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Transanal, full-thickness, Swenson-like approach for Hirschsprung disease

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Abstract

Purpose: Swenson's procedure for Hirschsprung disease (HD) was thought to disturb fecal, urinary, and ejaculatory functions leading to other approaches including the Soave and Duhamel techniques. Given our Center's experience with a full-thickness rectal dissection for anorectal malformations, and using the new transanal concept, we chose to apply these ideas to the primary treatment of HD, and describe technical aspects and impact on fecal, urinary, and sexual function.

Methods: We reviewed our series of HD patients who underwent a transanal, Swenson-like rectosigmoid dissection, assessing for postoperative stricture, anastomotic leak, enterocolitis, and long-term results for bowel, urinary, and sexual function.

Results: Of 67 patients, 28 had a transanal resection, 5 had transanal plus laparoscopy, and 34 had transanal plus laparotomy, of those, 28 patients had a leveling colostomy prior to referral. The average length of resection was 27 cm ± 12.7 cm. Mean follow-up was 17.2 months (range 1–96 months). 44 patients were at least three years old at follow-up and were assessed for urinary and fecal continence; all (100%) had voluntary bowel movements and urinary continence. Enterocolitis occurred in 9 patients (14%) and constipation (requiring laxatives) occurred in 21 (32%). Of 24 male patients, 21 (88%) reported the occurrence of spontaneous erections post-operatively.

Conclusion: Our data support the fact that a modification of Swenson's original transabdominal dissection concept using the recently described transanal approach is an excellent technique for Hirschsprung, and produces excellent long-term outcomes for fecal and urinary continence, and seems to preserve erectile function.

It has been more than 60 years since the original description of the etiology of Hirschsprung disease (HD), elucidated by Dr. Ovar Swenson [1]. Since then, numerous techniques have been described for removal of the aganglionic distal colon, but debate remains over which technique provides the best short and long-term results. The operative management of HD has evolved dramatically, from full-thickness rectosigmoid dissection (Swenson) [2], an endorectal dissection (Soave) [3], a retrorectal pouch procedure (Duhamel) [4,5], and a low anterior resection (Rehbein) [6] to more recently a primary repair [7,8] that can be done transanally [9,10], and using laparoscopic

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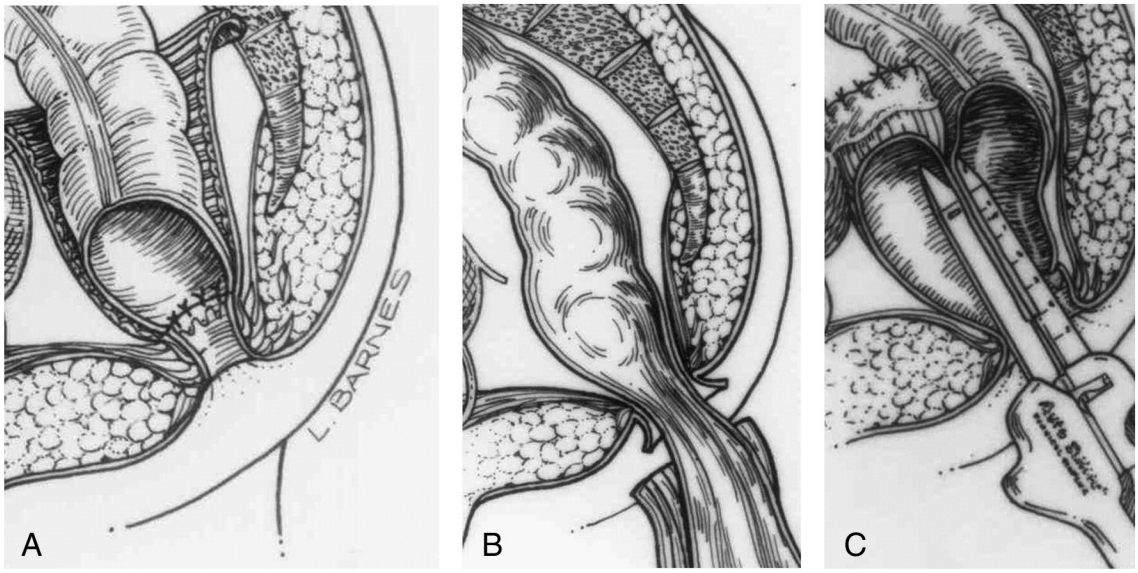

Fig. 1 Diagrams of the Swenson, Soave, and Duhamel procedures.

techniques [11]. Removal of aganglionic bowel, pulling through ganglionic bowel, and preserving the anal canal and sphincter mechanism, remain the principles to surgical repair regardless of technique. The Soave is most commonly done nowadays, with Duhamel next, which is particularly used for total colonic HD. Swenson is the least often chosen technique (Fig. 1).

