With the rapid change of technology and increasing volumes of information, review articles are increasingly valuable. In an effort to provide this service for our readers, we solicit and publish review articles which summarize the current status of diagnosis and treatment of colonic disease.

Hirschsprung's Disease in Adolescents and Adults

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Hirschsprung's disease in the adolescent and adult is a rare and often misdiagnosed cause of lifelong refractory constipation. Two adolescent and three adult patients with Hirschsprung's disease treated between 1973 and 1987 at the University of Michigan Medical Center are reported. Each patient presented with chronic constipation requiring enemas, cathartics, and multiple hospital admissions for management. Diagnosis in each case was made with barium enema and full-thickness rectal biopsy. Four patients underwent endorectal pull-through procedures, all with good long-term results. The fifth patient, initially treated with a Duhamel retrorectal pull-through procedure, required reoperation for constipation secondary to a retained rectal septum. Review of 199 cases of adult Hirschsprung's disease enables comparison of the various operative procedures for this disorder with respect to postoperative complications and functional outcomes. Anorectal myectomy with low anterior resection, the Duhamel-Martin procedure, and the Soave endorectal pull-through procedure are the most acceptable methods for surgical management. [Key words: Hirschsprung's disease; Constipation; Adult; Adolescent]

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HIRSCHSPRUNG'S DISEASE IN its classic form consists of aganglionosis of the rectum and colon and characteristically presents in childhood with symptoms of constipation, colonic obstruction, or sepsis due to enterocolitis. Rarely does Hirschsprung's disease remain undiagnosed until adolescence or adulthood. In such patients, the prolonged partial colonic obstruction is overcome by hypertrophy of the active, normally innervated, proximal bowel.[1] In addition, patients can compensate for the obstructed aganglionic bowel by using cathartics and enemas. Ultimately, however, the dilated colon may decompensate secondary to the distal physiologic obstruction, and the patient may experience rapidly worsening constipation or even acute obstruction. Rarely, it is at this advanced stage of Hirschsprung's disease that the older child or adult will present for definitive diagnosis and surgical management.

In 1948, Swenson and Bill[2] described an abdominoperineal pull-through procedure in which the distal aganglionic bowel was resected with preservation of continence by protecting the anorectal sphincters. In 1960, Duhamel[3] proposed a retrorectal transanal pull-through procedure, and in 1963, Soave[4] described the endorectal pull-through procedure. These latter procedures were developed in an attempt to eliminate the anastomotic complications, incontinence, and persistent constipation occasionally associated with the Swenson procedure. In 1966, Lynn[5] reported his experience with the posterior rectal myectomy to correct

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short segment aganglionosis. These procedures have all subsequently undergone modifications,[6,7] underscoring the lack of an ideal operation for management of this disease.

The surgical treatment of Hirschsprung's disease in the adult or adolescent (adult Hirschsprung's disease) remains controversial. Each of the procedures developed for the childhood disorder have been applied to adolescent children and adults with varying success. Herein, we report our experience over the past 15 years with five patients in whom the diagnosis of Hirschsprung's disease was made in adolescence or adulthood and describe their surgical treatment. In addition, a complete review of the surgical literature relevant to adult Hirschsprung's disease is reported and a collective experience of the surgical treatment of this disorder is presented.

Clinical Material

The records of all patients over the age of 12 years with histologically proven Hirschsprung's disease treated at the University of Michigan Medical Center between 1973 and 1987 were reviewed. Twelve years was chosen as the lower age limit since the clinical presentation of Hirschsprung's disease in adolescents is very similar to that of adults yet clearly different from neonates and infants. Records of five patients were reviewed. The diagnosis was suggested in all patients by means of barium enema and confirmed with rectal biopsy. Rectal biopsy performed 2 cm above the anal verge was considered the standard for diagnosis, with histologic absence of ganglion cells and the presence of hypertrophied nerve trunks in a full-thickness biopsy necessary for definitive diagnosis. Clinical features, operative procedures, and results were recorded. Follow-up evaluation was obtained through postoperative clinic visits and subsequent phone interviews for all but one patient.

