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DISEASES OF THE ESOPHAGUS

Original article

Congenital esophageal stenosis associated with esophageal atresia

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SUMMARY. **Congenital esophageal stenosis (CES) is a rare clinical condition but is frequently associated with esophageal atresia (EA). The aim of this study is to report the diagnosis, management, and outcome of CES associated with EA. Medical charts of CES-EA patients from Lille University Hospital, Sainte-Justine Hospital, and Montreal Children's Hospital were retrospectively reviewed. Seventeen patients (13 boys) were included. The incidence of CES in patients with EA was 3.6%. Fifteen patients had a type C EA, one had a type A EA, and one had an isolated tracheoesophageal fistula. Seven patients had associated additional malformations. The mean age at diagnosis was 11.6 months. All but two patients had non-specific symptoms such as regurgitations or dysphagia. One CES was diagnosed at the time of surgical repair of EA. In 12 patients, CES was suspected based on abnormal barium swallow. In the remaining four, the diagnostic was confirmed by esophagoscopy. Eleven patients were treated by dilation only (1–3 dilations/patient). Six patients underwent surgery (resection and anastomosis) because of failure of attempted dilations (1–7 dilations/patient). Esophageal perforation was encountered in three patients (18%). Three patients had histologically proven tracheobronchial remnants. CES associated with EA is frequent. A high index of suspicion for CES must remain in the presence of EA. Dilatation may be effective to treat some of them, but perforation is frequent. Surgery may be required, especially in CES secondary to ectopic tracheobronchial remnants.**

KEY WORDS: **children, congenital stenosis, esophageal atresia.**

INTRODUCTION

Congenital esophageal stenosis (CES) is a rare clinical condition defined as a fixed intrinsic narrowing of the esophagus present at birth. The incidence is approximately 1 in 25 000–50 000 live births.[1] It is often associated with other malformations including esophageal atresia (EA), chromosomal anomalies, cardiac anomalies, intestinal atresia, and anorectal malformations.[2] In the largest case series, the association of CES and EA ranges from 3% to 14%.[3–6] Diagnosis of CES associated with EA is difficult, and treatment may be delayed. Three subtypes of CES have been described: ectopic tracheobronchial remnants (TBR), segmental fibromuscular stenosis, and membranous stenosis. Different treatments have been proposed in regard of the subtype of CES, but this still remains controversial.

The goal of this study is to report the diagnosis, management, and outcome of CES associated with EA.

METHODS

Medical charts from Lille University Hospital (n = 180), Sainte-Justine University Health Centre (n = 165), and Montreal Children's Hospital (n = 132) were retrospectively reviewed. Data were obtained from the patient's clinical, radiologic, and surgical charts. From 1990 to 2012, 17 patients were diagnosed with CES associated with EA. CES may have been diagnosed: (i) during the first surgery; (ii) when the esophagogram showed persistent smooth and segmental narrowing of the esophagus with proximal dilatation the anastomosis and the gastroesophageal

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junction; or (iii) endoscopically. In all cases, no evidence of an acquired cause of stenosis was found. Agreement to be part of a multicenter database of EA patients was obtained through the consent of each child's parents or primary caregiver.

RESULTS

Overall, the incidence of CES in patients with EA was 3.6%.

Characteristics of the patients

The characteristics, management, and outcome of the 17 patients are summarized in Table 1. There were 13 boys and 4 girls. Fifteen patients (88%) had a type C EA, one (6%) had a type A EA, and one (6%) an isolated tracheoesophageal fistula (type E). Seven patients had associated additional malformations: two had trisomy 21, four had VACTERL (association of at least 3 of the following malformations: vertebral, anorectal, cardiac, tracheal, esophageal, renal and limb), and two had ventricular septal defect. The mean age at diagnosis was 11.6 months, ranging from 1 day to 60 months. All patients but two had non-specific symptoms before the diagnosis of CES, including regurgitations, dysphagia, respiratory problems, and growth retardation. One patient presented with esophageal food impaction (Fig. 1).

Diagnosis

One CES was diagnosed at the time of surgical repair of the EA because of inability to pass an 8 Fr. tube through the distal esophagus to the stomach during surgery.

In 12 patients (71%), CES was suspected on abnormal barium swallow (Figs 1,2). In seven (41%), the CES was detected on the first postoperative barium swallow 7–10 days after the initial surgery. In five (30%), the diagnosis of CES was delayed and found on a second or third esophagogram. In these patients, the retrospective review of the previous esophagograms showed clearly the CES indicating a misinterpretation and a low index of suspicion for a possible associated CES.

