BRONCHOSCOPIC AND ESOPHAGOSCOPIC PROBLEMS IN INFANTS

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The indications for bronchoscopy and esophagoscopy in infants are constantly increasing. During the past ten to fifteen years, many new problems have arisen that are primarily associated with the examinations of the small air and food passages of patients under one year of age. The demands have become more complex and interesting, and the need for diagnostic studies or therapeutic procedures greater, largely as the result of the spectacular advances of pediatric surgery of the heart, lungs and esophagus. This discussion is intended as an evaluation of the endoscopic aspects of some of these problems.

INSTRUMENTARIUM

Laryngoscopes, bronchoscopes and esophagoscopes must vary in design from simple miniatures of the adult instruments to be satisfactory for endoscopic examination of infants. Using the smallest lamps and carriers, bronchoscopes of 3 mm. to 3.5 mm. in diameter and 20 to 30 cms. in length have a special smooth tip to provide an adequate visual lumen without causing laryngeal trauma during insertion through the larynx. Similarly, small diameter infant esophagoscopes with small aspirating channels permit safe examination of the esophagus of an infant of any age. Such examinations necessitate the smallest of accessory instrumentarium such as bougies, aspirators, forceps and even miniature telescopes.

Bronchoscopic and esophagoscopy examinations in infants are preferably done in a

Fig. 1

Bronchoscopic and Esophagoscopy Problems in Infants

completely equipped endoscopic operating room where all types of respiratory emergencies have been anticipated and the equipment for their management is immediately available. Of greatest importance is an adequate tracheotomy set with appropriate size of instruments and tracheotomy tubes for infant respiratory problems. While the endoscopic procedure itself should not necessitate a tracheotomy, surgical relief of an obstructed airway may be neces-

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sitated by the pathology disclosed by the examination. A short tracheotomy tube with a shallow curve has been found more satisfactory in infants than the full curved tubes. Simplification of the plate on the tube facilitates cleansing and routine care.

TECHNIQUES

Endoscopic examinations of the newborn and infant age groups require no anesthesia. Procedures must be rapid, and particular care taken to insure an adequate oxygen exchange. Oxygen should flow into the infant bronchoscope through the oxygen arm on the tube to permit examination in a high oxygen concentration. Esophagoscopy may be complicated by respiratory obstruction because of compression of the trachea unless the procedure is done with a 3.5 or a 4 mm. diameter esophagoscope. Introduction of an intratracheal airway is a safe alternative although rarely necessary if small esophagoscopes are used. Miniature bougies are essential, as are small diameter aspirators and forceps. A No. 8 (French) bougie serves as a lumen finder, this is an indispensable guide into the cervical esophagus, a guide through a tight stricture, or through the cardia into the stomach.

Bronchography in infants under one year of age necessitates special consideration of technic, instrumentarium and media. Lipiodol appears to be most satisfactory radiopaque medium in that it is neither too viscid nor too free-flowing. It is relatively non-irritating and small amounts cast a satisfactory shadow. Anesthesia for bronchography in infants is unnecessary and is merely an additional hazard. A No. 7, 8, or 9 (French) polyethylene tube with very thin walls is passed through the nose and guided into the larynx and trachea under direct vision using an infant laryngoscope. An appropriate sized hypodermic needle serves as an adapter between the tubing and the syringe containing the oil. The oil is injected under fluoroscopic guidance. There is, of course, danger of respiratory obstruction during the examination caused by the catheter or the oil itself. This danger may be enhanced by the pathology for which the procedure is indicated. A laryngoscope, suction pump, tracheal aspirator and oxygen should be available and prepared for use in the fluoroscopic room during all bronchographic examinations of infants. Radiographic record of the examinations are made by spot films and conventional PA, lateral and oblique films.

