CHAPTER 26

Hirschsprung's Disease

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INTRODUCTION

Hirschsprung's disease (HD) is characterised by an absence of ganglion cells in the distal bowel and extending proximally for varying distances. The absence of ganglion cells has been attributed to failure of migration of neural crest cells. The earlier the arrest of migration, the longer the aganglionic segment. The pathophysiology of Hirschsprung's disease is not fully understood. There is no clear explanation for the occurrence of spastic or tonically contracted aganglionic segment of bowel.

The aganglionosis is confined to rectosigmoid in 75% of patients, sigmoid, splenic flexure or transverse colon in 17% and total colon along with a short segment of terminal ileum in 8%. The incidence of HD is estimated to be 1 in 5000 live births. The disease is more common in boys with a male-to-female ratio of 4:1. The male preponderance is less evident in long-segment HD, where the male-to-female ratio is 1.5–2 to 1.

Of all cases of HD, 80–90% produce clinical symptoms and are diagnosed during the neonatal period. Delayed passage of meconium is the cardinal symptom in neonates with HD. Over 90% of affected patients fail to pass meconium in the first 24 h of life. The usual presentation of HD in the neonatal period is with constipation, abdominal distension and vomiting during the first few days of life. About one-third of the babies with HD present with diarrhoea. Diarrhoea in HD is always a symptom of enterocolitis, which remains the commonest cause of death.

The diagnosis of HD is usually based on clinical history, radiological studies, anorectal manometry and in particular on histological examination of the rectal wall biopsy specimens. Barium enema performed by an experienced radiologist, using careful technique, should achieve a high degree of reliability in diagnosing HD in the newborn. It is important that the infant should not have rectal washouts or even digital examinations prior to barium enema, as such interference may distort the transitional zone appearance and give a false-negative diagnosis. A typical case of HD will demonstrate flow of barium from the undilated rectum through a cone-shaped transitional zone into dilated colon. In the presence of enterocolitis complicating HD, barium enema may demonstrate spasm, mucosal edema and ulceration.

The diagnosis of HD is confirmed on examination of rectal biopsy specimens. The introduction of histochemical staining technique for the detection of acetylcholinestrase activity in suction rectal biopsy has resulted in a reliable and simple method for the diagnosis of HD. Full thickness rectal biopsy is rarely indicated for the diagnosis of HD. Once the diagnosis of HD has been confirmed by rectal biopsy examination, the infant should be prepared for surgery. Biopsies for frozen sections are taken to determine the extent of aganglionosis and level of transition zone.

In recent years, the vast majority of cases of HD are diagnosed in the neonatal period. Many centres are now performing one-stage pull-through operations in the newborn with minimal morbidity rates and encouraging results. The advantages of operating on the newborn are that the colonic dilatation can be quickly controlled by washouts and at operation the calibre of the pull-through bowel is near normal, allowing for an accurate anastomosis that minimizes leakage and cuff infection. Recently, a number of investigators have described and advocated a variety of one-stage pull-through procedures in the newborn using minimally invasive laparoscopic techniques. More recently, a transanal endorectal pull-through operation performed without opening the abdomen has been used with excellent results in rectosigmoid HD.

A number of different operations have been described for the treatment of HD. The four most commonly used operations are the rectosigmoidectomy developed by Swenson and Bill, the retrorectal approach developed by Duhamel, the endorectal procedure developed by Soave and deep anterior colorectal anastomosis developed by Rehbein. The basic principle in all these procedures is to bring the ganglionic bowel down to the anus. The long-term results of any of these operations are very satisfactory if they are performed correctly.

Figure 26.1

The abdomen is opened via the Pfannenstiel incision. The biopsy site is selected by observing the apparent transitional zone. In the usual case of rectosigmoid aganglionosis, three seromuscular biopsies are taken along the antimesenteric surface without entering the lumen. One biopsy is taken from the narrowed segment of bowel, a second biopsy from the transition zone and a third biopsy from the dilated portion above the transition zone. Biopsies are assessed intra-operatively by frozen section, to determine the level of ganglionic bowel.

Figure 26.2

Many surgeons prefer right transverse colostomy; others advocate performing colostomy just above the transition to ganglionic bowel. Ileostomy is indicated in patients who have total colonic aganglionosis. A right transverse colostomy is convenient in usual cases. We perform a loop colostomy over a skin bridge. A V-shaped incision is made in the right upper quadrant. The V-skin-flap is reflected upwards. The external oblique is split and the internal oblique and transverse abdominis muscles are divided with diathermy. The peritoneum is opened.

