

**33<sup>rd</sup> International Meeting of the  
Pediatric  
Colorectal Club**

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**ABSTRACT BOOK**

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**17-19 MAY, 2026  
VIENNA, AUSTRIA**



## SHOULD PULL THROUGH BE OFFERED TO PATIENTS WITH CLOACAL EXSTROPHY?

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**PURPOSE:** Cloacal exstrophy is a rare and severe congenital anomaly impacting the gastrointestinal and genitourinary tracts, requiring complex multidisciplinary surgical reconstruction. Around the country, pull through is uncommon in patients with cloacal exstrophy with heterogeneous outcomes data, however it is frequently performed at our center. We aimed to characterize this population and their longitudinal outcomes.

**METHODS:** A review of a prospectively maintained database (1981-present) of all patients with cloacal exstrophy was conducted, noting demographics, associated anomalies, surgical history, and outcomes reported at most recent follow up.

**RESULTS:** There were 91 patients with cloacal exstrophy (67 classic, 24 covered) during the study period. Pull through, offered based on a trial of bowel management through the stoma, was performed in 58 patients (63.7%) at median age 2.46 years (IQR 1.44, 5.08). The most frequent reasons to not undergo pull through were failed stoma bowel management (42.4%) and patient/family preference (33.3%). After pull through, 80% of patients were clean for stool and 4/58 patients underwent reversal. Outcomes were reported in 40/58 patients, assessed at median 4.84 years post pull through (IQR 0.65, 10.69).

**CONCLUSION:** With preservation of colon and incorporation into the gastrointestinal tract at initial operation, followed by a successful bowel management through the stoma, 80% of patients with cloacal exstrophy who underwent pull through are clean for stool. As such, patients with cloacal exstrophy should be offered a pull through, ideally prior or simultaneously to bladder reconstruction.

## THE "FEED AND WRAP" MRI FOR PATIENTS WITH ANORECTAL MALFORMATIONS: DIAGNOSTIC VALUE AND LIMITATIONS

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**PURPOSE:** Magnetic resonance imaging (MRI) is essential in the preoperative assessment of patients with anorectal malformations (ARM). The "feed-and-wrap" (FW) technique allows MRI to be performed in infants without general anaesthesia (GA). This study evaluated the feasibility and diagnostic quality of FW-MRI in ARM patients compared with MRI performed under GA.

**METHODS:** ARM patients who underwent MRI using the FW technique between 2017 and 2020 were compared with a control group examined under GA. The groups were matched for age and sex. All examinations were performed on a 1.5-T MRI scanner. Image quality was assessed using a three-point scale across seven anatomical regions (3 = fully evaluable, 2 = evaluable with limitations, 1 = non-evaluable). Accuracy of fistula identification was evaluated using intraoperative findings as the reference standard.

The linear-by-linear association test was used to compare individual anatomical regions. Fisher's exact test was employed to compare the accuracy of fistula type classification. Agreement between MRI findings and operative findings was assessed using Cohen's kappa coefficient.

**RESULTS:** A total of 44 patients were included, of whom 75% were boys. Patients in the GA group were significantly older at the time of MRI (82 vs. 67.5 days;  $p = 0.030$ ). FW-MRI achieved a mean evaluability score of 2.64 across the seven anatomical regions. Although scores in the GA group were higher (3.00), the majority of FW patients (18/22) had sufficient evaluability (score  $>2$ ) in all regions. Four FW patients had at least one non-evaluable region, most commonly the spinal canal, vertebrae, or pelvic floor. Overall agreement between MRI and intraoperative findings for fistula identification was 91.3% ( $\kappa = 0.874$ ,  $p < 0.001$ ), with no significant difference between GA-MRI (92.3%) and FW-MRI (90.0%).

**CONCLUSION:** The "feed-and-wrap" technique provides adequate diagnostic quality for most infants with anorectal malformations. However, visualization of spinal structures may be limited, and MRI under general anaesthesia should be considered when spinal dysraphism is suspected on ultrasound scan.

## RADIOLOGICAL AND CLINICAL OUTCOMES FOLLOWING MULTICENTRE PROTOCOLISATION OF HIGH-PRESSURE COLOSTOGRAM FOR MALE ANORECTAL MALFORMATION

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**PURPOSE:** The impact of standardising high pressure colostogram (HPC) for assessment of male anorectal malformation (ARM) has not been investigated. Using a protocol measured against a validated 10-point criteria (HPC-10), this study evaluated the impact of protocolisation on the quality of radiology and clinically relevant adverse events resulting from HPC.

**METHODS:** Between 2023 and 2024, the HPC-10 protocol was implemented across three tertiary paediatric surgical centres. All patients who undertook HPC and completed subsequent anorectoplasty were evaluated, comparing retrospective (pre-implementation) and prospective (post-implementation) cohorts. Radiological quality was assessed using HPC-10 criteria, encompassing reporting adequacy (5 points), diagnostic accuracy (3 points), and technical adequacy (2 points). Adverse events included radiological complications, repeat studies (additional radiation), unplanned intraoperative changes, or procedure-related injuries. Continuous variables were represented as median (range) and analysed using the Mann–Whitney U-test. Logistic regression assessed associations between HPC-10 scores and adverse events, while ROC analysis identified the optimal cut-off.

**RESULTS:** Eighty-eight boys aged 12 weeks (2-55) were included (51 pre- and 37 post-protocol). Anomaly distribution was comparable between cohorts. After protocol implementation, there was a significant increase in HPC-10 score (8 vs. 9,  $P<0.001$ ). Reporting adequacy improved from 3 to 4 ( $P<0.001$ ), particularly on increased reporting of rectal pouch position relative to the pubococcygeal line (16% vs. 65%,  $P<0.001$ ) and to coccyx (22% vs. 51%,  $P=0.004$ ). Diagnostic accuracy remained unchanged (each cohort scored 3,  $P=0.322$ ). Technical adequacy improved overall ( $P<0.001$ ), with higher rates of adequate pressure (67% vs. 95%,  $P=0.002$ ) and adequate exposure (84% vs. 100%,  $P=0.012$ ). Adverse events decreased 5-folds post-protocol (16% vs. 3%,  $P=0.047$ ). The HPC-10 showed excellent discriminatory value (AUROC 0.894), with higher score independently predicted reduced adverse events (OR 0.586, 95% CI 0.434–0.792;  $P<0.001$ ). A HPC-10 score  $\geq 7$  predicted absence of adverse events (sensitivity 0.80, specificity 1.00).

**CONCLUSION:** Protocolising HPC had improved radiological quality (reporting and technique). Despite no change in diagnostic accuracy, procedure-related adverse events had reduced. The HPC-10 criteria offer an objective and reproducible assessment tool with strong clinical value. It supports a structured quality-improvement implementation, radiology evaluation and audit of HPC outcomes to enhance surgical safety in male ARM.

## USE OF THE 5MM ENDOSCOPIC STAPLER FOR LIGATION AND DIVISION IN LAPAROSCOPIC ASSISTED REPAIR OF ANORECTAL MALFORMATION

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**METHODS:** A retrospective chart review was performed on patients who underwent LAARP for imperforate anus between March 2015 and December 2025.

**RESULTS:** Thirty-one patients underwent LAARP with division of the fistula using the 5-mm stapler. The average age was 3.2 months and average weight was 4.5 kg. The location of the fistula was rectoprostatic in 24 cases and rectobladder neck in 5 cases, and high rectovesicle fistula in 2. There were no complications. Follow-up cystoscopy in the first 8 patients showed no discernable fistula, nor any evidence of the metal clips adjacent to the urethral or bladder mucosa.

**CONCLUSION:** Division of a fistula at or above the level of the prostate can safely and effectively be performed with the 5-mm stapler. The stapler allows for division flush with the urethra or bladder ergonomically and quickly.

## INDIRECT EVIDENCE FOR THE VOLUME-OUTCOME RELATIONSHIP FOR CORRECTIVE SURGERY FOR ANORECTAL MALFORMATIONS USING THE IQWiG V24-07 RAPID REPORT METHODOLOGY

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**PURPOSE:** Anorectal malformations (ARM) are rare congenital anomalies requiring demanding corrective surgery. Previous studies have been unable to conclusively determine whether higher hospital caseloads improve surgical outcomes for ARM due to low case numbers, heterogeneous cohorts, and challenges in measuring long-term results. To address this evidence gap, this study applies the IQWiG V24-07 framework to assess whether indirect evidence from other conditions can be transferred to evaluate a potential volume–outcome relationship in corrective surgery for ARM.

