

# Per-Anal Endoscopic Myotomy as Rescue Therapy for Hirschsprung Disease After Unsuccessful Surgical Myectomy

Dervis Bandres, MD<sup>1</sup>, Carlos Prada, MD<sup>2</sup>, José Soto, MD<sup>1</sup>, Marielvis Dávila, MD<sup>1</sup>, Maria Bandres, MS<sup>3</sup>, and Victoria García, MD<sup>4</sup>

<sup>1</sup>Division of Gastroenterology and Hepatology, Department of Medicine, Centro Medico Docente La Trinidad, Caracas, Venezuela

<sup>2</sup>Division of Pediatric Surgery, Department of Surgery, Centro Medico Docente La Trinidad, Caracas, Venezuela

<sup>3</sup>School of Medicine Luis Razetti, Universidad Central de Venezuela, Caracas, Venezuela

<sup>4</sup>Department of Pathology, Centro Medico Docente La Trinidad, Caracas, Venezuela

## ABSTRACT

Hirschsprung disease (HD) may affect short, ultrashort, or long segments of the rectum. Invasive endoscopy has gained popularity in its treatment. We report a case of a 9-year-old girl diagnosed with HD at 4-year-old, at which time she underwent a myectomy. Nonetheless, because of the persistence of clinical symptoms such as bowel movements every 5–7 days and abdominal distension, per-anal endoscopic myotomy was recommended. An anterograde myotomy in the posterior wall was performed to the circular layer of the muscularis propria. At the 3-year follow-up, she has continued daily bowel movements without laxatives.

