Long-Term Esophageal Function Following Repair of Esophageal Atresia

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Primary repair of esophageal atresia restores gastrointestinal continuity, but does not ensure normal esophageal function. To date 22 patients, six to 32 (average 15) years after repair of their esophageal atresias, have been evaluated by personal interview and esophageal manometrics and acid reflux testing. Previous barium swallow examinations had demonstrated varying degrees of anastomotic narrowing (12 patients), abnormal esophageal motor function (11 patients), gastroesophageal reflux (two patients), and hiatal hernia (one patient). Ten patients experience intermittent dysphagia for solid foods. Seven have typical symptoms of gastroesophageal reflux. Esophageal function tests including manometry and intraesophageal pH recording, have demonstrated varying abnormalities of esophageal motility in 21 patients and moderate to severe gastroesophageal reflux in 13. Two patients have required reconstruction of the esophagogastric junction for control of severe reflux esophagitis. The unexpected high incidence of gastroesophageal reflux in these patients, coupled with their abnormal esophageal motility which impairs normal acid clearing, renders them more prone to reflux esophagitis. Careful long-term evaluation for gastroesophageal reflux and its complications is indicated following primary repair of esophageal atresia. Evaluation of esophageal function with intraesophageal pressure and pH recordings is a far more sensitive indicator of esophageal physiology than the barium swallow examination.

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IN 1941 CAMERON HAIGHT performed the first successful extrapleural ligation of the fistula, with primary anastomosis of esophageal segments, in a female infant with congenital esophageal atresia and tracheoesophageal fistula.[14] This monumental technical achievement formed the basis of the modern surgical approach to infants with this lesion and was responsible for converting a condition which carried a 100% mortality rate to one which is now associated with a survival rate in excess of 60%.[17]

During the two decades following Dr. Haight's initial report, numerous articles documenting and refining the preoperative, intraoperative and postoperative management of infants with esophageal atresia were published. It soon became apparent, however, that while primary repair of esophageal atresia restored gastrointestinal continuity, it did not ensure normal esophageal function. Cinefluorographic and later esophageal manometric studies in infants and young children documented disordered esophageal motility in almost all of these patients.

In recent years, with refinements in techniques of esophageal manometry and the use of the intraesophageal pH probe to measure gastroesophageal reflux directly, more precise data regarding the nature of various motor disturbances of the esophagus have emerged. The almost simultaneous development of severe reflux esophagitis in two young adults, 16 and 27 years of age, who had had repair of esophageal atresia, prompted this study, which was undertaken to define more precisely the long-term esophageal motor disturbances and incidence of gastroesophageal reflux in these patients.

Material and Methods

Patients being followed in the Thoracic Surgery Clinic at the University of Michigan, who had survived primary repair of esophageal atresia in infancy for at least six years, were contacted to determine their willingness to undergo interviews and esophageal function tests. To date, 22 patients have been evaluated by review of their medical history, personal interview, barium swallow examination when indicated, and esophageal function tests. The age range of these patients was six to 32 years (average 15). Seven patients were between six and eight years of age, seven were between 12 and 16 years, and the remaining eight were between 17–32 years of age. All but one of these patients had had a tracheoesophageal fistula in association with esophageal atresia. The anomalies were of the common variety, with a blind proximal

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esophageal pouch and the fistula between the proximal end of the distal esophageal segment and the tracheobronchial tree. One patient had had primary repair of an esophageal atresia without a tracheoesophageal fistula.

Seven patients had new barium swallow examinations because of increasing esophageal symptoms; eight had had barium swallows within five years of this study and the remaining seven had the results of earlier barium examinations in their medical records. Because esophageal symptoms in these latter 15 patients had not changed recently, repeat roentgenographic studies were not obtained.

Esophageal function tests, which were obtained in all patients, included manometry and intraesophageal acid reflux testing. In addition, acid clearing ability[4] was tested in 19 patients, and a Bernstein acid perfusion test[3] was performed in 14. Intraesophageal pressures were measured through triple-lumen constantly perfused catheters with openings spaced five centimeters apart. A pH probe was attached to the manometry catheters. The combined unit was inserted through the mouth in the seven patients younger than 12 years and through the nose in the remaining 15 patients. No sedation was used in any patient. Using standardized procedure described elsewhere,[29] the location, length, amplitude, and ability to relax reflexly with deglutition were determined for both esophageal sphincters in each patient. Esophageal motor activity with swallowing was recorded at three to five centimeter intervals.