In the remaining four (23%), systematic barium swallow was unable to show the CES, and the diagnostic was made by esophagoscopy (Fig. 3).

The CES was localized distal to the esophageal anastomosis in all patients.

Management and outcome

Dilatation of the CES was attempted as a first-step treatment in all patients.

Ten patients (59%) were treated successfully by dilatation only (one to three dilatations per patient). Of these 10 patients, six had hydrostatic dilatation with balloon, and four had dilatation by bouginage. In these patients, esophageal perforation was encountered in two patients: One was following bouginage and was treated medically without any further complication. The other patient was dilated during EA surgery at 2 days of life and needed to be redilated at 15 months. A perforation occurred following pneumatic dilation without further complication after medical treatment.

Six patients (35%) underwent surgery (resection and anastomosis) because of failure of dilatations (hydrostatic dilatation, four patients; bouginage, four patients – one to seven dilatations per patient) due to persistent and protracted symptoms or perforation in one patient. Failure of disappearance of waist under fluoroscopy was predictive failure of dilatation in three out of four patients who underwent hydrostatic dilatation (Fig. 4).

Three patients had histologically proven ectopic TBR on resected specimen, and all three had surgery following failed dilatations. One patient who underwent surgery after esophageal perforation following a dilatation by bouginage had membranous stenosis proven on resected specimen.

DISCUSSION

Although CES is rare in the general pediatric population, its association with EA is frequent ranging between 3% and 14% in the largest series.[3–6] Therefore, in all children operated for EA, a high index of suspicion is required in order to detect as early as possible an associated CES to prevent the occurrence of symptoms and possible complications such as anastomotic leaks, respiratory problems, dysphagia, food impaction, and failure to thrive.[5]

As reflected by the age at diagnosis (mean 11.6 months, range 1 day–60 months), the diagnosis of CES can be difficult. Clinicians and radiologists must be aware of the possible association with EA. Moreover, the characteristics of the patients presenting with the association of CES with EA are not specific. There is no predictive clinical feature that can suggest the presence of a CES, although we found a slight predominance of males (13/17; 75%) and two patients with trisomy 21 as reported in other case series.[4–6] It is noteworthy that CES can be found in patients with isolated type C EA without any other associated anomalies. The symptoms (regurgitations, dysphagia, and respiratory problems) found in children with CES are also frequent in EA patients, are not specific, and are the same than those found with an anastomotic strictures. Thus, because in EA patients no particular symptom or clinical feature can suggest the presence of a CES, it is essential to interpret all imaging studies with a special attention to CES.

Table 1 Characteristics of the 17 patients with congenital esophageal stenosis (CES)

Patient no	Gender	Type of esophageal atresia	Associated anomalies	Age at diagnosis	Diagnostic exam	Primary treatment	Outcome
1	M	C	None	14 months	Barium swallow	Dilatation (balloon)	Failure of dilatation Surgery
2	M	C	None	9 months	Barium swallow	Dilatation (balloon)	Failure of dilatation Surgery
3	M	A	VACTERL, imperforated anus	9 months	Esophagoscopy	Dilatation (balloon)	Success
4	M	C	None	2 days	Surgery of EA	Dilatations (#1 bougienage during surgery, #2 balloon at 15 months)	Perforation (during second dilation at 15 months) Success
5	F	C	VACTERL	27 months	Barium swallow	Dilatation (balloon)	Success
6	M	C	Trisomy 21	13 months	Barium swallow	Dilatation (balloon)	Success
7	M	C	None	8 days	Barium swallow	Dilatation (balloon)	Failure of dilatation Surgery
8	F	C	VACTERL, duodenal atresia	16 months	Esophagoscopy	Dilatation (balloon)	Success
9	M	C	None	22 days	Barium swallow	Dilatation (bougienage)	Success
10	M	C	VACTERL, imperforated anus	16 months	Esophagoscopy	Dilatation (balloon)	Failure of dilatation Surgery
11	M	C	None	1 month	Esophagoscopy	Dilatation (bougienage)	Failure of dilatation Surgery
12	M	C	None	1 day	Barium swallow	Dilatation (bougienage)	Success
13	M	C	None	33 months	Barium swallow	Dilatation (bougienage)	Success
14	M	C	None	60 months	Barium swallow	Dilatation (bougienage)	Success
15	F	C	None	8 days	Barium swallow	Dilatation (bougienage)	Perforation during dilation #3 Surgery
16	M	C	VSD	7 days	Barium swallow	Dilatation (bougienage)	Perforation during dilation #2 Success
17	F	D	Trisomy 21 VSD	27 months	Barium swallow	Dilatation (balloon)	Failure of dilatation Surgery

EA, esophageal atresia; VACTERL, association of at least 3 of the following malformations: vertebral, anorectal, cardiac, tracheal, esophageal, renal and limb; VSD, ventricular septal defect.