Results

The clinical findings, diagnostic results, operative procedures and follow-up of the five patients treated at the University of Michigan Medical Center are presented in Table 1. Each patient had a history of abdominal distention as well as severe, lifelong constipation necessitating regular cathartics or enemas. In one patient, pregnancy exacerbated her symptoms of Hirschsprung's disease, leading to a definitive diagnosis and eventual postpartum surgical treatment. In all patients, barium enema suggested the diagnosis of Hirschsprung's disease (Fig. 1), and rectal biopsy confirmed the diagnosis. The initial rectal biopsy in one patient was equivocal, and diagnosis was delayed until persistence of symptoms mandated a repeat biopsy, which confirmed the diagnosis. Four patients underwent the endorectal pull-through procedure, all with excellent long-term results, although one patient's postoperative course was complicated by an anastomotic leak, which was successfully managed conservatively. One patient was referred having previously undergone a Duhamel procedure and subsequently required reoperation for a retained rectal septum. His short-term result was excellent, but he has since been lost to follow-up. Three of the five patients underwent initial diverting colostomy. The endorectal pull-through was then performed approximately six months later, after the proximal dilated colon had returned to normal caliber. The colostomy was then closed as a third procedure two to six months after the pull-through. One patient was given a diverting colostomy at the time of his endorectal pull-through, and this was closed at a second operation two months later. The final patient underwent a single-stage endorectal pull-through without fecal diversion. The decision to perform a diverting colostomy was made individually for each patient and was based on degree

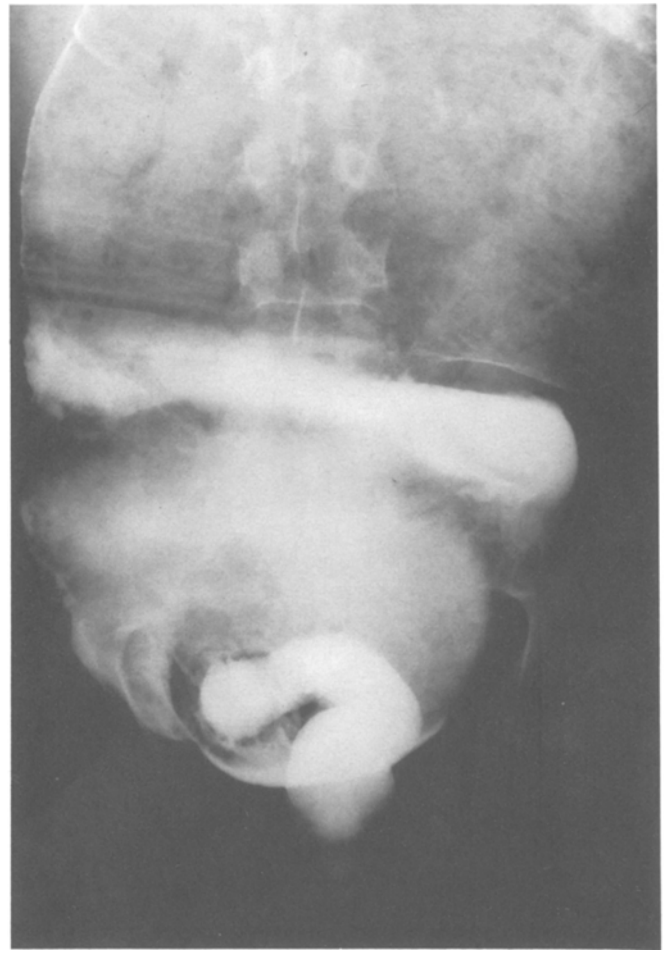

FIG. 1 (Patient 4). Unprepared barium enema showing megacolon and lower rectal narrowing.

