



The problematic Soave cuff in Hirschsprung disease: Manifestations and treatment

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ABSTRACT

Purpose: Following a Soave pull-through for Hirschsprung disease (HD), some children struggle with obstructive symptoms. We hypothesized that these symptoms could result from a functional obstruction of the pull through caused by the Soave cuff, and that cuff resection might improve bowel emptying.

Methods: We reviewed patients referred to our center from 2008 to 2012 with obstructive problems following a Soave pull-through for HD (CCHMC IRB # 2011–2019). Only patients with an obstructing Soave cuff were analyzed. Patients with other reasons for obstruction (anastomotic stricture, transition zone, aganglionic segment) were excluded.

Results: Thirty-six patients underwent reoperation at our center for obstructive symptoms after an initial Soave pull-through. Seventeen of these patients had a Soave cuff only as the potential source of obstruction. Pre-operative symptoms included enterocolitis (10), constipation (6), and failure to thrive (1). Nine patients (53%) required irrigations to manage distension or enterocolitis pre-operatively. 14/17 patients (82%) had a palpable cuff on rectal exam. Eight patients (47%) had radiographic evidence of a cuff demonstrated by distal narrowing (4) or a prominent presacral space (4). Four children (23%) underwent excision of the cuff only. Thirteen (76%) had removal of the cuff and proximally dilated colon [(average length 7.2 cm) (12 performed transanally, and five needed laparotomy as well.)] Post-operatively, episodes of enterocolitis were reduced to zero, and need for irrigation to treat distension was reduced by 50%. Nine patients have voluntary bowel movements, and five are clean on enemas. 3/6 patients with pre-operative constipation or impaction now empty without enemas. (Follow up 1–17 months, mean 7 months.)

Conclusions: Recurrent enterocolitis, constipation, or failure to thrive can indicate a functional obstruction due to a Soave cuff when no other pathologic cause exists. Physical exam or contrast enema can identify a problematic cuff. Reoperation with cuff resection can dramatically improve bowel emptying.

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In 1948, Orvar Swenson described the first operative approach to the management of Hirschsprung disease (HD) [1]. Subsequently, other surgical procedures were developed, including the endorectal dissection (Soave) [2], retrorectal procedure (Duhamel) [3,4], and the full-thickness rectosigmoid dissection (Swenson) [1]. In addition, there have been the development of the transanal technique, and the addition of laparoscopy to all of these procedures [5–8].

The Soave procedure was developed specifically to protect the surrounding structures, outside of the rectal wall, from damage, by performing an endorectal dissection, leaving a muscular cuff behind. The procedure was initially performed transabdominally, but was later adapted to a transanal approach. Although the denominator is unknown, it is clear that most children do well after their surgical correction. However, there is a group of patients who suffer from poor emptying of the pull through [9]. In particular, following a Soave procedure, we have identified a group of children who have recurrent obstructive symptoms; manifested by recurrent enterocolitis, severe constipation, and overflow incontinence. A cited complication, specific to the Soave procedure, and a potential reason to explain

these symptoms, is an obstructing muscular cuff [10]. This is defined by the rectal cuff from the residual muscular sleeve causing a narrowing around the pull through and subsequently a functional obstruction.

This review reports our Center's experience with patients referred for post-surgical obstructive symptoms following a Soave pullthrough in which we felt that an obstructing cuff was the pullthrough's anatomic problem. We intend for this report to alert the clinician to the fact that a patient with persistent obstructive symptoms following a Soave pullthrough may have a problematic Soave cuff as the cause of the trouble.

1. Methods

Charts of all patients referred to our Center for obstructive symptoms following a previous Soave procedure for surgical correction of HD were reviewed [CCHMC IRB #2011–2019]. We performed a redo pullthrough in all of these patients, with removal of a muscular cuff. Patients with any other reason for obstruction, including anastomotic stricture, transition zone, or aganglionic segment, were excluded from the study. The patients who had a Soave cuff problem, in addition to any other pathological or

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anatomical reason, were also excluded. Patients with only an obstructing Soave cuff problem, with or without proximally dilated colon, were analyzed.

2. Results

2.1. Demographics

There were thirty-six patients identified from our database who presented to our Center with symptoms of obstruction after an initial Soave pullthrough in whom we performed a redo procedure. Seventeen of the patients had an obstructing Soave cuff as their only reason for their symptoms (identified preoperatively by clinical exam, contrast enema and postoperative confirmation from pathology). The nineteen other patients who had a Soave cuff plus an additional anatomic or pathologic issue were excluded from this analysis and have been described elsewhere [11,12].

The seventeen patients analyzed included seven females and ten males. Their ages ranged from six months to eight years of age. The time of the original pullthrough surgery to redo surgery ranged from 0.2 to 8.5 years, with an average of 3.5 years.

