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PRACTICE GUIDELINE

Updated European Reference Network for rare Inherited and Congenital Digestive and Gastrointestinal Anomalies guidelines for the management of rectosigmoid Hirschsprung's disease 2025

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Abstract

Objectives: To revise the 2018 European Reference Network for rare Inherited and Congenital Digestive and Gastrointestinal Anomalies (ERNICA) clinical guideline for the management of rectosigmoid Hirschsprung's disease (HSCR) based on new evidence and evolving clinical priorities, ensuring continued relevance, trustworthiness, and consistency in care.

Methods: The update followed a structured five-step process: identifying new evidence (2018–2024), prioritizing recommendations using the Update Priority Tool, and revising selected topics using the Grading of Recommendations Assessment, Development and Evaluation-Evidence to Decision (GRADE EtD) framework. A multidisciplinary panel of pediatric surgeons, patient representatives, and methodologists from 12 countries oversaw the process. Revised recommendations were developed through evidence review and consensus meetings.

Results: Ten prioritized clinical topics were revised. Key updates include refined recommendations on histopathological staining in diagnosis, preoperative screening for renal anomalies and malnutrition, postoperative anastomotic calibration, individualized dietary guidance, sexual and fertility health during follow-up, transition to adult care, and the role of botulinum toxin in managing Hirschsprung's-associated enterocolitis (HAEC). Routine preventive anal dilatations were no longer recommended. The guideline highlights the importance of structured, multidisciplinary, and patient-centered care throughout the lifespan and identifies several research gaps, including nutrition, transition of care, fertility, and HAEC definitions.

For affiliations refer to page 203.

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[Correction added on 5 June 2026, after first online publication. Article format and title have been updated.]

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Conclusion: This updated guideline reflects current evidence and expert consensus, supporting equitable, patient-centered HSCR care. Implementation will be supported via the European Pediatric Surgical Audit (EPSA), and future updates will be informed by registry data and ongoing research.

KEYWORDS

consensus, long-term outcomes, multidisciplinary care, pediatric surgery, rare diseases

1 | BACKGROUND

Hirschsprung's disease (HSCR) is a congenital malformation of the enteric nervous system, marked by the absence of ganglion cells in the myenteric and submucosal plexuses of the distal intestine. This lack of innervation disrupts normal peristalsis and leads to chronic functional bowel obstruction.[1] The condition typically affects the rectum or rectosigmoid region, though in 5%–10% of cases, aganglionosis may extend through the entire colon or even into the small intestine.[2] HSCR occurs in approximately 1.43 (1.37–1.50) in 10,000 live births[3] and shows a strong male predominance, with a male-to-female ratio of about 4:1. Surgical removal of the aganglionic segment remains the cornerstone of treatment, with procedures such as the Swenson, Soave, and Duhamel techniques commonly employed.[1] Despite definitive surgical intervention, postoperative bowel dysfunction and associated disease-related morbidity remain prevalent, underscoring the need for highly specialized, long-term follow-up care and a coordinated transition of care to adult healthcare services.

The establishment of European Reference Networks (ERNs) was driven by the need to overcome the challenges posed by the geographical spread of both expert healthcare providers and patients across Europe. A central objective of the ERNs is to reduce healthcare inequalities and to promote access to high-quality, specialized care for individuals affected by rare or low-prevalence, complex diseases. Within this framework, the ERN for rare Inherited and Congenital Digestive and Gastrointestinal Anomalies (ERNICA) was created, covering a range of conditions including HSCR.[4]

Clinical guidelines are intended to guide clinical practice by offering recommendations to healthcare professionals based on the best available evidence, a careful assessment of benefits and harms, and the opinions and experiences of both patients and clinical experts.[5] Implementation of such guidance is expected to reduce unwarranted practice variation, thereby improving care quality, patient outcomes, and experiences.[5,6] An ERNICA clinical guideline for the management of HSCR in Europe was developed in 2018 and published in 2020. It was the product of an international expert workgroup from eight European countries within the ERNICA ERN, including specialists in surgery, medicine, histopathology, microbiology, genetics, as well as representatives from patient organizations: the German support group for anorectal malformations and HSCR, and Associazione Italiana Morbo di Hirschsprung (A. Mor. Hi), the Italian Society for HSCR.[7]

As new research evidence continues to emerge, including changes in available interventions, outcomes, and costs, maintaining the trustworthiness of clinical guidelines becomes increasingly complex and requires regular monitoring and critical appraisal of the literature. Given the continuous evolution of knowledge, regular updates are necessary to ensure reliability and clinical relevance. Guideline updating typically involves staged approaches to assess the need for revision and to integrate new findings.[8] In line with ERNICA's scheduled 5-year cycle of clinical guideline revision,[6] the 2018 guideline for the management of rectosigmoid HSCR was selected for an update. The aim of this study is to describe the systematic approach and outcomes of the revision process.

What is Known

- A European, multidisciplinary ERNICA guideline for the management of rectosigmoid Hirschsprung's disease (HSCR) was published in 2018.
- The guideline aimed to reduce practice variation but required updating due to emerging evidence and evolving clinical priorities.

What is New

- Targeted revision of key domains: diagnosis, pre- and postoperative care, poor functional outcomes, long-term follow-up, and Hirschsprung's-associated enterocolitis.
- New recommendations for routine screening for congenital anomalies of kidney and urinary tract (CAKUT) and systematic assessment of nutritional status.
- Clear distinction between anal calibration and dilatation, with removal of routine preventive dilatations.
- Expanded focus on patient-centered, lifelong care, including diet, sexual health, fertility, transition to adult care, and implementation via ERNICA–European Pediatric Surgical Audit.

2 | METHODS

2.1 | Ethics statement

This article is a clinical practice guideline developed through expert consensus and does not involve the collection of new human data, animal experimentation, or any procedures requiring ethical approval. No institutional review board approval was therefore required.

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2.2 | Update process

The update process followed established stages of guideline updating, including (i) identification of new scientific evidence, (ii) assessment of the need to update, (iii) prioritization of recommendations to update, (iv) updating the recommendations, and (v) transparent documentation and publication of the update.[8] In this update cycle, a pragmatic and focused approach was adopted: only prioritized clinical recommendations were subject to revision based on newly available evidence or a shift in expert opinion based on recent experiences, while non-prioritized recommendations remained unchanged. Organizational recommendations were not reassessed, as changes in these areas after 5 years are unlikely and more challenging to evaluate. The methodological approach combined the Grading of Recommendations Assessment, Development and Evaluation (GRADE) Evidence-to-Decision (EtD) framework,[9] which was applied to reassess and formulate individual recommendations, with the Checklist for the Reporting of Updated Guidelines (CheckUp) framework to structure documentation of the updating process.[8] Recommendations marked as "unchanged" in the tables below were not selected for revision during the prioritization process and remain valid for another 5 year period, unless new evidence warrants evaluation sooner.

2.3 | Panel composition

A multidisciplinary panel was assembled to oversee the revision process, and a steering committee was established. The panel included 28 pediatric surgeons and one dietician from 25 centers across 12 European countries (Belgium, Czech Republic, Denmark, Finland, France, Germany, Italy, Netherlands, Norway, Poland, Spain, and Sweden) four patient representatives representing three patient groups (SoMA, the German support group for anorectal malformations and HSCR, A. Mor. Hi, the Italian Society for HSCR, and the Dutch Association for Hirschsprung's disease) and an implementation coordinator. The steering committee included six pediatric surgeons from three ERNICA centers across three countries. All expert centers for HSCR and patient organizations recognized by ERNICA were invited to participate in the guideline update. All ERNICA expert centers' representatives were invited to nominate one panel member from relevant disciplines, preferably a pediatric surgeon or pediatric gastroenterologist. Selection emphasized ensuring multidisciplinary expertise, previous experience in HSCR management, and a demonstrated commitment to evidence-based care. All nominated panel members were pediatric surgeons. Even after explicitly asking centers to consider delegating a pediatric gastroenterologist, no pediatric gastroenterologists signed up as panel members. Participating centers reported that, based on the prioritized topics for this update cycle, they considered it more relevant for their pediatric surgeons to participate. All panel members were required to disclose potential conflicts of interest prior to their participation. The panel's diversity in terms of discipline, country, and representation aimed to ensure a comprehensive, patient-centered, and clinically relevant update process.

2.4 | Study design

A systematic literature search was conducted to identify new evidence published between January 2018 (the search cut-off date for the previous guideline) and April 2024. Searches were performed in PubMed, Embase, and other relevant databases, using the full search strategy detailed in Supporting Information S1: Data 1. Titles and abstracts were screened independently by two reviewers (D.R. and W.I.). Studies were categorized according to their relevance to existing recommendations, emerging new topics, or as general supportive references. Full-text review was performed for all studies deemed potentially relevant to the guideline update (see Preferred Reporting Items for Systematic Reviews and Meta-Analyses [PRISMA] flowchart in Supporting Information S1: Data 2).