**METHODS:** An interdisciplinary expert panel conducted a structured, three-step evaluation. First, potential evidence source populations with strong direct evidence for a volume–outcome relationship were identified. Second, differences between the selected evidence source population and the target population (children undergoing corrective ARM surgery) were systematically analysed according to the 12 criteria defined by IQWiG V24-07. Third, structured group discussion and open voting were performed to assess the transferability of the volume–outcome relationship using predefined categories (A–E).

**RESULTS:** Adult rectal resection was unanimously selected as the most appropriate evidence source population due to robust evidence of a volume–outcome relationship and relevant surgical commonalities, including deep pelvic dissection. Prostatectomy and vaginoplasty were excluded due to limited comparability or insufficient direct evidence. Of the 12 transferability criteria, consensus was reached in six. Differences in comorbidities, concomitant treatment, and setting were judged irrelevant for transferability. Differences in sociodemographic and disease-specific characteristics were considered relevant but estimable in their impact. Divergent ratings were observed for diagnosis, intervention, endpoints, specialization, and follow-up care. Short-term technical outcomes such as anastomotic leakage were deemed directly transferable, whereas mortality was considered largely non-informative in ARM due to low event rates. Functional outcomes, including fecal continence and bladder dysfunction, required subtype-specific interpretation. Overall, the panel concluded that the volume–outcome relationship demonstrated in adult rectal cancer surgery is transferable to corrective ARM surgery.

**CONCLUSION:** Using a structured and transparent methodological approach, this study supports the transferability of a volume–outcome relationship to corrective surgery for ARM. These findings provide an evidence-informed rationale for centralization strategies, structured training, and the development of specialized multidisciplinary care pathways in ARM management.

## BEYOND THE INITIAL HOSPITALIZATION: HEALTH CARE COST DRIVERS IN INFANTS WITH ANORECTAL MALFORMATIONS

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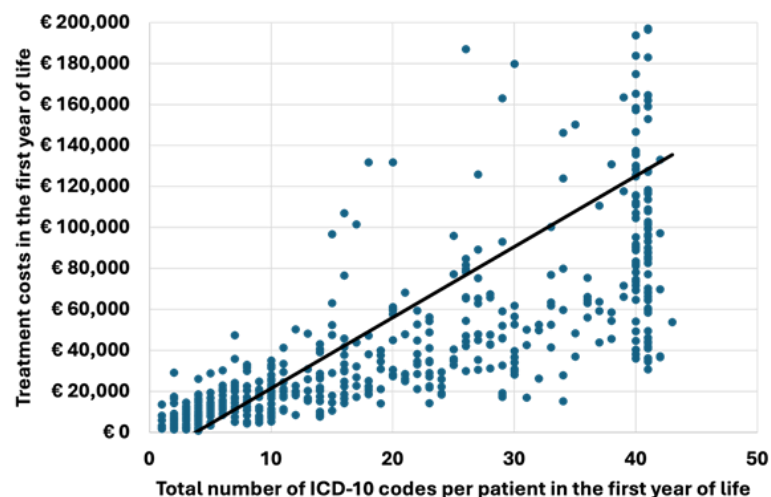
**PURPOSE:** Anorectal malformations (ARM) are rare congenital anomalies requiring surgical repair within the first year of life. Affected patients often present with associated malformations, including the VACTERL association (vertebral, anal, cardiac, tracheoesophageal, esophageal, renal, and limb anomalies). Data on the economic burden of treating patients with ARM and VACTERL are limited.

**METHODS:** Ethical approval was obtained. Pseudonymized health insurance claims data from January 2020 to June 2024, representing approximately 15% of the German population, were analyzed. Data from patients with ARM who underwent surgical reconstruction, and associated congenital anomalies, including those consistent with VACTERL association, were identified using specific ICD-10 and OPS codes. Multiple linear regression models were used to analyze treatment costs and influencing factors, including length of hospital stay, duration of ventilation, and comorbidities during initial hospitalization and the first year of life. Analyses were exploratory, and significant p-values interpreted accordingly.

**RESULTS:** In total, 530 infants with ARM were identified. Cumulative length of hospital stays in the first year of life was  $52 \pm 68$  days. Mean treatment costs were  $\text{€}32,553 \pm 49,171$  for the initial hospitalization. Mean total costs in the first year of life were  $\text{€}61,757 \pm 87,970$ . Each additional day in hospital and of ventilation increased mean treatment costs by  $\text{€}588$  and  $\text{€}78$ , respectively ( $p < 0.0001$ ). Each additional surgical procedure was associated with a mean increase of  $\text{€}734$  ( $p < 0.0001$ ). The presence of esophageal atresia was associated with significantly higher treatment costs, resulting in an additional  $\text{€}19,581$  ( $p = 0.0112$ ). Cumulative treatment costs in the first year of life correlated positively with the number of ICD-10 codes (**Figure**). 71 patients (13.4%) showed clinical features of VACTERL association. Of these, 29 (40.9%) were identified by ICD-10 coding. Conversely, 18 (38.3%) of 47 VACTERL-coded patients did not meet diagnostic criteria based on claims data.

**CONCLUSION:** Management of infants with ARM is associated with substantial healthcare expenditures beyond the initial hospitalization. Length of hospital stay, duration of ventilation, number of ICD-10 codes and surgical procedures, and concomitant esophageal atresia were identified as major cost drivers. ICD-10-based coding appears to incompletely capture clinically defined VACTERL association and may cause misclassification in claims data.

**Figure.** Association between diagnostic burden and treatment costs of patients with ARM in the first year of life. Each dot represents one individual patient.



## EDUCATIONAL RELATIONSHIP BETWEEN HEALTH CARE PROFESSIONAL AND CAREGIVER IN COLOSTOMY MANAGEMENT IN PATIENTS WITH ANORECTAL MALFORMATIONS: AN OBSERVATIONAL STUDY

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**PURPOSE:** Anorectal malformations (ARM) are rare congenital anomalies managed either with primary reconstruction or preliminary colostomy. Effective home care requires adequately trained caregivers. This study aimed to evaluate the impact of the educational relationship between healthcare professionals and caregivers on colostomy management in ARM patients.

**METHODS:** A cross-sectional observational study was conducted at our center. Parents of patients with ARM with preliminary colostomy under active follow-up completed a specifically designed and validated questionnaire. The survey assessed sociodemographic characteristics, surgical details, complications, home management, perceived support, and the impact of nurse-led education on caregiver confidence. Descriptive and statistical analyses were performed.

**RESULTS:** The questionnaire was dispensed to 102 caregivers of patients with ARM who received colostomy; 65 (64%) completed it. Most respondents were mothers 75%, consistent with the fact that in 83% of cases they were primarily responsible for stoma care. Additionally, 55% were employed full-time.

In 89% of patients the stoma was created within the first week of life and in 50% it was maintained for 6–12 months. No stoma-related complications were reported in 37% of cases, and 15% of caregivers reported no management difficulties. The most common issues were stool bag detachment (40%) and faecal leakage (36%). Despite the reported challenges, 58% felt sufficiently confident in managing them at home.

A very high impact on daily life was reported by 37%, while 28% reported minimal and 17% no impact.

Overall, 52% of caregivers felt adequately supported, 29% very well supported, nevertheless and 74% rated the educational support provided by the paediatric nurse as highly important; however, 38% expressed the need for additional practical training.

**CONCLUSION:** Parents identified a dedicated healthcare professional as essential for practical and educational support. Structured training and effective communication improved caregivers' knowledge, confidence, and quality of life. A family-centered approach with continuous support reduced emotional and caregiving burden. These findings highlight therapeutic education as a cornerstone in the management of patients with ARM and colostomy.