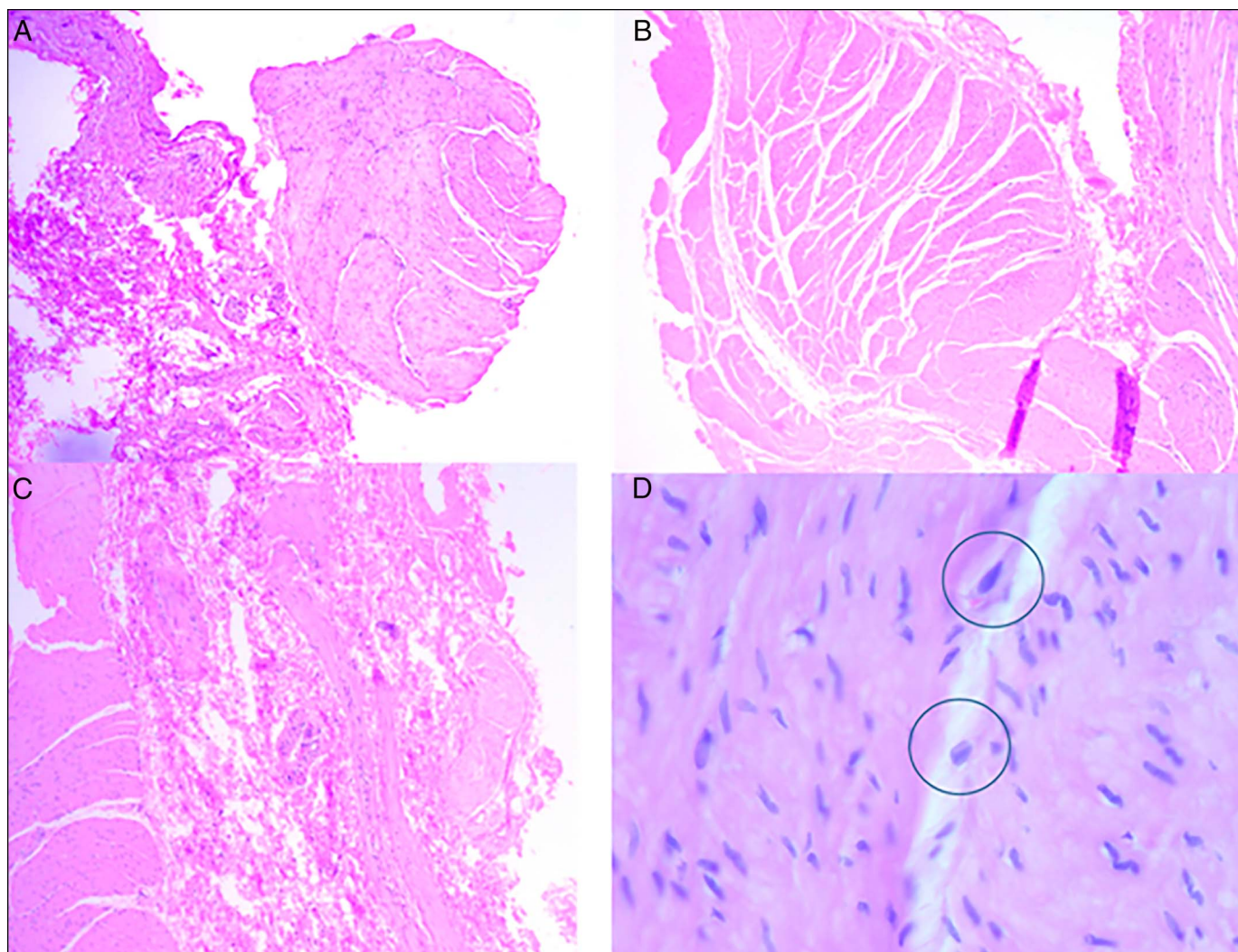
## INTRODUCTION

Hirschsprung disease (HD) is characterized by the absence of ganglion cells in the myenteric and submucosal plexuses in variable segments of the intestine. It affects the rectosigmoid region (short segment), although ultrashort and long variants have been described. Current treatments include surgical procedures;<sup>1</sup> however, endoscopy has gained popularity, given its success in treating achalasia and refractory gastroparesis. In 2016, Bapaye et al<sup>2</sup> reported the first per-rectal endoscopic myotomy (PREM) as a treatment for ultrashort HD in an adult; later in 2018,<sup>3</sup> he reported a second case in a pediatric patient. We present the first successful per-anal endoscopic myotomy (PAEM) in rescue of unsuccessful previous surgical myectomy in a patient with previous diagnosis of ultrashort HD. This is the first case reported in the Americas and the third worldwide.

## CASE REPORT

This is the case of a 9-year-old girl whose mother reported onset of symptoms in the neonatal period, presented by a decreased frequency of bowel movements (every 5–7 days) and abdominal distension, which partially improved with polyethylene glycol. At 4 years of age, she experienced an exacerbation of the symptoms despite laxatives. She was evaluated by a pediatric gastroenterologist and pediatric surgeon and was diagnosed with typical features of ultrashort HD. She subsequently underwent a 5-cm-length rectal myectomy by Lynn procedure. The pathology report described hypertrophy of the circular layer of the muscularis propria, few hypertrophic nerve fibers, and scarce myenteric ganglion cells shown in 3/55 cuts with hematoxylin and eosin stain (Figure 1). During the immediate and intermediate postoperative period, she showed considerable improvement in the rhythm and consistency of the stools. Five months after her intervention, she experienced recurrent symptoms. At age 9 years, she presented again with abdominal distention and severe constipation. This prompted her visit to our health center.

Physical examination revealed a globose abdomen, hyperactive bowel sounds, and a palpable painless descending and sigmoid colon. Laboratory work was within normal limits. A standing abdominal X-ray showed dilation of the entire colon (Figure 2). Barium

