Surgical Techniques for the Treatment of Hirschsprung's Disease—A Historical Systematic Review and Current Status Quo

Ann. Ital. Chir., 2025 96, 12: 1592–1612
https://doi.org/10.62713/aic.3943

Josef Hager[1], Consolato M. Sergi[2,3]

[1]Department of Pediatric Surgery, Medical University of Innsbruck, 6020 Innsbruck, Austria
[2]Department of Laboratory Medicine and Pathology, University of Alberta, Edmonton, AB T6G 2R3, Canada
[3]Anatomic Pathology Division, Children's Hospital of Eastern Ontario, University of Ottawa, Ottawa, ON K1H 8L1, Canada

AIM: Hirschsprung's disease, or aganglionosis of the colon, is a congenital disease characterized by a lack of neurons in the enteric plexus of the large bowel due to either migration failure or defective differentiation of neural crest cells. We aimed to conduct a historical systematic review of available literature and personal experience to exhibit the surgical techniques performed in the past and the current status quo of surgery for Hirschsprung's disease.
METHODS: We searched PubMed, EMBASE, Google Scholar databases for studies examining surgery in aganglionosis or Hirschsprung's disease. The search included all studies published from inception until 9 November 2024. Eligible criteria included all peer-reviewed articles, not necessarily written in English, but also German, French, Italian, and Spanish items dealing with surgery and the prognosis of patients with Hirschsprung's disease after surgical intervention.
RESULTS: A total of 2527 studies were reviewed, and the studies relevant to the surgery were extracted for this historical review. This article delivers a unique historical path and an analysis of some of the most critical surgical techniques for the approach to Hirschsprung's disease, with two outstanding sections on innovative robotic-assisted surgery and life quality after surgery. Currently, surgeons often tailor their approach to Hirschsprung's disease by combining their knowledge and expertise on several surgical procedures and incorporating subtle adjustments based on each patient's imaging, inspection, and pathological findings. The conventional wisdom held that surgeons should perform whichever surgery they felt most comfortable with, regardless of whether a particular technique produced better results. Although the core elements of surgical care have been identified and surgery has been shown to improve outcomes in patients affected with Hirschsprung's disease, the precise etiology and correct treatment for reconstitution of ganglion cells in the aganglionic portions of the bowel are lacking. The surgical treatment options for Hirschsprung's disease have been conceived and implemented after the etiology of Hirschsprung's disease was successfully clarified in the 1940s.
CONCLUSIONS: There is still no internationally valid agreement on which techniques should be used for the various forms of intestinal aganglionosis, especially for total colonic and ultra-short intestinal forms. On the other hand, minimally invasive surgery, artificial intelligence, and machine learning are quickly entering medicine and surgery. These approaches will probably modify the surgical approaches to Hirschsprung's disease. In the future, surgeons may integrate new knowledge derived from proteomics and genomics into current surgical procedures. This integration pinpoints a therapeutic approach that may eventually entail aspects of personalized medicine.

Keywords: intestinal aganglionosis; Hirschsprung's disease; surgery; abdomen

Introduction

Hirschsprung's disease, or aganglionosis of the colon, is a congenital disease characterized by a lack of neurons in the enteric plexus of the large bowel due to either migration failure or defective differentiation of neural crest cells [1]. The Danish pediatrician Harald Hirschsprung reported at the 4th meeting of the German Society for Pediatrics, held in Berlin in 1886, on two children who had died from what he called megacolon congenitum, a disease caused by enterocolitis in this section of the gastrointestinal tract, although he could not explain the circumscribed colon enlargement or wall hypertrophy [2–4]. The reason for his observation was not identified until 1945 by Theodor Ehrenpreis [5] based on clinical and radiological studies, according to correct physiological assumptions. The pathogenesis of Hirschsprung's disease was verified in 1948, namely through the detection of an existing enteric aganglionosis in the aboral (distal) part of the megacolon (lack of ganglion cells in the Auerbach's myenteric plexus and Meissner's or Schabadasch's submucosal plexus), which caused dysfunction of the peristalsis. Despite the fact that this

Submitted: 3 January 2025 Revised: 2 April 2025 Accepted: 11 April 2025 Published: 29 October 2025
Correspondence to: Consolato M. Sergi, Department of Laboratory Medicine and Pathology, University of Alberta, Edmonton, AB T6G 2R3, Canada; Anatomic Pathology Division, Children's Hospital of Eastern Ontario, University of Ottawa, Ottawa, ON K1H 8L1, Canada (e-mail: csergi@cheo.on.ca).

condition may have been recognized before Hirschsprung, his name is inextricably linked to this complex affection. It is primarily a disruption of the migration of the neural crest cells from the neural crest to the intestinal wall during the 5th to 12th embryonic week [1]. Today, the exact cause of congenital colonic aganglionosis, which can occur in one in 3000–5000 children, has not yet been clarified despite numerous studies over two centuries [6–8]. This study aims to offer a historical overview of the procedures used for the surgical treatment of Hirschsprung's disease. It will also involve a short overview of minimally invasive surgery with some data arising from robotic-assisted surgery, as well as some considerations of the long-term outcome of patients affected with Hirschsprung's disease, which has been surgically treated. The following paragraphs will highlight "classical" surgical interventions for treating Hirschsprung's disease according to their chronological beginning and further developments. Despite trying to be objective in the choice of the articles following our systematic review, there is an obvious high degree of subjectivity in assessing surgical techniques for Hirschsprung's disease, which is derived from the authors' personal experiences. This disclosure is evident as a limitation of this historical article.

Classification of Aganglionosis

The aganglionosis affects a colonic segment of individually different lengths, depending on where the neuroblast migration stopped. This malformation is usually limited to the rectosigmoid (about 72% of all patients [range: 64–83%]; so-called short segment aganglionosis) [6,7]. However, it can also affect a more extensive section of the colon. It may involve up to the right colonic flexure (long-segment aganglionosis, approximately 15% of patients [range: 5–24%]), or it may affect the entire colon (Zuelzer-Wilson syndrome, approximately 8% of patients [range: 1–20%]). Very rarely, aganglionosis can extend even further orally and, in extreme cases, even affect the entire gastrointestinal tract. A special form of aganglionosis occurs in about 5% of cases and is so-called ultra-short segment Hirschsprung disease, limited to the anorectal zone below the pelvic floor [9–14]. Kawaguchi *et al*. [15] proposed the following terminology, which has been quite broadly accepted worldwide, although not uniformly:

- Short-Segment Hirschsprung's Disease: Aganglionosis up to the sigmoid colon-descending colon junction.
- Long-Segment Hirschsprung's Disease: Aganglionosis proximal to the sigmoid colon-descending colon junction but with ganglion cells in some portion of the colon.
- Total Colonic Hirschsprung's Disease: Aganglionosis of the entire colon and less than 5 cm of the terminal ileum.
- Small Intestinal Hirschsprung's Disease: Aganglionosis extending proximally to more than 5 cm of the terminal ileum.
- Total Intestinal Hirschsprung's Disease: Aganglionosis of nearly the entire intestine, with less than 20 cm of ganglionated small intestine beyond the Ligament of Treitz.

The term "ultra-short segment Hirschsprung disease" has an unclear definition and a very controversial approach, and it will be treated separately below.

Materials and Methods

We searched PubMed, EMBASE, and Google Scholar databases for studies investigating the use of surgery in aganglionosis or Hirschsprung's disease. The search included all studies published from inception until 9 November 2024. Articles in English, German, French, Italian, and Spanish were retrieved and studied (eligibility criteria). Several techniques have changed over the last two centuries following the discovery of aganglionosis or Hirschsprung's disease, and an additional criterion was the presentation of a surgical technique. The surgical methods were grouped according to the authors, and all databases, registers, websites, and organizations were reported. The Preferred Reporting Items for Systematic reviews and Meta-Analyses (PRISMA) 2020 flow diagram for new systematic reviews, which included searches of databases, registers, and other sources, was used (**Supplementary Material**) [16].

Results and Discussion

A total of 2527 studies were reviewed, and the studies relevant to surgery were extracted in this historical review. Fig. 1 displays the PRISMA flow chart used for the current historical review. The results of the search and selection process are in the flow chart. The number of records identified in the search and the number of studies included in the review are shown. We indicated studies that might appear to meet the inclusion criteria but were excluded, and we explained the reason for the exclusion. The assessment of the risk of bias for each included study is difficult to determine because some studies were conducted in the last century. We cannot offer summary statistics for each group or an effect estimate and its precision due to the temporally different performance of these procedures. The causes of heterogeneity among studies were not investigated. Sensitivity analyses to assess the robustness of the synthesized results were not systematically performed, but an ad hoc assessment of the risk of bias for each synthesis was formulated. The evaluations of certainty (or confidence) in the body of evidence for each outcome are difficult to determine due to the heterogeneity of the studies. In the following paragraphs, we will highlight the therapeutic options in common settings, the therapeutic options in special settings (total colonic aganglionosis, ultrashort-segment of Hirschsprung's disease, short bowel syndrome [SBS]), the use of robotic-assisted surgery in the setting of Hirschsprung's disease, and the quality of life following a surgical procedure for Hirschsprung's disease.

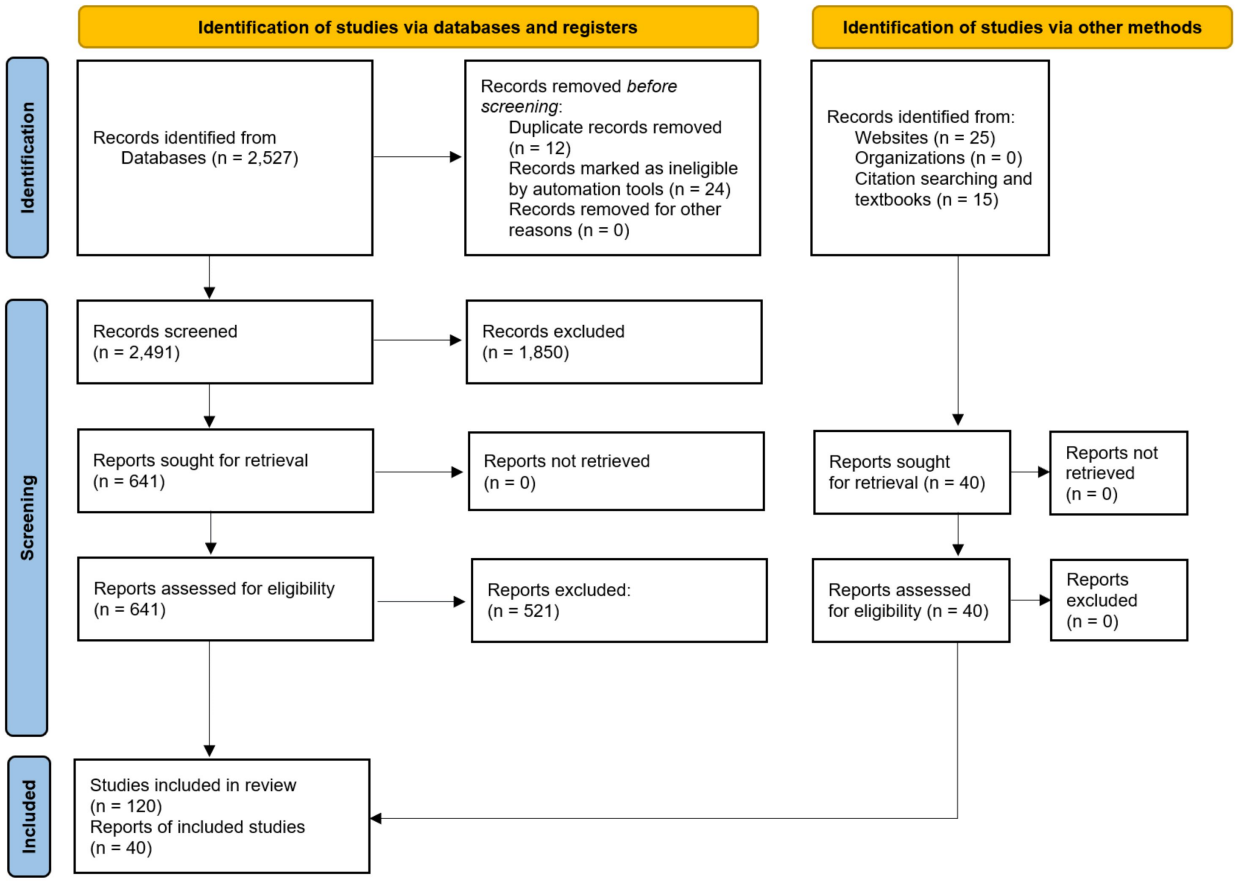

Fig. 1. PRISMA flow chart used to carry out the current historical narrative review. PRISMA, Preferred Reporting Items for Systematic reviews and Meta-Analyses.

Therapeutic Options

If the diagnosis of Hirschsprung's disease is performed, the current therapy is the resection of the hypoganglionic area of the megacolon and the aganglionic colonic section to eliminate the passage obstruction. The first person to suggest that this disorder might be associated with a lack of ganglion cells in the distal rectosigmoid colon was probably Tittel in 1901 [8,17]. Complete excision of the descending colon, sigmoid, rectum, and anus was performed in 1898 by Frederick Treves [18], who believed that mechanical obstruction had clogged the colon. In 1900, Fennwick [19] proposed the concept of lower-end spastic contraction. In 1920, Alberto Dalla Valle [20] found that these patients lacked ganglion cells in the distal, narrow part of the colon. The sole treatment available to these individuals then was a sympathectomy and daily enemas, often with unsatisfactory outcomes. In 1938, Robertson and Kernohan [21] detailed the aberrant peristalsis of the distal aganglionic bowel. Two years later, Tiffin's team [22] carried out a comparable procedure. A functional abnormality of the rectosigmoid region caused by a deficiency or absence of ganglion cells was first reported by Zuelzer and Wilson [23] in 1948. Subsequently, it was verified by Whitehouse and Kernohan [24] in the same year. Regardless of these results, beliefs on the importance of ganglion cell absence evolved slowly. Instead of ganglion cells producing colonic dilation, Ehrenpreis [25] proposed in 1970 that colonic dilation caused the lack of ganglion cells.

Orvar Swenson was developing a surgical cure for megacolon, a condition that was almost always lethal at the time. At the same time as the pathophysiology of Hirschsprung's disease was being uncovered, and for patients suffering from Hirschsprung's disease, he established an experimental study in 1948 that suggested removing the rectosigmoid colon and rectum while keeping the sphincter intact [26,27]. For the first time, it seemed that ganglionic dysfunction was the culprit component of the disease [8]. With pediatric radiologist Neuhauser, surgeons also defined "congenital megacolon" and its causes, symptoms, and treatments [27]. Importantly, Swenson found that those with Hirschsprung's disease did not have mechanical blockage but functional obstruction. The procedure proposed by Swenson was successful, but it was a lengthy and risky operation with a high infant mortality rate. In 1956, a French surgeon named Bernard Duhamel [28] cut the rectum and proximal normal colon in half, creating an anastomosis that ran side to side. Duhamel [29] suggested moving the ganglionated segment to the back of the ano-rectum via the retro-rectal gap, thus

avoiding the rectum. Two clamps, the handles of which protruded from the child's anus, were used in a side-to-side crushing technique to join the retained ano-rectum and the pull-through segment until the walls annealed.

