Congenital Atresia of the Esophagus

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ATRESIA of the esophagus ranks second only to imperforate anus as a cause of neonatal alimentary obstruction. Also, it is the most common of all esophageal anomalies, the most frequent cause of neonatal difficulty in swallowing and, when in its usual association with a tracheo-esophageal fistula, is a major cause for respiratory distress in the neonatal period. Although the earliest survivors treated by Leder, Ladd, and Haight are now of voting age, 6, 12, 18 this congenital lesion with or without tracheo-esophageal fistula continues to present a major challenge in surgical management. Koop 14 recently stated it clearly: "...management of atresia of the esophagus has become a criterion of competence for both individual surgeon and institution in the field of pediatric surgery."

We recently have reviewed the experience with 42 consecutive cases of esophageal atresia diagnosed clinically at the University of Oklahoma Medical Center from 1940 through 1961, covering the 22 years since the first successes were achieved in this country. Our purpose today is to report a belated but gratifying trend in local results, to review the mortality, morbidity, and survival factors for the last five years, and to outline our current plan of management.

Incidence

Review of this small series suggests that the cases be considered in three chronological groups. In the first eight years (1940-1947) the diagnosis was made infrequently and late and the treatment should not really be called surgical. There were only five cases recognized, the average age at diagnosis was almost eight days, and one infant died without operation while three of the four gastrostomies were considered palliative. During the nine-year middle period (1948-1956), there were 19 cases, and the admission age averaged only 2.8 days despite one 12-day old referral. A definitive if not aggressive surgical plan was evident throughout this period although during the first half, one infant died in 36 hours without operation, five were considered at exploration to be impossible to anastomose and only three had an anastomosis completed. The first primary anastomosis performed here was on Case 7 (1948). The first surgical survival (i.e., an infant who left the hospital able to take oral feedings well) was No. 9 in 1950; the next was No. 25, seven years later. Of the 18 cases diagnosed in the third period covering the last five years (1957 through 1961), 17 had a primary anastomosis and 12 (70%) survived. The average age at admission was still excessive, 2.3 days.

In this small series, contrary to the usual reported sex incidence, girls outnumbered boys 24 to 18. It is of interest but of questionable significance that 37 were recorded as white, three were Indian, and only two were Negro. Hospital admission racial incidence details are not adequate for comparison. The scattered intrastate distribution of birth places is in keeping with the statewide referral pattern of the University Medical Center patient clientele. If an average quoted incidence of one case per 2,500 live births 8 is applied to Oklahoma's current annual live birth rate (50,000-55,000), it is suggested that even with a recent increase in referrals, we currently treat only about one-sixth of the Oklahoma cases. Brief communication with the other large hospitals in the area suggests that probably half of the state's cases may still die unrecognized or with suboptimal, palliative, or no treatment.

Historical and obstetrical information details was scanty and unrevealing. Only four times was polyhydramnios recorded although excessive amniotic fluid may be demonstrated in two-thirds or more of infants with esophageal atresia. 

Fig. 1. Varieties of esophageal anomaly identified by (a) an esophageal atresia, (b) an esophageal atresia with a tracheo-esophageal fistula, (c) an esophageal atresia with a tracheo-esophageal fistula and tracheo-esophageal fistula.
Types of Esophageal Anomaly

Figure 1 includes the varieties of esophageal atresia encountered in this small series. Isolated stenosis and isolated tracheo-esophageal fistula are not included. It seems preferable to avoid the confusion of the oft-quoted classification labels (such as Vogt,²² Ladd,¹²) and to identify by anatomical description. The common or usual type will be used to indicate the predominant pattern of blind proximal esophageal pouch with fistula between distal esophagus and trachea or main bronchus. Figure 2 indicates several of the less fundamental variations of local anatomy from this series which seem to possess surgical significance or interest.

Diagnosis

It has been noted in several large series that only about one-fourth of operative survivors come from the majority group operated on after the first 48 hours of life. Average age at admission for the entire series here was three days but the first few cases averaged six days when admitted and nearly eight days before diagnosis was established. A marked improvement in the 1948-56 period is reflected by an average admission age of 2.8 days with a decreased hospital delay in diagnosis. We consider the average age of 2.3 days during the recent five-year period to be excessive even though the referring physician had more often approached the correct diagnosis prior to consultation. In fact, the improved accuracy of recent referral diagnoses may suggest an ex-

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Fig. 1. Varieties of esophageal anomalies usually described. Standard classification labels are avoided. The anomaly is identified by its anatomical characteristics. A, (3 cases) isolated atresia; length of lower segment variable but nearly always short. Incidence probably should be 10%, B, (none) atresia with proximal fistula; very rare. C, (37 cases) atresia with distal fistula, the usual or common type; length of lower segment, site and size of fistula variable, but gap is very rarely insurmountable. D, (1 case) atresia with proximal and distal fistulae; rare. E, isolated fistula ("H-fistula") and F, isolated stenosis, not covered in this report.

