

Per oral endoscopic myotomy in a pediatric patient with achalasia

Miotomía endoscópica por vía oral (POEM) en un paciente pediátrico para tratamiento de la acalasia esofágica

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Abstract

Introduction: Achalasia is the most common primary motor disorder of the esophagus. Its reported incidence is low, even more in pediatric patients. Laparoscopic Heller myotomy is the current standard of treatment. During the last years, per-oral endoscopic myotomy (POEM) has been positioned as a safe and effective therapeutic alternative as the Heller procedure for esophageal achalasia. **Objective:** To describe the POEM technique and report the first pediatric case in our country. **Clinical Case:** 11-year-old patient, previously healthy, who presented with progressive dysphagia for solids and liquids and weight loss. The study concluded a type II achalasia. The patient underwent a POEM and had a postoperative course without incidents. One year after the intervention, symptomatic, endoscopic and manometric resolution have been documented. **Conclusions:** The described case is the first POEM in a pediatric patient in our country. Esophageal achalasia is uncommon in pediatrics and POEM has demonstrated clinical success and safety comparable to laparoscopic Heller myotomy in short and medium term. Long-term follow-up will determine its definitive role in the treatment of pediatric patients with esophageal achalasia.

Keywords:

Esophageal achalasia;
POEM;
Heller myotomy;
therapeutic endoscopy

Introduction

Achalasia is the most common primary esophageal motor disorder. It is a rare disease characterized by the absence of esophageal peristalsis and incomplete or absent lower esophageal sphincter relaxation¹.

Its reported incidence is low, in pediatric patients it is estimated annually at 0.11 per 100,000 inhabitants, without predilection by race or gender² and the average age at diagnosis is 10 years³.

The most common symptoms are dysphagia, regurgitation, retrosternal pain, and vomiting after feeding⁴. It can be associated with weight loss, failure to thrive⁴, and even appear atypically with cough or recurrent pulmonary infections secondary to aspiration⁵.

Partly because of the limited number of pediatric patients with achalasia, the standard of treatment has been controversial³. The traditional management includes endoscopic balloon dilation and laparoscopic Heller myotomy with or without partial fundoplication⁶. The botulinum toxin injection, use of calcium channel blockers or long-acting nitrates are less effective therapies in the long term and are probably considered as a bridge to surgery or palliation in those patients that definitive surgical management is contraindicated⁷.

In 2007, Pasricha described the endoscopic myotomy technique in a porcine model⁸, and in 2010, Inoue reported the first results in humans⁹. Since then, the POEM (peroral endoscopic myotomy) has been installed as a new therapeutic alternative in the achalasia management⁷. The accumulated experience in adults has shown results that position it as a safe technique and as effective as Heller's myotomy. The procedure has been gradually introduced in the pediatric population, with initial results⁷.

The objective of this report is to describe the first case, in our country, of a pediatric patient with achalasia undergoing POEM and his follow-up to date.

Clinical case

11-year-old male patient with history of allergic rhinitis and atopic dermatitis, eutrophic and with good psychomotor development. He consulted on an outpatient health center due to two-month progressive dysphagia for solids and liquids, without vomiting, heartburn or regurgitation. He has lost four kg in the last four months, without abdominal pain.

As part of the diagnostic study, an upper endoscopy was performed which was within normal limits and the esophageal biopsy was reported without alterations.

An esophageal contrast study was requested, which

identified findings compatible with esophageal achalasia (figure 1). It was complemented with a high-resolution esophageal manometry, whose report described an integrated relaxation pressure median of 36 mmHg (VN < 15 mmHg), panesophageal pressurization phenomena after multiple swallows and esophageal aperistalsis, consistent with type II achalasia according to the modified Chicago classification (figure 2).

Once the diagnosis was confirmed, the case was discussed in a multidisciplinary team and it was decided, after agreement with the parents, to resolve it by endoscopy using POEM. The preoperative Eckardt score¹⁰ was 10 points (dysphagia 3 points, retrosternal pain 3 points, regurgitation 3 points, and weight loss 1 point).

The procedure was carried out in the operating room, under general anesthesia, and in the supine position. A diagnostic endoscope with a hood, a CO₂ insufflator, and an ERBE® electrosurgical unit was used. The initial endoscopy showed food content in the esophagus, which was aspirated prior to initiating the intervention. The gastroesophageal junction (GEJ) was 38 centimeters from the dental arch.

