Objective: To characterize a successful approach to the management of infants with long-gap esophageal atresia (EA) with tracheoesophageal fistula (TEF), significant prematurity with respiratory distress syndrome (RDS), or both, so as to preserve the native esophagus.

Design: A review of the medical records and office charts of a cohort of patients with EA and TEF.

Setting: A tertiary care children’s hospital affiliated with a major university.

Patients: A total of 118 children with EA and TEF admitted from February 1986 through December 1996. All of the patients diagnosed as having EA and TEF during this period were included.

Intervention: Of the 118 infants, 88 received primary repair of EA and TEF within 48 hours of birth. An additional 23 children had the TEF divided and a gastrostomy placed secondary to (1) severe RDS and prematurity (n = 6), (2) long-gap EA (gap length > 4 cm or the upper pouch above the thoracic inlet (n = 10), or (3) associated cardiac defects (n = 7). Delayed primary EA repair was done when the RDS resolved or the gap length was 2 cm or less.

Main Outcome Measures: Successful anastomosis of native esophagus. Comparison of incidence of gastroesophageal reflux, anastomotic complications, or survival between groups undergoing primary or delayed repair.

Results: Primary EA was accomplished in 88 patients. Delayed EA was successfully accomplished in 18 of the 19 surviving patients within 5 months, thereby preserving the native esophagus in all surviving infants. There was no difference in anastomotic complications, gastroesophageal reflux, or survival when the delayed group was compared with those who had a primary repair.

Conclusions: Using delayed EA repair, all children with EA and TEF, regardless of gap length, can have their esophagus preserved. The primary cause of mortality was the association of a severe cardiac anomaly with EA and TEF.

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The ideal management of esophageal atresia (EA) with tracheoesophageal fistula (TEF) includes division of the fistula and primary esophageal repair performed in a single operation during the newborn period. This approach is successful in most patients born with EA and distal TEF today. The premature infant with significant respiratory distress syndrome (RDS) or the newborn with associated congenital anomalies, specifically cardiac lesions, who is difficult to effectively provide support with mechanical ventilation may not tolerate the lung retraction or operative time necessary for complete repair during a single setting. The finding of a long gap between esophageal segments or of a high upper pouch may preclude primary anastomosis or result in an anastomosis under significant tension, which carries an increased risk of anastomotic complications, recurrent TEF, and gastroesophageal reflux (GER). The purpose of this study was to determine the ideal approach to the management of infants with long-gap EA with TEF, significant RDS and prematurity, or both, or associated cardiac anomalies.

RESULTS

The cases of 23 infants who had only TEF division as newborns are summarized in Table 1. Nineteen infants (83%) survived to undergo a delayed esophageal reconstruction. The repair was successfully accomplished in 18 (95%) of 19 patients; 3 patients had a myotomy to com-
At 4 months of age, the infant had a cervical esophagosophageal fistula and gastric repair with gastrostomy emergently. A large distal TEF. This infant underwent division of the esophagus and upper esophageal pouch were then mobilized and a primary end-to-end esophagoesophagostomy was performed using a single layer, interrupted technique.

In the delayed group where primary EA repair was not performed, the proximal end of the distal esophagus was oversewn and then sutured to the prevertebral fascia. A gastrotomy was also created. Postoperatively, these patients had upper pouch nasoesophageal suctioning and were fed via the gastrotomy. Gap length was assessed by fluoroscopy, using a transgastric catheter or probe to identify the uppermost extent of the distal esophagus.

Delayed primary esophageal repair was performed when the proximal pouch was below the thoracic inlet, the RDS had resolved, or both. At the time of delayed EA repair, a transpleural approach was generally used, although a few patients did have a second extrapleural dissection. The mediastinal pleura overlying the distal esophagus was opened, and the esophagus was mobilized. The bulbous, previously oversewn end was excised, and a transgastric catheter or probe to identify the uppermost extent of the distal esophagus.

In the group where primary repair was not accomplished, secondary to (1) severe RDS and prematurity (n = 6), (2) EA with long gap or the upper pouch above the thoracic inlet (n = 10), or (3) associated cardiac anomaly (n = 7).