Following the initial review of new evidence, a prioritization exercise was conducted to identify which of the existing guidelines' clinical recommendations should be revised. Each existing section of recommendations was evaluated against four priority criteria: the risk to patient safety if outdated, the possibility of a shift in the balance between benefits and harms, the availability of new evidence, and the contextual relevance to current practice. Panel members scored each criterion using a 7-point Likert scale, following the structure of the Update Priority Tool (UPTool).[10] For each section of the guideline, percentage scores were calculated for each priority domain (safety, harms, evidence, expertise), and an overall domain score was derived from these four measures. The resulting scores were used to rank the sections and guide the selection of recommendations for full evidence review. Recommendations that achieved high aggregate priority scores were selected for full evidence review. To ensure the feasibility of the update process within the available timeframe and resources, the panel agreed to limit the number of recommendations undergoing full revision to a maximum of 10.

For each prioritized topic, evidence tables were prepared, summarizing study characteristics, key findings, and quality assessments. Articles were assigned an evidence level (I–III) based on the Canadian Task Force on the Periodic Health Examination classification system (1979),[11] as summarized in Supporting Information S1: Data 3. Comparative questions were evaluated according to GRADE methodology,[12] rating the certainty of evidence per outcome measure and formulating recommendations accordingly. Non-comparative clinical questions were assessed using the Oxford Center for Evidence-Based Medicine (OCEBM) levels of evidence.[13] All summarized evidence was distributed to panel members prior to the final consensus meeting.

During the face-to-face consensus meeting, the panel jointly completed a structured GRADE EtD framework for each prioritized recommendation.[9] The discussion was facilitated to systematically address the balance of benefits and harms, stakeholder values, resource implications, feasibility, acceptability, and equity considerations. Panel members contributed their views and deliberations in real time, and a designated rapporteur documented the group's judgments and discussion points. Panelists could also contribute additional recent scientific papers to support specific points raised during the discussions. Based on these discussions, draft revised recommendations were formulated and recorded.

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TABLE 1 Recommendations for the diagnosis of HSCR.

Recommendations	Level of evidence and strength of recommendation	Revision status
The diagnosis of HSCR should be based on representative rectal histology and should be confirmed before pull-through surgery.[14–16] • RSB and open biopsy are equally accurate if they provide enough submucosal tissue. The least invasive, feasible method should be chosen. • Biopsies should be taken from the posterior and/or lateral rectal wall at least 2 cm proximal to the dentate line or 3 cm from the anal orifice, and must contain a representative amount of submucosa.[17–19] • A minimum of one histologically representative tissue sample is required. One open biopsy usually provides sufficient tissue, but with RSB, it is advisable to take 2–3 biopsies.	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
Rectal biopsy is indicated if the clinical history and physical signs are suggestive of HSCR. • The classic triad of symptoms is delayed passage of meconium (>24 h in a term infant), abdominal distension, and bilious vomiting.[20,21] • The majority of patients present during the neonatal period or early infancy.[20–22] • The threshold for biopsy is lowered by the presence of a syndrome associated with HSCR or family history of HSCR.[21]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
Rectal biopsy should also be considered for the exclusion of HSCR in: • Early-onset constipation associated with failure to thrive. • Older children with persistent constipation or symptoms of more generalized intestinal motility disorders. • Patients with an absent RAIR on anorectal manometry.	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged
Biopsies should be evaluated by an experienced consultant histopathologist, with external consultation if needed. • The presence of any number of ganglion cells on H&E staining excludes HSCR.[16,17,21] • If ganglion cells are not seen, further histologic evaluation is required before confirming the diagnosis of HSCR. • **A combination of H&E and calretinin is recommended to improve diagnostic accuracy, especially in challenging samples of premature infants where the ganglion cells are small and not well visualized on H&E.**[23–25] • In HSCR, acetylcholinesterase activity is increased, and calretinin immunohistochemistry is negative.[26,27] Within Europe, external consultation can be requested from an ERNICA center. The CPMS e-platform is fully operational and currently used within ERNICA for cross-border case discussions.	Level of evidence III. Strength of recommendation: Strong, for	**Modified**

Abbreviations: CPMS, Clinical Patient Management System; ERNICA, European Reference Network for rare Inherited and Congenital Digestive and Gastrointestinal Anomalies; H&E, hematoxylin and eosin; HSCR, Hirschsprung's disease; RAIR, recto-anal inhibitory reflex; RSB, rectal suction biopsy.

An overview of the methodological workflow is visually summarized in the implementation section at the end of this paper.

Updated recommendations are reported in this paper, with each recommendation categorized as "new," "modified," or "unchanged." For each change, a narrative explanation is provided, summarizing the rationale informed by evidence and panel discussions. All panel members were invited to review the proposed changes and submit feedback. Feedback received from the ERNICA-wide stakeholder review was discussed during a final online meeting. All major comments were reviewed, and panel members deliberated collectively to reach final conclusions. Where appropriate, recommendations were adjusted based on the discussion.

3 | RESULTS

3.1 | Results of prioritization process and topics selected for the revision

The prioritization exercise identified the sections with the highest scores for potential revision as follows: post-operative management (19.3%), poor functional outcome (19.0%), diagnosis (18.8%), operative management (15.9%), follow-up (15.9%), and Hirschsprung's associated enterocolitis (HAEC) (15.5%). For these highest-scoring domains, the panel formulated clinical questions based on the most frequently raised free-text comments, excluding comments that were mentioned only once. These draft questions were discussed and, when deemed necessary, adapted during an online meeting with all panel

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TABLE 2 Recommendations for who should operate on patients with HSCR.

Recommendations	Level of evidence and strength of recommendation	Revision status
Patients with HSCR should undergo pull-through surgery in centers with at least two pediatric colorectal surgeons and pathological, radiological, and anesthetic expertise, including pediatric and neonatal intensive care and specialized nursing 24/7.[28,29] • Concentration of interdisciplinary experience is associated with better outcomes in complex or rare pediatric surgical conditions.[30–33] • Accurate primary assessment of the disease phenotype in HSCR permits appropriate surgical management.[33] • The need for redo surgery is low in centers that regularly manage HSCR, and complications are appropriately identified and managed.[2,28,34–37]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
Centers performing pull-through surgery for HSCR should have the capabilities to manage the entire care pathway.[28] • This includes management of surgical complications and primary surgical management of all forms of HSCR, provision of multidisciplinary care until adulthood, and specialist nursing.[33–35,38,39] • Ability to deliver comprehensive follow-up until adulthood, including provision of transition of care.[33,35]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
Centers that operate on HSCR patients should demonstrate active involvement in quality control and improvement.[28,29] • Maintaining prospective registries permits assessment and monitoring of case volumes and outcomes.[33] • Commitment to training surgeons, pathologists, and nurse practitioners in diagnostics and management of HSCR ensures continuity of local expertise.[33] • Up-to-date care practices and understanding of the disease process through networking and participation in continued medical and surgical education.[33] • Information should be given about the availability of patient support organizations as early as possible, and patients should be informed about the availability of current guidelines.[28,33]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged

Abbreviation: HSCR, Hirschsprung's disease.

members, after which a voting session was conducted using Mentimeter. The clinical questions selected for full review and potential inclusion in the updated guideline were:

- What type of histopathological staining is most appropriate in the diagnosis of HSCR? (section *diagnosis*; two new literature sources included) (see Supporting Information S1: Data 4 and 5).
- Should a pre-operative urinary tract assessment be advised in all patients with HSCR? (section *preoperative care*; one new literature source included) (see Supporting Information S1: Data 6).
- What best practices in pre-operative care regarding the optimization of nutritional status can be identified in patients with HSCR? (section *preoperative care*; two new literature sources included) (see Supporting Information S1: Data 7).
- Should (a) anastomotic calibrations and (b) routine dilatations by the surgeon be advised after pull-through surgery for HSCR? If yes, what are the most important considerations regarding Hegar size, pain management, and anxiety management? (section *postoperative care;* respectively, five and three new literature sources included) (see Supporting Information S1: Data 8–11).
- What is the role of dietary habits in the self-management of bowel function during follow-up in patients with HSCR? (section *follow-up*; five new literature sources included) (see Supporting Information S1: Data 12).
- How can sexual and fertility problems in patients with HSCR be addressed during follow-up? (section *follow-up*; four new literature sources included) (see Supporting Information S1: Data 13).
- What are the indications for treatment with botulinum toxin in patients with HSCR presenting with HAEC or obstructive symptoms? (section *HAEC*; two new literature sources included) (see Supporting Information S1: Data 14).
- Should biofeedback be advised in patients with HSCR and poor functional outcomes? (section *poor functional outcomes*; two new literature sources included) (see Supporting Information S1: Data 15 and 16).
- What is the optimal design for transition of care in patients with HSCR? (section *follow-up*; two new literature sources included) (see Supporting Information S1: Data 17).

In the following sections, only new, modified, or eliminated recommendations are discussed. These can also be identified in the section-specific tables, where they are marked as "new" or "modified." Recommendations that were not reviewed or changed are not discussed further.