Acid reflux testing in the 15 patients 12 years of age or older was performed as described by Kantrowitz and his associates,[19] by placing 250 to 300 ml of 0.1 normal HCl into the stomach and monitoring intraesophageal pH during standard postural maneuvers by means of an electrode positioned five centimeters above the distal high pressure zone (HPZ), or the pressure inversion point (PIP) when no HPZ was present. In the younger, smaller patients, 100 to 175 ml of 0.1 normal HCl were used, depending upon the subject's complaint of abdominal fullness. Even with the shorter esophageal length in the children, the pH electrode was positioned five centimeters proximal to the HPZ or PIP. Thus measured acid reflux in these children was of relatively greater magnitude than a similar grade of reflux in the adults.

Results

Past History

Twelve of the 22 patients had required esophageal dilatations during the first two years of life, one between ages five and seven, two between ages seven and nine, and five after ten years of age. Ten patients had required esophagoscopy and removal of impacted food during their first ten years of life, but none after this age. In one patient an upper thoracic esophageal perforation had occurred during instrumentation of an anastomotic stricture at age four. One boy had undergone resection of the esophageal anastomosis and primary reanastomosis at one and one-half years of age for a persistent anastomotic stricture. The stenosis had recurred and required yearly dilatations until age ten. Recurrent respiratory infections were indicated in the histories of eight patients. One patient had had an asymptomatic H-type TEF divided four years after his original repair in infancy.

TABLE 1. *Esophageal Symptoms After Repair of Esophageal Atresia (22 patients)*

Symptom	No.	Per Cent
Substernal dysphagia	10	45
Delayed emptying	7	32
Acid regurgitation	7	32
Food regurgitation	6	27
Heartburn with postural aggravation	4	18
Heartburn from spices, citrus fruits, etc.	4	18
Cervical dysphagia	4	18
Painful swallowing	1	5

Measurements of the distance the lower esophageal segment had to be mobilized for the anastomosis had been recorded in the operative notes of 12 of the patients. The length of distal esophageal mobilization was one to one and one half centimeters in four patients, two to two and one half centimeters in two patients, three to three and one half centimeters in four patients, and more than four centimeters in two patients.

Current History

Chronic respiratory symptoms, "asthma" or recurrent infections, were present in only four patients, aged six, seven, nine, and 14 years. Esophageal symptoms, however, were found in 17 of the 22 patients (Table 1). Using the clinical status evaluation proposed by Des Jardins and associates,[10] five (23%) patients represented excellent results of operation (completely asymptomatic); 13 (59%), good results (occasional dysphagia particularly for breads or meats), and four (18%), only fair results (frequent dysphagia and/or recurrent respiratory infections).

Barium Swallow Examination

The barium swallow findings reported in these patients are shown in Table 2. While some degree of anastomotic narrowing or abnormal esophageal motility was noted by the radiologist in about one-half of the patients, radiologic reports such as "satis-

TABLE 2. *Barium Swallow Findings Reported After Repair of Esophageal Atresia (22 patients)*

Symptom	No.	Per Cent
Anastomotic narrowing	12	55
Abnormal motility with delayed emptying	11	50
Gastroesophageal reflux	2	9
Hiatal hernia	1	5

factory appearance of the esophagus following repair of esophageal atresia" were equally common and provided little or no information regarding the status of esophageal function.

Esophageal Function Tests

Esophageal function tests proved to be the most sensitive indicator of esophageal abnormality (Table 3). In five patients there was complete loss of tone of the distal esophageal sphincter mechanism with no identifiable HPZ. The average mean and peak HPZ pressures (four and 9.17 mm Hg, respectively) in the remaining 17 patients tended to be approximately one-half the comparable values (nine and 20 mm Hg) for asymptomatic, healthy young adult controls who have been evaluated in our laboratory with esophageal function tests. In all patients with an identifiable HPZ, however, normal reflex relaxation of this sphincter mechanism with deglutition was demonstrated (Fig. 1). Similarly, normal relaxation of the upper esophageal sphincter with swallowing occurred in all patients and mean and peak pressures within this sphincter were comparable to those of normal controls.

All but one patient had abnormal esophageal motor function. In 18 patients, there was no progressive peristalsis within the entire esophagus, esophageal contractions after swallowing being simultaneous and either of low (11 patients) or normal (seven patients) amplitude (Fig. 2).

