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# Ultrashort-segment Hirschsprung's disease complicated by megarectum: A case report☆

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ABSTRACT

Ultrashort-segment Hirschsprung's disease (USHD) is a rare variant of HD that is controversial in terms of diagnostic criteria and acceptable therapy. Idiopathic megarectum, also known as an intractable idiopathic constipation, is a functional disorder, which ordinarily is not surgically treated. Reported herein is a case of USHD complicated by a megarectum. The patient was an 8-year old male who presented with long history of constipation and abdominal distension. Contrast enema revealed massive dilatation of rectum, with no discernible transition zone. Absence of ganglion cells in a full-thickness rectal biopsy taken 2 cm proximal to the dentate line confirmed a diagnosis of USHD. The entire length of dilated bowel (delineated on preoperative contrast enema) was resected, using a Soave type transanal approach, with pull-through of ganglionated normal-caliber colon. In instances of USHD where megarectum prohibits use of technically less rigorous procedures (i.e., Duhamel pull-through or internal sphincter myectomy), a Soave type transanal pull-through may be a reasonable alternative.



Hirschsprung's disease (HD) usually presents in the neonatal period or at early infancy, typically with the transition zone at rectosigmoid junction. However, ultrashort-segment Hirschsprung's disease (USHD) has been known to present as a rare variant in a particular subset of patients. As indicated, the culprit aganglionic segment is ultrashort, often escaping detection by contrast enema. Not only does this delay diagnosis, but the appropriate means of surgical treatment may be equally unclear [1]. By contrast, idiopathic megacolon, also known as an intractable idiopathic constipation, is the result of stool-holding behavior. Because there is no underlying organic disorder, the latter is generally treated without surgery [2]. Reported herein is a case of USHD in an 8-year old male who presented with long-standing constipation, growth retardation, and megarectum. His condition was initially viewed as idiopathic megacolon. Optimal treatment in this setting is discussed.

## 1. Case report

An 8-year-old male presented with long-standing constipation and abdominal distension. The constipation began soon after birth, requiring intermittent enemas and laxatives. At physical examination, his height (110 cm) and weight (16 kg) both were below 3rd percentile, and his abdomen was distended. Contrast enema revealed a massively dilated rectum, without a definitive transition zone (Fig. 1A, B). To investigate potential organic disease, a full-thickness biopsy of rectum was obtained 2 cm proximal to the dentate line. Tissue examination revealed aganglionosis, establishing a diagnosis of USHD complicated by megarectum (Fig. 2A). Subsequently, a transanal pull-through procedure was elected, calling for complete resection of the megarectum and a portion of sigmoid colon (about 18 cm overall by preoperative contrast enema) in order to achieve a normal-caliber intestinal tract.

In knee-chest position, circumferential sutures were placed 2 cm proximal to the dentate line for uniform traction, and submucosal dissection began. After entering the peritoneal cavity, full-thickness dissection was undertaken to straighten the sigmoid colon for resection. Posterior myectomy was then performed, followed by bowel resection (megarectum and sigmoid colon, roughly 18 cm) and colo-anal anastomosis (Fig. 3). On intraoperative frozen



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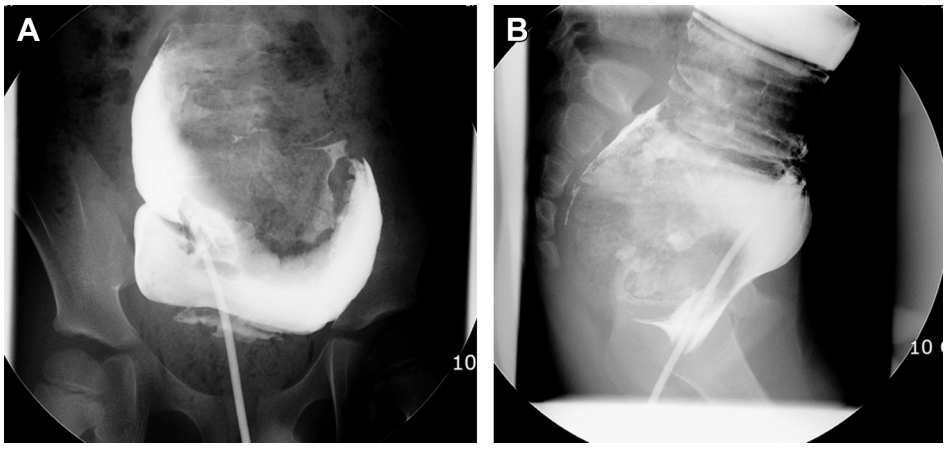


