

Thoracoscopic management of congenital esophageal stenosis secondary to tracheobronchial remnant in pediatric patients

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ABSTRACT

Introduction. Congenital esophageal stenosis (CES) is an extremely rare pathology in children, with an incidence of 1/25,000-50,000 live births. According to its histopathological classification, there are three types of CES: fibromuscular hyperplasia, membranous diaphragm, and tracheobronchial remnants.

Clinical case. We present the clinical case of a 39-month-old male patient diagnosed with CES secondary to tracheobronchial remnants, with multiple vomit and reflux episodes since he was 4 months old. He was admitted at the emergency department with respiratory distress. An upper GI endoscopy and an esophagogram were initially carried out. Stenosis resection and thoracoscopic esophageal anastomosis were performed.

Conclusions. Tracheobronchial remnants are the second most common presentation of congenital esophageal stenosis. They can be managed through dilatations or surgery according to etiology.

KEY WORDS: Congenital esophageal stenosis; Thoracoscopy; Histology; Treatment.

MANEJO TORACOSCÓPICO EN ESTENOSIS ESOFÁGICA CONGÉNITA SECUNDARIA A REMANENTE TRAQUEOBRONQUIAL, EN POBLACIÓN PEDIÁTRICA

RESUMEN

Introducción. La estenosis congénita de esófago es una patología extremadamente rara en niños, con una incidencia de 1/25.000-50.000 nacimientos. Según la clasificación histopatológica se encuentran tres tipos: hiperplasia fibromuscular, diafragma membranoso y remanentes traqueobronquiales.

Caso clínico. Se presenta un caso clínico de un paciente masculino de 39 meses con diagnóstico de estenosis congénita del esófago secundario a remanentes traqueobronquiales, que presentó múltiples episodios de vómito y reflujo desde los 4 meses del nacimiento. Ingresó en el Servicio de Urgencias por presentar signos de dificultad

respiratoria, realizándosele estudios iniciales de endoscopia de vías digestivas altas y esofagograma. Se practicó resección de estenosis y anastomosis esofágica toracoscópica.

Conclusiones. Los remanentes traqueobronquiales son la segunda causa de presentación de la estenosis esofágica congénita. El manejo de esta patología puede ser de dos formas, ya sea por medio de dilataciones o quirúrgico, y la elección de una u otra va a depender de su etiología.

PALABRAS CLAVE: Estenosis congénita del esófago, toracosopia, histología, tratamiento.

INTRODUCTION

Congenital esophageal stenosis (CES) is a rare pathology globally, with an incidence of 1/25,000-50,000 live births. Even though sex-related prevalence has not been demonstrated, some studies have revealed it is more frequent in male, white patients⁽¹⁻⁴⁾.

There are three types of CES according to histological findings: fibromuscular hyperplasia (FH) – the most common one, accounting for 54% of cases –, tracheobronchial remnants (TR) – 30% –, and membranous diaphragm (MD) – 16%^(2,5). Treatment can be conservative – using endoscopic pneumatic dilatation – or surgical^(3,6).

According to the literature, the use of either technique is determined by the type of CES^(1,3). We present our experience in the minimally invasive management of an infant diagnosed with CES secondary to tracheobronchial remnants.

CASE REPORT

This 39-month-old male patient was admitted at Fundación San Vicente Children's Hospital, in Medellín (Colombia). He had been referred from Chocó Colom-

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Date of submission: February 2020 *Date of acceptance:* October 2020