With an intraesophageal pH probe, moderate to severe acid reflux was documented in 13 (59%) of these patients, including the 5 in whom no distal esophageal HPZ was found (Fig. 3). This degree of reflux occurred in four of the seven patients younger than 12 years and nine of the 15 patients 12 years or older. Of the five patients in this series who had no esophageal symptoms, four had no detectable gastroesophageal reflux and one had minimal (1+) gastroesophageal reflux (considered within normal limits). The only patient with normal acid clearing ability was the single individual in this group whose peristaltic activity was normal on manometric evaluation. Instillation of acid into the

TABLE 3. *Results of Esophageal Function Tests after Repair of Esophageal Atresia (22 patients)*

Test	Finding	No. of Patients	Per Cent
I. Manometry			
A. Characteristics of HPZ (mean ± S.D.)			
Mean pressure (mmHg)	4 ± 2.42		
Peak pressure (mmHg)	9.17 ± 5.63		
Length (cm.)	2.18 ± 1.39		
No HPZ		5	23
B. Motility Disorder			
None		1	5
Loss of progressive peristalsis		21	95
Simultaneous normal amplitude contractions—entire esophagus		7	33
Simultaneous low amplitude contractions—entire esophagus		11	52
Normal peristalsis-proximal esophagus; sumultaneous contractions in distal two-thirds		3	14
C. Characteristics of upper esophageal sphincter (mean ± S.D.)			
Mean pressure (mmHg)	7.55 ± 2.17		
Peak pressure (mmHg)	17.77 ± 5.45		
Length (cm.)	2.5 ± .65		
II. Acid-Reflux Test			
Negative		8	36
Minimal (1+)		1	5
Moderate (2+)		3	14
Severe (3+)		10	45
III. Acid-Clearance Test			
Abnormal		18	95
Normal		1	5
IV. Acid-Perfusion Test			
Positive		3	21
Negative		11	79

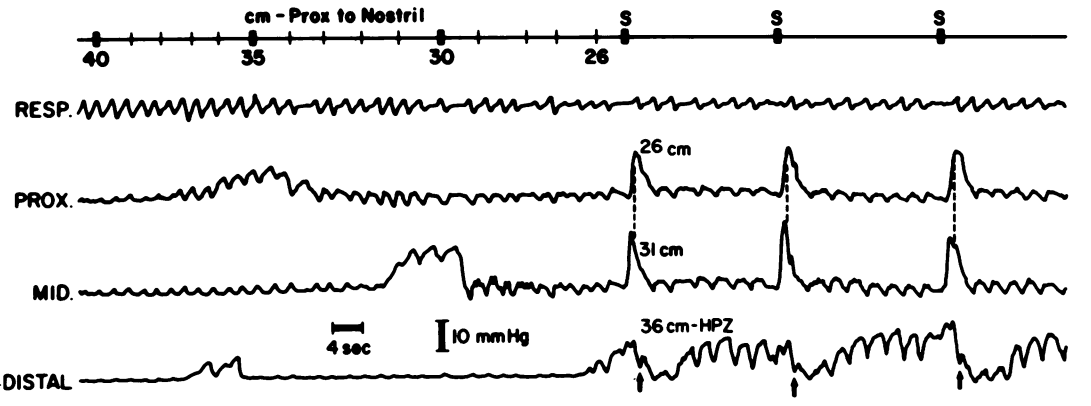

FIG. 1. Pullback resting-pressure profile of the gastroesophageal junction in 16-year-old patient who had had repair of esophageal atresia. Note the weak distal esophageal high pressure zone (HPZ) between 36 and 34 cm, normal reflex relaxation (arrows) of the HPZ with swallowing (S), and the normal amplitude, but simultaneous esophageal contractions with swallowing.

esophagus produced no symptoms in 11 of the 14 patients in whom the acid perfusion test was performed. Table 4 shows the relationship of the degree of distal esophageal mobilization required at operation to the presence and extent of gastroesophageal reflux found. Two of the six patients who required between one and two and one half centimeters of distal esophageal mobilization were found to have moderate or severe reflux, in contrast to five of the six patients who required mobilization of three or more cm of distal esophagus.