TABLE 1. *Five Patients with Adult Hirschsprung's Disease Treated at the University of Michigan Medical Center*

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age at diagnosis	57	16	33	51	16
Sex	Male	Female	Female	Male	Male
Onset of symptoms	Birth	Birth	Birth	Birth	Birth
Constipation	Severe	Severe	Severe	Severe	Severe
Barium enema	Narrow rectosigmoid	Dilated colon	Narrow rectosigmoid	Narrow rectosigmoid	Narrow rectosigmoid
Rectal biopsy	Diagnostic	Initially negative, repeat diagnostic	Diagnostic	Diagnostic	Diagnostic
Procedure	ERPT*	ERPT	ERPT	ERPT	Duhamel
Initial colostomy	Yes	Yes	No	Yes	Yes
Postoperative course	Anastomotic leak and stricture	Normal	Normal	Normal†	Referred with constipation secondary to retained septum; septectomy with subsequent resolution
Result	Good 4 to 6 bowel movements/day	Good 2 to 3 bowel movements/day	Good 5 to 6 bowel movements/day	Good 2 to 4 bowel movements/day	Good
Follow-up	54 months	79 months	85 months	18 months	Lost to follow-up

*Endorectal pull-through.
†Bilateral compartment syndrome due to prolonged time in stirrups; resolving.

of dilatation of the colon proximal to the aganglionic segment as well as the adequacy of the mechanical bowel preparation and ease of operation.

Literature Review

Review of the English-language surgical literature relevant to adult Hirschsprung's disease over the past 30 years is presented in Table 2. Reports on adult Hirschsprung's disease have advocated operative management with one of the following procedures: 1) Soave endorectal pull-through as modified by Boley,[4,6] 2) Duhamel retrorectal pull-through[3] with or without the Martin modification,[7] 3) Swenson procedure,[36] 4) posterior anorectal myectomy as described by Lynn,[5] 5) posterior anorectal myectomy combined with low anterior resection, 6) low anterior resection (State procedure), and 7) colectomy.

For the purposes of the review, surgical complications were classified as: major, consisting of anastomotic leak, abscess, impotence, and stricture requiring reoperation; and minor consisting of wound infection, stricture requiring dilatation, temporary urinary retention, and rectal hematoma. Long-term results were classified as: good, consisting of complete fecal continence with rare use of laxatives or enemas; fair, consisting of occasional fecal incontinence or routine use of laxatives or enemas; and poor consisting of continued reliance on mechanical disimpaction with no improvement over preoperative condition. Patients needing further surgery were considered to have a poor result. The associated complications and long-term results of each surgical procedure are described below and summarized in Table 3.

Soave Endorectal Pull-Through: Thirty-two cases of adult Hirschsprung's disease managed with the Soave endorectal pull-through as modified by Boley are reported. Combined long-term results are acceptable, although a significant number of perioperative complications occurred (Table 3). In evaluating these results, however, it is important to note that three patients treated by one group[10,15] had poor results, and two of these patients are reported to be impotent. Impotence is a very unusual complication with the endorectal pull-through, since the dissection should be carried out within the submucosal plane of the rectum, away from the pelvic autonomic nerves. If these cases are excluded, the incidence of major postoperative complications remains high, but 85 percent of the patients have good long-term results. The endorectal pull-through procedure is frequently criticized because of an increased incidence of postoperative strictures requiring dilatation.[1] This occurred in one of the three patients treated with this procedure at our institution and a total of four patients in the collected literature review (13 percent); all responded to dilatation. The most common major complication reported was anastomotic leakage (16 percent) leading to cuff or perirectal abscesses. This complication also occurred in one of our patients.

