Atresia of the Esophagus: *

Increased Survival with Staged Procedures in the Poor-risk Infant

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In the last 5 years, primary end-to-end anastomosis of the esophagus for atresia has resulted in a 92 per cent operative survival in a full-term baby, with no associated severe anomaly and without severe pneumonia. Yet the same procedure carried out in a premature or critically ill baby resulted in only a 38 per cent survival.

After Haight's [7] report of the first successful end-to-end anastomosis of the esophagus in a patient with esophageal atresia, it soon became the standard operation for most cases. With increasing experience it became obvious that the definitive operation carried out as the primary procedure usually resulted in success in a full-term baby but produced a low survival rate in the premature and critically ill patient. In 1962 the report of Holder, McDonald, and Woolley [11] on staging the operative procedures revived interest in an approach which had been suggested by Richter [22] in 1913.

Newborns tolerate one major surgical insult remarkably well in the first few days of life. Complications are tolerated poorly. Thus a simplified operation relatively free of complications is a necessity for the poor-risk patient with esophageal atresia. A review of 249 cases of esophageal atresia at The Children's Hospital of Philadelphia revealed an increased mortality with prematurity, severe pneumonia, severe associated congenital anomalies and in babies critically ill from acquired problems such as septicemia, anoxia, *etc.*[15] If selected patients with additional problems complicating esophageal atresia could be sustained during the first few days or weeks of life, until they could tolerate not only an end-to-end anastomosis of the esophagus but also the potential complications associated with this procedure, the survival rate should increase. Only about one half of the patients with esophageal atresia represent the ideal full-term baby without severe pneumonia or a severe associated anomaly (Fig. 1).

Throughout this report the complicating diagnoses associated with atresia of the esophagus are rigidly defined. *Severe pneumonia* is identified x-ray opacification of one entire lung, of one lobe in each lung or of one lobe in one lung only if there are associated symptoms of respiratory distress such as cyanosis, retraction, or tachypnea. *Hyalin membrane disease* is diagnosed only microscopically at postmortem. *Severe associated anomalies* are those which are immediately incompatible with life and could prove fatal within the operative period of 30 days. All other anomalies are in separate groups and called major or moderate.[7]

Management of 41 Cases During 15-Year Period

Between 1950 and 1964 the initial procedure in the management of esophageal

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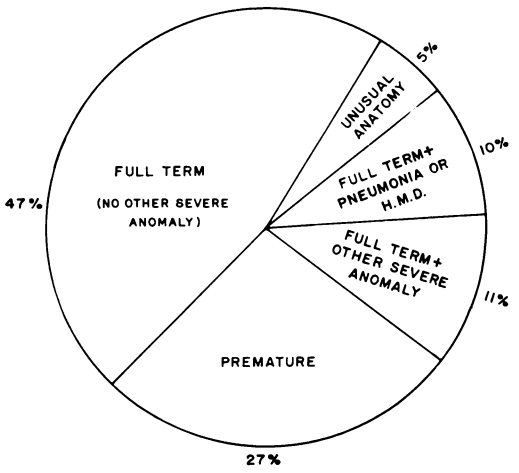

FIG. 1. Distribution of infants in a series of 249 patients with atresia of the esophagus.

atresia was but the first of several operations in 41 patients. Five patients were essentially moribund and had a gastrostomy only to prevent gastric regurgitation through the fistula into the tracheobronchial tree and did not survive long enough to have a second procedure. Our earlier cases were staged by doing a cervical esophagostomy, division of the tracheoesophageal fistula and a Stamm feeding gastrostomy. Such management required an interposition operation at a later date. Patients with delayed colon transplants have uniformly survived but the morbidity has been high. Morbidity usually is associated with angulation of the esophagus in the neck, along with leaks and strictures at the esophagocolon anastomosis.

Since 1962 we have discontinued cervical esophagostomies as the initial procedure in staging these cases. By leaving the upper esophageal pouch intact and not sacrificing any esophageal length, we have been able to perform an end-to-end anastomosis of the esophagus at a later date, or at least able to perform a colon anastomosis in the mediastinum to the undisturbed upper esophageal pouch. Four patients with severe pneumonia and overwhelming tracheobronchial secretions we intended to stage with a long delay between first and definitive operation; however, after gastrostomy in all and tracheostomy in three, an end-to-end anastomosis was carried out when the operative intent was to do only a division of the fistula. The types of staged proceedures which had been scheduled are summarized in Table 1.

Technic

A staged procedure should be short, atraumatic and as free of potential complications as possible. The first operation should insure that the pulmonary problems can be cleared and nutrition maintained until the baby is vigorous enough for an end-to-end anastomosis of his esophagus and all that it entails.

