



Hirschsprung's Disease: A Review of Its Management Options

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Review Article

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ABSTRACT

Objectives: This article evaluates current data in literature on the definition, outcomes, and current management strategies for patients with Hirschsprung's disease (HD).

Methods: Different questions about the definition, management, and outcomes of patients with Hirschsprung's disease (HD) had been created. English-language articles published until 2022 had been gathered by searching Scopus, PubMed, Google Scholar and Web of Science.

Results: 61 manuscripts had been included in this review article. The Three well known operations which had been and still available for management of HD are: Soave, Swenson, and Duhamel operations. Transanal endorectal pull-through and laparoscopically assisted Swenson pull-through procedures for Hirschsprung's disease offer a minimally invasive alternatives to traditional open abdominal operations.

Conclusion: The outcomes of laparoscopic assisted trans-anal or complete transanal are slightly superior to other procedures.

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1. INTRODUCTION

Hirschsprung's disease (HD) is considered a functional obstruction of intestine which is caused by ganglion cells absence in the submucosal and myenteric plexuses with the subsequent result of absence of peristalsis in the affected intestinal segment [1]. HD is the most common congenital intestinal motility disorder [2]. HD incidence is nearly 1: 5000 live births, Short-segment Hirschsprung's disease, in which the aganglionosis is restricted to the rectosigmoid area is present in over 80% of cases [3]. The extent of aganglionosis in long-segment HD may involve the entire colon causing a condition named total colonic aganglionosis (TCA) [4]. No available consensus on the best surgical strategy for treatment of HD.

2. METHODS

The aim of this review article is to outline the pathophysiology, diagnosis, surgical approaches, long-term results, new strategies for treatment of patients with HD. Different questions about the definition, management, and outcomes of patients with Hirschsprung's disease (HD) had been created. English-language articles published until 2022 had been gathered by searching Scopus, PubMed, Google Scholar and Web of Science.

3. PATHOPHYSIOLOGY

During normal embryological development, the nervous system of the intestine is formed by neural crest cells proximal to distal migration along the intestine. These pluripotent stem cells differentiate into the submucosal and myenteric plexuses ganglia. HD occurs when there is an interruption of this migration process with subsequent absence of normal innervation to the most distal portion of the intestine. The aganglionic segment is becomes spastic, incapable of dilatation, and causes an obstruction within the proximally dilated normal bowel. Long-segment HD is defined as aganglionosis extending proximally beyond the splenic flexure. The rectosigmoid colon is the most commonly affected area with 80% incidence, followed by transverse colon or the splenic flexure with 17% incidence and finally the entire colon with 8% incidence [5].

4. DIAGNOSIS

Diagnosis of HD depends mainly on clinical history, imaging, and rectal biopsy histopathological examination [6].

4.1 Clinical Presentation

The most important symptom in neonates having HD is delayed passage of meconium beyond 48 hours of life [7]. Distal intestinal obstruction is a common presentation in neonates with HD within the first few days after birth. Vomiting, Distended abdomen, empty rectum and explosive offensive discharge of stool may be present [8]. Chronic constipation in elder children with HD may be a later presentation together with failure to thrive and gross abdominal distention [5]. Children having (HAEC) Hirschsprung's Associated Enterocolitis may present with fever, abdominal distension, and foul-smelling or explosive stool [9].

4.2 Radiological Investigations

4.2.1 Plain X ray of the abdomen

Erect posture view of abdomen may show air fluid levels and absence of rectal air [10].

4.2.2 Contrast enema

A narrow rectum with a dilated proximal colon on contrast enema is diagnostic of (rectosigmoid) short-segment aganglionosis. A transition zone with a variation in lumen size can be easily demonstrated in lateral views of the enema film [11].

4.3 Histopathological Diagnosis

Rectal biopsy is the gold standard for diagnosing HD [12]. The main histopathological finding in HD are: absence of ganglion cells in the myenteric and submucosal plexuses, marked hypertrophy of nerve fibers and abnormally increased acetylcholinesterase activity are [13].

4.4 Anorectal Manometry

The absence of the rectoanal inhibitory reflex (RAIR) in HD patients had been formerly used in diagnosis, but it is of a limited value currently, except for diagnosing anal sphincter achalasia [14].

4.5 Variant Hirschsprung Disease

This term is used to describe a clinical presentation suggestive of HD but with presence of ganglion cells on rectal biopsy [15].

4.5.1 Intestinal Neuronal Dysplasia (IND)

There are two types of IND. The less common type A has a characteristic absent or reduced sympathetic innervation of the myenteric and submucosal plexuses with hyperplasia of the myenteric plexus. The more common type B consists of dysplasia of the submucosal plexuses, nerve fibers thickening, increased acetylcholinesterase staining, giant ganglia and ectopic ganglion cells identification in the lamina propria. The later type B can occur alone or accompanying HD. IND may be either diffuse or focal [16].

