Esophageal Atresia and Tracheo-Esophageal Fistula

25 Years' Experience and Current Management

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■ *A review of the experience with esophageal atresia and tracheo-esophageal fistula over a 25-year period appears to lead to the advisability of the following procedures in surgical management:*

- *Emergency gastrostomy under local anesthesia in all patients.*
- *Extrapleural interruption of tracheo-esophageal fistula and end-to-end esophago-esophagostomy in patients who have the common type of upper esophageal atresia with distal tracheo-esophageal fistula.*
- *Upper esophageal stretching and eventual esophago-esophagostomy in patients with proximal and distal esophageal atresia with or without proximal tracheo-esophageal fistula.*

IT HAS BEEN almost 30 years since Ladd and Leven[1,2] treated the first surviving patients who were born with esophageal atresia. In the intervening years the anesthetic management of neonates has improved significantly, the general supportive measures for the surgically treated have become more specific and effective, and various surgical approaches have been tried and either discarded or continued depending upon their effectiveness.[3-6] For these reasons the overall survival of such patients now generally exceeds 50 percent and is more or less dependent upon the well known incidence of associated anomalies and the degree of prematurity. The term infant without significant congenital anomalies should survive with present management.

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During the past few years, our approach to the patient with esophageal atresia and tracheo-esophageal fistula has changed in several respects. The reasons for these changes and the results will be presented in this retrospective review.

Material of Study

During the past 25 years there have been 30 patients with esophageal atresia at the White Memorial Medical Center. The first three of these patients died before receiving any definitive treatment. The remaining 27 were treated surgically. Twenty-eight of the patients had proximal esophageal atresia with distal tracheo-esophageal fistula. One had proximal and distal esophageal atresia without fistula. One had proximal esophageal atresia with fistula and distal esophageal atresia without fistula. The last mentioned patient will be discussed in detail later in this communication.

From 1953 to 1957 there were nine patients, with one survivor. The subsequent decade, 1958 to 1968, is divided into two five-year periods. During the first five-year period (1958 to 1963) there were 12 patients, three surviving. From 1964 to 1968 there were nine patients, with eight surviving (Table 1).

Early in the history of tracheo-esophageal fistula and esophageal atresia the high incidence of associated congenital anomalies was recognized. Fourteen infants in this series had one or more additional anomalies (Table 2). The incidence and distribution were in agreement with most reported series.[4,5,7]

Treatment

It is well known that (1) pulmonary complications, (2) prematurity, (3) associated major congenital anomalies, and (4) anastomotic leaks are the major factors leading to death in these infants.[3-5,7,8] The deaths in this series further emphasize the above causes of death (Table 3). The importance of gastrostomy has become more widely accepted in recent years, and all patients in our care now have emergency gastrostomy under local anesthesia as soon as the diagnosis is established. The reasons are as follows: Diaphragmatic motion and pulmonary function are better after gastric decompression, endotracheal anesthesia is easier and safer, postoperative feedings can be given through the gastrostomy until the infant is vigorous enough to swallow via the esophagus, and retrograde esophageal dilation, using the Tucker dilators, is easily performed through the opening. In this series of patients there were 19 who had gastrostomy, and 11 of them survived. Of the seven who did not have gastrostomy, one survived.

Following gastrostomy, upper pouch suction is instituted with the Replogle tube connected to constant suction. In addition, members of the nursing staff perform manual pharyngeal suction frequently to protect the trachea from overflow of saliva.

Although extrapleural dissection for interruption of the tracheo-esophageal fistula and anastomosis of the esophagus requires approximately 20 minutes additional operating time, this time is well spent in comparison with the quicker transpleural approach to the trachea and esophagus. Even if a small opening is made in the pleura, it is advisable to proceed with the extrapleural dissection. There are fewer pulmonary complications with this approach but, more important, if there is an anastomotic leak this is well tolerated through the extrapleural space, but virtually a mortal insult if the esophagus leaks transpleurally. With the extrapleural approach, staging (which we formerly recommend for premature infants) is rarely indicated or necessary.[8] The fourth rib is resected subperiosteally and the extrapleural space is approached through the bed of the fourth rib. By so doing we hope to obviate the scoliosis which has been a sequel to a vertical incision with multiple rib resections. Of 12 patients with the extrapleural dis-

TABLE 1.—*Survival of 30 Infants with Tracheo-Esophageal Fistula and Esophageal Atresia*

Time	Total Patients	Survivors Number	Survivors Percent
1944-57	9	1	11.0
1958-63	12	3	25.0
1964-68	9	8	88.8
25 Years	30	12	40.0

TABLE 2.—*Associated Congenital Anomalies in 14 Patients with Tracheo-Esophageal Fistula and Esophageal Atresia*