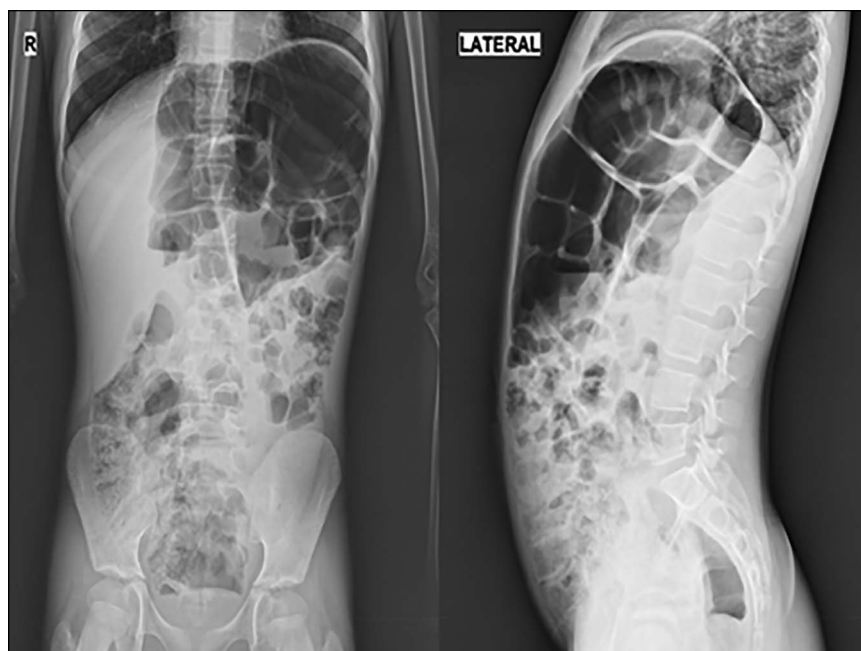


**Figure 1.** (A) Hypertrophy of circular layer of the muscularis propria (hematoxylin and eosin stain, 4× magnification). (B) Nodular disorganized arrangement of muscle fibers of the muscularis propria (hematoxylin and eosin stain, 10× magnification). (C) Hypertrophic myenteric nerve (hematoxylin and eosin stain, 10× magnification). (D) Isolated ganglion cells (circled; hematoxylin and eosin stain, 25× magnification).

enema demonstrated dilation of the colon, a transition zone, and a narrow area in the anorectum (Figure 3). Magnetic resonance imaging showed similar findings (Figure 4). The ultrashort HD diagnosis was finally made, after taking in consideration the clinical presentation, the previous pathology report, and the radiographic findings, which displayed a narrow area in the lower rectum. Two options were proposed for this patient, surgical, or endoscopic intervention; the patient's relatives opted for the latter.

The procedure was performed under general anesthesia in lithotomy position; ceftriaxone and metronidazole were administered perioperatively. Rectoscopy was performed with a gastroscope (GIF-HQ180; Olympus Corporation, Japan) (9.8 mm, channel 2.8 mm) and revealed a narrow zone 3 cm proximal to the pectinate line. The submucosa was elevated, using a mixture of 10 mL of 0.9% saline, 10-mL mannitol 18%,

and methylene blue, 1.5 cm proximal to the pectinate line. The pediatric surgeon then made a transverse incision in the posterior wall creating a small pocket that allowed for the introduction of the endoscope in the submucosal space to make a tunnel that could reach 5 cm proximal to the narrow area (Figure 5). The dissection of the submucosa was performed using the needle knife and IT Knife Catheter and EndocutQ (50W Effect 2, Vio 200D; ERBE, Tübingen, Germany). Once the tunnel was completed, a small incision was made in the muscularis propria with a needle knife to expose the longitudinal muscle layer. An antegrade myotomy of the circular muscle layer was performed with an IT Knife and EndocutQ while withdrawing the endoscope (Figure 6). The length of the myotomy was calculated on the extent of the transition zone, which according to the radiographic findings seemed to be 5 cm long. To check whether the myotomy was sufficient, the scope was withdrawn outside the tunnel to visualize the lumen,