Fig. 2. Surgically significant variations in local anatomy of the anomalies encountered in this series. A, the common type with upper portion of the lower segment attenuated, joining a narrowed but patent fistula. Trimmed and dilated at operation and sutured under moderate tension after maximum proximal and extensive distal mobilization, there was no anastomotic leak but stricture required dilation. B, usual type, with high proximal pouch (at low cervical level). C, (2 cases) usual type; fistula enters a right main bronchus rather than trachea. No special technical difficulty although distal segment seemed shorter. D, usual type; fistula enters under-surface of carina. E, (several cases) usual type but gap is nearly zero and segments of esophagus are in partial muscular but not luminal continuity. Technically favorable; in one case, posterior wall was maintained intact, diaphragm-like obstruction and indented anterior wall were excited prior to end-to-end suture of about 300° circumference. F, usual type with large diameter fistula. Technical advantage was overshadowed by severity of associated pneumonitis and abdominal distortion. (Martin described an infant who could cry audibly only with gastrostomy tube occluded.) G, (2 cases) usual type plus diaphragmatic stenosis of distal esophagus. Calibration with catheter or blunt curved hemostat and limited dilation prior to anastomosis. Postoperative esophagogram and swallowing function will guide further therapy. H, atresia with both proximal and distal fistulae. Former is attached to esophagus above the end of pouch so can be treacherous. One example here suspected but unverified by x-ray, operation, or autopsy. I, fistula at low cervical level of tight esophageal stenosis. Anastomosis and division of fistula in neck after thoracotomy. Only case here expired, no autopsy.
Table 1. Clues to Early Diagnosis of Esophageal Atresia

<table>
<thead>
<tr>
<th>Clue</th>
<th>Description</th>
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<tbody>
<tr>
<td>Prematurity</td>
<td>Rapid distention (postnatal)</td>
</tr>
<tr>
<td>(1 in 3)</td>
<td>Gasless abdomen</td>
</tr>
<tr>
<td>Polyhydramnios (2 in 3)</td>
<td>Green sputum</td>
</tr>
<tr>
<td>Other anomalies (1 in 3)</td>
<td>R.U.L. atelectasis</td>
</tr>
<tr>
<td>Umbilical artery absent</td>
<td>Pouch air bubble</td>
</tr>
<tr>
<td>Resuscitation (need for)</td>
<td>Catheter block (or coil)</td>
</tr>
<tr>
<td>Secretions (frothy excessive)</td>
<td>Sclerema neonatorum (not so early)</td>
</tr>
</tbody>
</table>

...isions in the delivery room and nursery and would follow a provisional diagnosis based on alimentary intubation and minimal roentgenography by immediate telephone referral and transportation to a pediatric surgical center rather than by time-consuming, unnecessary and often dangerous contrast X-ray study.16

Some of the common pitfalls observed to cause unwarranted diagnostic delay are related to the items listed in Table 1. Two of the most frequent have been a tendency to blame early choking spells and respiratory distress on a difficult labor and the failure to exclude deception by a catheter coiled in a blind esophageal pouch before considering a gastric aspiration to be negative. Only the curled tube will give an audible air puff localized over the neck. Such errors are not unique to this series and deserve more emphasis in the instruction and supervision of the obstetrical or pediatric practitioner. The
diagnosis of esophageal atresia is simple if the attending physician thinks of it. Most of the clues or maneuvers listed in Table 1 are evident so promptly that we believe that any infant whose obstruction is not suspected until after he has difficulty with his first feeding has already seen suboptimal medical care.

Although the prompt removal of 0.5 cc of lipiodol or aqueous contrast medium from the pouch catheter can be safely performed, it may be that the only way to discourage fruitless, excessive prerereferal x-rays (some still use barium!) is to advocate abandonment of the use of any radio-opaque material in diagnostic procedures in this condition.10 Simple films showing a coiled or blocked catheter in the cervico-thoracic region and presence of abdominal gas confirm the diagnosis of the usual anomalous pattern but may miss the very rare upper fistula, especially if its neck is proximal to the blind pouch and thus less promptly fatal.

The newborn with clinical evidence of esophageal obstruction plus a gasless abdomen by x-ray will prove with very rare exceptions to have isolated esophageal atresia, (i.e. no tracheal fistula). In fully 90 per cent of cases with isolated atresia, the distal esophageal segment has proved to be hypoplastic and unsatisfactory for primary anastomosis.

Some authors have advocated exploratory thoracotomy for all such infants to rule out a plugged fistula or to anastomose the rare adequate distal esophageal segment.8 Others have proceeded as though the gasless abdomen was sufficient evidence of hypoplastic lower esophageal segment.

We do not consider biphacotomy justified and posterior contrast delineation phageal and possible fistulas.

**Survival I**

The factors usually ex...renal anomaly. This incidence increase if autopsies had more complete. Of died in the hospital, auto on 17, but only two inc...hanced a...There are these six can theoretically amenable to significant anomalies occurring relative frequency) i alimentary, neurologic, at...tions, as has been the exp...large pediatric centers.

**Pneumonia.** The prec...nary disease was difficult recent familiar cases and the...should not be ignored when there was radiography infiltration or at Significant changes in both lungs have generally y The presence of pneun...ctasis predominantly in has been so common as nastic clue. Our infants were respiratory distress a

**Fig. 3A. Rationale for initial gastrostomy. Routes of aspiration contributing to high incidence of atelectasis and pneumonitis in infants with common type of anomaly.**

Refill of highly acidic gastric juice through fistula is considered much more important than overflow and inhalation of saliva and is the basis of our increased use of initial decompressive gastrostomy.
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**Survival Factors**

The factors usually expected to be of major serious prognostic significance were verified in this review: low birthweight, additional congenital anomalies, and pneumonia (including atelectasis and consolidation).