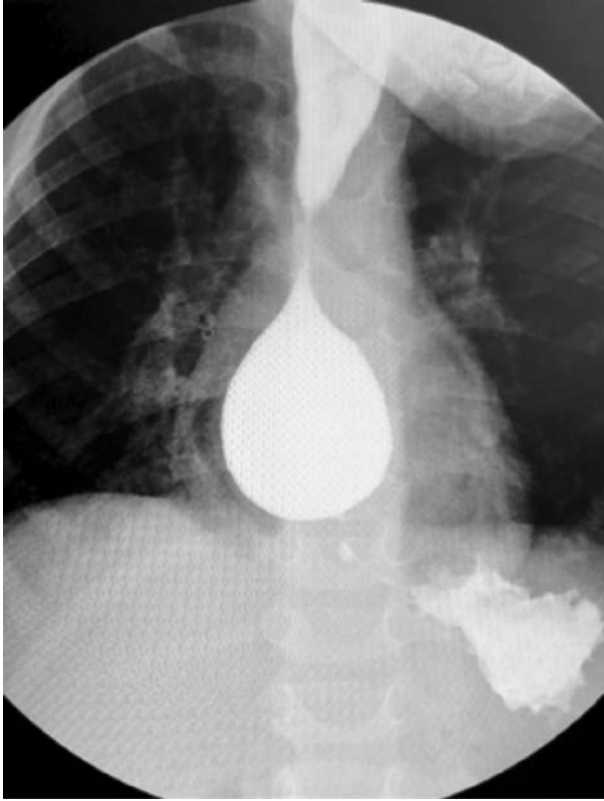


Figure 1. Esophagogram: esophageal narrowing.

bian department with 4-day respiratory distress associated with multiple episodes of emesis starting at 4 months of age. In addition, he had been suffering from solid-liquid dysphagia since he was 18 months old, and he also had reflux – predominantly at night – and poor weight gain for his age. His relatives said no previous clinical studies had been performed prior to hospitalization as a result of their religious beliefs.

They provided the results of an upper GI endoscopy (UGIE) which demonstrated 80% stenosis of the lower

third of the esophagus. Therefore, an esophagogram was carried out, which revealed esophageal distal dilatation above the gastroesophageal junction (Fig. 1). A new UGIE was performed at our institution, which demonstrated a pinpoint esophageal stenosis of approximately 25 cm at the level of the upper dental arch (UDA) (Fig. 2), so pneumatic dilatations were suspended. Given the suspicion of cartilaginous etiology, surgery was decided upon. A three-port video-thoracoscopy was carried out with the patient in a right lateral position and selective intubation of the right bronchial artery? vein? The first 5mm port was placed at the confluence of the sixth intercostal space and the left anterior axillary line, with a 5 mmHg pneumoperitoneum and a 2 l/min flow. The 3mm ports were placed at the confluence of the fifth intercostal space and the posterior axillary line, and at the confluence of the eighth intercostal space and the posterior axillary line. The distal esophagus was dilated, and an 18 Fr dilator was introduced orally down to the stenotic site. The lower pulmonary ligament was freed until the left inferior pulmonary vein was identified. An incision was carried out in the mediastinal pleura, with identification of the distal esophagus, which was subsequently isolated and differentiated with a ligation for circumferential dissection and repair purposes. The resection area was marked using diathermy. A 1cm esophageal segment was resected using the scissors and submitted for pathological examination. Anastomosis was achieved using separated Polyglyconate (Maxon™- Covidien) 5-0 stitches, and a 10Fr feeding tube was introduced before completing the anterior side of the anastomosis. The pleural cavity was then aspirated. A 14Fr thoracostomy tube was left in place and fixated with 3-0 nylon, leaving a water seal (Fig. 3). On postoperative day 7, an esophagogram was carried out (Fig. 4), demonstrating contrast medium passage and no narrowing. Oral feeding was resumed, and the patient was discharged. One month later, a follow-up assessment was performed, with the parents confirming the diet had been observed, and no episodes of vomit had been recorded. The patient also had gained weight. A con-