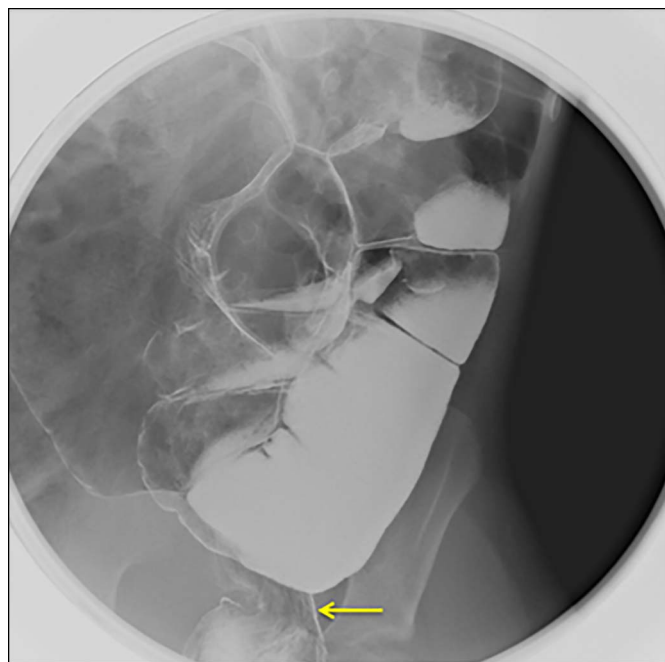


**Figure 2.** Standing abdominal X-ray before per-anal endoscopic myotomy (9yo).

mucosal integrity, and disappearance of the rectal narrowness, in combination with a digital rectal examination. Hemostasis was achieved with a Coagrasper. The incision was stitched above the pectinate area with absorbable suture (Vicryl 4-0 mm). The patient remained clinically stable and was discharged 24 hours postoperatively with oral antibiotics and soft diet for 5 days. At the 3-year follow-up, she continued to have daily bowel movements without laxatives.

## DISCUSSION

Since the first report of endoscopic myotomy in 1978 by Ortega et al,<sup>4</sup> this technique has evolved to the third space endoscopic surgery, which includes per-oral endoscopy myotomy (POEM) for esophageal achalasia,<sup>5-7</sup> G-POEM for gastroparesis,<sup>8</sup> Z-POEM for Zenker diverticulum,<sup>9</sup> and, more recently, PAEM or PREM for HD and its variants.<sup>3,4</sup>

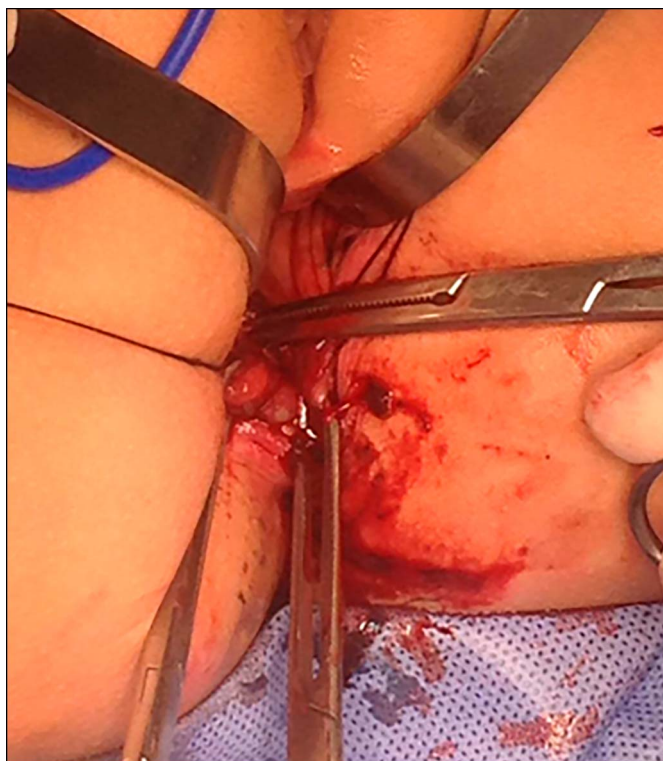


**Figure 3.** Barium enema before per-anal endoscopic myotomy showing the transition zone (arrow).



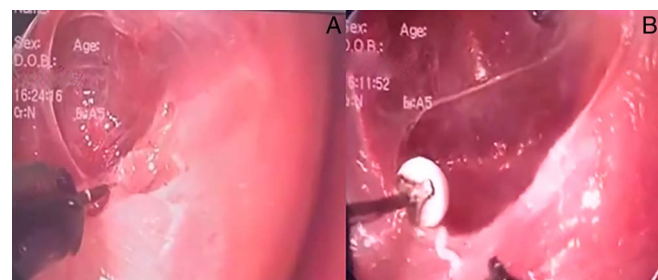
**Figure 4.** Magnetic resonance imaging before per-anal endoscopic myotomy showing the transition zone (arrow).