**Prematurity.** Although more than one-third of the infants were classified as premature because of birth weight under 5½ pounds, only one weighed less than four pounds. This incidence of prematurity in the early, middle, and recent periods was zero (0/5), 53 per cent (10/19) and 24 per cent (4/17), respectively.

**Associated Anomalies.** Half of the infants were noted to have at least one other congenital anomaly. This incidence would decrease if autopsies had been more frequent and more complete. Of the 29 infants who died in the hospital, autopsies were performed on 17, but only two included the brain. Although the associated anomalies are considered significant in 10 cases, in only six did they appear to be the actual cause of death. Three of these six can now be considered theoretically amenable to surgical therapy. The significant anomalies occurred (with decreasing relative frequency) in the cardiovascular, alimentary, neurologic, and genito-urinary systems, as has been the experience reported from large pediatric centers.

**Pneumonia.** The precise extent of pulmonary disease was difficult to assess even in the recent familiar cases and almost impossible in the earlier cases, and the distinction between collapse and consolidation is often not clear. Pneumonia has been considered moderate when there was radiographic or clinical localization of infiltrate or atelectasis to one lobe. Significant changes in both lungs in all of that one lung have generally justified a severe label. The presence of pneumonitis or lobular atelectasis predominantly in the upper lobe has been so common as to constitute a diagnostic clue. Our infants who appeared in severe respiratory distress at the time of surgical consultation often proved to be late referrals with marked abdominal distention or congenital heart disease or with a history of persistent, traumatic endoscopic or radiologic diagnostic efforts, or of excessive prior parenteral fluid administration. It is considered highly significant that the infants with no fistula, both here and elsewhere, presented with relatively little pulmonary difficulty even though characteristically diagnosed later than the usual type.

There has been much difference of opinion as to the major cause of the ubiquitous respiratory complications (featuring pneumonitis) in patients before and after operation for atresia of the esophagus: gastro-esophageal regurgitation via the fistula or aspiration of excessive nasopharyngeal secretions. It has been our opinion for several years that the chief cause of preoperative atelectasis and pneu-
monitis is reflux of highly acidic gastric juice through the distal esophagus and fistula into the tracheobronchial tree. The newborn patient is undoubtedly exposed to the danger of overflow aspiration of nasopharyngeal mucus and ingested feeding during attempted swallowing, but we suspect that even the oral feeding causes its major harm by prompting a coughing, choking episode which triggers gastro-esophageal reflux aspiration by spasmodic compression of a distended stomach (Fig. 3).

It is fairly well accepted that aspiration pneumonitis in adults has a primarily chemical, rather than bacterial etiology, because of its distinctive clinical and pathological syndrome. It has been shown, at least in adults and experimental animals, that the lesions can be caused by food particles and by liquid vomitus having a pH less than 2.5. Gastric enzymes, bile and liquids of pH greater than 2.5 proved relatively innocuous. Pathologic descriptions from fatal cases involving primarily acid vomiting emphasized extensive, hemorrhagic consolidation and microscopically extensive peribronchial infiltration, destruction of lung parenchyma, alveoli filled with hyaline exudate and red cells, and irregular sloughing of bronchial epithelium. Food particles evoke a predominantly foreign body reaction in lung tissue quite unlike the respiratory difficulties in our neonatal patients. The pathologic similarity of the acid pneumonitis and that encountered in the infant with tracheoesophageal fistula is considered significant.

The stomach contents of the normal newborn and premature have been shown to be so nearly neutral in reaction at birth that their acidity seems unlikely to be an irritant factor should they reach the lungs in late prenatal life. The gastric acidity then increases very rapidly so that within 4.5 hours, nearly all samples show a pH of between 3 and 1, averaging titration equivalent of a free acidity of 30 to 40 ml. N/10 HCl/100 ml, although range was from zero (achlorhydria) to above 70 ml. The early alimentary reaction was found most acid in the gastric fundus, less in the pylorus and nearly neutral in the mouth and esophagus. (In the two cases in which we have sampled the gastric content at the time of gastrostomy, the pH values were 2.3 and 1.2). This prompt but unexplained postnatal hyperacidity tends to decline after the second day, falling to near zero by the eighth day, then rising by the third week to the generally low level of infancy. Smith comments that pepsin and cathepsin are both present at birth and, partially activated by the sufficient HCl (optimum digestive pH is 2.0, still active at 4.0), could prove irritating in the respiratory tract. Acid secretion may increase during periods of hypoventilation and hypercarbia; in these infants it goes without the dilution of swallowed saliva but can still be buffered to some extent by mucus and regurgitated duodenal content.