Figure 26.1

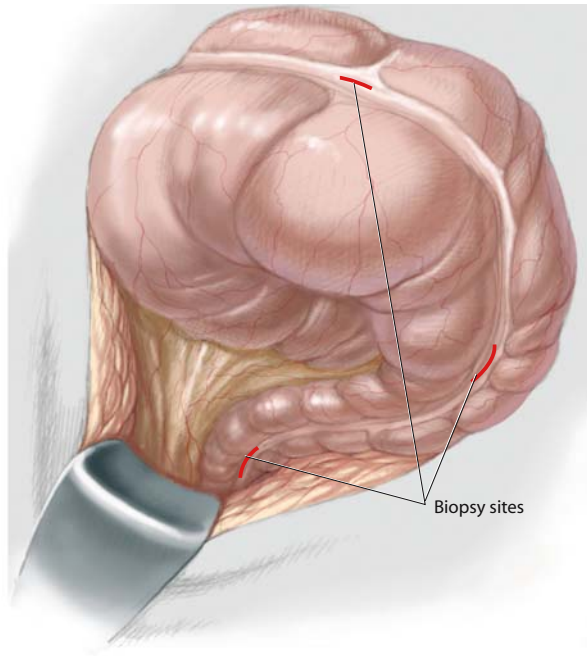


Figure 26.2

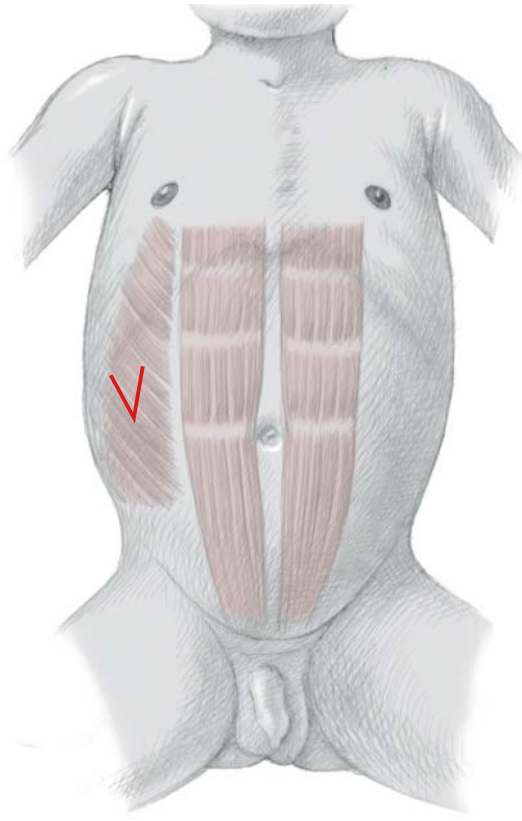


Figure 26.3–26.5

An opening is made in the mesocolon of the selected segment of transverse colon. The skin flap is pulled through the opening in the mesocolon and sutured to the opposite skin margin. A few interrupted absorbable sutures of 4/0 or 5/0 are placed between the peritoneum, the muscle layers of abdominal wall and the seromuscular layer of colon. The colon is opened longitudinally along the antimesenteric border using diathermy. The bowel is sutured to the skin using interrupted 4/0 absorbable sutures.