We prefer a transpleural approach under general endotracheal anesthesia. The fistula is divided and the tracheal end oversewn. The esophageal end of the divided fistula is securely closed with interrupted sutures of black silk and sutured high onto the posterior chest wall (Fig. 2). During thoracotomy the surgeon has his best op-

TABLE 1. *Summary of Four Categories of Patients in Whom Some Sequence of Staged Procedures Was Planned*

Procedure		Patients
1. Gastrostomy only		5
Intent (early experience): Prevent gastric regurgitations until desperately ill patient able to withstand definitive procedure.		
2. Cervical esophagostomy, gastrostomy		23
With tracheoesophageal fistula	16	
Without tracheoesophageal fistula	7	
Intent: Eventual colon transplant		
3. Upper pouch intact, gastrostomy		9
With division of tracheoesophageal fistula	7	
No division required; no fistula	2	
Intent: Subsequent end-to-end anastomosis of esophagus when warranted by patient's weight and condition		
4. Upper pouch intact, gastrostomy on suction		4
Tracheostomy for overwhelming secretions	3	
Pneumonia—severe	1	
Intent: As in No. 3, but on thoracotomy for division of tracheoesophageal fistula, end-to-end anastomosis performed		

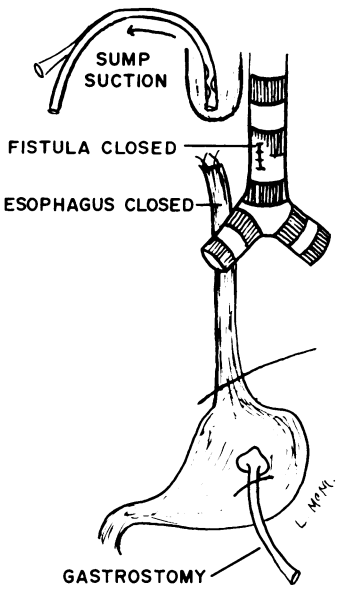

FIG. 2. (Left) Diagramatic sketch of the essential steps in the first of the staged procedures: closed tracheoesophageal fistula, closed distal esophagus tacked to posterior chest wall, gastrostomy and sump catheter in proximal esophagus.

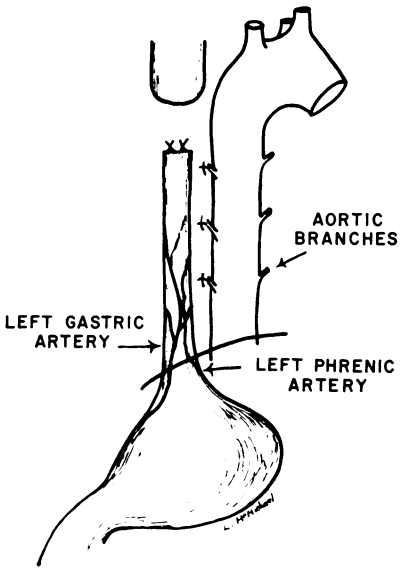

FIG. 3. (Right) Diagramatic sketch of the blood supply to the distal esophagus from the left gastric and left phrenic arteries after division of the aortic branches.

portunity to learn the anatomy of the defect and to make plans for eventual definitive operation. If the anesthetist passes a firm plastic catheter down the oral pharynx into the esophagus to outline the lower limits of the upper pouch, the surgeon can get a good idea of the gap between the two esophageal segments which eventually must be bridged. The later delineation of the size of the upper pouch with radiopaque dye may be hazardous.[8]

Postoperatively the baby is maintained on antibiotics until the tracheal culture taken on admission and its sensitivities to antibiotics are reported. At that time appropriate changes in antibiotic coverage are made. The baby is maintained in a high humidity, warm Isolette®* in the head-up position. The upper pouch is kept empty with a sump catheter ** which is maintained on constant suction. The patency of the catheter must be insured with irrigations of 3 cc. of saline once or twice an hour. Occasionally the use of mucolytic enzymes such as papaine or acetylcystine might be helpful. The irrigating solution will return immediately if the catheter is patent.

Special precautions are carried out to prevent and correct any postoperative pulmonary problems because of 50 per cent of the deaths with this anomaly are still due to pneumonia. The patient is turned from side to side at hourly intervals. The nasopharynx is aspirated with a red rubber catheter as a double precaution against accumulations of secretions. Routine tracheal aspirations are carried out every 12 hours with direct exposure of the cords by means of a laryngoscope. The frequency of endotracheal aspiration may be altered depending upon the amount and consistency of the tracheal secretions. Gastrostomy feedings are started on the second or third postoperative day, there being no fear of aspiration of vomitus.

The extrapleural approach [11] is used by some, and at times under local anesthesia. We believe that the speed and improved exposure obtained by the transpleural route, as well as the advantage of controlled respirations and the ease of endotracheal toi-

* Air Shields, Hatboro, Pennsylvania.

** Replogle Sump Catheter, Aloe Medical, 1831 Olive Street, St. Louis 3, Missouri.

lette, outweigh the possible advantage of local anesthesia. We have not had trouble with lung adhesions at the subsequent operation. Early in our series of staged procedures a triply ligated fistula recanalized and the ligature cut through, leading to the demise of the patient from aspiration pneumonia. Similar reports confirm the dangers from ligated fistulas [4, 9, 11, 34] and the procedure should be discarded.