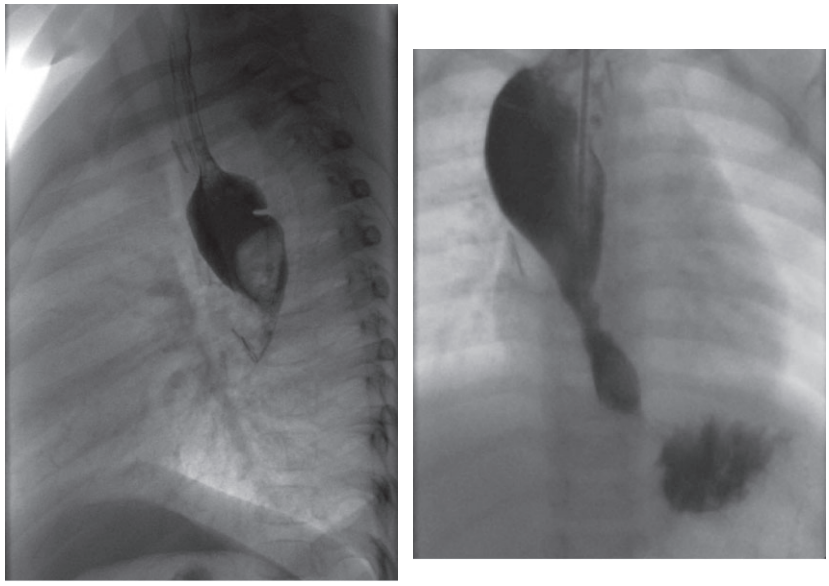

Fig. 1 Patient 2. Boy 9 months, esophageal atresia (EA) type C. (Left) Food impaction; (right) esophagogram after disimpaction showing the congenital stenosis. Dilations (balloon) were attempted but failed. The patient was operated. Histological analysis showed tracheobronchial remnants.

Barium swallow was diagnostic of CES in 12 out of the 17 patients. However, it is noteworthy that in five of them, more than one radiological examination was required because the previous ones either missed the diagnosis or were misinterpreted. Indeed, mild CES can be interpreted as transient spasm, dysmotility, or esophageal narrowing due to reflux. Moreover, in four patients because esophagogram was not informative, the CES was diagnosed with esophagoscopy. This shows that a normal barium swallow does not exclude the possibility of a CES, and additional investigation should be pursued if clinical suspicion is present.[6]

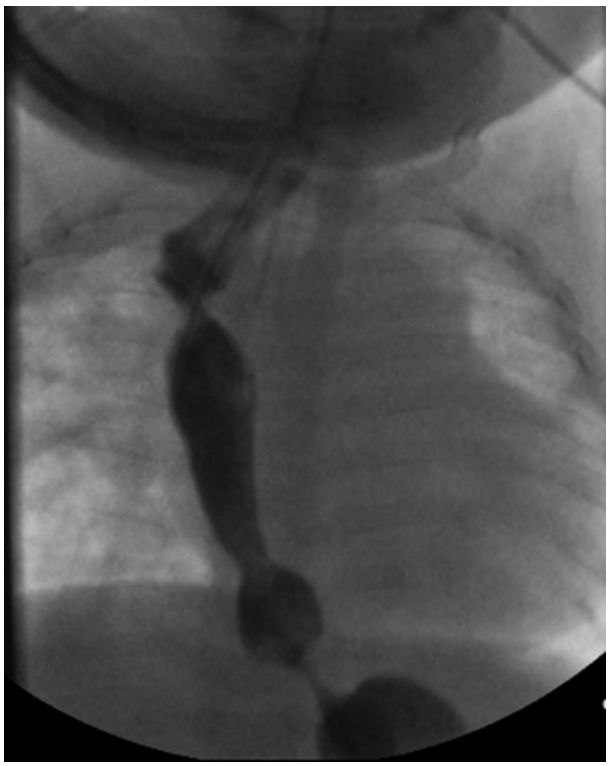

Fig. 2 Patient 3. Boy 9 months, esophageal atresia (EA) type A, VACTERL (association of at least 3 of the following malformations: vertebral, anorectal, cardiac, tracheal, esophageal, renal and limb). Anastomotic stricture and congenital esophageal stenosis (CES). The latter was successfully dilated with complete improvement after one single balloon dilation.

