

Long-Term Analysis of Children With Esophageal Atresia and Tracheoesophageal Fistula

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Background/Purpose: For children with esophageal atresia (EA) or tracheoesophageal fistula (TEF), the first years of life can be associated with many problems. Little is known about the long-term function of children who underwent repair as neonates. This study evaluates outcome and late sequelae of children with EA/TEF.

Methods: Medical records of infants with esophageal anomalies (May 1972 through December 1990) were reviewed. Study parameters included demographics, dysphagia, frequent respiratory infections (> 3/yr), gastroesophageal reflux disease (GERD), frequent choking, leak, stricture, and developmental delays (weight, height < 25%, < 5%, respectively).

Results: Over 224 months, 69 infants (37 boys, 32 girls) were identified: type A, 10 infants; type B, 1; type C, 53; type D, 4;

type E, 1. Mean follow-up was 125 months. During the first 5 years of follow-up, dysphagia (45%), respiratory infections (29%), and GERD (48%) were common as were growth delays. These problems improved as the children matured.

Conclusions: Children with esophageal anomalies face many difficulties during initial repair and frequently encounter problems years later. Support groups can foster child development and alleviate parent isolationism. Despite growth retardation, esophageal motility disorders, and frequent respiratory infections, children with EA/TEF continue to have a favorable long-term outcome.

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INDEX WORDS: Esophageal atresia, tracheoesophageal fistula.

IN 1943, using an extrapleural approach, Haight and Towslet¹ reported the first successful repair of esophageal atresia (EA) with tracheoesophageal fistula (TEF). The incidence of EA and TEF ranges from 1 in 2,500 to 4,500 live births.^{2,3} Previous investigators have reported on the perioperative challenges faced by these neonates⁴, however, few have focused on the long-term outcome after repair. Furthermore, complete evaluation of gastrointestinal and respiratory function and morbidity of these growing children is lacking. The aim of this study was to identify the incidence and clinical significance of the long-term complications occurring in children treated for this congenital anomaly as a neonate.

MATERIALS AND METHODS

From May 1972 to December 1990 (18.7 years), 69 children with EA/TEF have been treated at our institution and were available for long-term follow-up. Study approval from the institutional review

board (IRB) was obtained. The medical records of these infants were reviewed to discover residual symptoms or complications that arose during infancy, elementary years, or teenage years. Children were seen annually in clinic. Symptoms and parental concerns were evaluated at 5-year intervals. The study was restricted to children who were available for follow-up for at least 10 years after surgical repair so that long-term data could be retrieved.

Study parameters included incidence of dysphagia, respiratory infections, gastroesophageal reflux, choking episodes, anastomotic leak or stricture, and developmental delays. Dysphagia was judged based on a careful history, frequency and extent of vomiting, and sensation of a food bolus becoming lodged in the esophagus. Respiratory infections were defined as at least 3 infections per year requiring examination and treatment by a physician. Only events thought to be related to aspiration were included in the current study. Children were further evaluated with regards to the number requiring anastomotic dilatations or anti-reflux procedures. Weight and height were taken at each visit and plotted on a standardized growth curve from the National Center for Health Statistics.

RESULTS

Thirty-seven boys and 32 girls were included in the study. Initial repair was carried out through a right extrapleural thoracotomy with division of TEF and primary single layer esophagoesophagostomy, often with prior gastrostomy placement. Esophageal replacement with colonic interposition was utilized in 7 of 10 type A patients. Forty-nine infants (71%) received gastrostomy tubes at the discretion of the attending surgeon. Gastrostomy was maintained until the child showed adequate nutritional support with good oral intake. Subsequently,

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Table 1. Description of Anomalies in Study Cohort

Description of Anomaly	Gross Classification	No. of Cases
Esophageal atresia without tracheoesophageal fistula	A	10
Esophageal atresia with proximal tracheoesophageal fistula	B	1
Esophageal atresia with distal tracheoesophageal fistula	C	53
Esophageal atresia with fistula to both pouches	D	4
Tracheoesophageal fistula without esophageal atresia	E	1
Total		69

NOTE. Study cohort consisted of 37 boys and 32 girls.

