SURGERY OF THE ESOPHAGUS

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Surgery of the Esophagus

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Preface

This revision of *Surgery of the Esophagus,* more than 15 years after the first publication, has been prompted by a number of factors; among them are improved treatment of some esophageal diseases and a discouraging lack of progress in the treatment of others. While the physiology of the esophagus is much better understood, the etiology and pathogenesis of many lesions remain obscure. Certain technical aspects of esophageal surgery are decidedly better, so a revised summary of esophageal surgery may be of value.

Twenty years ago, the *Index Medicus* contained one or two pages of references concerning the esophagus; today the list approaches 30 pages. Half or more of these are in a language other than English. The task of obtaining and translating these references would require years. For this reason, only a limited number of foreign language references are quoted.

My debts in the preparation of this book are many. First, it is a pleasure to acknowledge the superb care the residents have given the patients under discussion, as well as the stimulus derived from their questions. Second, I appreciate the cooperation of my associates in the Department of Surgery, particularly the Chairman, Dr. David C. Sabiston, Jr., who obviously provided the opportunity for my work as well as an impressive example of precise medical authorship.

Numerous radiographs were reviewed and selected by Dr. William F. Barry, Jr., Dr. Irvin Johnsrude, Dr. Reed P. Rice, Dr. Frederick M. Kelvin, Dr. M. W. Stannard, and Dr. Herman Grossman. Helpful and authoritative corrections were provided by Dr. Howard C. Filston in his review of the chapters on congenital anomalies. Dr. Jerry S. Trier kindly reviewed the section on the columnar-lined lower esophagus.

Aid was provided in the onerous task of abstracting patient records by Dr. Wayne Flye, Dr. Steves Ring, and Dr. Walter Wolfe. Dr. James Lowe helped with translations and Dr. Lawrence H. Muhlbaier with statistical analyses. A major contribution of the manometric records was made by Dr. Andre Duranceau.

Information from the Duke Cancer Registry was promptly supplied by Lou Woods. Our Librarian, Michael Blanton, helped assemble the bibliography.

The first drawings were made by William B. McNett, Orville A. Parkes, and James M. Goodman. Recent drawings and graphs were made by Donald G. Powell with the aid of Linda Kohl-Orton. Photographs were made by Paul C. Greenwood, David G. Hong, and Ronald G. Kovacs. I am deeply indebted to all of them, and especially to the Chief of the Division, Ronald L. Mitchell, for his personal attention.

The stimulus for this task originally came from Dr. William G. Anlyan, who has continued his encouragement throughout the years. My collaborator, Dr. Will C. Sealy, provided the foundation, based on his extensive clinical experience, his objective evaluation and judgments, and his insistence on clarity and reason.

The reprints, references, and revisions have been kept in strict order by Betty Howell, with provision of additional help and advice. The many reference abstracts and the repeated revisions of the manuscript have been typed with speed and accuracy by Penny Ennis, to who I am indeed grateful.

Finally, I thank my wife for her patience, tolerance, and understanding.

R.W.P.