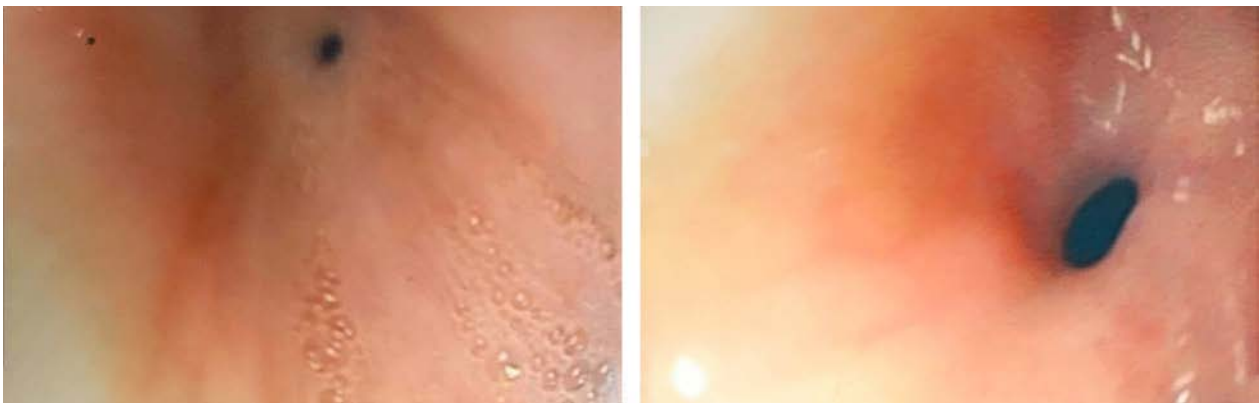


Figure 2. Upper GI endoscopy: 80% stenosis of the lower third of the esophagus.

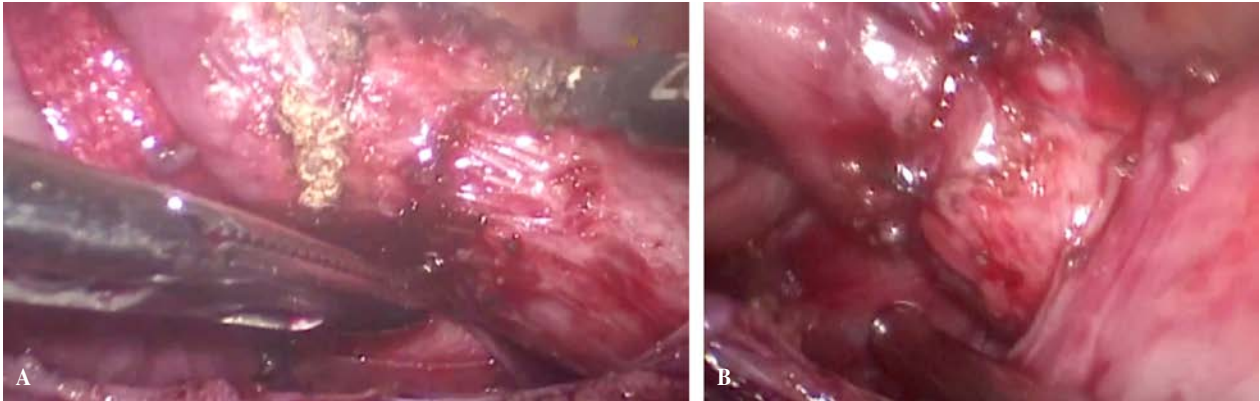


Figure 3. Video-thoracoscopy. A) Remnant resection. B) Esophageal anastomosis.

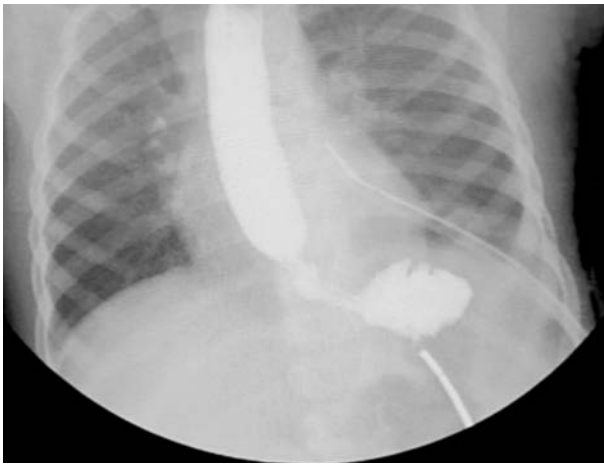


Figure 4. Postoperative esophagogram, with adequate passage of the intestinal lumen.

tol digestive endoscopy was carried out on postoperative month 3. It demonstrated esophageal permeability and a slight narrowing 25 cm away from the dental arch, without any dilatation required. The patient has not visited our institution any more as the family lives in a secluded rural area. Over the phone, they said the child has not suffered from dysphagia for the time being.