There were 134 patients in the delayed group; of these patients, 67 (50.0%) had associated cardiac anomalies, including patent ductus arteriosus in 23 (34.3%), ventricular septal defect in 15 (22.4%), atrial septal defect in 15 (19.4%), tetralogy of Fallot in 8 (12.0%), and aortic arch anomalies in 8 (12.0%). Twenty-one patients had 2 or more cardiac lesions, which were severe enough in 7 patients to warrant a delayed EA repair approach.

Patient data collected included birth weight, estimated gestational age, age at first operation, and the presence of associated anomalies. The operative approach, findings, and specific procedure performed were recorded. The patient age and weight at delayed EA repair, the interval between TEF division and EA repair, and delayed operative findings and procedure were also noted. The patients were grouped according to the indication for delay, and data and outcomes were compared.

OPERATIVE MANAGEMENT

Patients admitted with EA and TEF were initially managed with upper pouch suctioning; the patients were positioned with the head elevated slightly. After physical examination, an echocardiogram was done to assess congenital structural defects and the position of the aortic arch.

Regular radiographic images were taken, and an ultrasound was done to assess vertebral or renal anomalies. At initial operation, a right thoracotomy and an extrapleural approach were used. The distal TEF was divided, and the tracheal side was closed with interrupted sutures. The distal esophagus and upper esophageal pouch were then mobilized and a primary end-to-end esophagoesophagostomy was performed using a single layer, interrupted technique.

In the delayed group where primary EA repair was not performed, the proximal end of the distal esophagus was oversewn and then sutured to the prevertebral fascia. A gastrotomy was also created. Postoperatively, these patients had upper pouch nasoesophageal suctioning and were fed via the gastrotomy. Gap length was assessed by fluoroscopy, using a transgastric catheter or probe to identify the uppermost extent of the distal esophagus.

Delayed primary esophageal repair was performed when the proximal pouch was below the thoracic inlet, the RDS had resolved, or both. At the time of delayed EA repair, a transpleural approach was generally used, although a few patients did have a second extrapleural dissection. The mediastinal pleura overlying the distal esophagus was opened, and the esophagus was mobilized. The bulbous, previously oversewn end was excised, and a primary end-to-end esophageal anastomosis was performed using a single layer, interrupted technique. A contrast study was obtained at 1 week postoperatively to evaluate the anastomosis for narrowing or leakage. Oral diet was started and advanced as tolerated. The gastrotomy was used to maintain adequate energy intake.

OUTCOME

Patient survival up to and after delayed EA repair was calculated. The incidence of anastomotic complications, recurrent TEF, and clinically significant GER, as well as the need for fundoplication, were recorded. Anastomotic stricture was defined as that requiring 1 or more dilatations or another operation. Anastomotic leakage was defined as either a subclinical finding noted on the results of a radiograph of the digestive tract using barium sulfate as the contrast medium or clinical anastomotic leakage. Patient survival to discharge and tolerance of oral nutrition were recorded. In follow-up, the time to full oral diet was calculated. These outcome parameters for patients who had delayed EA repair were compared between groups and with those patients who had primary EA and TEF repair during the newborn period.

STATISTICAL ANALYSIS

Categorical data were compared using χ² analysis, and continuous data were compared using the Student t test. Statistical significance was achieved at P < .05.

Overall, 107 (90.7%) of 118 patients with type C (EA with distal TEF) (90.7%) survived. Ten of 11 of the non-survivors had severe associated cardiac anomalies, which contributed to their deaths (Table 2). Multiple cardiac defects (≥3 defects) were more common in the delayed repair group (17.4% vs 2.3%) (P < .01). Ventricular septal defect (21.7%) and tetralogy of Fallot (13%) occurred more frequently in the delayed repair group, al-
though this was not statistically significant. All 18 patients who had successful delayed EA repair survived.

In the 18 patients who had delayed EA repair, there was no difference in anastomotic stricture (16.6% vs 12.0%), leak (11.0% vs 2.3%), recurrent TEF (5.5% vs 2.3%), GER (22.2% vs 14.7%), or need for fundoplication (11.1% vs 2.3%) as compared with primary repair ($P_{.35}$). Postoperative complications and outcome parameters are listed in Table 3. At discharge, 6 patients were tolerating full oral diet and 8 were receiving a combination of oral diet and gastrostomy nutrition, which was advanced to full oral diet within 4 months. Five patients was prescribed gastrostomy nutrition at discharge, and 4 were converted to full oral diet within 6 months; the 1 remaining child had frequent aspiration with poor swallowing coordination and continues to receive a combination of oral diet and gastrostomy nutrition at 7 months following discharge.

