

Reoperation for Hirschsprung's Disease

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Background/Purpose: Reoperation for Hirschsprung's disease traditionally has been used for patients with anastomotic leaks or stricture or with severe constipation from retained aganglionic segment or neuronal dysplasia, but there is little information regarding its use for other complications and the long-term outcome in these patients.

Methods: In a 23-year period, 107 infants and children underwent Soave (68 patients) or Duhamel (39 patients) pull-through procedures. The age at operation was newborn to 6 years (mean, 10 months). Eighty percent had aganglionosis limited to the rectosigmoid colon. Follow-up was by office visit or telephone (mean, 8.5 years).

Results: Twenty-three of the 68 patients with Soave pull-through (34%) underwent reoperation for intractable enterocolitis (10 patients, all 10 cured); anastomotic stenosis (four patients, three cured, one continued diversion); anastomotic leak (four patients, four cured); retained aganglionic segment (three patients, three cured); one necrosis of pull-through converted to Duhamel and cured; and one rectal prolapse that was diverted. Fifteen of the 39 patients with Duhamel procedure (38%) underwent reoperation for severe constipa-

tion (seven patients, six cured, one diverted); persistent rectal septum (four patients, 4 cured); and intractable enterocolitis (four patients, three cured, one diverted). Overall, 21 of 23 patients (91%) with reoperation after Soave procedures were cured, whereas 13 of 15 patients (87%) who underwent reoperation after Duhamel procedure were cured, and four patients remain diverted.

Conclusions: These data show that aggressive reoperation can result in a high cure rate in Hirschsprung's disease. Although there is no significant difference in the rate of reoperation after Duhamel and Soave procedures, the patients with Soave pull-through required more complex reoperations, with several requiring more than one procedure. An aggressive approach to reoperation in patients with Hirschsprung's disease clearly is justified.

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INDEX WORDS: Hirschsprung's disease, enterocolitis, constipation.

HIRSCHSPRUNG'S DISEASE is one of the most common congenital anomalies that pediatric specialists manage. Although the clinical presentation has been well known for over 100 years and the anatomic abnormality of absence of ganglion cells recognized for 50 years, the outcome after surgery remains less than ideal. Recently we reviewed the results of proctectomy and Soave and Duhamel pull-through procedures in a group of infants and children,¹ demonstrating a satisfactory outcome (no further operations or hospitalizations) in only 60% to 70% of patients. The current report analyzes the further surgical treatment for the remaining patients in this series and suggests that aggressive surgical therapy for complications after the initial procedure can result in long-term cure in a high percentage of cases.

MATERIALS AND METHODS

From 1975 to 1997, 107 infants and children with Hirschsprung's disease were treated at Cardinal Glennon Children's Hospital, Saint Louis University Health Sciences Center, St Louis, MO. The characteristics of the patients, extent of disease, symptoms at presentation, and associated disorders are presented in Table 1. This population is typical for a large series of patients with Hirschsprung's disease, with the majority presenting at birth with signs of bowel obstruction or at older ages with chronic constipation. Aganglionosis was limited to the

rectosigmoid or left colon in 87%, and a small percentage of patients had a positive family history or Down's syndrome.

Sixty-eight patients (64%) underwent Soave pull-through procedure at age newborn to 5 years (mean, 9 months), using the Boley modification, whereas 39 patients (ages 6 months to 6 years) underwent modified Duhamel procedure. The patients were followed up postoperatively for 6 months to 22 years (mean, 8.5 years) by routine office visit for the first 5 years, then by telephone. The development of complications that prompted an unscheduled office visit, emergency room visit, rehospitalization, or reoperation was recorded. Specific symptoms or complications that eventually resulted in reoperation were also recorded. These data were further subdivided depending on the type of pull-through procedure that had been performed. For the purposes of this study, a child was considered "cured" if he or she was not diverted (colostomy or ileostomy), had a normal defecation pattern for age with little or no oral medications, and required only occasional (1 to 2 per month) rectal irrigation or enema.