8 children underwent formal closure of the gastrocutaneous fistula.

Mean follow-up was 10.5 years. One child continued to be seen for 26 years. Fifty-three children (77%) were found to have EA with distal TEF. Anatomic description including fistula type and Gross classification can be found in Table 1. Infants were seen periodically for the first few years of life and then returned annually for follow-up evaluation. All of the children returned for postoperative visits during the first 5 years. During the elementary school ages of 6 to 10 years, 56 children (81%) returned to the clinic. Finally, 42 children (61%) were seen back in the clinic more than 10 years after their original operative repair. The families of children lost to follow-up had often relocated out of state and sought medical care at other facilities.

Incidence of dysphagia, respiratory infections, gastroesophageal reflux (GERD), and choking can be found in Fig 1. Interestingly, these complications occurred most frequently during the first 5 postoperative years and tended to improve as the child grew older. Heights and weights are noted in Fig 2. Almost half of the children weighed less than the 25th percentile during the first 5 years; however, the children's weight and height steadily improved. By the time the children had reached 10 years of age, less than one third of them continued to have weights or heights less than the 25th percentile.

Eighteen children (26%) required multiple dilatations during the first 5 years of life. Sixty percent of the children who subsequently had GERD had gastrostomy

tubes as infants. Nineteen children (28%) with GERD required Nissen fundoplication for severe gastroesophageal reflux, 2 of whom had had Barrett's esophagitis. Twelve children (17%) were shown to have an anastomotic stricture on upper gastrointestinal series. Two children (3%) had an anastomotic leak. Two children (3%) had documented tracheomalacia. Because of their infrequency, anastomotic leaks and tracheomalacia did not contribute significantly to overall morbidity.

DISCUSSION

Children with EA/TEF have a significant incidence of gastroesophageal reflux. Postoperatively, esophageal motility disorders are observed. Esophageal peristalsis is impaired, whereas the lower esophageal sphincter may be incompetent.⁵⁻⁷ Several studies have reported an increased incidence of gastroesophageal reflux after EA/TEF repair; however, the definitions have not been uniform. Furthermore, discrepancy between subjective reflux and documented gastroesophageal reflux disease is likely to be present. Using retrospective and prospective methods, Koch et al⁸ objectively studied the incidence of GERD with endoscopic, manometric, and radiographic studies. He suggested that GERD was the result of delayed gastric emptying, superior displacement of the esophagogastric junction, and decreased esophageal clearance in the hypoperistaltic esophagus. Following up patients for up to 23 years, he noted that three fourths of the children had radiographic evidence of gastroesopha-

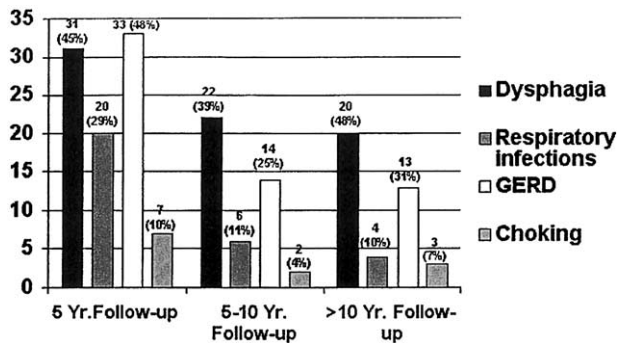


Fig 1. Bar graph shows incidence of dysphagia, respiratory infections, GERD, and choking at 5-year intervals.