In this review, we demonstrate our experience with a transanal, full-thickness, Swenson-like dissection, which we began to do because of our Center's experience with the full thickness rectal wall dissection for anorectal malformations [12], and using the idea of the transanal approach [9,10]. We surmise that the Swenson approach lost favor after its initial description because surgeons were performing the dissection too wide, leading to the described complications. With an elegant technique in the proper plane, we feel that this approach is safe, reproducible, and preserves urinary, fecal, and sexual function. We also favor this approach because it allows complete removal of essentially the entire original aganglionic bowel (except for the preserved 1 cm above the dentate line), without leaving behind a cuff (Soave) or a pouch (Duhamel).

1. Methods

We reviewed all patients between July 2000 and July 2010 who underwent a pull-through for Hirschsprung disease by the authors. Patient characteristics were collected and included age, gender, associated anomalies, and other medical problems. We excluded five patients with significant associated anomalies, as it would be difficult to compare their long-term outcomes to children with isolated Hirschsprung disease. We also excluded two that have yet to have their ostomies reversed, and 11 who required treatment for total colonic Hirschsprung disease. Preoperative contrast imaging of each patient was assessed for level of transition zone. Information on early and late post-operative complications was evaluated as well as long-term outcomes regarding fecal and urinary continence. Following approval from our Institutional Review Board, a telephone and email survey was conducted to assess the male patients for inferred preservation of sexual function, i.e. spontaneous erections. Rates of stricture formation, pelvic abscess, constipation, and enterocolitis were noted as well. Enterocolitis, for the purpose of this review, was considered to have occurred if the patient had foul smelling stool in the presence of abdominal distention which required irrigations, whether in an inpatient or an outpatient setting. All caregivers, were taught how to perform colonic irrigations prior to their definitive operation, and are carefully counseled to recognize and intervene on post-operative enterocolitis, which we have found reduces the readmission rate.

Our patients were divided into groups including those who underwent transanal resection only, transanal resection plus laparoscopy, or transanal resection plus laparotomy. The technique for rectal dissection was a transanal, full-thickness, Swenson-like approach with careful preservation of the dentate line and was uniform throughout all groups. Preoperative contrast imaging was performed in all patients, rectal biopsy was used to confirm the diagnosis of Hirschsprung disease, and a full-thickness intraoperative biopsy was used to assess that the pull-through segment had ganglionic cells and normal sized nerve trunks (<40 microns) [13]. We also collected data regarding the length of each resected specimen.

Patient's hospital courses and follow-up at 4 weeks, 3 months, 6 months, and then yearly were evaluated. Data are expressed as mean and range ± SD.

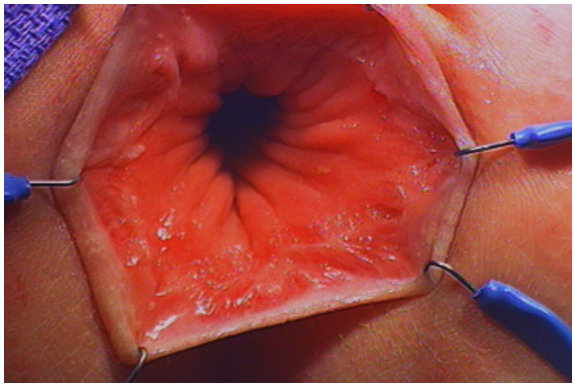

Fig. 2 Lonestar retractor demonstrating the dentate line.

2. Surgical technique

All patients were given a mechanical bowel preparation along with rectal irrigations. Preoperative antibiotics including cefoxitin and metronidazole were given to all patients with provisions made for allergies. All patients underwent a total body preparation from the nipples to the toes, and urinary catheterization was performed. The transanal dissection was performed using a prone, buttock elevated position.

