29 Early and Late Complications Following Operative Repair of Hirschsprung's Disease

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29.1 Overview

Since Harald Hirschsprung's classic description in 1886, over 100 papers on complications following repair of Hirschsprung's disease have been published. Original works by Swenson (1948), Rehbein (1953), Duhamel (1956), and Soave (1964) and their predecessors emphasized large single-institution or even single-surgeon experiences rendering comparative outcome analysis difficult. Contemporary surgical management has evolved from the traditional three-stage approach to the more recent introduction of minimally invasive laparoscopic techniques [1, 2] and neonatal one-stage reconstruction [3, 4]. Initial results of these procedures have been limited to single-center or small multicenter series with relatively short follow-up. Although multiple studies have suggested that the popular endorectal technique is safe and efficacious [5–7], the influence on the incidence of late complications is yet to be fully determined. Many of the techniques can also be done in an open or laparoscopically assisted manner. Different risks and benefits are attendant with each of these choices.

The majority of children with Hirschsprung's disease have satisfactory results following definitive pull-through reconstruction. Complications occurring after the surgical repair of Hirschsprung's disease can be temporally categorized into early and late complications. However, there is significant overlap in regard to the time period during which these may occur. Some complications (e.g., wound infection, bleeding, stricture, bowel obstruction, dehiscence, stomal complications) are not unique to Hirschsprung's disease, and are discussed only briefly. Overall, most children with HD do not develop complications within the first 30 days postoperatively. The most commonly encountered late complications are chronic constipation, enterocolitis, and encopresis. Most will present within the first few postoperative months, and symptoms will gradually improve with time. Other complications such as fistulae, and genitourinary and sexual dysfunction, will infrequently be encountered.

29.2 Early Complications

29.2.1 Wound Infection

By definition, surgical repairs of Hirschsprung's disease are classified as clean-contaminated cases. The risk of infection should be low in most cases. Skinner reviewed

over 2500 operative cases and documented a 1.7–19.2% incidence of wound infection for all four primary repairs [8]. Factors contributing to the incidence of wound infections include adequate preoperative bowel preparation, perioperative antibiotics, adequate preoperative nutrition, meticulous hemostasis, length of operation and sterile surgical technique.

29.2.2 Bleeding

Significant postoperative bleeding after definitive repair of Hirschsprung's disease is rare. Obviously, preexisting coagulopathy, sepsis, inadequate intraoperative hemostasis all are potential contributing factors. Hematoma in the early postoperative period may increase the risk of infection and anastomotic complications. With careful technique, this complication should be avoided.

29.2.3 Anastomotic Complications

29.2.3.1 Leak

Anastomotic leak is the most serious of the early postoperative complications. Factors increasing the risk of this complication include: tension, ischemia, technical (inadequate repair), poor nutritional status and other general wound-healing problems (steroids, etc), residual aganglionosis, and distal obstruction. Down's syndrome may be associated with an increased leak rate. One study suggests that the risk of anastomotic leak is independent of patient age or the length of aganglionic bowel [9]. Postoperative rectal manipulation (temperature, examination, or medications) or examination in the early postoperative period may lead to anastomotic problems. A sign should posted at the bedside prohibiting such manipulations. Suspected leaks are usually evaluated with water-soluble contrast enemas (Figs. 29.1 and 29.2).

The incidence of anastomotic failure varies from 1% to 10%. Leaks may be subclinical, resulting in stricture formation. Some studies suggest that a large percentage of strictures result from a small anastomotic leak. Major anastomotic leaks can lead to localized abscess formation or free peritoneal leakage and sepsis. Obviously, more severe leakage may require percutaneous drainage, surgical exploration, diverting proximal colostomy, and eventual anastomotic revision.

29.2.3.2 Pelvic Abscess

The overall incidence should be less than 5%. The same factors resulting in leak are also implicated in this complication. CT scan is the diagnostic procedure of choice. Diagnosis of pelvic abscesses requires a high index of suspicion and subsequent prompt intervention to avoid further morbidity including extension of the infection, systemic sepsis, and necrosis of the pull-through segment. Treatments may range from percutaneous drainage to stomal diversion.

29.2.3.3 Cuff Abscess

The incidence is usually under 7% [10–12]. This complication may occur after the Soave-Boley operation. An abscess is located between the rectal muscularis and the colonic pull-through segment. Factors leading to an increased risk for this complication include: ischemia, retained rectal mucosa, bleeding, pelvic contamination, and tension. Some authors feel that transabdominal peritoneal drainage for the first two to three postoperative days reduces the risk of cuff abscess [13]. Treatment varies from simple broad-spectrum antibiotic coverage (with or without percutaneous drainage) to diversion. Small fistulas or sinus tracts may resolve spontaneously.