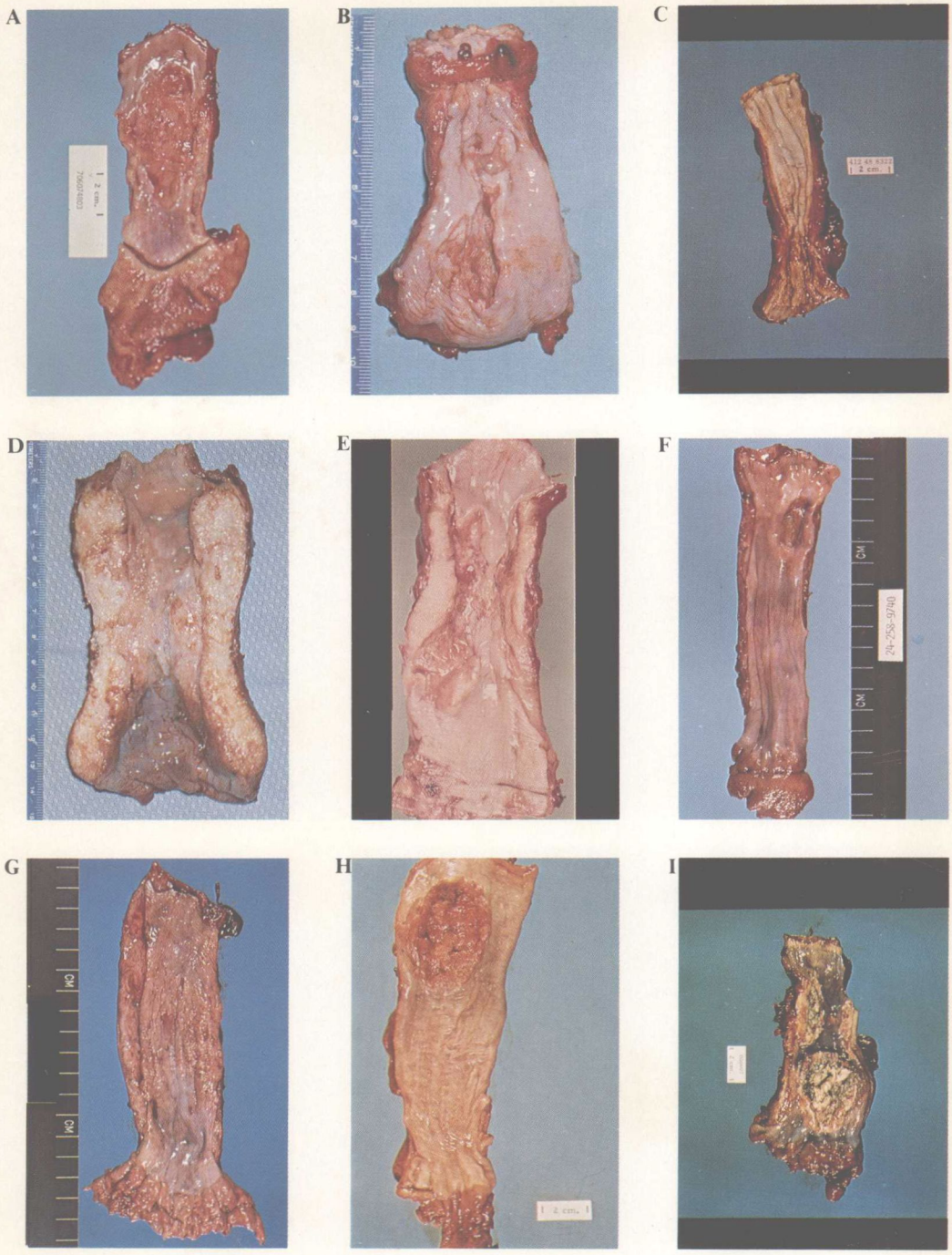

PLATE I

Examples of carcinoma of the esophagus. **A, B.** Typical ulcerative lesions. **C.** Constricting lesion. **D,E.** Marked thickening of wall and constriction. **F.** Response of carcinoma after preoperative irradiation. **G.** Diffuse, superficial carcinoma. **H.** Raised rolled edge. **I.** Large necrotic polypoid tumor.