Duhamel Retrorectal Pull-Through: Excellent results have been described using this procedure for the treatment of adult patients. The largest experience with this procedure has been reported in three articles from St. Marks Hospital, London.[10,14,15] To avoid duplication of patients when tabulating the combined reported

TABLE 2. *Published Reports of Adult Hirschsprung's Disease*

	Procedures					
Author, year	Soave	Duhamel	Swenson	Myectomy	LAR* + Myectomy	LAR*/Colectomy
Lynn and Van Heerden,[8] 1975				8		
Outcome:				5 Good		
				1 Fair		
				2 Poor		
Complications:				None		
Lee and Koh,[9] 1977				3		
Outcome:				3 Good		
Complications:				None		
Todd,[10] 1977‡	3	14				3
Outcome:	1 Good	14 Good				3 Good
	2 Poor					
Complications:	2 Major	None				None
McCready and Beart,[11] 1980	3	4	17	13	0	13
Outcome:	3 Good	3 Good	14 Good	7 Good		8 Good
		1 Fair		1 Fair		1 Fair
			3 Poor	5 Poor		4 Poor
Complications:	2 Major	1 Minor	6 Major	1 Minor		2 Major
Bower,[12] 1983	3					
Outcome:	3 Good					
Complications:	1 Minor					
Hamdy and Scobie,[13] 1984				3	2	
Outcome:				2 Good	2 Good	
				1 Poor		
Complications:				None	None	
Elliott and Todd,[14] 1985‡		39				
Outcome:		36 Good				
		3 Fair				
Complications:		5 Major				
Barnes *et al.*,[15] 1986‡	7	17		1		
Outcome:	4 Good	13 Good				
	2 Fair	2 Fair				
	1 Poor	2 Poor		1 Poor		
Complications:	None	1 Major		None		
Fishbein *et al.*,[16] 1986				5	6†	
Outcome:				2 Good	6 Good	
				3 Poor		
Complications:				None	None	
Starling *et al.*,[17] 1986	6	1	2	3		1
Outcome:	6 Good	1 Good	1 Good			
			1 Poor	3 Poor		1 Poor
Complications:	1 Major	None	1 Major	None		1 Major
	2 Minor		1 Minor			
Natsikas and Sbarounis,[1] 1987		6				
Outcome:		6 Good				
Complications:		None				
Wheatley *et al.*, 1989	4	1				
Outcome:	4 Good	1 Good				
Complications:	1 Major	1 Major				
	1 Minor					
Various authors,[2, 12, 18-35] 1948–1986	6	7	11	2		16
Outcome:	6 Good	6 Good	9 Good	2 Good		12 Good
						2 Fair
		1 Poor	2 Poor			2 Poor
Complications:	1 Major	None	3 Major	None		2 Major
			2 Minor			

*LAR = low anterior resection.
‡Includes patients with poor results after myectomy alone as an initial procedure.

TABLE 3. *Combined Results of the Surgical Treatment of 199 Adults with Hirschsprung's Disease*

Endorectal pull-through
32 operations
8 major complications (25 percent)
4 minor complications (13 percent)
Results
27 Good (85 percent)
2 Fair (6 percent)
3 Poor (9 percent)
Duhamel procedure
58 operations
6 major complications (10 percent)
1 minor complication (2 percent)
Results
53 Good (91 percent)
4 Fair (7 percent)
1 Poor (2 percent)
Swenson procedure
30 operations
10 major complications (33 percent)
2 minor complications (7 percent)
2 impotence (7 percent)
Results
24 Good (80 percent)
6 Poor (20 percent)
Posterior anorectal myectomy
38 operations
0 major complications (0 percent)
1 minor complication (3 percent)
Results
21 Good (55 percent)
2 Fair (5 percent)
15 Poor (40 percent)
Posterior anorectal myectomy and low anterior resection
8 operations
0 major complications (0 percent)
0 minor complications (0 percent)
Results
8 Good (100 percent)
Low anterior resection
18 operations
1 major complication (6 percent)
0 minor complications (0 percent)
1 death (6 percent)
Results
13 Good (72 percent)
2 Fair (11 percent)
3 Poor (including 1 death) (17 percent)
Colectomy
15 operations
4 major complications (27 percent)
0 minor complications (0 percent)
1 death (7 percent)
Results
10 Good (67 percent)
1 Fair (7 percent)
4 Poor (including 1 death) (26 percent)

results, only the largest series of Duhamel results reported from this institution[14] is included in Table 3. Although a retained rectal septum leading to postoperative constipation has been reported in children after the Duhamel procedure,[37] only one adult patient is described who required reoperation for a retained rectal septum.[15] Our patient treated with a Duhamel procedure elsewhere required reoperation for constipation secondary to a retained septum but the result after this second procedure was excellent at the time of discharge. The Martin modification eliminates the rectal septum,[7] thereby preventing this complication.