To alleviate postoperative soiling, the Duhamel procedure was modified to place the anastomosis above the internal sphincter. Stapling devices were used to connect the native ano-rectum with the pull-through segment, relieving the infant of the discomfort of clamps dangling from the anus. In 1958, a German surgeon, Fritz Rehbein [30], performed a procedure comparable to an anterior sigma-rectum resection for Hirschsprung's disease. He preserved a generous amount of tissue corresponding to the aganglionic rectum in its original location. Various European researchers, Romualdi, Ehrenpreis, and Pellerin, documented their experiences with recto-sigmoid resection in 1960, 1961, and 1962, respectively [8,31–33].

The initial surgical description of submucosal dissection for Hirschsprung's disease was published in 1952 by Asa Yancey [8,34,35]. To safeguard the adjacent pelvic nerves and genitourinary organs, an Italian surgeon, Franco Soave [36], published a study in 1964 on endorectal (between the mucosa and the muscularis propria) rectosigmoid dissection. This procedure was also described at the same time by Scott Boley [37] in the United States of America.

The distinction between their two methodologies was that Dr. Soave exteriorized the normoganglionic bowel through the anus for several days before subsequently conducting the anastomosis to the anus. Boley, on the other hand, executed a primary anastomosis to the abbreviated residual rectum. For unspecified reasons, Dr. Soave's work was published in the same issue immediately before Dr. Boley's publication. Dr. Yancey initially described this submucosal dissection, and it is crucial to note that Dr. Boley's publication did not provide a variation of the Soave technique but rather a modification of the Swenson technique.

The Martin technique [38], a variant of the Duhamel operation, was created in 1962. Martin was the first Surgeon-in-Chief at Cincinnati Children's Hospital Medical Center, Ohio, USA. An extended aganglionic bowel tract was left *in situ* after his modification, essentially an extension of Duhamel's procedure. To diagnose Hirschsprung's disease, a rectal suction biopsy tube was invented in 1969 by pediatric surgeon Helen Noblett of Melbourne, Australia [39]. While working at the Jewish Long Island Hospital in New York, NY, USA, Henry So, a surgeon born in the Philippines, adopted an endorectal pull-through procedure in 1980 for neonates with Hirschsprung's disease that did not include a colostomy [40].

In their description of a single-stage transanal pull-through surgery, Luis De la Torre-Mondragón and his colleagues [41,42] performed a Soave procedure using a transanal route, which involved beginning a submucosal dissection right above the dentate or pectinate line, which is the junction between the superior and inferior anal canal. To remove the sigmoid colon and rectum from the anus, a muscle sleeve was constructed. Full-thickness biopsies were then taken to locate the transition zone, which was defined as a zone with a poor representation of ganglion cells and a few hypertrophic nerve fibers. The ganglionated section was sutured to the rim of the anal mucosa once the ganglion cells had been identified. Compared to previous colostomy techniques that used laparotomy, on transanal endorectal resection and details of the pull-through technique were later published [8,43]. At the same time, a vast multi-center study proved that patients who had the 1-stage transanal Soave pull-through had a safe operation with minimal pain, early discharge, early feeding, and a low rate of complications overall [44]. Jacob Langer *et al*. [45,46] had previously published their experience with this procedure. At the same time, the groundbreaking article on laparoscopic-assisted endorectal pull-through for Hirschsprung's disease was published by Keith Georgeson *et al*. in 1995 [47] and 1999 [48]. In the following paragraphs, the techniques are described in detail.

Pull-through Technique ("Swenson's Procedure")

Orvar Swenson (Boston, MA, USA) was probably the first pediatric surgeon to present a groundbreaking correction of Hirschsprung's disease [8,26,27,49–51]. His technique was based on correctly interpreting the connection between megacolon congenitum and the colon/rectum aganglionosis existing aborally (opposite to or away from the mouth). The abdominal pull-through operation (initially with a preceding colostomy) involved the following procedure steps. First, the section of the colon, which is located proximally to the megacolon, the hypoganglionic megacolon, and the aganglionic section of the colon, are mobilized up to the rectosigmoid junction. After clinical or radiological findings have been used to determine the beginning of the oral ganglionic section, the colon is resected in this area, as well as the area of the rectosigmoid transition. The remaining sections are blinded and closed. The peritoneal fold is then opened, and the rectoanal section is dissected near the wall to protect the branches of the pudendal plexus and the rectal blood vessels. The closed rectum is pulled through perianally until the dentate line can be seen on the protruding mucosa. The anterior wall of the rectum is then divided obliquely between about 2 cm and the posterior wall about 1 cm above the dentate line. With this oblique anastomosis, the nerve fibers and vessels located anterior to the lowest rectum should also be spared. After removal of the rectum, the normal-ganglionic colon is pulled through the anal canal, and an extra-anal anastomosis is created with the anal mucosa. The anastomosed bowel is then repositioned into the pelvis. The original surgical procedure was revised over the years by Swenson himself and other (pediatric) surgeons because of the complications inherent in the technique (infection, anastomosis leak, incontinence, bladder emptying disorders, and impotence) [51,52].

During the last decade of the 20th century, new surgical-technical options emerged that became relevant to the Swenson method. At the beginning of the 1990s, Swenson's procedure was performed for the first time as a laparoscopically assisted intervention. The principle corresponded to an open procedure. Subsequently, the lowest section of the rectum was more accessible. However, despite reported satisfactory results, the colo-recto-anal anastomosis still created extra-anal issues [53,54]. This procedure was subsequently performed with laparoscopic robot assistance in children aged around 16 weeks and weighing approximately 5.5 kg [55]. In 2003, Weidner and Waldhausen [56], as well as Peterlini and Martins [57], reported on the first successful one-stage transanal procedure in 15 children, albeit with a small left-sided lower abdominal laparotomy to obtain extramucous colon biopsies to define the beginning of the ganglionic colon, the latter using a modified anastomosis technique. In 2008, a Chinese group reported a one-stage transanal procedure using the usual Swenson procedure in 135 children between 9 days and five years, without additional mini-laparotomy, with excellent results [58]. These results were also confirmed by other authors [50,59]. The issue of possible postoperative complications constituted one problem with the Swenson procedure. They were due to extensive preparation of the rectum, especially the ventral-distal section, which was associated with lesions of the pudendal plexus or branches of the A. rectalis superior and A. rectalis media and A. rectalis inferior. In addition, the position of the colo-rectal oral-anal anastomosis was a sticking point in Swenson's procedure. This anastomosis must be located just above the anal ring to trigger the stimulus to defecate and a normal defecation process. The results of long-term follow-ups varied greatly in this respect, not least depending on the technical skills and experience of the respective surgeons and the type of procedure (open, laparoscopic, or transanal) [59]. Nevertheless, the Swenson procedure is still the procedure of choice for many pediatric surgeons worldwide when approaching a patient with Hirschsprung's disease. Just a few years after the Swenson procedure was introduced, the complications mentioned led to new operational and technical considerations to be implemented to avoid these issues. In the following paragraphs, we present techniques which were implemented over the years and their extraordinary developments.

Low Anterior Resection Techniques

"Anterior (partial) Sigma/Rectum Resection" (State, Hiatt)

In 1950s, David State [60–62] (Minneapolis, MN, USA) presented a new concept for treating Hirschsprung's disease, which he called a "physiological" surgical method. He recommended fully resecting the radiologically determined megacolon (the extended hypoganglionic colon section, including transition zone) and the aganglionic recto-colonic segment. Moreover, he formed a colorectal anastomosis at a height of about 7–10 cm from the dentate line [60–62]. In 1951, Robert B. Hiatt [63] (New York, NY, USA) reported on an identical procedure, albeit only as a temporary project, since he soon became involved with the Swenson procedure. He performed an anterior sigmoid/rectal resection with an anastomosis between the middle sigmoid and the remnant of the rectum, with the anastomosis being at a height of 4–6 cm orally (proximally) to the dentate line. However, the length of the aganglionic rectum remaining *in situ* with the State procedure and the remaining sigmoid colon with the Hiatt method caused Hirschsprung's disease to relapse after surgery. This failure is at the root of labeling these procedures as unsuccessful despite their principle remaining partly preserved.

"Deep Anterior Resection" (Rehbein)

Fritz Rehbein [30,64–66] (Bremen, Germany), who had used the Swenson procedure in 1951 and 1952, abandoned this technique because of the abovementioned complications. In 1953, he took up the idea of the resecting procedure propagated by David State. In contrast to State, the German surgeon oriented himself to the anterior deep rectal resection customary in adults for treating colo-rectal carcinomas located more than 6 cm from the ano-cutaneous line [30,64–66]. He aimed to perform a resection of the hypo- and aganglionic colon/rectum section significantly more extensively than State (or Hiatt) after the clinical-visual determination of the oral resection border (usually in the transverse colon). The remaining aganglionic rectal remnant should have been 3 to 5 cm long. The end-to-end anastomosis is located extraperitoneally, about 2 cm below the peritoneal fold. In addition, he recommended a forced dilatation (bougienage) of the anal canal (including the anastomosis) for several weeks after the procedure was performed to reduce any obstruction symptoms. Bougienage is the dilation of a tubular cavity (such as a constricted esophagus) with a bougie, a tapering cylindrical instrument for introduction into a tubular passage of the body.

Rehbein and Wernicke [65] reported on the first (positive) technical experiences as early as 1955. In 1958, Rehbein presented his results for 67 children arising from several experiences [30] and published a follow-up analysis in 1960 [64]. In 1963, Rehbein and Nicolai [66] reported on 110 successfully operated children. Rehbein was convinced that the complete resection of the rectum was probably unnecessary. He suggested that the existing achalasia of the internal sphincter muscle should be treated with bougienage or a partial myectomy [67,68]. His method, using the open technique, was the method of choice for Hirschsprung's disease in the German-speaking world for years. This, even though a deep recto-colic anastomosis, is not a very easy procedure from a technical point of view.

In addition, as mentioned above, the procedure can often result in obstructive problems, which can be attributed to the non-innervated internal anal sphincter and relatively narrow

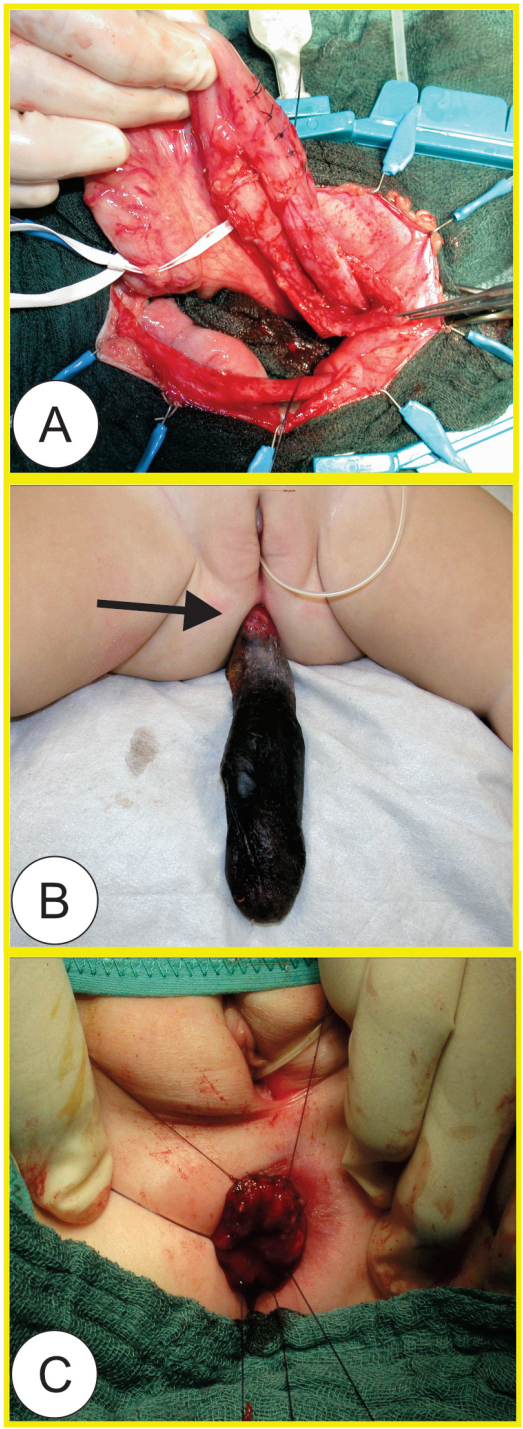

Fig. 2. Soave's procedure for aganglionosis (Hirschsprung's disease). (A–C) Surgical site for endorectal pull-through surgery according to Soave - (A) Sheathing of the rectal mucosa. (B) Transanally pulled through the aganglionic colon section (arrow: rectal stump) on the 6th postoperative day. (C) End-to-end descending rectostomy on the 8th postoperative day. The surgical site gross photographs belong to the personal archive of Professor Dr. J. Hager. Written informed consent was provided by the parents/guardians at the time of the surgical procedure.

colo-rectal anastomosis. With the advent of gastrointestinal stapling techniques to create an end-to-end anastomosis, the remaining aganglionic rectal segment was reduced to approximately 2 cm. However, the procedure was challenging, particularly in young infants, due to the tiny size of the circular stapler (the smallest stapler head had a diameter of 21 mm) [69,70]. In our opinion and personal experience, appropriate anal bougies must be carried out preoperatively to be able to insert the device transanally and connect it to the counter-pressure plate in the oral section of the colon. In 2003, a study involving Rehbein's patients was published, in which 22 pediatric surgical institutions in Germany, Austria, and Switzerland took part [71]. In this study, the surgical results and the postoperative outcomes of the patients were presented in detail. Compared to the standard of other surgical methods, Rehbein's procedure proved to be equally good or even better than previous surgical interventions for Hirschsprung's disease. A renewed Hirschsprung's disease symptomatology was not an issue due to the possible growth in the individual case's relatively long aganglionic rectum segment [72]. This problem, which the author of this work observed in 3 out of 27 of his patients (length of the remaining rectum: 6, 7, and 9 cm), made a redo necessary (resection of the rectal stump and then proceed as with the endorectal pull-through with coloanal anastomosis). Despite many positive long-term results [73], apart from the increased obstructive issues in individual cases, the Rehbein method seems to have lost relevance in pediatric surgery. The transanal endorectal pull-through surgery (TERPT), with no additional stoma installation and faster food build-up, seems to harbor fewer obstructive problems [74,75].

Endorectal Pull-through Procedures

Yancey Technique

In 1952, the American surgeon Asa Yancey [34] (Tuskegee, AL, USA) presented a new surgical technique with which he wanted to avoid the complications associated with Swenson's operation and its extensive rectal mobilization. Yancey described his surgical technique as a modification of Swenson's procedure. His idea was not to dissect the extraperitoneal section of the rectum but to leave it in situ and to remove its mucosa up to about 1.5 cm above the dentate line. After the removal of the hypo/aganglionic sigmoid segment, the *in situ* rectum should have served as a matrix for pulling through the colon containing ganglion cells, which in turn is anastomosed to the anal mucosa. Surprisingly, this technique, which he tested in dogs as experimental animals and used successfully in an adult patient with a rectal disease unrelated to Hirschsprung's disease, found no interest in the USA or Europe. It is unknown why this technique found no favor, but some racial prejudice may have played a significant role, because Yancey was a black surgeon and the Civil Rights Act, a comprehensive U.S. legislation intended to end discrimination based on race, color, religion, or national origin, was not guaranteed until 1964 [8,35].