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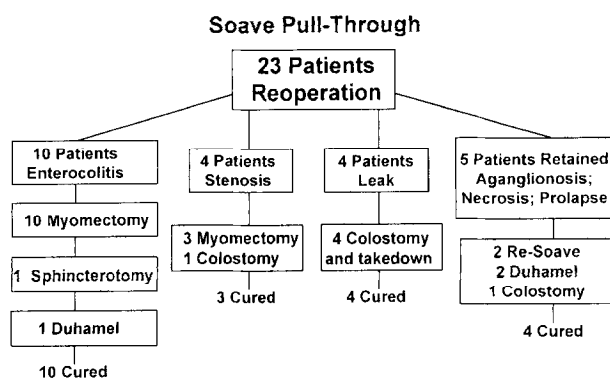
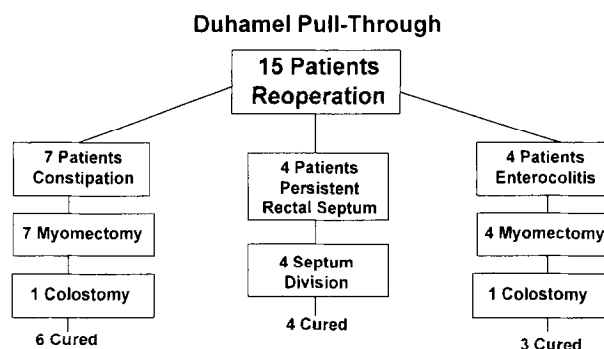
Table 1. Characteristics of 107 Patients With Hirschsprung's Disease

	No. of Patients	Percentage
Age at diagnosis		
Newborn (<30 d)	62	58
Infant (30 d-1 yr)	28	26
Child (>1 yr)	17	16
Positive family history	8	7
Extent of disease		
Rectosigmoid/left colon	93	87
Transverse colon	7	6
Total colon/small bowel	7	6
Initial clinical presentation		
Intestinal obstruction/constipation	81	76
Diarrhea (enterocolitis)	18	17
Perforation	4	4

RESULTS

One child died in this series (1%) of recurrent enterocolitis and dehydration approximately 2 years after Duhamel pull-through procedure. This child had a colostomy at the time of death and had not undergone any reoperation except for fecal diversion. There were no other early or late deaths in this series.

The specific reasons for reoperation, the procedures performed, and the outcomes after reoperation for Soave pull-through procedure are shown in Fig 1. Persistent or recurrent enterocolitis (two to six episodes) requiring rehospitalization for intravenous fluids, antibiotics, and rectal irrigations was the most common reason for further operation after Soave procedure. All 10 of these patients were cured, however, with anorectal myomectomy (10 patients), additional sphincterotomy in one patient, and conversion to a Duhamel procedure in another. Frequently, the results of the myomectomy were dramatic with almost immediate cessation of diarrhea, improved appetite and weight gain, and normal stool pattern with little or no oral medications. Stenosis at the coloanal anastomosis developed in four additional patients, none

**Fig 1. Indications, procedures, and outcomes in 23 patients who underwent reoperation after Soave pull-through procedure.****Fig 2. Indications, procedures, and outcomes in 15 patients who underwent reoperation after Duhamel pull-through procedure.**

of whom responded to repeated dilatations. Three of these patients responded to anorectal myomectomy, whereas one required colostomy because of rectal perforation during dilatation and remains with fecal diversion. Leak at the coloanal anastomosis developed in four patients and required subsequent fecal diversion. All of these colostomies have been closed, and all of the children are cured. Finally, five patients had either retained aganglionic segment ($n = 3$), necrosis of pull-through ($n = 1$), or severe rectal prolapse ($n = 1$). These were treated with re-pull-through procedure in four patients (two Soave, two Duhamel), all cured, whereas the child with severe rectal prolapse remains diverted. Overall, 21 of 23 patients (91%) treated with these various reoperative procedures were eventually cured. Combined with the patients who required no reoperation, the rate of patients with excellent results is more than 90%.