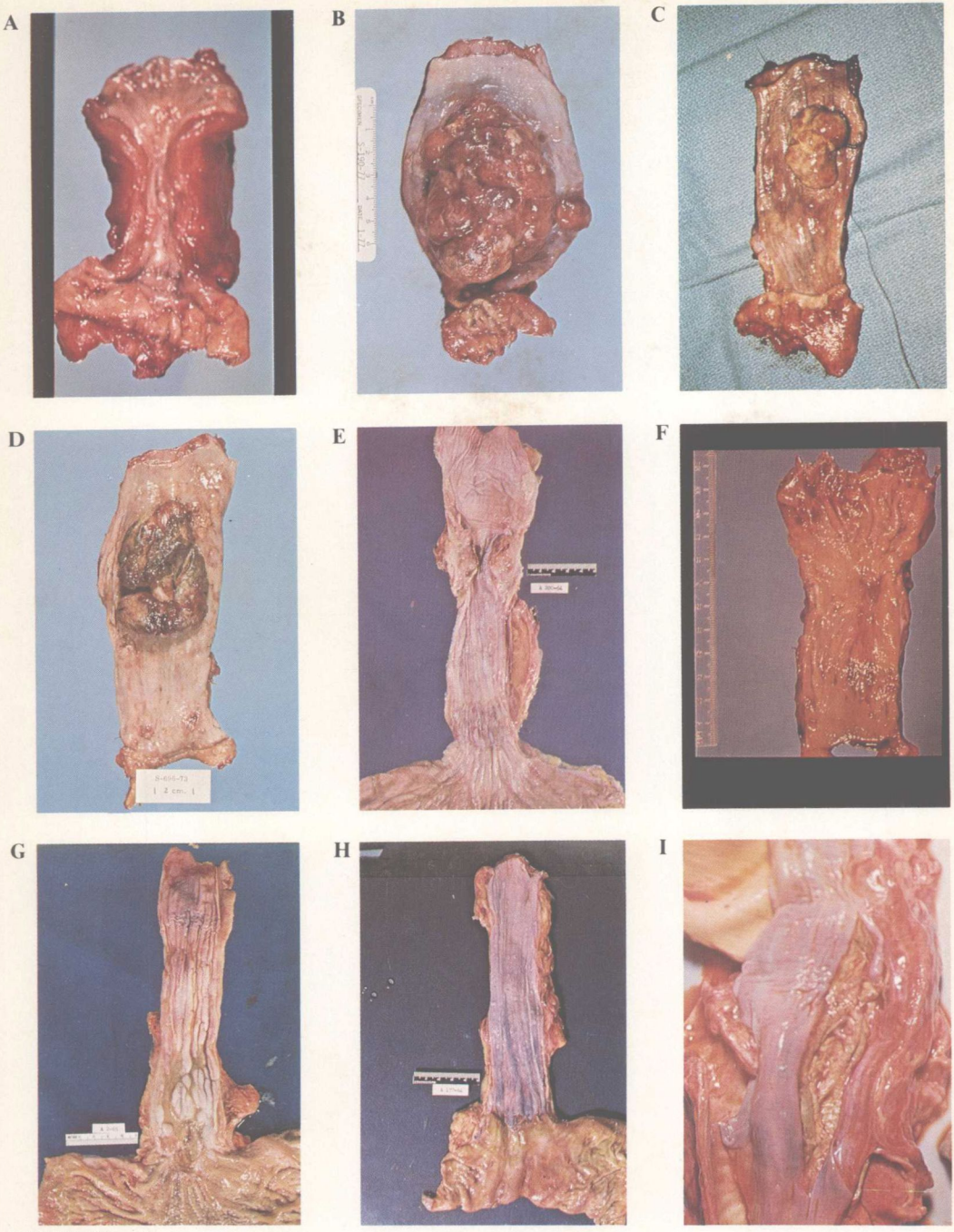

PLATE II

A. Mucoepidermoid carcinoma. **B.** Leiomyosarcoma. **C.** Carcinosarcoma. **D.** Pseudosarcoma. **E.** Bronchogenic carcinoma metastatic to nodes compressing esophagus. **F.** Barrett's ulcer in columnar-lined lower esophagus. **G.** Autopsy specimen of hiatal hernia, marked esophagitis, and penetrating ulcer at junction. **H.** Esophageal varices. **I.** Rupture of esophagus due to forcible insertion of Sengstaken-Blakemore tube.

Contents

1. Congenital Atresia and Tracheoesophageal Fistula

Congenital anomalies of the esophagus are fortunately not common. The most frequently encountered is congenital atresia, usually with a fistula from the distal segment into the trachea. Effective treatment dates from the successful one-stage anastomosis performed by Haight. Since then, hundreds of infants who would have died have recovered with nearly normal alimentary tract function.

Classification. The atresia and various types of fistula have been identified by numbers, roman numerals, and letters. Holder and Ashcraft (1966) in the most definitive review published, show in a table a list of 10 different such classifications proposed by various authors. The chances for confusion are obvious, particularly when surgeons reporting their experiences do not designate which classification is used. Perhaps it would be clearer simply to describe the anomaly. Five major categories describe adequately most patients: (1) atresia without fistula, (2) atresia with fistula from proximal segment, (3) atresia with fistula from distal segment, (4) atresia with fistula from both segments, and (5) fistula without atresia (Fig. 1-1).

History. Durston, in 1670, apparently was the first to record a malformation of the esophagus: a simple atresia of the lower third without fistula in the right component of conjoined twins. Gibson, in 1697, reported the first patient with esophageal atresia with tracheoesophageal fistula of the typical form. Mackenzie (1884) found reported 40 cases of atresia with tracheoesophageal fistula. Lozach, in 1816, described the first case of complete absence of the gullet. Tenon in 1791 first recorded a case of complete membranous obstruction of the upper part of the esophagus. Blasius in 1674 described a double esophagus. Vogt (1929) described the radiologic findings and suggested the classification generally quoted.

Steele (1888) reported one of the earliest operations for esophageal atresia. Holmes, in 1869, first suggested that anastomosis of the two segments of an atretic esophagus might be possible. Keith (1910) and Richter (1913) also noted that operative measures to unite the two segments were theoretically possible. One of the

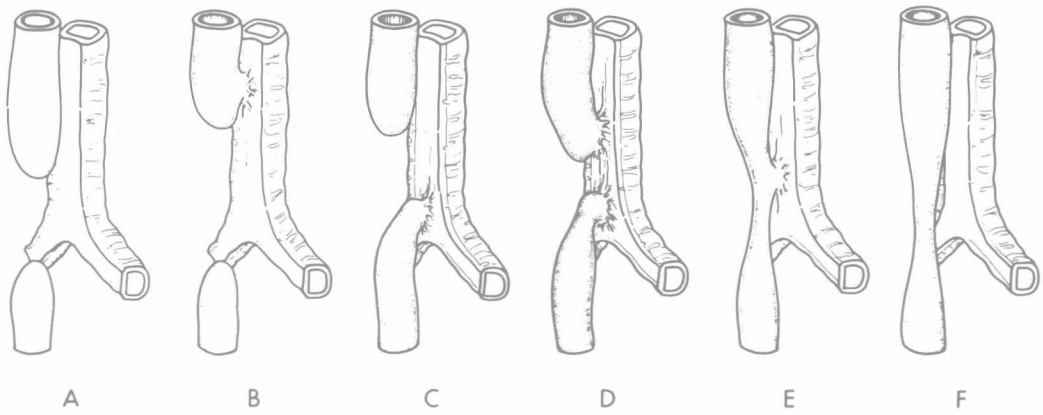

FIGURE 1-1

Major types of anomaly. **A**. Atresia without fistula. **B**. Atresia with fistula into proximal segment. **C**. Atresia with fistula into distal segment. **D**. Atresia with fistula into both segments. **E**. Fistula without atresia. **F**. Congenital stricture.

earliest attempted anastomoses was that by Wing. In the discussion of a paper by Vinson (1924) LeWald stated that recently, Dr. L. A. Wing had operated on a baby who had congenital atresia of the esophagus with tracheoesophageal fistula. He succeeded without much difficulty in dividing the fistula and uniting the two ends of the esophagus. The infant did not recover but survived the operation a sufficient length of time to justify the opinion that the procedure might possibly succeed in the future.