The lone star retractor hooks were placed initially just inside the anal canal at the level of the mucocutaneous junction (Fig. 2). The hooks were then replaced deeper, just proximal to the dentate line so that it was no longer visible; assuring that it would be protected during the dissection (Fig. 3). In this way the distal 1.5 cm of anal canal is preserved. Interrupted silk sutures were placed 1.0 cm proximal to the dentate line in a circumferential fashion to provide uniform traction (Fig. 4). Monopolar electrocautery was used to perform a full-thickness dissection along our traction line of silk sutures (Fig. 5). Very precise coagulation of vessels right along the rectal wall was performed. The

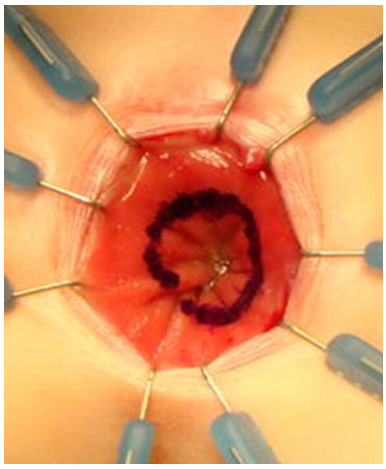

Fig. 3 Dentate line hidden by placing the Lonestar hooks deeper. Planned line of dissection marked in purple, in Cincinnati affectionately known as the purple line of Lee.

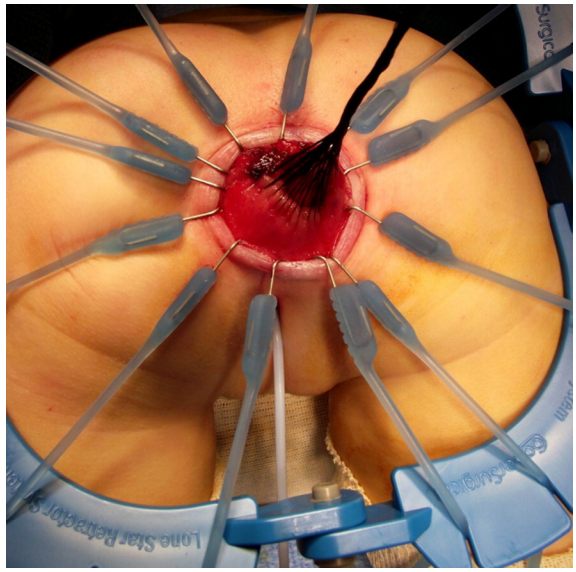

Fig. 4 Silk sutures placed 1.0 cm proximal to the dentate line and used for traction.

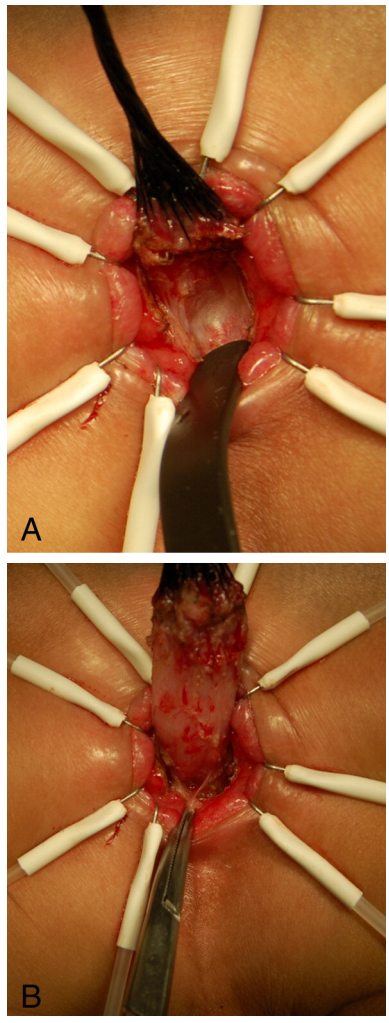

Fig. 5 Full-thickness dissection. (A) Note the areolar tissue visible in the full thickness plane. (B) The mosquito demonstrates the peritoneal reflection.