29.2.4 Dehiscence

Wound dehiscence occurs in less than 3% of children undergoing definitive repair [11]. Attention to technique, adequate hemostasis, good nutrition, and avoidance of ischemia, tension and infection are preventative.

29.2.5 Retraction of the Pull-Through Segment

The incidence of retraction is less than 10% [10, 14, 15], usually occurring in the early postoperative period. If retraction is suspected, examination under anesthesia will confirm the diagnosis. If very minimal, transanal repair may be attempted. Incomplete retraction can be managed with a proximal diverting colostomy and delayed revision in several months.

29.2.6 Stomal Complications

Stomal problems such as retraction, stenosis, parastomal hernia, skin breakdown, and prolapse can occur. However, the incidence is no different for patients with Hirschsprung's disease than for other diseases. Stomal therapists, working under the direction of a pediatric surgeon, often will detect problems at earlier stages and thus provide for earlier intervention. Avoidance of stomal complications is one of the arguments used by advocates of primary one-stage repair.

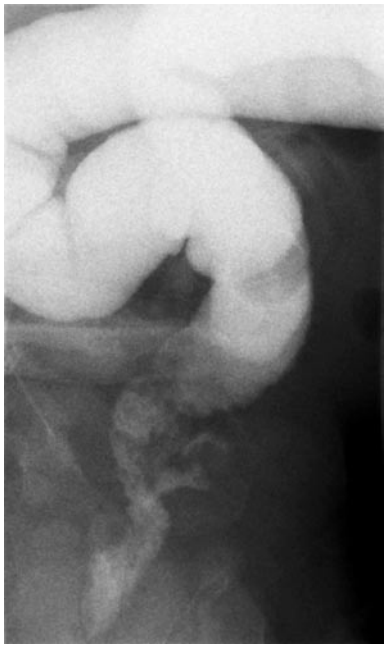

Fig. 29.1 A 1-year-old patient with anastomotic leak following Soave's procedure

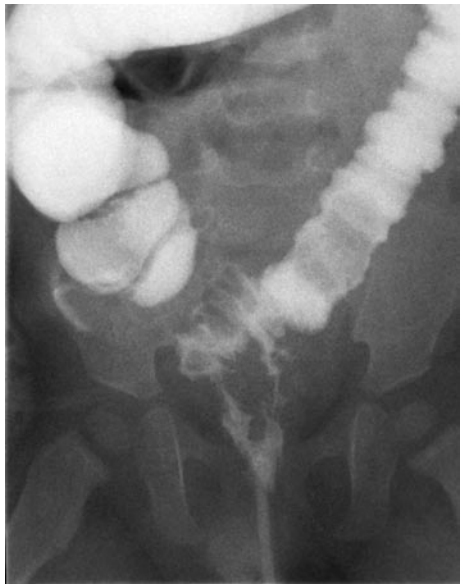

Fig. 29.2 Same patient as in Fig. 29.1 2 months later. A fistula from the distal to proximal rectum has developed

29.3 Late Complications

29.3.1 Enterocolitis

Hirschsprung's enterocolitis is the most serious and potentially life-threatening complication of Hirschsprung's disease. It may occur before or after definitive repair. Enterocolitis of Hirschsprung's disease was first recognized by Hirschsprung in 1886 [16] and Swenson and Fisher in 1956. Bill and Chapman are credited with the first detailed description in 1962 [17].

Despite significant advances in elucidating the genetic etiology of Hirschsprung's disease [18] and improved surgical techniques, little progress has been noted in discovering the etiology or prevention of Hirschsprung-associated enterocolitis (HAEC) (Figs. 29.3 and 29.4). Many theories have been proposed including mechanical dilatation and fecal stasis, alterations of mucin components, increased prostaglandin activity, *Clostridium difficile* infection [19], rotavirus infection, and impaired mucosal immune defense mechanism. Unfortunately, the pathogenesis of enterocolitis is poorly understood. The presence of stasis or relative obstruction may be causative in some patients (residual aganglionic colon, stricture, pelvic inflammation, sphincter achalasia). This perplexing problem includes a wide range of clinical presentations including abdominal distension, explosive diarrhea, vomiting, fever, lethargy, rectal bleeding, and shock [20]. The cost of caring for an infant with HAEC is more than 2.5 times that of an infant with Hirschsprung's disease and no enterocolitis [21].