**CAREGIVER BURDEN AND ANXIETY SYMPTOMS IN PARENTS OF CHILDREN WITH ANORECTAL MALFORMATIONS: THE MEDIATING ROLE OF ENVIRONMENTAL MASTERY**

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**PURPOSE:** Parents of children with complex congenital conditions such as anorectal malformations often experience high caregiving demands, which may lead to increased caregiver burden, defined as the physical, emotional, and psychological strain associated with providing long-term care, and may negatively affect psychological well-being. Notably, caregiving burden has been associated with high anxiety symptoms. However, the psychological mechanisms through which caregiving burden contributes to anxiety symptoms remain understudied among parents of children with anorectal malformations. The present study aimed to examine the association between caregiver burden and anxiety symptoms and to test whether environmental mastery—a dimension of psychological well-being reflecting the perceived ability to manage life circumstances—mediates this relationship.

**METHODS:** A cross-sectional study was conducted with 106 parents of children diagnosed with anorectal malformations. Caregiver burden was assessed using the Zarit Caregiver Burden Scale, environmental mastery using the Environmental Mastery subscale of the Ryff Psychological Well-Being Scales, and anxiety symptoms using the Hospital Anxiety and Depression Scale. Hierarchical linear regression analyses were performed to examine predictors of anxiety symptoms. A Sobel test was conducted to assess the significance of the indirect effect of caregiver burden on anxiety through environmental mastery.

**RESULTS:** Caregiver burden significantly predicted anxiety symptoms in the first step of the regression model ( $\beta=.47$ ,  $SE=0.03$ ,  $t=5.43$ ,  $p<.001$ ), with the model being statistically significant ( $F(1,104)=29.51$ ,  $p<.001$ ). In the second step, both caregiver burden and environmental mastery were significant predictors of anxiety symptoms ( $F(2,103)=25.36$ ,  $p<.001$ ). Specifically, caregiver burden remained positively associated with anxiety symptoms ( $\beta=.31$ ,  $SE=0.03$ ,  $t=3.42$ ,  $p=.001$ ), whereas environmental mastery was negatively associated with anxiety symptoms ( $\beta=-.37$ ,  $SE=0.08$ ,  $t=-4.09$ ,  $p<.001$ ). The Sobel test indicated a significant indirect effect of caregiver burden on anxiety through environmental mastery ( $z=5.85$ ,  $SE=0.04$ ,  $p<.001$ ).

**CONCLUSION:** These findings suggest that higher caregiver burden is associated with greater anxiety symptoms among parents of children with anorectal malformations and that lower environmental mastery may represent a key psychological pathway linking caregiving burden to anxiety. Interventions aimed at strengthening parents' sense of environmental mastery and perceived control over daily challenges may help mitigate anxiety symptoms in this population, supporting caregivers' mental health.

## A NEW CLASSIFICATION OF MEGARECTUM IN ANO-RECTAL MALFORMATIONS - DESCRIBING 4 SUB-TYPES WITH DIFFERENT ETIOLOGIES AND DIFFERENT MANAGEMENT STRATEGIES: 17 YEARS EXPERIENCE

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**PURPOSE:** Megarectum(MR) in anorectal malformation (ARM) causes severe morbidity. Patients who need surgery often have severe complications and often need redo surgery. To analyse why megarectums occur, propose a new classification based on different sub-types and propose different management strategies for each sub-type so we can reduce the incidence of megarectum.

**METHODS:** Over a 17-year period, we reviewed all ARM to identify megarectum, defined by radiological recto-pelvic ratio > 0.61. A new classification was proposed:

Primary megarectum: pre-anorectoplasty. Subtypes: a) Congenital rectal ectasia b) Late diagnosis of ARM.

Secondary megarectum: post-anorectoplasty. Subtypes: a) Anal stricture b) Constipation and dysmotility.

Complications and Krickenbeck bowel function were compared between conservative management (CM) and surgery.

**RESULTS:** Of 124 ARM, 22 (18%) developed megarectum; of these, 7 underwent surgery. There was no difference in functional outcomes when comparing CM vs surgery—voluntary bowel movement (both 86%), soiling (40% vs. 57%) and constipation (both 86%). However, surgery was associated with major complications (43%) and the requirement for invasive bowel management, compared to CM (85% vs. 27%,  $P = 0.02$ ). 6/7 surgical patients needed antegrade continence enema (ACE), one of these has a permanent ileostomy.

The strategies involved in reducing the incidence of megarectum included- judicious tapering of rectum during PSARP, newer techniques to reduce anal strictures and aggressive post-operative bowel programs in patients with severe constipation and dysmotility.

With strategic changes, incidence of megarectum reduced from 20/77 (26%) before 2013, to 2/47 (4%) after 2013 ( $P = 0.002$ ). More importantly, no patient has needed surgery for megarectum in last 11 years.

**CONCLUSION:** The best treatment of megarectum in ARM is prevention! Surgery for megarectum carried a high risk of complications, with patients often needing ACE or stoma. By adhering to newer strategies discussed, we reduced the incidence of megarectum and have avoided surgery for megarectum since 2014.

**LONG-TERM OUTCOME AFTER VACUUM-ASSISTED CLOSURE IN PATIENTS WITH ANORECTAL MALFORMATIONS: A MULTI-CENTER COMPARATIVE STUDY**

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**PURPOSE:** Perineal wound complications following posterior sagittal anorectoplasty (PSARP) remain a relevant clinical challenge in patients with anorectal malformations (ARM) with an occurrence of 22%-43% after PSARP according to the literature. In adults, vacuum-assisted closure (VAC) therapy has been used to support wound healing after colorectal procedures. In pediatric patients, there is only limited evidence, which suggests a potential benefit of VAC therapy after ARM surgery. We aimed to evaluate the use of endorectal VAC therapy after PSARP and to compare postoperative and long-term outcomes between patients treated with and without VAC therapy in a multicenter comparative study.

**METHODS:** All consecutive patients who underwent PSARP from 01/2018-12/2025 in two participating pediatric surgical departments were included. In one department, all patients (n=26) received VAC therapy following PSARP, in the other PSARP was performed without VAC therapy (n=26). Postoperative perineal wound dehiscence was analyzed.

**RESULTS:** A total of 52 patients were included. Baseline characteristics were comparable between the cohorts. Overall follow-up duration was similar between groups (median 1 [IQR 0-2] vs. 1 [IQR 1-3.8] year, p=0.26). The interval between surgery and first follow-up examination was comparable between cohorts (median 18.0 [IQR 14-20] days in VAC cohort vs. 18.5 [IQR 15-29.5] days in non-VAC cohort, p=0.25). Postoperative outcomes did not differ significantly between the VAC and non-VAC cohorts. Postoperative stool smearing occurred in one patient in the non-VAC cohort and in none of the VAC cohort, without a significant difference between groups (p>0.99). No cases of incontinence were observed in either cohort (p>0.99). Perineal wound dehiscence occurred in 2 patients (7%) in the VAC cohort and in 3 patients (10%) in the non-VAC cohort (p>0.99).

**CONCLUSION:** In this multicenter comparative study, VAC therapy after PSARP was associated with a marked lower rate of postoperative wound dehiscence compared to previously reported rates of wound dehiscence following PSARP and compared to the non-VAC cohort of our present study. Based on our results, VAC therapy might help to reduce postoperative wound dehiscence following PSARP. However, prospective studies with larger patient numbers are required to confirm our findings.

## TRANSITION OF CARE FROM PAEDIATRIC TO ADULT SERVICES FOR PATIENTS WITH ANORECTAL MALFORMATIONS: A QUALITATIVE STUDY USING RICH PICTURES

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**PURPOSE:** Little is known about how patients with anorectal malformations (ARMs), their caregivers and healthcare providers perceive and experience transition from paediatric to adult care (transition of care) in low- and middle-income countries. This study aimed to explore the perceptions and experiences of young adults, adolescents, their caregivers, and healthcare providers regarding transition of care, as well as their perceptions of an ideal transition of care at the Johannesburg Paediatric Colorectal Clinic.

**METHODS:** A qualitative, exploratory study was conducted employing rich pictures. A workshop was held for each of the four stakeholder groups. Participants were asked to draw a rich picture illustrating their perceptions and experiences of the transition of care, followed by a second picture depicting the ideal transition of care. Data were analysed using Braun and Clarke's thematic analysis.