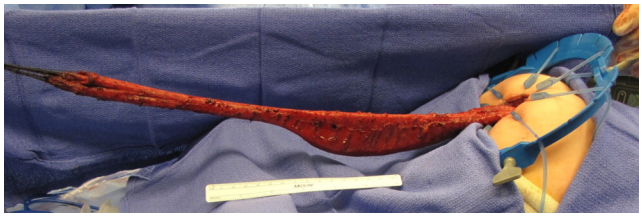

Fig. 6 Mobilized rectosigmoid showing transition to healthy bowel.

dissection was carried into the peritoneal cavity and the colon was pulled-through to the appropriate level, confirmed by intraoperative frozen section pathology (Fig. 6). Full-thickness biopsies were sent to pathology, assuring analysis of the muscularis and submucosa. Laparoscopy or laparotomy was used to start in order to assist the dissection in patients with disease that extended above the mid-sigmoid, for patients whose transition zone was not obvious on contrast study, and for those who had a previous leveling colostomy. The anastomosis was created in two layers, 1.0 cm proximal to the dentate line (Fig. 7). At one month the patient undergoes rectal exam in the clinic using Hagar dilators to assess for the need for dilations, often used in children under age one.

3. Results

After employing our exclusion criteria, 67 patients were evaluated, with age at the time of operation ranging from 6 days to 111 months (mean 18.8 ± 26 months). There were 51 male and 16 female patients, a ratio of 3.2:1. Associated anomalies in the series included five patients with Down syndrome (7.5%), and one with an associated defect in the RET proto-oncogene (who underwent prophylactic thyroidectomy at two years of age). There were two patients who had fathers with Hirschsprung disease.

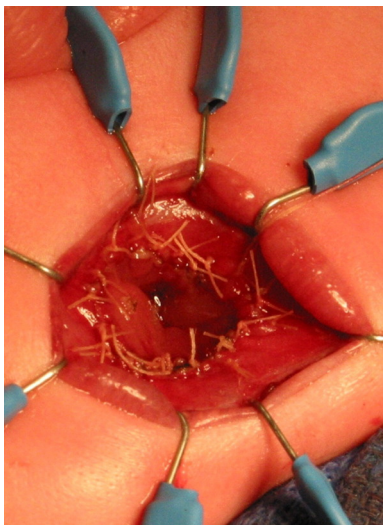

Fig. 7 Completed anastomosis.

Preoperative evaluation with contrast enema revealed a transition zone in the rectosigmoid in 47 patients (70%), left colon in 3 (4.5 %), transverse colon in 2 (3%), and in a nondiagnostic location in 15 (22.5%). Twenty-eight patients (42%) underwent fecal diversion (at another institution) prior to definitive repair by us and in 18 (27%) we performed a transanal resection in the neonatal period. We performed a purely transanal resection for those patients without preoperative diversion who had a well-defined sigmoid, (or lower) transition zone on contrast enema. Twenty-eight patients underwent a purely transanal resection, two of whom had a preoperative transverse colostomy which was left untouched for later reversal as a third stage. For those patients with nondiagnostic contrast enemas, we performed laparoscopy first to aid in identification of the transition zone and to assist in colonic mobilization. Thirty-four patients underwent laparotomy along with their transanal resection, many of whom presented early in our series, prior to the widespread use of laparoscopy for treatment of Hirschsprung's disease [14–16], or they required a laparotomy to pull-through their diverting colostomy.

The average length of resected specimen was 24.6 ± 10.9 cm for the transanal only group, 22.6 ± 11.4 cm for the laparoscopic assisted group, and 31.1 ± 13.6 cm for the laparotomy assisted group.

Operative complications were evaluated. There were no patients who required perioperative blood transfusions. There were two incidences of anastomotic leak (3%) which were managed with diverting ileostomies, one in a 5 year old and the other in a seven year old. There were no anastomotic strictures or intraabdominal abscesses. One patient experienced a prolonged post-operative ileus and on final pathology, had ganglion cells at the level of the anastomosis but also hypertrophic nerves (greater than 40 microns). He required a repeat pull-through and subsequently has done well.

Long-term outcomes were assessed at office follow-up as well as via a telephone/email questionnaire. Four patients (6%) were readmitted to the hospital in the early post-operative period secondary to enterocolitis, while 5 more (8%) reported symptoms of enterocolitis but were managed with an aggressive regimen of metronidazole, rectal irrigations, and rectal dilations if indicated, at home, a total rate of enterocolitis of 14% occurring within the first six months after surgery.

Forty-two patients were at least 3 years of age (with an equal mix of boys and girls) at last follow-up and were assessed for fecal and urinary continence. All 42 patients (100%) reported voluntary bowel movements and all voided spontaneously with urinary control. There were no episodes of postoperative urinary retention. A telephone/email questionnaire of the parents of 24 male patients who could be reached revealed 21 (88%) had spontaneous erections on a regular basis. Three patients have not demonstrated spontaneous erections post-operatively, according to their parents, but in these three boys an erection had never been noted pre-operatively, so it was hard for us to draw any conclusion.