Swenson and Bill Abdominoperineal Pull-Through: In 1975, Swenson *et al.*[38] reported their collected results with 282 patients treated with the abdominoperineal pull-through procedure. This review included 71 adults treated for Hirschsprung's disease. Unfortunately, the complications and long-term results are reported only for the collected group of patients, and therefore the results with these 71 adult patients can not be included in the literature review. Two adult patients described in detail by Swenson and Bill[2] have been included in the review. In total, thirty adult patients treated with this procedure have been reported. One third of the patients had major complications and 20 percent have had poor long-term results. Major complications were related to anastomotic leakage leading to fistula or abscess formation.[11,36] Strictures were also a frequently noted complication.[11,17] Due to the extensive pelvic dissection, impotence is often mentioned as a potential complication of this procedure.[37] Two adult patients in the collected series are reported to have developed impotence.[11]

Posterior Anorectal Myectomy: This technique was originally described by Lynn[5] for the treatment of short-segment Hirschsprung's disease in children. This procedure has recently been advocated for adults, particularly as an initial, low morbidity procedure in patients with probable short-segment aganglionosis.[16] In this procedure, a mucosal incision is made just above the dentate line, the submucosa is separated from the muscularis, and the muscularis is then divided or a strip excised. The excised specimen should include a segment of the internal sphincter and should demonstrate ganglion cells present at the most cephalad portion of the myectomy.[8] Should the entire specimen be devoid of ganglion cells, the patient most likely will require further surgery to resect the remaining aganglionic intestine.

Thirty-eight adolescent and adult patients managed with anorectal myectomy are described in the literature. Of note is that the procedure is relatively complication-free, with only one minor complication reported (Table 3). Long-term follow-up demonstrates poor results in

40 percent of patients. These patients subsequently required further surgery, presumably to resect aganglionic intestine not completely removed with the myectomy. Nonetheless, 55 percent of the patients had excellent results from this low-risk, relatively simple procedure.

Posterior Anorectal Myectomy with Low Anterior Resection: Due to the poor long-term results with anorectal myectomy alone, a combined procedure of anorectal myectomy with low anterior resection has been proposed.[16] Anorectal myectomy is initially performed, and if the cephalad resection margin demonstrates aganglionosis, a subsequent anterior resection can be performed to remove the remaining aganglionic intestine. Long-term results with this combined approach have been good (100 percent good results), and the procedures have been complication-free.

Low Anterior Resection (State Procedure): Anterior resection alone has been described in the management of 18 adult patients. Only 72 percent of the patients have had good results and there has been one reported death (6 percent).[11]

Colectomy: Left hemicolectomy or subtotal colectomy has been used to treat 15 adult patients. Although the extent of the colectomy has not been carefully defined, 67 percent of the patients have had good results, and one death has been reported (7 percent).[11,18]

Discussion

Adult Hirschsprung's disease is an uncommon disorder, since congenital aganglionosis is most often diagnosed in infancy and early childhood.[39] Nonetheless, several reports document this entity in adults, and it must be considered in any adult with prolonged, refractory constipation.