Other sequences of procedure may be used in staging. Some prefer a preoperative gastrostomy under local anesthesia until the child is better able to tolerate the division of the fistula. This procedure has several disadvantages: 1) aspirations can easily occur despite gastrostomy, 2) feedings cannot be started until the fistula is closed and 3) certain fistulas are so large that the loss of air through the gastrostomy may further embarrass respiration. Martin described an infant who could cry audibly only with his gastrostomy occluded.[19]

Early in our experience, endotracheal aspiration in patients with a gastrostomy—but with the tracheoesophageal fistula intact—resulted in repeated aspiration of bile from the tracheobronchial tree. The increased abdominal pressure from straining during the procedure resulted in gastric content being poured into the lungs and made the fistula division necessary.

Anatomy of the Esophagus in Esophageal Atresia. The esophagus is a tenuous structure because it lacks a serosal layer and consequently supports sutures poorly. In esophageal atresia the upper pouch gets a rich blood supply from the inferior thyroid artery. The distal esophagus is often frail and extremely thin from disuse and gets its blood supply from the left gastric and phrenic arteries, along with the small direct branches from the aorta. The tracheal end of the fistula may be supplied by the bronchial artery.[17] The blood supply of the distal esophagus is at best minimal and may be destroyed further by being mobilized for anastomosis to the proximal pouch. The caliber of the fistula and the thickness of its wall varies considerably from patient to patient.

Some surgeons prefer some tension on the anastomosis with minimal dissection [31] to a comfortable anastomosis afforded by greater mobilization of the distal esophagus,[25] with the concomitant risk to the distal blood supply. In staging—if the patient's condition permits—the problem of blood supply can be overcome, at least in part, by dividing the aortic branches at the initial operation in order to encourage an increased intramural blood supply to the lower esophagus from the left gastric and phrenic arteries, which will not be destroyed in freezing up the esophagus for the definitive procedure (Fig. 3). The absence of anastomosis accompanying the freshly destroyed blood supply to the distal esophagus should enhance healing of the suture line and minimize a leak at the next procedure.

Experimentally it has been shown [27] that the gastric arteries alone are sufficient to keep the esophagus viable and heal an anastomosis. The proximal pouch in patients with esophageal atresia has normal co-ordinated peristalsis. The distal esophagus almost invariably has a motor dysfunction,[3, 14] resulting either in no peristaltic movements or dysrhythmic spastic contractions. There is no uniformity of opinion as to whether this results from a congenital defect in the vaginal innervation or whether it is a result of trauma at the time of operation.

Results

Five patients who had gastrostomy only are all dead. Four had severe congenital heart disease and were managed before our commitment to staged procedures. The gastrostomy was carried out only to lessen gastric regurgitation, a phenomenon which we now realize occurs readily despite gastrostomy in many patients. The fifth patient was a 2 pound, 11 ounce infant

with esophageal atresia and a ruptured myelomeningocele. He died after gastrotomy under local anesthesia and before we could divide his fistula.

Of 23 patients who had a cervical esophagostomy and gastrostomy with division of the tracheoesophageal fistula where present, 13 died before the intended colon transplant could be carried out. Analysis of the causes of death indicate that nine of 13 were beyond our control and that four might be classified as preventable. Some of these patients without tracheoesophageal fistula died even though no thoracic procedure was undertaken. All 10 patients who had a colon transplant survived.

Table 2 summarizes the nine patients who had an initial gastrostomy and division of the tracheoesophageal fistula when present but whose upper esophageal pouches were left intact and maintained free of secretions by suction. Only one was not a premature baby (F. A.) and his associated problems were a clear indication for staging. This child died, as did all Mongols in this series (in contrast to survival with other gastrointestinal anomalies). Death was due to congested heart failure from a large ventriculoseptal defect. The death due to recanalization of a ligated fistula has already been discussed.

The two remaining deaths were unfortunate. One infant died after compression of his airway at 6 weeks due to an enlarging pulmonary artery aneurysm. He had already survived pyloromyotomy for pyloric stenosis at the age of 6 weeks. The other patient died of anaphylactic shock a few minutes after injection of penicillin for upper respiratory infection at the age of 6 months.

Four patients had definitive operations, although scheduled for staged procedures, a few days after the initial operation. All were in serious distress from tracheobronchial secretions associated with pneumonia and probably due in part to aspiration of diagnostic radiopaque material before admission. Three had tracheostomies; one was repeatedly aspirated endotracheally. When thoracotomy was done for division of the fistula, the close proximity of the two esophageal segments led the surgeon to do definitive end-to-end anastomosis instead. All survived.

Complications of Staging. Though the staged procedures are relatively free of complications constant, skilled, experienced nursing remains essential. The sump catheters can get plugged and result in aspiration pneumonia unless attention is paid to the hourly irrigations. Once a catheter is plugged it should be replaced. We have noted that after the first 2 weeks of life, some babies taking gastrostomy feedings well and, maturing normally, can handle their saliva almost without the aid of a suction catheter. As they get older, cough reflexes are stronger and they can spit up any excess saliva. Nevertheless we have maintained suction on the pouch as long as it remains intact. Before we began using the sump catheters, we kept several patients in the head-down position after the fistula had been divided and aspirated the nasopharynx every hour or two intermittently. The infants handled their secretions remarkably well after 2 weeks and suctioning became almost unnecessary. One infant in the head-down position developed a leak of the distal esophageal stump, resulting in a mediastinal abscess which required subsequent extrapleural drainage. After this experience, we discarded feeding patients in the head-down position in preference to the head-up position with a sump catheter.