Usually gastrostomy or jejunostomy only was utilized in treating these patients, but those surgeons particularly interested devised numerous methods of multiple-stage operations other than by direct anastomosis. These procedures have been reviewed by Gage and Ochsner (1936). It therefore became apparent that technically operative treatment would be successful and eventually direct anastomosis could be performed. As a better understanding of infant physiology developed, particularly the administration of blood and intravenous fluids, the chance of success rapidly increased. In addition, improvement of anesthesia in the infant was a parallel advance which further enhanced the possibility of recovery of the infant. Finally, the first survivors were operated upon only a day apart in November 1939; the first by Leven in Minneapolis and the second by Ladd in Boston. Haight, on March 15, 1941, for the first time successfully performed ligature of the fistula and end to end anastomosis.

Etiology. Kreuter (1905) suggested that epithelial occlusion at the 19- to 20-mm stage of embryonic development, followed by lack of recanalization, might lead to esophageal atresia. Subsequent work, however, failed to confirm this. Pressure was also suggested as a possible cause: from the heart by Schmitz (1923), and from abnormal vessels by Fluss and Poppen (1951) and Langman (1951). The importance of mechanical pressure is doubtful as the extremely gelatinous nature of the embryo would appear to permit considerable displacement of the tube.

Yamasaki (1933) noted the rarity of tracheal atresia as compared with esophageal atresia and called attention to the difference between the thin dorsal layer of cells and the thicker epithelium on the ventral side of the primitive foregut. He postulated that the rapid growth of the trachea and pulmonary primordium used up so much of the "growth potential," as expressed in available cells, that the posterior digestive segment could not provide enough cellular material to complete the esophagus. Gruenwald

(1940) suggested a similar chain of events wherein a delay in separation accompanying a rapid elongation of the trachea might carry the developing digestive tube caudally so rapidly that it lost the ability to differentiate into a separate and normal esophagus over the distance that the trachea had grown. With a decrease in rate of growth of the trachea, the cranial segment of the digestive tract might begin to differentiate and even make up for growth difference and overlap the lower segment. The different degrees of the malformation, i.e., fistula at different levels of the trachea, develop according to the degree of separation occurring prior to the onset of rapid growth. Therefore, the opening of the lower esophageal segment into the bifurcation represents the highest degree of anomaly and the length of the upper esophageal segment is independent of the level of the fistula. Rosenthal (1931) felt that the cause rested in an altered or deficient growth of the entodermal cells that eventually provide the tracheoesophageal septum.

Smith (1957), in his extensive studies (see Chapter 17), pointed out that during the period from 21 to 32 days (4 to 12 somite to 7 to 8 mm), three rapid, complex, and major changes are occurring in the embryo: elongation and growth; separation of the trachea from the foregut; and branching and growth of the bronchial tree. During this time, abnormalities may be due to retardation of growth or excessive growth, or, more probably, both. Smith's investigations led him to suggest that the development of both esophageal atresia and tracheoesophageal fistula may involve hyperplasia of the lateral esophageal ridges. The atresia would appear to result from the too rapid elongation of the trachea, combined with diminution in esophageal substance so that the esophagus "cannot make ends meet," as suggested by Gruenwald. The thickened tissue of the wall of the lateral esophageal groove provides cellular material for the continuation of the esophagus, at the lower part of the trachea, with the development of a fistula (Fig. 1-2). Atresia without fistula may arise, as suggested by Yamasaki, from failure of the fistula to persist or to develop. If no suggestion of a fistulous tract exists, Smith assumed only the factor of too rapid elongation of the trachea without abnormality of the lateral esophageal grooves. Fistula without atresia and atresia with fistula to both segments may be due to localized or limited failure of development of the tracheoesophageal ridges, perhaps caused by imbalance between tracheoesophageal division and elongation (Fig. 1-3).

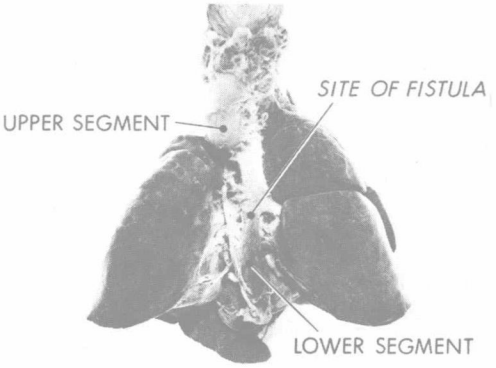

FIGURE 1-2
Most common type of atresia and fistula. This infant was admitted at 12 days of age and died the following morning. Posterior view shows the dilated upper pouch, gap between segments, site of fistula, and long lower segment of esophagus.

