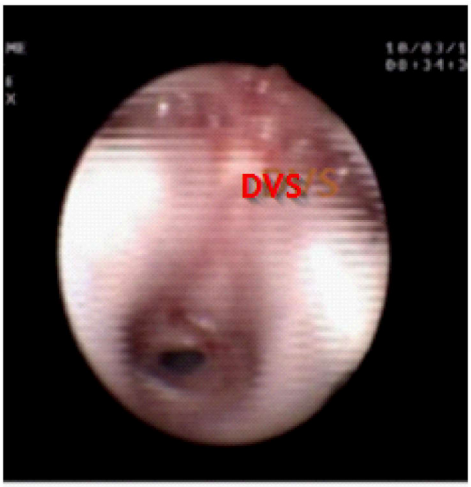

Fig.2. Bronchoscopic view of subglottic stenosis

Membranous type is mild, usually circumferential and consists of fibrous soft-tissue. The cartilaginous type is the severe one and requires corrective surgery. If the stenosis is severe, it may present with noisy breathing and respiratory distress since birth.

Tracheomalacia

Tracheomalacia is characterized by flaccidity of the tracheal support cartilage which leads to tracheal collapse especially when increased airflow is demanded. Due to defective cartilage support, in airway malacias, the contour of airways is maintained by the bronchial smooth muscle tone. If the condition extends further to the main bronchi, it is termed tracheobronchomalacia and if localized to one main bronchi without involving trachea, termed as bronchomalacia.Primary tracheomalacia is caused by congenital immaturity of the tracheal cartilage,when previously normal cartilage undergoes degenerationit is named as secondary tracheomalacia.

Tracheomalacia should be considered when unexplained symptoms of wheezing or coughing are present in young infants, especially if the symptoms start four to eight weeks after birth and persist without signs of viral infection. Unlike the polyphonic wheezing that is heard in bronchiolitis, in infants with airway malacia, low-pitched monophonic wheezing is demonstrated over the central airways. In a child with tracheomalacia the monophonic wheeze is loudest over the trachea.

Infants with tracheomalacia presenting with wheeze may not improve with beta agonist nebulization as beta agonists relax the bronchial smooth muscle. In fact, repeated use of beta agonists can actually aggravate the wheeze in these situations by reducing the muscle tone further.Flexible bronchoscopy done under local anesthesia is the gold standard for the diagnosis of dynamic airway anomalies. Airway malacia is diagnosed when there is a 50% reduction in luminal diameter during expiration(Fig.3).

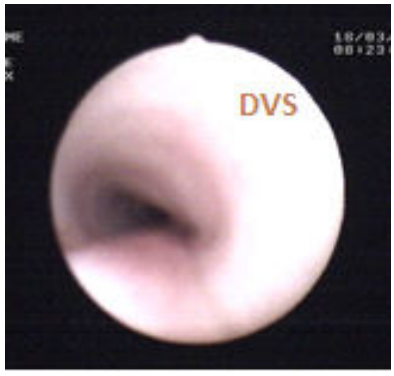

Fig.3. Bronchoscopic view of tracheomalacia

H-type of tracheoesophageal fistula

H-type fistula is the third most common (4-6%) variation of tracheoesophageal fistula (4-6%). H-type fistula which consists of a tracheoesophageal fistula

without esophageal atresia. The fistula is oblique, usually runs upwards (from esophagus to trachea) and occurs anywhere between the level of the cricoid cartilage and the middle part of the esophagus (Fig.4).

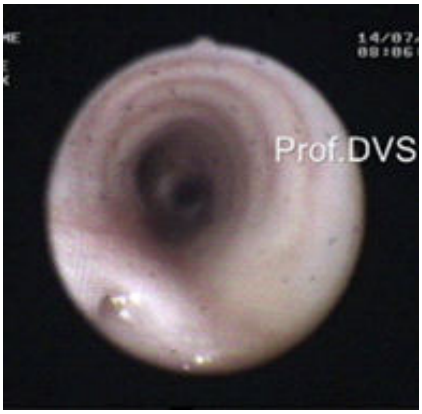

Fig.4. Bronchoscopic view of H- type of tracheoesophageal fistula

Recurrent respiratory infections and hospitalisations for recurrent pneumonia are the common features. Strong suspicion and fiberoptic bronchoscopy helps in the early diagnosis, otherwise the condition may be missed for many years

Tracheal diverticulum andtracheal bronchus

Congenital tracheal diverticulum is a supernumerary bronchus presenting as an out pouching of the right tracheal wall and end blindly. It may be asymptomatic or present with cough, recurrent respiratory infection and wheeze (Fig.5).

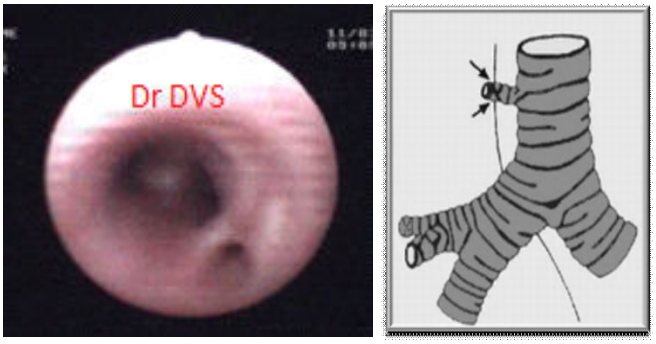

Fig.5. Bronchoscopic view of Tracheal diverticulum and figure depicting tracheal bronchus

If the anomalous bronchus originates from the right lateral wall of trachea, and supply the entire upper lobe or its apical segment, it is named as tracheal bronchus. The diagnosis of tracheal bronchus should be considered in cases of persistent or recurrent pneumonia of right upper-lobe. Most children with tracheal bronchus can be treated conservatively but if they are symptomatic surgical excision is necessary.

Imaging techniques such as high-resolution computed tomography and three-dimensional reconstruction of the airway may suggest the diagnosis, but bronchoscopy invariably confirms the diagnosis.

Vascular ring due to double aortic arch

The term vascular ring is used to describe vascular anomalies that result from abnormal development of the aortic arch complex.The most common forms are double aortic arch and a right aortic arch with an aberrant left subclavian artery.

The double aortic arch is the most common complete vascular ring, encircling both the trachea and esophagus which compresses both. Respiratory symptoms predominate and dysphagia may be present (Fig.6).

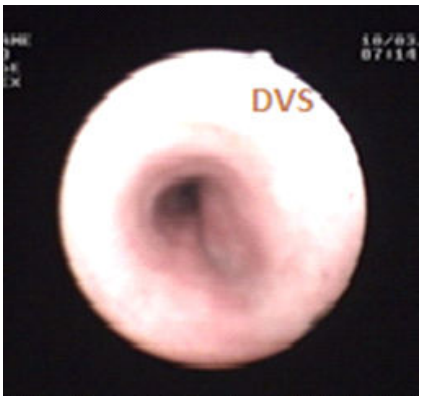

Fig.6. Bronchoscopy demonstrating extrabronchial mass

Several methods can be employed to diagnose and evaluate the underlying vascular and tracheo-bronchial anatomy. Barium oesophago-graphy, echocardiography, cardiac catheterization, and bronchoscopy are used to evaluate the functional component of the compression and detect airway abnormalities.The diagnosis may be suggested by barium esophagram that shows an indentation of the esophagus by the vascular ring. Bronchoscopy by demonstrating extrabronchial pulsatile mass suggests vascular etiology.