3. Presentation of patients

All seventeen patients in our group exhibited obstructive symptoms. Ten of the patients presented with symptoms of recurrent enterocolitis. Six had severe constipation and two of these six patients had recurrent admissions for impactions. Two patients in this group presented with soiling due to overflow incontinence related to their constipation. Two patients had chronic abdominal distention. One patient had failure to thrive. One patient did not have his original ostomy closed due to a distal narrowing (caused by the Soave cuff) and was undergoing routine dilations. Because of symptoms of recurrent enterocolitis or abdominal distention, nine patients performed regular rectal irrigations to control their symptoms.

As part of our workup for a patient with Hirschsprung disease not doing well following his or her primary repair, all of the patients underwent a contrast enema, examination under anesthesia and rectal biopsy [13]. We do not routinely use colonic or anorectal manometry in our Center during our assessment, as we have found that they do not provide any additional information from what we learn from the contrast study and examination under anesthesia. A Soave cuff was suspected in eight of the seventeen patients on the contrast enema. This was seen as a prominent presacral space (four patients, as seen in Fig. 1a); or distal narrowing of the pullthrough segment (four patients, Fig. 1b). On examination, there was a palpable cuff in fourteen of the seventeen patients (82%) – felt as a fibrous thickening or a band/ring around the distal pullthrough on digital exam. There were 2 patients who had a partially destroyed dentate line in addition to a palpable cuff. The rectal biopsies of all 17 of these patients showed normal ganglion cells and no hypertrophic nerves. There were no anastomotic strictures.

4. Surgical correction

After discussion with the family, redo surgery was offered to the patients because of symptoms and findings on clinical exam. It was explained that the only anatomic reason identifiable to account for the symptoms was a problematic Soave cuff. All patients underwent a transanal dissection around the pullthrough colon (Fig. 2). This plane defined the space between the previous pullthrough segment and the previous retained muscular cuff. Six patients also had a laparotomy. One patient, because of an absent dentate line and concern for continence pre and post-operatively [13], also had a Malone appendicostomy done as part of the procedure [14].

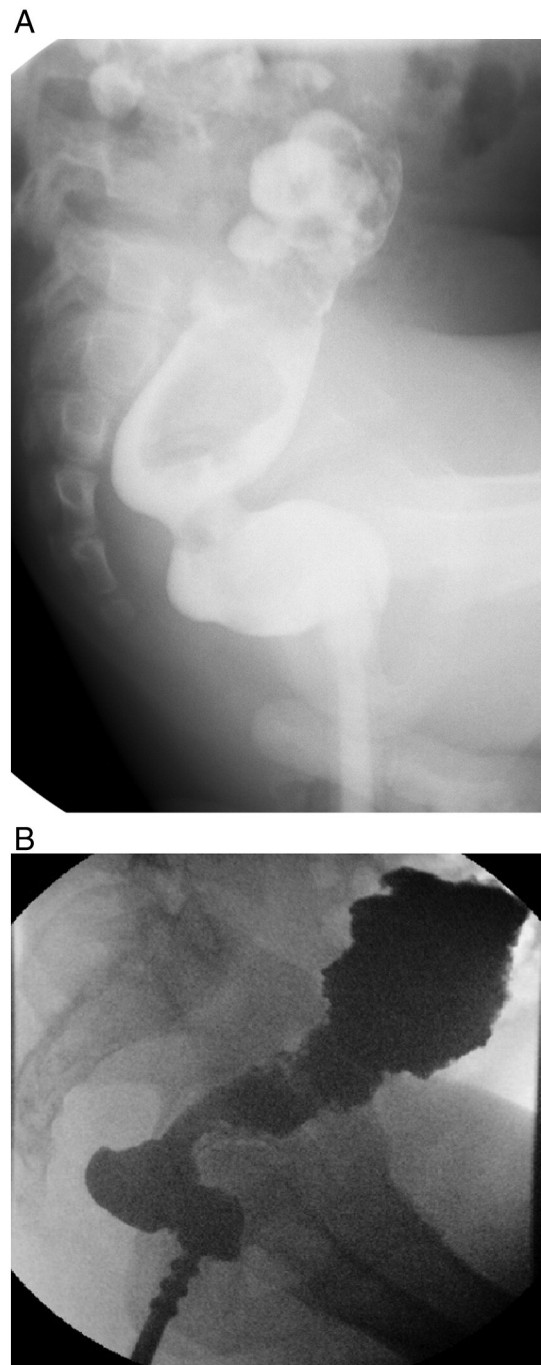


Fig. 1. (A) Indentation of distal pullthrough by a Soave cuff. (B) Distal narrowing of the pullthrough and compression by a Soave cuff.

During the transanal dissection, the muscular cuff was palpated, visualized, and excised (Fig. 3). Four of the patients had removal of the muscular cuff only. The remaining patients had resection of proximally dilated colon (mean length 7.2 cm, range of 3–30 cm).

5. Postoperative results

Patient follow-up was between one and seventeen months with a mean of seven months. After surgery, there were no reported episodes of enterocolitis. Of the nine patients who required routine irrigations preoperatively for enterocolitis or abdominal distention, only three patients reported needing irrigations, but now only

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