

# Esophageal Function in Esophageal Atresia

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*In order to determine the spectrum of esophageal dysfunction in repaired esophageal atresia, 14 patients were evaluated with esophageal manometry, intraluminal pH recording, and radiology. Nine patients had no difficulty in swallowing but six had symptoms suggestive of gastroesophageal (GE) reflux. On pH recording, six had evidence of GE reflux. Basal sphincter pressure was 22 mm Hg in both reflux and nonreflux patients. No patient had manometric evidence of peristalsis in the proximal esophagus, but six had peristalsis in the distal esophagus. On radiology all had a normal peristaltic stripping wave in the cervical esophagus, and peristalsis was absent in the proximal thoracic esophagus in all patients but present in the distal esophagus in five of the 10 patients studied. Esophageal dysfunction is present in all patients with repaired esophageal atresia even when symptoms are absent.*

Abnormalities of esophageal peristalsis and of lower esophageal sphincter (LES) function have been reported in children with repaired esophageal atresia, either with or without an associated tracheoesophageal fistula (1-8). Many children surviving repair of an esophageal atresia frequently have symptoms related to esophageal motor dysfunction including regurgitation, vomiting, heartburn, dysphagia, and chronic respiratory symptoms such as recurrent pneumonia and nocturnal wheezing (4-9). Several studies show a correlation between abnormal esophageal manometric findings and esophageal symptoms, but in some instances the manometric instrumentation was suboptimal (1-3, 5-7). Our aim in this study was to evaluate esophageal motor

function using high-fidelity manometric instrumentation in a cohort of patients with repaired esophageal atresia and to correlate the manometric findings with roentgen findings.

## MATERIALS AND METHODS

The study group consisted of 14 patients (7 boys and 7 girls), mean age 7.5 years (range 2-20) with repaired esophageal atresia. Patients were selected solely on the basis of their willingness to participate. Informed consent was obtained from the older subjects and from the parents of children. The study was approved by the Human Research Review Committee of the Medical College of Wisconsin. Eleven of the patients had a history of an associated tracheoesophageal fistula. Two patients had had a Nissen fundoplication. One had required gastric-tube esophageal replacement, and seven had required esophageal dilatations. A detailed questionnaire was completed for all patients concerning swallowing function, dysphagia, esophageal obstruction, heartburn, gastroesophageal reflux, growth rate, and respiratory infections.

The instrumentation and recording techniques for esophageal manometry have been detailed previously (10-14) with the modification that we used a 9-lumen manometric assembly with an outer diameter of 4.5 mm. This small assembly is extruded as a single multilumen tube (13), and is well tolerated by children (15). The tube has 8 circumferential recording lumens, each 0.8 mm in

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diameter and a larger central lumen 1.6 mm in diameter. A micro pH electrode of 1.5-mm diameter (Microelectrodes Inc., Londonderry, New Hampshire), when inserted through the central lumen, exited the assembly through a side hole, located 3.5 cm above a sleeve device (14) that is used to monitor the LES pressure. The sleeve device, 4 cm in length was fused to the end of the tube. When positioned straddling the LES, the sleeve device records LES pressure continuously despite axial movement of the LES along the sleeve during respiration and swallowing (14). The sleeve method improves patient cooperation because once the sleeve is located in the LES high pressure zone, further tube positioning is unnecessary. Lateral recording orifices were positioned at either margin of the sleeve assembly and at 3-, 6-, 9-, 12-, and 15-cm distances above the sleeve. During manometry each recording lumen was infused with water at a rate of 0.5 ml/min by a hydraulic pump (12).

After transnasal insertion of the manometric tube, the children were placed on a padded x-ray table. Continuous simultaneous LES pressure and intraluminal pH monitoring were performed for 15–30 min in each child. Stress maneuvers were performed in children who did not demonstrate spontaneous gastroesophageal reflux (GER) during the observation period. These stress maneuvers included abdominal compression (20–40 mm Hg), positioning 30° head down, and rolling the patient into a left lateral position. Esophageal motor activity in response to wet and dry swallows was recorded. Two of the patients had a Mecholyt<sup>R</sup> test. The complete study lasted less than 1 hr. Patients under age 5 years were mildly sedated with intravenous diazepam (15).