An elevation of the mucosa was performed with a Voluven® and indigo carmine solution, then a longitudinal mucotomy was performed in the anterior face of the esophagus between the two and three o'clock positions (figures 3A and 3B), approximately 11 centimeters proximal to the GEJ. Subsequently, a submucosal tunnel (figure 3C) was created using a submucosal dissection technique and an endoscopic Flush Knife BT® of 2.5 mm, extending 41 centimeters to the dental arch. The distal extension to the GEJ was verified through endoscopic retroflexion in the gastric area to observe the subendocardial submucosa staining, added to the endoscopic measurement, identification of the oblique muscle fibers of the stomach and the vessels of the gastric submucosa.

The myotomy of the internal circular fibers of the esophageal muscle was performed with a triangle tip electrosurgical knife (TT Knife®), from 30 to 41 centimeters from the dental arch (figure 3D). Hemostasis was then assured and the tunnel was instilled with a gentamicin solution. The mucotomy was closed with endoscopic clips (figure 3E) and the final stage ensured easy passage of the instrument through the GEJ.

Complete fasting was kept until an esophageal study with water-soluble contrast was performed within the first 24 hours, which ruled out filtration (figure 4). The patient was then fed with a liquid diet, with good tolerance, and was discharged on the second day, with indication of proton pump inhibitor (PPI) in two daily doses.

The patient was controlled one week after the intervention, observing the resolution of symptoms and



Figure 1. Preoperative esophageal contrast study. A short segment stenosis with the characteristic morphology of a bird's beak, dilation of the esophagus in its middle third and filiform passage of the contrast to the gastric chamber is evident at the gastroesophageal junction.

good diet tolerance. He started eating mash and it was indicated that other foods should be progressively incorporated.

Three months after the surgery, he was controlled with an upper endoscopy, which showed a good passage of the instrument through the GEJ, absence of poor esophageal emptying or evidence of gastroesophageal reflux (GER). The high-resolution esophageal manometry showed no esophageal pressurization phenomena and an integrated relaxation pressure median of 15 mmHg. The patient remains asymptomatic, with no GER symptoms or PPI requirement. The upper endoscopy performed after one year does not show any changes compared to the previous one. His Eckardt score at follow-up is one (occasional dysphagia).

Discussion

Esophageal achalasia is a rare diagnosis in pediatrics¹¹. Available treatments include pharmacological, endoscopic, and surgical alternatives, the most commonly used is the endoscopic balloon dilation and laparoscopic Heller's myotomy. Between the latter two, the superiority of Heller's myotomy is recognized because it is related to lower recurrence rates than balloon dilation². Currently, optimal management in pediatric patients has not been defined and the literature still lacks standardized follow-up protocols⁴.