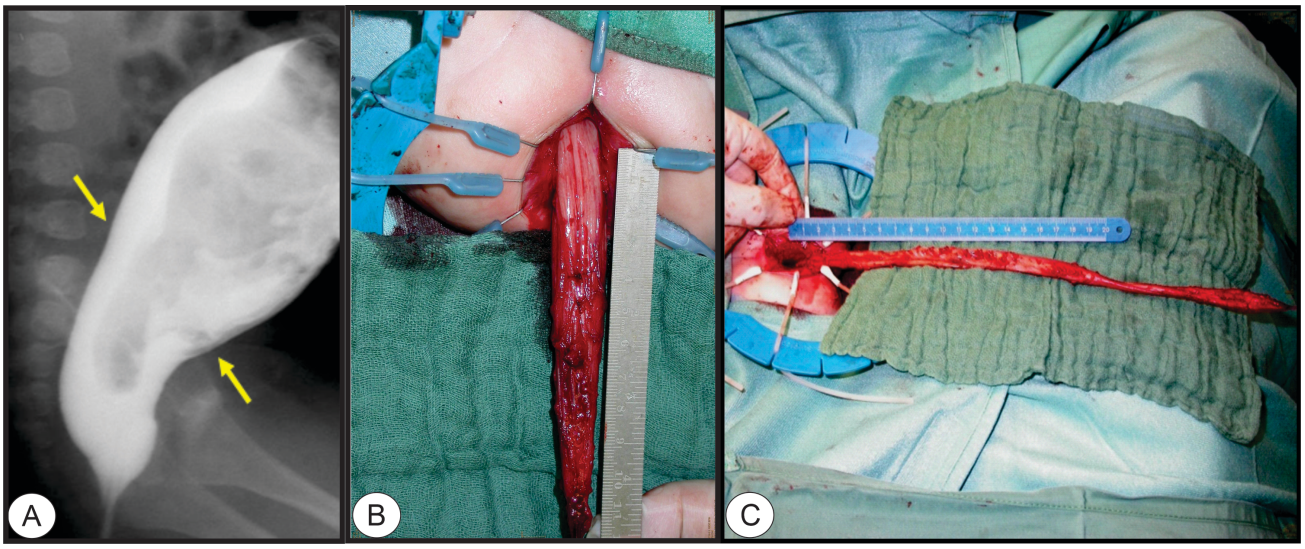

Fig. 3. De la Torre's procedure for aganglionosis (Hirschsprung's disease). According to de la Torre, four steps are critical, including (1) spreading the anal canal, exposing the dentate line and making a circular incision of the rectal mucosa approximately 1 cm orally from it and attaching sutures; (2) sheathing the rectal mucosa; (3) opening the peritoneal fold and mobilizing the sigmoid colon, and (4) taking extramucosal biopsies for determination of the resection limit. In (A) is the X-Ray-imaging of the transition zone between aganglionosis of the rectum and ganglionosis (euganglionotic segment) of the sigmoid colon (arrows). In (B) is shown the operation site of the transanal endorectal rectosigmoidectomy and in (C) is shown the recto-descendectomy performed according to de la Torre. The surgical site gross photographs and X-ray photographs belong to the personal archive of Professor Dr. J. Hager. Written informed consent was provided by the parents/guardians at the time of the surgical procedure.

Soave and Boley Techniques

In 1963, the Italian pediatric surgeon Franco Soave [76–78] (Genoa, Italy) published a new surgical method for treating Hirschsprung's disease (Fig. 2). Soave did not make the free to avoid pelvic vessel or nerve injuries. His technique is referred to as transrectal anal pull-through surgery. This method was probably already used in a similar form in 1952 by Yancey [34]. Whether Soave knew about this technique is not known. Soave labeled his work as "Hirschsprung's Disease: A New Surgical Technique", suggesting that he borrowed his technique intellectually from Rehbein (Bremen, Germany) and Romualdi (Rome, Italy) with data obtained during the surgical correction of anorectal atresia [31,36,37,79]. Both Rehbein and Romualdi recommended leaving the affected section of the rectum in situ after removing the mucosa and pulling the sigmoid through this section of the rectum to the neo-anus [31,80]. This consideration was based, as Rehbein formulated, on the publications of the general surgeons William W. Babcock [81] (Philadelphia, PA, USA) in 1947 and Harry E. Bacon [82] (Philadelphia, PA, USA) in 1949. These surgeons used a similar approach in the treatment of deeply located rectal carcinoma to preserve the functionality of the anal sphincters. In this context, however, Soave's statements in his guest lecture "Endorectal pull-through 20 years' experience" at the 15th annual meeting of the American Pediatric Surgical Association (9–12 May 1984) in Marco Beach (Florida, USA) are thought-provoking. On that occasion, Soave stated that in 1957, he had considered the partial rectal passage allegedly presented by Romualdi as early as 1955. He did not mention Rehbein but emphasized only Romualdi as his "mentor" for his technique, which he attributed to Mark M. Ravitch (Baltimore, MD, USA) [31]. In addition, Soave stated that he regarded the method he had developed as an innovation before meeting Yancey personally in 1971 during a Sabbatical period in the USA. Jasonni *et al.* [83], in their "Soave's Chapter" of the book "Hirschsprung's Disease and Allied Disorders" edited by Holschneider & Puri, emphasized the "Soave's procedure", but did not highlight the potential Yancey's controversy. The Soave's procedure includes the following steps: After a paramedian laparotomy, the hypo/aganglionic and the orally adjacent ganglionic colon are mobilized to determine the oral resection border. Then, the rectal wall is incised circularly about two cm above the peritoneal fold up to the mucosa. The mucosa is then detached from the rectal muscles up to about 1–1.5 cm above the dentate line (depending on the child's age). Soave avoided pelvic nerve or vascular injuries by keeping the serosa or adventitia around the rectal muscle cuff in place. After dilating the anal canal, the released rectal mucosa and the hypo/aganglionic section of the colon are pulled through endorectally and transanally until the beginning of the normoganglionic colon, which needs to be verified by a pathologist during an intraoperative frozen section. The aganglionic bowel section is removed, but no colo-rectal anastomosis is made. In 1964, Soave [36] published a paper in which he detailed his thoughts on the endorectal

passage of the ganglionic colon beyond the anus without primary anastomosis [84]. In the same volume, following Soave's paper, a paper by the American pediatric surgeon Scott Boley [37] (New York, NY, USA) was published, in which he described an identical three-stage procedure similar to the Soave method but with the immediate creation of a colon-anal anastomosis. In the literature, Boley's method is considered a modification of Soave's technique. However, Boley insisted that he developed his method without knowledge of the Soave method. He considered only the techniques borrowed from Ravitch (1948) [85] and Rehbein (1963) [66,86]. Various follow-up studies have shown that the Soave method has inherent complications, e.g., anastomosis issues, strictures of the through colon, frequent episodes of diarrhea, incontinence periods, and varying severity of enterocolitis. Apart from a few mild changes, e.g., affecting the anastomosis or the "mucosectomy" technique, the muscular rectum was the main target of such modifications. It often tends to shrink, and prolonged stretching treatment is necessary. A few "improvements" were requested, e.g., splitting the muscular rectum up to the puborectal loop or extensive cuff resection, sometimes using a posterior approach. Later, Soave's procedure was also carried out with laparoscopic assistance during the 1990s to reduce surgical trauma and the recovery phase after the operation and minimize the complications. On the other hand, Keith Georgeson *et al*. [47] (Birmingham, AL, USA) were the first to report a laparoscopic procedure using the one-stage technique. They properly defined the beginning of the normoganglionic colonic section using extramucosal biopsies for intraoperative frozen section examination, with skilled pathologists able to differentiate immature ganglion cells from non-ganglionic cells. Then, they mobilized this section and the hypo/aganglionic colon up to the upper third of the rectum. The rest of the procedure was performed using transanal access, i.e., the mucosa was incised about 5–10 mm above the dentate line in a circular manner, and the upper third of the rectum was encapsulated. The entire mobilized colon, including the rectal mucosa, was then pulled through endorectally and transanally, and, finally, the hypo/aganglionic section was resected. The ganglionic colon was anastomosed extra-anally with the anal mucosa [47]. In 1999, Georgeson *et al*. [48] called this approach the gold standard for Hirschsprung's disease. Subsequently, Georgeson indicated that the transanal endorectal passage, without laparotomy or laparoscopic assistance, may be based on the works of Luis de La Torre-Mondragón and José Arturo Ortega-Salgado [41] (Mexico City, Mexico [1998]) and Jacob Langer *et al*. [46] (Toronto, ON, Canada [1999]). Interestingly, the working group that initially monitored their procedure using a laparoscope stated that such an approach may be the method of choice for short-distance Hirschsprung's variants [87]. Georgeson's procedure did not lose its importance in children with long-distance aganglionosis, even if it has been modified in the meantime. The laparoscopic single-incision and/or robot-assisted variants of the Georgeson technique presented by various authors are technically interesting and will be discussed further (please see below).

De la Torre and Langer Technique

The working groups around Luis De la Torre-Mondragón and Jacob Langer [41,46] described the technique known as TERPT in 1998 and 1999. This technique is based per se on the Soave's and Boley's procedures, but in reverse form, i.e., the procedure is performed from the anus without laparotomy. Briefly, the anal canal is first carefully dilated and then stretched with an anal retractor to open the anal canal and expose the dentate line. Epinephrine, NaCl, or air are applied to the submucosa about 3–5 mm proximal to the dentate line to simplify the planned "mucosectomy". Mucosectomy is a surgical procedure involving the removal of a circular section of mucosa from the rectum, leaving a 1 cm margin above the dentate line. The rectal mucosa is incised circularly in the area just mentioned with electrocautery and carefully cleared to attach several retaining sutures here so that further endorectal submucosal dissection can be carried out under tension. The muscular sheath of the rectum is incised anteriorly at a height of about 6–7 cm, thereby opening the rectouterine pouch (rectovaginal pouch, pouch of Douglas, or cul-de-sac), which is the extension of the peritoneum into the space between the posterior wall of the uterus and the rectum in the human female. In the human male, the region corresponding to the rectouterine pouch is the rectovesical pouch, which lies between the urinary bladder and rectum. The rectal muscles are divided circularly. The rectosigmoid is mobilized by cutting all the blood vessels that supply these two sections close to the intestinal wall. After reaching the peritoneal fold, the rectosigmoid is pulled through the anus and, depending on the determination of the resection margin utilizing serial seromuscular biopsies or full-thickness biopsies, is removed. The normoganglionic colon is anastomosed to the remaining mucosa superior to the dentate line (Fig. 3). Langer *et al*. [46] emphasized that they initially monitored the abdominal part of the operation using a laparoscope. TERPT has become established in most pediatric surgical departments to treat short, short-segment aganglionosis patients. Our personal experience has shown that this surgical method is not limited to the rectosigmoid but can be used to resect the aganglionic colon up to the left flexure without laparoscopic support and even further to the right hemicolon with laparoscopic support or a transanal mesenteric resection in the sense of a NOTES procedure (Natural Orifice Transluminal Endoscopic Surgery) [88–92]. However, as the follow-up showed, the TERPT implies the same obstruction problems (soiling, defecation disorders, enterocolitis, etc.) as the Soave/Boley techniques due to the remaining muscular rectum [55,56,61]. Stretching the sphincter apparatus during the procedure should not cause any challenges [91–93]. The rectum cuff was first split, but it was demonstrated to be

insufficient [45,94]. Its length was consequently reduced to minimize the obstruction, resulting in a site similar to that of Swenson's procedure [43,45,95–97].

Retrorectal Transanal Pull-through Surgery (Duhamel)

In 1956, Bernard Duhamel [28] (Paris, France) described a technique with a completely different approach to Hirschsprung's disease. He used a retrorectal passage of the colon. He considered keeping the aganglionic rectum as a partial reservoir for receiving or voluntarily emptying feces, which had not been considered in the other techniques. Initially, the hypoganglionic and aganglionic sections of the colon are mobilized and resected after defining the oral resection margin while leaving a rectal stump. The ganglionic colon and rectal stump are closed blindly. The peritoneal fold between the urinary bladder or uterus and the rectum is cut, and the distal rectum is dissected, accessible from the dorsal side up to approximately 1.5 cm above the dentate line. The posterior wall of the surgical anal canal is incised about 1–1.5 cm proximal to the dentate line. After the resection, the ganglionic colon is placed in the avascular retrorectal plane and sutured side-to-side to the native aganglionic rectum. A clamp is inserted through this incision into the retro-rectal space and advanced into the abdominal cavity. With the help of this clamp, the sutures attached to the distal end of the ganglia-containing colon are pulled through the rectal stump. A side-to-side anastomosis occurs between the rectal stump's avascular area and the normoganglionic colon. The anterior wall of the ganglionic intestine is sutured to the proximal posterior wall of the opened rectum and, subsequently, the posterior wall of the ganglionic segment to the distal posterior rectal wall. Duhamel's procedure was instrumental because of the retrorectal/anal anastomosis that was initially too deep, but also because of the different-sized rectal stump that remained *in situ*.

The criterion is to keep the rectal stump small, especially since it remains aperistaltic as an aganglionic portion and represents a distensible blind sac. It was prone to complications, e.g., anastomosis issues, incontinence, proctitis, fecaloma, and enterocolitis. Therefore, various modifications (e.g., by using stapler devices for the colo-rectal anastomosis) took place regarding the penetration to protect the M. sphincter ani internus and the rectal pouch as well as the sphincter achalasia [38,98–104]. Apart from these modifications to address issues inherent to the technique, in 1984, Hickey and Guiney [101] suggested creating a protective colostomy to minimize surgically related local infections in the small pelvis and wound healing disorders. This proposal was revisited by Peters *et al*. [102] some 40 years later. Duhamel's procedure, despite various other modifications, including the interim laparoscopic procedure, was still loaded with some minor complications [99,102–109]. Thanks to its problem-free laparoscopic applicability for failed Swenson's surgery, long-segment Hirschsprung's disease, total colonic aganglionosis, and in cases of difficult mucosectomy, the Duhamel technique is currently considered to be one of the best procedures for most of the infants [102,106,110–114].

Therapeutic Options in Special Settings

The surgical treatment of two forms of Hirschsprung's disease is briefly presented below, i.e., the complete aganglionosis of the colon, including the lower ileum (Zuelzer-Wilson syndrome or better Jirásek-Zuelzer-Wilson syndrome) and the so-called ultra-short segment of Hirschsprung's disease.