Contrast-enhanced CT angiograms with 3D multiplanar reformat (3D-reconstruction) of children with a double aortic archnot only confirms the diagnosis but also provides the information needed for surgery.

Other vascular anomalies include the pulmonary artery sling, which also requires surgical correction. The most common open (incomplete) vascular ring is the aberrant right subclavian artery.

Tracheal stenosis

Tracheal stenosis comprises a wide range of tracheal abnormalities but the common denominator is congenital narrowing of the trachea. Based on the features like complete tracheal ringsand the involvement of the pars trachealis, the narrowness of the trachea, and the extent of tracheal involvement, many distinct types were identified(focal, generalized, funnel-shaped).

Congenital subglottic stenosis, a variant of tracheal stenosis, usually presents immediately after birth. Other forms of tracheal stenosis include funnel shaped trachea, stenosis with complete cartilage rings and tracheal webs.

In our experience the circular trachea (long-segmentminor variant tracheal stenosis with complete tracheal rings with less pars trachealis muscle) are frequently observed, presenting with noisy breathing(biphasic stridor), respiratory distress requiring repeated hospitalisation without a definitive diagnosis. Multidisciplinary approach avoid repeated hospitalizations.

Pulmonary hypoplasia and agenesis

Pulmonary hypoplasia denotes incomplete development of the lungs, resulting in abnormal low number of alveoli. It may be secondary to other fetal abnormalities that interfere with normal development of the lungs which include diaphragmatic hernia, congenital cystic adenomatoid malformation, fetal hydronephrosis, and mediastinal tumor.

Chest radiograph shows ipsilateral shift of the mediastinum due to volume loss oflung. In Pulmonary hypoplasia, the orifice of involved main bronchus is narrow and get pruned due to underdevelopment which maybe demonstrated by fiberoptic bronchoscopy (Fig.7.)

Agenesis of lung is a primary defect in organogenesis where the affected side demonstrates complete absence of the bronchial system and lung. Here the trachea continues to bronchial system without carina (Fig.7). Pulmonary aplasia is category in between hypoplasia and agenesis where bronchial stump and carina are visualized without bronchial

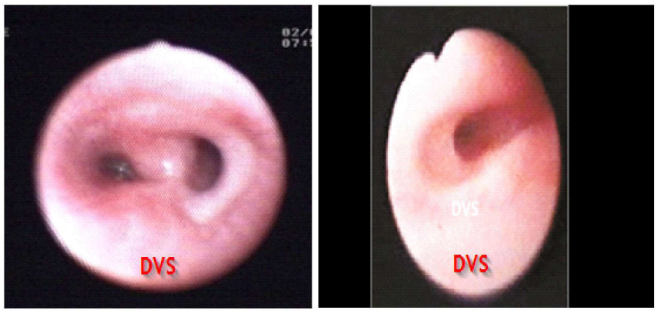

Fig.7. Bronchoscopic view of pulmonary hypoplasia left and right lung agenesis

system on the affected side. Pulmonary hypertension complicates lung agenesis. Right lung agenesis has a higher morbidity and mortality.

Multidisciplinary approach involving pediatrician, pulmonologist (bronchoscopist), and intensivist before bronchoscopy, may result in early diagnosis andavoid unnecessary investigations.

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Chapter 6

Understanding Interventional Bronchoscopy

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Keywords : Bronchoscopy • Interventional bronchoscopy • Flexible bronchoscopy • Endobronchial biopsy • Transbronchial biopsy • Airway stenting • Diffuse alveolar hemorrhage • Airway foreign body • Fetal bronchoscopy

Introduction

Over the last 3 decades, flexible bronchoscopy has replaced rigid bronchoscopy as a diagnostic tool. It is widely used for multiple diagnostic and therapeutic/ interven- tional indications across all ages, and has become an integral component in the training and armamentarium of most pediatric pulmonologists. The routine diag- nostic flexible bronchoscopy includes inspection of the airway, with the obvious advantage that its flexibility affords visualization of distal airways that cannot be reached by the rigid scope. Additionally, flexible bronchoscopy is often performed under various levels of sedation/ anesthesia but mostly without the need for intubation; as such it offers an advantage over the rigid scope. Bronchoalveolar lavage (BAL) has become an integral element of routine bronchoscopy, and offers insight into inflammatory and infectious processes in the airway. This chapter does not deal with diagnostic bronchoscopy or BAL but rather with interventional proce- dures that constitute the demarcation line between the role of the endoscopist and the surgeon. Due to the types of pathologies and respective size of instruments, such interventional uses are much more limited in pediatrics compared to adult practice.

Our target audience is pediatric pulmonologists, who are mostly trained with flexible scopes, yet are frequently requested to weigh on decisions that are on the interface between rigid and flexible bronchoscopy. Specific circumstances that generate this discussion are the approach to retrieval of foreign bodies and placement of airway stents. These topics are broadly addressed in this chapter with the attempt to provide the most updated literature review. A recent review on the topic of Interventional Bronchoscopy in Pediatrics[1] offers a differently weighted perspec- tive by authors whose expertise straddles both rigid and flexible bronchoscopy.

For this chapter we organize the topics into the following broader groups:

(a) Use of Flexible Bronchoscopy for Acquisition of Diagnostic Material.

(b) Bronchoscopy for Removal of Obstructive, Noxious, or Damaging Materials from the Airway or the Lung.

(c) Management of the Narrowed or Obstructed Airway: Debridement, Dilation, and Stenting.

(d) Use of Bronchoscopy for Other Procedures.

(e) New Horizons.

Reference is made to a book chapter on Special Procedures in a textbook of Pediatric Bronchoscopy co-authored by one of us[2], in our attempt to minimize repetition of material that has been previously addressed.

Use of Flexible Bronchoscopy for Acquisition of Diagnostic Material

Endobronchial Biopsy (EBB)

While it is unequivocal that EBB has contributed greatly to the understanding of lung diseases when used as a research tool, a careful review of the literature revealed limited documented role for EBB in clinical practice. This statement is limited to

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diagnostic advantage offered by EBB of tissue taken from the mucosal surface over standard BAL, and excludes biopsies taken from intraluminal lesions or lesions of the bronchial wall. A review of the literature spanning the years since the publication of the chapter on Special Procedures in Pediatric Bronchoscopy[2], revealed no study or publica- tion to alter the opinion expressed in the cited chapter with regard the limited utility of EBB. In a recent review on the role of rigid and flexible bronchoscopy in chil- dren, Nicolai[3] reached a similar conclusion that no clear indications have yet been identified for bronchial wall biopsies, and citing a single paper[4] commented that such role "is currently being elucidated." It is of note, however, that in the above chapter[2] as well as in a previous Editorial on Endobronchial Biopsy in Childhood[5] the authors alluded to the very same paper by Salva et al. They commented that this series of 170 children is the largest study ever published for clinical rather than research indications for EBB; the study offered no information on the clinical utility of the procedure but rather only a conclusion on safety of EBB.

Transbronchial Biopsy (TBB) and Transbronchial Needle Aspiration (TBNA)

Transbronchial biopsy (TBB) is a technique used to obtain lung tissue specimens from distal regions of the lung predominantly for histopathological examination. TBB has become the standard tool for diagnosis of acute rejection in lung transplant recipients with a high sensitivity and specificity. In a retrospective review of 61 pediatric lung transplantation patients who underwent 179 TBB; the procedure yielded clinically useful information—specific pathologic diagnosis in 54% of cases and alteration of medical treatment in 64%. The procedure was deemed a low-risk diagnostic procedure[6].