The specific reasons for reoperation, the procedures performed, and the outcomes after reoperation for Duhamel pull-through are shown in Fig 2. In contrast to the Soave pull-through, the most common indication for reoperation after Duhamel procedure was persistent constipation unresponsive to nonoperative therapy (seven patients). These patients all underwent anorectal myomectomy, but one patient showed no improvement and subsequently underwent colostomy. Four additional patients had a persistent rectal septum, even though septum division was felt to have been completed at the time of the original procedure. All four underwent repeat septal division with a stapling device, which cured each child. Finally, four patients had persistent repeated episodes of enterocolitis, which prompted anorectal myomectomy. Three children were cured almost immediately with this procedure, whereas one child subsequently underwent colostomy and eventually died of enterocolitis. Overall, 13 of 15 (81%) patients who underwent these various

reoperative procedures were eventually cured. When combined with the patients who required no reoperation, the rate of patients with excellent results is more than 90%.

DISCUSSION

The discovery that Hirschsprung's disease is associated with aganglionosis, and the subsequent design of operations by Swenson,² Soave,³ and Duhamel⁴ is one of the major success stories in pediatric surgery. With refinement and modification of these three basic surgical procedures, there has been steady improvement in the outcome after surgical repair of Hirschsprung's disease. Mortality, except that associated with severe enterocolitis is practically nonexistent. However, in the present series, only 65% to 76% of patients did not have a complication that required reoperation or rehospitalization.

The most common postoperative complications, enterocolitis and recurrent constipation, are frequently blamed on patient disease rather than technical mistakes.⁵⁻⁷ Children who have enterocolitis before pull-through procedure are at higher risk for continued enterocolitis postoperatively. Because postoperative enterocolitis has a mortality rate of 30% or more, aggressive management of patients with enterocolitis, especially in patients with recurrent episodes necessitating hospitalization, is advantageous and may result in a reduced mortality rate if the episodes can be abated or eliminated.

Few publications of large series of pull-through procedures in children suggest that these postoperative problems might be indicators of technical errors and thus amenable to reoperation. In a review of a large series by Swenson et al,⁸ reoperation was mentioned as a treatment for rectal stricture and anastomotic leak, but was discouraged as a treatment option for enterocolitis. Other large series by Joseph and Sim,⁹ Rescorla et al,¹⁰ and Reding et al¹¹ mention reoperation as an option for specific postoperative complications (stricture, stenosis) but not as definite treatment for other complications such as enterocolitis. In contrast, several other investigators have advocated reoperation, particularly myomectomy or sphincterotomy, for the treatment of recurrent enterocolitis. Marty et al¹² used anorectal myomectomy in eight patients who had persistent postoperative enterocolitis with excellent results. Elhalaby et al¹³ used myomectomy to treat postoperative enterocolitis in nine patients, with eight showing good to excellent results.

In the current series, we have taken an aggressive reoperative approach to not only the well-recognized postoperative pull-through complications of anastomotic stricture, anastomotic leak, persistent rectal septum, or retained aganglionic segment, but also the problem of

chronic recurrent enterocolitis. Repeated episodes of enterocolitis are probably secondary to colonic fecal retention, either because of a short segment of retained aganglionosis, hypoganglionosis, or internal sphincter "achalasia." Anorectal myomectomy¹⁴ has been an effective additional surgical procedure in our experience for these patients, resulting in cure in most instances. Myomectomy has also been an effective treatment for anastomotic stricture after Soave procedure.

The development of constipation after pull-through procedure is complex and can be difficult to diagnose and manage. Both Duhamel and Soave procedures can result in a persistently aganglionic segment of rectum that can cause refractive constipation. In addition, ischemia of the pull-through colon may result in ganglion cell death, which can appear identical to retained aganglionosis on barium enema and rectal biopsy.⁶ Early rectal biopsy in the child with persistent constipation should allow early detection and therapy in this setting.

The surgical treatment of retained aganglionosis can include either anorectal myomectomy or redo pull-through operation. For the treatment of recurrent aganglionosis after pull-through procedure, Shermeta and Nilprabhassarn,¹⁵ Banani and Faroolan,¹⁶ and Heij et al¹⁷ favor myomectomy, whereas Tariq et al¹⁸ have found redo pull-through procedures to be efficacious in the treatment of this postoperative complication.

When myomectomy or rectal dilation have failed, or when a long segment of aganglionosis is suggested by contrast enema and proven by biopsy results, then redo pull-through operation usually is required. In our series, five patients, all of whom had a Soave procedure initially, required a second pull-through procedure. Two patients underwent another Soave procedure, whereas three others underwent conversion of Soave to a Duhamel operation. All of these patients had excellent outcomes, suggesting that even extremely aggressive reoperative procedures are well tolerated and can be curative.