One patient developed an aspiration pneumonia at 7 weeks from radiopaque dye as we tried to estimate the distance between the upper pouch and the distal esophagus. We now believe that the size of the upper pouch can be determined best at the time of original operation by means of a catheter inserted in the pharynx. If

TABLE 2. *Summary of Nine Patients Whose Upper Esophageal Pouch Was*

Patient	Birth Wt.	Operative Weight	Age Dx Made	Reason for Staging	Age at Division of Fistula
D. E. S.	3 lb. 0 oz.	2 lb. 13 oz.	2 days	Prematurity	
A. Y.	3 lb. 5 oz.	3 lb. 2 oz.	2 days	Prematurity, respiratory distress	3 days
F. I.	4 lb. 4 oz.	3 lb. 8 oz.	5 days	Prematurity, pneumonia, duodenal atresia, septicemia	5 days ligated
D. E. A.	3 lb. 14 oz.	3 lb. 12 oz.	2 days	Prematurity, aspiration pneumonia	5 days
A. P.	4 lb. 1 oz.	3 lb. 15 oz.	1 day	Prematurity, respiratory distress	2 days
B. R.	4 lb. 8 oz.	4 lb. 2 oz.	1 day	Prematurity, penumonia; dextrocardia	2 days
F. L.	4 lb. 3 oz.	4 lb. 3 oz.	1 day	Prematurity	1 day
D. I.		4 lb. 6 oz.	4 days	Aspiration pneumonia—barium; prematurity	4 days
F. A.	7 lb. 1 oz.	7 lb. 2 oz.	1 day	No fistula, perforation of stomach, duodenal atresia, Mongolism, congential heart disease—severe genitourinary anomalies	

there is a relative certainty of a wide gap between the two ends of the esophagus, plans should be made not to perform the definitive operation until the child is strong enough to tolerate a colon transplant if necessary. Other complications have been reported prior to second procedure, such as gastric perforation [6, 23] and recanalization of ligated fistulas, already mentioned.[4, 9, 11, 34]

The consequences of gastrostomy in the newborn should be considered. Although it has been recommended [12, 30] for routine use, Connor [1] reported a complication rate of 38.5 per cent in gastrostomies in the newborn, with a 30 per cent mortality in patients with gastrostomies performed for esophageal atresia. Nonabsorbable sutures should be used. Gastrostomies have been considered as a possible etiologic factor in pyloric stenosis. We have had four cases of pyloric stenosis in this series, two with gastrostomy. An additional 16 cases of pyloric stenosis with gastrostomies have been reported in the literature.[2, 5, 24, 26] One of our patients treated by staged procedures developed pyloric stenosis at 5 weeks and required a pyloromyotomy. When indicated,

Left Intact in the Mediastinum During the Course of Staged Procedures

Age at Definitive Operation	Weight at Definitive Operation	Definitive Operation	Course and Result
9 months	17 lb. 8 oz.	Intrathoracic colon transplant	Uncomplicated, living and well
4 months	9 lb. 2 oz.	End-to-end anastomosis of esophagus	Distal esophageal leak at 6 weeks; pleural abscess drained; died at 6 months—anaphylactic shock from penicillin
			Duodenojejunostomy; recanalized fistula and died on 18th postoperative day
4 months	9 lb. 6 oz.	End-to-end anastomosis of esophagus	2 days—gastrostomy, 2 days—tracheostomy, living and well
70 days	7 lb. 9 oz.	End-to-end anastomosis	Tracheostomy—lipiodal inspiration pneumonia, living and well
			Pyloromyotomy for pyloric stenosis 6 wk.; respiratory distress 8½ wks. from large pulmonary artery aneurysm; died
3 months	6 lb. 12 oz.	End-to-end anastomosis of esophagus	Recurrent fistula at 5 mo.; empyema, C. esophagostomy, colon transplant—3½ years; living and well
33 days	6 lb. 10 oz.	End-to-end anastomosis of esophagus	Tracheostomy, stricture of anastomosis revised 2½ wks. later, dilatations, living and well
2 months	7 lb. 8 oz.	Creation of small hiatal hernia, end-to-end anastomosis of esophagus	Died at 3½ months from congestive heart failure; autopsy—large ventriculoseptal defect

the live-saving value of the gastrostomy cannot be challenged. However, its routine use in all cases seems inadvisable. Forty per cent of patients in this series had a gastrostomy during the course of treatment.[4]

Discussion

Between 1950 and 1954, a primary end-to-end anastomosis of the esophagus was carried out in 93 per cent of our patients with the common type of esophageal atresia. Since 1963 when we began leaving the upper esophageal pouch intact, only 60 per cent of the patients with the common type of esophageal atresia have had a primary end-to-end anastomosis. During the same period only 25 per cent of premature infants had a primary anastomosis. The remaining patients were all staged. Poor-risk patients are now allowed time to recover from any pulmonary complications and grow more mature and vigorous before definitive esophageal repair is carried out.