While intuitively attractive, the use of TBB for interstitial lung disease (ILD) is less well established; and in general, while typically including both alveolar and peribronchial tissue, sample volumes are not adequate for diagnosis[7,8]. Interestingly, however, a questionnaire based review from 38 centers including 131 children with ILD reported utilization of TBB alone or in combination with other procedures in 19.8% of the cases. The report does not clarify, however, to what extent TBB was a key contributor for the diagnosis[9]. In an analysis of the diag- nostic methods used for children with ILD by Fan[8], TBB was used in 6 out of 30 patients, with 3 (50%) being diagnostic for sarcoidosis and bronchiolitis obliterans. The authors did not specify the reason for choosing TBB over VATS or open lung

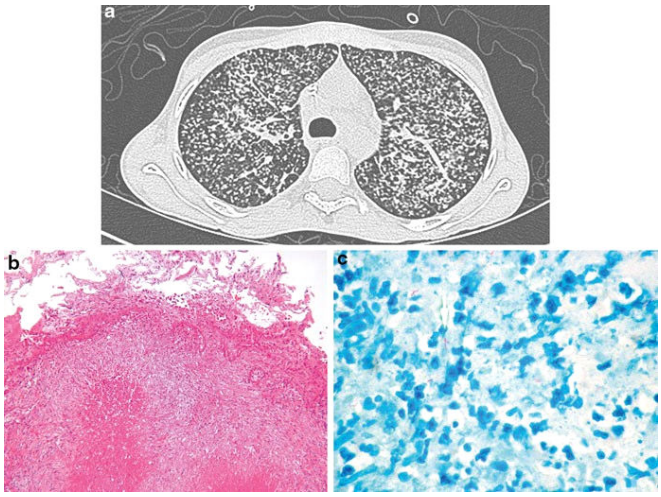


Fig.1. A 21-year-old with HIV infection. (a) Chest CT scan with diffuse nodular disease sug- gestive of miliary tuberculosis. (b) Transbronchial biopsy specimen revealing caseating granu- loma. (c) Ziehl–Neelsen stain revealing acid-fast bacilli

biopsy. A large prospective study of 500 consecutive patients, predominantly adults, in an ILD clinic over a 10-year period reports definitive diagnosis in 85 % of patient – 60% on the basis of invasive procedures. The diagnostic yield of TBB was limited, but 19 % of patients had their diagnosis confirmed by TBB. The yield of TBB for the few children included in this study was also disappointing[10]. Thus, the clinic's current practice is to prefer transbronchial cryo-bioptic techniques (see below) (Personal communication: Ferran Morell, MD).

The size limitation of the instrument that can be used to obtain TBB from small children imposes inherent limitations as well for tissue diagnosis in diffuse lung disease; however, Fig.1 depicts an example of a transbronchial bioptic specimen obtained from an older HIV infected patient that led to conclusive diagnosis of mili- ary tuberculosis.

The technique, safety, and complications (e.g., pneumothorax, hemorrhage, tran- sient pyrexia, and transient dyspnea) of TBB are beyond the scope of this chapter and are reviewed in detail by Tagliaferro *et al*.[11]. The precautions to be kept in mind, however, are that only one lung should be sampled in order to avoid the occurrence of bilateral pneumothorax or hemorrhage[12]; it is also recommended that patients

be hospitalized overnight following the procedure[11].

In a recent review article on childhood bronchoscopy[3], mention is made of new applications and techniques that are being introduced to the pediatric bronchoscopy practice such as endobronchial ultrasound and transbronchial needle biopsy of lymph nodes. The potential uses of endobronchial ultrasound in pediatrics was recently reviewed[13]. It is a minimally invasive technique that allows tissue sam- pling of peripheral lung lesions or mediastinal/hilar masses with high diagnostic accuracy and significantly lower morbidity and mortality compared to alternative approaches. Radial probe endobronchial ultrasound is used in adults for the investi- gation of peripheral lung lesions and could be adopted in children to achieve accu- rate biopsy of such lesions. Linear probe endobronchial ultrasound allows minimally invasive biopsy of mediastinal and hilar lesions.

Ultrasound-guided transbronchial needle aspiration (TBNA) of mediastinal lymph nodes is widely used in adults for cancer diagnosis[14]. This technique is not commonly utilized in pediatric patients; however, successful use of the minimally invasive technique of endobronchial ultrasound-guided transbronchial needle aspi- ration (EBUS-TBNA) was reported when sarcoidosis was diagnosed via material from hilar adenopathy in a 13-year-old child[15]. Given the size of the EBUS bron- choscope, application to younger children is not feasible; the largest report of pedi- atric TBNA for mediastinal lymphadenopathy did not use EBUS[16]. In this prospective study of 28 children (median age 41 months; range 9–168 months) guidance to the site of the biopsy was based on presence of enlarged subcarinal lymph nodes on chest CT scan reconstruction and the visual appearance of the carina. Definitive diagnosis by TBNA was found in 54 % of cases and in 36 % of the cases, cytology performed in the bronchoscopy suite led to the diagnoses. The authors concluded that TBNA is a safe procedure that adds value to flexible bron- choscopy in the diagnosis of mediastinal lymphadenopathy in children.

The limitations discussed above in regards to TBB for the diagnosis of parenchy- mal disease, predominantly in ILD, led to recent successful introduction of trans- bronchial lung biopsy by flexible cryo-probe. The technique allows acquisition of large biopsy samples of lung parenchyma that exceed the size and quality of samples obtained by forceps biopsy[17]. No pediatric reports are yet available, but in an adult study comparing historical controls to transbronchial cryo-biopsy in lung transplan- tation patients, no significant bleeding or pneumothorax occurred following trans- bronchial cryo-biopsy. The mean duration of bronchoscopy using cryo-probe was significantly shorter than the traditional forceps biopsy technique (5 vs. 8 min, respectively). The mean diameter of the specimen taken by forceps in historical con- trols was 2 mm compared to 10 mm obtained using the cryo-probe with no crush artifacts observed; ultimately, overall improved diagnostic value was reported[18].

Bronchoscopy for Removal of Obstructive, Noxious, or Damaging Materials from the Airway or the Lung

Bronchoscopy, both rigid and flexible, has been used for removal of various endog- enous and exogenous materials in the airways that interfere with gas flow or exchange. This segment will cover in detail foreign body aspiration and also touch on less common conditions.

Bronchoscopy for Aspirated Foreign Bodies in Children

Removal of foreign bodies (FB) is by far the most common procedural challenge for the bronchos- copist. The US Centers for Disease Control (CDC) report almost 200,000 accidents per year resulting in nonfatal injury from foreign body aspiration in children less than 10 years of age[19]. These numbers may be even larger in other parts of the world, with a recent report from Algeria, where the authors state: “Foreign body aspiration is a real public health problem in Algeria”[20].

The decision about the need for intervention for suspected FB was addressed in a retrospective study of 160 children[21] aimed at exploring the best clinical and radiological predictors for finding a FB via bronchoscopy. Foreign body aspiration (FBA) was proven bronchoscopically in 122 (76%). In multivariate analyses inde- pendent predictors of FBA were focal hyperinflation on chest radiograph, witnessed choking, and white blood cell count greater than 10,000/mL. Once there is suspi- cion of FBA, Martinot[22] proposed a management algorithm to assist in the deci- sion between flexible versus rigid bronchoscopy based on the experience with 83 children. The authors propose rigid bronchoscopy to be performed first in case of asphyxia, a

radiopaque FB, or association of unilaterally decreased breath sounds and obstructive emphysema. In any other case, flexible bronchoscopy is to be per- formed first for diagnostic purposes. They comment that if the algorithm was applied retrospectively to the 83 children in their study, it would have decreased the negative first rigid bronchoscopy rate to 4%. They concluded that flexible bron- choscopy was a safe and cost-saving diagnostic procedure in children with sus- pected FB aspiration.