Since the first successful repair of EA with tracheoesophageal fistula in 1941, the outcome has steadily improved.$^{1,5}$ Advances in neonatal care, earlier recognition and management of associated congenital anomalies, improvements in neonatal anesthesia, and refinement of surgical technique are principally responsible for the improved outcomes reported. The classification system of Waterston et al,$^4$ which defined risk categories for patients with EA and TEF based on birth weight, gestational age, pulmonary status, and the presence of associated anomalies, was critical in identifying high-risk infants and their associated poor outcome. Surgical management differed according to patient classification. In 1962, Holder et al$^6$ reported improved survival in high-risk premature infants using a staged operative approach. A gastrostomy was initially placed, which then allowed time for resolution of pulmonary problems, followed by a delayed thoracotomy with division of TEF and EA repair. This strategy was widely used, with several other centers$^7,8$ reporting increasing success, including our own institution.$^9$ As neonatal care continued to improve and more effective ventilators became available, many of the criteria outlined by Waterston et al became less significant and surgical management became based on the clinical status of the infant.$^6$ Pohlson et al$^{10}$ re-

### Table 1. Characteristics of Patients Who Had Delayed Esophageal Atresia (EA) Repair*

<table>
<thead>
<tr>
<th></th>
<th>RDS (n = 6)</th>
<th>Long-Gap EA (n = 10)</th>
<th>Severe Cardiac Lesions (n = 7)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Estimated gestational age, wk</td>
<td>32.8 ± 3.9</td>
<td>34.9 ± 3.4</td>
<td>34.4 ± 3.1</td>
</tr>
<tr>
<td>Birth weight, kg</td>
<td>1.94 ± 0.72</td>
<td>2.34 ± 0.79</td>
<td>1.97 ± 0.88</td>
</tr>
<tr>
<td>Weight at EA repair, kg</td>
<td>2.74 ± 0.5</td>
<td>4.09 ± 1.35</td>
<td>3.36 ± 1.52</td>
</tr>
<tr>
<td>EA repaired successfully, No.</td>
<td>5</td>
<td>10</td>
<td>3</td>
</tr>
<tr>
<td>TEF-EA interval, d†</td>
<td>53.8 ± 22.1</td>
<td>89.8 ± 35.4†</td>
<td>59.2 ± 33.2</td>
</tr>
</tbody>
</table>

*Data are given as mean ± SD unless otherwise indicated. There was no significant difference in estimated gestational age, birth weight, or weight at delayed EA repair.

†The interval between tracheoesophageal fistula (TEF) division and EA repair was longer in patients with a long gap compared with premature infants with respiratory distress syndrome (RDS) ($P_{.03}$).

### Table 2. Patient Characteristics of Nonsurvivors*

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Initial Procedure</th>
<th>Time Until Death, d</th>
<th>EGA, wk</th>
<th>Birth Weight, kg</th>
<th>Associated Defects</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>No surgery</td>
<td>1</td>
<td>28</td>
<td>1.0</td>
<td>Ventricular septal defect, R arch</td>
</tr>
<tr>
<td>2</td>
<td>No surgery</td>
<td>2</td>
<td>34</td>
<td>1.9</td>
<td>Transposition of great arteries, dextrocardia</td>
</tr>
<tr>
<td>3</td>
<td>No surgery</td>
<td>1</td>
<td>36</td>
<td>3.2</td>
<td>Hypoplastic left heart syndrome</td>
</tr>
<tr>
<td>4</td>
<td>Primary EA repair</td>
<td>266</td>
<td>38</td>
<td>3.5</td>
<td>VATER, tracheomalacia, sepsis</td>
</tr>
<tr>
<td>5</td>
<td>Primary EA repair</td>
<td>8</td>
<td>36</td>
<td>2.3</td>
<td>Atrial septal defect, congestive heart failure, renal failure</td>
</tr>
<tr>
<td>6</td>
<td>Primary EA repair</td>
<td>8</td>
<td>37</td>
<td>2.9</td>
<td>Transposition of great arteries</td>
</tr>
<tr>
<td>7</td>
<td>Gastrostomy</td>
<td>1</td>
<td>37</td>
<td>1.2</td>
<td>Trisomy 18 syndrome, VATER</td>
</tr>
<tr>
<td>8</td>
<td>TEF division, gastrostomy tube</td>
<td>86</td>
<td>31</td>
<td>1.1</td>
<td>Atrial septal defect, ventricular septal defect, tricuspid atresia</td>
</tr>
<tr>
<td>9</td>
<td>TEF division, gastrostomy tube</td>
<td>19</td>
<td>35</td>
<td>1.9</td>
<td>Ventricular septal defect, R arch, truncus arteriosus</td>
</tr>
<tr>
<td>10</td>
<td>TEF division, gastrostomy tube</td>
<td>65</td>
<td>34</td>
<td>2.0</td>
<td>Tetralogy of Fallot</td>
</tr>
<tr>
<td>11</td>
<td>TEF division, gastrostomy tube</td>
<td>12</td>
<td>29</td>
<td>1.6</td>
<td>Respiratory distress syndrome, patent ductus arteriosus</td>
</tr>
</tbody>
</table>