The thin-walled, flimsy distal esophagus hypertrophies and dilates as the result of reflux gastric contents due to the incompetent gastroesophageal junction of an infant[29] and presents eventually as a thick-

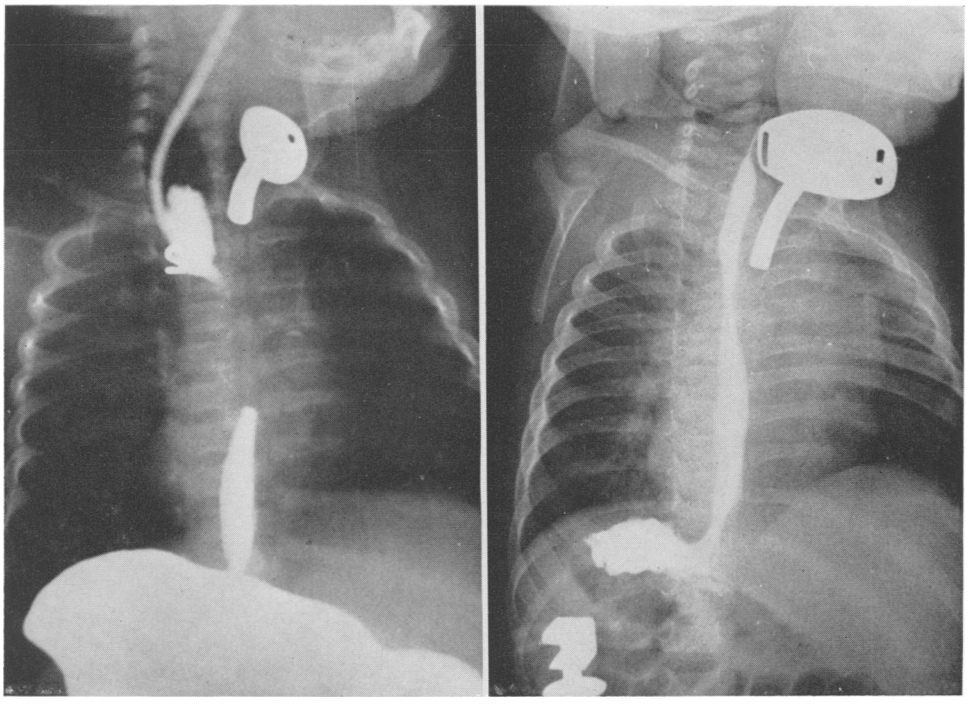

FIG. 4. Patient DEA (Table 1). Left. Illustrates gap between upper blind pouch and the distal esophagus in an infant born at 3 lb. 14 oz. Right. Esophagus after end-to-end anastomosis carried out at 4 months.

walled, lengthened structure with a good blood supply, somewhat equivalent to the proximal pouch in caliber. Wide gaps can be approximated rather easily because of the better blood supply and better quality of the esophageal tissue (Fig. 4a, b).

With the exception of two patients, those who had the common type of esophageal atresia and the upper pouches left intact with the intent to stage had an end-to-end anastomosis of the esophagus performed eventually. Of the two cases with no fistula, a small hiatal hernia was created in one to facilitate anastomosis and a colon transplant was performed in the other. At least three procedures are available to gain the added length when a wide gap needs to be bridged. Howard [13] has stretched at least one upper pouch periodically with a No. 12 mercury bougie and produced enough elongation of the upper pouch to complete a satisfactory end-to-end anastomosis to a short lower segment.

By cutting a part or all of the phrenoesophageal ligament, added esophagus can be gained to facilitate the anastomosis by the creation of a small hiatal hernia. If this is done the baby should be maintained in the head-up position postoperatively in order to cut down on reflux into the esophagus. This should not be confused with previous attempts [18] to pull the entire stomach into the chest to bridge a wide gap as in patients with no fistula. Severe respiratory embarrassment and reflux esophagitis resulted in the abandonment of this procedure. We eventually produced a small hiatal hernia on an extremely poor-risk Mongol with duodenal atresia, esophageal atresia without a fistula and a gastric perforation. The perforation was closed and a gastrojejunostomy was performed along

with the gastrostomy. After the baby became more vigorous and mature a definitive operation was carried out by anastomosing the two ends of the esophagus after creating the small hiatal hernia. Our plan was to repair the hernia subsequently and restore the gastroesophageal junction to its normal position. This baby, however, died at 14 weeks from congestive heart failure due to a large ventriculoseptal defect. Zachary [35] has created such a hiatal hernia in an infant in order to facilitate end-to-end anastomosis of the esophagus. The hiatal hernia was repaired at 2 years of age. The patient is now swallowing normally and does not have a hiatal hernia.

An interposition operation with an isoperistaltic transplant can be carried out in the neonatal period. We have discontinued colon transplants in the newborn, as have others,[18, 20] because of the unacceptable high mortality.