The fundamental lesion of Hirschsprung's disease is absence of ganglion cells in Auerbach's and Meissner's plexuses.[40] The absence extends from the anus for a varying distance proximally, which may involve as little as the distal rectum or as much as the entire colon,[39] and even the entire intestine. Skip areas are believed not to occur, implying that a rectal biopsy containing ganglion cells rules out the diagnosis of Hirschsprung's disease. Balloon motility studies have demonstrated that lack of relaxation and absence of peristalsis in the aganglionic bowel results in a functional intestinal obstruction.[41] It is presently believed that proximal hypertrophy of the normal intestine may overcome the distal obstruction, especially in short-segment disease, such that some patients may survive into adulthood with congenital megacolon before they require surgical treatment.[10]

Characteristically, adolescent and adult Hirschsprung patients present with lifelong, refractory constipation. Many rely on cathartics and enemas, and multiple hospitalizations for abdominal pain, distention, and partial bowel obstruction are not uncommon. Despite multiple medical visits, the diagnosis typically remains elusive, as was the case in each of our patients. Of interest is that, in one of our patients, Hirschsprung's disease was exacerbated by pregnancy, thereby leading to a definitive diagnosis. This is a rarely reported phenomenon.[19,20]

Any adult with lifelong constipation and no fecal soiling should be evaluated for Hirschsprung's disease. Abdominal radiographs, unprepped barium enema, anorectal manometry, and rectal biopsy are all utilized to confirm the diagnosis.

Plain abdominal x-ray often demonstrates massive dilation of the colon proximal to the aganglionic segment. In such patients, unprepped barium enema is indicated so that the transition zone is not altered by a mechanical preparation.[21] Barium enema will generally reveal a narrow rectum representing the aganglionic segment, and a markedly dilated colon proximal to this segment.[21] A cone-shaped transition zone is classically described.[39] A normal examination does not rule out the diagnosis, however, with approximately a 20 percent false-negative rate reported.[11,42,43] In our series, one of five patients had a nondiagnostic barium enema.

Although not used in our series, anorectal manometry has been increasingly advocated as a screening test to differentiate idiopathic megacolon from adult Hirschsprung's disease.[44] In normal individuals, transient rectal distention causes relaxation of the internal anal sphincter. In Hirschsprung's disease, sphincter contraction rather than relaxation occurs. Greater than 90 percent diagnostic accuracy has been reported.[15,16,42]

Rectal biopsy is currently regarded as the standard diagnostic test for Hirschsprung's disease at any age, with absence of ganglion cells establishing the diagnosis. Due to the thickness of the rectal mucosa in adults, suction rectal biopsy commonly used in infants is usually insufficient, and full-thickness biopsy is required. Increased acetylcholinesterase activity in the aganglionic specimen may also be used to confirm the diagnosis.[22] Rectal biopsy can occasionally prove eqivocal.[23,43] One of our patients initially had a biopsy with ganglion cells reported, but on repeat biopsy six years later, Hirschsprung's disease was confirmed with aganglionosis demonstrated 2 cm above the anal verge. This underscores the need to use barium enema and manometric studies and to repeat rectal biopsies in patients with equivocal results but suggestive clinical histories.

The surgical management of adult Hirschsprung's disease remains controversial. Even in an extensive

review, statistically significant differences in results can not be advanced due to the variety of procedures used, the varying skill of multiple surgeons involved, and the improvements in patient care since the first studies were presented. Further, there exists a definite bias toward the publication of favorable results, such that for a rare disease as this, a number of different procedures appear to be effective. Nonetheless, general conclusions may be drawn regarding postoperative complications and functional results.

The Soave endorectal pull-through has been associated with good results in the management of pediatric patients[37,45] and has the advantage of eliminating the risk of pelvic nerve injury as all of the dissection is carried out within the rectal muscular cuff. Long-term results with this procedure in adults are generally good, although the incidence of postoperative complications in adults is much higher than that reported in children.[37,45] This may mitigate against its use in adults, particularly for a surgeon with little experience with pull-through procedures.