Anomaly	Number
Congenital Malformations of the Heart and Great Vessels	13
Genitourinary System	6
Musculoskeletal	6
Gastro Intestinal Tract	4
Respiratory	4
Mongolism	4
Miscellaneous	2

TABLE 3.—*Factors Responsible for Death in 18 Patients with Tracheo-Esophageal Fistula and Esophageal Atresia*

Time	Total Patients	Cause of Death	Number	Total Deaths
1944-57	9	Pulmonary Complications	6	
		Major Congenital Anomaly	1	
		Cardiac Arrest Intraoperative	1	
			—	8
1958-63	12	Pulmonary Complications	4	
		Cardiac Arrest Intraoperative	2	
		Anastomotic Leak	2	
		Major Congenital Anomaly	1	
			—	9
1964-68	9	Interstitial Pneumonia	1	1

section, eight have survived, and of 14 with the transpleural dissection four have survived.

An Unusual Sequela

In one patient with esophageal atresia, achalasia developed at age six years. It is well known that esophageal motility in patients with esophageal atresia is abnormal. Lind[9] demonstrated that esophageael motility in patients who have had surgical correction of esophageal atresia is similar to that of achalasia in adult patients. It is somewhat puzzling, then, that more patients with esophageal atresia do not develop clinical and x-ray findings of achalasia. The patient in our series in whom achalasia did develop was one of a set of premature twins who was treated by surgical staging.[10] Initially, the fistula was ligated in continuity and subsequently the esophagus was reconstructed anatomically. A recurrent tracheo-esophageal fistula developed with a rather severe esophageal stricture, requiring resection and esophageal reanastomosis. When the patient was six years old it became apparent he had achalasia (Figure 1). The time-honored Heller type myotomy[11] was temporarily beneficial, but achalasia recurred and the second time was treated with the Jackson-Mosher dilator, which corrected the defect.

Esophageal Atresia Without Fistula

An infant with esophageal atresia without fistula from either esophageal segment, or with fistula from the upper esophageal segment only, can be treated in a similar staged fashion. The common method of bridging the gap between the two atretic segments of the esophagus has been to interpose a segment of colon.[3,12] This affords a satisfactory conduit for the passage of food and fluids from the mouth to the stomach, but the operation is long and somewhat difficult, and often there are serious complications. Stricture formation, anastomotic leaks, lengthening of the colonic segment and reflux of fluids into the tracheo-bronchial tree, resulting in bronchitis and pneumonia, are common ones. More recently a number of such patients have had eventual anatomic esophageal reconstruction after stretching of the upper pouch.[13,14] In the past it was felt that in some of these patients the gap was too wide to permit end-to-end anastomosis. The majority of such patients can eventually have the advantage of an anatomical esophagus rather than interposition of a colon segment. The following case illustrates the point.