Surgery on Total Colonic Aganglionosis

Ultra-short segment and long segment Hirschsprung's disease were the clinical subtypes of Hirschsprung's disease, at least prior to extensive research. Total colonic aganglionosis (TCA) and total colonic aganglionosis with small bowel aganglionosis (TCSA), the latter of which may entail a very long segment of Hirschsprung's disease, are likely subtypes of the long segment one. TCA, usually extending into the ileum, is a rare affection (2–13%, probably approximately 8% of patients affected with Hirschsprung's disease). It usually becomes symptomatic in newborns or infancy but can rarely become relevant until childhood and, in individual cases, even later. Aganglionosis that extends from the anus to the ileocecal valve, but no more than 50 cm proximal to it, has been characterized as TCA due to the unique symptoms and complications linked with extremely long aganglionic segments in TCSA. Therefore, it is distinct from the relatively uncommon duodenal-to-anus form of aganglionosis and the extended intestinal form, also known as TCSA. From a pathophysiological and biological perspective, it is still unclear whether these two entities should be considered independently [115]. Clinically, the event presents an incomplete small bowel obstruction (subileus) with or without diarrhea and without signs of a megacolon. The radiological finding may just be as untypical as the clinical picture. Despite these challenges, unique patterns of radiological characteristics have been discovered that could suggest the presence of TCA. Microcolons (apart from the microcolon-hypoperistalsis syndrome, which has a different histopathology background detected by the smooth muscle actin immunostaining [116] and the absence of characteristics in a normal colon) were classified as three separate radiological images in TCA on imaging [117]. Preliminary research indicated that barium retention for at least 24 hours was highly indicative of Hirschsprung's disease. However, water-soluble contrast has mainly replaced barium in current practice, rendering it less useful as an indication [118]. The data we evaluated from the literature highlights that if this syndrome is suspected, the removal of rectal (suction) biopsies is indicated to detect aganglionosis. Early diagnosis and appro-

priate management of intestinal obstruction are considered essential prognostic criteria [1,119–126].

The surgical treatment of this disease is typically multistage. An ileostomy or an ostomy carried out above 50 cm proximal to the ileocecal valve is usually planned initially. It is crucial to have a pathologist with experience in diagnosing immature and mature ganglion cells [1]. After the child has recovered, one of the numerous corrective operations is carried out weeks later. So far, there is no consensus as to which of the various methods is the best for this specific extended segment of aganglionosis. A consensus may be helpful for both surgical and prognostic settings (e.g., perioperative morbidity, mortality, functional outcome, and enterocolitis or HAEC-Hirschsprung's disease-associated enterocolitis) [127]. Initial considerations were devised to preserve part of the non-innervated colon. The antimesenteric enteroplasty and the Kimura patch should be mentioned, even if these techniques gained less importance later because of their complications. Our review of the literature and personal experience indicates that all surgical operations known for treating Hirschsprung's disease have been used.

The outcome may be difficult to predict after surgical treatment of patients with TCA due to the various surgical interventions used and the different follow-up times described in the literature. Enterocolitis episodes are not uncommon (up to 40% of cases), while daily frequent bowel movements (up to approximately 80% of patients, depending on the study), and soiling, as well as perianal excoriations (approximately 20%), are typical issues. However, they may decrease over the years. Although constipation is also possible, it seems comparatively rare after surgery. Developmental deficits (low body mass index, shortness) are also observed in up to 30% of children with TCA. It may be critical to separate TCA from megacystis-microcolon-hypoperistalsis syndrome, as indicated above.

Surgery on So-called Ultra-short Segment of Hirschsprung's Disease

The transition zone at the rectosigmoid junction is a common symptom of Hirschsprung's disease, which often manifests in the neonatal period or early infancy. But in other cases, ultrashort-segment Hirschsprung's disease (ultrashort-Hirschsprung's disease) manifests as a highly sporadic form [11,14,128–132]. The aganglionic segment is extremely short, so it can often go undetected by contrast enema. Not only does this put off diagnosis, but it also makes it harder to determine what kind of surgery would be most effective [133,134]. It is debatable if ultrashort-Hirschsprung's disease is a suitable word when describing a condition with symptoms comparable to typical Hirschsprung's disease [9]. The current standard for diagnosing ultrashort-Hirschsprung's disease is a rectal segment that is less than one to two centimeters long and has no ganglionic structures. Although there are easier options, such as an internal sphincter myectomy or a Duhamel-type pull-through, these procedures still leave the megarectum in place, which is unacceptable. The Swenson pull-through would be a good method for treating the ultrashort segment, but is not usually used because of its potential lesions of the structures in the small pelvis and the associated urological and sexual dysfunctions. While there is no evidence to support the claims that Swenson dissection increases the risk of urinary or sexual dysfunction, a recent study by Hong *et al*. [135] used a Soave transanal pull-through procedure with wide posterior myectomy and had positive outcomes in an 8-year-old boy. While the Swenson transanal pull-through may be better regarding cuff-related complications, even for seasoned surgeons, the procedure's technical requirements can be challenging [136]. In our experience and reviewing the literature data critically, we found that the different recommendations for treating ultrashort-Hirschsprung's disease are linked to the controversial nature of this condition. We suggest that the (partial) posterior anal sphincter myectomy is a simple method. Depending on the age of the patient and considering that the inner anal sphincter is the method. Depending on the age of the patient and considering that the inner anal sphincter is about 4 cm long in adults and correspondingly shorter in children, depending on their age, a horizontal arcuate incision about 1–2 cm long is performed exactly in the dorsal mucocutaneous junction of the anal canal for about 0.5–1 cm above the ano-cutaneous line. From here, the dorsal section of the M. sphincter ani internus is dissected orally at approximately 3–5 cm (depending on the patient's age), and a muscle strip of 0.5–1 cm is resected. The typical complications of this method are recurrence of constipation or the development of fecal incontinence. Apart from the posterior anal sphincter myectomy, one of the various pull-through operations can also be considered as a method, as reviewed by Hong *et al*. [135] and Glasgow *et al*. [136].

Short Bowel Syndrome

If Hirschsprung's disease requires an extensive small/large bowel resection, it causes SBS (residual bowel length <35 cm in a neonate). A surgical treatment of this issue may be necessary [119,137]. An autologous intestinal reconstructive surgery (AIRS) is currently planned based on the experience that the remaining intestinal remnant usually develops lumen distension, which in turn is a prerequisite for AIRS. Two techniques are currently being used: longitudinal intestinal lengthening and tailoring (LILT) and serial transverse enteroplasty (STEP). If these methods are not successful, the last resort is an intestinal transplant. The LILT method described by Adrian Bianchi [138] in 1980 is the first AIRS technique [139]. Its principle is that the mesentery consists of two layers of tissue. Two tubes can be formed by splitting the mesentery and the attached dilated intestine longitudinally, which are then anastomosed [138–140]. Although this method is efficient, it does imply serious potential for complications: devascularization

of the intestine after splitting the mesentery, leakage along the longitudinal rows of sutures and the anastomoses in the small intestine, and recurrent intestinal dilatation or dysmotility of the extended intestinal segment. The STEP method, first published in 2003, is technically much more straightforward than Bianchi's operation [141]. This setting is because the mesentery remains intact, and an anastomosis is unnecessary. Lengthening of the dilated bowel was performed by serial transverse applications of a stapler from opposite directions to create a zig-zag channel [141]. With this method, very short, dilated small intestinal segments can be lengthened, and the intestinal lengthening can be repeated after a while. Today, the STEP method is primarily used for SBS treatment because of its simplicity [142–144]. With this method, however, the orientation of the muscle fibers is changed, i.e., the circular muscle fibers become longitudinal muscle fibers and vice versa, which leads to motility disorders of the intestine [145,146]. To address the problems mentioned, some modifications have been added, e.g., the spiral intestinal lengthening and tailoring (SILT) technique, which was tested in experimental animals in 2013 and used clinically in 2014 [145,147] or the one presented in 2022 Saeki Spiral Shark (3S) method, also a spiral technique [143]. The extent to which these techniques will achieve clinical significance remains to be explored [140].

Robotic-assisted Surgery

The use of surgical robots has increased recently. The most recent development in minimally invasive methods for treating Hirschsprung's disease is probably robotic-assisted surgery [89,146,148–165]. The advantages of laparoscopic surgery are combined with improved accuracy and this state-of-the-art method of visualization, which could lead to better surgical results. The Da Vinci Surgical System is the most common robotic platform for Hirschsprung's disease robotic-assisted surgeries [151,157]. The major components are a high-definition 3D vision system, a patient-side cart with robotic arms for surgical instruments, a console where doctors operate via foot pedals and hand controls, and wristed tools with an improved articulation angle for enhanced dexterity. Patient positioning, robotic arm port placement, system docking, transition zone identification (with intraoperative biopsies), colon mobilization, endorectal muscular cuff creation transanally, and critical transanal pull-through and anastomosis are the procedural steps for robotic-assisted pull-through in high definition. For the abdominal section, several institutions have used robotic assistance, and for the pull-through, they switched to a transanal method [89,146,148–164,166–168].

There are several advantages to robotic-assisted surgery over traditional laparoscopic methods, the most important of which are better visualization and increased precision. During dissection and suturing, the robotic system scales the surgeon's movements and successfully filters hand tremors, leading to exceptionally accurate actions. Second, compared to conventional laparoscopy, the 3D high-definition vision system provides better depth awareness, allowing surgeons to more precisely navigate anatomical regions. Particularly helpful for delicate treatments in small areas like the pelvis, the robotic system's wristed tools offer excellent dexterity and a wider range of motion. Moreover, the seated console's ergonomic design lessens surgeon fatigue, which is advantageous during lengthy procedures. In the future, robotic technology may make telesurgery possible, enabling surgeons to work remotely. However, this capability is still in its early stages and is not frequently used. Finally, these systems have a remarkable educational component. They will often have tools that make it easier to train others, so more seasoned surgeons can keep an eye on their students. Although Hirschsprung's disease robotic-assisted surgery is still in its infancy, encouraging new data is mounting. Compared to traditional laparoscopy, the success rate of robotic-assisted pull-through procedures and the number of patients requiring open surgery afterward are lower than those of earlier studies. In addition, studies have shown that robotic surgery may have a lower learning curve than traditional laparoscopic procedures. It is because robotic systems provide better visualization and intuitive controls. While long-term data are still in the works, preliminary findings show that functional outcomes, such as continence and bowel function, after robotic surgery are comparable to other minimally invasive methods. Although larger-scale studies are needed to confirm these discoveries, preliminary results indicate that the complication rates of robotic-assisted surgery are equivalent to or lower than those of traditional laparoscopy, which is good safety news [146,148–156].

Similar to other minimally invasive treatments, there have been reports of specific problems such as anastomotic leaks, enterocolitis, and constipation. Rates seem similar among methods, but more data is needed to compare accurately. The more significant ports needed to accommodate robotic devices have prompted worries about an uptick in port-site problems, especially hernias. Continued research and clinical experience are crucial for understanding robotic-assisted surgery's relative safety and efficacy for Hirschsprung's disease, especially while the field is still in its early stages. As with any new medical technology, there are several factors to consider when deciding whether or not to use robotic surgery. The significant upfront investment and continuing maintenance costs of robotic systems are major obstacles. Due to the associated costs, many medical facilities may be reluctant to incorporate this technology into their daily operations. Second, surgeons utilizing robotic techniques may have longer operating times, particularly during the early learning curve. Efficiency, patient outcomes, and the usage of operating rooms are all impacted by this aspect. The lack of tactile feedback in existing robotic systems is another primary concern affecting surgical accuracy and the ability to

identify tissue features. Surgeons frequently depend solely on visual signals, which might not completely replace the tactile input offered by conventional surgery [89,146,148–163,167,168]. If we want to know how long robotic-assisted treatments last and how effective they are, we must keep collecting data like this. More research, better technologies, and cost-effectiveness evaluations are required to overcome these obstacles and improve the integration and results of robotic surgery in clinical practice. Robotic-assisted surgery is anticipated to become more significant in treating Hirschsprung's disease as technology improves and more surgeons acquire proficiency with robotic systems. This method may become even more helpful in treating Hirschsprung's disease as technological advances, such as smaller devices and platforms for children, occur worldwide.

Quality of Life

A large number of individuals with Hirschsprung's disease suffer from surgical morbidities that extend beyond bowel dysfunction [169]. Doctors and patients are now more concerned with the long-term effects of Hirschsprung's disease, which can change the bowel, lower urinary tract, sexual function, fertility, psychological and social health, and, ultimately, quality of life in both childhood and adulthood [169,170]. The duration of aganglionosis and any accompanying symptoms determine the severity of these long-term complications, which sometimes seem causally related and cluster in specific individuals. The hereditary abnormality that causes Hirschsprung's disease can also put patients at risk for other diseases down the road, like "medullary thyroid carcinoma". On the other hand, the pathophysiology of the increasingly common inflammatory bowel disease associated with Hirschsprung's disease is still not well understood. Issues with bowel regularity and fecal control are most common in the years immediately after surgery, but they do continue into adolescence and adulthood, albeit to a reduced extent. Even in patients whose medical therapy and self-coping mechanisms are inadequate, the long-term prognosis appears to be positive since competent bowel control can lead to social continence and a decent quality of life. Interdisciplinary specialized approaches considering psychosocial factors are the most effective in managing bowel dysfunction and its possible long-term problems [169,171–173]. This approach also aids in accurately identifying areas where adult healthcare must continue to provide input for the benefit of young adult patients.

Problems in recognizing the need to defecate, fecal soiling, and a decrease in the frequency of fecal accidents are common long-term symptoms of decreased bowel control following pull-through surgery for Hirschsprung's disease. The number of patients reporting fecal accidents is fewer than 25%. However, 10% of those patients had more severe cases, meaning they soiled themselves at least once a week or needed protective gear [169,172–175]. Most adult patients had undergone Duhamel or Rehbein reconstruction; however, this did not translate to a significantly higher prevalence of constipation compared to normal controls. Some neurological causes include (1) a transition zone pull-through when the abnormally innervated transition zone is not accidentally removed, (2) colon dysmotility caused by abnormal enteric innervation of the normal ganglionated bowel, and (3) persistently abnormal innervation of the internal anal sphincter with no rectoanal inhibitory reflex and inadequate relaxation. The patient's quality of life may be improved with a permanent diverting ostomy if they have intractable fecal incontinence or bowel dysfunction that is unresponsive to other treatments. Sexual dysfunction, reduced fertility, and abnormal genitourinary function may occur in youth, and these issues need to be considered by family doctors or general physicians. An iatrogenic risk is higher in patients with low rectal dissection. After endorectal pull-through, most teenage patients with Hirschsprung's disease seem to have minimal damage to their pelvis, lower urinary tract, and erectile function. According to the literature, 18% of the men who received the Duhamel's procedure showed some erectile dysfunction, and 4.3% had problems with ejaculation. There is a correlation between erectile dysfunction and intestinal dysfunction. It is recommended to evaluate erectile function and screen for bladder dysfunction using urine flowmetry before moving on to other therapies [175,176]. When compared to the general population, teenage girls may report a decline in sexual quality of life and infertility, which may be associated with postoperative pelvic adhesions. We strongly suggest that young adult females with Hirschsprung's disease should routinely be offered a gynecologist consultation with information on probable fertility concerns.