Historically, a younger age at diagnosis and repair has implied an increased risk of HAEC. Teitelbaum et al. noted a significantly increased incidence of low-grade enterocolitis in infants undergoing a primary endorectal pull-through [6]. These patients are felt to have a more severe disease process. Furthermore, infants who experience enterocolitis before operation have an increased risk of occurrence of HAEC following operation [17]. The older child that 'escapes' the newborn period without detectable disease may have a milder variant. These older children show a different pattern of presentation and a consistently shorter transition zone compared with neonatal disease. A recent report noted that Hirschsprung's disease in the older child did not portend a worse outcome compared with younger children [22]. Enterocolitis is also more common in children with long-segment disease (two- to threefold increase). There is no racial predilection, but it may be more common in boys than girls.

Reports of enterocolitis following operative repair of Hirschsprung's disease varies by publication and operative procedure. Duhamel's repair is probably associated with the lowest rate of enterocolitis. In 4000 cases, Duhamel patients were noted to have a 7.1% incidence of enterocolitis. A Japanese study of 1628 patients noted a considerably higher incidence with 35% following Sw-

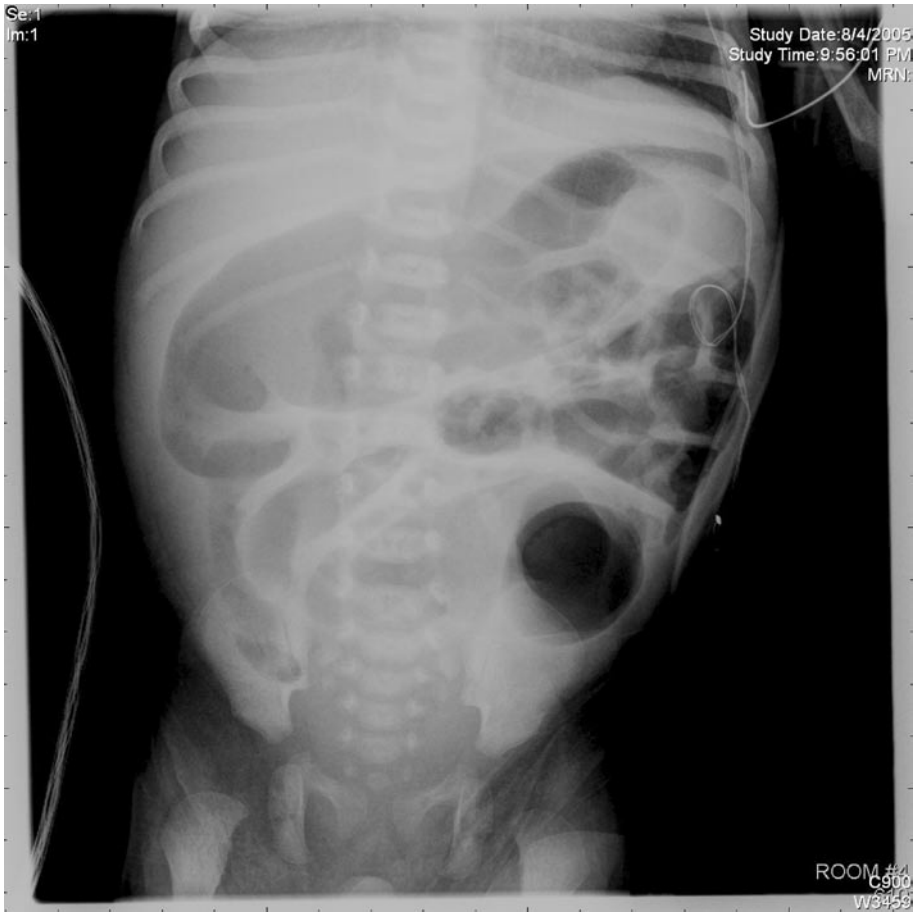

Fig. 29.3 Anteroposterior radiograph demonstrates classic findings of enterocolitis including moderate distension of bowel lumen and edema of bowel wall

enson, 14% following Duhamel, 20% following Soave, and 12% following Boley's procedure [23]. In a survey from the surgical section of the American Academy of Pediatrics, enterocolitis was noted to occur in 16% of children undergoing a rectosigmoidectomy procedure such as Swenson or Rehbein. However, this same group of surgeons reported a 6% incidence following Duhamel pull-through and 25% following the Soave-Boley procedure [15]. Hackam et al. noted a 32% incidence of postoperative enterocolitis in their review of 105 consecutive patients from the Hospital for Sick Children [24]. This incidence correlated with patients having anastomotic complications and intestinal obstruction. Moore et al. note that the incidence of enterocolitis is higher for patients with total colonic aganglionosis than for those with short-segment disease [25]. Patients with trisomy 21 may have a higher risk of HAEC [21], felt to be related to humoral and cellular immune deficiency [26]. In one study, almost 45% of infants with trisomy 21 developed HAEC [21]. Associated anomalies and difficulty in diagnosis may impact the severity of the enterocolitis.