When performed in adults, the Duhamel procedure appears to have a lower major postoperative complication rate (10 percent) than the endorectal pull-through procedure (25 percent). As with the endorectal pull-through, the primary complication is anastomotic disruption leading to abscess or fistula formation. Long-term results are also excellent despite these complications (91 percent good results). This procedure also has the advantage of avoiding an extensive pelvic dissection. The classic Duhamel procedure has been criticized because of the risk of postoperative constipation due to a retained rectal septum or stenosis at the colorectal anastomosis.[11] The Martin modification reduces the incidence of these complications.[1]

The Swenson procedure has been associated with excellent results in the hands of Swenson and his associates, but these results have been difficult for other surgeons to reproduce in both children and adults. In 1975, Swenson *et al.*[38] reported their collective results with 282 patients including 71 adults treated with the Swenson procedure and noted a 5 percent incidence of anastomotic leak and 90 percent excellent long-term results. No patient became impotent. These results have not been duplicated in adults by other surgeons who have experienced a combined 33 percent major complication rate, a 7 percent rate of impotence, and only 80 percent good long-term results. The need for an extensive pelvic dissection and the high incidence of anastomotic complications make the Swenson procedure risky for surgeons without a previous successful experience with this procedure.

Recent reports have advocated anorectal myectomy as an initial approach to adult Hirschsprung's disease due to its low morbidity and technical ease of performance.[7,16] Although 40 percent of the patients had poor results, many of these patients were subsequently successfully treated with the addition of anterior resection. Therefore, anorectal myectomy has been advocated as a safe low morbidity procedure in which pelvic dissection is not necessary and treatment failures are relatively easily managed with subsequent bowel resection. Long-term results with anorectal myectomy and anterior resection are excellent and no major complications have been reported, although the patient series is small. Lynn and Van Heerden[8] report that patients in whom ganglion cells are found at the upper end of the myectomy can be considered cured and will not require a subsequent procedure. McCready and Beart,[11] however, found this not to be the case in their series, with two of five myectomy treatment failures having ganglion cells at the proximal resection margin. This fact makes it difficult to separate patients who need only a myectomy from those who could benefit from both myectomy and anterior resection. Some patients may undergo needless anterior resections while others will have definitive treatment delayed with the attendant morbidity of possible recurrent megacolon, acute obstruction, or perforation. Nevertheless, the excellent long-term results and low complication rate make myectomy with anterior resection a very attractive procedure for adult Hirschsprung's disease, particularly for surgeons without extensive experience with pull-through procedures.

Low anterior resection alone as well as left hemicolectomy or subtotal colectomy have been advocated in some early reports.[11,18] These procedures, however, do not address the need to correct the rectal aganglionosis and therefore are predisposed to recurrent megacolon. They are no longer commonly used to treat Hirschsprung's disease. Surprisingly, two thirds of the reported patients treated with these procedures had excellent results. In all likelihood, these patients either were originally misdiagnosed as having Hirschsprung's disease and actually had functional constipation, or had short-segment aganglionosis such that resection of the megacolon and anastomosis of normally innervated colon provided sufficient propulsive force to overcome the residual short-segment rectal aganglionosis. Regardless, because they leave residual aganglionic intestine behind, low anterior resection and left hemicolectomy generally do not provide adequate treatment for Hirschsprung's disease.

Clearly, there is no obvious best choice of a surgical procedure for the treatment of Hirschsprung's disease in the adolescent and adult. The endorectal pull-through procedure, the Duhamel-Martin procedure, and anorectal myectomy with low anterior resection are all

associated with very good long-term results. As in the treatment of childhood congenital megacolon, a successful outcome depends as much on the surgeon's experience as with the procedure chosen. It must also be noted that for the successful management of adult Hirschsprung's disease, diversion with colostomy before a definitive procedure is often essential to allow for nutritional improvement, adequate colonic cleansing, and return of the hypertrophied, distended megacolon to normal caliber. Bower,[12] in fact, contends that adequate preoperative preparation with a colostomy, and not the type of definitive procedure, may be the most important contributor to a successful outcome. Regardless, the Soave procedure, the Duhamel-Martin procedure, and anorectal myectomy with anterior resection are all appropriate procedures in the hands of well-trained surgeons.

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