Treatment

The 13 patients in whom moderate to severe gastroesophageal reflux was documented have been instructed to elevate the head of their bed, and avoid eating or drinking for two hours before retiring at night. In addition, those with symptomatic gastroesophageal reflux have been placed on small, frequent feedings and antacids after meals and at bedtime. Because of the severity of their esophageal symptoms, however, three patients underwent esophagoscopy, and in each, mild anastomotic narrowing with distal grade II (ulcerative) esophagitis and free gastroesophageal reflux were seen. Two of these patients have required surgical reconstruction of the esophagogastric junction to control reflux and its complications. The first of these, a 16-year-old female, had required yearly esophageal dilatations for recurrent anastomotic narrowing. Her acid regurgitation and heartburn with postural aggravation were refractory to medical therapy. Intraoperative dilatation of her anastomotic stricture and a combined Collis gastroplasty and Belsey hiatal hernia repair have eliminated her dysphagia and symptoms of reflux for more than one year.

The second patient, a six-year-old female, had required esophageal dilatations at three to four month intervals since repair of her esophageal atresia, and despite sleeping in an almost sitting position, experienced recurrent bouts of nocturnal regurgitation of gastric contents and pneumonitis. Although she was

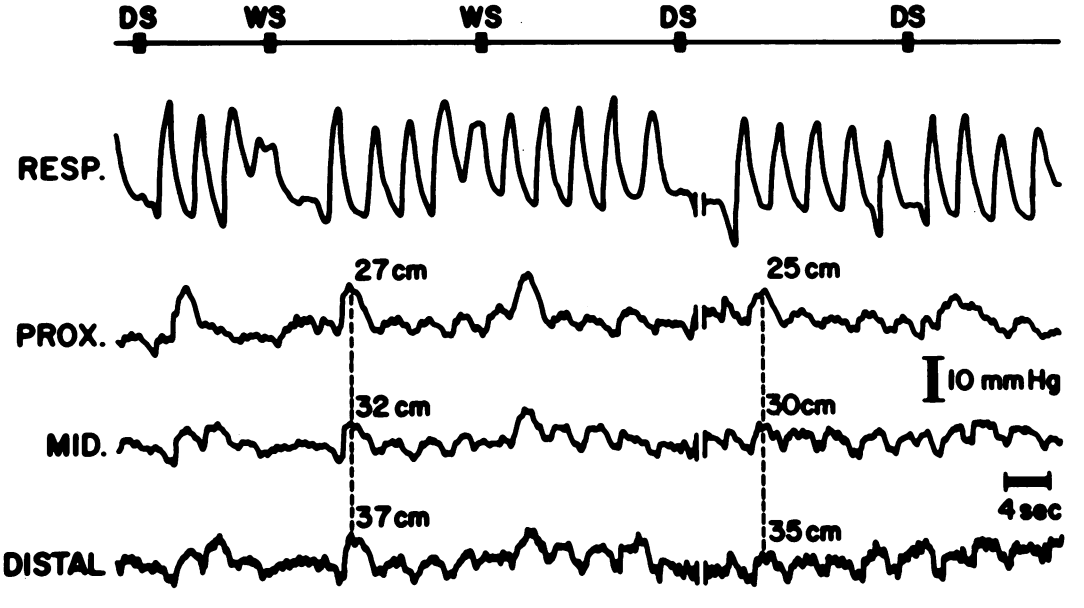

FIG. 2. Aperistalsis in esophageal atresia patient. There is lack of effective peristalsis with swallowing, contractions being weak and simultaneous. (DS = dry swallow; WS = wet swallow.)