4.5.2 Hypoganglionosis

Hypoganglionosis which is characterized by small and scattered ganglia with accompanying acetylcholinesterase distribution abnormalities [17].

4.5.3 Internal sphincter achalasia

Internal sphincter achalasia which is characterized by presence of symptoms of HD in spite of presence of normal ganglion cells on rectal biopsy together with lack of normal (RAIR) recto anal inhibitory reflex on anorectal manometry [18].

4.6 Treatment of HD

Surgical intervention is the definitive treatment of HD with resection of the aganglionic intestine and anastomosing the normally ganglionated intestine to the rectum with preservation of continence [19].

4.6.1 Preoperative considerations

Preoperative considerations preoperative resuscitation, correction of any electrolytes abnormalities and rectal irrigation with warm saline every six hours to relieve any obstruction [20]. Colostomy may be needed to decompress severely dilated colon [21]. Triple intravenous antibiotic coverage should be given one hour before surgery and repeated postoperatively three times daily [22].

4.7 Surgical Procedures

4.7.1 Swenson procedure

The aim of this operation is to resect the aganglionic bowel with an end-to-end anastomosis above the anal sphincter. It is extremely important to maintain the dissection very near to the rectal wall to avoid injury to the deep pelvic vessels, nerves, vas deferens, seminal vesicles, prostate and vagina [23].

4.7.2 Soave procedure

It had been described by Franco Soave in 1964 and its principle depends on removal of the mucosa and submucosa of the rectum and pulling normally ganglionated bowel through aganglionic muscular cuff [24]. The main advantage of this procedure is minimizing the risks of pelvic structures injury [25].

4.7.3 Duhamel procedure

It involves bringing the normally ganglionated colon between the rectum and sacrum and anastomosing the two walls with a stapler to produce a new lumen that is aganglionated anteriorly and normally ganglionated posteriorly. It is felt to be safer, easier than the Soave or Swenson operations and create a large anastomosis with reduction of stricture incidence. In spite of these advantages, It leaves a large area of aganglionic intestine with the risk of enterocolitis, obstructive symptoms and constipation [26-28].

4.7.4 Transanal Endo-Rectal Pull Through (TERPT)

Transanal mucosectomy for HD is a modification of the Soave's procedure. It had been first described in 1998 by De La Torre-Mondragon and Ortega [29,30]. Pelvic nerves damage can be obviated if the dissection is done directly on the rectal wall. A short muscular cuff is left, so cuff abscesses occurrence is minimized and rectal compliance is increased [31]. No abdominal opening is needed; so, adhesion risks are reduced with excellent cosmetic results [29].

4.7.5 Laparoscopic assisted pull through for HD

This operation had been first described by Georgeson et al., in 1995 in 12 patients with excellent results [32]. The benefits of this

technique are preservation of the pelvic nerves, simplicity and skipping the obstructing muscle cuff that could happen following the Soave procedure [33].

4.7.6 Laparoscopic assisted Swenson pull through

In the laparoscopic part of this technique full thickness rectal and colonic mobilization is being done using unipolar diathermy in a plane directly on the bowel wall to avoid injury of any pelvic organ [34]. In the perineal part, a full thickness rectal incision is being done above the pectinate line and a full thickness Swenson plane is being created circumferentially, the dissection is being continued until reaching the peritoneal reflection, then coloanal anastomosis is being done [35].

4.7.7 Laparoscopic Duhamel procedure

In addition to the advantages mentioned in the open Duhamel technique, the laparoscopic version of the procedure adds a lower risk of injuring pelvic nerves, good visualization, minimizing postoperative pain and quicker recovery [36].

4.7.8 Laparoscopic soave procedure (laparoscopic assisted transanal endorectal pull through)

This procedure avoids dissection in the pelvic cavity and theoretically lower the incidence of pelvic organs and nerves injuries [37].

4.8 Complications Following Repair of Hirschsprung's Disease

The main postoperative complications include enterocolitis, incontinence and obstructive symptoms, regardless of which operation is performed [38].

4.8.1 Specific complications of laparoscopic pull through surgeries

4.8.1.1 Trocar associated complication

Abdominal hematoma, incision site bleeding, abdominal organs injuries or large blood vessels injuries can cause extensive bleeding, fistula formation and even death [39].

4.8.1.2 Electric and ultrasonic associated complications

Improper use of ultrasonic or electric instruments as shear, electric hook or ultrasonic scalpel, can

cause severe thermal injuries to the tissues nearby [40].