Incidence. The reported incidence of esophageal atresia varies from the two extremes of 1:800 births reported by Belsey and Donnison (1950) to 1:38,375 births noted by Raffensperger and his colleagues (1964). From all the reported series, a reasonable estimate would seem to be 1 in 3000 to 4000 live births.

The familial incidence of esophageal atresia was summarized by Sloan and Haight (1956) in their report of congenital esophageal atresia with tracheoesophageal fistula in brothers born 1 year apart. Sloan and Haight accepted five other instances of atresia in siblings. Hausmann et al (1957) later reported three consecutive siblings with atresia. Engel and his associates (1970) reported the same type of atresia with fistula into the distal segment in a mother and daughter. Dennis et al (1973) described two sisters with atresia; the son of one sister had atresia with fistula. On the basis of their survey, they

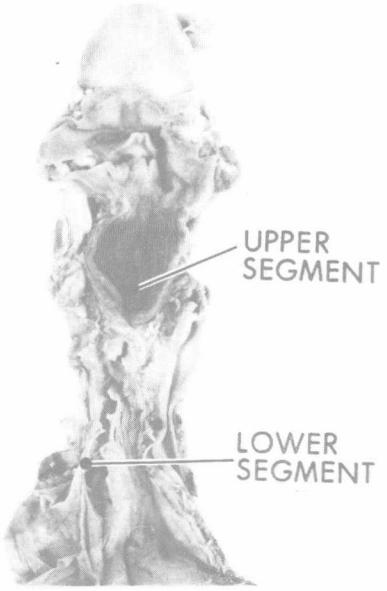

FIGURE 1-3
This infant was admitted at 1 week of age and expired after gastrostomy. Note thickened upper segment and short, narrow lower segment.

conclude that the genetic contribution is relatively nonspecific.

Esophageal atresia has been noted in one of a set of twins by many observers, although it is not clear in some of the series whether or not the twins were identical. Ingalls and Prindle (1949) found that 4 of their 107 patients were one of a set of twins. Brown and Brown (1950) found this in 2 of 24. Leven et al (1952) found this in 2 of 102. In Haight's experience (1957) with 201 patients, 10 were found to be 1 of a set of twins; 6 of these sets of twins were considered to be identical. Three reports (Wooley et al, 1961; Blank et al, 1967; Bolam et al, 1973) record that both twins in their sets of twins had both atresia and fistula.

Haight states that although the answer is not clear, their observations suggest the importance of nongenetic factors in the etiology of the anomaly.

In our group of patients at Duke, one patient was one of twins. Another patient had a sibling with the anomaly.

The incidence of each type of anomaly is shown in Figure 1-4. These collected series include the 1068 we had found reported to 1958, the 1058 patients from Holder's survey, and 2292 reported since then. Duplication of cases has undoubtedly occurred but the percentages are reasonable approximations. The atresia with tracheoesophageal fistula from the distal segment is by far the most frequent type, comprising almost 90 percent. An appreciable number are seen with esophageal atresia alone, followed in frequency by those with fistula and no atresia, then fistula from both distal and proximal esophageal segments, and lastly fistula from the proximal segment. The distribution of these anomalies according to sex in three collected series of 292 cases was 60 percent in males and 40 percent in females.

Anatomy and Pathology. In the common type of atresia with fistula into the distal segment, the proximal pouch varies in length from 1 to 3.5 cm.

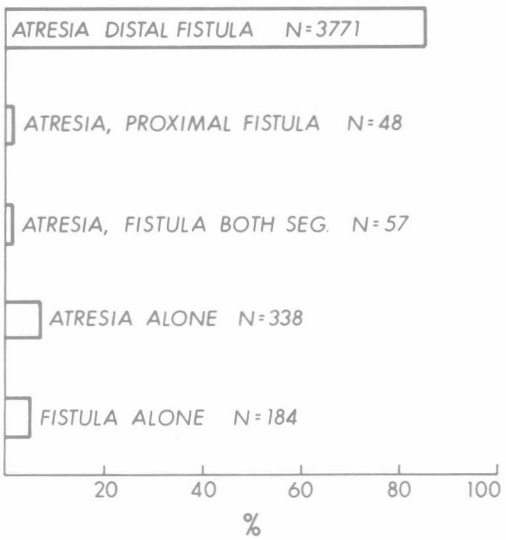

FIGURE 1-4
Incidence of the various types of atresia and tracheoesophageal fistula in 4418 patients reported.