*Of 23 patients, 4 (17.0%) died before esophageal atresia (EA) repair. This is significantly higher than the 2 (2.3%) of 88 patients who died following primary EA repair with tracheoesophageal fistula (TEF) ($P_{.<.01}$). EGA indicates estimated gestational age; VATER, vertebral defects, imperforate anus, tracheoesophageal fistula, and radial and renal dysplasia.

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often associated with EA and TEF,14 were frequently ob-

were attempted primary EA repair if the patient's condition 

carded early thoracotomy with TEF division and at-

sisted more than 4 dilatations. McKinnon and Kosloske3 re-

sulting gastric distension further compromises lung capacity and 

limits effective ventilation. Early management with gas-

trostomy tube may result in a loss of positive ventilating 

pressure and lead to increased flow through the TEF.12 

Gastroesophageal reflux through the TEF directly into 

the tracheobronchial tree occurs and is not addressed by 

initial gastrostomy alone. Attempts at enteral feeding via 

the gastrostomy, although reported by some,13 increase 

aspiration risk and may only contribute to the develop-

ment of chronic lung disease. We, therefore, have advo-

cated early thoracotomy with TEF division and at-

tempted primary EA repair if the patient's condition 

allows.

Using this approach, we report an overall survival of 

93% in surgically treated patients in the present study. Nineteen 

(83%) of 23 patients who had initial TEF division sur-

vived to undergo delayed EA repair. This delayed EA re-

pair group represents a high-risk patient population as 

pair group represents a high-risk patient population as 

we believe that their poor outcome would have been impacted 

by primary EA repair. We also did not experience signifi-

cant morbidity related to the need for a second thoraco-

tomy, a possibility inherent in this delayed repair ap-

proach. The 7- to 8-week interval between procedures 

resulted from the time necessary for resolution of the pul-

monary condition and for patient growth. All of the pa-

tients with RDS or cardiac lesions were hospitalized be-

 tween procedures.

Patients with long-gap EA historically have been 

reated with initial cervical esophagostomy and delayed 

esophageal replacement.7 Attempts at native esophageal 

reconstruction have been associated with anastomotic complica-

ions as a result of tension in bringing the distant ends together. Efforts aimed at preserving the 

ative esophagus have included esophagomyotomy,15 up-

per pouch bougienage,16 staged extrathoracic elongation,17 

and delayed primary repair. Boyle and colleagues18 re-

ported primary esophageal anastomosis in 8 patients with 

ultrashort-gap EA (gap length, >3.5 cm) without a length-

ening procedure. They note significant anastomotic tension 

in most cases with associated early complications, 

but overall eventual good outcome. Of the 4 patients in 

the present study with long-gap EA with TEF, 50% re-

quired fundoplication for GER refractory to medical 

therapy and 50% developed anastomotic stricture requir-

ing more than 4 dilatations. McKinnon and Kosloske3 re-

ported an increasing incidence of anastomotic compli-

actions and significant GER requiring fundoplication that 

correlated with increasing gap lengths.