Our preference for esophageal replacement when the upper pouch is intact is the transverse colon brought up through the normal esophageal hiatus. If a short segment is needed the middle colic blood supply is used. Recently, however, we have used the left colic artery[28] when a long segment of colon is needed. This gives a better blood supply and more equal diameter of colon to esophagus and a maximum length of bowel can be transplanted.

Single layer anastomosis with decreased strictures (27%) but increased leak rate (17%) now is more valuable since there is no urgency for the surgeon to get the ends of the esophagus together. This should significantly decrease the incidence of leaks and strictures because the anastomosis under tension will not be necessary. We have not had a single leak in a primary anastomosis of the esophagus in the past 2½ years—since we began to stage cases and leave the upper pouch intact.

The patient with esophageal atresia without a fistula always has been considered to carry a high mortality [10, 21] because of his increased prematurity,[6] increased number of associated anomalies [32] and the added difficulty of getting the two ends of the esophagus together because of the long gap. These patients do have a decreased incidence of pneumonia to outweigh other risk factors.

Exploratory thoracotomy has been commonly practiced in patients with esophageal atresia and an airless abdomen in an attempt to find a lower esophagus long enough for a primary anastomosis. In our series only three of 249 patients with esophageal atresia had no air in the abdomen by x-ray and yet had a connection of the lower esophagus to the trachea. One of these had a fistula which was blocked with a mucus plug; the other two had atretic esophageal attachments to the trachea. The rarity [33] of a fistula in the absence of air in the stomach by x-ray should discourage an exploratory thoracotomy in these patients since very few have a patent connection to the trachea. After thoracotomy has been omitted the lungs should be examined carefully to detect that rare case with a mucus plug blocking the fistula.[16] Instrumentation of the lower esophagus to determine its length at the time of gastrostomy can be a difficult procedure and is not recommended for the premature infant.

Our lightest patient (2 pounds, 13 ounces) had esophageal atresia without a fistula. A gastrostomy was performed and the upper pouch handled in the usual manner. This patient subsequently was sent home with a sump catheter in his upper pouch. He was admitted one month later because of an incarcerated inguinal hernia which was repaired and again he was sent home with a sump catheter until he was scheduled for his colon transplant. Not every mother can be trusted with this type of home care, but we have met several whom we thought competent after careful, lengthy instruction.

TABLE 3. *Summary of Patients Whose Initial Operation (Tracheostomy, Gastrostomy) Was Done with Intent to Stage Procedures but Who Had Definitive Operations after a Few Days Delay*

Patient	Birth Weight	Operative Weight	Age Dx Made	Reason for Staging	Age at Definitive Operation	Weight at Definitive Operation	Definitive Operation	Course and Result
S. H.	5 lb. 0 oz.	4 lb. 10 oz.	1 day	Pneumonia, severe atelectasis	2 days	4 lb. 10 oz.	End-to-end anastomosis of esophagus	Living and well
L. E.	5 lb. 12 oz.	5 lb. 10 oz.	2 days	Pneumonia, severe	5 days	5 lb. 0 oz.	End-to-end anastomosis of esophagus	Tracheostomy, living and well
S. T.	6 lb. 11 oz.		1 day	Pneumonia, severe	6 days	6 lb. 1 oz.	End-to-end anastomosis of esophagus	Tracheostomy, resection of stricture of anastomosis, living and well
N. O.	7 lb. 8 oz.	7 lb. 0 oz.	2 days	Pneumonia, severe	4 days	6 lb. 13 oz.	End-to-end anastomosis of esophagus	Tracheostomy, 2 months resection of stricture, living and well

The sump catheter can be used to manage the rare patient in whom anastomosis becomes so strictured as to prevent feeding. We have so treated one patient until his pneumonia cleared and he was considered strong enough to withstand resection of his stricture and a second anastomosis.

The operation of choice in patients with esophageal atresia and tracheoesophageal fistula is a primary end-to-end anastomosis of the esophagus following division and oversewing of the fistula. In the poor-risk patient experience has shown this operation to give an unacceptably high mortality. Our brief experience with staged procedures in this type of patient has been gratifying.

Although staging involves a prolonged hospital stay, increased expense and the skilled personnel necessary to implement the program, survival will be improved in a group of patients formerly considered to be hopeless.

Summary

The results of staging operative procedures in 41 patients with esophageal atresias are presented. Twenty-three patients had a cervical esophagostomy and gastrostomy performed as the first stage. Thirteen had the upper esophageal pouch left intact but had division and closure of the tracheoesophageal fistula and gastrostomy. Five patients had a gastrostomy only.

Primary end-to-end anastomosis of the esophagus yields good results in full-term babies without severe pneumonia or severe associated anomalies (92%) and discouraging results (38%) in prematures and poor-risk patients.

Staging of operative procedures should be considered in premature babies, in patients with an unsatisfactory esophageal anatomy to permit an end-to-end anastomosis of the esophagus, in those with other major anomalies incompatible with life but

amenable to correction and in critically ill patients.