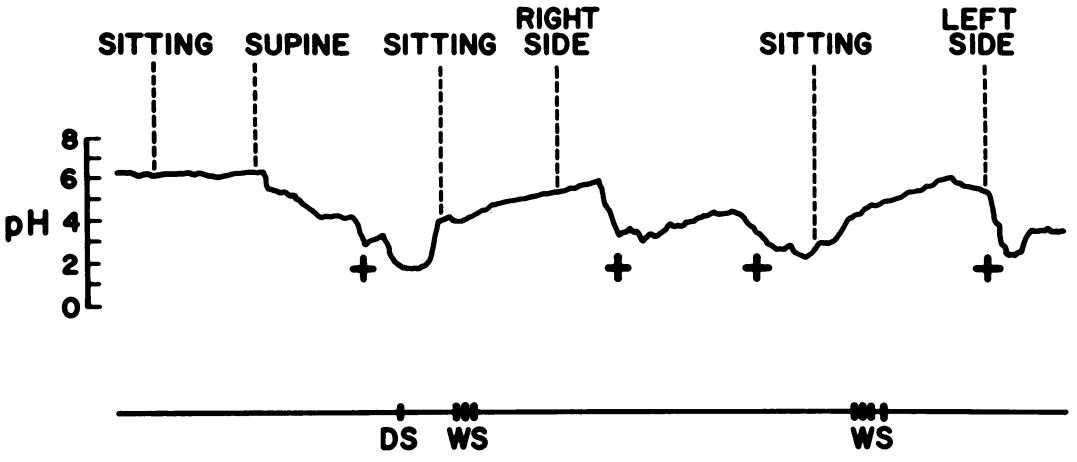

FIG. 3. Strongly positive (3+) acid reflux test in esophageal atresia patient. Gastroesophageal reflux (+), indicated by drops in intraesophageal pH below 4, occurs whenever the patient lies supine, on the right side or on the left side. (DS = dry swallow; WS = wet swallow; HPZ = high pressure zone.)

unable to relate experiencing substernal pain, she was terrified of lying supine, and would immediately clasp her hands over her epigastrium and vomit in this position. Now five months since her hiatal herniorraphy, this girl sleeps supine and uninterrupted throughout the night, with only occasional dysphagia for meats.

The third patient with endoscopic ulcerative esophagitis, a 27-year-old female who has required numerous esophageal dilatations of an anastomotic stricture, has moderate symptoms from gastroesophageal reflux, but has refused operative intervention at this time.

Discussion

In 1957, Haight noted abnormal motor activity of the esophagus distal to the anastomosis in patients with repaired esophageal atresia and tracheoesophageal fistula.[12] Following this, reports emerged confirming radiographically demonstrable abnormalities of esophageal motility in children who had survived repair of esophageal atresia with or without tracheoesophageal fistula. Cine-esophagograms typically demonstrate lack of a coordinated esophageal peristaltic stripping wave, either throughout the entire esophagus or beginning at the level of the anastomosis.[7,10,20,21]

TABLE 4. *Relationship Between Degree of Distal Esophageal Mobilization and the Presence and Extent of Gastroesophageal Reflux (12 patients)*

Length of Distal Mobilization (cms.)	No. of Patients with Degree of Reflux		
	0–1+ (none–mild)	2+ (moderate)	3+ (severe)
1–1.5	3	0	1
2–2.5	1	1	0
3–3.5	1	1	2
4 or more	0	0	2

Manometric studies in children and young adults who have had repair of esophageal atresia and TEF have defined the abnormality in esophageal motor function further. Progressive peristalsis is lacking, and esophageal contractions tend to be simultaneous, repetitive, and of either weak or normal amplitude.[22,27] The aperistaltic segment of esophagus may extend both proximal and distal to the level of the anastomosis.[5] Pressures in both esophageal sphincters have been normal. Although Lind and associates[22] reported failure of relaxation with deglutition of the distal HPZ in these patients, this observation has not been substantiated in either our laboratory or by the results of others, who have found manometrically normal-appearing lower esophageal sphincter function.[5,27]

Our manometric studies demonstrated lack of a distal HPZ in five (23%) of our patients and in the remaining patients, HPZ pressures approximately one-half those seen in asymptomatic young adult control subjects. In every patient with a demonstrable esophageal HPZ however, normal reflex relaxation with deglutition occurred. All but one of our patients manifested the typical motility disorder which has been documented by others.

In 18 patients, aperistalsis was evident throughout the entire esophagus, and in three it was limited to the distal two-thirds. One of our patients showed normal amplitude, progressive peristalsis throughout the esophagus, and this was the only patient who demonstrated normal acid clearing ability. As other investigators have found, upper esophageal sphincter pressures and motor function were normal in all of these patients.

Gastroesophageal reflux in adults is associated with a decrease in distal HPZ pressure.[30,31] The detection of moderate to severe gastroesophageal reflux with the intraesophageal pH probe in 13 (59%) of our patients correlated well with their abnormally low HPZ pressures, and emphasizes that the manometric demonstration of a distal esophageal HPZ does not necessarily indicate competence of the lower esophageal sphincter mechanism. The fact that gastroesophageal reflux was seen radiographically in only two patients and a hiatus hernia in only one in this series is not surprising. The limitations of the barium swallow examination in documenting gastroesophageal reflux are increasingly apparent.[2] Further, it is now known that gastroesophageal reflux can occur in the absence of a radiographic hiatus hernia.[15]