The multicenter analysis of Teitelbaum et al. compared primary endorectal pull-through with a two-stage approach and noted a trend towards a higher incidence of enterocolitis in the primary endorectal pull-through group (42%) compared with those with a two-stage approach (22%) [6]. These authors note that a lower threshold in diagnosing enterocolitis in the more recent years may explain the difference between the two procedures.

The incidence of enterocolitis depends on the type of repair, presence or absence of predisposing factors, and institutional diagnostic criteria for enterocolitis. The reported incidence varies widely in the literature. These factors limit comparative analysis. Table 29.1 shows the incidence of enterocolitis in collected series.

Early recognition with prompt treatment are important for successful outcome. In 1956 Swenson and Fisher advocated rectal tube decompression for the initial treatment of enterocolitis [27]. Rectal decompression and irrigations are still advocated by many in the absence of signs of necrosis or peritonitis. Aggressive fluid resuscitation, bowel rest, and administration of broad-spectrum antibiotics are administered, and resection with diversion is necessary if peritonitis or clinical worsening occurs.

If repeated bouts of enterocolitis persist after definitive pull-through, investigation into mechanical causes should

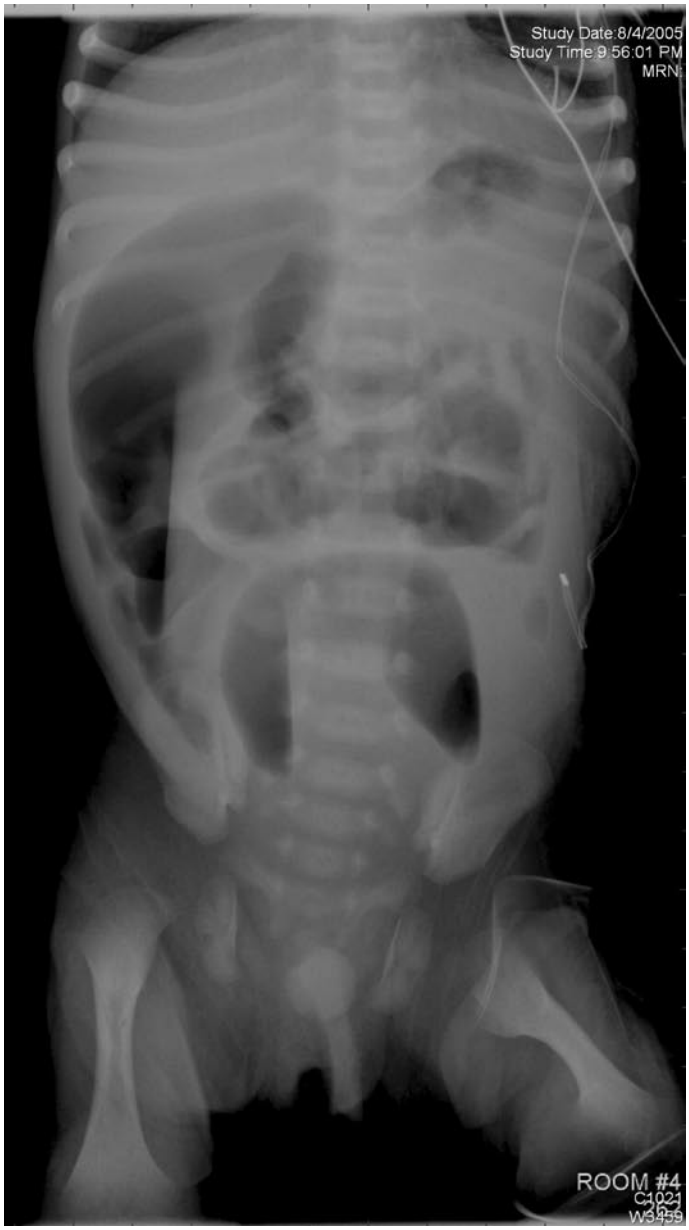

Fig. 29.4 Lateral radiograph demonstrates significant air-fluid levels in a patient with enterocolitis

be considered. Contrast enema, manometry, and rectal biopsy may be necessary. Most patients with enterocolitis will improve over time. Polley et al. and Marty et al. have recommended internal sphincterotomy for those who have persistent enterocolitis despite appropriate investigation [28, 29]. Children with enterocolitis secondary to obstruction may be treated either temporarily by botulinum toxin injection or more permanently with sphincterotomy. In Swenson's series of 880 patients, sphincterotomy was eventually necessary in 6.8% of children [30].