**RESULTS:** Four overarching themes were identified: (1) Born to shine – living with ARM as a lifelong condition that shapes, but does not define, identity; (2) Golden gloves – paediatric services as trusted, emotionally safe spaces, contrasted with fear and uncertainty regarding adult care; (3) Growing up, letting go – transition experienced as both developmental progression and relational loss; and (4) Overwhelmed joint passion – system fragmentation, limited adult expertise in congenital colorectal conditions, poor information transfer, and reliance on informal paediatric workarounds.

Transition of care was experienced as a fragile, relational and system-level process rather than a discrete transfer event. It was marked by the loss of trusted paediatric relationships, uncertainty regarding adult expertise, and fragmented information transfer. These experiences shaped participants' perceptions of an "ideal transition" as one that is relationally anchored, developmentally appropriate, and coordinated across services. Participants identified feasible, low-resource strategies, including: adolescent-focused clinics, joint paediatric adult consultations to build trust, identifiable adult "champions", and structured information-handover tools.

**CONCLUSION:** Co-designed transition pathways offer a pragmatic opportunity to strengthen lifelong care for patients with ARMs in resource-constrained settings.

## PERMANENT OSTOMY IN HIRSCHSPRUNG DISEASE: CAN WE PREDICT WHICH PATIENTS ARE AT RISK?

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**PURPOSE:** Hirschsprung disease is frequently associated with other comorbidities. Chromosomal variants occur in 8–12% of patients, with trisomy 21 present in approximately 12%. When comprehensive screening is performed, associated anomalies are detected in up to 57.5%. Neurodevelopmental delay has been demonstrated even in non-syndromic patients and is associated with worse functional outcomes. Although permanent ostomy has historically been reported in up to 14%, contemporary rates and predictive factors remain unclear. We aimed to determine the rate of permanent diversion among patients who underwent pullthrough for Hirschsprung, identify factors associated with creation of permanent ostomy post pullthrough, and evaluate risk stratification parameters.

**METHODS:** Following Institutional Review Board approval, we performed a retrospective review of all patients diagnosed with Hirschsprung disease and treated at a single tertiary referral center between 2016 and approval date. Patients who underwent permanent ostomy creation were identified. Variables included segment length, associated syndromes, associated malformations, neurodevelopmental and mental health disorders, failed reconstructive surgery, and failed bowel management with antegrade enemas. The primary outcome was permanent ostomy. Patients with total intestinal aganglionosis were excluded.

**RESULTS:** A total of 635 patients with Hirschsprung disease were identified of which 15 patients underwent creation of end ostomies after pullthrough (2.4%). Within this group total colonic Hirschsprung disease was the most common diagnosis (53%) followed by short (20%) and long segment disease (13%). Primary extent of disease was not documented in two cases.

Among permanently diverted patients, 13 of 15 (86.7%) had at least one identified risk factor. Associated syndromes were present in 46.7%, associated malformations in 66.7%, neurodevelopmental or mental health disorders in 60%, failed reconstructive surgery in 20%, and failed bowel management with antegrade enemas in 6.7%.

**CONCLUSION:** In this cohort study, creation of permanent ostomy after pullthrough was less frequent compared to historical reports. It was primarily associated with syndromic conditions, congenital anomalies, and neurodevelopmental comorbidities. Given the high prevalence of associated anomalies in Hirschsprung disease, structured risk stratification may improve counseling and targeted management.

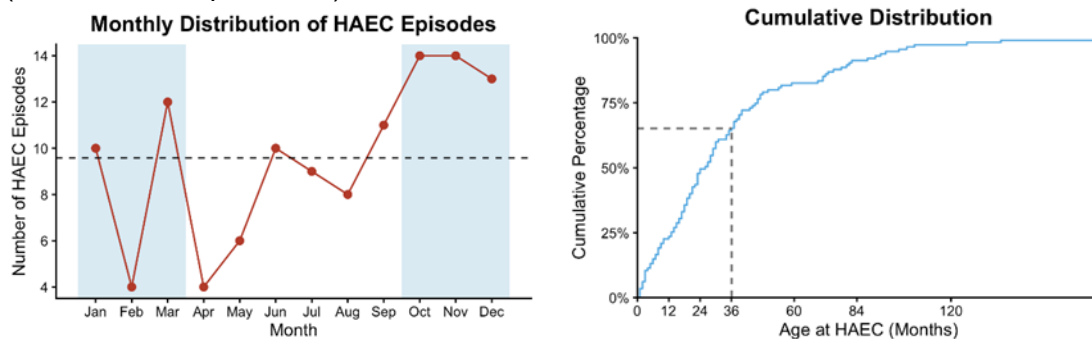
## SEASONAL DISTRIBUTION AND POSTOPERATIVE TIMING OF HIRSCHSPRUNG-ASSOCIATED ENTEROCOLITIS: A SINGLE-CENTER RETROSPECTIVE REVIEW

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**PURPOSE:** Hirschsprung-associated enterocolitis (HAEC) is the most serious complication of Hirschsprung disease. Although HAEC can occur from the neonatal period through adulthood, its frequency appears to decrease with increasing age. Current models of pathogenesis describe a multifactorial process involving impaired epithelial barrier function, dysregulated innate immunity, and alterations in the intestinal microbiome. Respiratory viral infections induce systemic immune responses that may extend beyond the respiratory tract. In children with Hirschsprung disease, who already have compromised mucosal defenses, these may act as systemic stressors capable of precipitating HAEC episodes.

**METHODS:** A retrospective descriptive analysis was conducted of patients presenting with HAEC at Children's Hospital Colorado between 2010 and 2025. HAEC episodes requiring hospital admission were identified, recording admission date and patient age to evaluate temporal and age distribution. Seasonality was analyzed by comparing winter (October–March) and non-winter months. Patients without pull-through surgery or presenting within 30 postoperative days were excluded.

**RESULTS:** A total of 115 HAEC episodes were identified. The median age at presentation was 26 months (IQR 13–46), with 65.8% of episodes occurring within 36 months of age. Episodes occurred most frequently during early childhood (41.7%) and infancy (22.6%). The median time from pull-through surgery to HAEC was 565 days (IQR 179–1065). By postoperative interval, 46 episodes (40.0%) occurred within the first year, 41 (35.7%) between 1–3 years, 17 (14.8%) between 3–5 years, and 11 (9.6%) more than 5 years after surgery. Regarding seasonality, 67 episodes (58.3%) occurred during cold months compared with 48 (41.7%) during warmer months (exact binomial  $p = 0.046$ ).



**CONCLUSION:** A higher proportion of HAEC episodes occurred during colder months, with peaks observed in October, November, December, and March. This seasonal pattern suggests a potential association between respiratory viral infections during colder months, systemic inflammatory responses, and HAEC episodes. Additionally, most HAEC episodes occurred by 36 months of age. We hypothesize that this pattern may reflect increasing immune maturation with age, which could influence susceptibility to HAEC. These findings highlight potential environmental and temporal factors associated with HAEC and may guide future investigations into disease triggers and prevention strategies.

## CROSSING THE 100-PATIENT MILESTONE: OUTCOMES FROM A HIRSCHSPRUNG DISEASE REFERRAL CENTRE IN A RESOURCE-LIMITED SETTING

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**PURPOSE:** To report our experience with Hirschsprung disease (HD) in a low-middle income country, after reaching 100 patients, with the help of a systematic and structured electronic database.

**METHODS:** A retrospective analysis was performed on all HD patients seen in our dedicated clinic from 2019 to 2025, including those primarily treated at or referred to our centre. Data on demographics, surgical management, complications, and follow-up were extracted from the database. Functional outcome was assessed using the Rintala score in patients who underwent pull-through and reached toilet training age. Comparisons between groups were performed using appropriate statistical tests ( $p < 0.05$ ).

**RESULTS:** A total of 102 patients were included. Among these, 24 (23.5%) patients were referred from other hospitals (20/24 after pull-through surgery). Diagnosis was established through rectal suction biopsy or surgical biopsy, according to patient age. Thirty-four patients (33.3%) had delayed diagnosis ( $> 1$  year of age).