Although each patient had prior x-ray films available for evaluation, fluoroscopic evaluation of pharyngo-esophageal motor function was performed immediately after manometry in 10 of the subjects. Appropriate spot-films were obtained. The x-ray examinations were performed by one of two radiologists experienced in evaluating esophageal motor function. The patients were examined prone, supine, and in some instances upright. Specific attention was given to motor function of the pharynx, upper esophageal sphincter (UES), esophageal body, and LES. Five or more separate swallows of barium were evaluated in each subject. In some instances swallows were observed both during manometry recording and after removal of the manometric tube.

For purposes of data analysis, the LES pressure tracing from each patient was divided into 1-min periods. The mean LES pressure for each minute was scored by a planimetric method (16) that subtracted end-expiratory gastric pressure from end-expiratory LES pressure. The average of the minute values gave a mean value for the 10- to 15-min observation period. The high-low LES pressure range was calculated as the difference between the highest and lowest minute LES pressure values. The basal LES pressure at the onset of a reflux episode was scored as the mean LES pressure during the 10-sec interval just prior to the reflux episode. GE reflux was scored when the intraluminal pH fell 2 or more pH units for 10 or more seconds (15). The findings from manometry were correlated with the roentgen findings.

TABLE 1. RESPIRATORY SYMPTOMS IN CHILDREN WITH REPAIRED ESOPHAGEAL ATRESIA

Always asymptomatic	4
Pneumonia, 1–4 episodes	7
Nocturnal	2
Wheeze with URI	1

## RESULTS

**Clinical Findings.** All 14 patients studied had normal growth values for height and weight. Nine had no recent difficulty in swallowing, three had occasional or minor difficulties with swallowing, and two needed to eat slowly or drink excessive amounts of liquid to wash down solids. Six patients had recent clinical symptoms suggesting GER. In two children repeated episodes of regurgitation had required Nissen fundoplication which alleviated the reflux symptoms. In one child symptoms of reflux had resolved spontaneously. Symptoms suggestive of pulmonary aspiration were present for 10 of the patients (Table 1).

**Manometric Findings.** During esophageal manometry and pH monitoring, GE reflux was detected in six of the 14 patients (Table 2). Patient 7, with the gastric tube esophageal reconstruction, had numerous reflux episodes. This patient is not included in the statistics below as she did not have a true LES and most of her esophageal body was not present. Neither of the two patients who had had Nissen fundoplications demonstrated acid reflux.

The mean basal LES pressure for a 10-min monitoring period for the group of 13 patients was  $22 \pm 14$  SD mm Hg (range 1–52 mm Hg). The mean basal LES pressure for the eight patients without demonstrable GER was  $22 \pm 13$  mm Hg (range 6–52 mm Hg), whereas the mean basal LES pressure for the five patients with GER was  $22 \pm 16$  mm Hg (range 1–47 mm Hg). The LES pressures did not differ significantly between the two groups. Considerable minute-to-minute variation in the LES pressure occurred in most of the subjects.

The underlying mechanism for reflux episodes differed among the patients. The patient with the gastric-tube esophageal reconstruction had no LES, and acid was present continuously in the gastric tube. One patient had free acid reflux associated with a low basal LES pressure (1 mm Hg), and one patient refluxed only with stress maneuvers. Three patients had reflux episodes associated with transient inappropriate relaxation of the LES (15, 16). Of the six patients with demonstrable GER, three

TABLE 2

Patient	Age (yr)	LES pressure (mm Hg $\pm$ SD)	History of GER symptoms	Present GER symptoms	History of respiratory symptoms	Present respiratory symptoms	GER on pH monitoring	Comment
1	2	18 $\pm$ 6	-	-	+	+	-	
2	13	19 $\pm$ 3	+	-	-	-	-	Nissen
3	6	6 $\pm$ 6	-	-	-	-	-	
4	4	16 $\pm$ 4	+	+	+	-	-	Nissen
5	9	21 $\pm$ 2	+	-	+	+	-	
6	3	1 $\pm$ 0	+	+	+	+	+	
7	7	14 $\pm$ 0	+	+	+	+	+	Gastric tube
8	15	26 $\pm$ 5	+	-	-	-	+	
9	2	28 $\pm$ 7	+	-	+	-	+	
10	19	52 $\pm$ 7	-	-	-	-	-	
11	6	47 $\pm$ 14	-	-	+	-	+	
12	4	28 $\pm$ 7	+	-	+	+	-	
13	13	9 $\pm$ 0	+	+	+	-	+	
14	2	18 $\pm$ 10	-	-	-	-	-	
Totals			9	4	9	5	6	

were asymptomatic, three had heartburn, and two had wheezing. Of the eight patients without demonstrable GER, three had recurrent wheezing or pneumonia.