The incidence of enterocolitis directly correlates with mortality. Several series have noted that approximately 50% of deaths are directly related to an enterocolitis episode [12, 15, 29]. In a survey of members of the AAP concerning 1196 patients with Hirschsprung's disease, enterocolitis occurred at the time of diagnosis in 168 patients (14%) with an alarming 30% mortality [15]. In Swenson's series of 880 patients, death after discharge from enterocolitis occurred in about 1% [30].

29.3.2 Constipation

Constipation is probably the most common complaint following surgery (Fig. 29.5). The assessment of severity is highly subjective. The actual rates of constipation may be

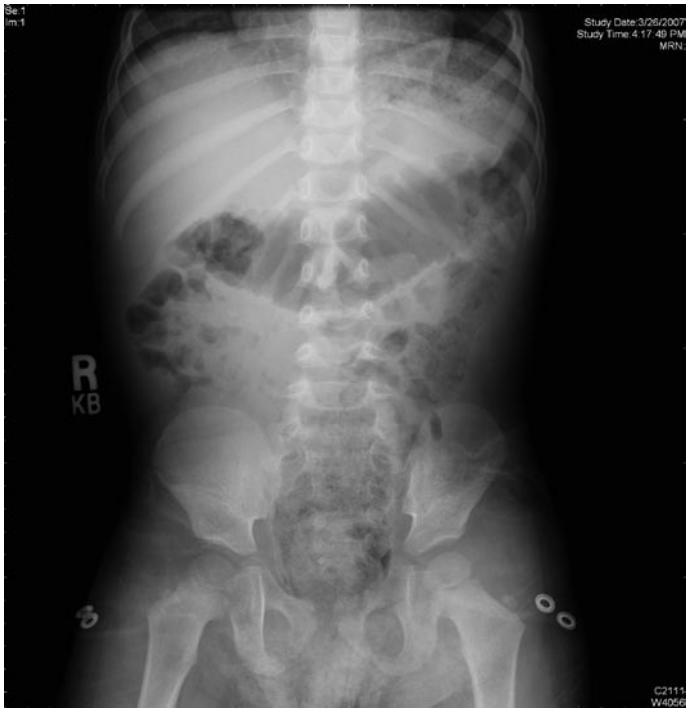

Fig. 29.5 Moderate constipation following Soave's procedure is noted throughout the ascending and descending colon in this 2-year-old patient

underestimated given that many patients are maintained on stool softeners and/or enemas. Rates of constipation between the Swenson, Duhamel, and Soave procedures are roughly equivalent. However, the Rehbein procedure showed a higher rate of constipation necessitating treatment with sphincter dilatation, further resection, or sphincteromyectomy [31]. An increased rate of constipation is not surprising following the Rehbein procedure given that there is a 4–5 cm aganglionic segment left in situ which can become obstructive. A decreased rate of sphincter insufficiency is balanced with increased rates of constipation.

Constipation may result from incomplete resection, sphincter achalasia, stricture formation, fecaloma, neuropathic ganglionic bowel, acquired proximal aganglionosis or may be "functional". Table 29.1 demonstrates the incidence of constipation in collected series. In a pooled sample of almost 8000 patients, the overall incidence of constipation was 7.9%.