Over the past several years, a gastrostomy has been performed in nearly all cases usually at the closure of the thoracotomy incision or one to two days later. For over 10 years the senior author has favored gastrostomy as a separate initial procedure under local anesthesia on the very ill or severely distressed infant. The decompressive efficiency of a large tube left open without suction has been impressive, adding to our list of causes of gas-deficient abdominal x-ray pattern in the infant. The apparent therapeutic value in treatment of atelectasis and pneumonitis, which we had considered limited to prevention of further reflux prompted its more liberal use as an initial step. The rather striking improvement in clinical and radiologic pulmonary changes (Fig. 4) noted in several cases is attributed to the increased efficiency and safety of the patient's own aerative efforts plus our gentle pharyngeal suction and stimulation with the gastrostomy as a pop-off valve avoiding further reflux aspiration. Also gastrostomy can be done promptly at any hour and the definitive operation deferred until the infant and operating team are in optimum condition (Table 2). If the critically ill patient is slow to improve, the thoracotomy can be further delayed, even with the fistula still open, provided gastrostomy feeding is avoided and the blind pouch emptied at frequent (15 to 30 min.) intervals while the other systems are being treated intensively, Martin has used this approach with remarkable results; some of his patients have apparently benefited by gastrostomy decompression periods without fistula ligation as long as 21 days prior to successful primary anastomosis and repair of fistula.

Preoperative Management

Our currently preferred management reflects several during the past few years our approach to preoperative general concepts of treatment consideration of supportive care: 1) Primary anatomic emergency middle-of-the-nose Reflux of gastric secretions agastric fistula is the operative pulmonary common transpleural esophagus desirable, but staging in cesions will salvage more in ageal substitution is neccessary retrosternal colonic transplastic

General aims of preoperative care are: 1) treatment of existing of further pneumonia and establishing optimum metabolic infant operative candidate; the best surgical team available steps found beneficial in this period are listed:

1. Incubator (Isolette)
2. High humidity
3. Oxygen—usually 3.0 to 4.0 per cent permitted if the ilization or oxygen transport
4. Position—head of body 25 to 30 degrees to minimize
Preoperative Management

Our currently preferred plan of operative management reflects several changes in thinking during the past few years and has modified our approach to preoperative care. Several general concepts of therapy are basic to consideration of supportive as well as operative care: 1) Primary anastomosis is not an emergency middle-of-the-night procedure. 2) Reflux of gastric secretions through the esophagotracheal fistula is the major cause of preoperative pulmonary complications. 3) Primary transternal esophageal anastomosis is desirable, but staging in certain high risk situations will salvage more infants. 4) If esophageal substitution is necessary, the delayed retrosternal colonic transplant is preferred.

General aims of preoperative management are: 1) treatment of existing and prevention of further pneumonia and atelectasis; 2) establishing optimum metabolic status for the infant operative candidate; and 3) providing the best surgical team available. Some of the steps found beneficial in the preoperative period are listed:

1. Incubator (Isolette)
2. High humidity
3. Oxygen—usually 3.0 to 5.0 L/min., over 40 per cent permitted if there is impaired ventilation or oxygen transport.
4. Position—head of body platform elevated 25 to 30 degrees to minimize further tracheobronchial aspiration by both decreasing gastroesophageal reflux and increasing the collecting efficiency of proximal pouch aspiration, as well as to decrease impairment by distention of the predominantly diaphragmatic respirations. Optimum position of head and neck will vary with each patient, governed by ease of respirations. Micrognathic infants may need lateral or prone position to prevent glossectopic obstruction; one of our two with this complicating feature should probably have had anterior glossectomy performed.18
5. Constant nursing attention
6. Antibiotics—penicillin and streptomycin initially until report of admission nasopharyngeal culture and sensitivities is available.
7. Vitamin K (2.0 mg./day i.m. × 3). Vitamin C only if parental fluids.
8. Serial body weights
9. Body temperature control—36.1° to 36.6°C. range, requires monitoring and special care during x-ray studies, i.v. therapy, etc.
10. Parenteral fluids—usually none prior to operative therapy unless there is clear evidence of moderate or severe dehydration by clinical and/or weight loss observation (usually only in full-term infant over two days old), fever, and perhaps severe "physiologic" jaundice with serum bilirubin up to 12 to 15 Gm. % range. Only 5.0 per cent D/W, no salt.
11. Aspiration of the blind proximal esophageal pouch—inwelling transnasal small caliber (No. 5 or 8 Fr.) soft plastic catheter with several holes limited to 1.5 cm. end space, secured by tape and with intermittent suction applied by low
### Table 2. Outline of Current Operative Management Plan for Infant with Esophageal Atresia

