Management of Congenital Esophageal Stenosis

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Background/Purpose: The authors report the incidence, diagnosis, and treatment methods of congenital esophageal stenosis (CES) at their institution.

Methods: A retrospective analysis of 123 patients with tracheoesophageal anomalies in a pediatric hospital between 1980 and 1999 was performed. Charts were reviewed for patient demographics, presence of true CES, associated congenital anomalies, method of diagnosis, methods of treatment, and histopathology.

Results: Six of the 123 patients (4.9%) had true CES (one patient had 2 separate stenoses). Two patients had isolated CES, one patient had CES with isolated tracheoesophageal fistula (TEF), one patient had CES with isolated esophageal atresia (EA), and 2 patients had CES with EA/TEF. Diagnoses were made with an upper gastrointestinal (GI) contrast study in 5 patients, and one patient had one of 2 stenoses diagnosed by prenatal ultrasound and the other diagnosed intraoperatively. Four of the 7 stenoses were treated with surgical resection, and the remainder was treated with esophageal dilatation. Histopathology from the 4 resected stenoses showed tracheobronchial remnants in 3 specimens and submucosal thickening in 1 specimen.

Conclusions: Although isolated CES is rare (2 of 123 = 1.6%), CES associated with other tracheoesophageal anomalies has a higher incidence (4 of 123 = 3.25%). Patients with this lesion should be treated first with dilatation. If ineffective, resection is required.

INDEX WORDS: Congenital esophageal stenosis, tracheoesophageal fistula, esophageal atresia.

CONGENITAL esophageal stenosis (CES) is a rare yet significant entity because of its clinical manifestation, and it is also associated with tracheoesophageal fistula (TEF) or esophageal atresia (EA). The incidence of CES is estimated at 1:25,000 to 50,000 live births, and the incidence of other congenital anomalies associated with CES ranges from 17% to 33%.[1] There are 3 pathologic/histologic types of CES that have been described in the literature: (1) fibromuscular thickening, (2) cartilaginous ring, and (3) membranous web.[2] The purpose of this study is to present our experience with diagnosis, management, and treatment of this rare yet important lesion.

MATERIALS AND METHODS

A retrospective analysis was performed in a designated pediatric hospital by collecting all cases of congenital tracheoesophageal abnormalities between 1980 and 1999. IRB approval was obtained from Emory University (IRB ID 904-2001). The search resulted in 123 patients, who were all reviewed for the presence of CES. The defining criteria set by Nihoul-Fékété et al.[2] was used for those patients who had an available surgical pathology specimen. Those who did not undergo a surgical resection had their disease defined as CES based on radiologic confirmation of an intrinsic esophageal stricture not attributed to extrinsic or caustic etiologies.

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RESULTS

Six of the 123 patients (4.9%) reviewed in our series were shown to have true CES. Patient demographics and associated congenital anomalies of these 6 patients are presented in Tables 1 and 2.

Diagnosis

In all but one of the patients, the diagnosis was made by an upper GI contrast study (UGI). A prenatal ultrasound scan was performed in patient 6 that resulted in the diagnosis of an intrinsic stenosis in the midesophagus as well as, duodenal atresia, duodenal web, ventricular septal defect (VSD), and atrial septal defect (ASD). The patient underwent an operation for repair of EA/TEF, and, incidentally, a second intrinsic stenosis was found in the distal esophagus. Patients 2 and 5 had UGI studies done on day 1 of life because of intolerance to feedings, which prompted analysis. The remaining patients in the series had their studies done at later dates because of a delayed onset of symptoms. All of the CES lesions that we had found in these patients, except for one, were located in the distal esophagus.

Table 1. Patient Characteristics

Patient No.	Gender	Race	Gestational Age (wk)	Maternal Age (yr)	Birth Weight (g)
1	F	W	?	17	3,005
2	F	W	32	17	?
3	F	AA	33	19	1,150
4	F	W	34	32	2,126
5	F	A	42	35	3,120
6	F	W	38	32	2,200

Abbrevations: W, white; AA, African American; A, Asian; ?, data was not available.