In the present study, we were able to achieve a pri-

ary esophageal anastomosis in all patients with a long gap 

or an upper esophageal pouch initially located above the 

thoracic inlet after approximately 3 months. Patients were 

reated with upper pouch suctioning and gastrostomy bol-

us feeds. Seven patients remained hospitalized between 

procedures, whereas 3 were cared for at home. No specific ef-

forts were taken to enhance elongation of the upper pouch 

in the interval. Spontaneous growth of the upper pouch 

may result from continuous swallowing of saliva as has been 
suggested by Puri et al.19 By tacking the pouch to the ver-

tebra, there is also some elongation of the esopha-

gus associated with linear growth of the infant. All pa-

tients demonstrated GER on contrast study radiographic 

images, and we believe that this contributed to the growth 

of the distal esophageal segment.

Marked wall thickening and overall growth of both 
esophageal segments were uniformly noted at delayed

* There was no significant difference in the instances of anastomotic complications, gastroesophageal reflux, or survival in patients who received delayed EA repair compared with those who received primary repair (P<.35). TEF indicates tracheoesophageal fistula.

<table>
<thead>
<tr>
<th>Complication</th>
<th>Primary Repair (n = 88)</th>
<th>Respiratory Distress Syndrome (n = 9)</th>
<th>Long-Gap EA (n = 18)</th>
<th>Severe Cardiac Lesions (n = 3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stricture</td>
<td>11 (12.5)</td>
<td>1 (20)</td>
<td>4 (20)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Leak</td>
<td>2 (2.3)</td>
<td>1 (20)</td>
<td>1 (10)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Recurrent TEF</td>
<td>2 (2.3)</td>
<td>1 (20)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>&gt;1 Operation</td>
<td>2 (2.3)</td>
<td>1 (20)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Gastroesophageal reflux</td>
<td>13 (14.7)</td>
<td>1 (20)</td>
<td>4 (20)</td>
<td>1 (33)</td>
</tr>
<tr>
<td>Fundoplication</td>
<td>2 (2.3)</td>
<td>1 (20)</td>
<td>1 (10)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Death</td>
<td>4 (4.6)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
</tbody>
</table>
esophageal reconstruction. The intramural blood supply was excellent and allowed further mobilization of the esophageal ends to achieve a tension-free anastomosis. We speculate that prior dissection and mobilization ultimately enhance intramural esophageal blood supply much in the way of a delayed pedicle flap, commonly used in reconstructive surgery. Two patients developed anastomotic stricture, 1 of whom had had a subclinical leak revealed on a prior contrast study image and the other who had had a myotomy. One required revision of the anastomosis and eventually did well. The observed incidence of anastomotic leak (10%) or stricture (20%) in these patients with long-gap EA was similar to that reported in other large series and certainly less than the 50% reported by Boyle et al and the 44% reported by Brown and Tam in similar patients who underwent a primary repair.

Symptomatic GER developed in 2 patients; 1 patient with associated duodenal stenosis required fundoplication. As discussed above, we had previously demonstrated GER on contrast study results in all patients and thought that it was important in the growth of the distal esophagus. Following EA repair, all patients were treated with histamine type 2 receptor blockers, upright positioning on a wedge, and prokinetic (motility) agents. Parker et al documented significant GER and manometric abnormalities in the distal esophagus in 14 of 17 patients with EA and TEF; these include disordered motility, aperistalsis, and simultaneous contractions. Six patients in that study required fundoplication. Tension on the distal esophagus, especially with a prior gastrotomy, alters the position of the gastroesophageal junction and the angle of His, both of which contribute to the development of GER. We have generally been reluctant to perform fundoplication in our patients with EA and TEF because of the risk of worsening esophageal motility and have favored instead a more gradual advancement of oral diet postrepair. While most patients were discharged while still receiving a combination of gastrostomy nutrition and oral diet, all except 1 were tolerating a full oral diet within 6 months.

We conclude that primary esophageal repair can be performed in all patients with EA and distal TEF. The initial finding of a long gap does not commit the child to a cervical esophagostomy and esophageal replacement. The staged approach is also useful in premature infants with RDS or associated cardiac defects. In these high-risk infants, the native esophagus can be preserved for the long term and their acute pulmonary condition status can be optimized during the newborn period.

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