Future Directions

Rapid evolution characterizes the field of minimally invasive surgery for Hirschsprung's disease, which shows promise for future improvement in several areas. These advancements are made to enhance surgical results, decrease complications, and elevate patient living standards. There is a lot of hope for Hirschsprung's disease surgery in combining cutting-edge imaging methods with new surgical navigation systems. Indeed, more accurate transition zone and critical structure identification may be possible with intraoperative near-infrared fluorescence imaging. Surgeons could be given 3D/4D, real-time patient anatomy representations overlaid on the operating field using this technology in conjunction with augmented reality systems. These developments can lessen the likelihood of partial resection and enhance the preservation of vital structures, such as pelvic nerves. An intriguing new area of research is using artificial intelligence (AI) and machine learning (ML) in surgical planning and execution. These innovations can potentially improve preoperative risk assessment, leading to more informed decisions about which minimally invasive

Table 1. Procedures for Hirschsprung's disease with their improvements, advantages, and current applications.

Procedure (Name)	Year	Original steps	Improvements	Pros	Current application
Pull-through procedure ("Swenson's procedure")	1048–1949	Open transabdominal approach: Resection of the hypo- and aganglionic segment at the level of the rectosigmoid junction and blind closure of the ganglionic segment or the rectum close to the wall. Mobilization of the rectum up to the rectoanal junction, perianal pulling through of the rectal stump, exposure of the dentate line, resection of the rectal stump, pulling through the ganglionic colon, and extra-anal creation of a coloanal anastomosis.	Preparatory problems in protecting the nervous, vascular, and muscular structures of the pelvic floor (consequence: incontinence, bladder emptying disorders, impotence). Torsion of the continuous colon. Anastomotic dehiscence with abscess formation in the abdomen and small pelvis—see [63,185].	No stoma necessary. No technology-related muscle cuff.	Laparoscopic procedure [53,56].
Anterior (deep) rectal resection (Rehbein)	1953	Transabdominal with resection of hypo and aganglionic segments, including either end-to-end anastomosis across the normoganglionic colon and rectal stump.	(1) Symptoms of rectal incontinence. (2) Colostomy. (3) Stenosis of the Anastomosis. (4) Sphincter Achalasia [73].	Infraperitoneal resection (1) Issue of the pelvic floor. (2) Intact sphincter. (3) Extraperitoneal location of anastomosis [75,182,183].	Surgery after Rehbein is rarely used today [75].
Retrorectal transanal pull-through (Duhamel)	1956	Transabdominal and transanal access with Resection of the hypo- and aganglionic segment, blind closure of the colon containing the ganglion bone and the rectum, clearing the posterior wall of the rectum (up to approximately 1.5 cm above the dentate line), the ganglionic colon is placed in the avascular retrorectal plane and sutured side-to-side to the native aganglionic rectum (preservation of the native rectum and longitudinal anastomosis between the ganglionic colon and rectum).	Stoma (one-stage operation carried out very late*) and numerous modifications due to: (1) Problems with the depth of the anastomosis in the area of the posterior wall of the anal canal, (2) Anastomotic dehiscence with abscess formation in the small pelvis, (3) Aganglionic rectum grows with it - result: fecal cessation or pouchitis (Modified Duhamel Procedure) for Hirschsprung's Disease.	Aganglionic rectum preserved for feces. No vascular lesions in the pelvis. Use of staplers to create anastomosis [179,180].	Duhamel laparoscopy in One-Stage-Technique [181].

Table 1. Continued.

Procedure (Name)	Year	Original steps	Improvements	Pros	Current application
Endorectal pull-through (Soave)	1963	Transabdominal approach: clearing the aganglionic colon segment, Incise the rectal serosa/muscles approximately 2 cm above the peritoneal fold down to the mucosa, and bluntly push away the seromuscular layer to 1–1.5 cm above the dentate line pulling the aganglionic and ganglionic segments through the muscular tube and resection of the aganglionic segment Soave did not perform an anastomosis, but instead waited for the serosa of the continuous ganglionic colon to adhere to the muscular tube and the remaining mucosa of the distal rectum.	(1) Tendency to stenosis of the muscle cuff with signs of obstruction or constipation. (2) Anastomosis problems (dehiscence, stenosis) [184].	There are no problems with the structures in the pelvic floor area.	Laparoscopic procedure since 1995 (still used today for long-range aganglionoses) [47,87].
Transanal endorectal pull-through (De la Torre)	1998	Transanal approach: circumcision of the rectal mucosa approximately 3–5 mm proximal to the dentate line, endorectal dissection of the submucosa, circular division of the rectal muscles (height 3–5 cm from dentate line) and mobilization of the rectosigmoid to the peritoneal fold, pulling the rectosigmoid through the anus. Determine the extent of resection, dorsal incision of the rectal muscle cuff, resection of the aganglionic portion, and creation of an End-to-End Coloanal Anastomosis.	The length of the muscle cuff is chosen differently. The longer, the higher the risk of stenosis of the muscle tube and torsion of the continuous colon [97].	No laparotomy, no preliminary colostomy with their associated complication, reduced operating time, less blood loss, no pelvic structure damage, single hospital admission, short hospital stay, and faster nutrition are possible.	Laparoscopically for long-range aganglionosis [47,48,87,177,178].

Notes: Please see the text for the single techniques.

procedures to recommend to patients. AI-driven systems can analyze tissue properties in real-time during surgery, which could lead to more precise transition zone detection. More individualized patient care could be possible using ML algorithms to forecast and avoid postoperative problems like enterocolitis. Although robotic-assisted surgery for Hirschsprung's disease is in its infancy, future robotic technology advancements will address existing restrictions in this decade. Further reduction of surgical trauma and improvement of cosmetic outcomes could be achieved through refinement of single-port laparoscopic procedures. One possible approach to Hirschsprung's disease surgery, especially for short-segment disease, is the NOTES method, which involves doing the entire procedure through natural orifices instead of external incisions [90]. Although these methods seem technically difficult, they may significantly reduce postoperative pain and speed up recovery. Tissue engineering has the potential to completely alter how Hirschsprung's disease is treated.

Possible new therapeutic options may emerge from studies aiming to create modified neural crest cells, also known as enteric neurons. There is a collection of databases, including microRNA transcriptomics, which may favor some neural commitments of stem cells [7]. The repopulation of aganglionic segments could be achieved using these synthetic tissues alone or combined with minimally invasive surgical procedures. Optimized pain management, early eating, and movement are all possible components of these strategies that can shorten hospital stays and improve the recovery of our patients. Thanks to developments in robotics and telecoms, potentially usable telesurgery for Hirschsprung's disease management is on the horizon. This can improve access to specialist treatment in poor, developed regions by allowing skilled surgeons to execute or assist in surgeries remotely. Finally, we strongly hope that improved surgical training and center-wide technique standardization are two potential benefits of remote proctoring systems.

Final Remarks

The curative treatment of Hirschsprung's disease still consists of complete resection of the hypoganglionic part of the megacolon and the aganglionic colon/rectum. This can be done as a one-stage, two or three-stage procedure. After the advent of surgical treatment of Hirschsprung's disease, it took years for a two- or three-stage surgical procedure to be practiced. Table 1 (Ref. [47,48,53,56,63,73,75,87,97,177–185]) summarizes this progress, which has been listed in detail in the previous paragraphs. During the 1980s, a one-stage approach was increasingly targeted for clinically uncomplicated cases to avoid stoma problems. In addition, thanks to the earlier diagnosis, fewer symptoms occurred. The time for the operation was increasingly shifted to the newborn and infant period, and a single-stage procedure was propagated. However, this view has different opinions: Subsequently, no advantage in a one-stage versus a multi-stage approach in infants was seen, despite multi-institutional studies disclosing that multiple interventions result in significantly more re-hospitalizations and re-operations. From the various initial surgical methods, i.e., those to be carried out by laparotomy, four have emerged that are referred to as "classic" Hirschsprung's disease procedures:

- Swenson's abdominal-anal swipe (1948),
- Retrorectal transanal passage of Duhamel (1956),
- Rehbein's anterior deep recto-sigmoidectomy (1958), and
- Endorectal transanal passage of Soave (1963).

Following the general surgical trend, minimally invasive techniques were introduced during 794 the 1990s for treating Hirschsprung's disease. Hirschsprung's disease minimally invasive surgery has come a long way, greatly enhancing 796 patient outcomes and quality of life. Surgeons have shown that laparoscopic, transanal, and 797 robotic-assisted procedures can lessen surgical trauma, speed recovery, and keep functional outcomes over the long run. Still, there are obstacles to overcome, most notably regarding method standardization, long-segment disease, and long-term result optimization. Combining state-of-the-art imaging with AI and robotic devices is how Hirschsprung's disease surgery will be done in the future. These advancements can pave the way for new treatment modalities, better decision-making, and more precise surgeries. New methods for patient treatment may emerge from the growing trend of genetic and molecular profiling-based personalization. There is an urgent need for large-scale, long-term research to assess functional results and quality of life as the area advances. Working on high-technology-supported telesurgery and standardized training programs will also be critical to increasing access to specialized treatment. Future surgical treatments for Hirschsprung's disease may provide even better results for patients if researchers keep developing less intrusive methods.

Availability of Data and Materials

All review data have been downloaded from the platforms for biomedical references as indicated in Fig. 1 and original photographs of the specific procedures are occasionally obtained by contacting the first author if needed.

Author Contributions

JH selected the studies from the literature and wrote the first draft of the manuscript. CMS validated the data, compiled statistics, and edited the draft. Both authors contributed to important editorial changes in the manuscript. Both authors read and approved the final manuscript. Both authors have contributed sufficiently to the work and agreed to be accountable for all aspects of the work.

Ethics Approval and Consent to Participate

Not applicable.

Acknowledgment

This work is dedicated to Professor Vincenzo Jasonni, who directed the division of Pediatric Surgery of the Genoese Gaslini Children's Hospital, Italy, for over 20 years. He is remembered as "*A person always open to innovations and new technologies, a courageous, decisive man, and a natural leader*" (GenovaToday, 27 June 2022). Professor Jasonni was the director of the division, just before Professor Francesco Soave, who is considered an innovator in the surgical techniques for treating Hirschsprung's disease.

Funding

This research received no external funding.

Conflict of Interest

The authors declare no conflict of interest.

Supplementary Material

Supplementary material associated with this article can be found, in the online version, at https://doi.org/10.62713/aic.3943.

References

[1] Takawira C, D'Agostini S, Shenouda S, Persad R, Sergi C. Laboratory procedures update on Hirschsprung disease. Journal of Pediatric Gastroenterology and Nutrition. 2015; 60: 598–605. https://doi.org/10.1097/MPG.0000000000000679.

[2] Hirschsprung H. Constipation in the newborn as a result of dilation and hypertrophy of the colon. Jahrbuch für Kinderheilkunde und physikalische. 1887; 27: 1–7. (In German)

[3] BODIAN M, CARTER CO, WARD BCH. Hirschsprung's disease. Lancet (London, England). 1951; 1: 302–309. https://doi.org/10.1016/s0140-6736(51)92290-8.

[4] Sergi C. Hirschsprung's disease: Historical notes and pathological diagnosis on the occasion of the 100(th) anniversary of Dr. Harald Hirschsprung's death. World Journal of Clinical Pediatrics. 2015; 4: 120–125. https://doi.org/10.5409/wjcp.v4.i4.120.

[5] EHRENPREIS T. Megacolon in the newborn; a clinical and röntgenological study with special regard to the pathogenesis; a preliminary report. Acta Paediatrica. 1945; 32: 358–370.

[6] Sergi CM, Hager J. Editorial: Hirschsprung disease: genetic susceptibility, disease mechanisms and innovative management in the multi-omics era. Frontiers in Pediatrics. 2023; 11: 1274735. https://doi.org/10.3389/fped.2023.1274735.

[7] Sergi CM, Caluseriu O, McColl H, Eisenstat DD. Hirschsprung's disease: clinical dysmorphology, genes, micro-RNAs, and future perspectives. Pediatric Research. 2017; 81: 177–191. https://doi.org/10.1038/pr.2016.202.

[8] Jensen AR, Frischer JS. Surgical history of Hirschsprung disease. Seminars in Pediatric Surgery. 2022; 31: 151174. https://doi.org/10.1016/j.sempedsurg.2022.151174.

[9] Neilson IR, Yazbeck S. Ultrashort Hirschsprung's disease: myth or reality. Journal of Pediatric Surgery. 1990; 25: 1135–1138. https://doi.org/10.1016/0022-3468(90)90748-x.

[10] Duci M, Santoro L, Dei Tos AP, Loss G, Mescoli C, Gamba P, *et al*. Postoperative Hirschsprung's associated enterocolitis (HAEC): transition zone as putative histopathological predictive factor. Journal of Clinical Pathology. 2025; 78: 111–116. https://doi.org/10.1136/jcp-2023-209129.

[11] Slaczka F, Uruthirakumar R, Slaczka M, Bozeman A. Refractory Chronic Constipation in an Adolescent Female Later Diagnosed With Internal Anal Sphincter Achalasia. Cureus. 2024; 16: e57135. https://doi.org/10.7759/cureus.57135.

[12] Ouladsaiad M. How to manage a late diagnosed Hirschsprung's disease. African Journal of Paediatric Surgery: AJPS. 2016; 13: 82–87. https://doi.org/10.4103/0189-6725.182562.

[13] Ziad F, Katchy KC, Al Ramadan S, Alexander S, Kumar S. Clinicopathological features in 102 cases of Hirschsprung disease. Annals of Saudi Medicine. 2006; 26: 200–204. https://doi.org/10.5144/0256-4947.2006.200.

[14] Bentley JF. Ano-rectal achalasia of ultra-short segments in Hirschsprung disease. Minerva Chirurgica. 1971; 26: 310–314. (In French)

[15] Kawaguchi AL, Guner YS, Sømme S, Quesenberry AC, Arthur LG, Sola JE, *et al*. Management and outcomes for long-segment Hirschsprung disease: A systematic review from the APSA Outcomes and Evidence Based Practice Committee. Journal of Pediatric Surgery. 2021; 56: 1513–1523. https://doi.org/10.1016/j.jpedsurg.2021.03.046.

[16] Page MJ, McKenzie JE, Bossuyt PM, Boutron I, Hoffmann TC, Mulrow CD, *et al*. The PRISMA 2020 statement: An updated guideline for reporting systematic reviews. Journal of Clinical Epidemiology. 2021; 134: 178–189. https://doi.org/10.1016/j.jclinepi.2021.03.001.

[17] Tittel K. Ueber eine angeborene Missbildung des Dickdarmes. Wien Klin Wochenschr. 1901; 14: 903–907. (In German)

[18] Treves F. Idiopathic dilatation of the colon. Lancet. 1898; 29: 276–279.

[19] Fennwick WS. Hypertrophy and dilatation of the colon in infancy. The British Medical Journal. 1900; 2: 564–567.

[20] Dalla Valle A. Histological investigation of a case of congenital megacolon. Pediatria. 1920; 28: 740–52. (In Italian)

[21] Robertson HE, Kernohan JW. The myenteric plexus in congenital megacolon. Mayo Clinic Proceedings. 1938; 13: 123–125.