A series of staged procedures is presented which consists of division and closure of both ends of the tracheoesophageal fistula, tacking the distal esophagus to the thoracic wall, the performance of a Stamm feeding gastrostomy and constant suction by a sump catheter on the proximal esophageal pouch. Eventual end-to-end anastomosis can be carried out on the esophagus when the baby is mature and vigorous enough to tolerate the procedure.

Our operative survival has improved from 38 to 92 per cent in poor-risk patients using the staged procedures as described. We believe that staged procedures have much to offer those patients who have had such a low survival with a conventional approach.

References

1. Connar, R. G. and W. C. Sealey: Gastrostomy and Its Complications. Ann. Surg., **143**:245, 1956.
2. Clatworthy, W. H.: Esophageal Atresia, Importance of Early Diagnosis and Adequate Treatment Illustrated by a Series of Patients. Pediatrics, **16**:122, 1955.
3. Desjardins, J. G., C. A. Stephens and C. A. F. Moes: Results of Surgical Treatment of Congenital Tracheo-esophageal Fistula with a Note on Cine-Flourographic Findings. Ann. Surg., **160**:141, 1964.
4. Fevre, M. and B. Duhamel: Atresia de l'oesophage. Arch. Franc. Ped., **8**:615, 1957.
5. Gross, R. E.: The Surgery of Infancy and Childhood. Philadelphia, W. B. Saunders Co., 1953.
6. Haight, C.: Some Observations on Esophageal Atresia and Tracheoesophageal Fistula of Congenital Origin. J. Thor. Surg., **34**:141, 1957.
7. Haight, C. and H. A. Towsley: Congenital Atresia of the Esophagus with Tracheoesophageal Fistula. Surg. Gynec. & Obstet., **76**:672, 1943.
8. Hamilton, J. P., C. E. Koop and J. W. Hope: The Danger of Radiopaque Matèrial in the Diagnosis of Atresia of the Esophagus. To be published.
9. Hayes, D. M. and W. H. Snyder: Results of Conventional Operative Procedures for Esophageal Atresia in Premature Infants. Amer. J. Surg., **106**:19, 1963.
10. Hays, D. M.: An Analysis of the Mortality in Esophageal Atresia. Amer. J. Dis. Child., **103**:765, 1962.
11. Holder, T. M., V. G. McDonald and M. M. Woolley: The Premature or Critically Ill Infant with Esophageal Atresia: Increased Success with a Staged Approach. J. Thor. Cardiov., **44**:344, 1962.
12. Holder, T. M. and R. E. Gross: Temporary Gastrostomy in Pediatric Surgery. Pediatrics, **26**:36, 1960.
13. Howard, R.: Personal Communication. To be published in Surgery.
14. Kirkpatrick, J. A., S. L. Cresson and G. P. Pilling, IV: The Motor Activity of the Esophagus in Association with Esophageal Atresia and Tracheo-esophageal Fistula. Amer. J. Roentgenol., **86**:884, 1961.
15. Koop, C. E. and J. P. Hamilton: Factors Affecting Survival in Esophageal Atresia—Experiences with 249 Patients. To be published.
16. Linberg, E. J. and B. Erfan: Esophageal Atresia and the Gasless Abdomen. J. Lancet, **84**:83, 1964.
17. Lister, J.: The Blood Supply of the Esophagus in Relation to Oesophageal Atresia. Arch. Dis. Child., **39**:131, 1964.
18. Longino, L., M. M. Woolley and R. E. Gross: Esophageal Replacement in Infants and Children with use of a Segment of Colon. J.A.M.A., **171**:1187, 1959.
19. Martin, L. W.: cited by Richardson *et al.*, Congenital Atresia of the Esophagus. Ann. Surg., **29**:166, 1963.
20. Rehbein, F. and S. Hofmann: Oesophagusatresie mit Duodenalverschlub und Analatresie, Zugleich ein Beitrag zur Primaren Kolonersatzplastik. Z. Kinderchir., **1**:57, 1964.
21. Rehbein, F. and F. Yanagiswa: Complications After Operation for Esophageal Atresia. Arch. Dis. Child., **34**:24, 1959.
22. Richter, H. M.: Congenital Atresia of the Oesophagus. An Operation Designed for its Cure. Surg. Gynec. & Obstet., **17**:397, 1913.
23. Rosenkrantz, J. G. and F. J. Cozzetto: Distal Esophageal Perforation: Complication of Staged Repair of Esophageal Atresia. Surgery, **54**:678, 1963.
24. Rutten, A. P. M.: Oesophageal Atresia, A Critical Study of the Literature and Report of Sixteen Cases. Academisch Proefschrift 1962, H. J. Koersen en Zonen, Amsterdam.
25. Sandblom, P. H.: Treatment of Congenital Atresia of Esophagus from a Technical Point of View. Acta Chir. Scand., **97**:25, 1948.
26. Schwartz, S. I. and W. A. Dale: Unusual Tracheo-esophageal Fistula with Membranous Obstruction of the Esophagus and Postoperative Hypertrophic Pyloric Stenosis. Ann. Surg., **142**:1002, 1955.
27. Shek, J. L., C. A. Prietto, W. Tuttle and E. J. O'Brien: An Experimental Study of the Blood Supply of the Esophagus and its Relation to Esophageal Resection and Anastomosis. J. Thor. Surg., **19**:523, 1950.
28. Sherman, C. D. and D. Waterston: Oesophageal Reconstruction in Children Using Intrathoracic Colon. Arch. Dis. Child., **32**: 11, 1957.
29. Strowczynski, H., I. T. Beck, R. D. McKenna and G. H. Nickerson: The Behavior of the Lower Esophageal Sphincter in Infants and

