ANNALS OF SURGERY
Vol. 226, No. 4, 533–543

Development of a True Primary Repair for the Full Spectrum of Esophageal Atresia

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Objective
To determine whether or not a true primary repair, without myotomies and with the gastroesophageal junction below the diaphragm, can be accomplished across the esophageal atresia (EA) spectrum. Our hypothesis is that the esophageal anastomosis can withstand significant tension. The consequences, particularly for those patients with a very long gap atresia, were assessed.

Summary of Background Data
Difficulties arise roughly in proportion to the size of the gap between esophageal segments. Reported surgical complications remain frequent, and particularly at the far end of the EA spectrum, not all children are left with a satisfactorily functioning esophagus or esophageal substitute.

Methods
The outcomes of all infants who had a true primary repair of EA from 1976–1997 were determined. Surgically, the methods used to achieve a reliable true primary repair were expanded to accomplish this, even for a very long gap EA.

Results
From 1976–97, 70 infants with or without associated tracheoesophageal fistula (TEF) had primary repairs performed with no surgery-related deaths and 11% later deaths. No interpositions were performed since 1983. There were no discernible anastomotic leaks and one late recurrent TEF related to the early use of balloon dilation. Ten infants had gaps of 5.0–6.8 cm and, among these, four had gaps of 5.5–6.8 cm that could not be pulled together initially. Traction sutures in the esophageal ends, however, produced sufficient lengthening within 6–10 days for a true primary repair. Very long gap repairs produced more reflux (10 of 10 required a fundoplication *versus* 24 of 70 overall) and more dilations to relieve strictures. Two infants underwent stricture resection with no recurrence. On follow-up, all patients over 2 years of age were eating well or satisfactorily, and none had a gastrostomy tube.

Conclusions
(1) The esophageal anastomosis can withstand considerable tension and allows a reliable true primary repair for the full EA spectrum. (2) Growth is rapid and traction sutures will

produce significant esophageal lengthening within days. (3) With increasing tension, gastroesophageal reflux (GER) and strictures are more common; however, both are treatable. Follow-up reveals the benefits of true primary repair over other solutions.

The repair of esophageal atresia (EA) is considered the epitome of pediatric surgery, yet continues to pose difficulties for the surgeon.[1] Although survival is now determined by the associated anomalies, not all infants are left with a method of repair that will function well for 70 or more years. There is general agreement that the child's own esophagus is best, but it also is believed that a primary repair is not always possible.[2-11] The central issue is the distance between the two atretic segments; the longer the gap, the greater the tension if a primary repair is carried out.

A basic tenet of surgery is to avoid tension on an anastomosis, and many strategies have been developed to severely reduce or eliminate it in EA repair. The circular myotomy is the most commonly used technique to lengthen the esophagus in the repair of medium length and longer atresias, but it may not result in a low-tension anastomosis.[12] The esophageal hiatus can be opened and part of the stomach can be pulled up; however, this results in the gastroesophageal (GE) junction being above the diaphragm.

When the gaps are judged too long for a primary repair, many different solutions continue to be used.[5,8-11] The current resurgence of popularity for the gastric transposition procedure, and the still-frequent use of the colon interposition, make these two popular choices for esophageal replacement.[5,8,10,13] Unfortunately, early complications are frequent, and only approximately 60% to 80% of these interpositions are satisfactory on follow-up. More important, most substitutions have severe, late problems built into them and the results will become even less favorable.

Our underlying hypothesis, however, has been that the esophageal anastomosis will withstand a great deal of tension, and the demonstration of this has allowed us to extend the indications for primary repair. As a result of testing this hypothesis, we have found the growth of short esophageal segments will be surprisingly rapid and greatly enhances the possibility of a primary repair. Because we believe that a true primary repair will provide the best long-term solution for these children, the purpose of this report is to present our evidence that it is possible throughout the EA spectrum.