Ninety-two patients underwent pull-through surgery, predominantly staged Swenson procedures: 78 colonic (84.8%) and 14 ileoanal (15.2%). The mean number of surgeries per patient was  $3.96 \pm 3.0$  in the colonic group and  $5.0 \pm 1.51$  in the ileoanal group ( $p=0.22$ ). Redo surgery was significantly more frequent in patients referred after undergoing pull-through in other centres (25% vs 5.2%,  $p=0.02$ ), and in patients who underwent ileoanal pull-through compared with colonic (28.6% vs 5.1%,  $p=0.02$ ). Botox injection was also more frequent in the ileoanal group (57.1% vs 20.5%,  $p=0.01$ ). No other significant differences were observed between groups for other individual procedures to manage complications, including strictures or anastomotic leak.

Post pull-through enterocolitis occurred in 21.4% of ileoanal and 9.0% of colonic patients ( $p = 0.047$ ). Overall mortality was 10/102 patients (9.8%), of which five deaths were HD-related (two post-surgical complications, three enterocolitis).

Functional outcome was assessed in 21 patients, with an overall score of  $15.6 \pm 3.7$  and showing comparable results between ileoanal and colonic pull-through groups ( $15.2 \pm 3.12$  vs  $15.81 \pm 3.19$ ,  $p=0.71$ ).

**CONCLUSION:** By serving as a referral centre for HD, including complex cases with prior surgeries elsewhere, we combine high surgical volume with systematic data collection and follow-up to continuously evaluate outcomes.

## ADOLESCENTS' WITH HIRSCHSPRUNG DISEASE AND THEIR PARENTS' EXPERIENCES WITH A PATIENT EDUCATION PROGRAM

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**PURPOSE:** Patient education programs (PEP) may improve health literacy and coping strategies in patients with chronic diseases. We have examined how adolescents with Hirschsprung disease (HD) and their parents experienced a half day PEP.

**METHODS:** HD adolescents and their parents were invited to a 3-hour gender divided PEP. A light meal was served during information about HD. Then adolescents and parents met in separate groups moderated by a multidisciplinary team. After the PEP, attendees answered questions anonymously on a Likert scale (1=very bad, 10=excellent). Ethical approval was obtained. Numbers are given as median with range.

**RESULTS:** 15/37 (41%) of invited HD adolescents and 22 parents (64% mothers) attended. 86% of the boys had rectosigmoid aganglionosis, while 63% of the girls had total colonic aganglionosis. All attendees answered the questionnaire. 29% boys and 75% girls either used bowel management or had an enterostomy. Both adolescents and parents gave high scoring of the PEP (Table). All attendees recommended the PEP to other families

	Boys	Girls	Parents
<b>Demographics</b>			
Rate of attendance	7/22 (32%)	8/15 (53%)	-
Age, years	13 (12-15)	14.5 (12-17)	46 (33-54)
<b>Rating of PEP</b>			
Meeting peers	8 (6-10)	9.5 (5-10)	10 (6-10)
Share experiences with others	8 (7-10)	9 (5-10)	9.5 (7-10)
<b>Rating of health care providers</b>			
Pediatric surgeon	10 (8-10)	10 (8-10)	10 (6-10)
Psychiatrist/psychologist	9 (5-10)	9 (2-10)	10 (3-10)
Stoma nurse	8 (5-10)	9 (4-10)	10 (5-10)

**CONCLUSION:** HD adolescents and their parents were highly satisfied with the PEP. Meeting peers and sharing experiences were especially appreciated.

## ANALYSIS OF REASONS AND RISK FACTORS FOR REOPERATION AFTER RADICAL SURGERY FOR HIRSCHSPRUNG'S DISEASE

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**PURPOSE:** To investigate the reasons of reoperation after radical surgery for Hirschsprung's disease (HD) and analyze risk factors associated with reoperation, providing a basis for optimizing clinical decision-making and reducing reoperation rates.

**METHODS:** This was a single center retrospective cohort study including 507 patients who underwent radical HD surgery from January 2018 to December 2024. Patients were divided into a Reoperation Group (n=108) and a Non-Reoperation Group (n=399) based on whether they underwent reoperation due to postoperative complications. The reasons and the surgical procedures performed for reoperation were collected and analyzed. Clinical data prior to the initial radical surgery were compared between the two groups using univariate analysis to identify potential risk factors. Multivariate logistic regression was further applied to determine independent risk factors for reoperation.

**RESULTS:** Reoperations were performed for the following indications: anastomotic leak (n=33), predominantly managed with enterostomy, with 17 cases subsequently undergoing 23 re-pull-through procedures.; residual aganglionic segment (n=16) and secondary megacolon (n=15), both requiring re-do pull-through; internal sphincter achalasia (n=13) treated with botulinum toxin injection; severe enterocolitis (n=8) requiring enterostomy; and intestinal obstruction (n=9) necessitating laparotomy. Fourteen cases were due to other causes, such as rectourethral fistula. On univariate analysis, significant factors associated with reoperation included the presence of associated anomalies, longer postoperative stay, long-segment colonic aganglionosis, preoperative enterostomy, initial surgery at a non-tertiary hospital, lower parental education, and a lower rate of combined transabdominal surgery (all P<0.05). Multivariate analysis identified long-segment disease as an independent risk factor (OR=3.3, 95% CI: 1.6-6.3, P=0.001). Protective factors were surgery at a tertiary hospital (OR=0.6, 95% CI: 0.5-1.1, P=0.014), a combined transabdominal approach (OR=0.7, 95% CI: 0.5-1.0, P=0.044), and higher parental education (OR=0.6, 95% CI: 0.4-1.1, P=0.048).

Table 1 Univariate analysis

Risk/Protective Factors	Non-Reoperation Group	Reoperation Group	P-value
Group	399	108	
Gender (male ratio)	67.9%	66.7%	0.813
Data prior to the initial radical surgery			
Enterocolitis $\geq$ 2 times	19.5%	24.1%	0.277
Intestinal management methods (without ostomy)			
Colonic irrigation	32.0%	37.4%	0.351
Glycerin/sodium phosphate enema	20.3%	26.8%	0.203
Anal dilatation	3.3%	6.5%	0.247
Multiple modalities ( $\geq$ 2 methods)	44.4%	29.3%	0.010
Co-existing malformations	9.0% (36)	16.7% (16)	0.020
Data of initial radical surgery			
Age at surgery (months)	4.2 (IQR <sub>1</sub> 3.2-7.6)	5.0 (IQR <sub>1</sub> 3.8-10.2)	0.111
Surgical weight (kg)	7.2 (IQR <sub>1</sub> 6.3-9.5)	7.0 (IQR <sub>1</sub> 4.7-12.8)	0.740
Initial Surg. at Tertiary A Hosp.	97.5% (389)	72.2% (78)	0.001
Postop. LOS (days)	8.4 $\pm$ 3.5	10.7 $\pm$ 6.0	0.001
Enterostomy	6.8% (27)	26.9% (29)	0.001
Combined abdominal-transanal approach	98.5% (393)	79.6% (86)	0.001
Long segment / total colonic HD	23.8% (95)	50.9% (55)	0.001
Long segment HD	12.8% (51)	24.0% (26)	0.001
Total colonic HD	11.0% (44)	26.9% (29)	0.001
Parental higher education	50.4%	24.1%	0.001

Table 2. Multivariate logistics regression analysis

Risk/Protective Factors	OR	95%CI	P-value
?2 bowel management methods	0.7	0.3-1.2	0.769
Co-existing malformations	1.1	0.7-2.8	0.793
Data of initial radical surgery			
Initial Surg. at Tertiary A Hosp.	0.6	0.5-1.1	0.014
Postop. LOS (days)	1.2	0.8-3.3	0.584
Enterostomy	1.1	0.7-3.5	0.821
Combined abdominal-transanal approach	0.7	0.5-1.0	0.044
Long segment / total colonic HD	2.6	1.6-5.4	0.003
Long segment HD	3.3	1.9-6.3	0.001
Total colonic HD	1.7	1.2-4.2	0.107
Parental higher education	0.6	0.4-1.1	0.048

**CONCLUSION:** Anastomotic leakage, residual aganglionic bowel, and secondary megacolon constitute the primary reasons for reoperation after radical Hirschsprung's disease surgery. Long-segment disease was identified as an independent risk factor. Protective factors included the initial surgery performed at a tertiary hospital, utilizing a combined transabdominal approach, and higher parental education.