All patients were initially treated by esophageal dilatation using bouginage or balloon according to the expertise of each center. As reported by others,[1] dilatations were successful in the majority of the cases (10/17), but failures occurred in six patients for whom the stenosis was related to TBR. Perforation was frequent and occurred in 3/17 (18%) patients. This is in keeping with other large case series in which the incidence of perforation ranged from 5% for Yoo *et al*.[6] to 11% for Romeo *et al*.,[1] 33% for Newman *et al*.,[4] and 44% for Kawahara *et al*.[5] irrespective of the technique of dilatation. Two patients who suffered from perforation underwent dilatation using bouginage, and the other one had hydrostatic balloon dilatation. It would be interesting to address this issue in future studies to further clarify the advantages and disadvantages of each method.

Three causes of congenital anomaly of esophageal wall architecture have been described: ectopic TBR, segmental fibromuscular stenosis, and membranous stenosis.[7] In this study, three patients had ectopic TBR, and one had membranous stenosis proven on resected specimen histopathological analysis. The other 13 patients had no biopsy performed. Of the three patients having ectopic TBR, 100% of them underwent failure of treatment after multiple

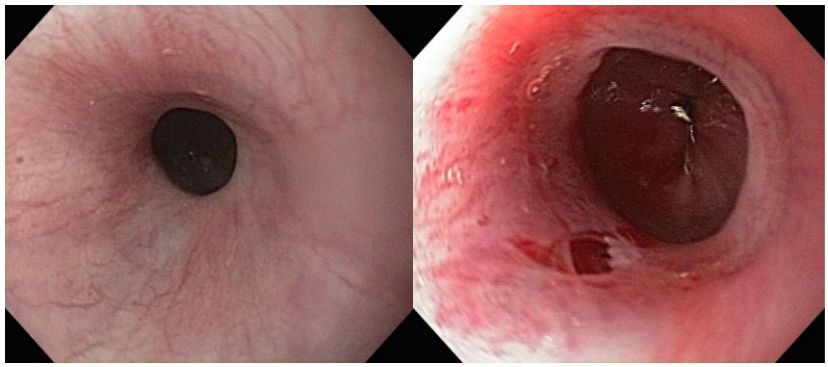

Fig. 3 Esophagoscopy showing the congenital esophageal stenosis (CES) before (right) and after (left) pneumatic dilation.

dilatations and ultimately had surgery. The only patient with membranous stenosis also had surgery after esophageal perforation following the second dilatation. Studies have suggested that endoscopic ultrasonography may help to demonstrate ectopic TBR.[1,8,9] We propose that patients with known ectopic TBR or patients who had failure after 2–3 dilations with failure of the waist to disappear on fluoroscopy (Fig. 4) should be considered for surgery because this type of CES tends to require surgical treatment as shown in this study and other studies.[3,6,8–12]

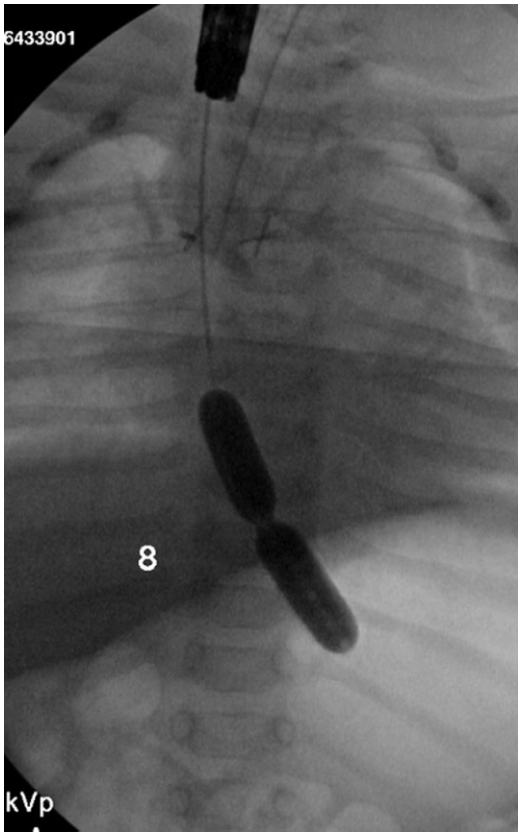

Fig. 4 Hydrostatic dilatation under fluoroscopy showing failure of disappearance of waist.

CONCLUSION

CES associated with EA is frequent. A high suspicion index for CES must remain in the presence of EA. It can be diagnosed at the time of EA repair or on the first postoperative esophagogram. Dilatation may be effective to treat some of them, but perforation is frequent. Surgery can be required, especially in CES secondary to ectopic TBR and is also indicated after failure of a program of dilatations.

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