<table>
<thead>
<tr>
<th>Anatomy</th>
<th>Patient</th>
<th>Sequence</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Usual (blind pouch, abdominal</td>
<td>Term, Healthy (over 5½ lbs,</td>
<td>(a) Immediate gastrostomy</td>
</tr>
<tr>
<td>gas in 1–6)</td>
<td>without severe pneumonitis or</td>
<td>(b) Anastomosis 12–36 hrs. later</td>
</tr>
<tr>
<td></td>
<td>anomalies)</td>
<td></td>
</tr>
<tr>
<td>2. Usual</td>
<td>Term, unhealthy (severe</td>
<td>(a) Gastrostomy (decompression only)</td>
</tr>
<tr>
<td></td>
<td>pneumonitis or major anomaly)</td>
<td>(b) Intensive supportive care</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(c) Emergency conservative (staging) approach to other anomaly, (e.g.,</td>
</tr>
<tr>
<td></td>
<td></td>
<td>colostomy for anal atresia)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(d) Anastomosis when improves progressively (6 lbs.)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(e) If progress unsatisfactory, interruption of fistula extra pleurally,</td>
</tr>
<tr>
<td></td>
<td></td>
<td>then</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(f) Defer anastomosis until gains to 6 lbs.</td>
</tr>
<tr>
<td>3. Usual</td>
<td>Big premature, healthy</td>
<td>(a) Gastrostomy (decompression)</td>
</tr>
<tr>
<td></td>
<td>(4½–5½ lbs, no severe pneumo-</td>
<td>(b) Fistula interruption next day</td>
</tr>
<tr>
<td></td>
<td>nitis or major anomaly)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>or combination of both moderate</td>
<td></td>
</tr>
<tr>
<td>4. Usual</td>
<td>Big premature, unhealthy</td>
<td>(a) Gastrostomy (decompression)</td>
</tr>
<tr>
<td></td>
<td>(4½–5½ lbs, (severe pneumo-</td>
<td>(b) Fistula interruption next day</td>
</tr>
<tr>
<td></td>
<td>nitis or major anomaly) or</td>
<td></td>
</tr>
<tr>
<td></td>
<td>combination of both moderate)</td>
<td></td>
</tr>
<tr>
<td>5. Usual</td>
<td>Small premature, healthy</td>
<td>Embark on same as (3) with longer (a)―(b) interval therapy, and if step</td>
</tr>
<tr>
<td></td>
<td>(under 4½ lbs,)</td>
<td>(c) progress lags, consider shift to late stage and probable</td>
</tr>
<tr>
<td>6. Usual</td>
<td>Small premature, unhealthy</td>
<td>substitution―with cervical esophagostomy.</td>
</tr>
<tr>
<td></td>
<td>(under 4½ lbs,)</td>
<td>Same as (4)</td>
</tr>
<tr>
<td>7. Usual, with T.E.F. occluded</td>
<td>All types</td>
<td>(a) Gastrostomy</td>
</tr>
<tr>
<td>(blind pouch, gasless abdomen</td>
<td></td>
<td>(b) X-ray delineation of distal esophagus → decision</td>
</tr>
<tr>
<td>in 7–9)</td>
<td></td>
<td>(c) Gastrostomy feeding</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(d) Pouch suction</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(e) 1st anastomosis at 6 lbs.</td>
</tr>
<tr>
<td>8. Distal esophagus adequate,</td>
<td>All types</td>
<td>Same as (7)</td>
</tr>
<tr>
<td>or T. E. F. occlusion evidence</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9. Distal segment hypoplastic,</td>
<td>All types</td>
<td>(a, b) Same as (7)</td>
</tr>
<tr>
<td>no fistula</td>
<td></td>
<td>(c) Gastrostomy feeding</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(d) Cervical esophagostomy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(e) Subternal right colon transplant at age 1½–2 yrs.</td>
</tr>
</tbody>
</table>

The immediate postoperative care of the patient varied according to the needs of the individual case. The immediate postoperative management plan for infants with esophageal atresia was designed to accommodate the variable requirements of each patient. The plan was divided into five categories based on the patient's anatomy, with each category having specific management criteria for patient care. The sequence of treatment included immediate gastrostomy, followed by various surgical interventions depending on the patient's condition. The plan aimed to provide a structured approach to the management of these infants, acknowledging the variability in their needs and ensuring a systematic approach to their care.
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pressure Gomco apparatus or nurse’s syringe every 15 minutes.
12. Blood—crossmatch 125 cc. for thoracotomy (operative loss measurements have ranged 25 to 60 cc.)
13. Cortisone see later discussions of resuscitation, aspiration, pneumo- nitis and selerema.
14. Digitalis

The immediate availability of a surgeon trained and equipped to manage the resuscitative needs that might appear at any time is a factor too often taken for granted, but its contribution to the improvement of survival rate in this series is inescapable.

Operative Management, Results

The treatment offered here prior to 1948 should not really be called surgical since one infant died without operation and three of the four gastrostomies were palliative. During the nine years from 1948 to 1957 a definitive surgical plan was evident throughout, though during the first four years one infant died in 36 hours without operation, five were considered at exploration to be impossible to anastomose and only three had a primary anastomosis. The lone surgical survivor prior to 1957 was a slightly premature two-day-old infant treated in 1950 by gastrostomy, extrapleural division of the fistula and end-to-end esophageal anastomosis. Her early course was stormy, an anastomotic stricture required dilation at age four months, and two months later she survived operative treatment for small bowel obstruction due to adhesions at the gastrostomy site.

The extrapleural approach for anastomosis was attempted a total of five times with but one anastomosis completed and no survivors. It was used on two other occasions when only an exploration for possible occluded fistula was planned in an infant whose x-rays showed a gasless abdomen. The transpleural exposure for anastomosis has been used in all cases since early 1952. Three patients had cervical esophagostomy with the gastrostomy, twice when the surgeon, after fistula closure, believed he could not achieve primary anastomosis and once when no fistula was found.

Although the sole survivor of the 19-patient middle period has been lost to follow up since age seven months, there is evidence of progress in the records of three infants who succumbed late and begrudgingly. One, in 1952, who had no fistula or distal segment was maintained with cervical esophagostomy and gastrostomy until her death from pneumonia and diarrhea at age 12 months. Two others in 1956 succumbed to pulmonary infection while still hospitalized, seven weeks and five months, postoperatively. The first had survived exchange transfusion for erythroblastosis, and the latter, a mongoloid with part of his stomach pulled into the chest because of a high-lying anastomosis, had clinical and autopsy findings of fibrocystic disease of the pancreas. The death of two infants was probably contributed to by failure of the surgeons to fully appreciate or treat an associated congenital stenosis of the diaphragmatic type located in the distal esophagus (Fig. 2G).