Its Relationship to Gastroesophageal Regurgitation. J. Pediat., 64:17, 1964.
30. Swenson, O., R. Lipman, J. H. Fisher and F. G. DeLuca: Repair and Complications of Esophageal Atresia and Tracheo-esophageal Fistula. New Engl. J. Med., 267:960, 1962.
31. Swenson, O.: End to End Anastomosis of the Esophagus for Esophageal Atresia. Surgery, 22:324, 1947.
32. Waterston, D. J., R. E. Bonham-Carter and E. Aberdeen: Congenital Tracheo-esophageal Fistula in Association with Oesophageal Atresia. Lancet, 2:55, 1963.
33. Waterston, D. J., R. E. Bonham-Carter and E. Aberdeen: Oesophageal Atresia: Tracheo-esophageal Fistula. Lancet, 1:819, 1962.
34. Woolley, M. M., R. F. Chinnock and H. P. Richard: Premature Twins with Esophageal Atresia and Tracheo-esophageal Fistula. Acta Paed., 50:423, 1961.
35. Zachary, R. B.: Personal communication.

Discussion

Dr. Frank Allbritten (Kansas City, Kans.): Dr. Tom Holder in our Department has been interested in this problem and asked that I discuss it because he could not be here. The information he has given me I am relaying to you.

We agree with Dr. Hamilton and Dr. Koop that certain patients with esophageal anomalies are better managed by a staged, rather than a primary, repair. There is now general agreement that esophageal atresia without tracheoesophageal fistula is best treated by gastrostomy and cervical esophagostomy in the newborn period, with a subsequent color interposition.

We believe a staged approach in the premature infant with the usual esophageal atresia and distal tracheoesophageal fistula does have advantages. The staged approach has consisted of immediate gastrostomy under local anesthesia, followed some 24 hours after admission by a retropleural division of the tracheoesophageal fistula under local anesthesia.

A subsequent esophagoesophagostomy is carried out transpleurally under general anesthesia when the patient has reached appropriate maturity and size. (Slide)

Our current indications for a staged approach are patients under four pounds; patients four to five pounds if not vigorous; patients over five pounds who are quite ill, even after gastrostomy.

Initially, all patients premature by weight were treated by a staged approach. It is apparent, however, that some of these patients of four to five pounds who are vigorous, have a lusty cry and minimal pulmonary complications can tolerate a primary repair.

This slide (Slide) shows the experience with premature infants treated by a staged approach. These are not in chronological order but are listed simply by their weight.

During this time, four patients weighing between four and five pounds were treated by a primary repair and survived. There were five deaths in the 15 patients treated by the staged approach. During this same period of time, four full-term, critically ill patients were staged, with two survivors.

You will note that the heavier patients in the previous slides actually had the highest mortality, but this slide gives some information why that occurred. The associated anomalies present in the five prematures who died are shown in this slide. (Slide)

Although we have not yet surgically attacked the associated anomalies, it seems reasonable that some of these patients might be palliated or corrected between the second and third stage of the esophageal repair. Since reduction of the mortality of the premature by the staged approach, further significant mortality reduction will have to be made by salvaging patients in the large group with associated anomalies.

Dr. Harris B Shumacker (Indianapolis): We have had generally good results and this is attributable to the early and sustained work of my associate, Dr. J. S. Battersby, who has had direct charge of patients with tracheoesophageal fitula.

From 1950 to date, 178 consecutive patients with esophageal atresia have been operated upon. No patient has been excluded, regardless of associated anomaly or disease, or the severity of any associated disease. There has been no reduction in the average age at the time of admission during the 15-year period. There has also been no increase in the average weight, in the first period the average weight being 6¼ pounds and in the last, 5½ pounds.

Except those who obviously required esophageal replacement, such as those with atresia without fistula, the aim has been a retropleural, one-stage primary anastomosis and this has almost invariably been achieved. The survival rate, in an over-all sense, has been related to patient size. Those few 7 pounds or over had a survival rate of 88%, those ranging from 5 to 7 pounds, from 55 to 60%, and those from 4 to 5 pounds, 31%.

During this period, it has been necessary to carry out esophageal replacement in all patients, all of whom have survived, though one late death occurred.

Further examination of the data shows that the survival rate of the entire group is related principally to the associated anomalies and associated disease. In one recent 5-year period, for example, there were nine deaths. Five of these were due to cardiac or genitourinary lesions which, in our opinion, were totally incompatible with survival. Two were due to advanced pneumonitis present at the time of admission, and the other two to unfortunate technical nursing errors. It is difficult to see