Fig. 2

BRONCHIAL OBSTRUCTION

The most important indication for bronchoscopy in the newborn as well as in adults is the diagnosis and treatment of tracheal or bronchial obstructions. Evidence of

obstruction of a lung, a lobe or a segment of a lobe is the atelectasis or emphysema of the anatomical unit. However, atelectasis in newborn infants has many implications. Normally, it takes from two days to two weeks after birth for complete aeration of the lung to take place and consequently patchy atelectasis is not in itself an indication for bronchoscopy in the newborn. But if excessive secretions are present on physical examination in addition to patchy atelectasis, rapid tracheal or bronchial aspiration is indicated. Diagnostically, there is more significance to the examination if a segment, a lobe or an entire lung is atelectatic. Bronchoscopic examination is indicated to determine the cause of the obstruction and to remove it if possible. A large quantity of secretion present in the tracheo-bronchial tree of a newborn infant requiring bronchoscopic aspiration should lead to studies to determine the presence of a congenital tracheoesophageal fistula or pharyngeal paralysis. The tracheoesophageal fistula may or may not be associated with a congenital esophageal atresia. A fistula without esophageal atresia may be located at any level between the cricoid cartilage and the tracheal bifurcation. Large fistulae are easily identified. Small

Fig 3

fistulae present more difficulties in diagnosis. They run obliquely with the tracheal opening often at a higher level than the esophageal opening. Once identified, they may be closed surgically, but severe pulmonary problems of aspiration pneumonia often precede the identification of the tract.

Pharyngeal paralysis due to a birth injury or a cerebral agensis may account for excessive bronchial fluids in the newborn. Similarly, laryngeal paralysis, unilateral or bilateral, may be associated with birth trauma, cardiac anomalies, or meningoceles. These paralysis are often responsible for the excessive tracheal mucus for which a bronchoscopic suction is requested. In a recent study, 45 infants with vocal cord paralysis were reviewed. Thirty one of these had bilateral paralysis of which fifteen had complete bilateral recurrent laryngeal nerve paralysis and sixteen partial bilateral paralysis. Of the fourteen unilateral

paralyses, seven involved the right and seven the left cord. The most common etiologic factor of the bilateral paralyses was some type of central nervous system lesion; cerebral hemorrhage, a congenital disease of the nucleus, such as mental retardation, hydrocephalus, meningoceles or cerebral agenesis. Improvement in function was noted in only 25% of cases, primarily in those with the peripheral lesions of birth injury. Unilateral left cord paralysis was most commonly due to an enlarged heart associated with an interventricular septal defect. The prognosis for development of normal cord action was poor. Right cord paralysis was usually due to a peripheral birth injury and recovery was noted in most cases by six months of age. In unilateral paralysis the cry is weak and breathy, the airway fair, but there were episodes of choking and cyanosis during feedings. Bilateral paralysis is characterized by inspiratory stridor, retractions and cyanosis, but the cry is clear. Respiratory obstruction may necessitate a tracheotomy in bilateral paralysis to establish an airway as well as for aspiration of tracheal mucus. Twenty-five percent of the infants with bilateral laryngeal paralysis required feeding tubes because of associated pharyngeal paralysis which rendered them unable to swallow.

CONGENITAL TRACHEAL AND BRONCHIAL ANOMALIES

Advances in pediatric surgery have presented many new bronchoscopic and esophagoscopic problems not known as recently as ten to fifteen years ago. Pulmonary, esophageal and cardiovascular surgery in infants have made great strides. This has required diagnostic and therapeutic endoscopic procedures to aid in determining the indications for such surgery and to assist in the management of some of the post-operative complications.

Evidence of lobar or main bronchus obstruction in infants is seen as atelectasis or obstructive emphysema of the lobe or the entire lung. Congenital anomalies such as absence or duplication of lobes and bronchi may account for the problem of dyspnea, but often additional pathology such as lung cysts or so-called idiopathic emphysema is found. Prior to resection for either persistent atelectasis or obstructive emphysema, the bronchi must be inspected. Search is made for a possible bronchial web or evidence of extra-bronchial compression which would indicate the etiology of the obstruction. From our own records at The Children's Memorial Hospital, it is apparent that the commonest cause for what appears to be segmental, lobar or total lung atelectasis is the absence of that anatomical part. Intraluminal obstruction is relatively rare in this age group. Emphysema of a specific segment, lobe or lung is most commonly due to a cardiovascular anomaly compressing the bronchus leading to the affected part of the lung. Next in frequency is idiopathic