This would result in a distance from the premaxilla of 10 to 13.5 cm. Attachment of the proximal pouch to the trachea and to the distal segment varies and is probably a factor in the length of the pouch. The diameter is increased from the normal of about 5 to 10 mm or more. The wall is usually thickened distally and the blood supply is good.

The attachment of the lower segment is usually in the membranous part of the trachea within 1 cm of the carina but may be above this or into the proximal portion of either main bronchus. Esophageal musculature and mucosa extend to the opening. In fact, Emery and Haddadin (1971), in 50 autopsies, found squamous metaplasia into the trachea in 40; in some this was extensive to the point of being circumferential. They emphasize the clinical importance of this deficiency in respiratory epithelium as the inability to move mucus would lead to increased respiratory complications.

The wall of the distal segment is thin; the diameter is decreased proximally but increases to normal toward the stomach. The blood supply is tenuous. Lister (1964), in his injection studies, found that the branches from the aorta may or may not be present, although those from the left gastric artery are seen. Lister suggests an anastomosis under tension might be safer than mobilization of the lower segment, which might interfere with the blood supply.

Major pathologic changes may be present in the lungs. In addition to aspiration of upper respiratory secretions and mucus from the proximal pouch, gastric secretions may enter the respiratory tract through the fistula and cause considerable pulmonary damage. Air from the trachea passes into the stomach, which becomes distended. In the absence of distal obstruction, some of the air passes into the bowel but some is also regurgitated, carrying with it gastric secretions. As the stomach becomes more distended with air, the chances of regurgitation and aspiration of gastric juice through the fistula increase. Hess (1913) found normal newborn infants to secrete a considerable quantity of hydrochloric acid. A representative study of the effects of acid aspiration is that of Greenfield and his associates (1969). They instilled 0.1 *N* hydrochloric acid into the trachea of dogs. Severe alveolar and capillary damage resulted. Arterial Po_2 fell, Pco_2 increased, and acidosis developed. In addition to this chemical damage, the distension in the abdomen may elevate the diaphragm sufficiently to interfere mechanically with respiratory function; this may be the major deleterious factor in some infants.

When the fistula is from the proximal segment only, in the presence of atresia, the opening is usually not from the distal end of the pouch at about the midpoint of the trachea. The distance to the distal segment varies but may be as much as 5 cm. Atresia with a fistula into the trachea from both proximal and distal pouches differs in that the proximal pouch may not be dilated and hypertrophied. If the proximal fistula is from the end of the upper pouch, it should be identified, but a number of infants have had the proximal fistula from the side of the esophagus. As a result, a successful anastomosis may be done and the proximal fistula missed at the original operation.

Atresia without fistula frequently presents a difficult situation because of the distance between segments, which may be short but more often is 4 to 5 cm. Tracheoesophageal fistula without atresia may occur at any level of the trachea but is usually above the midpoint. Variation in the size of the opening, as well as the fact that the fistula may extend caudad from trachea to esophagus, may delay the onset of symptoms for months or years.

DIAGNOSIS

Two situations should alert the clinician to the possibility of esophageal atresia: hydramnios and premature birth. Amniotic fluid swallowed by the fetus is normally absorbed in the gastrointestinal tract and transferred to the maternal circulation through the placenta. An obstructing lesion in the fetus, such as esophageal atresia without fistula, leads to excessive accumulation of amniotic fluid as normal absorption does not occur. In the review by Fransen and Lacquet (1974b) hydramnios was found reported in 57 to 85 percent of the mothers of infants with atresia but without fistula. The range was 17 to 32 percent when a fistula was present. Holder and Ashcraft estimate that the chances are about

1 in 14 that hydramnios is due to esophageal atresia.

Prematurity may be related to hydramnios. Holder and Ashcraft note that about 8 percent of all births in this country are premature but 34 percent of infants with atresia and distal fistula are premature. The incidence is as high as 75 percent in those without fistula, according to Fransen and Lacquet. The frequency of prematurity is less important in diagnosis than the relationship of birth weight to survival.

Symptoms and Signs. The first symptom noted immediately or soon after birth is excessive mucus which requires repeated and frequent aspiration. The amount of mucus may vary considerably in the first day or two of life; in some infants it may not be enough to arouse suspicion. In others, mucus may require almost constant removal to prevent respiratory distress. Characteristically, the first feeding results in a series of events which leads to diagnosis. The infant nurses hungrily, but this initiates an alarming episode of choking, coughing, regurgitation, respiratory distress, and even cyanosis. Each attempt to feed the infant produces the same manifestations.

In the usual type of atresia and fistula into the distal segment, air enters the stomach, as noted above, and abdominal distension is then a prominent sign. Infrequently, the fistula may be plugged by mucus or may be small and covered by a mucosal flap, so that air does not get into the distal segment. The absence of air in the stomach, however, usually indicates esophageal atresia without a fistula or, less frequently, atresia with fistula only into the proximal pouch. Abdominal distension may be a component of the manifestations of tracheoesophageal fistula without atresia (see below).