4.8.1.3 Complications related to creation of pneumoperitoneu

Subcutaneous emphysema is the most common complication [41].

4.8.2 Other complications of pull through procedures

4.8.2.1 Early complications

Early and late complications may be related to each other for example: (leaks may lead to scarring or stricture, anal canal destruction results in permanent incontinence) [42].

4.8.2.2 Wound infection

This can be lowered by perioperative antibiotics, preoperative bowel preparation, good hemostasis and sterile surgical technique [42].

4.8.2.3 Bleeding

Hematoma formation may increase anastomotic complications and the risk of infection [42].

4.8.2.4 Anastomotic leak

Factors aggravating the risk of anastomotic leak are: tension, bad technique, ischemia, inadequate nutrition, drugs (steroids, etc.), distal obstruction and residual aganglionosis, postoperative rectal manipulation in the early postoperative period [42].

4.8.2.5 Pelvic abscess

Same factors which cause leak may also cause pelvic abscess. If not adequately diagnosed and treated pelvic abscess may cause systemic septicemia and pull-through segment necrosis [43].

4.8.2.6 Perianal excoriation

The use of barrier creams may reduce the severity of this complication [42]. This usually resolves in six to eight weeks [44].

4.8.2.7 Anastomotic dehiscence of the pull through

This can occur due to compromised blood supply or inadequate mobilization [45,46].

4.8.2.8 Retraction of the pull-through segment

This problem can occur due to inadequate mobilization or tension of the pull-through segment. Minimal disruptions may be corrected by transanal repair. Severe retraction may need proximal diverting colostomy with a planned delayed revision after several months [42,47].

4.8.2.9 Fistula

This complication may follow leak or perforation. It may be enterocutaneous, rectovaginal or rectourethral. Fistulas can resolve spontaneously or may need a planned redo pull-through procedure [45].

4.8.2.10 Late complications

Incontinence, soiling, enterocolitis or obstructive symptoms can occur [42,44].

4.8.2.11 Bowel obstruction

The five main causes of bowel obstruction are mechanical as (stricture formation, twisted pull through, fecaloma, long rectal cuff, adhesions), pathologic (pull-through of aganglionic bowel or secondary aganglionosis), achalasia of the internal sphincter, motility disorders as (intestinal neuronal dysplasia (IND) and functional enlargement of colon) [48].

4.8.2.12 Constipation

It may result from sphincter achalasia, incomplete resection, stricture formation, neuropathic ganglionic bowel, fecaloma, acquired proximal aganglionosis or functional (stool withholding behaviors) [42].

4.8.2.13 Fecal soiling

Contrast enema, manometry, rectal biopsy and MRI of the pelvis may be needed for diagnosing its cause [49]. Bulking agents and dietary modifications should be the first line therapy before considering surgical intervention [50].

4.8.2.14 Stricture formation

This can be caused by technical complications, a narrow muscular cuff, compromised blood supply, not doing the postoperative dilatation program adequately or following an anastomotic leak. Digital rectal examination can detect it. Conservative measures should be tried first

followed by stricturoplasty or a redo pull-through [51].

4.8.2.15 Enterocolitis

Residual aganglionic segment, stricture, dysmotility, internal anal sphincter achalasia, twist in the pull-through or tight Soave muscular cuff can predispose the patient to enterocolitis [52,53]. Irrigations, dilations, botulinum toxin injections and antibiotic prophylaxis can be used as preventative measures [54,55]. Administration of broad-spectrum intravenous antibiotics, metronidazole, early resuscitation and bowel decompression or even colostomy may be needed for adequate management [56,57].

4.8.2.16 Voiding and sexual dysfunction

This may be caused by damage to the pelvic nerves. Denervation of the parasympathetic fibers to the pelvic splanchnic nerves will cause a flaccid bladder, on the other hand, denervation of the sympathetic fibers to the hypogastric nerves may cause loss of bladder compliance and bladder neck incompetence [58].

5. CONCLUSION

Hirschsprung's disease (HD) is considered a form of functional intestinal obstruction caused by absence of ganglion cells in the myenteric and submucosal plexuses of the intestine, which results in absent peristalsis in the affected bowel. It is also referred to as congenital megacolon or congenital colonic aganglionosis that occurs in 1 in 5,000-7,200 newborns. Hirschsprung's disease is being classified based on the length of involved bowel and location of transition zone. HD is one of the important pediatric diseases, which causes a significant morbidity. Open or laparoscopic Soave, Swenson and Duhamel procedures are available for the surgical treatment of HD. The outcomes of laparoscopic assisted trans-anal or complete transanal are superior to other older procedures. There several postoperative complications of the mentioned procedures that can be avoided by adequate technique and early detection of occurrence of complication.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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