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TABLE 3 Recommendations for preoperative care.

Recommendations	Level of evidence and strength of recommendation	Revision status
Patients should receive saline rectal irrigations 1–3 times per day to decompress the bowel until the definitive pull-through operation. • An additional colonic wash-out may be given for pre-operative bowel preparation.[2] • See below, if there is an inadequate response to rectal irrigations.	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
A stoma is indicated if rectal irrigations do not sufficiently decompress the bowel, or there are complications such as enterocolitis unresponsive to non-operative treatment, or bowel perforation.[2,40] • The safest empiric level is an ileostomy.[41] • In pneumoperitoneum, an ileostomy is also provided, if it is proximal to the site of perforation.[41] • A representative circumferential "doughnut" biopsy taken from the site of stoma formation is informative regarding the ganglionic status of the bowel at that level.[41]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
When possible, a pre-operative contrast enema is recommended to guide the likely level of aganglionosis. • A colonic caliber change suggests a histological transition zone at this level.[42,43] • Proximal to the rectosigmoid junction, colonic caliber changes are less accurate in predicting the disease level, and the possibility of long-segment HSCR should be considered.[42] • Contrast studies are complementary tools during the pre-operative workup. They do not replace the need for histological assessment to confirm the diagnosis.[42]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged
At pull-through surgery, one dose of broad-spectrum intravenous antibiotics should be given preoperatively. • The choice of antibiotics is determined by local regimens and regional resistance profiles, but should include coverage of both aerobic and anaerobic bacteria.[44,45] • No additional benefit has been shown for giving more than one pre-operative dose, but antibiotics may be continued for 24–48 h post-operatively.[44,45]	Level of evidence II–III. Strength of recommendation: Strong, for	Unchanged
The panel suggests that routine screening for CAKUT should be performed within the first year of life for all HSCR patients.[46] • **Ultrasound should be the primary screening tool.**	Level of evidence II. Strength of recommendation: Conditional, for	**New**
The panel recommends that malnutrition screening should be performed preoperatively in HSCR patients.[47] • **Growth should be monitored as the primary indicator of nutritional adequacy.** • **No deviation from standard feeding recommendations is necessary for HSCR patients.** • **In cases of deviating growth, targeted preoperative nutritional support should be initiated.**	Level of evidence II. Strength of recommendation: Strong, for	**New**

Abbreviations: CAKUT, congenital anomalies of the kidney and urinary tract; HSCR, Hirschsprung's disease.

3.2 | Recommendations for the diagnosis of HSCR (Table 1)

What type of histopathological staining is most appropriate in the diagnosis of HSCR? (see Supporting Information S1: Data 4 and 5)

The presence of any number of ganglion cells on hematoxylin and eosin (H&E) staining excludes HSCR. However, because the effective application of H&E is dependent on a sufficient sample, and ganglion cells may be small and immature in neonates and premature infants, further histologic evaluation with the addition of ancillary studies is required if ganglion cells are not identified. A combination of H&E and calretinin immunohistochemistry is recommended to improve diagnostic accuracy, particularly in challenging samples. Calretinin offers higher specificity compared to other stains[23]; however, its availability may be limited in some centers. In HSCR, acetylcholinesterase (AChE) is expressed in extrinsic cholinergic nerves, mostly in the mucosa (positive stain), while calretinin staining is negative (except within 1–2 cm of the transition zone).

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TABLE 4 Recommendations for operative management.

Recommendations	Level of evidence and strength of recommendation	Revision status
Centers should perform the type of pull-through in which they have the most experience, including management of post-operative complications and follow-up.[52–54] • Transanal ERP, including laparoscopy-assisted ERP and Duhamel pull-through, represent the most commonly performed operations.[43,55–59] Currently, there is no evidence for overall superiority of one method over another in terms of surgical complications or long-term bowel function.[54,60–67]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged
The pull-through operation should be performed when the patient is stable and growing well, and the bowel has been sufficiently decompressed. • Elective pull-through within 2–3 months after diagnosis is usual.[39,68–70] • No specific advantages have been identified for performing pull-through surgery during the neonatal period.[39,68–70] • Anesthetic considerations, clinical and nutritional status of the patient, parental concerns, and surgical risks/technical feasibility influence the timing of pull-through surgery.[39,68,69]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged
The anal canal must be preserved during pull-through surgery (Figure 1).[51] • The transitional mucosa above the dentate line contains the nerve endings responsible for the reflex arc in sensation and fecal continence, including the sampling reflex.[71] • Transanal dissection should be commenced 0.5–2 cm proximal to the dentate line[72–75] **to ensure it is consistently performed above the anorectal line.** • In endorectal pull-through, either no muscle cuff (Swenson) or a short muscle cuff (<2–3 cm) has comparable outcomes, but long seromuscular cuffs should be avoided.[51,68,71,74,76–79]	Level of evidence II–III. Strength of recommendation: Strong, for	**Modified**
The colon should be transected at least 5–10 cm proximal to the first normal biopsy to minimize the risk of a transition zone pull-through.[80] • If the level of disease remains intraoperatively uncertain, "mapping" biopsies should be obtained from different colonic levels.[42,81] • Intraoperatively, fresh frozen sections are a valid means for determining the presence of normal ganglionated bowel, but single samples may miss asymmetrical histologic extension of the transition zone.[71] • A circular "doughnut" biopsy from the level of transection permits circumferential (four-quadrant) optimal histologic assessment.[82] • If feasible, any abnormally dilated colon proximally should be resected to avoid a transition zone pull-through. • At the end of the operation, the resected specimen should be sent in full (marked oral to anal) to the pathologist.	Level of evidence III. Strength of recommendation: Strong, for	Unchanged

Abbreviation: ERP, endorectal pull-through.

3.3 | Recommendations for who should operate on patients with HSCR (Table 2)

The original recommendations pertaining to this section were found to remain valid and have been maintained without modification.

3.4 | Recommendations for preoperative care (Table 3)

Should a pre-operative urinary tract assessment be advised in all patients with HSCR? (see Supporting Information S1: Data 6)

Recent studies have demonstrated that congenital anomalies of the kidney and urinary tract (CAKUT) are present in approximately 22% of patients with HSCR, with around 7% requiring medical or surgical intervention. The most commonly diagnosed CAKUTs were kidney dysplasia or hypoplasia, hydronephrosis, and vesicoureteric reflux.[46,48] Early detection of renal abnormalities allows for timely monitoring and management; however, the consistency of prenatal screening varies between centers, and prenatal findings are often unavailable for review, particularly when patients are transferred for surgical care. On the other hand, in settings where prenatal assessment is thorough and reliable, additional postnatal screening may be unnecessary and could potentially contribute to parental anxiety. Nevertheless, given that CAKUT is often asymptomatic initially and symptoms may develop over time, early postnatal detection remains critical.

Consensus was reached that routine ultrasound screening is recommended for all patients with HSCR. Although optimal timing has not been firmly established, screening

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within the first year of life is recommended to facilitate early diagnosis while allowing flexibility based on local healthcare resources. Ultrasound is proposed as the preferred screening modality, being both widely accepted by clinicians and parents and effective for detecting urinary tract anomalies. Cases in which abnormalities are detected should be referred promptly for specialized urological evaluation.

In this context, the recommendation to perform ultrasound screening within the first year of life reflects a balance between the well-established association between HSCR and CAKUT and the current limitations of the evidence base. Although many renal anomalies are clinically silent, available data do not support a more precise age threshold or demonstrate improved outcomes with earlier screening. The panel therefore adopted a pragmatic and flexible timeframe, acknowledging substantial variation in clinical practice across centers. The intent of this recommendation is to support early identification and appropriate referral for monitoring, rather than to address detailed urological management, which lies beyond the scope of this guideline.

The panel highlighted the need for cardiovascular screening in all HSCR patients, as studies report a high incidence of cardiac comorbidities (6%–10%). However, current evidence only describes the presence of anomalies without specifying how many patients required treatment prior to any surgical intervention.[47,49,50] This knowledge gap underlines the necessity for future research efforts.