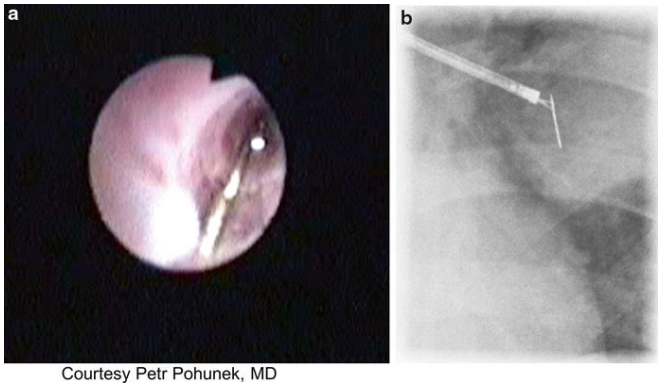
Rigid rather than flexible bronchoscopy has been advocated as the preferred instrument for extraction of foreign bodies since the early days of pediatric bron- choscopy[23] and continues to be the predominant practice[20]. Age appears to be a factor in decision-making. To this end, a study involving 102 infants (mean age 10.5 months, the youngest being 2 months old) with FBA, rigid bronchoscopy was used exclusively with a high success rate[24].

While the value of flexible bronchoscopy, as pointed out by Martinot *et al*.[22] is now widely accepted, the conventional teaching on extraction of aspirated FB points to the primacy of rigid over flexible bronchoscopy for such procedures because of its obvious advantages for visualization and instrumentation. It is con- ceivable, however, that this ongoing preference that emerges in literature is colored by fear of litigation if not abiding by "conventional" practice. This may create a distorted impression of limited value for flexible bronchoscopy. For the purist amongst the readers a "Cochranian" settlement of the question cannot emerge from literature that lacks any attempt for a controlled approach, neither is it likely that such evidence will emerge. The following segment attempts to provide experience on the role of flexible bronchoscopy for FBA.

Successful use of flexible bronchoscopy for extraction of FB has emerged over time, and some authors prefer flexible scopes for FBA. In a review of the Mayo Clinic Pediatric experience (1990–2001)[25] the authors preferentially used flexible bronchoscopy for extraction of FB in children. In their experience the procedure was successful and safe in children who underwent the procedure. The authors advise however that provisions be made to secure immediate rigid bronchoscope availabil- ity should the flexible bronchoscopic procedure be unsuccessful. Encouraged by previous reports of success and motivated by local circumstances or availability of relative expertise in flexible bronchoscopy, another publication[26] espoused flexible bronchoscopy for FBA. While this recommendation may be appropriate for some environments, our own experience is that we do not perform flexible bron- choscopy for suspected FB until immediate availability of rigid bronchoscopy is secured, along the lines of Swanson *et al*.[25].

A valid, and likely incontrovertible indication for preference of flexible over rigid bronchoscopy is FB that is lodged distally, beyond the reach of the rigid bron- choscope. Such conditions clearly justify an attempt of extraction with the more maneuverable flexible scope, yet may pose unique challenges as a result of angula- tion and depth of penetration into the bronchial tree with ever decreasing bronchial diameters as more distal branches are involved. Figure 3.2 depicts successful for- ceps extraction of a pin from a distal airway via flexible scope.

An important complicating factor can be posed by a FB that is imbedded in the surrounding tissue, often a granulation reaction, rendering the object invisible. Such circumstances may lead to surgical intervention and resection of the involved seg- ment. Two reports however address such conditions with the use of interim proce- dures or techniques in an attempt to loosen the embedded foreign body, in both cases an aspirated tooth, followed by successful extraction by using urologic baskets, balloon catheters or by forceps[27,28]. These techniques included the use of topical and parenteral steroids and the use of argon plasma coagulation. The authors did not comment on late outcomes or



Courtesy Petr Pohunek, MD

Fig.2 Flexible bronchoscopic extraction of a foreign body (pin) in a 10-year-old. (a) Bronchoscopic image of the pin within the airway lumen. (b) Fluoroscopic image of broncho- scopic extraction of the pin. (Courtesy: Petr Pohunek, MD. Prague, Czech Republic)

complications of the interim proce- dures. The comment to make however is that these reports potentially understate the risk of dislodgement of the FB, which is often the argument cited to act with mini- mal delay when suspected.

With peripheral location of the FB or with bronchial lumina that are narrow in pediatric patients, visual limitation can complicate use of the flexible scope and instrumentation for extraction of a distal FB. This occurs when forceps passed through the working channel obstructs the field of vision within the narrow bron- chus. Two studies describe the use of fluoroscopy[29] and image intensifier[30] to guide the grasping forceps for extraction of FB embedded in tissue past direct vision.

Sedation/Anesthesia for Foreign Body Associated Bronchoscopy

A detailed discussion on the sedation/anesthesia elements of flexible bronchoscopy is beyond the scope of this chapter. We will limit the comments on the topic to state that diagnostic procedures are mostly done through a nasal route, and recently often via laryngeal mask airway (LMA). A study of 1,947 procedures spanning the years 1988–2003 preferred use of LMA for flexible bronchoscopy in children 2 years of age and older, and complication rates were lower with the LMA (1.9 %) compared to the nasal route (3.5 %)[31].

In the context of this segment on FBA, while LMA is unlikely to be the choice approach when FBA is suspected, its use was reported in five cases in which FB was an incidental finding during a routine procedure and removed without difficulty and without the need to switch from the LMA to the conventional endotracheal tube[32].

Cast/Plastic Bronchitis

Cast or plastic bronchitis is a disorder characterized by formation of tenacious casts within the tracheobronchial tree. Spontaneously coughed up casts can draw attention to this uncommon condition. The distribution can be patchy or involving central seg- ments of the airways when casts assume the shape of bronchial branching. Severe and sometimes life threatening obstruction can result. A comprehensive review of the topic is offered by Madsen *et al*.[33]. The underlying mechanisms involved in formation of casts are varied and overall not well understood; however, children with asthma who have particularly tenacious secretions may be affected and often improve with aggressive asthma therapies. At risk, albeit uncommonly, are children with vari- ous congenital heart disease, and in particular those who undergo Fontan proce- dures. A variety of therapies, all anecdotal, have been suggested for the cardiac-related conditions. It has been claimed that Bronchoscopy for removal of casts that obstruct large airways can be lifesaving but our experience has often found it difficult and extremely time-consuming due to the gelatinous consistency of the deposited mate- rial that renders suctioning, lavage, or removal by forceps difficult or unsuccessful.

Pulmonary Alveolar Proteinosis

Pulmonary Alveolar Proteinosis (PAP) is a rare pediatric disorder consisting of accumulation of phospholipid-proteinaceous material in the alveoli. Primary variety generally presents in infancy and early childhood and the acquired variety manifests in the older age groups. The underlying pathology is related to abnormal surfactant homeostasis and predominantly to defects in GM-CSF signaling. Shah *et al*.[34] and Mallory[35] comprehensively reviewed the topic. High resolution chest com- puterized tomographic scans (HRCT) with “crazy paving” patterns and flexible bronchoscopy with bronchoalveolar lavage are typically the key to the diagnosis when it yields milky fluid from affected segments with the extracellular substance staining with PAS[36]. The cytology is dominated by foamy macrophages[34,35]. The most commonly considered procedure for a therapeutic intervention for PAP is a whole lung which is both challenging and time consuming. There are a number of approaches to the placement of the endotracheal tube (ETT) and isolation of the lung that is to be lavaged [37, 38]; The role of bronchoscopy is to secure the place- ment of the ETT and positioning of the balloon to prevent overflow of fluid into the ventilating lung. In essence a balloon catheter is placed in one main bronchus to seal off the entire lung that is to be lavaged with large amounts of saline, while ventila- tion is entirely dependent on the contralateral bronchus and lung. In exceptional cases where whole lung lavage is not feasible, a more arduous approach is that of direct segmental or subsegmental BAL[39,40].