During the past five years, all 18 cases diagnosed were of the common type. One premature infant admitted at age three days in a moribund state with clinical cyanotic heart disease and advanced selerema neonatorum, expired before gastrostomy was performed. Of the remaining 17 cases, all of whom had transpleural primary anastomoses, 12 survived hospitalization and 10 are known to be alive and well today. One mongoloid infant, operated on in 1957, had hydrocephalus diagnosed at age six months and died at home at age 13 months of unknown cause. Her recorded hospital and follow-up course suggests that more aggressive (though perhaps philosophically ill-advised) dilation therapy for what was almost certainly an early anastomotic stricture might have avoided malnutrition and even demise. Another patient was discharged to his home in a remote corner of the state weighing eight pounds at age five weeks with clinical and roentgenographic evidence of good swallowing function. We subsequently learned that he expired at home one month later after a one day illness suggesting acute respiratory infection. No postmortem was done so a stricture with aspiration or even a recurrent fistula cannot be ruled out.

Of the five postoperative hospital deaths, three were premature infants and three are attributed primarily to associated major anomalies although only one, an infant with persistent truncus arteriosus and cor bilocular, can be considered theoretically not amenable to
available therapy. Another infant with persistent postoperative cyanosis developed sclerema before death on the fourth postoperative day and at autopsy was found to have marked atelectasis of the right lung, moderate ascites, preductile aortic coarctation, large ductus arteriosus and a ventricular septal defect with thickened right auricular and ventricular walls. Perhaps early gastrostomy, improved right pulmonary ventilation and digitalization might have helped avoid sclerema and supported the infant during the period of excessive cardiopulmonary burden. Another premature infant might have survived had the obstructive features of his associated ileac atresia not been obscured by the decompressive efficiency of a gastrostomy proximal to a closed-loop, strangulated volvulus of the blind segment of ileum. Although his list of associated major anomalies requiring urgent neonatal operation is prognostically ominous, we prefer to consider this a preventable fatality.

One early postoperative death by respiratory failure is attributed partly to a technical error in hemostasis resulting in an obstructing localized retrotracheal hematoma. Initial gastrostomy followed by intensive prolonged supportive efforts might have allowed this critically ill infant to escape operation during the vulnerable period after his severe respiratory distress present at admission. He might then have tolerated the stress of a second thoracotomy for clot evacuation.

The last was a premature infant who had been subjected to a barium swallow before her admission here at age four days with bilateral diffuse pneumonia. An exceedingly stormy postoperative course requiring early and repeated resuscitation by laryngoscopy aspiration and complicated by difficulties associated with micrognathia and glossophtisis seemed to have been navigated successfully without the planned gastrostomy, but during her third postoperative week pulmonary infection spread and proved fatal (culture pseudomonas). Review of her course and the finding of a ventricular septal defect plus extensive confluent bronchopneumonia with evidence of aspiration suggest that salvage might have been possible with the help of gastrostomy, digitalization, and perhaps anterior glossopexy.

Operative Management, Technics

Gastrostomy. A simple Stamm gastrostomy performed under local anesthesia has been the only method used in this series. A detailed description of technic is not in order here. Meticulous technic and attention to details of wound care apparently have prevented the difficulties with hemorrhage, wound infection and leakage described by some authors. Two or usually three purse-string sutures of fineatraumatic suture are used to invert a No. 16 Fr. mushroom catheter with the roof of the tip excised.

Fistula Interruption (separate stage). With the gastrostomy available to avoid gastric overdistention, endotracheal general anesthesia is preferred. An extrapleural approach through the bed of the fourth or fifth rib posteriorly provides adequate exposure for division and suture of both ends and suture

fixation of the esophageal bral fascia. A suction dro the retropleural space for feeding and protection will permit weight gain pulmonary status to just to-and-fro anastomosis by 8 reaches six pounds.

Anastomosis. The sin here has usually included approach to the mediastinum right intercostal space (F principles stressed are:
1. Early division and suture.
2. Extensive mobilization of upper pouch to minimize distal segment to permit a tensionless anastomosis.
3. Usually two-layer or nonanastomosis of the layers.
4. Use of 5-0 or 6-0 interrupted sutures avoiding intramural bleeding.
5. Maximum gentleness of esophageal ends.
6. Calibration and polishing of the distal segment to exclude gross distortion.
7. Intermittent complete closure of upper and middle lobes.
8. Meticulous attention to
Fig. 5. Transpleural approach for correction of the common type anomaly. The excessive retraction of the vagus trunk is at risk of trauma.

Fig. 6. Step-wise division and suture closure of the fistula is the first intrathoracic maneuver in the preferred definitive operation. If the first layer is continuous, it is usually interlocking, of 6-0 arterial silk and catches a very narrow cuff of fistula. Second row buttresses with adventitia, avoiding excess cuff inversion. The obliquity pictured here has usually been lost when the tip of the lower segment has been trimmed back to healthier substance, so that transverse excision of the tip of the pouch usually provides the proximal opening for anastomosis.