Table 1. PRIMARY REPAIR OF ESOPHAGEAL ATRESIA

Number of infants	70
Birth weight (μg)	1.0–4.1
Age at operation (days)	1–322
Early deaths [n (%)]	1 (1)
Late deaths [n (%)]	7 (10)
Long-term survivors [n (%)]	62 (89)

PATIENTS AND METHODS

From 1976 to 1997, 70 infants underwent primary repair of EA at the University of Minnesota (Table 1). There were 60 infants (87%) with a distal fistula, eight (11%) with pure EA, and one with both an upper and a lower fistula and another with only an upper fistula. Two infants with an isolated H-type tracheoesophageal fistula, two who had a jejunal interposition before 1983, and one infant who did not undergo repair because of severe chromosomal abnormalities were excluded. Eight patients weighed <1.8 kg, 42 weighed between 1.8 and 3 kg, and 20 weighed >3 kg at time of repair.

The diagnosis was suspected after choking with feeding was observed and usually confirmed by the inability to pass a tube into the stomach. In some cases, either an air or a contrast pouchogram was used to secure the diagnosis. Before surgery, the patients were kept in a semiupright position with intermittent suction of the proximal pouch to prevent aspiration. Fluid resuscitation and antibiotics were given as indicated. Twenty patients had their primary repair delayed for more than 1 week after birth because of severe pulmonary distress, low birth weight, apparent pure EA, or when an unexpected right aortic arch led to repair through a left thoracotomy incision.

In this series, gap lengths were estimated at the time of repair and from x-ray studies.[14] This determination is admittedly imprecise and usually was made in the operating room both before dissection and after with mild traction applied either by suture or catheter. After dissection, the ends retract and the gap appears longer. Similarly, the gap length on x-rays was measured with a catheter at least in the upper pouch. The gaps ranged from insignificant to 6.8 cm (greater than seven vertebral

Presented at the 117th Annual Meeting of the American Surgical Association, Quebec City, Quebec, Canada, April 17–19, 1997.

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Accepted for publication April 23, 1997.

spaces). Vertebral spaces have the advantage of giving a relative estimate and gaps also were recorded by the number of spaces on x-ray (when the infant is approximately 5 kg in weight, vertebral spaces are approximately 1 cm apart).[14]

To assess the role of gap length in the results, the infants were subdivided into three groups. We defined short gaps as <1 cm, moderate gaps from 1 to 2.4 cm, and long gaps when 2.5 cm or more separated the esophageal ends. Given the imprecision of estimating gap length, we increased the usual requirement cited in the literature for a long gap from 2 to 2.5 cm.[4,5,7,14] There were 27 short gaps, 18 with moderate gaps, and 25 with long gaps.

A local vascular anomaly was associated with a long gap (>2.5 cm) in nine infants. A vascular ring comprised of a right aortic arch, retroesophageal aorta, and a patent ductus arteriosus was found in four infants. Five infants had an anomalously arising right subclavian artery, which coursed through the gap. The presence of a vascular ring or an anomalous vessel seemed to ensure a long gap. The remaining long gaps were found in infants with EA alone and EA with either a proximal or distal fistula and no other local anomalies.

Gaps of >3 cm are known to be associated with significant technical problems and often are considered to be beyond a true primary repair.[15,16] Consequently, the long gaps were further subdivided to examine separately the infants with very (or ultra) long gaps defined as >3.5 cm.[17] Examination of the ultralong group was done to prevent a large number of easier repairs obscuring the problems incurred by very long gap repairs.

Very long gaps commonly are associated with pure atresias without a distal fistula. Eight of the very long gap infants had pure EA with no fistula, one had a proximal but not a distal fistula, and only one had the most common form with a distal fistula. These ten had gap lengths greater than five vertebral spaces (range, 5–6.8 cm) and represented the extreme of the EA spectrum.