What best practices in pre-operative care regarding the optimization of nutritional status can be identified in patients with HSCR? (see Supporting Information S1: Data 7)

Nutritional optimization is a key component of pre-operative care in patients with Hirschsprung disease (HSCR), as undernutrition has been associated with adverse post-operative outcomes, including HAEC. Evidence in this area is limited, and only one multicenter study met the inclusion criteria for this guideline. In that study, moderate to severe undernutrition was present in approximately 16% of children at admission, and lower weight-for-age *Z* scores were independently associated with postoperative HAEC, particularly in patients with longer aganglionic segments and delayed diagnosis.[51]

Based on the available evidence, the panel recommends routine assessment of growth parameters at hospital admission using standardized anthropometry. Rather than reliance on a single metric, a broader evaluation including weight-for-age (WAZ), height-for-age (HAZ), and clinical assessment of growth trajectory is advised. In patients with comorbid conditions known to affect growth, such as Down syndrome, the use of condition-specific

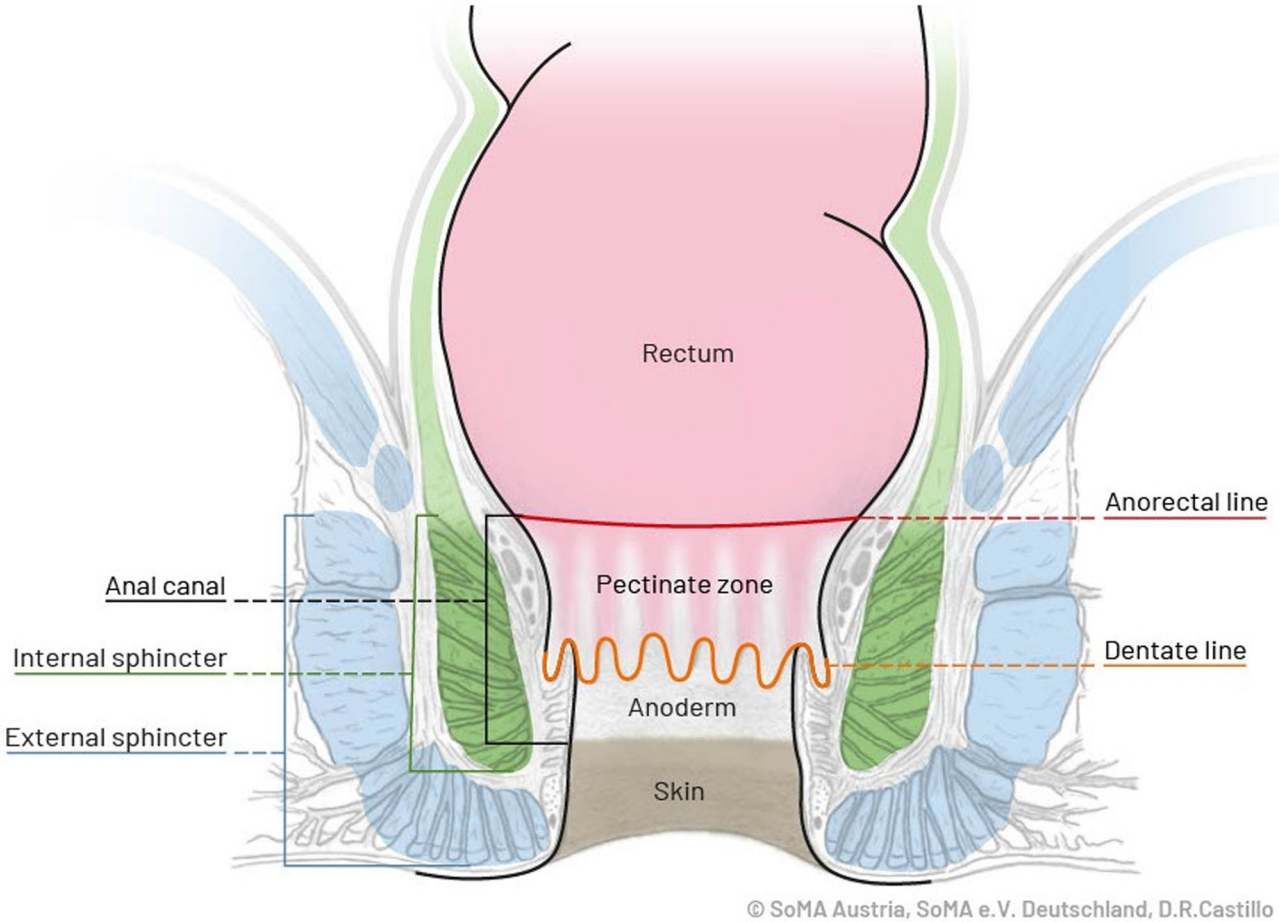

FIGURE 1 Anatomy of the anal canal.

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TABLE 5 Recommendations for early postoperative management after pull-through surgery.

Recommendations	Level of evidence and strength of recommendation	Revision status
Patients should receive specialist pediatric and nursing care during the early post-operative period, and anesthetic consultation should be available on request. • Use of ERAS principles may reduce length of stay, requirement for narcotic analgesia, and time to full enteral feeds.[84] • Parental counseling is important to ensure understanding and engagement with the care plan.[85] • Once bowel movements begin, perianal rash/skin excoriation is initially common and requires pre-emptive nursing.[65,86]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged
Enteral feeding can be started gradually when the patient has recovered from anesthesia and is clinically stable. • Within 24–48 h in most cases.[84] • Advance feeds as tolerated to normal diet.[84] • There is no evidence to suggest prolonged nil by mouth periods or prevent anastomotic complications.[84]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged
The panel recommends that calibration of the coloanal anastomosis should be performed after pull-through surgery to assess for strictures.[64] • **Calibration should preferably be done by a surgeon at a pediatric surgical center**.[87,88] • **No specific timing or Hegar size should be mandated, and the clinical picture should be leading in this decision**.[87,88]	Level of evidence III. Strength of recommendation: Conditional, for	**Modified**

Abbreviation: ERAS, Enhanced Recovery After Surgery.

growth charts should be considered. At present, no HSCR-specific nutritional screening tool has been validated. Available options, therefore, include serial anthropometric measurements, clinical dietary assessment, and referral for dietetic evaluation when growth deviation is identified. In cases of impaired growth (e.g., WAZ ≤ −2), targeted preoperative nutritional support should be initiated with the aim of achieving adequate growth and an anabolic state prior to surgery.

The potential role of extensively hydrolyzed formula (EHF) in reducing nutritional risk and postoperative complications was acknowledged during the expert panel discussions. However, the panel emphasized that the current evidence base is insufficient to support this intervention in clinical practice. Concerns were also raised regarding the bitter and potentially less-tolerated taste of EHF, the frequent need for nasogastric tube feeding, and the possible risk of discouraging breastfeeding in young infants. Therefore, at present, there is no recommendation to deviate from standard nutritional practices, including breastfeeding and the use of standard infant formula. The panel nevertheless identified this as an area of emerging clinical interest that warrants further investigation.

3.5 | Recommendations for operative management (Table 4)

In the original version of the ERNICA guidelines for rectosigmoid HSCR, the transanal dissection was described as being placed 0.5–2 cm above the dentate line. Although no formal recommendations from this section were selected for revision, an important concern was raised by members of the panel regarding the anatomical terminology used to describe the intraoperative resection line. Specifically, it was questioned whether the dentate line is the most appropriate reference point when describing the dissection plane. The term dentate line could be confusing, as it represents a two-dimensional structure, whereas the pectinate zone is three-dimensional. In children, due to the shortness and shallowness of the columnae anales, details of this area can be difficult to identify.

In the original version of the ERNICA guidelines for rectosigmoid HSCR, the transanal dissection was described as being placed 0.5–2 cm above the dentate line. However, this distance can vary depending on the size and anatomy of the patient. Because of this, the anorectal line, defined as the oral end of the anal columns, may offer a more anatomically accurate landmark.[51] Crucially, in all cases, the anastomosis must be constructed above the anorectal line to ensure preservation of the anal canal and the entire pectinate zone. From the anorectal line downward, all structures must be preserved, and the final anastomosis line must be positioned above the anorectal line as well. [Correction added on 22 May 2026, after first online publication. In the sentence beginning "In the original version of the ERNICA guidelines for rectosigmoid HSCR…" on page 193, reference citation 83 was has been corrected to 51.]

Therefore, the updated guideline includes the following illustration (Figure 1) to better clarify the relevant anatomical landmarks. Finally, it was agreed that further research is needed to better understand the anatomical factors influencing postoperative bowel function. Additionally, although this recommendation was not among those prioritized for revision according to the process described in the methods section, the extent of disagreement expressed during the discussion led to the formulation of a revised version. Upon request by a majority of the committee, the recommendation "*Transanal dissection should be commenced 0.5–2 cm proximal to the dentate line* ***to ensure it is consistently performed above the anorectal line***"

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TABLE 6 Recommendations for long-term follow-up.