Fig. 7. Technic of telescopic 2-layer anastomosis, originally described by Haight and used in many of our earlier cases, takes advantage of hypertrophy, dilation, and vascularity of blind proximal segment. Recently we have used 6-0 arterial silk in a 2-layer end-to-end technic.

Management, Technics

A simple Stamm gastrostomy or local anesthesia has been used in this series. A dozen of technics is not in order; technic and attention to detail apparently have prevented with hemorrhage, wound in-kage described by some usually three purse-string subcutaneous suture are used to include mushroom catheter with the cession (separate stage). With available to avoid gastric endotracheal and general anesthesia. An extrapleural approach of the fourth or fifth rib provides adequate exposure for incision of both ends and suture fixation of the esophageal end to the prevertebral fascia. A suction drainage tube is left in the retropleural space for a few days. Better feeding and protection from reflux usually will permit weight gain and improvement in pulmonary status to justify telescopic end-to-end anastomosis by the time the infant reaches six pounds.

Anastomosis. The single stage operation here has usually included a transpleural approach to the mediastinum via the fourth right intercostal space (Fig. 5). Some of the principles stressed are:

1. Early division and suture of the fistula (Fig. 6).
2. Extensive mobilization dissection of the upper pouch to minimize that needed about the distal segment to permit a tension-free anastomosis.
3. Usually two-layer end-to-end or Haight telescopic anastomosis of the esophagus (Fig. 7).
4. Use of 5-0 or 6-0 interrupted arterial silk sutures avoiding intramural knots.
5. Maximum gentleness and minimum handling of esophageal ends being sutured.
6. Calibration and possibly slight dilation of the distal segment to exclude the rare but treacherous distal stenosis (encountered twice in this series).
7. Intermittent complete expansion of the right upper and middle lobes or other atelectatic areas.
8. Meticulous attention to details of supportive care such as continuous monitoring and control of infant’s body temperature and the continuous measurement (sponge-weighing) and replacement of operative blood loss.

Substitution. We have never encountered an example of the usual type anomaly in which the mobilized esophageal segments proved of inadequate length for primary anastomosis but would in such a circumstance close the chest (fistula already divided) after closing the distal end and bring the proximal segment out above the left clavicle as a cervical esphagostomy. The favored reconstruction features retrosternal transplantation of a segment of right colon to connect the cervical esophagus with the lesser gastric curvature plus a drainage (Heineke-Mikulicz) pyloroplasty. The gastrostomy is maintained for intestinal decompression and feeding during the early postoperative period (Fig. 8).

Postoperative Considerations

The postoperative course following anastomosis for esophageal atresia is variable but, in the individual case fairly predictable. It should be relatively smooth in the vigorous full-term infant without other significant defects if early diagnosis led to prompt surgical correction before the onset of pneumonitis. More commonly the recovery is complicated and for success will require constant vigilance with frequent and critical evaluation by the surgical and nursing staffs. To the preoperative picture of pneumonitis and difficulty with
secretions will be added the factors of metabolic and circulatory imbalance and fatigue. The immediate recovery phase is the time of greatest mortality when some of the weaker infants, supported satisfactorily during the operation, eventually fail when on their own, especially when the residual depression of operative hypothermia is added. The surgeon must accept as fact that the infant's condition, from apparently excellent, may abruptly deteriorate within the hour, usually due to accumulation of secretions in the tracheobronchial tree. A too common sequence is that of the hypoxic, acidic prematurity who develops cardiovascular collapse and respiratory insufficiency and is resuscitated several times only to finally die when spontaneous respirations cease. A dedicated and skilled surgical and anesthetic staff can occasionally salvage one of these by continuous moment-to-moment observation and especially respiratory support.

There has been a recent increase in the number of postoperative complications recorded in our series with two major explanations apparent: 1) Infants are now surviving long enough to develop a wider variety of complications; and 2) Postoperative care and records are attended to more meticulously. It is often not clear whether a specific complication should be attributed to the disease or to the operation, but we have tabulated the major unfavorable developments of the operative and postoperative periods (Table 3). Those marked with an asterisk were encountered in the past five years. Preferred anticompliation measures are indicated along with several items of supportive care that possess preventive value.

A few complications deserve additional comment. Sclerema neonatorum is a rather mysterious condition of obscure etiology and grave prognosis. It shows a predilection for premature or term infants debilitated by dehydration, shock, infection, intracranial hemorrhage, or congenital heart disease. Classically it is characterized by the fairly sudden development of a nonbrawny induration of the skin and subcutaneous tissues, usually beginning in the lower extremities and ascending to involve the trunk and arms. Most cases have ended fatally with the role of the sclerema not always clear; e.g., limitation of respiratory excursion or perhaps just progression of the associated and usually serious underlying illness. An attractive but unproved theory not inconsistent with the characteristically unenlightening histologic studies has for many years implicated an alleged solidification vulnerability of neonatal subcutaneous fat due to the higher melting point associated with a relatively low oleic acid content (low iodine number and percentage unsaturation). Several reported survivals have been credited to cortisone therapy and raised environmental temperatures but the causal evidence is flimsy.