The cause of gastroesophageal reflux in these patients is a matter of conjecture at the moment, as is the debate on whether the esophageal motility disorder in these patients is a congenital abnormality or a result of vagal denervation of the esophagus at the time of repair of the atresia. Astley described "the development of gastro-esophageal incompetence due to operative traction on the cardia" in infants following repair of atresia and TEF.[1] Cameron Haight was concerned about the subsequent development of this complication and warned against routine mobilization of the distal esophageal segment to the level of the diaphragm "because of the possibility of producing an iatrogenic hiatus hernia."[13] Our data support this view and suggest that there is an increased incidence of subsequent gastroesophageal reflux in patients requiring mobilization of three centimeters or more of their distal esophageal segment for the primary anastomosis.

Gastroesophageal reflux is commonly seen in infants, but most tolerate this without difficulty and become symptom-free as they "outgrow" their reflux.[6,8,11] It has been well documented, however, that some infants and children may develop complications of gastroesophageal reflux that require operative intervention for their control.[18,26] After repair of esophageal atresia, gastroesophageal reflux in infants may be a more important factor influencing the patient's subsequent clinical course than has been appreciated generally. Chrispin and associates found radiographic sliding hiatal hernias and gastroesophageal reflux in 12 of 14 children with recurrent bouts of dysphagia and aspiration pneumonia following repair of esophageal atresia and TEF.[7] Holden and Wooler reported that five of their 33 surviving patients had hiatal hernias during their first six months of life.[16] At the time of their review, one was asymptomatic, one had no symptoms but had a pronounced hernia on barium swallow, one had a hiatal hernia which produced esophagitis, one had developed a peptic stricture, and one had died at six years of age from bronchiectasis. Several authors have suggested that resistant esophageal anastomotic strictures and recurrent respiratory infections in these infants and children may be perpetuated by gastroesophageal reflux and successfully treated with antireflux operations.[9,18,25]

The ability of the esophagus to clear refluxed acid back into the stomach, thereby limiting the duration of contact between the acid and the esophageal mucosa, is thought to play an important role in prevention of esophagitis.[4,28] Due to the lack of progressive peristalsis which characterizes the esophageal motility disturbance in patients with esophageal atresia, it was not surprising that 21 out of 22 of our patients demonstrated abnormal acid clearing ability. The implication of this combination of gastroesophageal reflux and impaired acid clearing ability is an increased incidence of complications secondary to gastroesophageal reflux–esophagitis and pulmonary symptoms. The precise role that gastroesophageal reflux plays in infants who, after operation, develop anastomotic strictures and pulmonary complications cannot be determined with certainty. But the documentation of so high an incidence of gastroesophageal reflux in young adults surviving repair of esophageal atresia can only lead to the assumption that the condition has been present since infancy.

It would seem prudent, therefore, beginning in the immediate postoperative period, to initiate accepted medical therapy for control of reflux in these infants, specifically, virtual constant upright posturing and small, frequent feedings of thickened formula.[7] As the children grow, elevation of the head of the bed at night should be routine. It must be remembered that these children have not experienced a period of normal esophageal function against which they can compare their symptoms and report them as being abnormal. When asked if they swallow normally, the response will almost always be "yes". But when questioned specifically about dysphagia for meat or bread, acid or food regurgitation, or nocturnal respiratory symptoms, what was reported as "normal" esophageal function often becomes clearly abnormal.[21]

Persistent or recurrent respiratory complications, bouts of severe dysphagia and failure to grow normally in these patients after six to nine years of age should not be attributed arbitrarily to their esophageal motility disorder. Patients in this age range rarely show more than minimal anastomotic narrowing on their barium esophagogram, and an atonic or poorly contracting but uninflamed esophagus should serve as an adequate conduit for most foods. It is noteworthy that of the five patients in this series with no esophageal symp-

toms, four had no demonstrable gastroesophageal reflux and one had only minimal reflux. An intensive effort to exclude gastroesophageal reflux, including a barium study with fluoroscopy, intraesophageal pH reflux testing, and esophagoscopy may be the only means of instituting proper therapy in these patients.