Operative Technique

Early in the series, a gastrostomy usually was included to reduce reflux and subsequent stricture formation. Because reports have indicated reflux actually may be increased by gastrostomy, recently it often has been omitted.[18] When included, the gastrostomy was placed low on the greater curvature in case a fundoplication was needed later. Although smaller in pure EA, the stomach was found to be quite adequate in strength as opposed to the conclusion in one study.[11]

Using a transthoracic, extrapleural approach on the side opposite the aortic arch, the dissection and anastomotic technique was carried out as has been described.[14] The fistula, if present, was divided and oversewn. Both the upper and lower ends of the esophagus were handled as minimally as possible. Because of the relatively rich submucosal blood supply, dissection of the upper pouch was extensive, and on two occasions, a neck incision was made for further mobilization. The lower end of the esophagus was minimally dissected to better preserve its blood supply, although it may not be greatly compromised by mobilization.[14]

The upper and lower pouches were opened and stay sutures placed at their lateral extent to aid in approximation. Tissue forceps were used without grasping to allow identification of the mucosa and the placement of deep suture bites without producing significant tissue injury. The tissue bites were very generous and were at least 6 mm from the edge, with care taken to include an equal depth of mucosa. The posterior row of 6–0 Prolene sutures (Ethicon, Somerville, NJ) was placed without tying. Depending on the gap, the posterior row of sutures was used to place traction until the ends came together. For some repairs, several minutes of traction was required until the ends came together and the sutures could be tied individually, off tension. The knots for the posterior row were within the lumen. The anterior row of 6–0 or 7–0 Prolene sutures (Ethicon) was placed and a temporary catheter stent passed by mouth across the anastomosis before these sutures were tied. In all cases, the temporary catheter stent was removed after the anastomosis was completed.

Six infants required a preliminary procedure so that a true primary repair could be carried out. Four patients, including two with cervical fistulas placed at other institutions, had gaps of 5.3 to 6.8 cm, and preliminary dissection confirmed that the ends could not be brought together initially. The longest gap (6.8 cm) infant is shown in the original study (Fig. 1A) and the eventual result (Fig. 1B). For these four, tissue-pledgetted traction sutures of 4–0 Prolene (Ethicon) were placed in the upper and lower segments before they were opened and brought out to the skin surface below and above the incision, respectively. Over the next 6 to 10 days, with increasing external traction, the segments lengthened surprisingly quickly. When the segments were within 1 to 2 cm of each other by x-ray examination, the patients were returned to the operating room and a true primary repair carried out.

One infant had an upper but no lower fistula. The cervical fistula was divided first, with the upper pouch pulled into the upper portion of the thorax and the primary repair performed 6 weeks later. One premature infant had the distal fistula ligated first to aid ventilator management, and 4 weeks later, the primary repair was done.

All 70 patients had a true primary repair of their EA without circular myotomies, and in all the GE junction

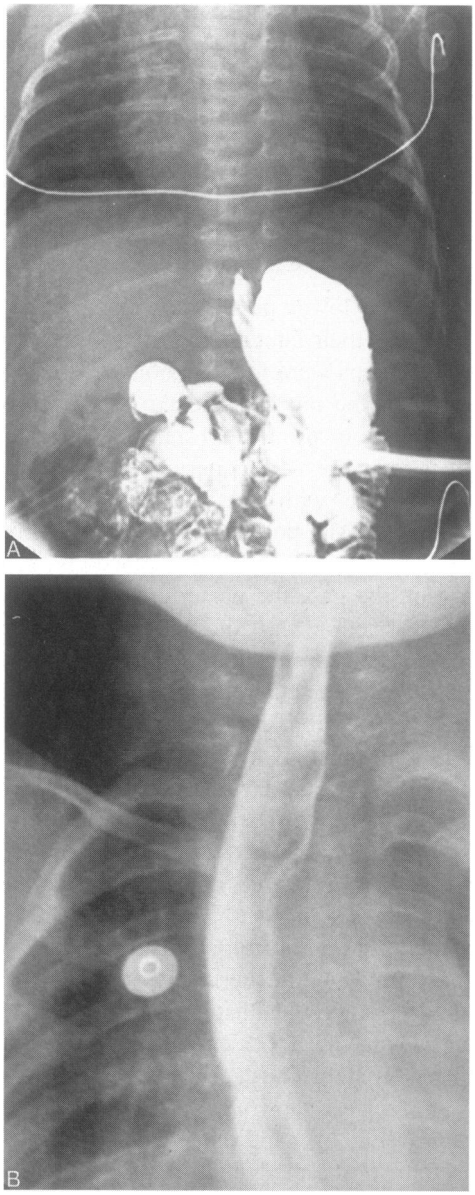

Figure 1. (A) Preoperative x-ray of a patient with pure esophageal atresia and a gap length of greater than seven vertebral spaces. A catheter is visible threaded to the end of the lower esophageal segment. (B) Most recent esophagram of same patient after initial traction sutures and subsequent true primary repair.

was left below the diaphragm. Since 1983, there have been no interposition grafts used to achieve continuity, even in infants with a very long gap.