**Fig. 1.** Preoperative contrast enema: (A) massive dilatation of rectum, with huge fecaloma; (B) no discernible transitional zone on lateral view.

section, ganglion cells were identified at 3 cm proximal to the dentate line (Fig. 2B).

The patient's postoperative course was uneventful. He resumed a normal diet on Day 5, with 20 g/day polyethylene glycol 4000 (Forlax; Ahn-Gook Pharmacy Co, Seoul, South Korea) as a laxative. Fecal soiling in the immediate postoperative period soon progressed to incontinence, so the laxative was tapered. A barium enema performed 1 month after surgery showed normal colonic contours (Fig. 4). At 3 months postoperatively, the patient was having normal bowel movements and no longer relied on laxatives or enemas.

## 2. Discussion

Traditionally, the term USHD is reserved for clinical manifestations similar to those of classic Hirschsprung's disease, but this definition has long been debated, and some have even questioned or denied its validity [3]. At present, the diagnosis of USHD generally corresponds with a documented aganglionic rectal segment $<$1–2 cm [4]. In this particular patient, idiopathic megacolon was suspected initially, given the lack of a transitional zone on contrast study. However, a rectal biopsy eventually confirmed that the distal 2 cm of rectum was aganglionic.

Treatment strategies for USHD are also controversial. Some prefer simple anal myectomy, whereas others advocate conventional pull-through procedures [4]. Conservative management is effective in most instances of idiopathic megarectum and megacolon, reserving surgical interventions, such as fecal diversion, transabdominal resection of rectum and sigmoid, antegrade continence enema, and internal sphincter myectomy, for those patients whose symptoms are refractory [5–9]. Often, however, these tactics are inadequate and are not reproducible, underscoring that the dilated and hypomotile state of residual rectum is largely responsible for limited clinical improvement. Subsequently, the core management of idiopathic megarectum has now shifted to include complete excision of the megarectum and any adjacent dilated colon [10]. More recently, a Swenson type transanal and full-thickness rectosigmoid resection is advocated in this setting, the primary advantage being that complete resection of the megarectum is achieved, without cuff-related complications or need for laparotomy [11].

One may legitimately question why a primary pull-through procedure was performed upfront in this patient with USHD, rather than electing a less-invasive approach such as myectomy and a bowel management program. The patient's long history of unsatisfactory medical management led us to believe that his problem more likely represented idiopathic intractable constipation and was not simply a consequence of passive dilatation secondary to USHD. Hence, we followed suggestions of Levitt et al. [11] in devising treatment, establishing two basic objectives for cure as follows: 1) complete resection of megarectum, delineating dilated bowel on preoperative contrast enema, and 2) pull-through of normal-caliber ganglionated colon.

Internal sphincter myectomy or a Duhamel type pull-through may have been technically easier, but these methods leave some or all of megarectum in place, which was not acceptable here. A Swenson type pull-through theoretically is ideal and would have satisfied our objectives. Recent reports linking the Swenson

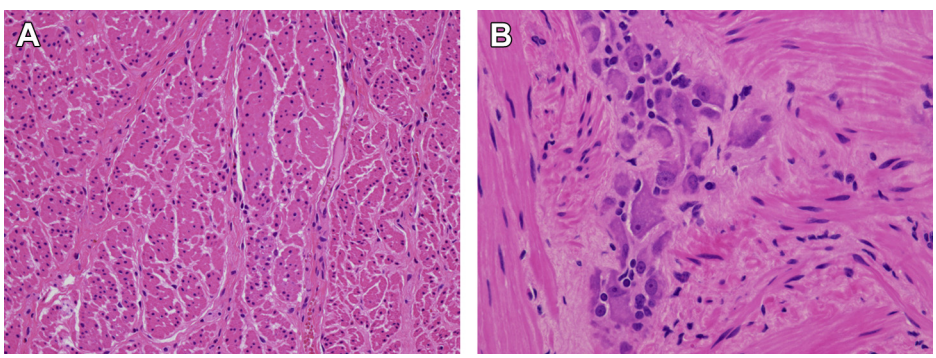


**Fig. 2.** Pathological microphotographs: (A) ganglion cells are absent at myenteric plexus (H&E, ×200); (B) ganglion cells are present (H&E, ×400).