DISCUSSION

CES was first described by Rosi in 1826, in a patient with MD. In 1828, Abel reported the first treatment with rubber tube dilatation⁽¹⁾. In 1936, Fredy and Dusch described the first case of TR in the autopsy of a 19-year-old female patient initially diagnosed with achalasia^(1,2).

This malformation occurs in the first three months of pregnancy as a result of an incomplete division between the primitive bowel and the respiratory tract^(1,2,7), causing

a permanent intrinsic narrowing of the esophageal circumference^(1,3). Patients are typically asymptomatic in the first months of life. At month 6, once solid feeding has been initiated, they start having vomit, reflux, and/or dysphagia, the latter being the most frequent symptom^(2,3,6,8). Less prevalent signs and symptoms include laryngeal stridor, malnutrition, repeated aspiration pneumonia, development disorders, and increased salivation^(2,9). This pathology is associated with gastrointestinal malformations in 33% of cases, with esophageal atresia being the most common one, described in 3-14% of cases^(1,3,10).

In case of suspicion of CES, clinical signs and imaging studies allow differential diagnoses to be ruled out. Imaging tests include esophagogram, which demonstrates stenosis grade and location. UGIE helps confirm esophageal narrowing is not secondary to esophagitis, achalasia, corrosive intake, or acid-peptic disease. PH measurement and manometry allow gastroesophageal reflux and endoscopic ultrasonography (EU) to be ruled out^(2,4,5,11). However, institutions should verify whether they have the resources required for this.

EU is the technique of choice for differentiating purposes between TR and FH, which are the most common causes of CES^(5,12). According to Keita Terui et al., differentiation allows esophageal wall perforation rate to be reduced by 16.5% during treatment⁽⁵⁾. Diagnosis is confirmed by histopathology^(2,13), which demonstrates cartilaginous, glandular, and/or epithelial tissue, either alone or combined, 3 cm away from the cardias, when etiology is TR^(1,2). In our patient, esophageal stenosis was secondary to TR as demonstrated by pathological examination.

Treatment varies according to patient etiology^(1,8). Non-surgical management is based on balloon dilatations⁽⁶⁾ and is recommended in cases where there is no cartilaginous etiology⁽¹⁾, owing to the greater risk of perforation or mediastinal abscess^(14,15). In a study published in 2012, F. Martin et al. described that some patients with stenosis as a result of corrosive intake benefit from the use of self-expandable prostheses in the presence of narrowings longer

than 2 cm (long stenoses), with tortuosity and recurrences following management with dilatations⁽¹⁶⁾. However, each cause should be individualized, since this will change the therapeutic algorithm used^(1,8). Surgery is indicated in TR cases, after dilatation failure or esophageal perforation^(1,5). It is aimed at identifying and resecting the stenotic defect, followed by an end-to-end anastomosis⁽¹⁾. Nowadays, CES is typically managed through conventional surgery, but minimally invasive surgery has demonstrated to be more beneficial, since it allows for earlier oral feeding resumption and better esthetic results^(1,14), as demonstrated in our patient.

CONCLUSION

Congenital esophageal stenosis (CES) is a rare pathology in the pediatric population, with tracheobronchial remnants being the second most common presentation. CES management varies according to etiology. Non-surgical management by means of dilatations can only be applied in cases without tracheobronchial remnants – otherwise, CES should be dealt with operatively. Other causes for surgery include dilatation failure and esophageal perforation. Nowadays, minimally invasive surgery stands as a feasible technique as it provides with numerous benefits in terms of pain reduction, operating site infection, intra-operative bleeding, and hospital stay.

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