Figure 26.3

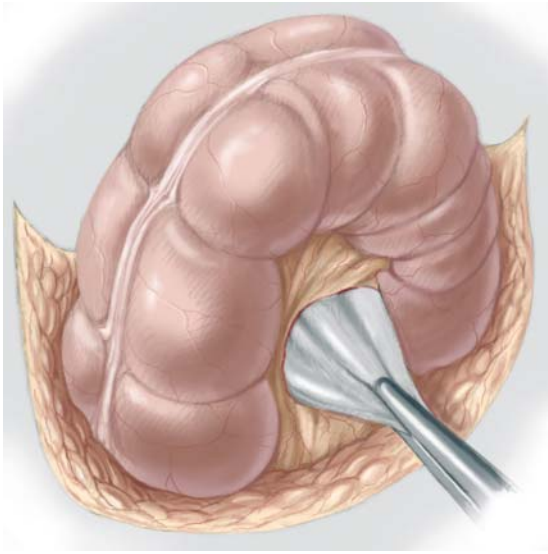


Figure 26.4

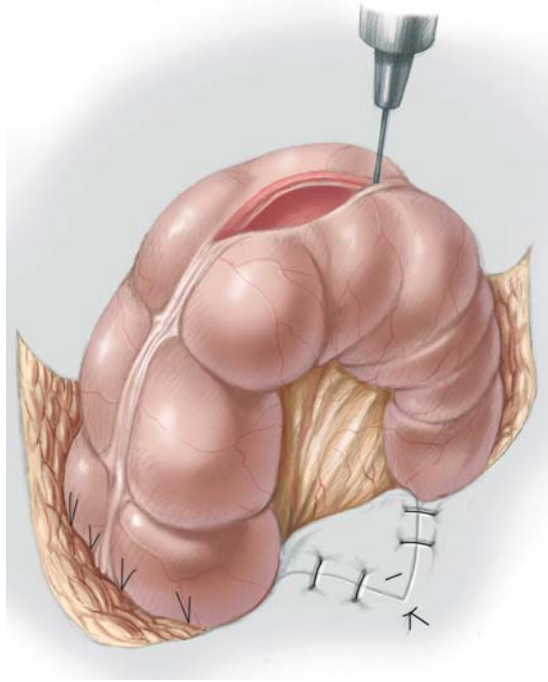


Figure 26.5

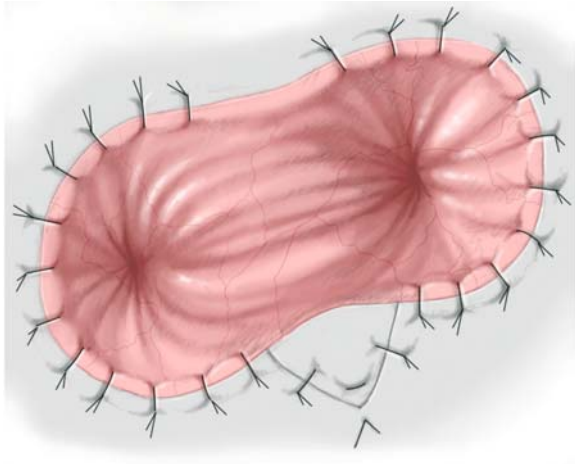


Figure 26.6

Many surgeons have reported good results with the primary neonatal pull-through operation for HD. The author prefers the one-stage transanal endorectal pull-through operation for classical recto-sigmoid HD and Swenson's pull-through operation for long-segment HD because of their simplicity and lack of complications. We have not used diversionary colostomy for usual cases.

Of patients with HD, 75–80% have rectosigmoid aganglionosis. A one-stage pull-through operation can be successfully performed in these patients using a transanal endorectal approach without opening the abdomen. This procedure is associated with excellent clinical results and permits early postoperative feeding, early hospital discharge and no visible scars. Once the diagnosis of HD is confirmed, rectal irrigations are carried out twice a day for 3 days before surgery. Intravenous gentamicin and metronidazole are started on the morning of operation.

The patient is positioned on the operating table in the lithotomy position. The legs are strapped over sandbags. A Foley catheter is inserted into the bladder. A Denis-Browne retractor or anal retractor is placed to retract perianal skin. The rectal mucosa is circumferentially incised using the cautery, approximately 5 mm above the dentate line, and the submucosal plane is developed. The proximal cut edge of the mucosal cuff is held with multiple 4/0 silk sutures, which are used for traction. The endorectal dissection is then carried proximally, staying in the submucosal plane.

Figure 26.7, 26.8

When the submucosal dissection has extended proximally to a point above the peritoneal reflection, the rectal muscle is divided circumferentially, and the full thickness of the rectum and sigmoid colon is mobilized out through the anus. This requires division of rectal and sigmoid vessels, which can be done under direct vision using cautery or ligatures.