## A NEW ANALYSIS OF PROGNOSTIC FACTORS IN HIRSCHSPRUNG'S DISEASE: FOCUSING ON THE AGANGLIONIC SEGMENT AND THE DILATED SEGMENT RESECTION LENGTH

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**PURPOSE:** To analyze aganglionic segment length and dilated segment resection length (DSRL) in children with Hirschsprung's disease (HSCR), and explore their impacts on short-term postoperative clinical outcomes.

**METHODS:** We retrospectively analyzed children who underwent radical HSCR surgery at Shanxi Children's Hospital from June 2016 to June 2024. Baseline characteristics including age at surgery, sex, and family history, as well as complications such as enterocolitis, soiling, and constipation were collected. Patients were divided into short-segment and long-segment groups based on pathological typing, and further subdivided into three subgroups according to DSRL:  $<10$  cm,  $10 \leq \text{DSRL} < 20$  cm, and  $\geq 20$  cm. Baseline data, DSRL, total intestinal resection length, and aganglionic segment length were compared among groups to determine the optimal DSRL for different pathological types (internal validation using Bootstrap). Anal function was evaluated by the Wingspread scoring system to analyze the relationship between these factors and short-term outcomes.

**RESULTS:** A total of 227 children were included, with 188 cases of short-segment HSCR and 39 cases of long-segment HSCR. The median age at radical surgery was 4 (IQR 3–6) months. Among children with short-segment HSCR, only preoperative anemia showed a statistically significant difference among different DSRL subgroups ( $P=0.037$ ). DSRL, total intestinal resection length, and aganglionic segment length were all associated with postoperative soiling ( $P=0.002$ 、 $P<0.001$ 、 $P=0.046$ ) and perianal erosion ( $P=0.007$ 、 $P=0.006$ 、 $P=0.049$ ). The excellent/good rate for anal function was 89.4%, with an optimal DSRL of 9.3 cm. For children with long-segment HSCR, there were no statistically significant differences in baseline characteristics among different DSRL subgroups. DSRL, total intestinal resection length, and aganglionic segment length were correlated with postoperative soiling ( $P=0.016$ 、 $P=0.003$ 、 $P=0.010$ ) and perianal erosion ( $P=0.023$ 、 $P=0.042$ 、 $P=0.004$ ). The excellent/good rate was 79.5%, with an optimal DSRL of 12.3 cm.

**CONCLUSION:** Both the length of the aganglionic segment and DSRL are key factors affecting the prognosis of HSCR. In both short-segment and long-segment HSCR, DSRL, total intestinal resection length, and aganglionic segment length are all associated with postoperative soiling and perianal erosion. The overall postoperative quality of life of HSCR children is satisfactory. Single-center clinical data suggest that better short-term clinical outcomes are achieved when the DSRL is 9.3 cm for short-segment HSCR and 12.3 cm for long-segment HSCR.

## FEASIBILITY OF A GANGLION CELL HISTOLOGY GRADING SYSTEM FOR INTRAOPERATIVE PROXIMAL RESECTION MARGIN ASSESSMENT IN HIRSCHSPRUNG DISEASE

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**PURPOSE:** Transition-zone pull-through from insufficient resection is a major cause of constipation and recurrent Hirschsprung-associated enterocolitis after pull-through surgery. Although histological criteria for defining the transition zone have been proposed, the “gold standard” requires full-thickness circumferential sampling, which is impractical for intraoperative leveling biopsy. In our clinical practice, ganglion cells within the proximal dilated segments frequently exhibit dysplastic and degenerative changes. We hypothesized that these morphologic alterations may correspond to transition-zone pathology. Building on our previous findings linking proximal-margin ganglion cell histopathology to outcomes, we developed a Ganglion Cell Histology Grading System (GCHGS) and explored its feasibility for intraoperative margin assessment compared to circumferential histopathological evaluation.

**METHODS:** Resected bowel segments from Hirschsprung disease cases were serially sampled at 2-cm intervals along the proximal direction. At each level, H&E-stained paraffin sections were evaluated using two approaches: (1) the proposed GCHGS, which stratifies ganglion cells into six grades based on specific histopathologic features, and (2) the reference standard of full-circumferential histopathological evaluation to exclude transition zone features. Two independent pathologists interpreted the slides in a blinded manner. Diagnostic performance (AUC, accuracy, sensitivity, specificity, PPV, NPV) and inter-reader agreement (ICC, Cohen’s  $\kappa$ ) were calculated.

**RESULTS:** A total of 137 level-specific assessments were analyzed. Compared with the reference approach, GCHGS demonstrated robust diagnostic performance, achieving an AUC of 0.832 and an overall accuracy of 0.825. Sensitivity was 0.782 (61/78) and specificity was 0.881 (52/59). Additionally, PPV was 0.897 (61/68) and NPV was 0.754 (52/69). The GCHGS yielded moderate inter-reader agreement (ICC = 0.652,  $\kappa$  = 0.650), which was superior to the agreement observed with the standard reference approach (ICC = 0.540,  $\kappa$  = 0.538).

**CONCLUSION:** In this exploratory study, GCHGS showed promising diagnostic performance in identifying transition-zone pathology, with improved inter-reader agreement relative to the reference standard. Larger prospective studies are warranted to refine this framework and validate its clinical utility in preventing morbidity related to transition-zone pull-through.

## IS THE APPENDIX A SUITABLE TOOL TO DIAGNOSE TOTAL COLONIC AGANGLIONOSIS: VISUALISATION OF GANGLION CELLS ACROSS AGE AND PATHOLOGY

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**PURPOSE:** The appendix has been proposed as a surrogate marker for TCA, but uncertainty regarding ganglion cell detectability across ages, pathological conditions and effects of integrity of tissue limits its clinical adoption.

**METHODS:** Prospective longitudinal study including both children and adults with resection of appendix or appendiceal tip biopsy between October 2022 and September 2024. 4 Subgroups were analysed: children with Hirschsprungs Disease (HD), children with TCA, non-HD children, and non-HD adults. Appendiceal specimens were evaluated with standard haematoxylin and eosin staining, with selective immunohistochemistry in equivocal cases. Ganglion cell presence was reported as present or absent by the Pathologist. The primary outcome was appendiceal ganglion cell detectability; secondary analyses evaluated the effects of age and inflammatory or malignant pathology and tissue destruction. Ethical vote available.

**RESULTS:** In 203 appendiceal specimens, ganglion cells were identified in 23/24 HSCR cases (96.0%) and absent in 18/19 TCA cases (94.7%). Ganglion cells were consistently present in all evaluable non-HSCR children and adults, with no false aganglionosis identified. Five non-HSCR specimens were non-evaluable due to severe tissue destruction. Cohort affiliation strongly predicted ganglion cell detectability ( $p < 0.0001$ ). Neither age nor inflammatory or malignant pathology significantly impaired assessment.

**CONCLUSION:** Appendiceal ganglion cells are reliably detectable across ages and pathological conditions in non-HSCR patients, with absence largely confined to TCA. Incorporating appendiceal histology into the diagnostic pathway may support earlier identification or exclusion of TCA, reduce diagnostic uncertainty, and complement systematic colonic biopsies in complex cases.

