Thirty-five–Year Institutional Experience With End-to-Side Repair for Esophageal Atresia

Robert J. Touloukian, MD; John H. Seashore, MD

Hypothesis: End-to-side repair (ES) with ligation of the tracheoesophageal fistula (TEF) reduces the risks of stricture and gastroesophageal reflux disease requiring operation compared with the end-to-end repair of esophageal atresia and distal TEF.

Design: Case series with institutional and historical control subjects.

Setting: Referral children’s hospital.

Patients: One hundred thirty-four infants diagnosed as having esophageal atresia and distal TEF between June 30, 1968, and July 1, 2003.

Interventions: Ninety-six infants having ES and 38 having end-to-end repair.

Main Outcome Measures: Patients were studied for overall survival, surgical complications, and well-being during the first year of life.

Results: Survival was 95% vs 90% (patients undergoing ES vs end-to-end repair). Complications included anastomotic leak, 8% vs 13%; recurrent TEF, 7% vs 3%, with only 1 recurrence in the last 28 patients having ES; anastomotic stricture (requiring dilatation), 5% vs 13%; gastroesophageal reflux disease requiring operation, 6% vs 18%; and esophageal dysmotility, which was present following nearly all ES and end-to-end procedures. Tracheomalacia-related respiratory symptoms following ES decreased from 50% to 11% at 1 year of age. Age-appropriate diet following ES was achieved in 93% by 1 year; 5% experienced occasional dysphagia or choking episodes.

Conclusions: The ES operation is accompanied by a reduced rate of stricture and gastroesophageal reflux disease requiring operation compared with end-to-end repair. Earlier concerns regarding an unacceptable risk of recurrent TEF were not substantiated.

Arch Surg. 2004;139:371-374

END-TO-END REPAIR (EE) with division of the tracheoesophageal fistula (TEF) has been the operation of choice to repair esophageal atresia for more than 50 years but is accompanied by a higher than 20% rate of anastomotic stricture and gastroesophageal reflux disease (GERD) requiring operation. Our institution has used the end-to-side repair (ES) with ligation of the TEF for 35 years and has sought to determine whether the risk of stricture and reflux previously reported could be reduced.

PATIENTS

One hundred sixty infants without previous surgery diagnosed as having esophageal atresia and a distal TEF were admitted to the Newborn Special Care Unit of the Yale–New Haven Children's Hospital between 1968 and 2003. One hundred thirty-four (84%) underwent primary repair, including 96 having ES with ligation of the TEF performed by us in a nonrandomized series. Thirty-eight patients received EE with division of the TEF by other pediatric surgeons on the hospital staff. The remaining 26 patients had staged repair, including 8 receiving an esophageal replacement operation. This study is limited to a comparison of the patients receiving ES at our hospital vs those having EE at our institution and others having EE reported from comparable institutions.

METHODS

DESIGN

A case series of 96 patients undergoing ES between June 30, 1968, and July 1, 2003, at the Yale–New Haven Children’s Hospital was compared with institutional and historical control subjects having EE with division of a TEF.

INTERVENTIONS

End-to-side repair with ligation of the TEF has been the subject of previous studies from our institution in which the technique has been presented and illustrated (Figure). Since the last
The overall survival of patients having ES was 95% compared with 92% and 90% for those having EE at our hospital and at other comparable institutions, respectively.\(^7\) When analyzed by the Waterston risk classification, the survival for ES drops to 84% vs 50% for group C classification patients. No statistical significance can be assigned because the number of patients having EE is small (Table 1).

Complications (Table 2) included anastomotic leak, defined as extension into the pleural space, which was reported in 8% for ES vs 13% for EE. The incidence of anastomotic stricture, defined as requiring multiple dilations to ensure patency, was 5% for ES vs 13% for EE.

The overall recurrence rate of TEFs following ES was 7%, with a decrease in incidence during the 35 years from 3 cases (17%) for the first 18 patients to a single case (4%) for the last 28 patients (Table 3). One (3%) of 38 patients undergoing EE at our institution had a recurrent TEF.

The incidence of GERD requiring an antireflux operation was 6% for ES vs 18% for EE. Esophageal dysmotility was almost uniformly observed on barium esophagram studies obtained after ES. This finding was similar to that observed following the EE operation. Tracheomalacia-related respiratory symptoms decreased from 50% to 11% at 1 year of age, and age-appropriate diet following ES was achieved in 93% by 1 year. Five percent experienced occasional dysphagia or choking episodes following ES (Table 4).