Figure 26.6

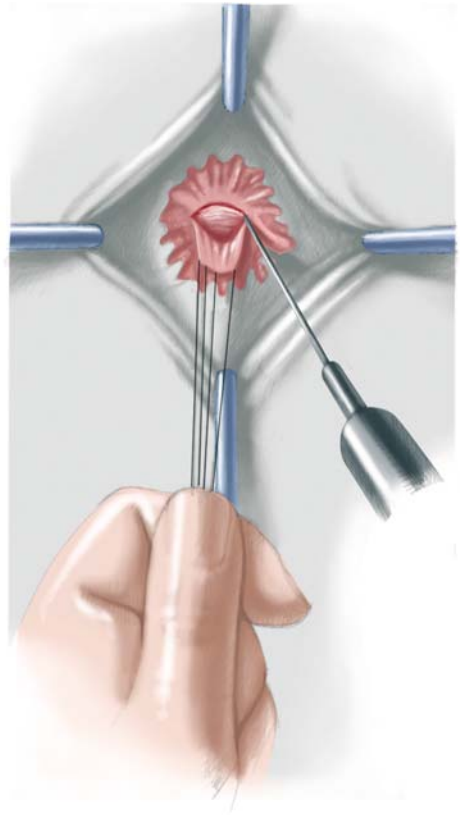
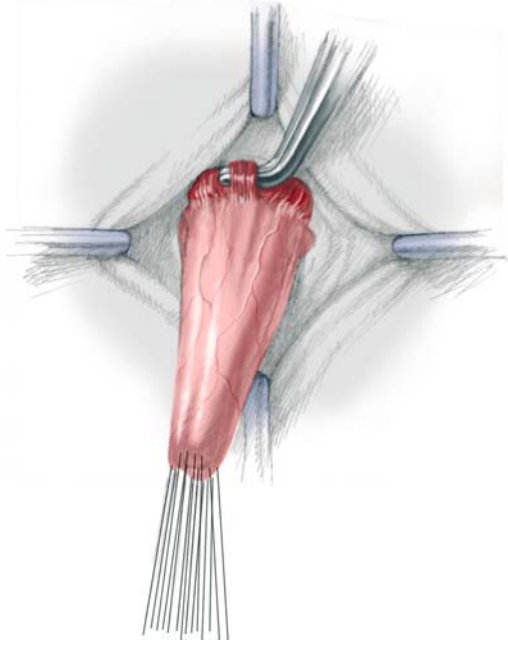


Figure 26.7



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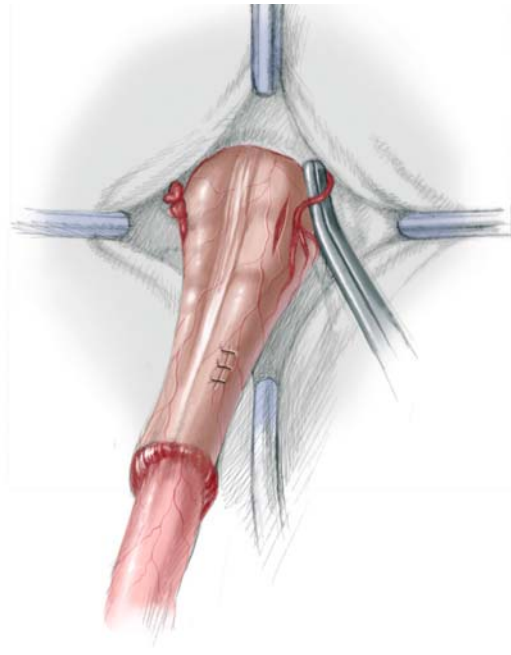


Figure 26.9

When the transition zone is encountered, full-thickness biopsy sections are taken and frozen section confirmation of ganglion cells is obtained. The rectal muscular cuff is split longitudinally either anteriorly or posteriorly. The colon is then divided several centimetres above the most proximal normal biopsy site.

Figure 26.10

A standard Soave-Boley anastomosis is performed. No drains are placed. The patient is started on oral feeds after 24 h and discharged home on the third post-operative day. Digital rectal examination is performed 2 weeks after the operation. Routine rectal dilation is not performed unless there is evidence of a stricture.