Postoperative Care

Infants who underwent repair with only moderate anastomotic tension were weaned from the ventilator as quickly as possible and kept in a semiupright position. When severe anastomotic tension was present, the infants were kept paralyzed and required the ventilator for an additional 3 to 6 days. No deep suctioning was performed. Total parenteral nutrition was provided for the first 7 to 12 days until oral or gastrostomy feeding was begun.

A postoperative esophagram was done on approximately day 10 for most infants to rule out a leak or anastomotic problem before the child was fed. An anastomotic leak would have been diagnosed by either the esophagram or the presence of saliva in the chest tube. Neither occurred. More recently, the contrast study has sometimes been omitted, except in the cases of very long gap atresias.

Most infants underwent routine, awake, outpatient, esophageal dilation at 6 weeks, 3 months, and approximately 5 months after repair. In addition, the gastrostomy was removed if no evidence for significant stricture existed. Additional dilations were performed as needed.

RESULTS

In this series, there were no apparent early surgical complications. Until 1987, postoperative barium swallows were performed in all cases, which showed nothing beyond an occasional irregularity at the suture line. All very long gap repairs have had contrast esophagrams, and no leaks have been found. For the entire series, there have been no clinically or radiographically apparent leaks.

The awake dilations have been done without difficulty. We have used fluoroscopic-guided balloon dilations when a fundoplication was present to avoid disruption of the wrap. Two complications occurred early in the series from the balloon technique. One late fistula recurred after a balloon dilation procedure when this method was beginning to be used. A low esophageal tear, well below the anastomotic site, was produced at another hospital from inflation of a misplaced balloon. Subsequently, there have been no complications from this technique.

There was one death within 30 days of the operation when support was withdrawn because of the consequences of severe birth asphyxia. Seven late deaths occurred that were related to associated anomalies or, in one case, the complications of low birth weight. The deaths were distributed throughout the birth weight groups.

Late operations were performed for a variety of indications. The subsequent procedures included 24 fundoplications for reflux, 5 aortopexies for tracheomalacia, and 2 resections of strictures in 2 infants who were resistant to dilation (Table 2). One infant underwent a later division of an unsuspected upper pouch fistula.

The relation between gap length, GE reflux, and stricture formation was examined. A Nissen fundoplication to control reflux was required in 24 (34%) of 70 infants from

Table 2. EARLY AND LATE RESULTS

	Number (%)
Early operative results	
Anastomotic leaks	0 (0)
Recurrent fistulas	0 (0)
Postoperative dilations	
0–4	51 (78)
5–10	9 (14)
>10	5 (8)
Late operations	
Excision of stricture	2 (3)
Undetected cervical fistula	1 (1)
Aortopexy	5 (7)
Nissen fundoplication	24 (34)
Surviving patients reached	51
Eating normal diet	51 (100)
Remaining gastric tubes	0 (0)

32 (46%) of 70 who had significant reflux. As reported previously, there was an association between reflux and late stricture formation.[19] Most patients had 2 to 4 prophylactic dilations of the esophageal anastomosis, and 21 (30%) of 70 received additional dilations for symptoms suggestive of anastomotic narrowing. These symptoms were more common the longer the gap length. In all cases except two, the strictures yielded to dilations, with or without fundoplication (Table 2).