Incomplete resection is more likely when frozen sections are relied upon to determine the level of proximal innervation for definitive repair. Accurate interpretation of seromuscular frozen biopsies is paramount in determining the success of the pull-through segment. Frozen sections are prone to sampling and interpretation error. Furthermore, the circumferential distribution of the transition zone is uneven creating a leading edge of ganglion cells extending into the aganglionic distal bowel [32]. Occasionally, these factors result in the use of transitional zone colon for the pull-through. The use of the transition zone for the pull-through segment is associated with an increased risk of enterocolitis, 61% in one series [33]. Treatment options include rectal myectomy and revision of the pull-through [34, 35]. Fecaloma is the presence of a large stool bolus in the aganglionic anterior segment of bowel. It may present with constipation. Alternatively, the obstruction may only allow more liquid material to pass in the form of diarrhea. It is usually associated with Duhamel's procedure, as a result of the partially functional reservoir that has been surgically created. The elimination of blind-ending aganglionic pouches/diverticulum has diminished this complication. The advent of laparoscopically stapled Duhamel procedures could potentially lead to this complication, unless steps are taken to eliminate the blind pouch. The pathogenesis of acquired aganglionosis remains obscure. Etiologies include vascular compromise of the pull-through with subsequent neuronal ischemia, viral infection with neuronal loss, or abnormally innervated proximal bowel. Cohen et al. described five patients (3% incidence) of acquired aganglionosis most of whom were treated successfully with myectomy [36].

Extensive evaluation of mild postoperative constipation is usually not indicated. For those failing a bowel regimen, a more detailed work-up is indicated. Contrast studies will identify pronounced rectal dilation and stricture. Repeat biopsy should be obtained to verify the presence of normal ganglion cells. Manometric analysis to rule out sphincter achalasia or other dysmotility should be obtained. Constipation may be caused by high anal

Table 29.1 Reported long-term complications of combined series: a review of the literature published from 1967 to 2004 (*ERPT* trans-anal endorectal pull-through procedure, *n* total number of patients in the series, † insufficient data)

Complication	Swenson		Duhamel		Soave		Rehbein		ERPT		All[a]	
	n	%	*n*	%	*n*	%	*n*	%	*n*	%	*n*	%
Enterocolitis	3531	13.4	4042	7.1	1268	4.5	440	8.2	†	†	10381	10.6
Constipation	2600	10.3	3567	7.0	571	3.7	367	15.5	149	4.1	7981	7.9
Bowel obstruction	1369	8.3	1288	7.6	1025	5.9	†	†	†	†	4012	7.5
Incontinence	2953	10.8	4010	4.7	1216	4.9	367	8.2	†	†	9063	7.1
Stricture	2188	7.1	3180	2.2	781	6.1	337	9.5	290	4.5	7198	5.0
Mortality	1373	2.8	3591	1.5	902	2.3	191	2.0	149	2.0	6532	2.0

[a]Includes combined series

resting pressure and a weak rectal peristalsis as noted on anorectal manometry and intestinal transit studies with imaging of the anal sphincter complex [37]. Colonic transit studies may provide further insight into the mechanism.

Constipation may be expected to improve over time. Rescorla et al. noted that 88% of patients had difficulty passing stools within the first five postoperative years [12]. However, these symptoms improved with longer follow-up. All patients had satisfactory stooling after 15 years [12]. Lifschitz and Bloss noted that 33% suffered from constipation after the initial operation but only 9% reported persistent constipation after an average of 5 years [38]. Patients with trisomy 21 reportedly have poorer bowel function [39]. Enemas are often necessary to control constipation or soiling after Hirschsprung's disease. Antegrade enemas via button cecostomy or appendicostomy are used in selected patients.

29.3.3 Bowel Obstruction

29.3.3.1 Adhesive

Violation of the peritoneal cavity leads to the formation of intraabdominal adhesions and the possibility of future bowel obstruction. Factors increasing the risk of adhesive obstruction include: prior operation, bleeding, leak, intraoperative contamination, and dehiscence. Early reports noted the incidence of postoperative bowel obstruction to be as high as 18% [23]. A combined review of over 4000 postoperative patients noted an incidence of adhesive bowel obstruction of 7.5% (Table 29.1). With many surgeons now favoring laparoscopically assisted procedures or complete endorectal procedures, the risk of postoperative bowel obstruction may decrease [40]. Fortunately, most bowel obstructions will respond to bowel decompression. In one study, only 20% of patients diagnosed with postoperative bowel obstruction required operative management [41].

29.3.3.2 Internal Hernia/Other

The incidence of internal hernia in most series is <2%. It is important to secure the mesentery of the pulled-through segment to the retroperitoneum in order to prevent this complication. Postoperative intussusception can occur after any operation. Patients with Hirschsprung's disease do not appear to be at any increased risk. If suspected, sonography is currently the diagnostic modality of choice. Another rare cause of early bowel obstruction is a twist of the pulled-through segment. When severe, vascular compromise may arise followed by anastomotic dehiscence.