Figure 26.9

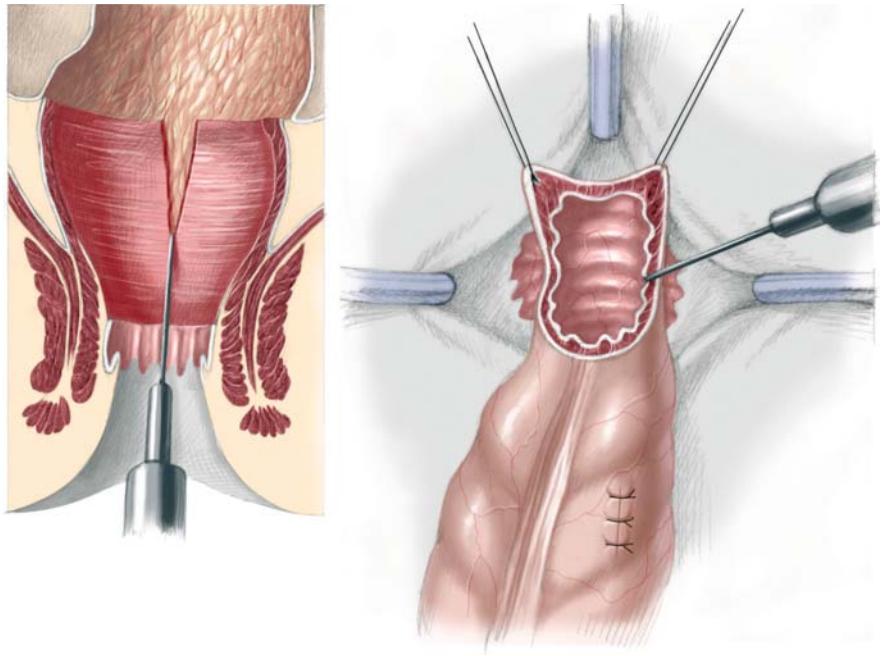


Figure 26.10

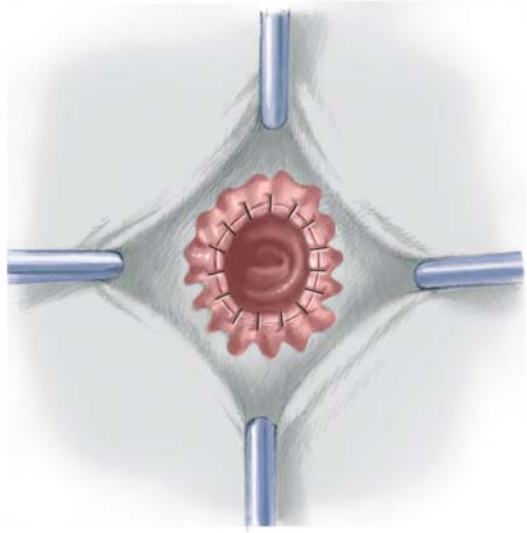


Figure 26.11

For Swenson's pull-through operation the patient is positioned on the operating table to provide simultaneous exposure of the perineum and abdomen. The pelvis is allowed to drop back over the lower end of the table and the legs are strapped over sandbags. A Foley catheter is inserted into the bladder. The abdomen is opened via a paramedian incision. Some surgeons prefer a Pfannenstiel incision when performing a Swenson's pull-through operation in the neonate. Extramucosal biopsies are taken at intervals along the antimesenteric border and assessed by frozen section to determine the level of ganglionated bowel. The sigmoid colon is mobilized by dividing the sigmoid vessels and retaining the marginal vessels. It may be necessary to mobilize the splenic flexure to obtain adequate length. The proximal level of resection above the ganglionated level, previously determined by frozen section, is selected and the bowel is divided between intestinal clamps or staples.

The peritoneum is divided around its lateral and anterior reflection from the rectum, exposing the muscle coat of the rectum. At this point, the bowel is divided at the rectosigmoid junction and removed. Dissection extends around the rectum, keeping very close to the bowel wall. It is essential to maintain the dissection close to the muscular wall in order to prevent damage to the pelvic splanchnic innervation. All vessels are electro-coagulated under direct vision. Sufficient tension-free length is obtained by dividing the inferior mesenteric pedicle, carefully preserving the marginal vessels. Dissection is carried down to the level of the external sphincter posteriorly and laterally, but does not extend as deeply anteriorly, leaving around 1.5 cm of intact rectal wall abutting against the vagina or urethra.