Follow-Up

To evaluate the longer term results of primary anastomosis in patients with tracheoesophageal fistula, we conducted two detailed telephone interviews with the parents of patients or with the patients themselves. The questions assessed the child's ability to swallow and dietary habits. This dietary and functional follow-up was carried out only for children 2 years of age and older. Patients younger than 2 years of age are often still progressing through feeding advancement and may be having dilations. Fifty-two surviving patients were older than 2 years of age (8 patients had died). One patient was lost to follow-up shortly after repair, and four more who were getting along well for several years were reached only for the first interview. The initial follow-up was 98%, and 90% were reached for current eating satisfaction. The average duration of follow-up was 8.8 years (range, 2–19 years). No gastronomy tubes remained.

All patients were reported to be eating satisfactorily or excellently, and 93% of the children were eating like their siblings. One fourth of the patients chewed their food carefully, especially meats or bread, but 91% could eat anything they wished or should for their age. Fourteen percent of the respondents had at least one episode of food sticking in the esophagus. Only 16% of the parents thought that the child might need additional dilations.

All surviving patients over 2 years of age are now eating a diet that is normal for age with a follow-up from 2 to 19 years. As a testimony to feeding well, 93% of patients were above the 10th percentile, 73% were above the 25th percentile, and for 59%, their weight percentile exceeded their height percentile.

DISCUSSION

The repair of EA has provided a long and fascinating story.[14] Within 60 years, the lesion has gone from being uniformly fatal to only rarely being the cause of death. Primary repair has become increasingly reliable following the first success by Haight in 1941, and the mortality is now largely determined by the severity of associated anomalies.[3,8,14,20] Although operative mortality has declined greatly, complications related to the EA repair continue to be frequent, and, particularly at the far end of the spectrum, a significant number of children have neither a good repair nor a satisfactory esophageal substitute. The difficulties and controversies cluster around the long gap solutions; therefore, we examined the consequences of doing a primary repair across the full EA spectrum and compared them with the outcomes reported for other approaches.

The gap length between the two esophageal ends largely determines the complication rate.[15,21] When a significant gap exists, the anastomosis will necessarily be under tension and a basic surgical principle violated. Moderate-to-long gaps are frequent, and some degree of tension will be present in more than half of primary EA repairs. Even the most recent series reported leakage rates of 9% to 21%, including major anastomotic disruptions, 5% to 12% recurrent fistulas, and strictures, which occurred in 15% to 29%.[2–7,13,15,20,22,23] Of interest, these complication rates do not significantly differ from the large, cooperative series reported in 1964.[24]

A long gap has been defined as either 2 or more cm or two vertebral spaces between the segments.[3,7,12,15,17,21] More than one author has noted, however, that too long is in the eye of the beholder.[8,22] Gaps 2 cm or less generally are considered amenable to primary repair, and when the gap is 3 cm, the complication rates are significantly higher.[7,10,11,15,16] One series included a subgrouping of EA with a 3-cm gap, and the reported complication rate (100%) was a testimony to the difficulty of treating this group of infants with primary repair.[16] But, because the

advantages of a native esophagus seem to outweigh the problems encountered, primary repair has been advocated strongly for all except for the longest gaps (>3 cm or three to four vertebral spaces).[9,11,16]

Despite the great interest and efforts made toward achieving a primary repair, a common admonition, in the face of a very long gap, has been that one should not try too hard to preserve the esophagus. A variety of substitutions have been advocated, and we have presented the only report of true primary repairs carried out with a gap length of >3.5 cm.[17] The results of our series counter the conclusion that esophageal substitution will be necessary at the very long end of the EA spectrum.

Our hypothesis has been that a well-constructed anastomosis can withstand significant amounts of tension.[17] This conclusion was supported by our initial results and allowed us to extend primary repair to the full spectrum of EA, even with gap lengths of > 5 cm.[25]

The technical aspects of EA repair rarely are discussed in any detail in recent reports. Sometimes it is stated that the surgeon should use fewer rather than more sutures to accomplish the anastomosis.[22] The suture type and size are little mentioned, although it has been noted that the highly reactive silk sutures are associated more frequently, as would be expected, with anastomotic leaks.[22,23] Anastomotic leaks usually seal spontaneously, but their presence signify problems that may be magnified in the presence of severe anastomotic tension. Recurrent fistulas virtually always require reoperation and also represent a significant technical difficulty.