The LES relaxed normally with swallowing in 12 patients. In one patient the LES relaxed only to 15 mm Hg with swallowing and, in the patient with the gastric tube, the LES was absent.

On manometry, no patient had demonstrable peristalsis in the middle third of the esophageal body (Figure 1). However, 11 patients had simultaneous nonperistaltic contractions in the body of the esophagus after swallowing. Peristaltic contractions were recorded in the distal 2-6 cm in six patients (Figure 1). When present, these waves were coordinated both with swallowing and with LES relaxation. In the two patients given Mecholyl, no abnormal responses occurred in the esophageal body.

**X-ray Findings.** In the 10 patients who were examined fluoroscopically at the time of manometry, all demonstrated normal motor function of the pharynx and UES. The primary peristaltic stripping wave invariably appeared normal in the cervical esophagus. Because the cervical esophagus is only 1-2 cm long in children, however, peristalsis in this segment was generally missed by manometry using an assembly with recording sites spaced 3 cm apart. In all 10 patients peristaltic and nonperistaltic contractions were absent in the proximal half of the thoracic esophagus, a finding that correlated with manometry. In five of the 10 patients peristalsis was absent in the distal esophagus, whereas in the remaining five peristalsis was observed fluoroscopically in the distal third of the esophagus. These latter findings also correlated with manometry. Ar-

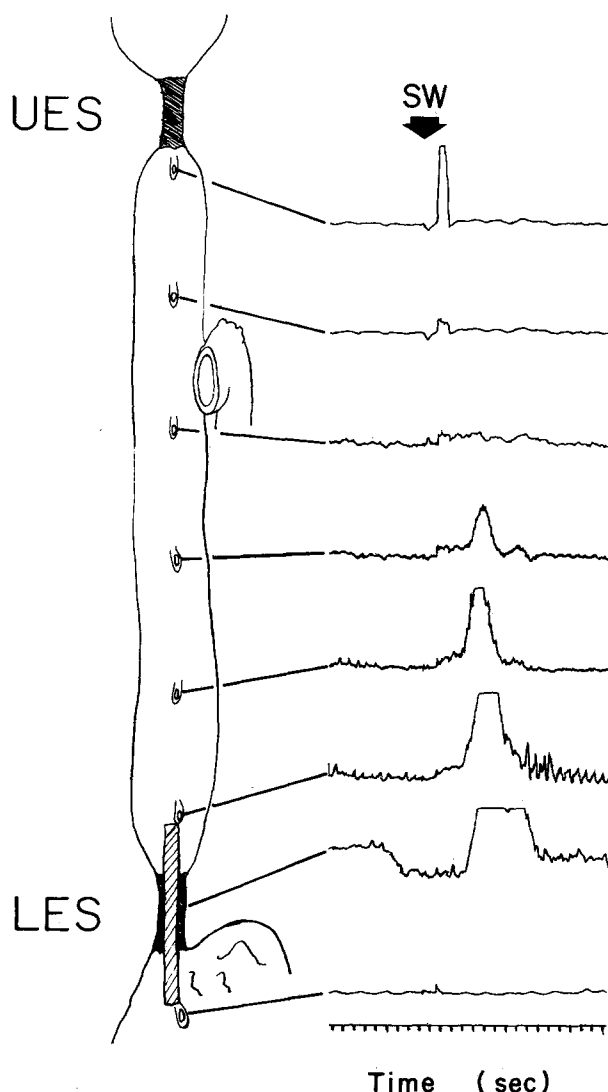
eas of aperistalsis were not cleared of barium with the patient recumbent, but drained by gravity when the patients were positioned upright. One patient demonstrated slight loss of compliance in the mid-esophagus, in the remainder the esophageal body had a normal diameter or was minimally dilated. In two patients a short segment of slight narrowing in the proximal thoracic esophagus suggested the site of surgical anastomosis. LES opening was normal in all 10 patients. One child had a small hiatal hernia, but no reflux. One patient had free GE reflux of barium.