29.3.4 Continence

Fecal soiling has the greatest negative impact on the quality of life in children with Hirschsprung's disease [42]. Soiling is physically, emotionally, and psychologically disabling [43]. Social withdrawal and poor academic performance are often the end result. Precise assessment of continence is difficult because of the retrospective nature of many of the published reports and lack of objective assessment of children's stooling pattern. Distinctions between occasional soiling and significant incontinence are difficult. Although uncommon, the surgeon should consider the possibility of retained aganglionosis. Additional work-up may include barium enema, manometry, and rectal biopsy. MRI of the pelvis and lower back can be useful in selected patients.

Most children will achieve satisfactory continence with time. Occasional soiling appears to improve over time. Rescorla et al. noted that 12% of their patients less than 5 years of age had some degree of soiling; however, be-

tween 10 and 15 years of age, the incidence declined to 6%. No patient older than 15 years suffered incontinence [12]. Another study found that fecal incontinence was more common in patients less than 15 years of age, but once the child reached late adolescence, bowel control improved significantly with only 8% having fair to poor continence [44]. A review of 880 patients undergoing the Swenson procedure noted that the incidence of soiling decreased from 8% at 5 years' follow-up to less than 2% at 20 years' follow-up [9]. Finally, a review of 2430 postoperative Duhamel patients noted that only 5.3% showed evidence of soiling [45]. Table 29.1 demonstrates the incidence of incontinence in collected series. The combined incidence was 7.1% in nine thousand patients.

Current literature clearly supports gradual improvement in stool continence. Surgeons should maximize medical treatment including the implementation of dietary modifications and bulking agents as the first line of therapy, prior to considering surgical intervention.

29.3.5 Strictures

The incidence of stricture is 8–24% historically, and is more common after Soave and Swenson repairs. Two multicenter reviews of endorectal procedures noted a 4.2–4.8% incidence of stricture [5, 46]. The etiology of anastomotic stricture can be multifactorial, including a narrow muscular cuff, technical complications, compromised blood supply, sequelae following an anastomotic leak, or failure to adhere to a dilation program. Constipation will usually result. Identification is facilitated by digital rectal or proctoscopic examination. Strictures not responding to conservative management may require stricturoplasty or a re-do pull-through procedure. In severe cases, formation of a colostomy and mucous fistula with antegrade dilation over a string may be useful [47].

Additional complications may result from strictures. Rectal dilation may lead to leak, increased constipation, stasis, bacterial overgrowth, dehiscence from tension, or colon retraction. Rectal spasm and colonic inertia can also cause similar problems. Historically, many strictures have responded to conservative outpatient treatment with rectal dilatations with Hegar dilators. Dilations should be avoided for at least 3–4 weeks after the pull-through. As many as one-third to one-half of all clinically significant strictures require surgical intervention [48]. In a review of 7000 patients, the overall incidence of strictures was 5%, with Duhamel procedures having the lowest stricture rates (Table 29.1).

29.3.6 Perianal Excoriation

This is very common after definitive repair and stomal takedown, but usually resolves within 2–3 months. The use of barrier creams beginning on postoperative day 1 may help to limit the severity of this problem. With resolution of postoperative diarrhea, the perianal skin will heal. The incidence of this complication can be expected to decrease with the trend towards neonatal primary repair. Coordinated care with a stomal therapist can be quite valuable in preventing or treating perianal excoriation.

29.3.7 Sphincter Achalasia

Sphincter achalasia is defined as failure of the internal sphincter to relax. Children may present with a myriad of symptoms including chronic constipation or overflow incontinence. Furthermore, children may have difficulty discriminating between solid, liquid and gaseous stools. The use of anal manometry is helpful in evaluating disorders of the internal anal sphincter. A review of patients undergoing postoperative manometry by Harrison et al. noted that persistent loss of normal relaxation of the internal anal sphincter with distension is common, regardless of the technique used [49]. Although many may be asymptomatic, those who are clinically symptomatic may benefit from repeated dilatations or lateral sphincterotomy [50]. Botulinum toxin injections into the internal anal sphincter may be used to asses the potential benefits of later myectomy [51], since the effect of the toxin is transient, usually less than 6 months [52].

29.3.8 Voiding and Sexual Dysfunction

Any operation requiring pelvic dissection places a child at risk for injury to nerves affecting bladder and sexual function. Duhamel's and Soave's modifications were designed to reduce the risk of injury to the delicate pelvic structures. Theoretically, the endorectal procedure should completely avoid injury to pelvic vessels and nerves while protecting the internal sphincter.