Recommendations	Level of evidence and strength of recommendation	Revision status
Children with HSCR should receive regular follow-up to adulthood within the context of an interdisciplinary care team, led by a pediatric surgeon. • Follow-up should be more frequent during the first year of life, but regular contact should be maintained 1–2 yearly thereafter.[35,89–93] • Opportunities to engage multidisciplinary resources, including gastroenterologists, nutritional therapists, psychologists, physiotherapists, specialist nurses, and social workers should be available.[89,90,94–97] • Attention to development of wider areas of social functioning, including self-efficacy and coping skills, should be addressed.[89,90,94–96,98,99] • Growth, nutrition, and development should be followed.[89,90,100]	Level of evidence III. Strength of recommendation: Strong, for	**Modified**
The panel suggests providing access to a dietitian experienced in HSCR for individualized dietary advice.[83,101–103] • **No generalized dietary restrictions are advised due to lack of strong evidence.**[83,90,101,102] • **Special consideration should be given to patients with impaired bowel function.**[83,90,101,103,104]	Level of evidence II–III. Strength of recommendation: Conditional, for	**New**
The panel recommends that female patients with HSCR should be informed that their condition may affect fertility.[89,94,98,105,106] • **Evidence indicates problems specifically in females, but that doesn't exclude males completely from developing the issues.**[106,107]	Level of evidence II–III. Strength of recommendation: Strong, for	**New**
The panel suggests that sexual function concerns should be addressed during follow-up, with referrals offered in case of detected problems.[89,90,94–96,98,105–107] • **A multidisciplinary care approach should be taken, involving gynecologists, fertility specialists, and pelvic floor therapists.**	Level of evidence III. Strength of recommendation: Conditional, for	**New**
Access to care and specialist consultation should be available. • Patients should have a named surgeon in charge of their care and clear information about where and how they should attend follow-up, including in adulthood.[108,109] • Instructions for where to seek emergency care should also be clearly defined.[108] • Patient support organizations and networks are active in many countries for information and peer support on lived experience of the disease.[109]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged

Abbreviation: HSCR, Hirschsprung's disease.

was put forward for vote. All healthcare professionals and patient representatives were invited to vote within a 3-week period, with the understanding that lack of response would be considered agreement. Of the eligible voters, a total of 24 (74%) participants voted in favor, one (3%) voted against—preferring a formulation that more explicitly emphasized the anorectal line and the preservation of structures distal to it—and 8 (24%) did not respond. As a result, the recommendation was amended in accordance with the majority vote.

3.6 | Recommendations for early postoperative management after pull-through surgery (Table 5)

Should routine anastomotic calibrations by the surgeon be advised after pull-through surgery for HSCR? If yes, what are the most important considerations regarding Hegar size, pain management, and anxiety management? (see Supporting Information S1: Data 8 and 9)

In this update, the previous recommendation regarding calibration after pull-through surgery has been revised. There is currently no high-quality evidence to support specific recommendations on the indication, timing, frequency, or Hegar size for anal calibration after pull-through surgery, and therefore, the previous recommendation has been removed. Additionally, calibration and dilatation were previously discussed under the same subsection, despite being distinct procedures. To improve clarity, calibration is now addressed within the current section, while dilatations will be discussed separately under the section *poor functional outcomes*.

Calibration, referring to the measurement of anal canal and anastomotic diameter, should be routinely performed to detect anastomotic strictures. This helps ensure an adequate anal canal, which is essential for optimal functional outcomes. Strictures may develop late in the postoperative course,

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suggesting that the optimal timing for calibration is variable and should be tailored to the clinical context. However, no consensus could be reached on a specific postoperative timeframe or objective criteria defining an "adequate" anal canal diameter, as anal dimensions evolve with age and growth, and strictures may present late in the postoperative course.

Given these uncertainties, the timing and need for calibration should be individualized and guided by the clinical context rather than performed routinely. It is also important to recognize that calibration may sometimes require anesthesia, adding procedural risks and burden for the patient. Finally, calibration should be performed by an experienced surgeon to ensure accurate and reliable evaluation.

3.7 | Recommendations for long-term follow-up (Table 6)

What is the role of dietary habits in the self-management of bowel function during follow-up in patients with HSCR? (see Supporting Information S1: Data 12)

Studies show that up to 80% of patients report dietary influence on bowel function, with nearly 70% noting symptom improvement after dietary changes.[101,102] However, these findings are largely based on single-center studies and patient-reported outcomes, and it remains uncertain whether dietary effects in HSCR differ significantly from those in the general population. In the absence of structured dietary guidance, families often rely on trial-and-error strategies and online information, contributing to stress, uncertainty, and a sense of being unsupported.[102,103] Specific foods, such as fruits and dietary fiber, are often perceived to improve stool consistency, while dairy, refined carbohydrates, legumes, high-fat foods, onions, and certain vegetables are frequently reported as symptom triggers.[101–103] These findings underscore the need for formal dietary screening and individualized guidance within HSCR care.

Recent studies further emphasize that dietary recommendations should not be generalized. High-fiber diets, while commonly used for constipation, may not be appropriate for all HSCR patients and may even worsen symptoms in some. Structured elimination diets, developed in collaboration with dietitians, may help identify individual trigger foods and optimize symptom control.[102,104] Moreover, dietary restrictions can negatively affect social participation and emotional well-being, highlighting the importance of psychological support as part of multidisciplinary care.[89,101,103] The panel also noted that bowel dysfunction often persists into adolescence and adulthood, reinforcing the need for long-term dietary monitoring.[89] The panel also suggests the integration of routine dietary assessments into HSCR follow-up care and recommends access to dietitians with specific expertise in HSCR. This individualized approach helps avoid unnecessary dietary restrictions and supports targeted symptom relief.[101,102,104] Future research should focus on developing standardized dietary protocols, investigating the role of probiotics, and conducting prospective trials, such as low Fermentable Oligo-, Di- and Monosaccharides, And Polyols (FODMAP) structured elimination diets, to evaluate their effect on gastrointestinal symptoms in HSCR.[102]

How to address sexual and fertility problems in patients with HSCR during follow-up? (see Supporting Information S1: Data 13)

Fertility concerns are increasingly recognized in the long-term follow-up of patients with HSCR, particularly among women. Available evidence, primarily derived from large cohort studies, indicates that female HSCR patients have significantly lower fertility rates compared to age-matched controls, are older at first childbirth, and more frequently have only one child.[105–107] These findings persist even after adjusting for confounding factors such as educational level and associated malformations. While the precise mechanisms remain unclear, hypotheses include pelvic adhesions, altered pelvic anatomy, chronic bowel dysfunction, and possible psychological factors. While several potential mechanisms have been hypothesized in the literature—including pelvic adhesions, altered pelvic anatomy, chronic bowel dysfunction, and psychological factors—there are currently no studies that systematically investigate the underlying causes of reduced fertility in patients with HSCR.

Although male HSCR patients do not show statistically significant reductions in fertility rates compared to controls, they should not be excluded from fertility discussions. Isolated cases of subfertility have been noted, and bowel dysfunction has been linked to poorer reproductive outcomes in men as well.

The panel emphasizes that fertility implications should be communicated early in care, ideally during adolescence or early adulthood, to support informed family planning. Patients experiencing delays in conception should be referred for evaluation, including gynecological work-up when appropriate, and have access to fertility specialists. Exploration of surgical strategies to minimize adhesions may also contribute to improving long-term reproductive outcomes. In this context, laparoscopic techniques may help preserve fertility by reducing postoperative adhesion formation.

Sexual function represents a critical yet under-addressed component of care for patients with HSCR. Among female patients, high rates of sexual dysfunction, including dyspareunia and sexual distress, have been reported, particularly in those with persistent bowel symptoms.[105] Up to 50% of sexually active women with HSCR report pain during intercourse, and studies consistently show lower sexual quality of life scores compared to controls.[105–107] These outcomes are strongly linked to poor bowel control, negative body image, and reduced self-esteem, factors that often emerge in adolescence and predict adult sexual function.[105–107] While male HSCR patients generally report normal sexual function, occasional issues such as erectile dysfunction, retrograde ejaculation, or anorgasmia have been documented.[107,110] Importantly, psychosexual wellbeing and relationship development during adolescence are predictive of adult sexual satisfaction in both sexes.[105,110]

Despite its importance, sexual health is often overlooked in clinical follow-up. Many pediatric surgeons feel unprepared to initiate these conversations, and access to sexual health professionals varies widely. In addition, panel members highlighted that cultural, religious, and societal norms across Europe influence how and by whom sexual health is discussed in clinical practice. In some settings, surgeons expressed hesitancy to address these topics directly, instead emphasizing the role of psychologists, gynecologists, and sexologists to ensure a more holistic and culturally sensitive approach to patient education. The panel underscores the need for structured, multidisciplinary support, including

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TABLE 7 Recommendations for HAEC.