In our experience here and elsewhere, by far the most common surgical lesion complicated by sclerema has been the usual type of esophageal atresia with tracheo-esophageal fistula. Perhaps this is not surprising since such
patients frequently concomitantly have several of the other conditions known to be associated if not contributory (prematurity, infection, hypothermia, shock, congenital heart disease, anoxia, etc.). Our records here show six diagnoses of sclerema in 42 cases. Of the eight atresias in 1959, three developed sclerema and one survived with help attributed to several factors: a successful surgical anastomosis, controlled environment, adequate hydration and I.V. cortisone. Various tissue temperatures being investigated and controlled in current cases. For several years we have preferred low-risk plasma rather than blood initially for the newborn who is hypothermic, in shock (especially peripheral circulation with marked hemococoncentration and viscid, perhaps sludged, blood). Low molecular weight dextran may prove of value, perhaps by delaying the fatal suffocative potential of the immobility of the tissues until the primary disease can be controlled.

Stricture. Postoperative stricture at the anastomotic site has been reported with varying frequency and therapeutic approaches. We use swallowing function as the major criterion for deciding the need for postoperative dilation except perhaps in the early postoperative period. Frequently children with an apparently narrow lumen by roentgenography have no functional difficulty. Recently Karkapatt completed that there was disturbed esophageal motor activity in all of 15 postoperative patients undergoing cinefluorographic study and postulated preexisting inherent vagal deficiency.\(^8\) A few similar examinations by our radiologists have not shown motor dysfunction with such consistency. Of 11 patients surviving six months to 4½ years, four have had one or more esophageal dilations and another has required endoscopic removal of a foreign body on two occasions although clinical swallowing was otherwise satisfactory. There is no apparent correlation of anastomotic technic with incidence of stricture.

Skilful gentle dilation of the anastomotic stricture may well be indicated in the early postoperative period in the infant troubled by what seem to be primarily respiratory symptoms. It is especially important for the surgeon to realize this and not relinquish the postoperative care to the pediatrician, few of whom appreciate the full gamut and significance of postoperative complications. Satisfactory emptying of the upper segment pouch resulting from instrumental dilation may possibly prevent development of a recurrent tracheo-esophageal fistula by permitting intraluminal drainage or reentry of a small abscess at the suture line. Other problems that may be eased by timely effective dilation are overflow aspiration, the characteristic brassy cough, and inability to avoid aspiration or choking following a burp.

If a gastrostomy is available, the Tucker rubber bougies tied in series are quite useful and probably the safest method available.\(^4\) If no string is swallowed, under anesthesia and guided by esophagoscopy, a woven filiform can easily and safely be passed through the stricture and fished out of the stomach for attachment of the series of Tucker dilators.

An exceedingly useful easy technic, especially when no gastrostomy is available, is the use of regular (urological) woven silk filiform Phillips bougies with gradually enlarging followers. This method is not adequately appreciated by surgeons and endoscopists although it has been used for many years.\(^4\) The four- to six-week-old full-term infant's anastomosis can usually be safely dilated to No. 18 Fr. Repeat dilation may be needed when food consistency increases at three to six months. At later dates, older children may have obstruction precipitated by incomplete mastication, careless swallowing, or swallowing of foreign objects.

Summary

A review of experience with the treatment of congenital esophageal atresia at the University of Oklahoma Medical Center during the past 22 years reveals a belated but gratifying improvement in results. Prior to 1957 there was only one isolated survival—1950.
During the past five-year period, 12 (70\%) of the 17 operated cases survived.

Major prognosis determinants proved to be those commonly reported by others: pneumonia, associated anomalies, prematurity and the experience of the therapeutic personnel. The latter two probably contributed significantly to the recently improving survival rate at this institution. A logical attempt to reduce mortality must recognize that immaturity, associated anomalies and the type of esophageal defect are unavoidable but not uncontrollable factors.

A staged regimen is advocated for the very small, weak, or ill patient, designed to defer definitive thoracotomy past his vulnerable, high-risk period and yet retain the preferred eventual reconstruction, end-to-end esophageal anastomosis. Reduction of mortality from associated congenital defects of other systems and organs will result from the productive efforts of progressively more trained pediatric and thoracic surgeons as well as from the decreased general operative risk permitted by staging of the esophageal procedures.

Pulmonary complications are a cause of serious difficulty in all stages of management. Education and encouragement of obstetricians and pediatricians can help reduce diagnostic delays and excessive preoperative studies.

Mounting evidence that the dominant cause of aspiration pneumonitis is reflux of acidic gastric secretions via the fistula has led to an increasing use of initial gastrostomy. Repeated observations of the increased effectiveness of tracheobronchial clearing maneuvers in the presence of a gastrostomy vent have prompted us to use immediate initial gastrostomy for all infants with esophageal atresia. The preferred management of the infant with an isolated atresia involves initial esophagostomy and gastrostomy followed later by retrosternal transplantation of a right colon segment.

References


During the past increasingly clear that the ideal vascular pedicle with occlusion of the lumen will control the problem of vascular grafts. The elastic fabric has created

Extensive bleeding during the time of surgery is less common. However, the elasticity of the graft will enable grafts to be used on any of the best means of

Materials

An apparatus was designed for use with respiration fusion rate (Fig. 1). Sigma-motor, which rate of 360 ml/min. tube was inserted into a 1/4-inch (internal diameter used for the re...

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* N.H. Student Research
** Model TM4. This investigation was supported by the National Institute of Health, U.S. Public Health Service Grant.