**COMMENT**

The ES operation was first proposed by Sulamaa et al\(^8\) in 1951 as a possible alternative to the EE for reducing the unacceptably high risk of anastomotic leak and stricture reported by Haight and Towsley.\(^9\) Further investigations, including the survey of the Surgical Section of the American Academy of Pediatrics published in 1964,\(^10\) indicated that the prevailing risk of leak was 17% and that the incidence of stricture was as high as 45%. Ty et al,\(^11\) in 1967, introduced the ES operation to North America, and studies\(^12-14\) during the next several years showed a lower rate of leak and stricture but a significant incidence of recurrent TEF compared with the EE operation. For these reasons, the ES operation was largely abandoned by pediatric surgeons.

Despite the poor results reported by others, we have continued to be enthusiastic about the ES. We report our 35-year experience with 96 patients to emphasize that...
with appropriate modification of the Ty et al. operation the risk of recurrent TEF could be minimized to an acceptable low level and the incidence of stricture, leak, and severe GERD requiring a modified antireflux procedure was lowered below that reported with the EE.

Surgeons agree that the risk of anastomotic stricture is increased with tension and local ischemia at the suture line. In a survey, the subjective estimate of the degree of tension at the EE anastomosis was reported as moderate to severe by more than 60% of surgeons. We experienced a degree of anastomotic tension similar to that associated with EE, necessitating a myotomy in 7 of 96 patients (7%) to accomplish the operation. Despite the subjective similarity in the degree of tension, the incidence of stricture following EE is significantly higher than with ES. In a recent study of 69 patients undergoing EE at James Whitcomb Riley Hospital for Children, Indianapolis, Ind, 26% required multiple dilatations for stricture. We propose that the reduced risk of esophageal stricture offered by the ES is based on advantages provided by the technique itself. Anastomosis to the side of the lower esophagus protects the submucosal blood flow because the segmental blood supply is not interrupted, thereby reducing the risk of ischemic injury. Postoperative tension at the anastomosis is also lowered by preserving the fistulous attachment that eventually becomes a fibrous cord. The ligated lower esophagus moves with the upper during swallowing and avoids the opposing forces observed at the suture line associated with the EE. The ES also provides an oblique anastomosis with a large enough circumference to facilitate free drainage of the upper pouch. The vector forces at the anastomosis are unequally distributed, reducing the tendency to separation.

An antireflux procedure to control GERD following EE is reported to be required in up to 28% of patients. The reduced incidence of GERD requiring an antireflux procedure following ES is difficult to explain, because radiographic evaluation of the lower segment following ES and EE showed a uniform tendency for moderate to severe dysmotility and mild to moderate gastroesophageal reflux. Nonetheless, by taking care to preserve the entering branch of the right branchial artery and the vagus nerve, as first advocated by Ty et al. some preservation of peristalsis is accomplished and the lower esophageal sphincter is protected. Scrupulous medical management also lowers the need for an antireflux operation.

The most contentious objection to the ES is an unacceptable high risk of recurrent TEF. Recurrence of the TEF occurred in 6 (21%) of the first 42 patients (14%) operated on by Pietsch and associates and in 17% (n = 3) of our first 18 patients. That number has dropped to only one recurrence for the last 28 patients in our series. We believe this success can be attributed to the learning curve and to modifications in our technique, such as flush ligation of the TEF within 1 mm of the tracheal wall and sufficient tightening of the stricture to the first branch and then crushing the muscular wall without cutting through the lumen. Our initial concern that overtightening may lead to disruption of the fistula has not materialized, as all recurrences were secondary to recanalization at the site of an intact ligature and none of the patients had an early pneumothorax. We also avoid tranfixion sutures, by scraping the mucosa or by double ligation, to decrease the risk of recurrence. Excessive fistula width, defined herein as greater than 6 mm, is unusual, is limited to fewer than 10% of cases, and is a contraindication to achieving permanent lumen obliteration. For these patients, fistula division and EE is recommended, as it is for patients with muscular continuity of the proximal and distal segments. Similarly, we have not found any difference in the indication for staging, and the same criteria are recommended as those used for the EE.

In conclusion, the ES operation is accompanied by a lower rate of stricture and GERD than the EE operation. The incidence of recurrent TEF is lower than previously reported by our institution and is attributed to strict adherence to modification of the technique. Therefore, earlier concerns regarding an unacceptable risk of recurrent TEF following ES have not materialized. The ES procedure is offered as an alternative and is not proposed as a replacement for those content with their acceptable results.