[22] Tiffin ME, Changler LR, Faber HK. Localized absence of ganglion cells of the myenteric plexus in congenital megacolon. American Journal of Diseases of Children. 1940; 59: 1071–1082. https://doi.org/10.1001/archpedi.1940.01990160145010.

[23] ZUELZER WW, WILSON JL. Functional intestinal obstruction on a congenital neurogenic basis in infancy. American Journal of Diseases of Children (1911). 1948; 75: 40–64. https://doi.org/10.1001/archpedi.1948.02030020047005.

[24] WHITEHOUSE FR, KERNOHAN JW. Myenteric plexus in congenital megacolon; study of 11 cases. Archives of Internal Medicine (Chicago, Ill.: 1908). 1948; 82: 75–111. https://doi.org/10.1001/archinte.1948.00220250085005.

[25] Ehrenpreis T. Hirschsprung's Disease. Year Book Medical Publishers Inc.: Chicago, IL. 1970.

[26] SWENSON O, BILL AH, Jr. Resection of rectum and rectosigmoid with preservation of the sphincter for benign spastic lesions producing megacolon; an experimental study. Surgery. 1948; 24: 212–220.

[27] SWENSON O, NEUHAUSER EBD, PICKETT LK. New concepts of the etiology, diagnosis and treatment of congenital megacolon (Hirschsprung's disease). Pediatrics. 1949; 4: 201–209.

[28] DUHAMEL B. New operation for congenital megacolon: retrorectal and transanal lowering of the colon, and its possible application to the treatment of various other malformations. La Presse Medicale. 1956; 64: 2249–2250. (In French)

[29] DUHAMEL B. A new operation for the treatment of Hirschsprung's disease. Archives of Disease in Childhood. 1960; 35: 38–39. https://doi.org/10.1136/adc.35.179.38.

[30] REHBEIN F. Intraabdominal resection or rectosigmoidectomy (Swenson's technic) in Hirschsprung's disease. Chirurg. 1958; 29: 366–369. (In German)

[31] ROMUALDI P. A new technic for surgical treatment of some rectal malformations. Langenbecks Archiv für Klinische Chirurgie vereinigt mit Deutsche Zeitschrift für Chirurgie.1960; 296: 371–377. (In German)

[32] EHRENPREIS T. Long-term results of rectosigmoidectomy for

Hirschsprung's disease, with a note on Duhamel's operation. Surgery. 1961; 49: 701–706.
[33] PELLERIN D. The surgical treatment of Hirschsprung's disease by resection and exterior anastomosis without sutures. The Journal of the International College of Surgeons. 1962; 37: 591–593. (In French)
[34] YANCEY AG, CROMARTIE JE, Jr, FORD JR, NICHOLS RR, Jr, SAVILLE AF, Jr. A modification of the Swenson technique for congenital megacolon. Journal of the National Medical Association. 1952; 44: 356–363.
[35] Woode D, Avansino J, Sawin R, Cornwell EE, 3rd, Waldhausen J, Gow KW. Asa G Yancey: The first to describe a modification of the Swenson Technique for Hirschsprung disease. Journal of Pediatric Surgery. 2022; 57: 1701–1703. https://doi.org/10.1016/j.jpedsurg.2022.03.030.
[36] SOAVE F. A NEW SURGICAL TECHNIQUE FOR TREATMENT OF HIRSCHSPRUNG'S DISEASE. Surgery. 1964; 56: 1007–1014.
[37] BOLEY SJ. NEW MODIFICATION OF THE SURGICAL TREATMENT OF HIRSCHSPRUNG'S DISEASE. Surgery. 1964; 56: 1015–1017.
[38] Martin LW, Altemeier WA. Clinical Experience with a New Operation (Modified Duhamel Procedure) for Hirschsprung's Disease. Annals of Surgery. 1962; 156: 678–681. https://doi.org/10.1097/00000658-196210000-00014.
[39] Noblett HR. A rectal suction biopsy tube for use in the diagnosis of Hirschsprung's disease. Journal of Pediatric Surgery. 1969; 4: 406–409. https://doi.org/10.1016/0022-3468(69)90606-x.
[40] So HB, Schwartz DL, Becker JM, Daum F, Schneider KM. Endorectal "pull-through" without preliminary colostomy in neonates with Hirschsprung's disease. Journal of Pediatric Surgery. 1980; 15: 470–471. https://doi.org/10.1016/s0022-3468(80)80755-x.
[41] De la Torre-Mondragón L, Ortega-Salgado JA. Transanal endorectal pull-through for Hirschsprung's disease. Journal of Pediatric Surgery. 1998; 33: 1283–1286. https://doi.org/10.1016/s0022-3468(98)90169-5.
[42] De la Torre-Mondragón L, Ortega-Salgado JA, Arroyo-Foneseca AP. Endorectal Pullthrough. Anal and laparoscopic access. Cirujano General. 1998; 19: 151–154.
[43] De La Torre L, Langer JC. Transanal endorectal pull-through for Hirschsprung disease: technique, controversies, pearls, pitfalls, and an organized approach to the management of postoperative obstructive symptoms. Seminars in Pediatric Surgery. 2010; 19: 96–106. https://doi.org/10.1053/j.sempedsurg.2009.11.016.
[44] Soave F. Endorectal pull-through: 20 years experience. Address of the guest speaker, APSA, 1984. Journal of Pediatric Surgery. 1985; 20: 568–579. https://doi.org/10.1016/s0022-3468(85)80003-8.
[45] Langer JC, Durrant AC, de la Torre L, Teitelbaum DH, Minkes RK, Caty MG, *et al*. One-stage transanal Soave pullthrough for Hirschsprung disease: a multicenter experience with 141 children. Annals of Surgery. 2003; 238: 569–83; discussion 583–85. https://doi.org/10.1097/01.sla.0000089854.00436.cd.
[46] Langer JC, Minkes RK, Mazziotti MV, Skinner MA, Winthrop AL. Transanal one-stage Soave procedure for infants with Hirschsprung's disease. Journal of Pediatric Surgery. 1999; 34: 148–51; discussion 152. https://doi.org/10.1016/s0022-3468(99)90246-4.
[47] Georgeson KE, Fuenfer MM, Hardin WD. Primary laparoscopic pull-through for Hirschsprung's disease in infants and children. Journal of Pediatric Surgery. 1995; 30: 1017–21; discussion 1021–22. https://doi.org/10.1016/0022-3468(95)90333-x.
[48] Georgeson KE, Cohen RD, Hebra A, Jona JZ, Powell DM, Rothenberg SS, *et al*. Primary laparoscopic-assisted endorectal colon pullthrough for Hirschsprung's disease: a new gold standard. Annals of Surgery. 1999; 229: 678–678–82; discussion 682–3. https://doi.org/10.1097/00000658-199905000-00010.
[49] Coran AG, Teitelbaum DH. Recent advances in the management of Hirschsprung's disease. American Journal of Surgery. 2000; 180: 382–387. https://doi.org/10.1016/s0002-9610(00)00487-6.
[50] Mohamed W, Elsawaf MI, Shalaby AI, Arafat AE, Marei MM, Aboulfadl MH, *et al*. Optimism for the Single-stage Transanal Swenson in Neonates. Journal of Indian Association of Pediatric Surgeons. 2021; 26: 16–22. https://doi.org/10.4103/jiaps.JIAPS_187_19.
[51] SWENSON O, RATHAUSER F. Segmental dilatation of the colon; a new entity. American Journal of Surgery. 1959; 97: 734–738. https://doi.org/10.1016/0002-9610(59)90338-1.
[52] Sookpotarom P, Vejchapipat P. Primary transanal Swenson pull-through operation for Hirschsprung's disease. Pediatric Surgery International. 2009; 25: 767–773. https://doi.org/10.1007/s00383-009-2428-5.
[53] Rigamonti W, Falchetti D, Torri F, Alberti D, Huscher C, Caccia G. The laparoscopic treatment of Hirschsprung's disease. La Pediatria Medica E Chirurgica: Medical and Surgical Pediatrics. 1994; 16: 499–501. (In Italian)
[54] Hoffmann K, Schier F, Waldschmidt J. Laparoscopic Swenson's procedure in children. European Journal of Pediatric Surgery. 1996; 6: 15–17. https://doi.org/10.1055/s-2008-1066459.
[55] Hebra A, Smith VA, Lesher AP. Robotic Swenson pull-through for Hirschsprung's disease in infants. The American Surgeon. 2011; 77: 937–941.
[56] Weidner BC, Waldhausen JHT. Swenson revisited: a one-stage, transanal pull-through procedure for Hirschsprung's disease. Journal of Pediatric Surgery. 2003; 38: 1208–1211. https://doi.org/10.1016/s0022-3468(03)00269-0.
[57] Peterlini FL, Martins JL. Modified transanal rectosigmoidectomy for Hirschsprung's disease: clinical and manometric results in the initial 20 cases. Journal of Pediatric Surgery. 2003; 38: 1048–1050. https://doi.org/10.1016/s0022-3468(03)00189-1.
[58] Xu ZL, Zhao Z, Wang L, An Q, Tao WF. A new modification of transanal Swenson pull-through procedure for Hirschsprung's disease. Chinese Medical Journal. 2008; 121: 2420–2423.
[59] Zhang SC, Bai YZ, Wang W, Wang WL. Long-term outcome, colonic motility, and sphincter performance after Swenson's procedure for Hirschsprung's disease: a single-center 2-decade experience with 346 cases. American Journal of Surgery. 2007; 194: 40–47. https://doi.org/10.1016/j.amjsurg.2006.10.018.
[60] STATE D. The surgical management of congenital megacolon (Hirschsprung's disease). The American Journal of Gastroenterology. 1954; 22: 47–54.
[61] STATE D. Physiological operation for idiopathic congenital megacolon (Hirschsprung's disease). Journal of the American Medical Association. 1952; 149: 350–355. https://doi.org/10.1001/jama.1952.02930210034010.
[62] State D, Rogers W. The Surgical Treatment of Idiopathic Congenital Megacolon. Bulletin of the University of Minnesota Hospitals and the Minnesota Medical Foundation. 1950; 22: 164–181.
[63] HIATT RB. The surgical treatment of congenital megacolon. Annals of Surgery. 1951; 133: 321–329. https://doi.org/10.1097/00000658-195103000-00006.
[64] REHBEIN F, von ZIMMERMANN H. Results with abdominal resection in Hirschsprung's disease. Archives of Disease in Childhood. 1960; 35: 29–37. https://doi.org/10.1136/adc.35.179.29.
[65] REHBEIN F, WERNICKE HH. Surgical experience in Hirschprung's disease. Bruns' Beitrage Zur Klinischen Chirurgie. 1955; 191: 18–34. (In German)
[66] REHBEIN F, NICOLAI I. OPERATION OF HIRSCHSPRUNG'S DISEASE. RESULTS OF INTRA-ABDOMINAL RESECTION IN 110 CASES. Deutsche Medizinische Wochenschrift (1946). 1963; 88: 1595–1597. https://doi.org/10.1055/s-0028-1112270. (In German)
[67] Rehbein F, Morger R, Kundert JG, Meier-Ruge W. Surgical problems in congenital megacolon (Hirschsprung's disease). A comparison of surgical technics. Journal of Pediatric Surgery. 1966; 1: 526–533. https://doi.org/10.1016/s0022-3468(66)80116-1.
[68] Regbein F. Anterior resection combined with anorectal myectomy in the treatment of Hirschsprung's disease. Journal of Pediatric Surgery.