Management/Treatment

For the purpose of reporting management and treatment each patient is presented individually.

Patient 1 at first underwent an esophageal dilatation with Maloney dilators. This initial dilatation session resulted in esophageal perforation confirmed by UGI. This was repaired with a 1-cm esophageal resection, end-to-end anastamosis, Nissen fundoplication, and pyloromyotomy because of transection of the anterior vagus with resection of the esophageal segment. The patient then underwent numerous esophageal dilatations and steroid injections secondary to an anastamotic stricture that later developed.

Patient 2 had numerous esophageal dilatations for the first 15 years of life. Because of unresolving dysphagia to solid foods, the patient eventually underwent a resection of the CES. Afterward, the patient had to undergo further dilatations with steroid injections because of the development of an anastamotic stricture.

In patient 3, the stricture was noted first at 55 days of life by UGI. Endoscopy showed a tight stenosis with no evidence of reflux esophagitis. Six separate esophageal dilatations had been performed, and there still was no radiographic resolution of the stenosis.

Patient 4 had an anastamotic stricture after repair of EA/TEF. This stricture, as well as the more distal intrinsic stenosis was noted on UGI postoperatively at 11 days of life. The patient underwent 2 esophageal dilatations, the second of which resulted in a distal esophageal perforation. The esophagus was repaired with primary closure of the perforation. Post operatively, the patient still showed evidence of the distal CES by UGI and no evidence of reflux. A formal pH-probe study or endoscopy was not performed on this patient. Because of the early diagnosis of the lesion, it was attributed to a congenital etiology.

Patient 5 underwent numerous esophageal dilatations that were ineffective in resolving the distal CES. The patient eventually underwent surgical resection of the stenosis.

Patient 6 had 2 intrinsic stenoses as previously described. The midesophageal lesion was resected at the time of EA/TEF repair, and the second distal lesion was not resected. Postoperatively, the patient underwent 3 dilatations of the distal stenosis.

Histopathology

Patients 1, 2, and 5 showed evidence of tracheobronchial remnants including cartilage, early columnar metaplasia, a disorganized/hyperplastic muscular layer, and ectopic glandular tissue. The mid esophageal stenosis that was resected from patient 6 showed a strictured lumen with scarring and a thickened submucosa consistent with fibromuscular thickening.

DISCUSSION

In the 19 years reviewed in our series of congenital tracheoesophageal abnormalities, only 6 patients were found with true CES. The numbers in this series are comparable with the other major series that have been reported in the last 20 years (Table 3). Thus, it can be said that this lesion is rare, yet one that we need to be made aware of because of the association with other congenital anomalies and the complexity of treatment.

In our experience, all of the patients with an intrinsic stenosis were first treated with attempted esophageal dilatations. In terms of definitive treatment, 3 of the 7 stenoses required surgical resection because of tracheobronchial remnants. These specific patients (1, 2, and 5) had ineffective esophageal dilatations, and one (patient 1) had a perforation caused by dilatation. Patient 3 had numerous dilatations that have yet to be effective in resolving symptoms. It is speculated that this patient has a tracheobronchial remnant causing the stenosis, and surgical resection actually has been planned for the near future. The majority of the other series (Table 3) reviewed as well as the results of this study show that the best treatment for a tracheobronchial remnant type of CES is with surgical resection.[2-5,8]

Three of the 7 stenoses (42.9%) were treated solely with esophageal dilatation. It is assumed, as other studies have done (Table 3), that the lesions treated effectively with dilatation fall under the category of fibromuscular

Table 2. Additional Congenital Anomalies Found With CES

Patient No.	Additional Congenital Anomalies
1	None
2	None
3	EA, distal TEF, APW, ASS, LSVC to coronary sinus, left pelvocaliectasis, L6 hemivertebra, right inguinal hernia
4	EA, distal TEF, tracheal malacia
5	H-type TEF
6	EA, duodenal atresia, duodenal web, VSD, ASD

Abbreviations: APW, aorto-pulmonary window; ASS, atrial situs solitus; LSVC, left superior vena cava; VSD, ventricular septal defect; ASD, atrial septal defect.