## DEFINING THE DISTAL MARGIN IN HIRSCHSPRUNG DISEASE SURGERY: WHY STANDARDIZED ANORECTAL ANATOMY MATTERS FOR CONTINENCE

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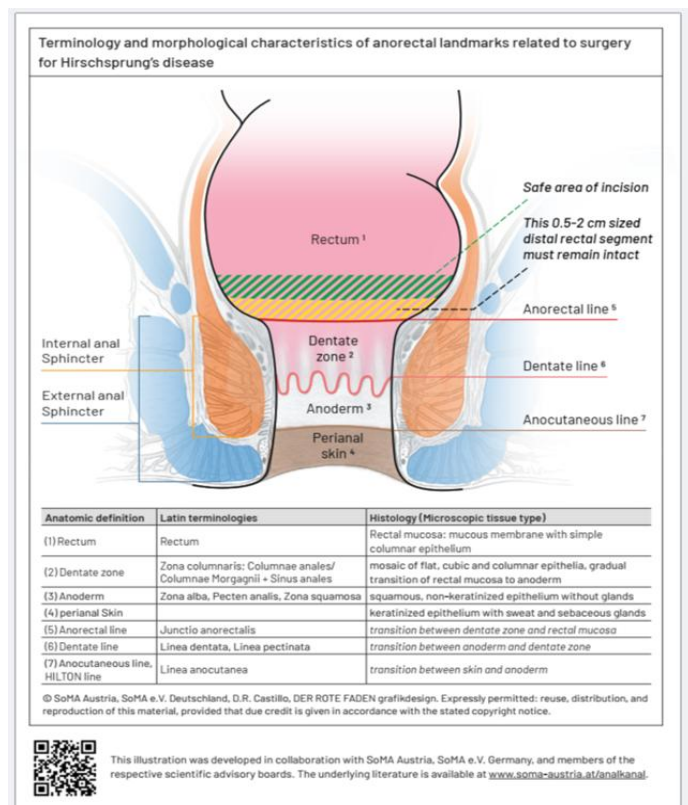
**PURPOSE:** Preservation of fecal continence mechanisms is a key objective of low rectal resection in Hirschsprung disease (HD). However, both historical and contemporary literature reveals considerable variability not only in surgical approaches but also in the anatomical terminology used to define the distal resection margin. Irreversible injuries to the anal canal continue to occur and are often associated with long-term functional consequences. This raises the question of whether imprecise anatomical nomenclature may contribute to avoidable errors and claims for a consented definition and standardized terminology of anorectal anatomy.

**METHODS:** We conducted a focused review of relevant surgical publications and anatomical nomenclature addressing the distal rectum, anal canal, dentate line, and anorectal line in the context of HD surgery. Terminological usage was compared with established anatomical definitions, and commonly used synonyms were systematically analyzed.

**RESULTS:** Substantial variability was identified in the description and demarcation of the distal rectum and anal canal. Terms such as dentate line, pectinate zone, and anorectal line are frequently used inconsistently, which may lead to uncertainty in defining the distal resection margin. To address this issue, we present a concise graphical overview and a structured list of anatomical synonyms intended to facilitate clearer identification and communication of these key landmarks.

**CONCLUSION:** Respecting the anorectal line requires a shared and precise anatomical understanding. Greater consistency in terminology may facilitate intraoperative orientation and support preservation of the continence mechanisms. Clearer definition of the distal margin may help reduce functional complications and improve long-term outcomes following Hirschsprung surgery.

**Figure.** Terminology and morphological characteristics of anorectal landmarks related to surgery for Hirschsprung’s disease.



## CLINICAL OBSERVATION OF DIFFERENT PREOPERATIVE BOWEL MANAGEMENT APPROACHES IN PATIENTS WITH LONG-SEGMENT HIRSCHSPRUNG'S DISEASE

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**PURPOSE:** To evaluate three preoperative bowel preparation methods—cleansing reflux enema (CRE), colostomy, and indwelling transanal tube (ITT)—for long-segment Hirschsprung's disease (LSHD).

**METHODS:** We retrospectively analyzed 68 LSHD patients undergoing radical surgery (2019–2025), divided into CRE, colostomy, and ITT groups. Postoperative recovery and complications were compared.

**RESULTS:** Compared to colostomy (n=16), CRE (n=45) showed larger preoperative intestinal diameter but shorter postoperative hospital stay and fasting duration, with no difference in operative time. ITT (n=7) had shorter postoperative fasting and hospital stay versus colostomy, with comparable preoperative intestinal diameter. CRE had larger preoperative intestinal diameter than ITT, but similar postoperative outcomes. No intergroup differences were observed in enteritis episodes or severity scores.

Table 1 Preoperative conditions

Variables	CRE (n=45)	Colostomy (n=16)	ITT (n=7)	P
Sex(male), n(%)	32(71.11)	13(81.25)	5(71.43)	0.726
Age(y), median(range)	106 (81,131)	188 (136,240)	94 (73,115)	<0.001
Weight-for-age Z-scores,mean(SD)	-0.06±0.22	-0.13±0.24	-0.68±0.24	0.508
Intestinal diameter(mm),mean(SD)	38.04±9.18	25.73±7.70	25.22±5.86	<0.001
Preoperative enteritis episodes,mean(SD)	0.24±0.49	0.23±0.60	0.29±0.49	0.708
Preoperative enteritis scores,mean(SD)	0.89±1.87	0.54±1.33	2.00±2.77	0.202

Table 2 Intraoperative conditions

Variables	CRE (n=45)	Colostomy (n=16)	ITT (n=7)	P
Operative time(min),mean(SD)	226.62±54.00	252.63±65.55	283.86±86.80	0.044
Bowel resection length(cm),mean(SD)	17.50 (14.25,20.75)	28.05 (21.95,34.15)	20.00 (13.5,26.5)	<0.001
Turnover(yes),n(%)	0 (0.00)	3 (18.75)	1 (14.29)	0.015

Table 3 Postoperative recovery conditions

Variables	CRE (n=45)	Colostomy (n=16)	ITT (n=7)	P
Hospital stay(d),median(range)	12 (9, 15)	14 (13,15)	12 (9, 15)	0.011
Postoperative fasting days(d),median(range)	2 (1.5,2.5)	4 (3,5)	2 (1.5,2.5)	<0.001
Postoperative enteritis episodes,mean(SD)	0.53±1.10	0.38±0.62	0.14±0.38	0.660
postoperative enteritis scores,mean(SD)	1.64±2.82	1.50±2.42	0.71±1.89	0.679

**CONCLUSION:** Colonoscopic transanal tube placement is a safe, effective, and minimally invasive decompression method, offering significant value for preoperative optimization in LSHD.

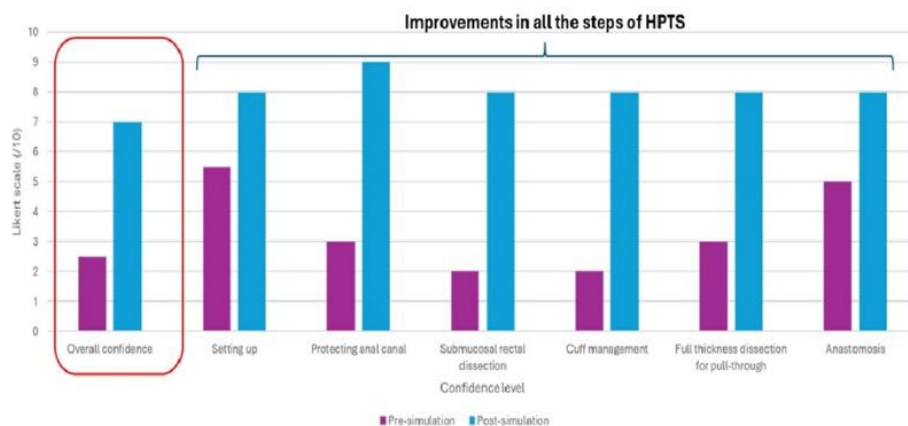
## IMPACT OF A LOW-COST LAMB ANUS HIRSCHSPRUNG'S PULL-THROUGH SURGERY MODEL: RESULTS FROM A STRUCTURED NATIONAL WORKSHOP

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**PURPOSE:** Hirschsprung's Soave pull-through surgery (HPTS) is technically demanding with limited trainee exposure, raising patient safety concerns during hands-on training. Following successful departmental introduction of a validated low-cost lamb anus model (BAPS 2024 presentation), we conducted a nationwide workshop for senior paediatric surgical trainees and junior specialists to evaluate the model's impact on operative confidence, skill acquisition and clinical application.

**METHODS:** A national HPTS workshop was funded by the UM Teaching Enrichment Research Grant (2024) using the lamb anus model (see Figure) (cost <USD 5) comprised: (1) fresh lamb anorectum with terminal perianal muscle and 3–5 cm perianal skin preserved; (2) a 20 cm rectum inserted into a mini-basket perforation and secured; and (3) dissection using Medtronic diathermy. Each trainee performed the five-step Soave procedure (Figure) twice (90–120 minutes per attempt) under 1:1 supervision. A consensus-derived Procedure-Based Assessment form was used for formative assessment. Operative confidence for each step was measured pre- and post-workshop using a 10-point Likert scale. Model realism and utility were similarly rated. A 3–6-month follow-up survey assessed clinical translation.