Lipoid Pneumonia

An extension of the concept of lung lavage was reported by Ciravegna *et al.*[41], in a case of an 8-

year-old diagnosed with exogenous lipoid pneumonia due to aspira- tion of mineral oil that was administered for constipation. The diagnosis was sup- ported by CT scan and BAL fluid (BALF) that was milky-appearing, yielding a high number of lipid-laden alveolar macrophages, as well as diffuse, free droplets of oil between alveolar cells on histology. Lung lavage was performed in the affected segments resulting in rapid clinical and radiologic improvement. A broader experience for the same diagnosis was reported in a study of 10 children with lipoid pneumonia secondary to mineral oil aspiration[42]. The authors took a stepwise BAL approach, which resulted in overall favorable outcomes.

Other Exogenous Foreign Material Aspiration

Sand aspiration to the lung in a 3-year-old with near-drowning was reported[43]. BAL was done when the child continued to have persistent wheezing and high venti- latory requirement and sand was detected in the BALF. Sequential lung washing fol- lowed by exogenous surfactant led to rapid improvement and subsequent recovery in PFTs. In a reported case of accidental instillation of activated charcoal into the lung by a misplaced gastric tube[44], an attempt was made to lavage the charcoal from the lung. While charcoal particles were observed in the BALF, the therapeutic effect could not be assessed since the case was complicated by severe pleural involvement.

Management of the Narrowed or Obstructed Airway: Debridement, Dilation, and Stenting

Impingement on airway lumen by tissue projecting into the lumen can result from various types of mechanical irritants and inflammatory processes both likely compounded by infection. Mechanical irritants and inflammatory processes in the airway lumen can both produce granulation tissues which with or without secondary infection can cause impingement on the airway lumen. Granulation tissue can follow irritation caused by an aspirated foreign bodies, endotracheal tubes, trache- ostomy cannulas, and at surgical sites. These conditions can be approached by debridement using the forceps via flexible bronchoscope, compression by high- pressure balloon catheters and ultimately laser photoresection.

An example of granulation tissue following bronchial anastomosis in lung transplantation being treated using gentle excision by forceps via flexible bronchoscopy is presented in Fig.3. Caution should be exercised since bleeding may be a complication and use of laser therapies that offer various option should be considered[1]. Soong *et al*.[45] described successful treatment of obstructive fibrinous tracheal pseudomembranes complicating central airways in 8 children following prolonged intubation using a combination of forceps, balloon and laser. Flexible bronchoscopic breaching, debridement, and dilation of what was assumed to be inflammation-related obstructive membranes was described in patients with CF and other post-infectious or inflammatory lesions[46,47].

The use of balloon dilation for airways can be considered for a variety of conditions both congenital and acquired in which the airway wall is narrowed[48,49]. The procedure can be done under bronchoscopic vision, but a radiologic approach has also been proposed[48]. An example of a bronchoscopic view of balloon dila- tion is presented in Fig.4. Such pathology may recur and eventually may require stent placement. Importantly, balloon dilation can be considered for congenital nar- rowing of central airways such as complete tracheal rings[50]. This procedure should however be viewed as a surgical intervention since the risk of laceration and fracture of tracheal rings would require extreme

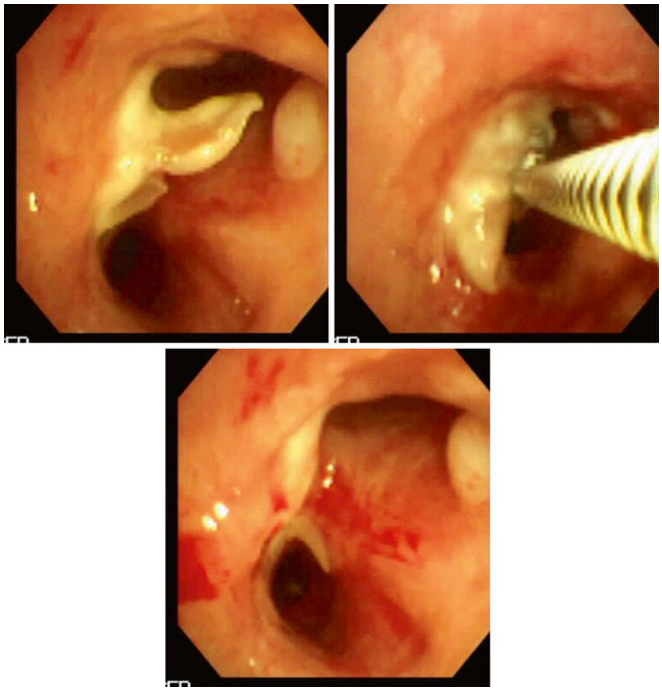


Fig. 3. Forceps debridement of postsurgical (anastomotic region after lung transplantation) scar tissue. The lower image shows patency of the airway lumen after the procedure

caution. While evaluation of long-term outcome of balloon dilatation in adults is published, the indications and conditions of the procedure are so widely different from those in pediatric patients such that it appears unreasonable to extrapolate the results. Thus, little information about the long-term results in pediatric patients is available[51].

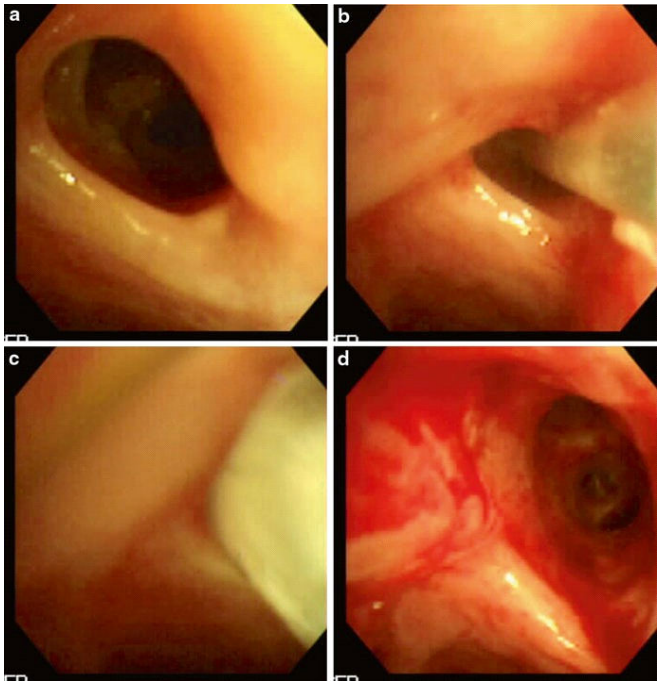


Fig.4 (a–d) Improved patency is gained by high-pressure balloon dilation (c depicts balloon in place) in the narrowed bronchial anastomotic segment of a patient who underwent lung transplantation

Placement of Stents in the Airway

Over the past two decades there has been a growing body of information on the use of tracheo-bronchial stenting in pediatrics that has slowly gained recognition as an accept- able technique for the treatment of central airway obstruction, however, there[52]. Stents though an attractive proposition continue to be a topic of discussion and debate in the pediatric pulmonary practice as their limitations generally render them unready for prime time.