The pulmonary changes depend on the severity of the factors noted above, i.e., aspiration of mucus, aspiration of gastric secretions, and limitation of diaphragmatic movement. The right upper lobe is usually involved first and to the most severe degree. Eventually pneumonitis extends throughout both lungs. Respirations become rapid and noisy. Cyanosis develops. If allowed to progress, infection supervenes. Today, most morbidity and almost all mortality results from this complication.

The symptoms and signs of atresia alone and atresia with fistula in the proximal pouch do not differ appreciably and are suspected when air is

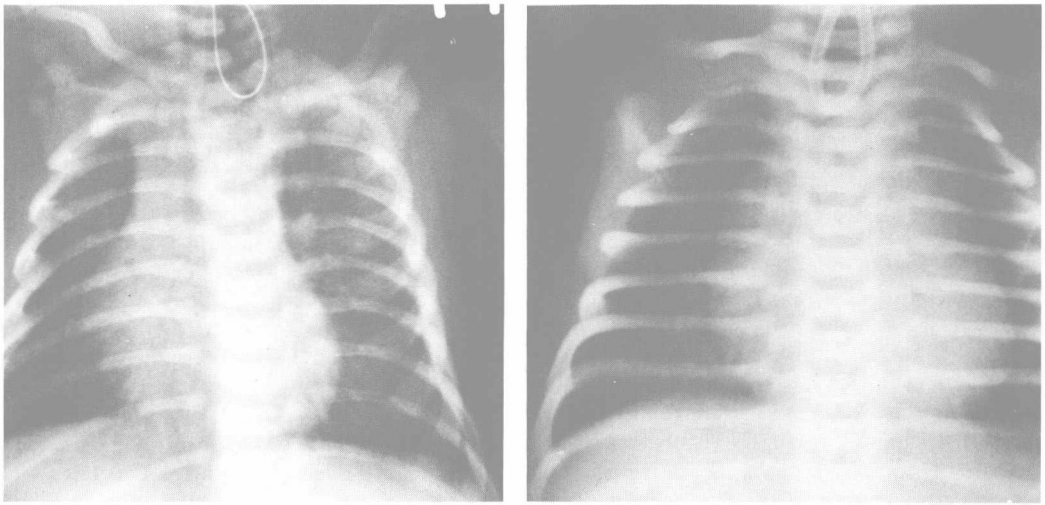

FIGURE 1-5
Catheter demonstrates the extent of the proximal segment in two patients with atresia.

absent from the stomach. Similarly, no major differences are apparent in atresia with distal fistula or with both proximal and distal fistula.

Radiologic Studies. When esophageal atresia is suspected, a sterile, stiff, radiopaque catheter, size 10 or 12 Fr, is well lubricated and passed through either nostril (Fig. 1-5). Obstruction will be met 9 to 13 cm from the nares and this essentially makes the diagnosis. Radiologic studies, however, are required for several reasons: (1) to confirm the location of the catheter and the extent of the proximal pouch, (2) to determine the severity of pulmonary involvement, (3) to examine for air in the stomach and bowel, and (4) to exclude some of the other concomitant anomalies.

Films of the chest will frequently show air in the proximal pouch, usually best seen on the lateral view. Selander (1941) noted the pouch may contain a fluid level and the trachea may be displaced anteriorly. A strip of air in the lower segment may occasionally be seen on the lateral film. The position of the catheter should be determined, particularly in the patient who is suspected of having atresia but in whom the catheter passes more than 12 or 13 cm. The usual explanation for this is that the catheter has coiled in the pouch but confusion and diagnostic delay have occurred when the catheter entered the trachea and passed through a fistula and then into the stomach.

The use of contrast material is probably ill-advised, although opinions do vary (Fig. 1-6). Holder and Ashcraft describe in detail the technique of instilling contrast. Koop (1971), who very strongly opposes use of any radiopaque material, states a mucosal coating of aspirated oily substance will reduce the cross sectional area of the trachea by 55 percent. The water-soluble materials are even more dangerous because of their high osmotic pressure, which can destroy the cells of the ciliated respiratory epithelium. The only reason radiopaque material might be of value would be to demonstrate a fistula from the proximal pouch. This is so infrequent, however, that it scarcely justifies jeopardizing the majority of infants with atresia (Fig. 1-7). Should the surgeon feel strongly that the information is necessary, the radiologist should instill a minute amount of material and aspirate this as soon as the film is exposed. With