1980; 15: 117.
[69] Holschneider AM, Söylet Y. Rehbein anterior resection in the treatment of Hirschsprung's congenital megacolon: Manual or stapler anastomosis - a comparative study. Zeitschrift für Kinderchirurgie. 1989; 44: 216–220. https://doi.org/10.1055/s-2008-1043238. (In German)
[70] Wester T, Hoehner J, Olsen L. Rehbein's anterior resection in Hirschsprung's disease, using a circular stapler. European Journal of Pediatric Surgery. 1995; 5: 358–362. https://doi.org/10.1055/s-2008-1066243.
[71] Rassouli R, Holschneider AM, Bolkenius M, Menardi G, Becker MR, Schaarschmidt K, *et al*. Long-term results of Rehbein's procedure: a retrospective study in German-speaking countries. European Journal of Pediatric Surgery. 2003; 13: 187–194. https://doi.org/10.1055/s-2003-41258.
[72] Heinrich M, Häberle B, von Schweinitz D, Stehr M. Re-operations for Hirschsprung's disease: long-term complications. European Journal of Pediatric Surgery. 2011; 21: 325–330. https://doi.org/10.1055/s-0031-1284423.
[73] Zganjer M, Cigit I, Car A, Visnjić S, Butković D. Hirschsprung's disease and Rehbein's procedure–our results in the last 30 years. Collegium Antropologicum. 2006; 30: 905–907.
[74] Gil-Vernet H JM, Royo GF, Brun N, Broto J, Gine C, Moreno A. Rehbein's procedure versus De la Torre in Hirschsprung's disease. Cirugia Pediatrica: Organo Oficial De La Sociedad Espanola De Cirugia Pediatrica. 2009; 22: 42–44. (In Spanish)
[75] Visser R, van de Ven TJ, van Rooij IALM, Wijnen RMH, de Blaauw I. Is the Rehbein procedure obsolete in the treatment of Hirschsprung's disease? Pediatric Surgery International. 2010; 26: 1117–1120. https://doi.org/10.1007/s00383-010-2696-0.
[76] Soave F. Sutureless colon-anostomy after mobilization and extramucosal lowering of the recto-sigmoid. A new surgical technique for the treatment of Hirschsprung's disease. Ospedale Italiano Chirurgia. 1963; 8: 285–294. (In Italian)
[77] SOAVE F. A NEW SURGICAL TECHNIC FOR THE TREATMENT OF HIRSCHSPRUNG'S DISEASE. COLOANOSTOMY WITHOUT SUTURE AFTER MOBILIZATION AND EXTRAMUCOUS LOWERING OF THE RECTO- SIGMOID. Journal De Chirurgie. 1963; 86: 451–464. (In French)
[78] Soave F. A new method for surgical treatment of the hirschsprung disease. Zentralbl Chir. 1963; 88: 1241–1249. (In German)
[79] SOAVE F. HIRSCHSPRUNG'S DISEASE: A NEW SURGICAL TECHNIQUE. Archives of Disease in Childhood. 1964; 39: 116–124. https://doi.org/10.1136/adc.39.204.116.
[80] REHBEIN F. Operation for anal and rectal atresia with rectourethral fistula. Chirurg. 1959; 30: 417–418. (In German)
[81] BABCOCK WW. Radical single stage extirpation for cancer of the large bowel, with retained functional anus. Surgery, Gynecology & Obstetrics. 1947; 85: 1–7.
[82] BACON HE. Cancer of the rectum. Surgery. 1949; 26: 584–589.
[83] Jasonni V, Pini Prato A, Martucciello G. Soave's Extramucosal Endorectal Pull-Through Procedure. In Holschneider A, Puri P (eds.) Hirschsprung's Disease and Allied Disorders (pp.337–348). Springer: Berlin/Heidelberg. 2008. https://doi.org/10.1007/978-3-540-33935-9_25.
[84] Soave F. Extramucosal endorectal pull through. Current Problems in Surgery. 1978; 15: 77–93.
[85] RAVITCH MM. Anal ileostomy with sphincter preservation in patients requiring total colectomy for benign conditions. Surgery. 1948; 24: 170–187.
[86] NICOLAI I, REHBEIN F. Management of imperforate anus with recto-urethral fistula. Archives of Disease in Childhood. 1963; 38: 167–169. https://doi.org/10.1136/adc.38.198.167.
[87] Georgeson KE, Robertson DJ. Laparoscopic-assisted approaches for the definitive surgery for Hirschsprung's disease. Seminars in Pediatric Surgery. 2004; 13: 256–262. https://doi.org/10.1053/j.sempedsurg.2004.10.013.
[88] Sholadoye TT, Ogunsua OO, Alfa Y, Mshelbwala PM, Ameh EA. Outcome of Transanal Endorectal Pull-Through in Patients with Hirschsprung's Disease. African Journal of Paediatric Surgery: AJPS. 2024; 21: 1–5. https://doi.org/10.4103/ajps.ajps_93_22.
[89] Li W, Lin M, Hu H, Sun Q, Su C, Wang C, *et al*. Surgical Management of Hirschsprung's Disease: A Comparative Study Between Conventional Laparoscopic Surgery, Transumbilical Single-Site Laparoscopic Surgery, and Robotic Surgery. Frontiers in Surgery. 2022; 9: 924850. https://doi.org/10.3389/fsurg.2022.924850.
[90] Muto M, Onishi S, Murakami M, Yano K, Harumatsu T, Ieiri S. Transanal Mesenteric Resection in Hirschsprung's Disease Using ICG under Concept of NOTES Technique. European Journal of Pediatric Surgery Reports. 2022; 10: e115–e117. https://doi.org/10.1055/s-0042-1751051.
[91] Till H, Heinrich M, Schuster T, V Schweinitz D. Is the anorectal sphincter damaged during a transanal endorectal pull-through (TERPT) for Hirschsprung's disease? A 3-dimensional, vector manometric investigation. European Journal of Pediatric Surgery. 2006; 16: 188–191. https://doi.org/10.1055/s-2006-924220.
[92] Bischoff A, Frischer J, Knod JL, Dickie B, Levitt MA, Holder M, *et al*. Damaged anal canal as a cause of fecal incontinence after surgical repair for Hirschsprung disease - a preventable and under-reported complication. Journal of Pediatric Surgery. 2017; 52: 549–553. https://doi.org/10.1016/j.jpedsurg.2016.08.027.
[93] Demir M, Akin M, Kaba M, Genc NM, Sever N, Karadag CA, *et al*. Assessment of Patients with Hirschsprung Disease and the Use of Laparoscopy. Sisli Etfal Hastanesi Tip Bulteni. 2020; 54: 218–221. https://doi.org/10.14744/SEMB.2018.84565.
[94] Yang L, Tang ST, Cao GQ, Yang Y, Li S, Li SW, *et al*. Transanal endorectal pull-through for Hirschsprung's disease using long cuff dissection and short V-shaped partially resected cuff anastomosis: early and late outcomes. Pediatric Surgery International. 2012; 28: 515–521. https://doi.org/10.1007/s00383-012-3071-0.
[95] Wester T, Rintala RJ. Early outcome of transanal endorectal pull-through with a short muscle cuff during the neonatal period. Journal of Pediatric Surgery. 2004; 39: 157–160. https://doi.org/10.1016/j.jpedsurg.2003.10.007.
[96] Li AW, Zhang WT, Li FH, Cui XH, Duan XS. A new modification of transanal Soave pull-through procedure for Hirschsprung's disease. Chinese Medical Journal. 2006; 119: 37–42.
[97] Zheng Z, Jin Z, Gao M, Tang C, Huang L, Gong Y, *et al*. Laparoscopic Complete Excision of the Posterior Muscular Cuff: Technique Refinements and Comparison With Stepwise Gradient Muscular Cuff Cutting for Hirschsprung Disease. Frontiers in Pediatrics. 2022; 10: 578843. https://doi.org/10.3389/fped.2022.578843.
[98] Steichen FM, Talbert JL, Ravitch MM. Primary side-to-side colorectal anastomosis in the Duhamel operation for Hirschsprung's disease. Surgery. 1968; 64: 475–483.
[99] Antao B, Radhwan T, Samuel M, Kiely E. Short-pouch and low-anastomosis Duhamel procedure results in better fecal control and normal defecation pattern. Diseases of the Colon and Rectum. 2005; 48: 1791–1796. https://doi.org/10.1007/s10350-005-0086-9.
[100] Grosfeld JL, Ballantine VN, Csicsko JF. A critical evaluation of the Duhamel operation for Hirschsprung's disease. Archives of Surgery (Chicago, Ill.: 1960). 1978; 113: 454–460. https://doi.org/10.1001/archsurg.1978.01370160112019.
[101] Hickey M, Guiney EJ. The Duhamel/Martin operation in the treatment of Hirschsprung's disease. Zeitschrift für Kinderchirurgie. 1984; 39: 110–111. https://doi.org/10.1055/s-2008-1044187.
[102] Peters NJ, Menon P, Rao KLN, Samujh R. Modified Duhamel's Two-Staged Procedure for Hirschsprung's Disease: Further Modifications for Improved Outcomes. Journal of Indian Association of Pediatric Surgeons. 2020; 25: 269–275. https://doi.org/10.4103/jiaps.JIAPS_55_19.
[103] Travassos DV, Bax NMA, Van der Zee DC. Duhamel procedure: a comparative retrospective study between an open and a laparoscopic technique. Surgical Endoscopy. 2007; 21: 2163–2165. https://doi.or

g/10.1007/s00464-007-9317-6.
[104] Vrsansky P, Bourdelat D, Pagés R. Principal modifications of the Duhamel procedure in the treatment of Hirschsprung's disease. Analysis based on results of an international retrospective study of 2,430 patients. Pediatric Surgery International. 1998; 13: 125–132. https://doi.org/10.1007/s003830050265.
[105] Chatoorgoon K, Pena A, Lawal TA, Levitt M. The problematic Duhamel pouch in Hirschsprung's disease: manifestations and treatment. European Journal of Pediatric Surgery. 2011; 21: 366–369. https://doi.org/10.1055/s-0031-1285875.
[106] Gharpure K, Jindal B, Jagadisan B, Naredi BK. Chronic Bleeding Following Duhamel Procedure Due to Staple Line Ulcers. Indian Journal of Pediatrics. 2018; 85: 792–793. https://doi.org/10.1007/s12098-017-2533-8.
[107] Penaloza CSQ, Barreto AC, Ortolan EVP, Zani A, Lourenção PLTDA. Management of Hirschsprung's Disease: A Survey with Brazilian Pediatric Surgeons. Children (Basel, Switzerland). 2024; 11: 1405. https://doi.org/10.3390/children11111405.
[108] Scholfield DW, Ram AD. Laparoscopic Duhamel Procedure for Hirschsprung's Disease: Systematic Review and Meta-analysis. Journal of Laparoendoscopic & Advanced Surgical Techniques. Part a. 2016; 26: 53–61. https://doi.org/10.1089/lap.2015.0121.
[109] Muller CO, Rossignol G, Montalva L, Viala J, Martinez-Vinson C, Mosca A, *et al*. Long-Term Outcome of Laparoscopic Duhamel Procedure for Extended Hirschsprung's Disease. Journal of Laparoendoscopic & Advanced Surgical Techniques. Part a. 2016; 26: 1032–1035. https://doi.org/10.1089/lap.2016.0152.
[110] Singh S, Wakhlu A, Ahmad I, Srivastava NK. The Laparoscopic Assisted Duhamel Pull through Procedure for Hirschsprung's Disease: Our Technique and Short Term Results. Journal of Pediatrics & Neonatal Care. 2017; 7: 00281. https://doi.org/10.15406/jpnc.2017.07.00281.
[111] Arafa A, Eltantawi HE, Ragab M. Laparoscopic-assisted duhamel for hirschsprung's children older than 3 years. African Journal of Paediatric Surgery: AJPS. 2022; 19: 27–31. https://doi.org/10.4103/ajps.AJPS_1_21.
[112] Bhandarkar K, De Coppi P, Cross K, Blackburn S, Curry J. Long-Term Functional Outcomes and Multidisciplinary Management after Ileorectal Duhamel Pull-Through for Total Colonic Aganglionosis-20-Year Experience in a Tertiary Surgical Center. European Journal of Pediatric Surgery. 2024; 34: 423–429. https://doi.org/10.1055/a-2181-2065.
[113] An YK, Yuan ZX, Wang SL, Cai J. Clinicopathological features and prognosis of 47 adults with Hirschsprung's disease and Hirschsprung's disease allied disorders. Zhonghua Wei Chang Wai Ke Za Zhi = Chinese Journal of Gastrointestinal Surgery. 2023; 26: 1154–1161. https://doi.org/10.3760/cma.j.cn441530-20230421-00131. (In Chinese)
[114] Han Y, Lin MB, Zhang YJ, Yin L. Total laparoscopic modified Duhamel operation in combination with transanal endoscopic microsurgery. JSLS: Journal of the Society of Laparoendoscopic Surgeons. 2014; 18: 128–131. https://doi.org/10.4293/108680813X13693422520288.
[115] Moore SW. Total colonic aganglionosis and Hirschsprung's disease: a review. Pediatric Surgery International. 2015; 31: 1–9. https://doi.org/10.1007/s00383-014-3634-3.
[116] Rolle U, O'Briain S, Pearl RH, Puri P. Megacystis-microcolon-intestinal hypoperistalsis syndrome: evidence of intestinal myopathy. Pediatric Surgery International. 2002; 18: 2–5. https://doi.org/10.1007/s003830200001.
[117] Stranzinger E, DiPietro MA, Teitelbaum DH, Strouse PJ. Imaging of total colonic Hirschsprung disease. Pediatric Radiology. 2008; 38: 1162–1170. https://doi.org/10.1007/s00247-008-0952-4.
[118] Cowles RA, Berdon WE, Holt PD, Buonomo C, Stolar CJ. Neonatal intestinal obstruction simulating meconium ileus in infants with long-segment intestinal aganglionosis: radiographic findings that prompt the need for rectal biopsy. Pediatric Radiology. 2006; 36: 133–137. https://doi.org/10.1007/s00247-005-0043-8.
[119] Rentea RM, Bokova E, Frischer JS, Gosain A, Langer JC, Levitt MA, *et al*. Evaluation and Management of Total Colonic Hirschsprung Disease: A Comprehensive Review From the American Pediatric Surgical Association (APSA) Hirschsprung Disease Interest Group. Journal of Pediatric Surgery. 2024; 59: 161677. https://doi.org/10.1016/j.jpedsurg.2024.08.017.
[120] Märzheuser S, Schulze F, Lindert J. Surgical Strategies in Total Colonic Aganglionosis: Primary Pullthrough-Pathway of Care. Children (Basel, Switzerland). 2024; 11: 911. https://doi.org/10.3390/children11080911.
[121] Granström AL, Irvine W, Hoel AT, Tabbers M, Kyrklund K, Fascetti-Leon F, *et al*. Ernica Clinical Consensus Statements on Total Colonic and Intestinal Aganglionosis. Journal of Pediatric Surgery. 2024; 59: 161565. https://doi.org/10.1016/j.jpedsurg.2024.04.019.
[122] Bokova E, Prasade N, Janumpally S, Rosen JM, Lim IIP, Levitt MA, *et al*. State of the Art Bowel Management for Pediatric Colorectal Problems: Hirschsprung Disease. Children (Basel, Switzerland). 2023; 10: 1418. https://doi.org/10.3390/children10081418.
[123] Wood RJ, Garrison AP. Total Colonic Aganglionosis in Hirschsprung disease. Seminars in Pediatric Surgery. 2022; 31: 151165. https://doi.org/10.1016/j.sempedsurg.2022.151165.
[124] Payen E, Talbotec C, Chardot C, Capito C, Khen-Dunlop N, Sarnacki S, *et al*. Outcome of Total Colonic Aganglionosis Involving the Small Bowel Depends on Bowel Length, Liver Disease, and Enterocolitis. Journal of Pediatric Gastroenterology and Nutrition. 2022; 74: 582–587. https://doi.org/10.1097/MPG.0000000000003415.
[125] Reinshagen K, Burmester G, Hagens J, Krebs TF, Tomuschat C. Colectomy Followed by J-Pouch Reconstruction to Correct Total Colonic Aganglionosis. Children (Basel, Switzerland). 2022; 9: 101. https://doi.org/10.3390/children9010101.
[126] Verkuijl SJ, Meinds RJ, van der Steeg AFW, van Gemert WG, de Blaauw I, Witvliet MJ, *et al*. Functional Outcomes After Surgery for Total Colonic, Long-Segment, Versus Rectosigmoid Segment Hirschsprung Disease. Journal of Pediatric Gastroenterology and Nutrition. 2022; 74: 348–354. https://doi.org/10.1097/MPG.0000000000003355.
[127] Pontarelli EM, Ford HR, Gayer CP. Recent developments in Hirschsprung's-associated enterocolitis. Current Gastroenterology Reports. 2013; 15: 340. https://doi.org/10.1007/s11894-013-0340-6.
[128] Redkar RG, Raj V, Bangar A, Hathiramani V, Chigicherla S, Tewari S. Role of ano rectal myomectomy in children with chronic refractory constipation. African Journal of Paediatric Surgery: AJPS. 2018; 15: 31–35. https://doi.org/10.4103/ajps.AJPS_99_17.
[129] Pattana-arun J, Ruanroadroun T, Tantiphalachiva K, Sahakitrungruang C, Attithansakul P, Rojanasakul A. Internal sphincter myectomy for adult Hirschsprung's disease: a single institute experience. Journal of the Medical Association of Thailand = Chotmaihet Thangphaet. 2010; 93: 911–915.
[130] Skába R, Rousková B, Simsová M, Kalousová J, Pýcha K. Treatment of the Hirschsprung's disease (HD) at the Department of Pediatric Surgery of Charles University, 2nd Faculty of Medicine, from 1979 to 2004. Casopis Lekaru Ceskych. 2004; 143: 748–751. (In Czech)
[131] Rahardjo TM, Nurzaman YA, Natalia J, Hapdijaya I, Devina L, Andrianto H, *et al*. Adult Hirschsprung's disease presenting as chronic constipation: a case report. Journal of Medical Case Reports. 2023; 17: 308. https://doi.org/10.1186/s13256-023-03986-y.
[132] Goto S, Ikeda K. Histochemical acetylcholinesterase activity in the mucosa of the resected bowel in Hirschsprung's disease. An analysis of 30 cases. Zeitschrift für Kinderchirurgie. 1985; 40: 26–30. https://doi.org/10.1055/s-2008-1059706.
[133] Bruder E, Meier-Ruge WA. Twenty years diagnostic competence center for Hirschsprung's disease in Basel. Der Chirurg. 2010; 81: 572–576. https://doi.org/10.1007/s00104-010-1924-4. (In German)
[134] Holschneider AM, Meier-Ruge W, Ure BM. Hirschsprung's disease

and allied disorders–a review. European Journal of Pediatric Surgery. 1994; 4: 260–266. https://doi.org/10.1055/s-2008-1066115.