Stenting of the airway has been used successfully in adults, and has been considered as an attractive alternative in children. Fundamental differences of pediatric compared to adult use include the benign nature of most stenoses which do not alter life expectancy,[52] the narrow and soft airways of children, that improve with airway growth and the shift of mediastinal vessels[53] and also the required long-term tolerance and adaptation to growth. These differences may significantly alter the therapeutic balance, calling into question the precise role stents play in the treatment of airway obstruction in children. However, recognition that situations exist in which no other options are available has led to increased use of this technique in pediatrics.

Obstruction of the airway is the result of abnormalities of the airway wall, intra- luminal causes or extrinsic compression[48]. Prior to stent placement, the airway is typically evaluated with bronchoscopy or bronchography and the chest evaluated for causes of compression with echocardiogram, CT scan or MRI[48].

Stents may be placed by either interventional bronchoscopists or invasive radiolo- gists with relative advantages and disadvantages to each, and often cooperation between them. Plastic/silicon stents were initially available. The first use of a metallic stent in pediatrics was reported in 1988[54]. Currently biodegradable stents are being introduced and offer potential new horizons[55]. The advantages and disadvantages of the various stents are further discussed below.

Most reports of use of stents in pediatrics are case reports or small series. In the absence of randomized clinical trials or larger series, it is difficult to compare the efficacy and tolerance of metal versus silicone airway stents in children; furthermore mortality rates in recipients of stents are generally high given that indications for stenting are mostly options of last resort[53].

Indications

The indications for stent placement as outlined in the adult literature include: extrin- sic stenosis of central airways with or without intraluminal components due to malignant or benign disorders; complex, inoperable tracheobronchial strictures, tracheobronchial malacia, palliation of recurrent intraluminal tumor growth, and cen- tral airway fistulae (esophagus, mediastinum, pleura) [56]. In pediatrics the common causes of obstruction leading to stenting are the following:

Congenital stenosis – This results from abnormal cartilage rings (small and com- plete), or compression by abnormal vessels, such as pulmonary artery slings. Operative repair is the standard of care, however, recurrent obstruction is often encountered, the result of malacia or restenosis. Balloon dilations and stenting is

thereafter sometimes the next step[48]. It may also be the result of accumulation of metabolic products such as seen in mucopolysaccharide storage disorders[57,58]. It is usually agreed that in cases involving vascular compression, relief of the compression is the initial step[58].

Tracheal or bronchial malacia – This usually resolves by 2 years of age; however, it may require intervention when diffuse and/or requiring treatment with long-term CPAP via tracheostomy[48]. In a series of 105 patients who underwent aortopexy for treatment of tracheo-bronchomalacia, five patients required stenting after failure of aortopexy[59]. Additionally, despite the generally favorable long term prognosis of airway malacia, severe "dying spells"[60,61], or severe growth retardation[58] may require a temporizing procedure.

Airway obstruction at a site of previous surgery – This usually results from granula- tion tissue and/or fibrosis following patch repair or over suture lines[62]. This condi- tion may occur after lung transplantation at the site of the bronchial anastomoses.

Palliative indications – Stenting is also considered for palliation. This includes patients in whom a lesion is unresectable because of anatomic constraints, meta- static disease or limitations due to overall medical condition; stent placement may be minimally invasive and may provide prolonged palliation[63]. Stenting may allow weaning of ventilatory support and subsequently allow hospital discharge, even if long term survival is not anticipated[48].

Types of Stents

There are several different types of stents with their respective advantages and dis- advantages, different methods of insertion, and varying requirements for follow-up and management.

A plethora of stents have been used in the airways. They can be divided into four major groups.

1. Polymer stents (Dumon, Polyflex)
2. Mettalic stents

   Balloon expandable (Palmaz) Self-expanding (Wallstent)
3. Covered Mettalic stents
4. Hybrid Stents

Stents can also be grouped based on indication, insertion technique, anatomical location or whether removable or not[64]. In 2011 use of a biodegradable polydiox- anone stent was first reported in children[55].

Silicone or silastic stents – These are long tubes that are easy to remove but have prob- lems with luminal occlusion and to a lesser degree migration[48]. The small radii of airways of children require thin walled stents, and when made of silicon these tend to be collapsible and prone to migration[58]. The continuous, non-fenestrated tube interrupts mucociliary clearance for which humidification and inhalation, inhalation of, mucolytic agents including DNase have been suggested, but their efficacy has never been documented[58]. Insertion is with a rigid bronchoscope, a device most pediatric pulmonologist are not familiar with such that placement is done only in centers with specialists trained in this procedure. Fayon et al. reported their experience with a custom manufactured polysiloxane (Tracheobronxane) stent in 14 children with success and failure rates equal at 43 %; the latter due to migra- tion or obstruction.

In summary, extreme caution is needed when using these stents that remain attractive mainly for short-term use in the hospital setting postoperatively. Stents with internal support structures in their walls (Polyflex) aim to resolve some of these problems; however, migration and mucous impaction remain significant and limit their use[58].

Metallic stents – These were initially developed for vascular lesions. They are relatively easy to deploy by bronchoscopy or bronchography, are thin walled and their mesh structure allows for continued mucociliary clearance and ventilation even when the stent covers bronchial openings[58]. Their main disadvantage is difficult removal as early as several weeks after implantation due to mucosal overgrowth[58] albeit removal has been documented up to 5 years after insertion [64]. Other problems include breaks due to material fatigue and migration into surrounding organs[58].

The most frequently used balloon expandable stent is the Palmaz stent. It is non- elastic and made of stainless steel. Its main advantage in pediatrics is the ability to overdilate as the child grows, while its disadvantages are fracture[48] or deforma- tion with cough[58]. They have the advantage over the self-expanding stents (such as the Wallstent; see below) in that they do not exert constant outward pressure after placement, which is implicated in erosion and hemorrhage[58]. In a 5-year pub- lished experience

with this stent; a total of 30 stents were placed via rigid bronchos- copy in 16 patients[62] with airway malacia as the most common indication. Repeat procedures were required in several patients due to obstruction, development of granulation tissue and migration.

The *Wallstent* is made of thin wire, is very flexible and compressible but re- expands after compression. Placement is not easy as it shortens by 20-40% during placement but its flexibility renders it easily adaptable to curved airways compared with the rigid Palmaz stent[58]. It is accepted that these stents are more appropriate for compression by vascular structures where the pulsatile mass against a rigid stent, such as Palmaz, could lead to vascular erosion[48]. The disadvantage of Wallstent is that it cannot be dilated as the child grows and may therefore result in stenosis if left in the airway of an infant[48] and likewise, in the trachea of young children[58].

Nitinol stents (Ultraflex) are another form of self-expanding metal stents made of a shape-memory alloy in either covered or uncovered forms and expand at body temperature. They do not exert continuous outward pressure but dislocation is less common than with silicon stents and is proposed as the optimal material for the human airway. While fewer complications have been reported with this kind of stent, it is not clear whether this reflects veritable superiority or the relatively infre- quent use[58]. They too cannot be dilated so they must be replaced as the child grows[64].

Polydioxanone biodegradable self-expanding stents were recently first reported in children[55]. Polydioxanone is a semicrystalline polymer of the polyester fam- ily. The predicted and observed degradation time was 15 weeks. These stents are inserted with a specific introducer that is too large to allow direct vision. As with the other self-expanding stents their final size is hard to predict.