We believe that whenever possible, every effort should be made to avoid hiatal hernia repair in infants and children. The long-term success rate of antireflux operation in children, particularly those with esophagitis and stricture, is not high.[23] And unlike adults, with conscientious medical management, symptoms of reflux in infants may be well-controlled and disappear completely as distal HPZ competence is gained with age. There will nevertheless be instances, as in our six-year-old female who required hiatal herniorrhaphy, in which life-threatening bouts of recurrent aspiration or impaired nutrition leave little choice other than operative intervention. We have been impressed, however, that both of our patients who had antireflux operations had moderate to severe periesophagitis and esophageal shortening which prevented a tension-free reduction of the esophagogastric junction well below the diaphragm. Mediastinal fixation of the midesophagus secondary to the initial repair of esophageal atresia also prevented cephalad mobilization to the aortic arch. In adults, with esophageal shortening from peptic esophagitis, the esophagus-lengthening Collis gastroplasty procedure in combination with either Belsey or Nissen fundoplication, has provided an excellent means of achieving a tension-free antireflux operation without the need to place sutures in the inflamed distal esophagus.[24] This approach has not been used extensively enough in children to determine if the gastroplasty tube will grow adequately with the patient.

Conclusions

The esophageal motility disorder present in infants and children after repair of esophageal atresia persists into adult life. Characteristically, there is loss of an effective primary peristaltic stripping wave and simultaneous, weak or normal amplitude esophageal contractions after swallowing. In addition, moderate to severe gastroesophageal reflux secondary to incompetence of the lower esophageal sphincter mechanism, usually without a radiographic hiatus hernia, is present in many young adults (59% in this series) with this condition. While gastro-esophageal reflux in these patients may be the result of a congenital abnormality of lower esophageal sphincter function, it is more common in those who require three centimeters or more of distal esophageal mobilization for their primary anastomosis. The combination of gastroesophageal reflux and impaired acid clearing ability due to the lack of effective peristalsis may be at least partially responsible for the high incidence of pulmonary complications and anastomotic strictures in these children. With age, reflux esophagitis may become increasingly severe.

Those who care for these patients in infancy and childhood must be diligent about maintaining an antireflux regimen. As these patients become adults, careful long-term evaluation for evidence of esophagitis is warranted. It cannot be assumed that these patients ever "outgrow" their abnormal esophageal function, and we are obligated to warn both parents and, later the patients themselves, of the potential hazards of gastroesophageal reflux and its complications they may face.

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Discussion

DR. ALEXANDER H. BILL (Seattle, Washington): I think this is a very worthwhile study, and it brings, then, the question of: How does one provide the best function for these children?

I have thought (and I have never been able to prove it) that this may go back to the anatomy that we see in the abnormality. As one looks to these, you see that the vagus comes down, and it then jumps the gap between the upper esophagus and the position of the lower esophagus by going along the trachea and then coming down and putting its fibers in on the medial side of the lower esophagus. I have felt that the key to good function may be in leaving these nerve fibers undisturbed.

In order to leave as much innervation as possible, it requires leaving undissected the medial side of the lower esophagus, because that's where the vagal fibers come in to the esophageal ganglia, from where they go to the esophagus. This is based on the possibility that the function of the esophagus is by reflex to the vagal fibers, rather than intrinsic, as in the bowel.

This is borne out by their figures, which show that the longer the dissection, the worse the function, suggesting that these lower fibers may have been damaged.

I would like to express appreciation for this very valuable study, and to ask their opinion of whether the vagal anatomy may be at the base of the problem.

DR. MARK B. ORRINGER (Closing Discussion): Dr. Bill has raised a question that has pervaded the literature regarding esophageal atresia for a number of years: the argument as to whether the functional abnormality of the esophagus in these patients is a congenital abnormality or is an acquired condition related to dissection of the vagus nerves at operation.

There is no easy solution to this problem. However, some detailed cine-fluorographic studies from the British literature have documented abnormal esophageal motor function in children without atresia but with small tracheoesophageal fistulae not diagnosed until age four and seven years of age. This suggests a congenital abnormality of esophageal motility in these patients in whom operations had not yet been performed.

On the other hand, although many of these patients have gastroesophageal reflux through an incompetent lower esophageal sphincter, in all of the patients that we studied, normal reflex relaxation of the lower esophageal sphincter mechanism with swallowing was identified. It is difficult to understand why in a congenital abnormality of esophageal function, one aspect of reflex esophageal activity should be preserved, while another, peristalsis, is lost.

Regardless of the cause of the impaired peristalsis in these patients, gastroesophageal reflux is of more clinical significance, not only in terms of feeding and pulmonary problems in infancy, but also because of the potential for esophagitis in childhood and adult life.