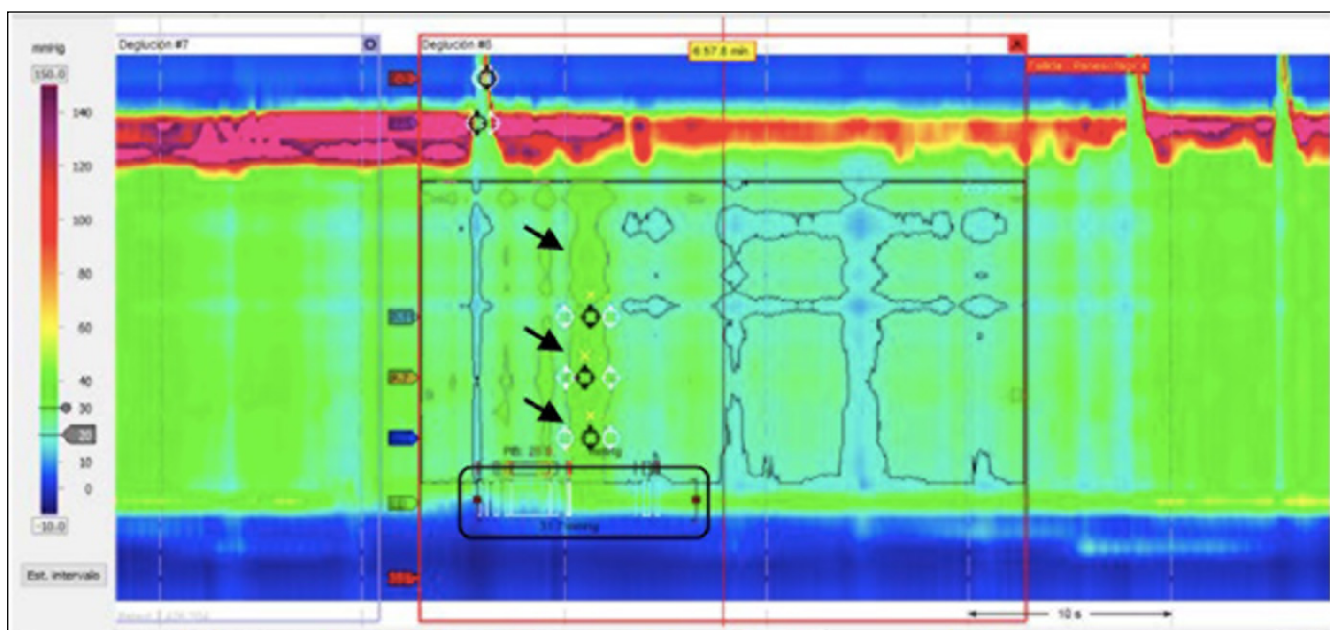


Figure 2. High resolution esophageal manometry. The absence of relaxation of the gastroesophageal junction (demarcated area) and panpressurization phenomena (arrows) can be identified in protocolized swallows.

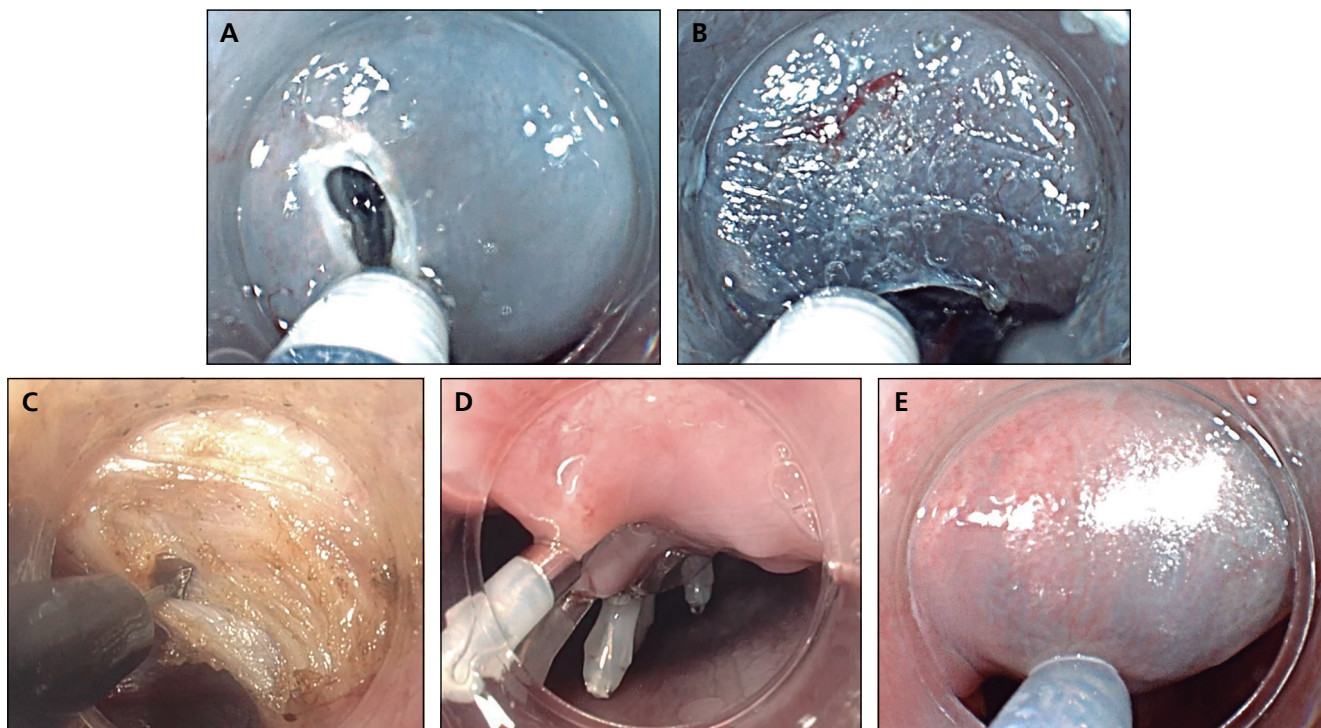


Figure 3. POEM technique. (A) Elevation of the mucosa. (B) Longitudinal mucotomy. (C) Creation of the submucosal tunnel. (D) Myotomy of the internal circular fibers of the esophagus. (E) Mucotomy closure with clips.