Fig. 4

Fig. 5

pulmonary emphysema, a condition characterized by pulmonary alveolar fibrosis. Five such cases have been seen in infants less than two months of age. This condition is characterized by severe dyspnea, physical and x-ray findings of lobar emphysema. Bronchoscopic examination showed essentially normal bronchi. Lobectomy was necessary because of the severe compression of lobes adjacent to the emphysematous lobe. Recovery was uneventful. In a sixth case, an infant 24 hours of age, a congenital bronchial web was found responsible for a partial obstruction. This was opened by a single dilatation with forceps and then the passage of the scope through the web. The differential diagnosis must consider such congenital anomalies as lung cysts,

bronchial or tracheal cysts which cause external compression of the airways, and the staphylococcal pneumonia with the rapidly changing pulmonary cavities characteristic of this disease. Confirmation of the diagnosis of this disease may be obtained from the bacteriologic examination of the bronchial secretions. Spontaneous pneumothorax, an occasional finding in a newborn infant may be responsible for severe dyspnea and cyanosis. This condition is, of course, a contra-indication to bronchoscopy and is usually promptly relieved by the aspiration of 50 to 75 cc. of air through the chest wall.

BRONCHOPULMONARY SUPPURATION

Bronchopulmonary suppuration in infants aged six months to a year should be investigated as possible fibrocystic disease of the pancreas. This has been recognized as a common clinical entity which involves not only the pancreas, but mucus and sweat secreting glands throughout the body. It is primarily a congenital anomaly of the pancreas characterized by a marked deficiency or complete absence of pancreatic secretions. The presenting clinical symptoms are a failure to gain weight on an adequate diet, under-nourishment, distended abdomen, steatorrhea with large foul stools, and the development of chronic bronchopulmonary suppuration. Cystic changes in the bronchial epithelium and the easy entry of infecting organisms into the respiratory tract results in extensive suppurative bronchitis, atelectasis, bronchiectasis and bronchopneumonia. The diagnosis is most commonly established by the demonstration of an absence of pancreatic enzymes in the duodenal secretions or in the stools, or by an absence of absorption of vitamin A into the blood stream. Sweat tests demonstrating a high secretion of electrolytes are further confirmatory indications of the disease. From the standpoint of the bronchologist the most important aspect of therapy is the management of the respiratory tract infection that is so prominent a part of the pathology of fibrocystic disease of the pancreas. The mechanical removal of secretions from the airway is assisted by liquefaction of the secretions through

the use of aerosols and placing the infant in a high humidity atmosphere of a steam room. Postural drainage and frequent catheter aspirations of the pharynx and nasopharynx 'are' of importance. Occasional exposure of the larynx with a laryngoscope and aspiration of the trachea with a catheter or small tracheal aspirator is of great value. Repeated bronchoscopic aspiration of the main bronchi and of the lobar bronchi assists in reducing the frequent episodes of broncho-pneumonia. Furthermore, it clears the airway of viscid material and allows freer entry of the aerosolized medications to the periphery of the bronchial tree.

ESOPHAGOSCOPY IN THE NEWBORN

Many of the esophagoscopic problems of the newborn infants have been recognized and accurately described in the past. However, until the external surgical approach to the esophagus was safe and feasible, relatively few of these problems had clinical significance. Paradoxically, with the advent of such surgery, many new problems have arisen, particularly in the management of a post-surgical stricture of both the airway and the esophagus.

ESOPHAGEAL ANOMALIES

Congenital anomalies of the esophagus are divided into three categories: 1. Those involving the esophagus alone. 2. Those in which the trachea is associated in the anomaly, and 3. those in which anomalies of other structures influence the anatomical configuration and thus the function of the esophagus. Anomalies of the esophagus alone include absence, duplication, atresia, webs, stenosis, short esophagus and achalasia. The strictures may be fibrous or merely a failure of epithelialization of a portion of the lumen. The achalasia, too, may be simple, but occasionally it is associated with extreme dilatation and fibrous stenosis in the cardia.

The combined esophago-tracheal anomalies consist of tracheoesophageal fistulae with or without esophageal atresia, esophageal atresia with a tracheoesophageal fistula is relatively common, whereas a fistula without esophageal atresia is relatively rare.

Esophageal anomalies associated with anomalies of other structures are of unusual interest. Commonest are the cardiovascular anomalies that cause esophageal obstruction through encirclement of the trachea and the esophagus. Another anomaly of this category is a lung bud having its origin in the esophagus.