Accepted for publication November 25, 2003.

This study was presented at the 84th Annual Meeting of the New England Surgical Society; September 20, 2003; Newport, RI; and is published after peer review and revision. The discussions that follow this article are based on the originally submitted manuscript and not the revised manuscript.

Corresponding author and reprints: Robert J. Touloukian, MD, Section of Pediatric Surgery, Yale University School of Medicine, 333 Cedar St, PO Box 208062, New Haven, CT 06520 (e-mail: robert.touloukian@yale.edu).

Table 4. Long-term Outcome of 96 End-to-Side Repairs With Ligation of Tracheoesophageal Fistula

<table>
<thead>
<tr>
<th>Recurrent Respiratory Symptom</th>
<th>No. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>First year of life only</td>
<td>54 (56)</td>
</tr>
<tr>
<td>Beyond first year</td>
<td>10 (10)</td>
</tr>
<tr>
<td>Age-appropriate diet</td>
<td>85 (89)</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>5 (5)</td>
</tr>
<tr>
<td>Choking spells</td>
<td>5 (5)</td>
</tr>
</tbody>
</table>

REFERENCES

And then, finally, why does simple ligation in continuity increase the risk of fistulization, while ligation in continuity with anastomosis does not? It is somehow related to the reflux issue as well, because with simple ligation there is increased intraluminal pressure immediately after the surgery right at that ligation, and I believe that has something to do with it. I would also hasten to add that we as surgeons find ligating hollow viscera extremely unorthodox. Nonetheless, one must ligate the fistula down tightly to obliterate the lumen and can’t allow it to recanalize. That is something that takes a little bit of courage for those of us who find ligating hollow viscera extremely unorthodox.

And then, finally, insofar as the Boston experience is concerned, I would encourage you to have those patients looked at, because with the long experience at Boston you should be able to demonstrate a significant improvement over time if the hypothesis that experience makes the difference is true.

Albert W. Dibbins, MD, Portland, Me: I want to focus on 2 things. One is gastroesophageal reflux. For our practice and, as you have shown, for many pediatric surgeons, reflux is the great difficulty after the repair. We have approximately the same rate that you showed with your end-to-end repair. Twenty percent of our children require Nissen fundoplication because the reflux is intractable and we can’t deal with it any other way. However, we are reluctant to do this because of the esophageal dysmotility problem. Have you done any studies of your patients to look at that lower esophageal sphincter pressures? Bill Sieber years ago took children with esophageal atresia and studied them before they were repaired and showed that the dysmotility in the distal segment was a function of the disease and the lower esophageal sphincter was intact. We have been impressed with 2 things. Our infants with repaired esophageal atresia have very low LES pressures and delayed gastric emptying. Gastric emptying studies in our children are prolonged, and this contributes to the reflux. Do you have any data on gastric emptying, because the low rate of significant reflux in your end-to-side patients is markedly different than most everyone else’s experience.

The other issue is the difficulty in finding recurrent fistulas. I have 2 children in my own practice with recurrent fistulas. I couldn’t tell you how many times I have bronchoscooped them and done other studies, knowing in my heart that they had a recurrent fistula. It took me 13 years to find one of them and 15 years to find the other, with really severe respiratory symptoms, and I never felt that I could operate on them until I could demonstrate the recurrent fistula. I wonder what your experience with that has been.

Dr. Touloukian: We have done esophageal motility studies and reported those in the early series simply to document the fact that the dysmotility exists principally in the lower esophagus. I think that is well known. However, with the results being as good as they have been, I have been a little reluctant to repeat the study, but as part of a research experience, that certainly is a good question. My presumption is that with minimal reflux lower esophageal sphincter tone is probably preserved.

The dysmotility issue leading to the need for an anti-reflux procedure is an important one. There is going to be some variability among various centers as to how frequently that operation is done, recognizing that any time you increase lower esophageal sphincter pressure in the face of dysmotility you are going to end up with a balloonied esophagus, increasing aspiration risks, so that with very good meticulous medical management a lot of these babies can be managed over the first year of life when this problem is a particularly significant one. I am not saying you can avoid needing to do the operation, but we have seen some of our patients with reflux that have actually done quite well after the first several months of life, so I think one has to be extremely patient and diligent with these so that you don’t do too early an operation. However, I am quite certain of the fact that the 20% rate is about the number.