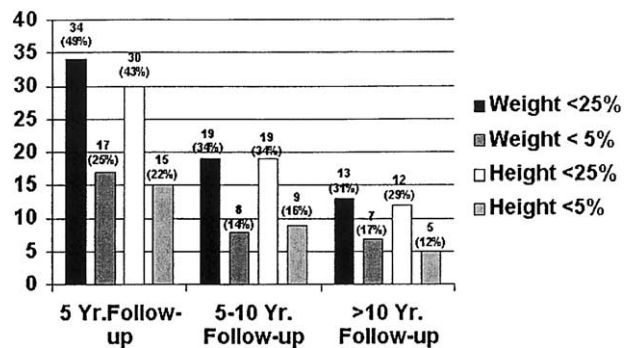


Fig 2. Bar graph shows the incidence of weight and height below the 25th percentile and 5th percentile at 5-year intervals. Weight and height data taken from the National Center for Health Statistics.

geal reflux during the early years of follow-up. This observation is similar to findings described by Fonkalsrud et al⁹ and Gauthier et al.¹⁰

Gastroesophageal reflux was found in only one fourth of the children followed into their late teen and early adult years. The current study also showed a declining incidence of gastroesophageal reflux as the child matured. Overall, 28% of the children underwent an antireflux procedure, most commonly Nissen fundoplication. Only 2 type A patients required an antireflux procedure. Most of these operations were performed in early childhood; however, a few of the funduplications were performed on teenagers, suggesting that disordered esophageal motility in some patients is a lifelong problem. All type A patients experienced gastroesophageal reflux. Four children did not respond to their first operation and required a second fundoplication, suggesting that there may be a subset of patients with poor motility or emptying. In these children, a partial wrap may be appropriate.¹¹ Schier et al¹² reported that 17% of their patients with EA/TEF required fundoplication, 9% in the study by Somppi et al.¹³ Six percent of the children in the review by Somppi et al¹³ had Barrett's esophagitis. Three percent of the children in this report had this serious complication.

The internet has become the second opinion for patients and families across all specialties. Several esophageal atresia support groups are currently available. The Chicago-based EA/TEF Child and Family Support Connection, circa 1992, and the United Kingdom-based TOFS, circa 1982, offer counseling and education to families and patients.^{14,15} Schier et al¹² reported their experience with the world's largest support group that included more than 1,100 children. The group, including families, patients, and pediatric surgeons, meets regularly and publishes a journal. The group focuses on providing emotional reassurance and information regard-

ing bougienage techniques, feeding patterns, and reflux management. Alarmingly, these investigators note that patients often found themselves isolated when making the transition from childhood to adult medical services, suggesting that pediatric surgeons should claim a larger role in preparing the adult physician to care for these patients. Furthermore, results of Schier's questionnaire notes that some parents had turned to alternative therapies to treat children with chronic respiratory infections, feeling that they had exhausted currently available allopathic practices. He also reported that two thirds of the children had no difficulties with feeding or only experienced difficulties during the first few postoperative years. With time, most children learned to eat and drink in an upright position and to avoid foods that might become lodged in the esophagus, such as hot dogs. Concerning gastroesophageal reflux, parents generally noted an improvement after fundoplication but also added that avoiding late meals was an effective way to reduce gastroesophageal reflux.

Children born with esophageal anomalies are likely to have problems well into their adult years. In some patients with severe esophageal motility disorders and poor esophageal emptying, dysphagia may be a long-term problem. Families not only face difficulties during the initial repair but also may encounter problems years later. A support group at the larger centers may be able to foster child development, alleviate parent isolationism, and provide a database to improve overall care. Few disorders offer the pediatric surgeon the ability to follow children so closely from early childhood well into their teenage years. Most serious complications occur within the first few years of life. Despite early growth retardation, a high incidence of esophageal motility disorders, and frequent respiratory infections, children with EA/TEF have a good long-term quality of life in the majority of cases.

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Discussion

A. Coran (Ann Arbor, MI): This is an important study because there are very few long-term studies that we have on most of the anomalies we take care of, and this is one of the few that looks at this particular anomaly. So, I want to congratulate the authors for this. I would like to ask one question. You said in the abstract that you had 10 Type A esophageal atresias. Of those 10, how many were repaired primarily and how many had an interposition? I would also like to make one comment. There is a long-term study in adults only, done in the late 1970s by Mark Orringer, who is an esophageal surgeon in Ann Arbor, in which he took all of Dr Haight's old patients who had reached adulthood and studied them with manometry and did motility studies. In that long-term follow-up, he found an incidence of about 25% of gastroesophageal reflux.