Twenty-one of sixty-seven patients (31%) reported constipation requiring at least intermittent laxative usage.

All had their pathology re-reviewed to assure that ganglion cells were present and hypertrophic nerves were absent at the anastomosis line. All of these have voluntary bowel movements. Two require occasional enemas.

4. Discussion

The etiology of Hirschsprung disease was originally described in 1948 as relating to a defective segment of distal colon producing a partial bowel obstruction [1]. In the 1950's, after thorough examination of surgical specimens, it was noted that the diseased segments of colon contained minimal, if any, evidence for an Auerbach's plexus, leading to the use of rectal biopsies for the purpose of diagnosis. Even as Dr. Ovar Swenson was only beginning to uncover the basis of the disease process, he sought to create a definitive surgical cure.

The original procedure, described by Drs. Swenson and Bill in 1948 [2], was a transabdominal approach where careful extra-rectal dissection was carried down to a level two centimeters above the anal canal. The aganglionic bowel was resected and an end-to-end anastomosis was fashioned. At that time, the only data on pelvic dissection and subsequent long-term outcomes were found in the adult literature following operations for rectal cancer, which suggested that this type of dissection could lead to urinary and fecal incontinence as well as sexual dysfunction [17]. Similar reports suggested that these outcomes could be extrapolated to the pediatric population [18]. In the early years, the original Swenson operation was considered a delicate and technically challenging operation and was perceived to have a high incidence of complications including urinary incontinence, fecal incontinence, and impotence [19]. The cause was felt to be due to a too wide dissection about the rectum. The fear of this operation led surgeons to search for procedures which were technically less difficult and avoided the complications associated with a full-thickness rectal dissection. Duhamel proposed resection of only the intraperitoneal portion of the aganglionic colon, leaving the rectal portion intact, creating a plane of dissection in the presacral space, and fashioning a side-to-side colorectal anastomosis [4]. Soave suggested an intramural plane of dissection which left an aganglionic muscular sleeve surrounding the anastomosis to protect the surrounding pelvic structures [3]. Long-term outcomes data were slow to accumulate as surgeons continued to develop these other types of operative treatments for Hirschsprung disease. Dr. Swenson's original full-thickness approach began to lose favor as the primary treatment for Hirschsprung disease.

A careful review of the long-term data shows that the outcomes following the Swenson operation are as good, if not better, than the outcomes following the Soave or Duhamel procedures [14,20]. Most notable is the manuscript from 1989 by Sherman et al. [20] describing the outcomes of 880 Swenson procedures. They reported no complications of urinary or sexual problems post-operatively and the rates of leak, reoperation, and post-operative enterocolitis were lower than historical data for other resection techniques [3,4,15,16,20,21].

We chose to take the idea of the transanal approach [9,10], and adopt the Swenson-like technique for our approach to Hirschsprung disease influenced by our Center's experience with a full-thickness, extrarectal dissection used in patients with anorectal malformation whose principle is to stay intimately near the rectal wall [12].

In this series, there were two patients who developed anastomotic leaks. None experienced strictures or intraabdominal abscesses. This compares favorably with historical literature which quotes leak rates of 5%–7%, stricture rates of 5%–24%, and abscess formation in 2%–6% [3,4,16,20,21]. Interestingly, both patients who developed anastomotic leaks were patients who had a much delayed diagnosis of Hirschsprung disease, one diagnosed at the age of five and the other at age seven. Both had very dilated proximal bowel and were managed with a primary transanal resection without fecal diversion. Perhaps in such patients, diversion of the pull through should be considered. One of our patients developed a prolonged post-operative ileus. Intraoperative frozen section results had revealed ganglion cells, but he was found to have hypertrophic nerves at the anastomosis on permanent section. He has subsequently undergone a repeat pull-through procedure and done very well. Currently, it is our practice to not only have confirmation of ganglion cells prior to performing an anastomosis, but also have pathologic assurance of normal appearing nerves without hypertrophied nerve trunks (<40 microns) in a circumferential biopsy sample which includes muscularis and submucosa [13].