The mobilized rectum is intussuscepted through the anus by passing a curved clamp or a Babcock forceps through the anal canal; an assistant places the closed rectal stump within the jaws of the clamp. When the dissection has been completed, it should be possible to evert the anal canal completely when traction is applied to the rectum. An incision is made anteriorly through the rectal wall about 1 cm from the dentate line, extending halfway through the rectal circumference. A clamp is inserted through this incision to grasp multiple sutures placed through the cut end of the proximal colon. An outer layer of interrupted 4-0 absorbable sutures is placed through the cut muscular edge of the rectum and the muscular wall of the pull-through colon. When the outer layer has been completed, the proximal bowel is opened and an inner layer of interrupted 4-0 absorbable sutures is placed. When anastomosis is completed, the sutures are cut, allowing the anastomosis to retract within the anus.

Figure 26.12

The advantage of the Duhamel pull-through is that very little manipulation of the rectum is performed anteriorly thus avoiding injury to the genitourinary innervation. The rectum is divided and closed just above the peritoneal reflection. The redundant aganglionic bowel is resected. The retrorectal space is created by blunt dissection down to the pelvic floor. The posterior rectal wall is incised 1.5 to 2 cm above the dentate line and sponge holding forceps is inserted into the retrorectal space and ganglionic bowel pulled through. The anterior half of the pulled-through ganglionic bowel is anastomosed to the posterior wall of the aganglionic rectum and remainder of the colo-rectal anastomosis completed by approximating the aganglionic rectum to the posterior wall of the pulled-through ganglionic bowel. Finally an extra long automatic stapling device is used to complete the side to side anastomosis between the aganglionic rectum and the ganglionic pulled-through bowel. Some surgeons complete the side to side anastomosis prior to closing the rectal stump, thereby preventing any residual septum.