Our position is that technique is of great importance in EA repair, particularly when dealing with the difficult end of the spectrum. We believe that many fine, nonreactive sutures will produce a far more secure anastomosis than the few, relatively heavy, reactive sutures usually advocated.[11,22] If the sutures are tied off tension, as described, then repair of even very long gaps under great tension appears to be reliable.[14,17]

The difficulties produced by long-gap EA are well recognized, and a number of approaches have been used to solve this problem. Time is commonly used to allow the segments to grow. A relative reduction in gap length may occur, but we and others have not found significant changes.[14] The infants are larger and seem sturdier at operation, however, so we wait until the infants are approximately 3.5 to 4 kg for a very long gap primary repair.

Bouginage has been advocated to increase upper pouch length and decrease the gap length to 2 cm so that a primary repair with or without myotomies could be carried out.[26] Despite the reported success, we have found bouginage is difficult for patients and families alike and could injure the upper pouch. Therefore, we do not use this approach.

Lengthening procedures also have been used to solve the long gap problem. A circular myotomy at one or more levels commonly is done to lengthen the upper pouch. Circular myotomies may only reduce, not eliminate, tension and complications remain. A compilation of 9 series (54 patients) showed circular myotomy was associated with a significant incidence of problems: stricture (33%), leak (22%), and recurrent tracheoesophageal fistula (6%).[12,27-33] In addition, a new problem arose and up to 20% experienced a dilated myotomy site, which, in some cases, caused ventilatory compromise.[30,32] Presumably, these dilated segments, unsupported by muscle, will continue to enlarge. Myotomies also will block peristaltic propagation and extend upward the level of dysfunction.[34]

A spiral myotomy has been proposed to lengthen the upper pouch; however, it also will extend the dysmotility, and there is little follow-up information available.[35] Finally, a flap created from the upper pouch has been used in some cases with encouraging results.[36] All lengthening techniques that involve dividing the muscle, however, have had significant complications and the long-term results are uncertain.

For gaps >3 cm, EA usually has been repaired using an interposition graft, most commonly colon or a variety of stomach-lengthening procedures. Jejunal interpositions have provided excellent long-term results; however, they rarely are used.[37] Colon interpositions seem to have a lower rate of satisfaction and gastric solutions are gaining favor.[38] Stomach solutions include creating a tube from the greater curvature, dividing the lesser curvature to elongate the stomach, or pulling up some portion of the stomach through the esophageal hiatus.[37-40] Gastric transposition is the most complete form in which virtually all of the stomach is pulled into the chest. The colon and stomach interpositions, however, have developed frequent early and late problems.[5,9-11,38-41] Most important, the presence of the GE junction in the chest or neck will ensure GE reflux, and the specter of metaplasia and eventual dysplasia seems inevitable.[42] Because a functioning, native esophagus should avoid most or all of these problems, the recommendation for primary repair has been made even in the face of frequent predictable complications.[11,16]

We believe a true primary repair without a myotomy or disruption of the esophageal hiatus should provide the best long-term solution. Although the tension was not quantitated, in the long gap group (24 infants), it ranged from significant to extreme. We believe the technical details including minimal tissue handling, generous tissue bites, and a single layer of fine, nonreactive sutures, tied off tension were important to success.[14,17] The relatively common complications of leak and recurrent fistula that have plagued all series were eliminated.

At the most difficult end of the spectrum, a true primary repair was accomplished in all ten patients with preoperative gaps of 5 to 6.8 cm. For six infants with gaps of 5.0–5.5 cm, steady traction over minutes allowed the true primary repair to be carried out initially. The preliminary traction sutures used in four infants revealed how rapidly growth is induced in the two esophageal segments. Although the traction suture requirement meant a two-stage rather than a one-stage primary repair and an additional 6 to 10 days receiving ventilator support, the results seem to justify this approach. A true primary repair was possible even in the most difficult patients, including those whose upper pouch was partially immobilized by a cervical fistula. As noted in other series, these children eat much sooner and better than after an esophageal substitution.[5,10,11] Now the most important influence on eating appears to be the presence or absence of food aversion.