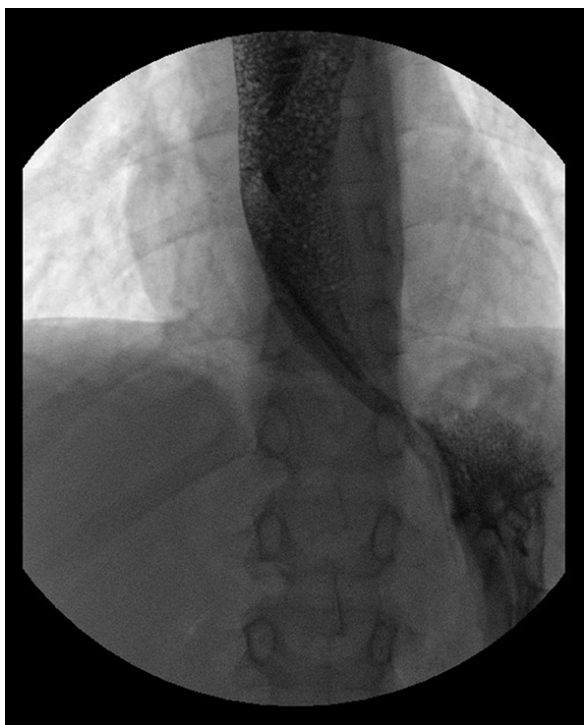


Figure 4. Postoperative esophageal contrast study. Clips are observed in the distal third of the esophagus, with contrast passage to the stomach. The area of obstruction previously described in the gastroesophageal junction is no longer visible and no extraluminal contrast, suggestive of filtration, is identified.

POEM has emerged as a competitive therapeutic alternative in the treatment of achalasia over the last decade because it combines the benefits of an endoscopic and, therefore, minimally invasive procedure with the surgical myotomy efficacy². Some authors consider in its favor that it avoids excessive dissection of the esophageal hiatus, scars and allows reintegration into activity in a few days^{12,13}, in addition to planning the length of myotomy according to the manometric and endoscopic study². The available follow-up studies report clinical success rates (Eckardt < 3) higher than 90% and improvement in the pressure profile of esophageal manometry¹³⁻¹⁵. In recent years, series have been published in pediatric patients, with good results in the resolution of symptoms, registering up to 100% at 24.6 months of the procedure, without serious perioperative complications (Clavien >III) or medium-term follow-up¹¹.

The technique has a good safety profile; capnoperitoneum and submucosal emphysema are unavoidable consequences and can be easily managed with intra-procedure if they are clinically significant^{9,16}.

The available data are not yet sufficient to establish recommendations for the technique application according to weight and age limits². The youngest patient reported corresponds to the case published by Maselli et al., a three-year-old patient with trisomy 21 and severe

malnutrition secondary to esophageal achalasia treated with POEM with success¹⁷.

It is important to recognize the learning curve associated with the procedure execution, which has been established in about 20 cases¹⁸; the best results are obtained in centers with experience in therapeutic endoscopy¹¹.

Currently, it is considered that all patients with achalasia can be treated with POEM, and it has even been established as a therapeutic alternative in other esophageal motor disorders such as diffuse esophageal spasm, nutcracker esophagus, and hypercontractile esophagus (*jackhammer*)¹¹. It would be the treatment of choice in type III achalasia because it allows myotomies of longer length and in patients with symptomatic relapse after Heller's myotomy or after a first POEM because it provides the alternative of varying the approach avoiding fibrosis zones².

While laparoscopic Heller's myotomy is currently the treatment of choice in children, POEM is increasingly used as a therapeutic alternative. It has promising results from the point of view of effectiveness and safety in pediatrics, and this report would constitute, to our knowledge, the first case made in the country. The results of the technique must be supported in multi-center studies of greater volume and with long-term follow-up, in order to constitute it as the alternative of choice in this population.

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

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Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

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