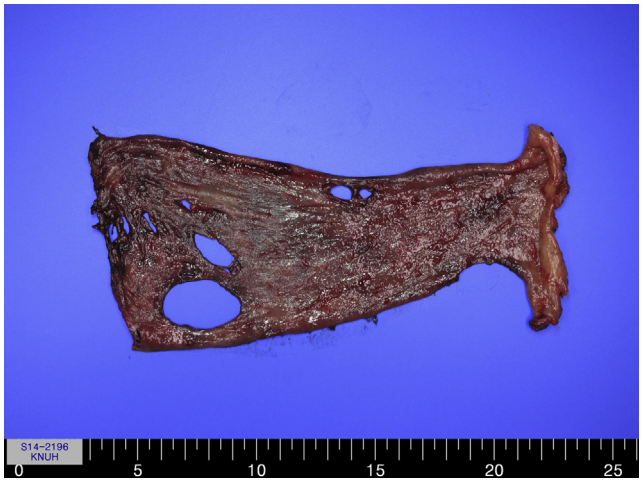


**Fig. 3.** Surgical specimen: mucosa of megarectum and full-thickness segment of sigmoid colon (lumen exposed).

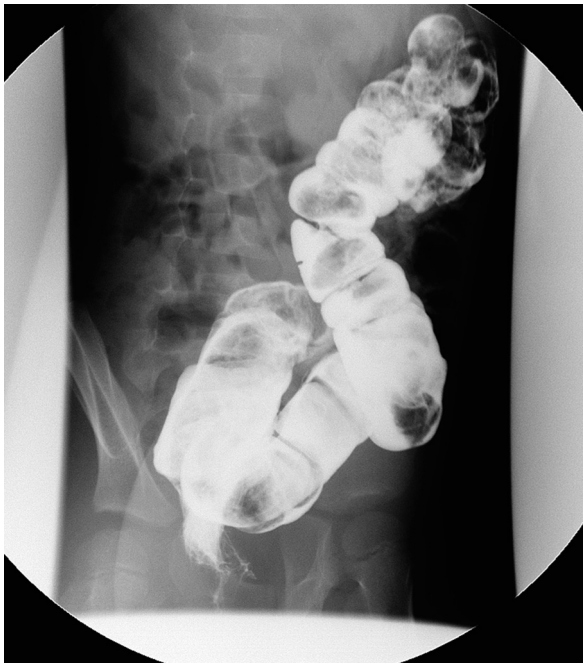

**Fig. 4.** Postoperative contrast enema: normal contour and caliber of pull-through colon (megarectum completely resected).

dissection with a risk of urinary or sexual dysfunction have been exaggerated, lacking any scientific data [12], but we had no prior experience with this procedure (transabdominal or transanal). Ultimately, we opted for a Soave transanal pull-through procedure with wide posterior myectomy, which produced satisfactory results. In terms of cuff-related complications, the Swenson transanal pull-through may be preferable by comparison, but its technical requirements are challenging, even for an experienced surgeon. In our hands, the Soave transanal pull-through proved to be a reasonable alternative for treatment of refractory constipation and megarectum.

## 3. Conclusion

This patient's condition was interpreted as intractable idiopathic constipation, in conjunction with USHD. Fearing more conservative treatments would be inadequate, a Soave transanal procedure, involving complete resection of the megarectum and pull-through of normal-caliber colon, was elected instead. This procedure dramatically improved the patient's clinical condition. Although a Swenson dissection ostensibly is ideal in such situations, the Soave transanal pull-through constitutes a reasonable alternative.

## Conflict of interest

There were no conflicts of interest.

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