The patient at the furthest end of the EA spectrum in this series is depicted in Figure 1A with a gap at time of initial operation of 6.8 cm (7.5 vertebral spaces). The lower esophageal segment was about as small as is seen in pure EA; nevertheless, satisfactory growth was induced by traction sutures to allow a true primary repair. He was one of two children to later require a stricture resection, and his final contrast study is shown in Figure 1B. This child is now 24 months old, eats whatever he wants, including pretzels and potato chips, and has not been dilated in 6 months. We believe this case shows that remarkable growth of even the smallest esophageal segment is possible and discarding similar small segments because they will be defective or useless is untenable.

Although our operative approach successfully accomplished primary repair throughout the EA spectrum, there may be later consequences of the initial tension. We found that gap length was related to the occurrence of GE reflux, stricture formation, and need for later fundoplications. We did not dissect the esophageal hiatus and kept the GE junction below the diaphragm; nevertheless, reflux is enhanced by the tension of a long gap primary repair.

It is well known that the lower esophageal segment has impaired function, which has been thought to be increased by tension.[43,44] Our follow-up studies, however, do not suggest that tension increases dysmotility or its consequences. The dysmotility appears to be a lack of coordinated peristalsis beyond the anastomotic site. The peristaltic wave normally is propagated through the muscle wall and will be interrupted wherever an anastomosis exists.[45] Similar dysmotility would be seen if a normal esophagus were transected and repaired and reflux may not occur. Once reflux is controlled, there does not appear to be a significant functional effect of the discoordinated motility and the children eat well.

Gastroesophageal reflux was more common in the long gap group (64%) than in the entire group (32/70 or 45%). Control of reflux has shown to be valuable in reducing stricturing.[19,46,47] Children who exhibited signs of significant GE reflux and did not respond to medical treatment underwent Nissen fundoplication. In this series, 24 (34%) eventually had a fundoplication and the majority were in the long gap group. These results suggest the degree of tension and amount of reflux are linked despite the imprecision in the clinical diagnosis of significant reflux and the variability in the criteria for operation.

The diagnosis of stricture also is somewhat subjective, but some generalizations seem justified. Virtually all patients in this series received two to four prophylactic outpatient dilations of the esophageal anastomosis in the first 5 months after surgery. Anastomotic strictures were defined by the need for additional dilations and were suggested by symptoms, including slow feeding or occasional choking. These symptoms do not necessarily prove the presence of a stricture, but a confirming contrast study was done in many of the patients. Additional dilations were required by 21 (30%) of 70 patients. The incidence of additional dilations was highest in the long gap group but occasionally was required in the minimal gap group. The stricture rate, however, was comparable to series using myotomies to reduce tension.[4,10,27,31]

Presumably, strictures are a consequence of several factors that include natural healing by fibrosis, the initial small size of the lower segment, the presence of tension, which further narrows the anastomosis, and GE reflux. The presence of a leak also would enhance stricture formation.[22,23] Significant reflux must be controlled if present and, without continuing reflux, the majority of these strictures responded to awake, outpatient dilations. The stricturing tendency usually relented within 6 to 12 months, but for two patients, it did not, and the stricture was resected. Although resection required another operation, it is a very effective method of treating this problem. Neither patient required continuing anastomotic dilations and both now eat well.

A less definite relation was seen between stricture formation and significant reflux. There were a few patients in whom operative control of GE reflux appeared to be useful in limiting stricture formation as has been reported. For the 23 infants who underwent a fundoplication, however, only 9 had more than 3 dilations. The apparent lack of a definite relation may be because we recommend a fundoplication early when significant reflux is present to minimize the stricturing tendency. With a relatively tight fundoplication, our reflux control appeared better than reported.[48]