[135] Hong SM, Hong J, Kang G, Moon SB. Ultrashort-segment Hirschsprung's disease complicated by megarectum: A case report. Journal of Pediatric Surgery Case Reports. 2014; 2: 385–387. https://doi.org/10.1016/j.epsc.2014.07.013.

[136] Glasgow WR, Lintzeris D, Stockton L, Harris Z. Ultrashort-Segment Hirschsprung's Disease Complicated by Megarectum and Obstructive Uropathy: A Case Report. Cureus. 2023; 15: e48851. https://doi.org/10.7759/cureus.48851.

[137] Rintala RJ, Pakarinen MP. Long-term outcomes of Hirschsprung's disease. Seminars in Pediatric Surgery. 2012; 21: 336–343. https://doi.org/10.1053/j.sempedsurg.2012.07.008.

[138] Bianchi A. Intestinal loop lengthening–a technique for increasing small intestinal length. Journal of Pediatric Surgery. 1980; 15: 145–151. https://doi.org/10.1016/s0022-3468(80)80005-4.

[139] Bianchi A. Intestinal lengthening: an experimental and clinical review. Journal of the Royal Society of Medicine. 1984; 77 Suppl 3: 35–41.

[140] Boroni G, Parolini F, Stern MV, Moglia C, Alberti D. Autologous Intestinal Reconstruction Surgery in Short Bowel Syndrome: Which, When, and Why. Frontiers in Nutrition. 2022; 9: 861093. https://doi.org/10.3389/fnut.2022.861093.

[141] Kim HB, Fauza D, Garza J, Oh JT, Nurko S, Jaksic T. Serial transverse enteroplasty (STEP): a novel bowel lengthening procedure. Journal of Pediatric Surgery. 2003; 38: 425–429. https://doi.org/10.1053/jpsu.2003.50073.

[142] van Praagh JB, Hofker HS, Haveman JW. Comparing bowel lengthening procedures: which, when, and why? Current Opinion in Organ Transplantation. 2022; 27: 112–118. https://doi.org/10.1097/MOT.0000000000000957.

[143] Saeki I, Kurihara S, Kojima M, Ohge H, Takahashi S, Hiyama E. A new surgical technique for short bowel syndrome. BMC Surgery. 2022; 22: 375. https://doi.org/10.1186/s12893-022-01823-5.

[144] Höllwarth ME. Surgical strategies in short bowel syndrome. Pediatric Surgery International. 2017; 33: 413–419. https://doi.org/10.1007/s00383-016-4043-6.

[145] Cserni T, Biszku B, Guthy I, Dicso F, Szaloki L, Folaranmi S, *et al*. The first clinical application of the spiral intestinal lengthening and tailoring (silt) in extreme short bowel syndrome. Journal of Gastrointestinal Surgery: Official Journal of the Society for Surgery of the Alimentary Tract. 2014; 18: 1852–1857. https://doi.org/10.1007/s11605-014-2577-2.

[146] Hou J, Feng W, Zhao H, Cui M, Wang Y, Guo Z, *et al*. Robotic-assisted Swenson procedure for Hirschsprung's disease with a median age of 35 days: a single-center retrospective study. Pediatric Surgery International. 2025; 41: 87. https://doi.org/10.1007/s00383-025-05988-x.

[147] Cserni T, Varga G, Erces D, Kaszaki J, Boros M, Laszlo A, *et al*. Spiral intestinal lengthening and tailoring - first in vivo study. Journal of Pediatric Surgery. 2013; 48: 1907–1913. https://doi.org/10.1016/j.jpedsurg.2013.01.048.

[148] Li Y, He S, Jin Z, Tang C, Gong Y, Huang L, *et al*. Comparison of robot-assisted and laparoscopic-assisted modified Soave short muscle cuff anastomosis surgeries for classical Hirschsprung disease. BMC Surgery. 2025; 25: 78. https://doi.org/10.1186/s12893-025-02799-8.

[149] Pini Prato A, Lacher M. Advancing Pediatric Robotic Colorectal Surgery: Trends, Outcomes, and Future Directions-A Comprehensive Review. European Journal of Pediatric Surgery. 2025; 35: 79–88. https://doi.org/10.1055/a-2506-6590.

[150] Delgado-Miguel C, Reparaz L, Clarkson W, Desai K, Camps JI. Colorectal Robotic-Assisted Surgery in Children. Long-Term Outcomes and Pitfalls. The International Journal of Medical Robotics and Computer Assisted Surgery. 2024; 20: e70032. https://doi.org/10.1002/rcs.70032.

[151] Almadhoun MKIK, Morcos RKA, Alsadoun L, Bokhari SFH, Ahmed Z, Khilji F, *et al*. Minimally Invasive Surgery for Hirschsprung Disease: Current Practices and Future Directions. Cureus. 2024; 16: e66444. https://doi.org/10.7759/cureus.66444.

[152] Duhoky R, Claxton H, Piozzi GN, Khan JS. Robotic Approach to Paediatric Gastrointestinal Diseases: A Systematic Review. Children (Basel, Switzerland). 2024; 11: 273. https://doi.org/10.3390/children11030273.

[153] Jin Y, Cai D, Zhang S, Luo W, Zhang Y, Huang Z, *et al*. Robot-assisted abdominal surgery in children less than 5 months of age: retrospective cohort study. International Journal of Surgery (London, England). 2024; 110: 859–863. https://doi.org/10.1097/JS9.0000000000000867.

[154] Zhang M, Zhang X, Chi S, Chang X, Zeng J, Bian H, *et al*. Robotic-assisted Proctosigmoidectomy Versus Laparoscopic-assisted Soave Pull-through for Hirschsprung Disease: Medium-term Outcomes From a Prospective Multicenter Study. Annals of Surgery. 2025; 281: 689–697. https://doi.org/10.1097/SLA.0000000000006172.

[155] Zhang S, Cai D, Zhang Y, Pan T, Chen K, Jin Y, *et al*. Comparation of robotic-assisted surgery and laparoscopic assisted surgery in children with Hirschsprung's disease: a single-centered retrospective study. BMC Surgery. 2023; 23: 294. https://doi.org/10.1186/s12893-023-02169-2.

[156] Huang J, Huang Z, Mei H, Rong L, Zhou Y, Guo J, *et al*. Cost-effectiveness analysis of robot-assisted laparoscopic surgery for complex pediatric surgical conditions. Surgical Endoscopy. 2023; 37: 8404–8420. https://doi.org/10.1007/s00464-023-10399-x.

[157] Zhang MX, Zhang X, Chang XP, Zeng JX, Bian HQ, Cao GQ, *et al*. Robotic-assisted proctosigmoidectomy for Hirschsprung's disease: A multicenter prospective study. World Journal of Gastroenterology. 2023; 29: 3715–3732. https://doi.org/10.3748/wjg.v29.i23.3715.

[158] Alsaawy SM, Gauci CM, Khalessi A, Phan-Thien KC. Robotic ultra-low anterior resection for idiopathic megarectum. ANZ Journal of Surgery. 2023; 93: 2241–2242. https://doi.org/10.1111/ans.18371.

[159] Xie X, Li K, Xiang B. Influencing factors of parental selections of minimally invasive procedure in children's abdominal surgery: a cross-sectional study in China. Journal of Robotic Surgery. 2023; 17: 1681–1687. https://doi.org/10.1007/s11701-023-01566-2.

[160] Ahmad H, Shaul DB. Pediatric colorectal robotic surgery. Seminars in Pediatric Surgery. 2023; 32: 151259. https://doi.org/10.1016/j.sempedsurg.2023.151259.

[161] Mottadelli G, Erculiani M, Casella S, Dusio MP, Felici E, Milanese T, *et al*. Robotic surgery in Hirschsprung disease: a unicentric experience on 31 procedures. Journal of Robotic Surgery. 2023; 17: 897–904. https://doi.org/10.1007/s11701-022-01488-5.

[162] Quynh TA, Hien PD, Du LQ, Long LH, Tran NTN, Hung T. The follow-up of the robotic-assisted Soave procedure for Hirschsprung's disease in children. Journal of Robotic Surgery. 2022; 16: 301–305. https://doi.org/10.1007/s11701-021-01238-z.

[163] Pini Prato A, Arnoldi R, Dusio MP, Cimorelli A, Barbetta V, Felici E, *et al*. Totally robotic soave pull-through procedure for Hirschsprung's disease: lessons learned from 11 consecutive pediatric patients. Pediatric Surgery International. 2020; 36: 209–218. https://doi.org/10.1007/s00383-019-04593-z.

[164] Mattioli G, Pio L, Leonelli L, Razore B, Disma N, Montobbio G, *et al*. A Provisional Experience with Robot-Assisted Soave Procedure for Older Children with Hirschsprung Disease: Back to the Future? Journal of Laparoendoscopic & Advanced Surgical Techniques. Part a. 2017; 27: 546–549. https://doi.org/10.1089/lap.2016.0337.

[165] Rickey J, Robinson CC, Camps JI, Lagares-Garcia JA. Robotic-assisted Soave procedure in an 18-year-old man with adult short-segment Hirschsprung's disease. The American Surgeon. 2013; 79: E223–5.

[166] Riccio A, Lisi G, Miscia ME, Di Paolo G, Lauriti G, Lelli Chiesa P. Secondary hydrosalpinx in adolescents: a challenging decision-making process for surgical choice and future fertility preservation. La Pediatria Medica E Chirurgica: Medical and Surgical Pe-

diatrics. 2021; 42: 10.4081/pmc.2020.233. https://doi.org/10.4081/pmc.2020.233.

[167] Pini Prato A, Faticato MG, Falconi I, Felici E, Casaccia G, Caraccia M, *et al*. Skipped aganglionic lengthening transposition (SALT) for short bowel syndrome in patients with total intestinal aganglionosis: technical report and feasibility. Pediatric Surgery International. 2020; 36: 1507–1510. https://doi.org/10.1007/s00383-020-04763-4.

[168] Pini Prato A, Arnoldi R, Faticato MG, Mariani N, Dusio MP, Felici E, *et al*. Minimally Invasive Redo Pull-Throughs in Hirschsprung Disease. Journal of Laparoendoscopic & Advanced Surgical Techniques. Part a. 2020; 30: 1023–1028. https://doi.org/10.1089/lap.2020.0250.

[169] Pakarinen MP, Mutanen A. Long-term outcomes and quality of life in patients with Hirschsprung disease. World Journal of Pediatric Surgery. 2024; 7: e000859. https://doi.org/10.1136/wjps-2024-000859.

[170] Lindert J, Day H, de Andres Crespo M, Amerstorfer E, Alexander S, Backes M, *et al*. Influence of Diet on Bowel Function and Abdominal Symptoms in Children and Adolescents with Hirschsprung Disease-A Multinational Patient-Reported Outcome Survey. Children (Basel, Switzerland). 2024; 11: 1118. https://doi.org/10.3390/children11091118.

[171] Evans-Barns HME, Hall M, Trajanovska M, Hutson JM, Muscara F, King SK. Psychosocial Outcomes of Parents of Children with Hirschsprung Disease Beyond Early Childhood. Journal of Pediatric Surgery. 2024; 59: 694–700. https://doi.org/10.1016/j.jpedsurg.2023.11.012.

[172] Koo FEC, Chan MCE, King SK, Trajanovska M. The early years: hirschsprung disease and health-related quality of life. Quality of Life Research: an International Journal of Quality of Life Aspects of Treatment, Care and Rehabilitation. 2023; 32: 3327–3337. https://doi.org/10.1007/s11136-023-03482-2.

[173] Judd-Glossy L, Ariefdjohan M, Ketzer J, Wehrli LA, Pena A, de la Torre L, *et al*. Long-term outcomes of adult patients following surgery for congenital colorectal conditions: analysis of psychosocial functioning. Pediatric Surgery International. 2022; 38: 1685–1692. https://doi.org/10.1007/s00383-022-05212-0.

[174] Verkuijl SJ, Meinds RJ, van der Steeg AFW, Sloots CEJ, van Heurn E, de Blaauw I, *et al*. Familial Experience With Hirschsprung's Disease Improves the Patient's Ability to Cope. Frontiers in Pediatrics. 2022; 10: 820976. https://doi.org/10.3389/fped.2022.820976.

[175] Feng X, Lacher M, Quitmann J, Witt S, Witvliet MJ, Mayer S. Health-Related Quality of Life and Psychosocial Morbidity in Anorectal Malformation and Hirschsprung's Disease. European Journal of Pediatric Surgery. 2020; 30: 279–286. https://doi.org/10.1055/s-0040-1713597.

[176] Gause CD, Krishnaswami S. Management of Anorectal Malformations and Hirschsprung Disease. The Surgical Clinics of North America. 2022; 102: 695–714. https://doi.org/10.1016/j.suc.2022.07.005.

[177] Georgeson KE. Laparoscopic-assisted total colectomy with pouch reconstruction. Seminars in Pediatric Surgery. 2002; 11: 233–236. https://doi.org/10.1053/spsu.2002.35361.

[178] Georgeson KE. Laparoscopic-assisted pull-through for Hirschsprung's disease. Seminars in Pediatric Surgery. 2002; 11: 205–210. https://doi.org/10.1053/spsu.2002.35350.

[179] van der Zee DC, Bax KN. One-stage Duhamel-Martin procedure for Hirschsprung's disease: a 5-year follow-up study. Journal of Pediatric Surgery. 2000; 35: 1434–1436. https://doi.org/10.1053/jpsu.2000.16407.

[180] van der Zee DC, Bax NM. Duhamel-Martin procedure for Hirschsprung's disease in neonates and infants: one-stage operation. Journal of Pediatric Surgery. 1996; 31: 901–902. https://doi.org/10.1016/s0022-3468(96)90406-6.

[181] Smith BM, Steiner RB, Lobe TE. Laparoscopic Duhamel pullthrough procedure for Hirschsprung's disease in childhood. Journal of Laparoendoscopic Surgery. 1994; 4: 273–276. https://doi.org/10.1089/lps.1994.4.273.

[182] Dohrmann P, Mengel W, Schaube H. Deep anterior resection with circular stapled anastomosis of congenital megacolon: clinical results. Progress in Pediatric Surgery. 1990; 25: 97–102. https://doi.org/10.1007/978-3-642-87707-0_12.

[183] Zani A, Eaton S, Morini F, Puri P, Rintala R, Heurn EV, *et al*. European Paediatric Surgeons' Association Survey on the Management of Hirschsprung Disease. European Journal of Pediatric Surgery. 2017; 27: 96–101. https://doi.org/10.1055/s-0036-1593991.

[184] Dickie BH, Webb KM, Eradi B, Levitt MA. The problematic Soave cuff in Hirschsprung disease: manifestations and treatment. Journal of Pediatric Surgery. 2014; 49: 77–80; discussion 80–1. https://doi.org/10.1016/j.jpedsurg.2013.09.034.

[185] WYLLIE GG. Treatment of Hirschsprung's disease by Swenson's operation. Lancet (London, England). 1957; 272: 850–855. https://doi.org/10.1016/s0140-6736(57)91389-2.