**Figure 5.** Transanal approach.

Two endoscopic myotomy techniques have been described for HD. Wang et al<sup>10</sup> described PAEM as a 7- to 9-cm myotomy, and Bapaye et al<sup>3</sup> defined PREM as a 20-cm myotomy, both of the rectal circular muscle layer. In our case, 2 treatment options were discussed with the parents, surgery (per-anal remyectomy or a pull-through surgery with a lower rectal resection), or endoscopic myotomy (PAEM or PREM), which was recently reported as a successful treatment for HD on naive patients.<sup>2,3</sup> The benefits of PAEM in contrast to surgery are better precision and control of the myotomy length, lower costs because of short hospital stay, less complications, and quick recovery. The challenge in our case was the possibility to encounter fibrosis caused by the previous myectomy, which could affect the



**Figure 6.** (A) Internal muscularis propria incision with needle knife. (B) Myotomy with IT-Knife.

creation of the submucosal tunnel. However, we avoided it because we made the tunnel 1 cm to the left from the previous surgical incision. The PAEM limitations are related to the newness of the technique and to the requirement of an invasive endoscopist. This technique seems to be a promising treatment for HD or its variants and also a rescue option for failed surgery.

## DISCLOSURES

Author contributions: D. Bandres wrote the manuscript and is the article guarantor. C. Prada and J. Soto edited the manuscript. M. Davila and M. Bandres reviewed the literature. V. Garcia revised the manuscript for intellectual content.

Financial disclosure: None to report.

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Informed consent was obtained for this case report.

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## REFERENCES

- Sharma S, Gupta DK. Hirschsprung's disease presenting beyond infancy: Surgical options and postoperative outcome. *Pediatr Surg Int*. 2012;28:5–8.
- Bapaye A, Waghlikar G, Jog S. Per rectal endoscopic myotomy for the treatment of adult Hirschsprung's disease: First human case (with video). *Dig Endosc*. 2016;28(6):680–4.
- Bapaye A, Bharadwaj T, Mahadik M, Ware S. Per-rectal endoscopic myotomy (PREM) for pediatric Hirschsprung's disease. *Endoscopy*. 2018; 50:644–5.
- Ortega J, Madureri V, Perez L. Endoscopic myotomy in the treatment of achalasia. *Gastrointest Endosc*. 1980;26(1):8–10.
- Nabi Z, Reddy N, Ramchandani M. Recent advances in third-space endoscopy. *Gastroenterol Hepatol (NY)*. 2018;14(4):224–32.
- Pasricha PJ, Hawari R, Ahmed I, et al. Submucosal endoscopic esophageal myotomy: A novel experimental approach for the treatment of achalasia. *Endoscopy*. 2007;39:761–4.
- Inoue H, Minami H, Kobayashi Y, et al. Peroral endoscopic myotomy (POEM) for esophageal achalasia. *Endoscopy*. 2010;42:265–71.
- Khashab MA, Stein E, Clarke JO, et al. Gastric peroral endoscopic myotomy for refractory gastroparesis: First human endoscopic pyloromyotomy (with video). *Gastrointest Endosc*. 2013;78:764–8.
- Li QL, Chen WF, Zhang XC, et al. Submucosal tunneling endoscopic septum division: A novel technique for treating Zenker's diverticulum. *Gastroenterology*. 2016;151(6):1071–4.
- Wang L, Cai Q, Fan C, Ren W, Yu J. Mo1628 A new potential method per anus endoscopic myotomy for treatment of internal anal sphincter achalasia. *Gastrointest Endosc*. 2013;77:AB451.

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