For the very long gap group, we found that primary repair resulted in increased reflux and the need for a fundoplication in all. All children older than 2 years of age,

however, now are doing well despite the severe initial tension. These children, who began with an ultra-long gap, now are eating a normal diet, despite the consequences of the primary repair and the expected dysmotility of the lower esophagus. Function appears satisfactory and should persist for life. We suggest that given the magnitude of the initial problem, reflux and strictures represent the minimum that could be expected from a true primary repair. The extra procedures required seem reasonable in attaining a functioning native esophagus, particularly when compared to the results with interpositions.[5,8–11,37,38,41,42,49]

In summary, we found that the entire spectrum of gap lengths, even ultra-long gaps, could be successfully repaired primarily. The risks of this approach are related to the amount of anastomotic tension that may be generated. When carefully done, however, the anastomosis seems capable of withstanding severe stress. Moreover, the placement of traction sutures will induce growth in the esophageal segments within days and allow a primary repair of the longest gaps. The consequences of increased tension will be a higher incidence of reflux and stricture formation with a likely need for fundoplication and dilations. These consequences can be effectively treated, however, within the first 2 years. Thereafter, this type of primary repair without myotomy and with the GE junction below the diaphragm will provide an excellent chance for a well-functioning native esophagus with life-long durability. The benefits should increase with time.

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Discussion

DR. KATHRYN D. ANDERSON (Los Angeles, California): Although I have spent my professional career in esophageal replacement, I agree that the native esophagus is best. But not at all costs.

Based on my overall personal involvement in 55 cases of esophageal replacement, 32, or 50%, were for pure atresia or esophageal atresia with tracheoesophageal fistula. However, in the past 10 years, the only cases in which replacement has been necessary are those in which another surgeon has abandoned the esophagus and performed an esophagostomy, or after multiple reoperations for stricture, the so-called "train wrecks" that all pediatric surgeons get.

Many years ago I lost a child in whom I had tried to save the esophagus for far too long in an extremely premature infant. He died at 1 year of age, after esophageal replacement, of chronic lung disease. Chronic aspiration may masquerade as asthma in these children.

So I would suggest that there is a hidden morbidity that may not be manifest for several years after primary repair. I would also suggest that measuring the gap in centimeters is misleading. A 5-cm gap in a full-term infant is very different from the same gap in the 1200-g preemie. Vertebral body number is a much more useful measurement, and we wait for the two pouches to approach to within a distance of three vertebral bodies.

Again, the time this takes will be very dependent on the size of the infant and may take many weeks in the extremely premature infant. A caution here: although rare, an upper pouch fistula in the absence of a lower pouch fistula does occur, and it will tether the upper esophageal pouch, preventing its growth.

I have several questions for you, Dr. Foker. How many of your infants were less than 1500 g? I notice you said they were 3.5 to 4.5 kg. Those are enormous babies. Did you have any less than 1500 g? Did the birth weight correlate with the length of the gap and the time it took the two ends to grow close enough to anastomose? How did you measure the gap? Were the pouches stretched by dilators in the upper and lower pouches when you measured them? This makes a big difference in the apparent distance between the two ends. Have you truly followed these children long term? If so, have you seen any complications of recurrent stricture after several years? Any incidence of airway disease? And what is your incidence of fundoplication failure?

DR. KEN KIMURA (Iowa City, Iowa): I think this report is unique and innovative. We pediatric surgeons have learned that a primary anastomosis of the esophagus must not be tried when the gap is greater than 2.5 cm, dictated by trial and error and "common sense." Dr. Foker and his colleagues have challenged our common sense gap length that has been achieved during our experience for the last 50 to 60 years.

Dr. Foker's series showed failure of circular myotomy to lengthen the esophagus 1 or 2 cm. Instead of circular myotomy, we advocate a spiral myotomy, which preserves continuity of the muscular layers in the proximal esophagus, elongates the esophagus, and allows satisfactory swallowing.

Dr. Foker dilated the esophagus very frequently. When we dilate the esophagus after repair of esophageal atresia, we intend to dilate the anastomosis. For this purpose, one or two dilatations are usually successful. The contrast study of the esophagus in your patient looked like a segmental fibrosis that was caused by local ischemia. Do you think multiple dilatations can handle it? Another question is a technical one. We have difficulty finding the distal esophagus at the definitive procedure, because it