CONGENITAL ESOPHAGEAL STENOSES

There are innumerable endoscopic problems associated with the congenital anomalies of the esophagus. Congenital stenosis of the esophagus not associated with a congenitally short esophagus may be a single web or multiple strictures involving segments of the esophagus varying in length from 1 to 10 cms. Congenital webs are not common, and respond immediately and usually permanently to a single dilatation or to a short series of dilatations. This procedure may consist of dilatation with an esophagoscope guided through

the web over a string or an esophageal bougie. Graduated mercury-filled rubber bougies may be used being rapidly increased in size because of the ease with which the web responds to dilatation. Strictures of whole segments of the esophagus are more common, and from an endoscopic standpoint may be subdivided into two further groups because of their different endoscopic appearance and their variation in response to therapy. In each, the distal esophagus is most frequently involved. Esophagoscopically, the walls of one type appear extremely soft, covered with granulation tissue, and apparently devoid of mucosa. These respond rapidly to dilatation by esophagoscopy or mercury-filled rubber bougies. However, the soft walls increase the danger of perforation during the dilatation and necessitate unusual caution. The second type of congenital segmental esophageal stenosis is a fibrous stricture, often extremely firm, and response to dilatation is slower; the stricture is more resistant to dilatation, and the lumen is not maintained for as long a post-dilatation period. Many of these are discovered when a foreign body causes complete obstruction. Roentgenograms of the esophagus taken after a foreign body has been removed often establishes the diagnosis. Analysis of the history of such infants or children may show that they had been considered "feeding problems." The stenosis associated with the short esophagus and cardiospasm are generally recognized at four to five months of age when the infant first is given semi-solid and solid foods. Here again, endoscopic dilatation should be given a most thorough trial before resorting to external surgical procedures because of the frequency of post-surgical stenosis.

ESOPHAGEAL ATRESIA

Combined esophagotracheal anomalies present the largest group of esophageal anomalies requiring endoscopic assistance. During the past fifteen years, 150 cases of esophageal atresia with tracheoesophageal fistulae have been operated upon at The Children's Memorial Hospital, with an overall survival of more than 50%. In recent years, the mortality has been reduced to approximately 30%. Improvement in surgical technic as well as the management of the immediate postoperative complications accounts for the steadily rising survival rate. Frequently, pre and post-operative bronchoscopic aspirations are performed. From the surgical standpoint, the most important improvement has been the decrease in operative time through the use of a transpleural rather than an extrapleural approach. Almost constant, weak pharyngeal suction is employed post-operatively. The principal cause of death in the most recent cases have been other anomalies incompatible with life, or prematurity with the infant weighing less than four pounds. The surgical procedure employed has been the direct anastomosis with the most meticulous mucosal suturing with 6-0 silk. This is used in an effort to avoid stricture formation at the line of anastomosis. Slow feedings have been started only after the fifth post-operative day. Approximately half of the infants who have survived have required some type of post-operative peroral or retrograde dilatation. This percentage has steadily decreased during the past five years. Nevertheless, the most important endoscopic problem in the post-operative management of congenital esophageal atresia is that of the stricture at the point

of anastomosis of the two segments. From a surgical standpoint this is often difficult or impossible to avoid since the upper portion of the lower segment may be extremely stenotic, occasionally its lumen is hardly more than a millimeter in diameter. Rarely do the two segments approximate each other sufficiently to provide mucosa for an adequately long suture line. Therefore, if the development of a stricture is anticipated due either to a narrow lower segment, a short suture line or infection at the site of anastomosis, it is imperative that a gastrostomy be done and a string be passed through the esophagus to be anchored in the stomach or intestine, eventually being brought out through the gastrostomy. Such a string serves as a guide through the irregular, fibrous anastomosis. Gastrostomy, which in early cases in the series was done routinely, but is now rarely necessary, greatly facilitates dilatations of the stricture since it is often impossible to find the lumen from above.