Recommendations	Level of evidence and strength of recommendation	Revision status
HAEC should be clinically suspected in the presence of diarrhea with explosive, foul-smelling stool, and/or >4 points from the Pastor et al. *HAEC score* items (Table 8). • The definition of HAEC remains imprecise even based on current understanding.[111–113] • A cut-off of >10 points has a reported sensitivity of 42% and specificity of 100% for HAEC.[112] • A cut-off of >4 points has a reported sensitivity of 84% and specificity of 98% for HAEC.[112]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
In suspected HAEC, there should be a low threshold for hospital admission. • In mild symptoms with no fluid or electrolyte balance disturbance and normal inflammatory markers, outpatient treatment with oral hydration +/– oral metronidazole and rectal irrigations may be appropriate, but prompt admission is indicated if symptoms do not improve. Recovery should be followed up.[112] • Admit all other cases for inpatient monitoring and treatment.[112] • Young age (<1 year) lowers the threshold for admission.[112]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
Following admission to hospital, patients with HAEC should be treated with intravenous fluid resuscitation, intravenous broad-spectrum antibiotics, and rectal washouts.[114] • Saline rectal washouts to decompress the bowel should be performed 2–3 times per day until the patient is well enough for discharge.[112,115] • Antibiotics may be changed to oral metronidazole once sufficient clinical improvement occurs.[112] • Vital functions, fluid and electrolyte balance, including urine output, should be closely monitored.[112] • Abdominal plain film x-ray should be considered.[112] • Consulting the colorectal surgical team responsible for the patient's care is recommended.[112] • Consult intensive care unit as appropriate.[112]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
Intersphincteric botulinum toxin injections are recommended for patients with recurrent or persistent symptoms of outlet obstruction and/or HAEC.[41] • In reports, 62%–89% of HSCR patients with HAEC and/or outlet obstruction improved after the first botulinum toxin injection.[116–120] • Injections may need to be repeated 3–6 monthly.[119,121] • The tendency to HAEC reduces over time; most episodes usually occur within the first few years after pull-through.[35] Principles of botulinum toxin administration. • Botox should be injected under a short general anesthesia.[116,122] • The patient is positioned in the lateral decubitus or lithotomy position.[122] • Injections are given in the four quadrants at the level of the dentate line into the anal sphincter musculature.[122] • Exposure of the dentate line with retractors, and/or ultrasound guidance can facilitate correct localization of the injections.[116,122]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged
The panel does not recommend the use of botulinum toxin in the prevention of HAEC.[110,123]	Level of evidence II–III. Strength of recommendation: Conditional, against	**New**
Prophylactic antibiotics may be considered for patients with frequently recurring or persistent HAEC.[124] • Antibiotics may be effective treatment of HAEC in individual patients, but it has not been shown that prophylactic antibiotics prevent recurrent HAEC.[124] • Recurrent courses of antibiotics interfere with the long-term composition of the gut microbiota, and therefore, rationalized use based on severity of symptoms is indicated.[124]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged

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TABLE 7 (Continued)

Recommendations	Level of evidence and strength of recommendation	Revision status
At present, there is insufficient evidence to support recommending the routine use of probiotics for the prevention of HAEC. • Although intestinal dysbiosis has been shown to be of importance in the etiology of HAEC, there are only two randomized controlled studies of probiotics and HAEC in the literature, showing conflicting results.[113,124–130]	Level of evidence I–III. Strength of recommendation: Conditional, against	Unchanged
In children with recurrent HAEC, consultation with a gastroenterologist and endoscopy should be considered.[114] • Patients with HSCR have an increased risk of developing inflammatory bowel disease.[131] • Fecal calprotectin is a non-invasive measure of intestinal inflammation in acute and chronic enterocolitis.[131]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged

Abbreviations: HAEC, Hirschsprung's-associated enterocolitis; HSCR, Hirschsprung's disease.

TABLE 8 Pastor et al.[111] HAEC score items.

Category	Item	Score
History		
	Diarrhea with explosive stool	2
	Diarrhea with foul-smelling stool	2
	Diarrhea with bloody stool	1
	Previous history of enterocolitis	1
Physical examination		
	Explosive discharge of gas and stool on rectal examination	2
	Distended abdomen	2
	Decreased peripheral perfusion	1
	Lethargy	1
	Fever	1
Radiologic examination		
	Multiple air fluid levels	1
	Dilated loops of bowel	1
	Sawtooth appearance with irregular mucosal lining	1
	Cut-off sign in rectosigmoid region with absence of distal air	1
	Pneumatosis	1
Laboratory		
	Leukocytosis	1
	Shift to left	1

Note: Using a cut-off of >4 points to identify suspected HAEC is more sensitive and may have better clinical applicability than Pastor and co-workers' original cut-off of >10 points.[100]

Abbreviation: HAEC, Hirschsprung's-associated enterocolitis.

pediatric surgery, gynecology, urology, psychology, and pelvic floor therapy, to address sexual function and body image concerns.[105,106,110] Sexual health education and psychosocial support should be integrated into adolescent care to foster confidence, address distress, and improve long-term outcomes.[107] As an expertise network, ERNICA can play a key role in pooling and disseminating educational resources to support this aspect of care across centers.

3.8 | Recommendations for HAEC (Tables 7 and 8)

What are the indications for botulinum toxin treatment in HSCR patients with persistent obstructive symptoms or recurrent HAEC? (see Supporting Information S1: Data 14)

Botulinum toxin (Botox) is used in the management of HSCR for children who experience persistent obstructive symptoms after pull-through surgery in the absence of mechanical causes, and for those with recurrent or ongoing symptoms of outlet obstruction and/or HAEC. Clinical studies report symptom relief in approximately 66%–71% of treated patients. However, the absence of a clear dose–response relationship complicates its use, and no standardized dosage can be recommended. In addition, botulinum toxin treatment carries a risk of adverse effects in around 17% of patients, including transient fecal incontinence, anal pain, and muscle fatigue.

Despite its therapeutic potential, the routine *preventive* use of botulinum toxin, particularly at the time of surgery or shortly afterwards, has not been shown to reduce the incidence or delay the onset of HAEC.[123,132] Current studies suggest that early botulinum toxin administration does not significantly alter long-term outcomes, and in some cases, may be associated with earlier HAEC onset or the need for additional botulinum toxin treatments. While the frequency of botulinum toxin use has increased across centers in recent years, the evidence does not support its use as a standard preventive strategy in asymptomatic patients or those without HAEC.

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TABLE 9 Recommendations on the management of patients with poor outcomes.

Recommendations	Level of evidence and strength of recommendation	Revision status
The panel strongly recommends against routine preventive serial dilatations after pull-through surgery in HSCR patients.[87,132] • **Dilatation remains an option for treating diagnosed strictures but should not be used preventively.**[87,132–134] • **When dilatation is indicated, a low threshold for performing the procedure under general anesthesia is advised.**[88] • Patient and family perspectives should always be considered in decision-making.[88]	Level of evidence III. Strength of recommendation: Strong, against	**New**
Children with normal intellectual development who are not continent of stool by 4 years of age should be considered for further evaluation. This should include: • A stooling history and stooling pattern to evaluate for tendency to constipation or diarrhea (for treatment, see below), and involuntary passage of flatus.[135,136] • Dietary history and growth.[135,136] • Examination under anesthesia +/– anorectal manometry to assess the integrity of the anal canal, sphincter complex, and dentate line, and for the presence of rolled muscle cuff, stricture, or rectal spur.[68,137] • Contrast enema to evaluate whether there is colonic dilatation, rectal spur, constipation, or a twisted pull-through.[137] • +/– Endorectal ultrasound to assess for sphincter defects.[137]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
The management of fecal incontinence should aim for age-appropriate continence in children with normal intellectual development.[138] • Primary prevention of the social consequences of fecal incontinence is a key goal of treatment.[96] • Enabling normal social integration, school attendance, and ability to participate in recreational activities from the outset is important for self-esteem, friendships, and long-term quality of life.[96] • Deficient fecal continence in a child is also a source of stress for caregivers, and psychological support should be available for patients and families.[96,139] • Cognitive impairment is associated with delays in achieving voluntary bowel control.[96,135]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged
Patients with an intact anal canal and appropriate pull-through but fecal incontinence should receive medical management as the first-line treatment. • For patients with a dilated colon and constipation (hypomotility), oral laxatives +/– a short course of enemas to ensure regular and complete colonic emptying.[140] • For patients without colonic dilatation and a tendency to loose stools (hypermotility), a constipating diet +/– loperamide +/– bulking agents (pectin, psyllium).[140] • Measure fecal calprotectin, consider ileo-colonoscopy, and repeat rectal biopsy.[140] • Proceed to bowel management if there is failure to respond, despite adequate dosing and compliance.[140]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged
Patients with fecal incontinence and damaged anal canal should receive bowel management.[138] • Maintaining an intact anal canal is a central goal in all standard operations for HSCR, and an indication for performing pull-through surgery in specialist units.[140] • An enterostomy is an option if bowel management fails to control symptoms.[140]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged
Children with persistent obstructive symptoms following pull-through surgery should undergo further evaluation and treatment[141,142]: • Rectal examination and contrast enema to rule out a mechanical cause and to assess for colonic dilatation.[143–148] • If no mechanical cause is found, a trial of intersphincteric botulinum toxin injections.[140,145]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged

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TABLE 9 (Continued)

Recommendations	Level of evidence and strength of recommendation	Revision status
• Review the histology of the proximal margins of the originally resected bowel.[82,143] • Repeat rectal biopsies to ensure normal innervation of the pulled-through bowel.[143,148] • If repeated botulinum toxin injections are ineffective, histology is normal, and there is no mechanical cause, bowel management can be offered.[140] • Consider redo surgery in patients with a recalcitrant stricture, twisted pull-through, rolled muscle cuff (Soave), rectal spur (Duhamel), or transition zone pull-through.[142,145,147]		
Bowel management program should comprise individualized care based on the symptom profile, local recommendations, and values/preferences of the patient/carer(s). • The goal of bowel management is to achieve regular and complete colonic emptying at predictable intervals.[140] • Options include regular retrograde enemas or antegrade colonic irrigation via an ACE appendicostomy or cecostomy +/– dietary modifications +/– laxatives.[140,149] • Psychological support can assist patients and families in coping with symptoms.[96] • An enterostomy may be required in isolated cases for intractable symptoms.[140]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged

Abbreviations: ACE, antegrade continence enema; HSCR, Hirschsprung's disease.

