Delayed Primary Repair of Esophageal Atresia With Tracheoesophageal Fistula

Is It Worth the Wait?

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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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PATIENTS AND METHODS

PATIENT POPULATION

The medical records of all newborn children admitted to Children's Hospital and Medical Center in Seattle, Wash, with the diagnosis of EA or TEF between February 1986 and December 1996 were reviewed. Using the classification system of Waterston et al.4 134 patients were identified: 5 patients (3.7%) were type A (pure EA); 1 patient (0.7%) had type B (EA with proximal TEF); 118 patients (88%) had type C (EA with distal TEF); 1 patient (0.7%) had type D (EA with proximal and distal TEF); and 9 patients (6.6%) had type H (TEF).

Eighty-eight patients (74.6%) with type C underwent fistula division and primary esophageal repair in a single operation; 4 of these patients required myotomy of the upper pouch for esophageal anastomosis. Three patients (2.9%) with severe associated congenital lesions died without any intervention. Three patients (2.5%) had gastrostomy placement as the initial procedure, 2 of which were performed at an outside hospital prior to referral. In 23 patients (19.5%), the TEF was surgically divided (ligated in 4) and a gastrostomy was placed. In this group, primary esophageal 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).

Sixty-seven of 134 patients (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 13 (19.4%), tetralogy of Fallot in 112 (17.4%), and aortic arch anomalies in 8 (12.0%). Twenty-one patients (15.9%) 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 gastrostomy was also created. Postoperatively, these patients had upper pouch nasoesophageal suctioning and were fed via the gastrostomy. 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 transternal 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 gastrostomy 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 nonsurvivors 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-

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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.

**COMMENT**

Since the first successful repair of EA with tracheoesophageal fistula in 1941, the outcome has steadily improved. 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 al6 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 centers7,8 reporting increasing success, including our own institution. 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.8 Pohlson et al10 re-

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

<table>
<thead>
<tr>
<th>Patients With</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 = .83).
ported 94% survival in premature infants who were managed with primary repair and emphasized that size alone need not dictate care.

The TEF assumes central importance in our management of the newborn with EA and TEF. This is especially true in the premature infant with RDS. As the need for positive pressure ventilation increases and as the lung compliance decreases because of RDS, the TEF allows for preferential ventilation of the fistula. The resulting gastric distension further compromises lung capacity and limits effective ventilation. Early management with gastrostomy tube may result in a loss of positive ventilating pressure and lead to increased flow through the TEF. 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, increase aspiration risk and may only contribute to the development of chronic lung disease. We, therefore, have advocated early thoracotomy with TEF division and attempted 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 survived to undergo delayed EA repair. This delayed EA repair group represents a high-risk patient population as the need for positive pressure ventilation increases and as the lung compliance decreases because of RDS, the TEF allows for preferential ventilation of the fistula. The resulting gastric distension further compromises lung capacity and limits effective ventilation. Early management with gastrostomy tube may result in a loss of positive ventilating pressure and lead to increased flow through the TEF. 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, increase aspiration risk and may only contribute to the development of chronic lung disease. We, therefore, have advocated early thoracotomy with TEF division and attempted 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 survived to undergo delayed EA repair. This delayed EA repair group represents a high-risk patient population as demonstrated by the higher incidence of complex cardiac defects and the higher early mortality, compared with those infants who had primary EA repair. Cardiac anomalies are often associated with EA and TEF, were frequently observed in our study population, and were present in all patients in the delayed EA repair group. In 4 patients, echocardiograms demonstrated an isolated patent ductus arteriosus, which would be expected in this newborn population; however, the cardiac defects were severe enough in 7 patients to undergo a delayed approach. Three of these 7 patients died before EA repair. Nearly all of the patients with EA and TEF who died at our institution during the past decade have had significant cardiac lesions. One infant (1.59 kg, 29 weeks’ estimated gestational age) with RDS and a severe patent ductus arteriosus with congestive heart failure died 12 days after TEF division. In this high-risk group (infants with severe cardiac defects), we do not believe that their poor outcome would have been impacted by primary EA repair. We also did not experience significant morbidity related to the need for a second thoracotomy, a possibility inherent in this delayed repair approach. The 7- to 8-week interval between procedures resulted from the time necessary for resolution of the pulmonary condition and for patient growth. All of the patients with RDS or cardiac lesions were hospitalized between procedures.

Patients with long-gap EA historically have been treated with initial cervical esophagostomy and delayed esophageal replacement. Attempts at native esophageal reconstruction have been associated with anastomotic complications as a result of tension in bringing the distal ends together. Efforts aimed at preserving the native esophagus have included esophagomyotomy, upper pouch bougienage, staged extrathoracic elongation, and delayed primary repair. Boyle and colleagues reported primary esophageal anastomosis in 8 patients with ultralong-gap EA (gap length, >3.5 cm) without a lengthening 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% required fundoplication for GER refractory to medical therapy and 50% developed anastomotic stricture requiring more than 4 dilatations. McKinnon and Kosloske reported an increasing incidence of anastomotic complications and significant GER requiring fundoplication that correlated with increasing gap lengths.

In the present study, we were able to achieve a primary 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 treated with upper pouch suctioning and gastrostomy bolus feeds. Seven patients remained hospitalized between procedures, whereas 3 were cared for at home. No specific efforts 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. By tacking the pouch to the vertebral fascia, there is also some elongation of the esophagus associated with linear growth of the infant. All patients 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

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Table 3. Clinical Outcome Following Delayed Esophageal Atresia (EA) Repair*

<table>
<thead>
<tr>
<th>Complication</th>
<th>Primary Repair (n = 88)</th>
<th>Respiratory Distress Syndrome (n = 5)</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>

*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 < .05). TEF indicates tracheoesophageal fistula.
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 stenosis, one 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 gastrostomy, 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 gastros-tomy 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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REFERENCES