Table 3. Comparison of Recent CES Series

CES Study	No. of Patients	Method of Treatment	Histopathology*
Nishina et al (1981)[3]	2	Surg res (2/2)	Trach-br rem (2/2)
Nihoul-Fékété et al (1987)[2]	20	Esoph dil (9/20)†; surg res (11/20)	Trach-br rem (4/20); mem diaph (6/20); fib-mus thick (1/20)
Neilson et al (1991)[4]	6	Esoph dil (3/6); surg res (3/6)	Trach-br rem (3/6)
Murphy et al (1995)[5]	3	Surg res (3/3)	Trach-br rem (1/3); fib-mus thick (2/3)
Newman and Bender (1996)[6]	18	Esoph dil (6/18)‡; surg res (2/18)§	Fib-mus thick (2/18)
Sarihan and Abes (1997)[7]	3	Esoph dil (2/3); surg res (1/3)	Mem diaph (3/3)
Diab et al (1999)[8]	6	Esoph dil (2/6); surg res (4/6)‖	Trach-br rem (1/6); fib-mus thick (1/6)¶

Abbreviations: esoph dil, esophageal dilatation; surg res, surgical resection; trach-or rem, tracheobronchial remnant; mem diaph, membranous diaphragm; fib-mus thick, fibromuscular thickening.

*This is a report of only pathology or surgically confirmed diagnoses.

†One of the 9 underwent surgical drainage caused by dilatation perforation.

‡Five of the 6 had an esophageal perforation secondary to dilatation; 3 were treated with surgical repair or drainage, and the remaining 2 were treated medically.

§One of these 2 patients underwent surgical resection due to dilatation perforation.

‖One of 4 had an esophageal perforation secondary to dilatation, which was treated conservatively, and the patient later underwent surgical resection.

¶The 2 other patients who had surgical resection had pathology reported as "normal."

thickening.[2,4] Pathologic analysis of this lesion is not as widely available as the other types because of the infrequent resection of this lesion. However, it is reasonable to attribute the stricture that is either diagnosed soon after birth, those that show no endoscopic evidence of reflux esophagitis, or those that have no radiographic evidence of external stricture to fibromuscular thickening. We propose that fibromuscular thickening is somewhat of a diagnosis of exclusion.

We did not have any patients with a confirmed membranous diaphragm as the cause of CES. However, some of the CES lesions treated effectively with dilatation may actually fall under this category. Sarihan and Abes[7] showed in their series that 2 of the 3 membranous diaphragm type CES were treated successfully with esophageal dilatation. Because this is a membranous and partially obstructing lesion, dilatation techniques should be used first to treat this form of CES as is used for the fibromuscular type of lesion.[1]

This series of 6 patients shows that CES is a lesion worth considering when faced with symptoms of dysphagia and foreign body trapping in infants after TEF/EA repair or in infants in whom these symptoms develop without the presence of other anomalies. In those patients that undergo repair of TEF/EA soon after birth, close attention should be paid to the postoperative UGI, which can reveal this lesion. The decision to resect at the time of surgery for the other anomalies, to resect at a later age, or to treat with conservative dilatation therapy should be guided by clinical suspicion for the histologic type. As can be seen in our series, resection is not always a curative option because of the development of postresection strictures in 2 patients. Additionally, dilatation is not without complication. Newman and Bender[6] reported perforation in 6 of 7 patients treated with dilatation. Special care must be taken to provide the most effective therapy for the patient when faced with this particular lesion.

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