Figure 26.11

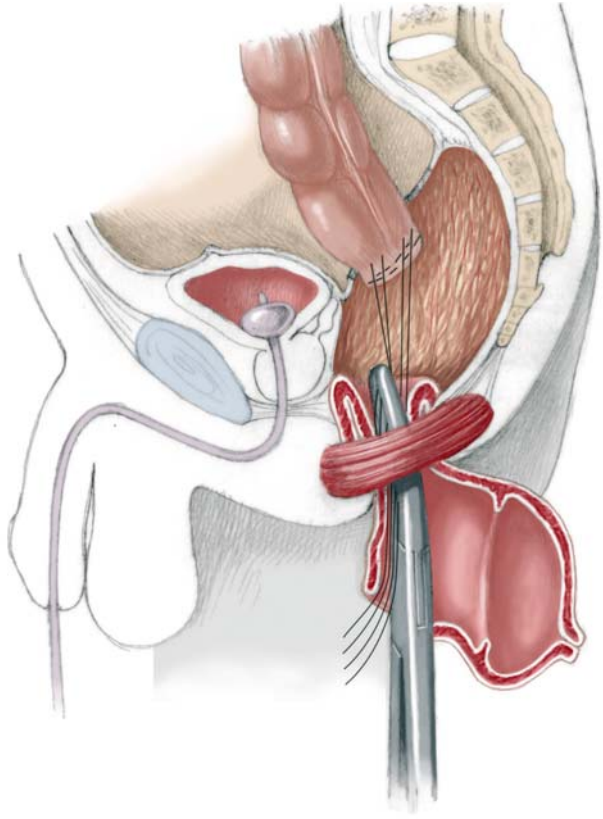


Figure 26.12

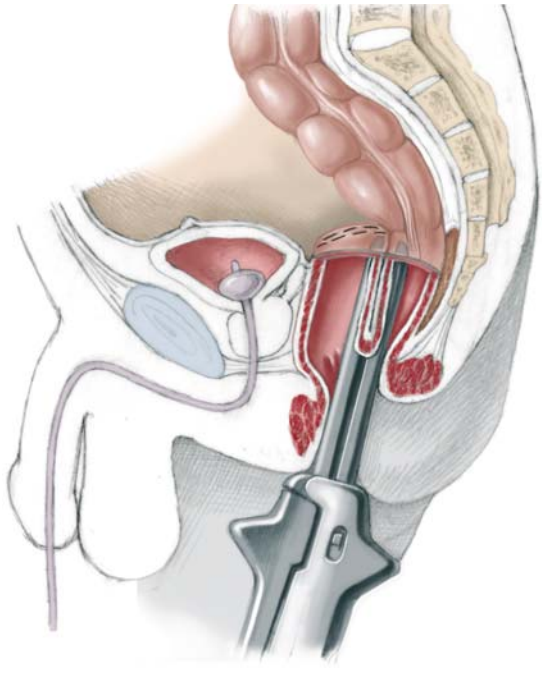


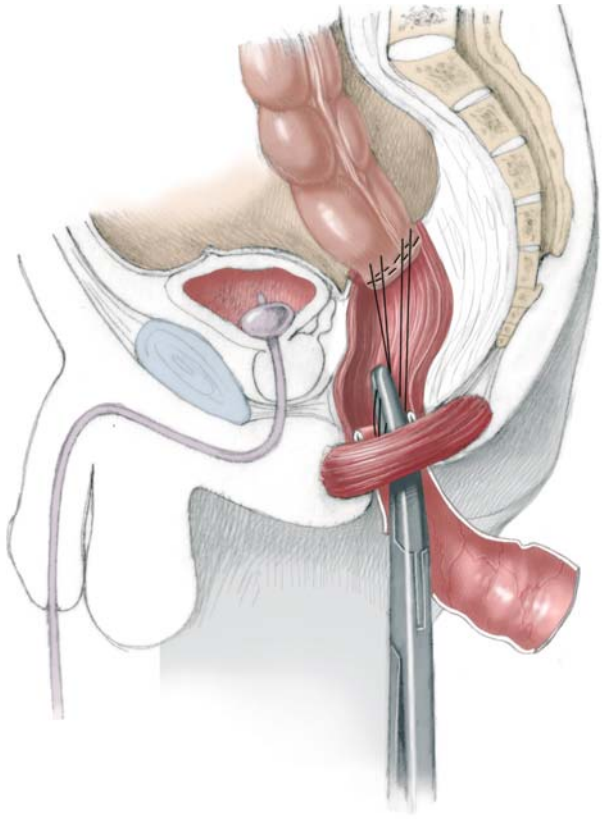
Figure 26.13

In Soave or endorectal pull-through the first steps of the operation are similar to those described for Swenson's or Duhamel operation. The colon is mobilized and resected about 4 cm above the peritoneal reflection. The endorectal dissection begins 2 cm below the peritoneal reflection. The seromuscular layer is incised circumferentially and the mucosal-submucosal tube is freed distally. The mucosal dissection is continued distally to the level of the dentate line. The mucosa is incised circumferentially 1 cm above the dentate line. A Kelly clamp is inserted from below and the ganglionic bowel is pulled through. Coloanal anastomosis is completed using 4/0 absorbable sutures.

Figure 26.14

Rehbein's technique differs from the Swenson's procedure, in that the anastomosis is a low, anterior colorectal anastomosis. In this procedure, 3 to 5 cm of the terminal aganglionic rectum is left behind, which is anastomosed to the ganglionic bowel.

Figure 26.13



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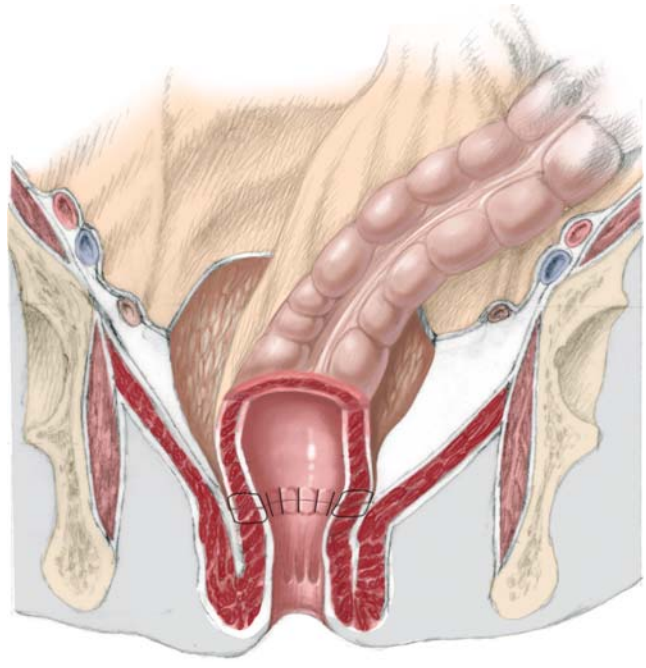

CONCLUSION

The vast majority of patients treated with any one of the standard pull-through procedures achieve satisfactory continence and function with time. The attainment of normal continence is dependent on the intensity of bowel training, social background and respective intelligence of patients. Mental handicap, including Down syndrome, is invariably associated with long-term incontinence.

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