One of the most difficult conditions to detect and manage after pull-through procedure is intestinal neuronal dysplasia.⁷ The histological criteria for this disorder are somewhat vague and nonspecific, which might explain the wide variation in prevalence of this disease. Although we had no instances of neuronal dysplasia in our series, other investigators have used combinations of myomectomy, colon resection, and pull-through with varying results.¹⁹

This series demonstrates that close follow-up and aggressive use of reoperation can result in low mortality rates and excellent long-term results in Hirschsprung's disease. However, further modification of the standard

surgical procedures for Hirschsprung's disease to reduce the length of the retained aganglionic segment will hopefully result in a decreasing need for these secondary procedures in the future. Recent innovations, such as

newborn pull-through²⁰ and laparoscopic pull-through procedures,²¹ although untested in long-term studies, hold promise for further improvements in surgical management of this common congenital abnormality.

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Discussion

P.K. Donahoe (Boston, MA): Can you tell us exactly how you perform your myomectomies? Do they differ from the sphincterotomies? Would you consider performing your myomectomies primarily?

T.R. Weber (response): We elevate the posterior mucosa, remove a strip of muscle from the dentate line proximally for 3 to 4 cm or longer, and 1 cm wide. Sphincterotomy is continued more proximally than myomectomy.

We have not performed primary myomectomy, but in selected cases it might be a good idea.

R. Touloukian (New Haven, CT): I want to compliment Dr Weber and his colleagues for a very honest and forthright report. It is distinctly unusual for surgeons to come before this audience with data suggestive that the initial operation is less than successful in more than 20% of their cases.

The evidence suggests that reoperation in a good

percentage of your patients is for retention of aganglionic colon. Apparently, myomectomy subsequently leads to a better result than you had with your initial procedure. What lesson is to be gained from this experience, and how do you now modify the Duhamel or endorectal procedure to lessen the risk of postoperative enterocolitis? Because the Swenson operation is the procedure leaving the least amount of retained aganglionic rectum, is there reason to move in that direction again?

T.R. Weber (response): We have modified our Soave technique to attempt to bring the anastomosis closer to the dentate line, to leave less of an aganglionic segment. I think the addition of a myomectomy, as Dr Donahoe suggested, would probably be useful as well.

J.J. White (Macon, GA): I just wanted to go further into your technique of myectomy. As I understand it, particularly with the Soave procedures, you are doing a myectomy of the normal ganglionic bowel that you have pulled

down; Hugh Lynn did his on the aganglionic bowel of the very short segment. Is that correct?

T.R. Weber (response): Yes, and I suspect our myomectomy goes up into the muscular cuff because we frequently enter an area of aganglionosis.

J.J. White: So, basically, you are going through the wall of the pull-through bowel into the wall of the rectal sheath, and you are doing a myectomy going through one complete wall and then partially into the residual rectal sheath.

T.R. Weber (response): That is correct.

S.J. Boley (Bronx, NY): I think the question of myomectomy should be cleared up. The problem with all of the procedures for Hirschsprung's disease is that we leave one third of the abnormal internal sphincter intact below our anastomosis, whether it be a Duhamel or a Swenson or an endorectal. That is the portion that causes enterocolitis. I was very surprised to hear you say that we do not think enterocolitis is amenable to surgery. We have been recommending myomectomy for the first episode of

enterocolitis after any of these procedures for more than 20 years.

T.R. Weber (response): I think selected investigators have recommended it, although there are a number who have written that there is no role for surgery in the patient with enterocolitis. But your point is well taken.

J. Langer (St Louis, MO): A number of groups have now performed long-term follow-up studies in patients with Hirschsprung's disease and have documented over a 10% incidence of incontinence long term. I have a great concern that myomectomies may be contributing or exacerbating that problem. Have you any long-term data on patients with myomectomies that might convince me that these patients are not at greater risk for long-term incontinence?

T.R. Weber (response): Our mean follow-up time is 8½ years. Our definition of cure is normal defecation pattern for age, and we have not seen a significant amount of long-term incontinence from myomectomy.