D. Little (response): Thank you for that question, Dr Coran. We had 10 patients who were described as Type A patients, and 7 of those required interposition to treat their esophageal atresia. So, 70% interposition, 30% primary for Type A.

D. Beals (Lexington, KY): In all cases, the symptomatology that you described increases from the 5- to 10-year age group to the greater than 10-year age group. So, the supposition that these patients have a good quality of life may be unfounded, because they may, in fact, have increasing complications as life goes on. Could you comment on that statistic?

D. Little (response): One of the reasons that this is the case is that these children are beginning to verbalize and now can describe their problems. As the children got older, older than 10 years, some of these symptoms got better. But, you are certainly right, many of these children will have complications that go for life, and that is one of the reasons I think this paper is important. This is not always a short-term problem. It is a long-term problem that these children may face.

D. Beals (Lexington, KY): You made a comment that the quality of life was excellent in these children, but how did you come to that conclusion since you did not show any quality of life data?

D. Little (response): Your point concerning quality of life is correct if you compare these children with esophageal atresia with children without anatomic anomalies; however, the first operative survivor would only be 62.

When compared historically, these children have a good quality of life. Dysphagia and reflux are the most common long-term problems and are usually medically managed. Operative intervention is sometimes required. We did not formally study quality of life as an objective variable. With the success of this study, we would like to design a study specific to quality of life.

R. Touloukian (New Haven, CT): I think it would be very useful for me to know how you stratified your results against any known risk classification schemes, such as the Waterston classification, which you mentioned, or the Spitz classification which uses the presence or absence of cardiac anomalies. Also, did you include your staged repairs in the long-term outcome? If so, in what way did staged repair affect the morbidity over the long term?

D. Little (response): Dr. Touloukian, I will address your second question first. We did include staged repairs in our data. Although these cases were few in number, results for perioperative complications and morbidity would probably be affected. However, we looked at long-term complications. I do not believe stratification would affect these results at 5 and 10 years postoperatively.

Regarding your first question, we did not stratify by the classification schemes of either Waterston or Spitz.

G. Holcomb (Kansas City, MO): Danny, that was a nice study. I am particularly interested in your 19 patients who underwent the Nissen fundoplication. As you know, there is some debate on whether the patients with esophageal atresia should undergo a partial fundoplication or a complete fundoplication. In that group of patients who underwent the Nissen fundoplication, did you notice an inordinate number of patients needing dilation or the need to take down the complete fundoplication and convert to a partial fundoplication?

D. Little (response): We did, in fact, look at that. It is important to note that many of the children that suffer from dysphagia and reflux disease may not tolerate complete fundoplication. In fact, four of our patients that initially underwent a Nissen fundoplication returned to the operating room with conversion to a different wrap, such as a Tha1. There certainly is ample evidence in the literature that these children with motility disorders may

benefit from a partial wrap or certainly a less tight wrap, if you will, to prevent this complication.

From the Floor (Amsterdam): Thank you very much for these interesting and relevant figures. One of our grave concerns is the effects of chronic reflux and the development of Barrett's esophagitis in these patients, and we have had several of those. Can you comment on that in your series?

D. Little (response): Of the 69 children that we treated during this 10-year follow-up, 2 of them did have Barrett's esophagitis. We have followed up with them. In fact, there is a study from Samppi showing a 6% incidence of Barrett's esophagitis in these patients. Our incidence was 3%.

S. Borenstein (Toronto): Can you comment on the

incidence of tracheomalacia in this group of patients and what proportion in follow-up required any type of surgical intervention for tracheomalacia.

D. Little (response): We had 3 patients in this study that had tracheomalacia, which is a bit lower than what you will see in the literature. None of those required operative intervention such as aortopexy.

K. Bass (Chicago, IL): My question is in follow-up to the Barrett's children that you diagnosed. Did those children undergo fundoplication before Barrett's developed? Or, did they undergo fundoplication after they Barrett's developed?

D. Little (response): It was subsequent to the Barrett's. They underwent fundoplication and then followed up appropriately.