3.9 | Recommendations on the management of patients with poor outcomes (Table 9)

Should routine dilatations by the surgeon be advised after pull-through surgery for HSCR? If yes, what are the most important considerations regarding Hegar size, pain management, and anxiety management? (see Supporting Information S1: Data 10 and 11)

Routine preventive dilatations after pull-through surgery in HSCR patients are not recommended, as current evidence does not support their effectiveness in preventing stenosis, HAEC, or constipation.[87,88,133,134] Moreover, these procedures may be associated with harm: they are often painful, emotionally distressing for both patients and parents, and may contribute to psychological trauma.[133,134] Aligning care by discouraging routine preventive dilatations may also reduce unwarranted variation and promote equity across centers.

It is essential, however, to distinguish between diagnostic calibrations (see above), prophylactic (or preventive) dilatations, and therapeutic dilatations. In the presence of a diagnosed anastomotic stricture, dilatation remains a valid treatment approach prior to considering more invasive surgical interventions. In such cases, serial dilatations may be attempted, with a low threshold for general anesthesia to minimize discomfort and trauma.[87,88,133,134] Decision-making should be guided by shared discussions that respect the perspectives of both patients and families.

Should biofeedback be advised in patients with HSCR and poor functional outcomes? (see Supporting Information S1: Data 15 and 16)

Biofeedback is not currently recommended in clinical guidelines for HSCR patients with persistent incontinence or poor functional outcomes, largely due to limited and indirect evidence. Most studies evaluate manometric improvements, which are not consistently linked to clinical symptom relief. Although some expert group members report selective use of biofeedback, its long-term effectiveness remains uncertain.[150] Additionally, the invasive nature of rectal probe-based biofeedback raises concerns, particularly for pediatric patients.

In contrast, pelvic floor physical therapy (PFPT) has emerged as a promising, non-invasive alternative and is typically delivered by trained pediatric physiotherapists. PFPT encompasses a range of techniques aimed at improving pelvic floor coordination, relaxation, and strength, together with abdominal core muscle training; in the referenced study, this included surface electromyography–based biofeedback using external electrodes to provide real-time, child-friendly feedback (e.g., video game–style interfaces), alongside prescribed home exercises to reinforce learning between sessions. PFPT has demonstrated significant reductions in incontinence, with one study showing improvement from 81.1% to 40.5% ($p = 0.001$).[151] However, PFPT represents a heterogeneous intervention, with treatment content, number of sessions, and use of biofeedback varying according to patient age, cooperation, symptom profile, and local expertise. This heterogeneity, together with barriers related to access, scheduling, and financial constraints, currently limits standardization of PFPT and complicates comparison of outcomes across studies.[151]

Given its potential benefits and lower risk profile, PFPT should be considered early in the management of HSCR patients with functional impairments, particularly incontinence, before pursuing more invasive interventions such as rectal probe–based biofeedback. Ideally, PFPT should be applied hand-in-hand with psychotherapy as part of a multidisciplinary treatment approach, addressing both physical and psychological aspects of care.[94]

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TABLE 10 Recommendations for transition of care.

Recommendations	Level of evidence and strength of recommendation	Revision status
The panel recommends to have a defined and approved transition protocol in place that fits the local organization of care. • **Before transition, a disease-specific checklist should be checked to ensure that everything has at least been named during the transition of care talks: bowel inflammation, HAEC, continence, bowel function, urological problems, and sexual health.**[94] • **The ERNICA transition working group advises that local guidelines address disease-specific points for HSCR.** • **Some patients will not need follow-up after transition because they are doing fine. Address in these cases where they could go if problems would arise later.**	Level of evidence III. Strength of recommendation: Strong, for	**New**
The introduction to adult medical disciplines should be prepared well before transition.[94] • Discussions concerning long-term follow-up should be initiated around adolescence/puberty, and individualized care plans involving the appropriate disciplines should be formulated.[89,94,152] • Maintaining continuity and consistency of the health care into adulthood is very important to patients.[89,90,94,152] • Patients should be given sufficient information and increasingly engaged in decision-making regarding their healthcare with age.[90,94,109] The future care provider should be clearly identified, with an opportunity for staged transition and liaison with pediatric services during the transition phase.[152]	Level of evidence III. Strength of recommendation: Strong, for	Unchanged

Abbreviations: ERNICA, European Reference Network for rare Inherited and Congenital Digestive and Gastrointestinal Anomalies; HAEC, Hirschsprung's-associated enterocolitis; HSCR, Hirschsprung's disease.

3.10 | Recommendations for transition of care (Table 10)

What is the optimal design for transition of care in patients with HSCR? (see Supporting Information S1: Data 17)

Due to an increasing awareness of patients with HSCR reaching adulthood, the need for structured transition of care protocols has become more evident.[153,154] While current guidelines recommend early preparation, they lack detailed instructions regarding timing, essential topics, and the minimum set of involved stakeholders. Patients consistently emphasize the importance of continuity, clarity, and a structured process during transition of care, yet practices vary widely across centers and countries, with many lacking formal programs.[153] Although some patients may no longer require routine follow-up, it remains essential to inform them where to seek care should new or recurring issues arise. Patient organizations have strongly advocated for clearer guidance to avoid vague recommendations and to safeguard continuity of care.

The transition of care process should be tailored to local healthcare systems but follow a standardized framework.[14] Ideally, preparation should begin between ages 10 and 12 to allow adolescents to gradually build knowledge, independence, and confidence in managing their condition.[153] Practical topics such as bowel function, continence, the risk of bowel inflammation, urologic complications, and sexual health should be explicitly addressed.[153] Education about the disease, strategies for long-term health management, and lifestyle considerations should also be included, involving the patient's associations in the proper communication and feedback acquisition. Prior to completing transition of care, the pediatric surgeon should review a locally approved disease-specific checklist with the patient to ensure key topics have been addressed.[14] Even for patients who appear stable and may not require continued follow-up, the plan should clearly outline where and how to access care if complications develop in adulthood.[153]

3.11 | Recommendations for genetic screening (Table 11)

The original recommendations pertaining to this section were found to remain valid and have been maintained without modification.

3.12 | Evidence gaps and targets for further research

The panel has identified several key research priorities to guide progress in the field of HSCR over the next 5 years. Further development and evaluation of artificial intelligence-based tools for interpreting histopathological samples from rectal biopsies could enhance diagnostic accuracy. Cardiovascular anomalies, reported in 6%–7% of HSCR patients, call for cross-sectional studies to assess the potential need for routine preoperative screening. In the nutritional domain, there is a need to validate screening tools for assessment of malnutrition specific to HSCR and compare their effectiveness with existing World Health Organization (WHO) *Z* score criteria. Research should also clarify the optimal timing for anal canal calibration after pull-through surgery. The role of diet in managing bowel function remains underexplored, and studies are needed to assess the impact of specific dietary interventions on symptoms, the

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TABLE 11 Recommendations for genetic screening.

Recommendations	Level of evidence and strength of recommendation	Revision status
In non-syndromic HSCR, genetic testing of RET should be considered. • RET remains the major gene in HSCR.[155–162] • Molecular testing allows a more accurate estimation of the risk of recurrence.[155,156,158,159,161] • Genetic testing of RET allows exclusion of the rare possibility of MEN2A-associated RET mutation that is associated with an increased risk of medullary thyroid cancer.[155,163,164] • Parents or patients who wish to have genetic screening should be offered referral for genetic consultation.[155] • Genetic consultation is also recommended for patients with a family history of HSCR, where the incidence of RET mutations is even higher.[155,158,159,161]	Level of evidence II–III. Strength of recommendation: Conditional, for	Unchanged
In syndromic HSCR, patients should be offered referral for genetic consultation and screening for the specific gene associated with the syndromic phenotype.[155,162]	Level of evidence III. Strength of recommendation: Conditional, for	Unchanged

Abbreviations: HSCR, Hirschsprung's disease; RET, rearranged during transfection.

potential risk of developing HAEC, microbiome alterations, and quality of life. and quality of life. The ERNICA Nutrition and Transition (of care) Working Groups may play an important role in facilitating and supporting research in these areas. Given the prevalence of sexual dysfunction and impaired fertility, particularly in women, it is essential to evaluate whether structured screening tools improve early detection and management of these issues. Transition of care is another key focus, with a need to investigate how different models influence long-term health outcomes and address gaps in adult care, including persistent continence and other functional issues. The use of intraoperative botulinum toxin for HAEC prevention should be further explored in prospective studies. Additionally, there is an urgent need to standardize and validate the definition and severity grading of HAEC to improve clinical consistency. Finally, the long-term comparative effectiveness of biofeedback and pelvic floor physical therapy in treating functional impairments should be studied using validated patient-reported outcome measures.[150]

3.12.1 | Methodological gap

Finally, although this guideline update was developed within a multidisciplinary European network and included patient representatives and invited topic-specific experts, the panel composition predominantly reflected pediatric surgical expertise. Broader representation from other disciplines involved in HSCR care—including pediatric gastroenterology, pathology, urology, psychology, and nutrition—may further strengthen future guideline development, particularly for complex disease phenotypes, long-term functional outcomes, psychosocial aspects, and comorbidity management. Expanding multidisciplinary involvement may also support the development of more detailed recommendations on areas such as enterocolitis management beyond surgical decision-making, nutritional interventions, urinary and sexual health, and psychological support. Future guideline updates should therefore aim to actively facilitate a model of multidisciplinary collaboration within rare disease networks, to ensure balanced perspectives across the full spectrum of HSCR care.