During the past fifteen years, we have had 67 infants who have required treatment for varying degrees of stricture following repair of esophageal atresia. Thirty of these had their original surgical corrective procedure elsewhere and were referred for dilatation of the esophageal stricture postoperatively. Most of these cases responded rapidly to one or two esophagosopic dilatation. In others, Plummer bougies or retrograde dilatations have been employed with frequent dilatations necessary to maintain the lumen. In most cases, however, graduated mercury-filled rubber bougies have been used perorally with reasonable good results. Five infants, all originally operated elsewhere, have required reoperation because of failure of dilatation methods.

Four cases presented an additional problem with important endoscopic features. These had been referred for dilatation of the esophageal stricture, but in addition were found to have a severe tracheal stenosis at the point from which the lower esophageal segment had been resected from the trachea and the opening into the trachea repaired. Respirations were loud, labored, and rasping in character and each infant had a brassy cough that was almost constant and uncontrollable. Examination of the trachea with the infant bronchoscope demonstrate the area of stenosis to be immediately above the bifurcation with granulation tissue on the posterior wall of the trachea. This area was gently dilated before dilatation of the esophagus could be begun. While the lumen of the airway was improved by dilatation, frequent upper respiratory infections caused alarming respiratory obstruction. Occasional tracheal dilatations were done during the intervals between upper respiratory infections. Symptoms have disappeared as these infants have grown. The first is now twelve years of age; she has no symptoms of either tracheal or esophageal obstruction.

From a theoretical standpoint, the infant with a complete esophageal atresia without a tracheoesophageal fistula should present a favorable surgical prognosis because of the absence of regurgitated gastric content in the tracheobronchial tree. The chemical pneumonia which complicates the aspiration in the infant with a fistula adds an additional hazard which is significant. However, this is more than offset by the fact that the two ends of the esophagus are so widely separated in complete atresia without a fistula, that direct

anastomosis without immobilization of the stomach is impossible. The surgeon must resort to alternatives such as gastrostomy and external pharyngotomy as the primary procedures, with reconstruction of the pharyngogastric continuity at a later date by intestinal or colon transplants.

Absence of all or a large part of the esophagus presents additional postoperative endoscopic problems. Early management of seven such cases has consisted of a gastrostomy and an external esophagostomy, direct anastomosis at two years of age at first consisted of raising the stomach to the upper segment, anterior to the sternum. Stenoses at the gastro-esophageal anastomosis necessitated attempts at dilatation over a thread. These were unsatisfactory and the use of a colon transplant in the later cases has been more successful. Again, the most careful mucosal suturing with 6-0 silk sutures has reduced or eliminated leakage, infection and stricture formation.

VASCULAR RINGS

Severe respiratory and esophageal obstruction may be produced by anomalies of the aort and its branches. During the past twelve years, 37 infants or children have been seen with vascular anomalies causing enough compression of the trachea, a bronchus or the esophagus to require surgical treatment. The symptoms were combined respiratory and esophageal obstruction. Respiratory symptoms varies from simple stridor to severe obstruction with acute cyanosis; these symptoms increased during feedings. Esophageal symptoms were dysphagia, in some from birth, and in most, beginning in the fourth and fifth months, when semi-solid foods were added to the infant's diet. Other symptoms were a brassy cough and repeated attacks of "croup". Most infants in this series were referred for bronchoscopic investigation of their stridor.

Vascular rings may be complete or incomplete. When both right and left aortic arches persist from embryonic life, a complete ring is formed about the esophagus and trachea. The anterior segment or left arch is usually small but occasionally the posterior segment may be the smaller vessel. If the aorta descends on the right, the left portion of ring is pulled even more tightly against the trachea and esophagus.

Incomplete vascular rings occur when a right aortic arch with a ligamentum arteriosum passes to the left to join the pulmonary artery. Though there is an anterior gap in this "ring", the pulmonary artery, ligamentum and right aortic arch cause enough esophageal and tracheal compression to produce symptoms. Another type of incomplete ring is formed when an anomalous right subclavian artery originates from the descending aorta and passed behind the esophagus. A third type of incomplete ring is due to a combination of a right aortic arch and an anomalous left innominate artery or left carotid artery passing from the right arch across the anterior surface of the trachea.

Surgical relief is obtained by dividing the encircling ring at a point that will relieve the pressure on the trachea and esophagus. Often several points of division are necessary, such as dividing the smaller of two arches, the ligamentum arteriosum or subclavian artery before optimal release is obtained.