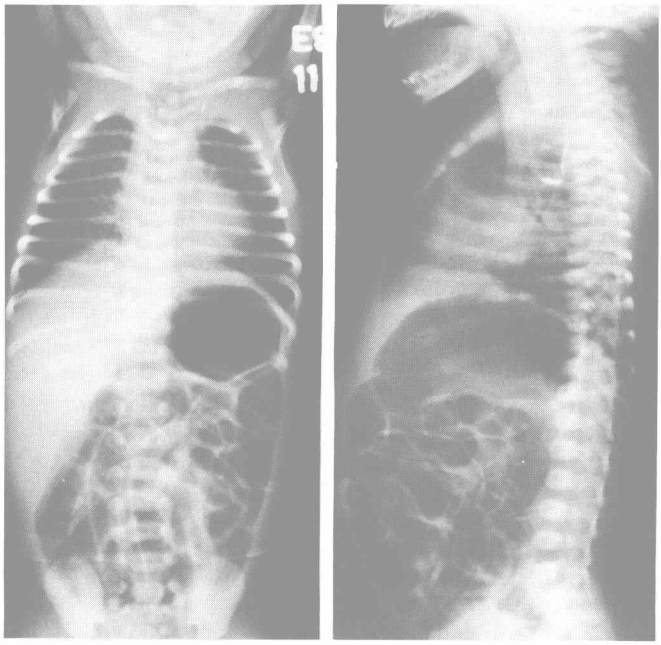

FIGURE 1-6
Small amount of radiopaque material was used to outline the upper esophageal segment, a step that is rarely necessary. Stomach and intestine distended with air, indicating the presence of a tracheoesophageal fistula.

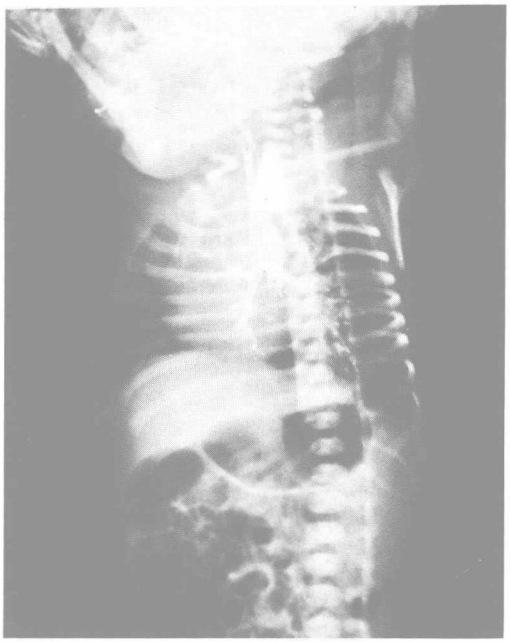

FIGURE 1-7
Danger of introducing contrast material shown by the inadvertent bronchogram.

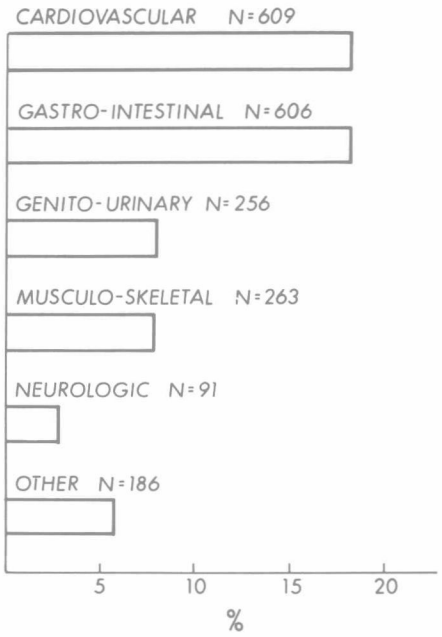

FIGURE 1-8
Incidence of associated anomalies in 3344 patients reported with esophageal atresia.

modern infant bronchoscopy utilizing optical telescopes, these fistulas can be clearly identified. Endoscopy is seldom indicated, however, in the usual patient with esophageal atresia and tracheoesophageal fistula (fistula without atresia will be noted later).

ASSOCIATED ANOMALIES

Malformations in addition to the esophageal atresia are common. In the 1058 patients surveyed by Holder and his colleagues, 505, or 48 percent, had 849 associated anomalies. From our collected cases through 1958, Holder's series and subsequent reports we show the frequency of these anomalies in 3349 patients (Fig. 1-8).

Cardiovascular and gastrointestinal anomalies are most common, and about one-third of cases with one will have both systems involved. Greenwood and Rosenthal (1976) found the 10 most common cardiovascular malformations, in decreasing order of frequency, reported to be: ventricular septal defect, patent ductus arteriosus, atrial septal defect, coarctation of the aorta, dextracardia, tetralogy of Fallot, truncus arteriosus, transposition of the great vessels, tricuspid atresia, and pulmonic stenosis. They determined that when esophageal atresia is present, the chances of a congenital heart anomaly were about 25 times greater than would otherwise be expected.

Imperforate anus is the most frequent of the gastrointestinal anomalies. Others include small bowel atresia or stenosis, hypertrophic pyloric stenosis, malrotation, duodenal bands, and biliary duct atresia. The most frequent anomaly of bone is hemivertebrae. Neurologic defects include spina bifida, hydrocephalus, and mongolism. Genitourinary malformations are horseshoe kidney, agenesis of the kidney and ureter, and various types of fistula formation. Atwell and Beard (1974) reported upper urinary tract anomalies in 19 of 40 neonates with esophageal