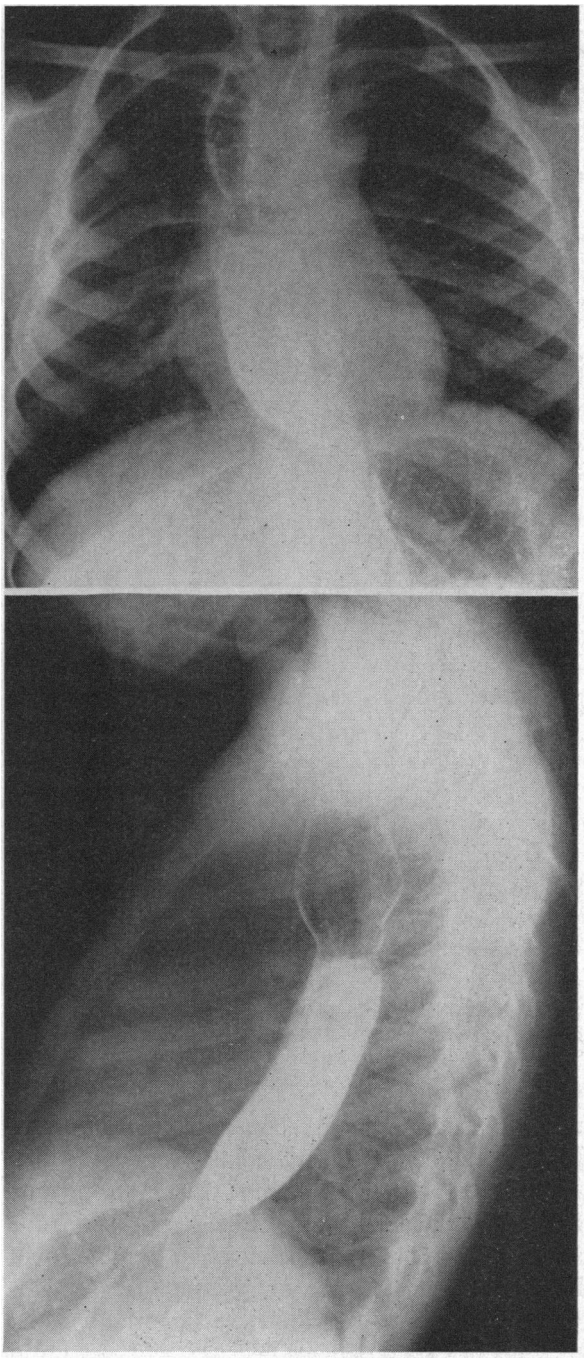

Figure 1.—Achalasia in a six-year-old twin previously operated upon for tracheo-esophageal fistula and esophageal atresia.

Report of a Case

The patient, a baby girl weighing 6 pounds 2 ounces at birth, which was attended by hydramnios, had feeding difficulties. X-ray films

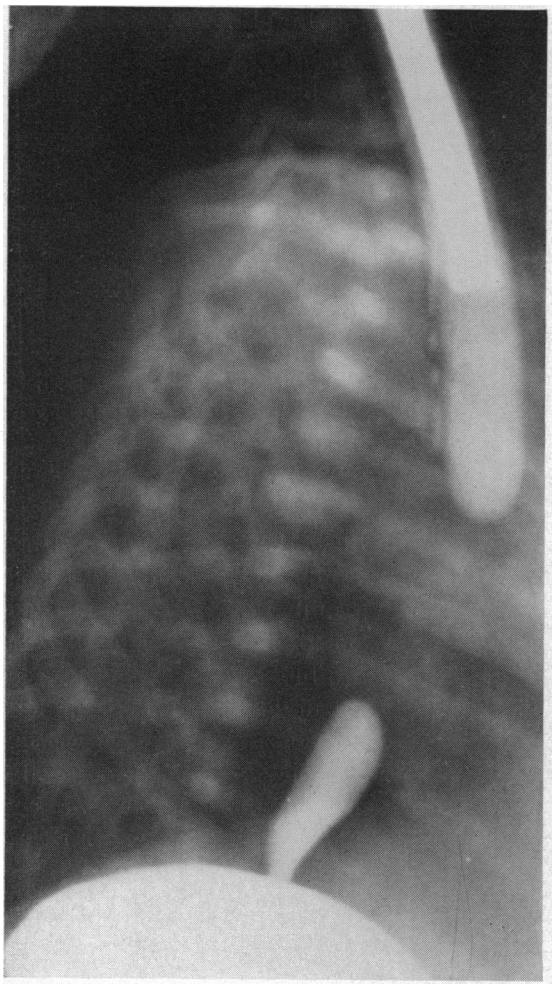

Figure 2.—Stretching of the proximal pouch, using a mercury-weighted bougie. Also seen is the distal atretic segment filled with barium.

revealed a blind upper esophageal segment and a gasless abdomen. Gastrostomy was performed under local anesthesia, after which an opaque medium introduced through the gastrostomy tube was noted to pass from the stomach into the small bowel without evidence of obstruction. (Radiographic studies of this kind are important in the treatment of such patients because, with no gas in the stomach or small bowel, one has no way of excluding the possibility of a small bowel atresia. If small bowel obstruction is present and feedings are attempted the gastrostomy is very likely to leak and peritonitis ensue.) The infant was fed through the tube, and suction was applied to the upper esophageal segment, which was stretched daily with a mercury-weighted bougie (Figure 2). This lengthened the segment, which was 11 cm from the gum line to the distal end at birth, to 14 cm at ten weeks of age. Esophago-esophagostomy was then performed by extrapleural dissection through the bed of the fifth rib. An unanticipated finding was a side fistula between the upper esophagus and the trachea. This defect was closed. Although the distal esophagus also was short, by mobilizing the upper esophagus and the distal esophagus it was possible to bring the two esophageal segments together under moderate tension with two layers of interrupted silk. Postoperatively the anastomotic junction leaked, but this was tolerated by the infant and eventually it closed spontaneously. An esophageal stricture that ensued was overcome by repeated dilation with Tucker dilators. At 10 months of age the patient weighed 16 pounds 1¾ ounces. She required occasional esophageal dilations and was eating food appropriate for her age.

Comment: The unusually wide gap between the esophageal segments in this patient—more than 3 cm at birth—would suggest that, since esophageal anastomosis was successful in this patient the majority of such patients can have an anatomical esophagus with its inherent advantages.

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