There have been nine deaths among the 37 patients operated. Most of these were young infants with severe low tracheal obstruction in whom anesthesia and management of tracheotomies necessitated by the low constriction, were extremely difficult. Relief of symptoms did not always immediately follow surgical severance of the constricting vessel. This was due to developmental narrowing of the airway at the site of constriction. Final improvement in symptoms was delayed until the tracheal cartilages could grow to their normal configuration.

SUMMARY

The indications for bronchoscopy and esophagoscopy in infants are constantly increasing. Problems associated with the examinations of the small air and food passages of patients under one year of age have become more complex, largely as the result of the spectacular advances of pediatric surgery of the heart, lungs and esophagus. Such surgery requires the most accurate diagnostic endoscopic pre-and postoperative endoscopic procedures that are of unusual interest. Using miniature lamps and light carriers, practical bronchoscopes and esophagoscopes for use in newborn infants are available. In addition, aspirators, forceps and even telescopes (optics) have been developed for special studies. No anesthesia is required or desirable for bronchoscopy, bronchography or esophagoscopy in infants; but team work to permit a rapid, thorough procedure is essential.

In the immediate post-partum period, bronchoscopic problems are essentially those of differential diagnosis and relief of bronchial obstruction. The most obvious condition is that of retained mucoid secretions; however, when bronchoscopic aspiration of the newborn is requested, there is usually an additional, serious underlying condition responsible for the retained secretions. Central nervous system changes such as cerebral agenesis or a birth injury resulting in loss of laryngeal and cough reflexes may account for the symptoms. However, congenital anomalies such as esophageal stenosis or atresia, with or without a fistula, pulmonary agenesis, lobar emphysema, anomalous vessels obstructing the trachea or bronchi, constitute the types of congenital anomalies that require endoscopic investigation. Corrective surgery for many of these conditions necessitates further endoscopy to assist in maintaining the patency of the air and food passages. Endoscopic problems in 150 cases of congenital atresia of the esophagus with a tracheoesophageal fistula have consisted of management of the stenoses at the site of anastomosis of the esophagus, the occasional severe tracheal stenoses occurring at the site from which the lower esophageal segment had been removed, and pulmonary atelectasis, particularly of the right upper lobe.

Severe respiratory and esophageal obstruction produced by anomalies of the aorta and its branches has been observed in 37 infants during the past 12 years. Diagnostic procedures have included bronchoscopy, esophagoscopy and, in some cases, bronchography. Intrabronchial support by means of polyethylene tubing following removal of the constricting vessel has been necessary. Persistence of bronchial narrowing is noted after surgery

in most cases, although there is improvement in symptoms because of relief of pressure. Reduction in lumen sized is due to the pressure of the vessel on the bronchus during development.

Bronchopulmonary suppuration in infants under one year of age presents a wide field for diagnostic and therapeutic bronchoscopy. Non-specific respiratory tract suppuration, the pulmonary aspects of pancreatic fibrocystic disease and specific infections such as tuberculosis have endobronchial and extra-bronchial aspects of particular bronchoscopic import. The correlation of general pediatric management with bronchial hygiene through endobronchial removal of obstructive secretions and exudates constitutes an important phase of the recognition and management bronchoscopic and esophagoscopy problems in infants.

LEGENDS

- FIG. 1 Infant tracheotomy tube with shallow curve and short length (33 mm.). The short length avoids contact of the end of the tube against the carina. The curve follows the tracheal path more accurately than does a shorter radius tube.
- FIG. 2 Equipment for obtaining a bronchogram of an infant. Polyethylene tubing is attached to rubber connecting tube and syringe by a large-bore needle. The infant laryngoscope, Samson infant tracheal aspirator and laryngeal forceps used to guide the polyethylene tubing into the larynx are shown.
- FIG. 3 Idiopathic emphysema of the left upper lobe of an infant two weeks of age. Left upper lobectomy performed following bronchoscopic evidence of absence of bronchial obstruction.
- FIG. 4 A. Apparent atelectasis of the left lung in a newborn infant.
B. Bronchogram following a bronchoscopy that had demonstrated a rudimentary left bronchus only 1 cm. in length.