Of note is that there are two stent types fraught with problems and are no longer in use: Strecker, a tantalum stent, and Gianturco a steel stent with external hooks[58].

Method of Insertion

Stents may be placed with bronchographic guidance. Bronchography has several advantages over bronchoscopy; it provides accurate measurements and can also asses the airway distal to the obstruction[48]. Bronchoscopy offers the advantage of direct visualization. The thicker silastic stents can only be inserted by rigid bronchoscopy. Imaging, usually by bronchography but also CT or MRI scans is necessary prior to the procedure for planning optimal placement and to determine the size of the stent. The ERS/ATS published a statement with the training requirements for bron- choscopists performing stent placement. This included ample experience with rigid/ flexible bronchoscopy and endotracheal intubation[56]. This implies that the procedure should only be offered in a few specialized centers[58].

Complications

Silicone stents. Occlusion and migration are the most common complications[48], the latter linked to the very nature of these stents that, in contrast to metal stents, do not become incorporated into the wall[53]. Migration was mostly encountered in the Fayon study with small caliber stents compressed by high-pressure vessels; pointing towards avoidance of the use of silicone stents in these circumstances. Granulation tissue is infrequent, moderate, and localized to the tip of the stent, and largely observed when the stent is too mobile[53].

Metallic stents. The potential for metallic stent erosion through the thin bron- chial wall is a subject of discussion, but with scant documentation. Wells *et al.*[65] described two patients with associated heart disease and stenting of the left main stem bronchus (LMSB). Both patients presented with ruptured pseudoaneurysms adjacent to the stented bronchus; this complication was likely compounded by adja- cent bronchial collateral vessels in patients with cyanotic heart disease. Stents might also erode into surrounding structures, with possible exsanguinating hemorrhage from bronchovascular fistulae[63]. Geller *et al.*[66] described three deaths due to massive tracheal bleeds in nine Palmaz stent placements occurring months after placement. All had concomitant tracheotomies. They concluded that tracheotomy and Palmaz stent placement in the airway might increase tracheal colonization/ inflammation and hence friability and proposed that tracheotomy be viewed as con- traindication to use of a Palmaz stent.

Granulation tissue formation is usually mild when the stent is placed against an intact airway wall [48]. It is best treated with intermittent ballooning, which

may be insufficient. Use of lasers should be avoided due to heat transfer through metallic objects [58]. Stent fracture is a specific complication of the metallic balloon expandable – stents.

Metallic self-expandable stenting requires long-term management to correct potential stent problems that also include migration or obstruction by inspissated secretions, granulation tissue, or tumor. Peng *et al*.[67] in a 5-year experience emphasize the role of flexible (vs. rigid) bronchoscopy in pediatric intensive care patients with stent repair as the second most common indication for procedures.

Information regarding status of the airways after stent placement can be obtained through imaging and pulmonary function testing. The need for routine bronchosco- pies to assess for complications and formation of granulation tissue is unclear, and while discouraged in adults, may be called for in pediatrics due to the smaller diam- eter of their airways. It remains unclear whether the finding of granulation tissue in asymptomatic patients warrants prompt treatment[58]. The risk exists that bron- choscopy through a stent may lead to its dislocation or damage[62].

Mitomycin C has been reported to inhibit fibroblast proliferation in granulation tissue formation in human cells[68] and bronchoscopic application of mitomycin C as adjuvant treatment for benign airway stenosis has been reported [69]. Curiously we have found no report of the use of this agent, for the prevention of granulation formation following stent placement, in the literature.

Since stenting is usually a last resort in patients that are a priori in critical straits, it is difficult to determine the mortality rate related directly to stent placement. A tentative estimate of 13 % mortality was stated by Nicolai [58] but remains difficult to ascertain.

Use of Bronchsocopy for Other Procedures

Sealing of Fistulae

Recurrence of tracheoesophageal fistula (TEF) after the original surgery is often dif- ficult to demonstrate and is mostly managed by repeated surgery. Literature in recent years[1,70,71] points, however, to a bronchoscopic option of sealing the fistula by collagen glue, fibrin, cyanoacrylate, or sclerosing agents. To improve results prepa- ratory mucosal priming via brush abrasion, laser or electrocautery is proposed, which is subsequently followed by the "glue." While more than one endoscopic procedure is often needed, the endoscopic repair is an attractive alternative to open surgical repair. The cited publications employ rigid bronchoscopy for the procedure. Goussard *et al.*[72] reported fibrin glue closure of a persistent bronchopleural fistula that complicated pneumonectomy in a 16-year-old girl with post-tuberculosis bron- chiectasis. While we were unable to find reports on the use of flexible bronchoscopy for primary or secondary closure of TEF, this report may open new avenues for such repair, in particular where H-type fistulae may be involved.

Bronchoscopic Sealing of Pneumothorax

The recent emergence of necrotizing pneumonia brought about a substantial increase in complications with bronchopleural fistulae (BPF)[73], as many as 12 % of hos- pitalized cases with necrotizing pneumonia were reported to suffer this complication[74]. Curiously, no reports of persistent leakage or the therapeutic approach thereof has been published. Endoscopic approaches to persistent pneumothorax with vari- ous sealing materials have been reported since the 1970s[75]. To identify the bron- chus leading to the air leak, a fiberoptic bronchoscope and a balloon catheter are used while diminution of the air leak with repeated inflations of the balloon is followed[76]. An interesting novel approach to identification of the leaking segment when balloon occlusion fails is the use of a capnographic catheter that is passed into the airway[77]. Once identified and with inflated balloon, the sealant can be injected through the distal port into the airway, or alternatively manipulated via forceps.

A recent review of the therapeutic transbronchial approach via use of bronchial valves[78] appears less feasible in the pediatric age group, but various sealants have been used in multiple reports of adult patients. Use of fibrin glue with rapid response was reported[76]. Wiaterek *et al*.[79] reported placement of several alternating layers of an absorbable hemostat (knitted fabric prepared by controlled oxidation of cellulose-Surgicel; Ethicon) within the segment of interest using bronchoscopy for- ceps followed by catheter injection of 3 mL of the patient's blood onto the absorb- able hemostat to create an occluding blood patch. Rigid bronchoscopy is predominantly used but some of these procedures utilize flexible bronchoscopy. The youngest patient we could identify in the literature who underwent such

procedure was an 11 month old who developed BPF 3 weeks after surgery for cystic adenomatoid malformation. The infant was successfully managed with porcine dermal collagen combined with fibrin glue plug[80].

Control of Diffuse Alveolar Hemorrhage

The topic of bronchoscopy for airway and pulmonary bleeding has been covered in a chapter previously alluded to [2]. At the risk of some repetition we wish, however, to highlight a novel bronchoscopic intervention for intractable lung bleeding.

Based on previous experience using systemic administration of recombinant fac- tor VII (rFVIIa) to effectively treat patients with pulmonary bleeding, its use has been extended to direct intrapulmonary instillation of rFVIIa in recalcitrant cases of diffuse alveolar hemorrhage (DAH)[81,82]. We recently used rFVIIa as an inter- vention of last resort to control unremitting diffuse pulmonary hemorrhage in two cases; a 16-year-old patient with acute myelogenous leukemia[83] and a 2-year-old patient with relapsed acute lymphoblastic leukemia[84]. We used the protocol pro- posed by Heslet[81] that entailed administration of rFVIIa into both main stem bronchi at a dose of 50 mcg/kg diluted in 50 mL of normal saline; for the smaller or younger patient, we opted to dilute in 25 mL of normal saline. The dose was divided in two equal aliquots and separately instilled to the main bronchi. To cite directly from our report: "Hemorrhage was visualized bronchoscopically, and its resolution following the treatment was immediate, unequivocal, and definitive"[83]. In a recent review of the topic, emphasis is made that multiple reported cases have been shown to respond promptly with the added safety of absence of thromboembolic complications when rFVIIa is administered bronchoscopically as opposed to systemically[85].