## DISCUSSION

Our study confirms and refines previous observations of esophageal dysfunction in children with repaired esophageal atresia. All the patients had abnormal esophageal peristalsis. We also found a high incidence of LES incompetency and gastroesophageal reflux on pH monitoring. Recurrent respiratory infections, however, were not necessarily associated with demonstrable GER.

Using the sleeve sensor we found considerable minute-to-minute variation of the LES pressure in both the GER and non-GER patients. We previously have observed similar LES pressure variability in children (15) as well as in adults (16). The low basal LES pressure in some of our children without demonstrable GER confirms our previous finding in children that GER cannot be predicted reliably by sample values of LES pressure (15). There was no difference in mean basal LES pressure between the

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**Fig 1.** Manometric recording of a peristaltic sequence. A 12-year-old girl underwent repair of a T-E fistula during infancy. Six manometric recording sites, spaced at 3-cm intervals, span the entire esophageal body. The sleeve device straddling the LES records sphincter pressure. After swallowing, a peristaltic pressure wave, 80 mm Hg in amplitude is seen at the most proximal recording site, disappears at the aortic arch level, but reappears in the distal half of the esophagus. The LES functions normally.

GER and non-GER patients. The incidence of respiratory problems was similar in GER and non-GER patients.

GER was associated with transient, inappropriate LES relaxation in three of the children who demonstrated acid reflux. We have observed this mechanism of GER previously in other children with demonstrable reflux (15) and also in adults (16). This newly described mechanism of GER appears

to be associated with a substantial number of reflux episodes in all groups of patients with GER whom we have studied.

Although others have found normal esophageal motor function in some children with repaired esophageal atresia (7), all of our patients had severe esophageal motor dysfunction. Peristalsis, when identified manometrically, was found only in the distal third of the esophagus. Due to the 3-cm spacing between manometric recording sites, however, peristalsis in the short 1- to 2-cm length of cervical esophagus was shown only on fluoroscopy. Previously we have shown that peristalsis over such short segments can be shown manometrically when the recording sites are spaced 1 cm apart (17).

Because most children with GER, but without esophageal atresia, do not develop pulmonary complications from aspiration, GER alone would not likely account for the frequent history of respiratory symptoms in our patients. Probably the combination of abnormal peristalsis and GER lead to pharyngeal regurgitation and aspiration in the 10 patients with pulmonary symptoms. Some patients with respiratory symptoms had normal LES function.

Antireflux regimens assist some patients with repaired esophageal atresia accompanied by GER or respiratory complications. Most antireflux regimens include upright positioning. If aspiration were secondary to poor esophageal clearance caused by abnormal esophageal peristalsis, elevation would be expected to assist esophageal clearance. While traditional medical regimens do not always succeed in improving symptomatology, bethanechol (5) and antireflux surgery (8) are successful in selected patients.

The incidence of demonstrable GER was 57% in our patients, including the two children who had a Nissen antireflux procedure. There has been much speculation concerning the increased incidence of GER that accompanies esophageal atresia. It may be caused by the initial reparative surgical procedure; however, children with tracheoesophageal fistula, but without atresia, often have GER (8). Secondly, surgical procedures on the esophageal body of experimental animals fail to have significant effects on the LES (18). On the other hand, it has been suggested that mobilization of the distal esophagus may contribute to LES incompetency (7). Therefore, in some patients, the surgical procedure itself may cause mechanical alterations that promote GER.

The etiology of the esophageal motor dysfunction in esophageal atresia remains unclear. Although some authors have suggested that damage to the esophageal branches of the vagus is the cause of motor dysfunction (3, 9), the finding of a negative Mecholyl test suggests the motor impairment is not due to simple denervation. The esophageal muscle may be abnormal, possibly secondary to ischemia. In most patients esophageal peristalsis is absent in the proximal thoracic esophagus, but may be intact in the cervical or distal esophagus or both. Because children with tracheoesophageal fistula without esophageal atresia often have esophageal motor dysfunction prior to surgery (19), it is likely that the motor disorder is part of the congenital abnormality.

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