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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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].

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
occasionally for distention. The patient with failure to thrive had resolution of symptoms.

With regards to continence, after reoperation, nine of the patients have voluntary bowel movements. Five patients are clean with bowel management and enemas. Of the six patients who had severe constipation and impactions, none have had recurrent impactions, and three of the six patients have resolved constipation completely and stool spontaneously with control.

6. Discussion

Dr. Swenson’s original surgical correction of Hirschsprung disease was described as a dissection carried out on the seromuscular coat of the rectosigmoid colon [1]. However, subsequent reports of damage to surrounding structures by some surgeons [15], led to the development of both the Soave and the Duhamel procedures. Swenson, in his paper from 2004, was concerned that the Soave modification left a long aganglionic muscular sleeve, which can extend from the peritoneal reflection down to the anal canal. He theorized, and we agree, that the residual sleeve, where the normal colon is pulled through, can remain in a contracted state, and can cause a compressive action, and may influence peristalsis in the normal colon [16]. Kimura recognized this and hypothesized that this was contributing to the severe constipation in some of his patients [10].

Obstructive symptoms can occur in 11%–42% of patients after undergoing a pullthrough procedure for Hirschsprung disease [17], and we believe that if a patient has undergone a Soave procedure, one must consider a residual Soave cuff as a culprit in the etiology of the obstructive symptoms. Over time, proponents of the Soave procedure have in fact modified their technique to limit the amount of residual aganglionic segment (the cuff) to 1–2 cm from the beginning of their dissection [17]. This has made the Soave more Swenson-like over time, and perhaps this technique could be called a “Soaveson” [18]. It will require further follow-up to determine whether these shorter Soave cuffs will prevent the development of obstructive symptoms in our Center we use the Swenson approach to avoid this problem [19].

Although not always obvious, a rectal cuff on clinical exam or on a contrast enema should be suspected in the algorithm of working up a patient who is not doing well following a Soave pullthrough. We believe that some of these patients may be the ones diagnosed with internal sphincter achalasia and treated with Botox [17]. Because the Botox temporarily relieves the spastic muscular cuff, the patients also experience temporary relief of their symptoms. Additionally, some surgeons advocate a posterior myectomy for relief of the distal obstruction. The effectiveness of a posterior myectomy varies with the underlying symptoms. 60% of children with chronic constipation and 75% of children with recurrent enterocolitis, following a pull-through procedure had improved symptoms after a myectomy [20]. Interestingly in this review, 27 of the 32 patients had a previous Soave and one could postulate that the myectomy is breaking an obstructing Soave cuff [20]. We avoid this procedure in our practice, as the risk of the myectomy is potential permanent injury to the sphincter resulting in fecal incontinence [20].

We have found that during the workup of the patient with a potential obstructing Soave cuff, if suspected, this can often be identified on clinical exam and contrast enema. We do not routinely use manometry in our institution, (anorectal or colonic in this situation) but this could potentially be a useful adjunct in the work up and follow-up after a redo pullthrough. Our management, in these cases, is to proceed with a redo transanal pullthrough, remove the muscular cuff and potentially any proximally dilated colon. We have found that this dramatically improves the patient’s symptoms. We theorize that although the cuff may have been split at the original surgery, fibrosis around the retained muscular cuff forming an obstructing ring or rolling up of the cuff may occur. Dividing and removing the cuff relieve the distal obstruction and resolve their symptoms. In patients with an obstructing cuff and proximal dilated colon, we feel that the dilation is due to the functional obstruction caused by the muscular cuff. Because the redo surgery in these cases includes resection of the cuff and the proximally dilated colon, improvement in symptoms could be due to the removal of both anatomical issues.

The goal of this paper is to highlight to the clinician that a patient with obstructive symptoms following a Soave procedure could have a problematic Soave cuff as the etiology of the obstruction.

References


[18] Personal communication, Daniel Von Allmen, MD.


Discussion

Discussant: Dr. Juda Jona (Chicago, IL): Marc, I would like to ask you, how many of the patients became incontinent after your procedure?

Response: Dr. Marc Levitt: None.

Prof Agostino Pierro: And how long did you follow up these patients for?

Dr. Marc Levitt: The longest follow-up I think is now nearly four years and the shortest follow-up is about six months.

Discussant: Dr. Juda Jona (Chicago, IL): May I interject first a historical vignette. When I was a fellow with Dr. Swenson in 1971 he received a patient that had a Soave procedure that presented just like he had Hirschsprung’s disease. In those days all operations were done transabdominally and I want to tell all the participants that the dissection was very easy because the previous operation was done through the lumen so that the exterior of the rectum was left intact. He showed beautiful histological sections of those two layers, the inner one with ganglionic cells and the other one with incontinence was basically zero with that approach. I just wonder whether your excision is a little more radical thus leading to this kind of incontinence.

Dr. Marc Levitt: I don’t think that is the cause. I don’t think we created incontinence with the redo because we make sure to note whether the anal canal and the sphincters were intact before. I think your idea is correct. I think why some myectomies succeed is essentially doing the same thing but from a slightly different approach. It is essentially cutting the cuff that is causing the obstruction. Both options would work. I think it’s important to note in most of the articles about myectomy they often don’t note which was the original Hirschsprung surgery procedure, so if you talk about myectomy for Soave only, then you are basically doing the same thing as I am suggesting. I actually think this is a little cleaner and neater than the myectomy because it has led to some incontinence in some of the series and I don’t think everyone does the same myectomy. If you lined up 10 surgeons doing myectomy, they would have a different concept of what a myectomy actually is and that concerns me, but I think physiologically you are doing the same thing. You’re breaking the cuff that is causing the extrinsic compression and that’s the concept here. I think if there are patients with obstructive symptoms after a Soave you have to be suspicious that there may be an anatomic cause and one of the anatomic causes I believe could be is an extrinsic cuff.

Discussant: Prof. Agostino Pierro (Toronto, ON): Marc, I would like to ask you, how many of the patients became incontinent after your procedure?

Response: Dr. Marc Levitt: None.

Prof Agostino Pierro Zero? Because this is a very extensive dissection that you do after ...

Dr. Marc Levitt: No, what I think is important to note is just like in a primary you must preserve the dentate line and do not overstretch the sphincters. We do not impose incontinence. Incontinence occurs if you destroy the sphincters by overstretching them or resect the dentate line. There are some patients that came to us without a dentate line but with obstructive symptoms, and in those patients we remove the cuff but those patients because of the inability to detect the stool because of loss of the dentate line needed enemas to treat their incontinence but we did not impose any incontinence by doing this redo. I do not think we should be afraid of doing redos because it might impose incontinence because if you maintain the principles of preserving the dentate line and not overstretching the sphincters, that’s where you are going to maintain continence. Incontinence is a result of hurting the sphincters or hurting the dentate line.

Prof. Agostino Pierro: And how long did you follow up these patients for?

Dr. Marc Levitt: The longest follow-up I think is now nearly four years and the shortest follow-up is about six months.
aganglionosis, and he obviously cured the patient with the Swenson dissection. You have to remember in those days also the Soave was done by cooling the bowel and having it dangling down the patient’s rear end so that the cuff invariably was long and caused a lot of troubles. 

*Dr. Marc Levitt:* Does everyone recognize each of these faces (slide with photos shown to audience)? Anyone want to note who these folks are?

*Moderator:* I’ll start with Swenson.

*Marc Levitt:* Very good. Next?

*Unidentified speaker:* Franco Soave.

*Marc Levitt:* Very good. In the middle? That’s Duhamel. And number four is Hirschsprung. Thank you all very much.