Segmental Bronchography

Bronchography is rarely used in the era of CT scan, and in particular since the advent of 3-D reconstruction. However, Bramson *et al*.[86] reported the use of flex- ible bronchoscopy and instillation of contrast material via the bronchoscope chan- nel. They pointed out that bronchogram elucidated findings that were unclear from other imaging procedures. A more recent study[48] of bronchography via flexible bronchoscopy emphasized the advantage of the procedure particularly when a laryngeal mask airway is used. In our previous publication[2], we stated the utility of segmental bronchography for cases in which CT scan failed to elucidate details of the finer internal structure of bronchi or their connections. The flexible broncho- scope was advanced peripherally and wedged into the bronchus feeding the area of interest, and the dye was injected through the working channel under fluoroscopy with images taken in rapid sequence.

In the study by Bramson *et al.*[86], Dionsil – a lipid based contrast material was used. This agent is not readily available at present; instead contrast materials used in arteriography such as loversol (Optiray, Mallinckrodt, Inc.) is used. McLaren *et al.*[48] report use of very small volumes of contrast, as little as 1 mL or less, for diagnostic bronchography in small children, except for interventional procedures where larger volumes of contrast (5 mL or more) may be required. They recommend isotonic contrast such as iotrolan (Isovist, Schering, Burgess Hill, UK), but point to safety of widely available, moderately hypertonic agents (such as Omnipaque, Nycomed, Nycoveien, Norway). In our experience the resolution of images with the non-lipid contrast materials is inferior to that achieved with the older materials; moreover, they fade rapidly, but with rapid sequence image acquisition, the information sought can usually be obtained. Figure 3.5 exemplifies segmental bronchography that established the diagnosis of unsuspected cardiac bronchus.

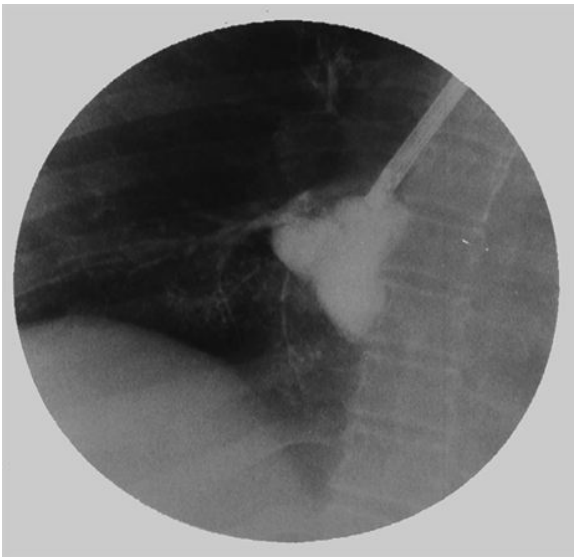


Fig.5 Segmental bronchography. Contrast material is injected via a wedged bronchoscope to establish airway connection, in this case revealing a cardiac bronchus

New Horizons

Fetal Bronchoscopy

A distinctly novel horizon in pediatric bronchoscopy is a recent report by Quintero *et al.*[87] of a first fetal bronchoscopy. This in utero bronchoscopy was undertaken with the hope of salvaging the lungs of a 32-week gestation fetus diagnosed with congenital lung abnormalities that were deemed incompatible with extrauterine sur- vival. The left lung was taken up by a mass that caused mediastinal shift and as a result, extremely small right lung (Fig.6). Ultrasound and fetal MRI suggested the possible presence of bronchial atresia or congenital cystic adenomatoid mal-formation (CCAM). Bronchoscopy resulted in intraoperative expansion of normal lung parenchyma in both the right and left lungs, with dramatically improved and normalized lung growth until birth. Postnatal CT and MRI were suggestive of extralobar pulmonary sequestration with cystic areas, with a feeding vessel stem- ming from the descending aorta. The lesions were eventually resected at 10 months of age confirming the presumptive diagnoses. The authors submit that the bronchos- copy established airway patency in obstructed airways and restored amniotic fluid flow to the lung periphery – a key element in lung development. This notion is based on increasing recognition that bronchial obstruction may be the common pathway to the frequently overlapping congenital anomalies of the lung, including CCAM, intralobar sequestration (ILS), extralobar sequestration (ELS), and lobar emphysema (LE). Several studies support bronchial atresia as the unifying mecha- nism responsible for these malformations, termed “bronchial atresia sequence” [88, 89]. This was corroborated by a report[90] stating that lesions already diag- nosed with peripheral bronchial atresia (radiologically and/or pathologically) were found to have frequent microcystic changes consistent with small cyst type CCAM. The same authors[91] reported a second fetal bronchoscopy at 30 weeks gesta- tion with similar presentation based on US and MRI findings. In this case the thera- peutic effect of the intervention was not as obvious, and only minor improvement occurred until delivery at 40 weeks. Perinatal CT-angiography demonstrated LUL congenital lobar emphysema (CLE), which was subsequently confirmed by pathology after postnatal resection.

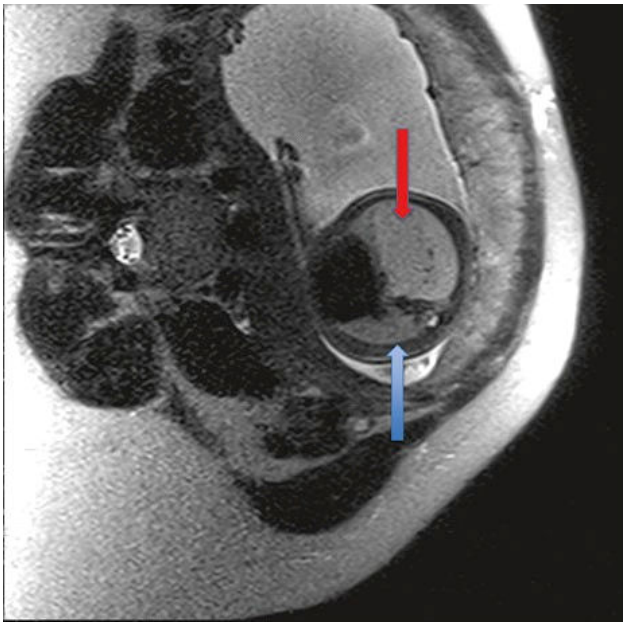


Fig.6 MRI of maternal abdomen with transverse section of fetal lungs in the uterus. Upper arrow points to a large left lung mass. Lower arrow points to a markedly diminished, normally structured right lung

In another study fetal intervention using the fetoscope (vs. bronchoscope) was undertaken to surgically breach an atretic bronchus in utero[92]. While the results are reported to have been favorable, little detail is provided regarding the technical elements of the intervention.

It is not possible to draw conclusion regarding the role of fetal bronchoscopy from this limited reported experience. It appears to be technically feasible with cur- rent instrumentation, and since fetal oxygenation is not dependent on the fetal lung, desaturation during the procedure is not a limiting factor. Cognizant that the intra- uterine course of congenital pulmonary lesions is not easy to predict, we think that fetal bronchoscopy should be reserved for cases in which the size or complexity of the lesion makes extrauterine viability unlikely. It is hoped that further experience will better define the potential role of improving or reversing progression of intra- uterine congenital anomalies with use of this novel intervention.

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