The etiology of voiding dysfunction is multifactorial and includes damage to the pelvic splanchnic nerves, the hypogastric nerves, or the pelvic nerve plexus. Parasympathetic denervation to the pelvic splanchnic nerves will lead to a flaccid bladder whereas sympathetic denervation to the hypogastric nerves may result in loss of bladder compliance and incompetence of the bladder neck and posterior urethra [43].

Postoperative enuresis for the different surgical techniques averages 9.5% [53]. Data for individual procedures are as follows: Rehbein 5.4%, Swenson 10.4%, Soave 15.3%, and Duhamel 14.3%. Endorectal follow-up studies of Elhalaby et al. [46] and Langer et al. [5] on a combined 290 patients do not specifically mention urological or sexual dysfunction.

Routine preoperative urodynamic screening is not recommended since children with Hirschsprung's disease are not at increased risk of urological problems. However, a large rectal reservoir may lead to outflow obstruction.

Patients with postoperative urinary complaints should be evaluated, usually initially with sonography and voiding cystourethrography. Urodynamic studies may be needed. Long-term voiding dysfunction is rare.

Discovery of sexual dysfunction requires extensive long-term follow-up. Moore et al. reported sexual dysfunction in 9% following Duhamel's operation and 10% following Swenson's operation [54], with a significantly lower incidence of sexual dysfunction and micturition disturbance following Soave's procedure. The main sexual difficulties identified in female patients were dyspareunia and primary infertility. Male patients voiced concern over poor erections, low sperm counts, or psychosexual problems. A review of 282 patients noted that 101 men with a prior Swenson's procedure had gone through puberty and none had developed impotence. Of these men, 80 were married with a total of 146 children [30]. Another study found a 2.4% incidence of ejaculatory dysfunction in 84 patients after Swenson's procedure [55]. Similar to the data concerning urinary dysfunction, overall assessment of complications demonstrated a significantly ($p<0.01$) lower incidence of sexual dysfunction and micturition disturbance following Soave's procedure when compared to Duhamel's and Swenson's procedures [54].

29.3.9 Mortality

Mortality is low (under 2%) with operative and early deaths being quite rare. Apart from children who die of associated cardiac anomalies or other major anomalies, toxic enterocolitis remains the most common cause of disease-related postoperative death. Additional etiologies include sepsis, abscess, hemorrhage, pneumonia, and embolism [45]. A significant reduction in mortality has occurred over the past 40 years. This may be attributed to improved resuscitation and management of comorbidities, use of parenteral nutrition, earlier detection and prevention of enterocolitis, and improved operative and perioperative care. Table 29.1 demonstrates the incidence of mortality in collected series.

29.3.10 Neonatal Reconstruction and Late Complications

Considerable controversy still exists concerning the efficacy of one-stage neonatal reconstruction. Complications from multistage procedures are well-known. Reliable data concerning the incidence of continence, sexual dysfunction, and constipation for one-stage procedures will require more time, since the procedures are relatively new. Also, as these patients mature they will be better able to voice concerns and thus physicians may more readily identify complications. A comparison of one- and two-stage procedures found a 63% complication rate with the one-stage open technique having a 30% incidence of postoperative enterocolitis [56]. Other studies have shown equivalent rates of complications between the one-stage and multistage approaches. One-stage enterocolitis rates have ranged from 10% to 14%. Another author noted an 8–10% incidence of postoperative enterocolitis [57]. Langer and Winthrop compared one-stage and two-stage Soave's procedures, and found a lower incidence of enterocolitis in babies weighing less than 4 kg [47]. Another study of the laparoscopic approach with one-stage neonatal reconstruction showed a very low incidence of adhesive bowel obstruction. Strictures were rare, and enterocolitis did not occur in this series [2].

29.4 Conclusion

Hirschsprung's disease is a neurogenic intestinal obstruction with potential for chronic illness. A wide spectrum of complications has been reported following definitive repair of Hirschsprung's disease. Enterocolitis remains the most serious late complication following definitive repair. Continued advances in our understanding of the disturbances in bowel motility and the immunological and neurohormonal forces involved in this disorder will result in an improving prognosis.

Traditional multistage procedures still have a role, especially in the very small, critically ill child. The long-term complications of one-stage and laparoscopically assisted procedures are currently not clearly known. Fortunately, the majority of patients with Hirschsprung's disease do quite well following definitive operation regardless of the technique employed. The great majority (94%) of children will become well-adjusted members of society [25]. Early development milestone deficiencies appear to improve over time. Appropriate preoperative conference with family members must include a candid discussion of the importance of realistic expectations and the need for close parental surveillance for late complications.

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