Enterocolitis is one of the most feared complications that can occur in the early post-operative period following surgery for Hirschsprung disease. Because of its severity, we are very aggressive regarding its management. While there is no precise definition of enterocolitis, we begin treatment with rectal irrigations and metronidazole when any of our post-operative patients develop abdominal distention, fever, abdominal pain, and/or foul smelling stool. Rectal dilatations are performed by the parents if indicated. We teach the irrigation technique to the family preoperatively. Irrigations and metronidazole are continued until the episode is resolved, and then weaned for a period of 3–4 weeks following the acute episode. We feel this aggressive approach avoids many hospital admissions. Though only four patients required admission for episodes of enterocolitis, we initiated treatment on five others, thus our overall rate of enterocolitis approaches 14%, which is on par with previously quoted data following a Swenson procedure and the 18%–20% quoted rate of enterocolitis following the Soave or the Duhamel procedures [3,4,15,16,20,21]. Most published reviews include only hospitalized patients so our broader definition may explain our higher incidence.

From a historical perspective, beginning as far back as 1950, there are manuscripts describing the dangers of the

Swenson dissection and the risks it poses to pelvic structures, as well as the long term risks to urinary and sexual function. However, there seems to be an absence of scientific data confirming these assertions. As we have noted, Sherman's review of 880 post-operative Swenson operations and found no patients who had urinary incontinence or sexual dysfunction [20]. Dr. Swenson himself, recently eulogized [22], often spoke of numerous patients whom he operated on who went on to have large families. He describes none who have complained of the aforementioned complications, and stresses, as we have affirmed, a meticulous dissection precisely on the rectal wall as key to avoiding complications. We surmise that such complications occur if the rectal dissection is performed too wide.

Urinary continence was noted in all patients in our series who were old enough to be assessed, and age related milestones for both urinary and fecal continence were not delayed. Bowel function is a primary endpoint in managing patients with Hirschsprung disease. All of the patients that we assessed for bowel function, who were at least three years old, demonstrate the capacity for voluntary bowel movements. There was a significant number who had constipation, 32% of our patients have constipation requiring laxatives but all have voluntary bowel movements without soiling. Given this high rate of constipation, we do not hesitate to begin senna-based laxatives post-operatively and obtain abdominal films often, to confirm that the colon is emptying adequately. Of course pathology needs to be reconfirmed, and it was.

We attempted to assess for the preservation of sexual function in our male patients, which is difficult to do in such a young patient population. We did note that 88% of the caregivers who were able to be contacted, confirmed that they had witnessed spontaneous erections in the male patients post-operatively. Those who had not witnessed an erection post-operatively, said that they had also not witnessed an erection pre-operatively. These data seem to confirm Dr. Swenson's view that the full-thickness dissection technique, when performed meticulously, does not jeopardize the nervi erigentes. Ultimate outcomes for preservation of sexual function will have to be continuously evaluated as these patients reach the age of sexual reproduction. Assessment of female sexual function will also need to wait until these patients are older.

Of patients who do poorly after their pull-through operations, a finite number will have mechanical issues which are specific to their individual surgical procedures. In patients who have had a previous Duhamel procedure, the rectal pouch, created partially of aganglionic bowel, can lead to stasis and episodes of recurrent enterocolitis [23]. Likewise, those patients with a previous Soave procedure can develop restriction by the muscular cuff, causing a functional obstruction leading to stasis, colonic dilatation, and emptying problems [24]. The Swenson-like, full-thickness dissection technique that we employ avoids leaving behind Hirschsprung's bowel altogether, except for the preserved 1 cm above the dentate line. We think there is great value to not leaving behind a cuff or a pouch [24].

The historical data regarding pelvic dissection techniques for Hirschsprung disease appear to suggest that Dr. Swenson's original operation compares very favorably to other operative techniques. Obviously, the original data refer to a transabdominal dissection which requires a laparotomy. We know that a deep pelvic dissection from above can be quite challenging, imprecise, and certainly carries a risk of injuring vital pelvic structures. Over the past 15 years, as emphasis has been placed on minimally invasive surgery, laparoscopy and the transanal dissection, either alone or in combination, have become the preferred operative techniques [9–11]. This, we believe, has led to a safer and more reproducible pelvic and rectal dissection. It appears there are little, if any, data to suggest that a full-thickness rectal dissection is less safe than other techniques and we believe there are key advantages, including excellent continence, and a purer operation without residual cuff or pouch left which can lead to obstruction. We feel that the full-thickness, Swenson-like dissection technique that we have described is safe, reproducible, and avoids many of the long-term complications associated with the other techniques.

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