3.13 | Implementation and monitoring

A central implementation support team (CIST) has been set up within ERNICA to support the successful implementation of ERNICA guideline recommendations in clinical practice. Deviations between the HSCR guideline recommendations and "current hospital-level practices" will be assessed to help prioritize and guide ongoing implementation efforts.

The European Pediatric Surgical Audit (EPSA) is a prospective, multinational clinical audit established in 2014 by the Dutch Association of Pediatric Surgeons to benchmark and improve the quality of care for rare congenital malformations, and was endorsed in 2019 by ERNICA to form the EPSA|ERNICA registry. The data set for HSCR is undergoing revision and will be aligned with the updated guideline recommendations where possible, enabling future monitoring of outcomes, quality of care indicators, and guideline adherence.

Providing performance feedback to clinicians, relative to defined standards, can be a very effective way to stimulate change and improve professional practice.[15] As an "audit and feedback" tool, the EPSA not only has the potential to monitor guideline adherence, but also to increase it across Europe.[16] Use of the EPSA for this purpose will be piloted as part of ERNICA's quality improvement system.[16] In addition to EPSA Audit & Feedback, other implementation strategies may be selected. To tailor such strategies, efforts will be made to understand the factors foreseen to hinder and/or facilitate successful implementation of the HSCR recommendations in practice.[16]

This updated guideline will undergo formal review for potential revision in 2030, or earlier if significant new evidence emerges. We intend to use EPSA data trends to inform the timing and scope of future updates.

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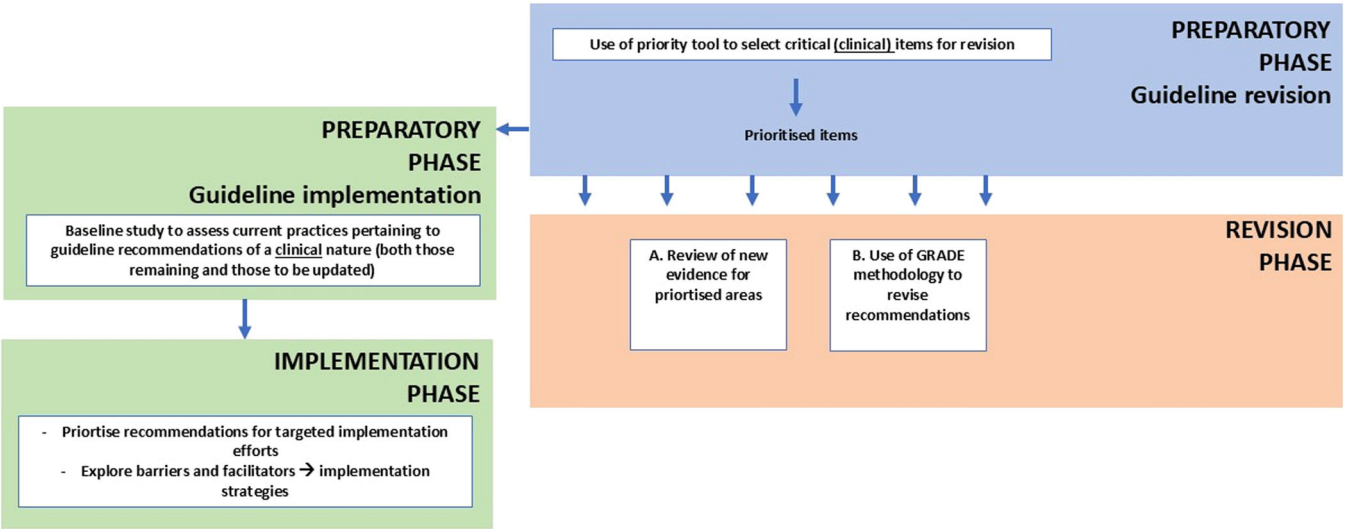

FIGURE 2 Overview of the revision process, including methodological workflow (right) and implementation planning (left). GRADE, Grading of Recommendations Assessment, Development and Evaluation.

4 | CONCLUSION

This updated version of the ERNICA guideline for recto-sigmoid HSCR reflects both advancements in clinical evidence and evolving priorities in patient-centered care. The systematic updating process and grounded methodology (Figure 2) allowed the panel to critically appraise new data and revise key recommendations to better support current practice. Importantly, this update addresses several previously under-represented aspects of care, including the role of nutritional optimization, individualized dietary advice, sexual health, fertility, and structured transition of care to adult services. The updated recommendations strengthen consistency, equity, and a holistic approach to care for patients with HSCR across the lifespan. In addition, the guideline explicitly highlights key evidence gaps, providing a clear direction for future research and further refinement of clinical practice.

Implementation of the updated recommendations will be supported through alignment with quality monitoring tools such as the EPSA, enabling benchmarking and continuous quality improvement across ERNICA centers. This work demonstrates that the ERNICA network remains committed to advancing the quality of care for patients with HSCR and to maintaining guidelines that are dynamic, trustworthy, and responsive to emerging clinical needs.

GLOSSARY

ACE – A surgical procedure that creates a small opening through which fluids can be used to flush the colon (used for bowel management).

Hirschsprung's disease – A birth condition where nerve cells are missing from part of the bowel, causing problems with passing stool.

Acetylcholinesterase – An enzyme found in the body that helps break down a chemical involved in nerve function; used in diagnosing nerve-related bowel conditions.

Aganglionosis – A condition where nerve cells (ganglia) are missing from part of the intestine.

Anastomosis – A surgical connection between two parts of the intestine.

Anastomotic calibration – A procedure to check the size of the connection made during bowel surgery to ensure it's not too narrow.

Anastomotic stricture – A narrowing at the site where two parts of the intestine were surgically joined.

Anorectal line – An anatomical marker used during surgery to preserve important bowel structures for continence.

Anorectal malformation – A condition where the anus and rectum do not develop properly.

Botulinum toxin – A medicine that relaxes muscles and is used to help children with bowel problems after surgery.

Bowel management – A treatment plan to help children empty their bowels regularly, often involving enemas or medications.

Calibration – A gentle measurement of the anal canal to check for narrowing or blockage after surgery.

Calprotectin – A protein found in stool that shows if there is inflammation in the intestine.

Calretinin – A special stain used in lab tests to detect nerve cells in tissue samples.

Colonic dilatation – An abnormal widening of the large intestine.

Colonic irrigation – A procedure that involves flushing the large intestine with liquid to help remove stool.

Constipation – Difficulty or delay in passing stool.

Contrast enema – An X-ray test where a liquid is put into the bowel to show its shape and structure.

Continence – The ability to control bowel movements.

Dilatation – A treatment to widen a narrowed area in the bowel.

Enterocolitis – An infection or inflammation of the intestines, common in children with Hirschsprung's disease.

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Enterostomy or stoma (colostomy, ileostomy) – A surgical opening made into the intestine that allows stool to leave the body into a bag.

Fecal incontinence – Inability to control bowel movements, leading to accidents.

Fertility – A person's ability to have children.

Intestinal dysbiosis – An imbalance in the bacteria living in the gut, which may cause digestive problems.

Manometry – A test that measures how well the muscles and nerves in the rectum and anus are working.

Obstructive symptoms – Signs that stool or gas cannot pass normally through the intestines, such as bloating or vomiting.

Pelvic floor physical therapy – Exercises and techniques to strengthen the muscles involved in bowel control.

Perianal rash – Skin irritation around the anus.

Peripherin – A protein used in lab tests to help identify nerve tissue.

Posterior sagittal anorectoplasty – A surgical method to correct anorectal malformations.

Probiotics – Live bacteria that can support gut health.

Pull-through surgery – The operation where the unhealthy part of the bowel is removed, and the healthy part is attached to the anus.

Rectal biopsy – A test that removes a small piece of tissue from the rectum to check for missing nerve cells.

Sexual dysfunction – Problems related to sexual activity, such as pain or difficulty with intercourse.

Sphincter complex – The muscles that help control bowel movements.

Transition of care – The process of moving from pediatric to adult healthcare services.

Transition zone – The area between normal bowel and bowel without nerve cells in Hirschsprung's disease.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

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