**RESULTS:** Seventeen participants and 14 trainers from nine paediatric surgical centres attended. Thirty-four simulated procedures were completed using the model described. Median overall confidence improved from 3.0 to 7.0 ( $P<0.0005$ ). Greatest gains were in submucosal dissection (2.0 to 8.0), cuff management (2.0 to 8.0) and anastomosis (3.0 to 8.0) (all  $P<0.001$ ) (**Figure**). Second attempts demonstrated greater fluency and reduced trainer input. Model realism and training utility were rated 8/10 and 9.5/10, respectively. Follow-up (12/17; 71%) showed 100% agreement that the workshop improved ability to assist or perform HPTS; 66% strongly agreed they were better operators and 60% better assistants. Qualitative feedback highlighted improved anatomical orientation and procedural clarity.



**CONCLUSION:** This structured simulation-based HPTS workshop significantly improved confidence, procedural understanding and workplace application. The model is high in fidelity, scalable, ethical and effective for HPTS training.

## POTENTIAL BENEFITS OF SIMULTANEOUS ANORECTO-VAGINOPLASTY FOR PATIENTS WITH PERSISTENT CLOACA: INSIGHTS FROM A NATIONWIDE SURVEY IN JAPAN

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**PURPOSE:** One of the key topics for patients with persistent cloaca is determining when and which procedure to perform vaginoplasty. The purpose of the present study was to clarify the treatment outcomes of simultaneous and metachronous anorecto-vaginoplasty, and to define limit for timing of simultaneous surgery.

**METHODS:** The present study was retrospective cohort study reviewed based on the results of a national survey in Japan. A total of 299 patients underwent both vaginoplasty and anorectoplasty were enrolled in the present study. We compared patient characteristics, surgical outcomes, and menstrual related surgical outcomes between the simultaneous (n=237) and metachronous surgery (n=62) groups.

**RESULTS:** The median follow-up term after vaginoplasty of all patients was 144 [77-223] months, and simultaneous surgery group and metachronous surgery group were 140 [78-217] months and 162 [63-261] months, respectively. The incidence of redo-vaginoplasty and redo-anorectoplasty showed no significant difference between the simultaneous and metachronous group. The ages at the first redo-vaginoplasty and anorectoplasty were significantly younger in the simultaneous group than in metachronous surgery group ( $p < 0.01$ ) as shown in **Table 1**. The incidence of surgical management for menstrual outflow obstruction showed a lower trend in the simultaneous surgery group ( $p=0.08$ ). The cutoff surgical timing based on defecation function in the simultaneous surgery group was 18 months (Odds:0.33,  $p < 0.01$ ).

**Table 1.** Comparison of surgical outcomes between different surgical strategies

	All patients (n=299)	Simultaneous (n=237)	Metachronous (n=62)	p-value
<b>Vaginoplasty</b>				
Age at primary vaginoplasty, (months old)	18 [11-35]	15 [10-22]	133 [63-164]	< 0.01
First redo-vaginoplasty	69 (23.1)	53 (22.4)	16 (25.8)	0.61
Age at surgery, (months old)	85 [45-146]	74 [43-123]	150 [86-222]	< 0.01
Second redo-vaginoplasty, (n, %)	15 (5.0)	13 (5.5)	2 (3.2)	0.74
Age at surgery, (months old)	135 [48-165]	135 [61-178]	93 [64-123]	0.91
<b>Anorectoplasty</b>				
Age at primary anorectoplasty(months old)	14 [9-24]	15 [10-22]	14 [8-28]	0.83
First redo-anorectoplasty, (n, %)	71 (23.6)	52 (21.9)	19 (30.6)	0.18
Age at surgery, (months old)	32 [21-59]	28 [19-40]	59 [34-94]	< 0.01
Second redo-anorectoplasty, (n, %)	5 (5.0)	3 (1.3)	2 (3.2)	0.28
Age at surgery, (months old)	120 [77-124]	77 [52-101]	133 [127-140]	0.25

**CONCLUSION:** Simultaneous surgery is comparably feasible to metachronous surgery without increasing the need for redo-vaginoplasty or redo-anorectoplasty. Anorecto-vaginoplasty should be performed before 18 months of age from the perspective of defecation function.

## ROLE OF LAPAROSCOPY IN THE MANAGEMENT OF ANORECTAL MALFORMATIONS: ONE CENTER EXPERIENCE

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**PURPOSE:** Current functional outcomes for anorectal malformations (ARMs) are unsatisfactory. According to recent report by American Pediatric Surgical Association, high incidence of patients suffers from constipation, urinary tract infection and fecal incontinence. The aim of this study is to investigate the role of laparoscopic assisted anorectoplasty (LAARP) in management of ARMs.

**METHODS:** A retrospective review of 610 ARMs patients treated with LAARP from 2001 to 2025 at our centers. Their age ranged from 2 days to 24 years ( $3.2 \pm 0.42$  months, 51 male neonates). The classification of the malformations included rectovesical fistula in 63 cases, prostatic fistula in 146, bulbar fistula in 275, no fistula in 17, rectovestibular fistula in 53, cloaca in 56, postoperative urethral diverticulum in 30 and postoperative megarectum in 31.

**RESULTS:** LAARP was successfully performed in all patients without conversion to open and necessity for blood transfusion. The average operating time was  $1.54 \pm 0.40$  hours, 8 (1.3%) and 12 (2.0%) patients suffered from rectal retraction and anal stenosis respectively and needed surgical intervention. Anorectal prolapse occurred in 54 cases (8.9%) and corrected by stratified surgical techniques. No male patient suffered from urethral injury, recurrent urethral fistula and urinary incontinence. However, the minor urethral diverticulum without symptom was found in 3 of 535 rectourinary fistula (0.56%) by protocol postoperative VCU. 30 patients with postoperative urethral diverticulum and 31 with postoperative megarectum were successfully treated. 503 patients had been followed up for 3 to 24 years, voluntary bowel movements achieved in 481(95.6%); no soiling or grade 1 soiling in 433 (86%), grade 2 soiling in 42 (8.3%), and grade 3 soiling in 28 (5.6%); no constipation or grade 1 constipation in 437 (86.8%), grade 2 constipation in 55 (11.0%), and grade 3 constipation in 11 (2.2%).

**CONCLUSION:** LAARP facilitates sufficient rectal mobilization, accurate fistula closure, and precise tunnel formation, playing an important role in ARMs management.

**CLOACAL MALFORMATION: POSTNATAL MANAGEMENT AND LONG-TERM OUTCOMES**

Z. Wang, J.-K. Yu, W. Zhong, H. Xia  
(Guangzhou, China)

**PURPOSE:** Cloaca is a rare and complex anorectal malformation in females, characterized by a single perineal opening. This report summarizes the clinical experience and management protocol for patients with cloacal malformation at our center over a 22-year period, emphasizing a comprehensive, multidisciplinary approach.

**METHODS:** We conducted a retrospective analysis of patients treated for cloacal malformation from June 2002 to October 2024. Since 2015, a systematic, protocol-driven management pathway was implemented, covering the entire lifespan from fetal diagnosis to adult care. Management stages include initial neonatal assessment for emergencies (vaginal hydrocolpos, urinary retention), fecal/urinary diversion if needed, definitive diagnostic imaging at 3-6 months to define anatomy (common channel length, vaginal/urethral length), and individualized radical surgery. Postoperative care involves long-term follow-up of functional outcomes and management of complications by a multidisciplinary team.

**RESULTS:** A total of 69 patients were included. Anatomical classification based on common channel (CC) length (cm) revealed: Type I (CC $\leq$ 1) in 28 (40.6%), CC 1-3 in 20 (29.0%), CC 3-5 in 15 (21.7%). Associated anomalies were frequent: cardiovascular (46.4%), urologic (43.5%), and spinal (33.3%). Radical surgery was performed in 64 patients at a mean age of 21.3 months. Surgical techniques included posterior sagittal anorectoplasty (PSARP) with vaginal pull-through (20.3%), total urogenital mobilization (TUM, 25.0%), and vaginal replacement (23.5%). Notably, we reported the world's first series of single-port laparoscopic-assisted vaginal replacement with anoplasty (9 cases, 14.1%). Postoperative complications included wound dehiscence (7.2%), rectal retraction/stenosis (5.8%), and vaginal retraction/stenosis (4.3%). With a median follow-up of 104 months, management spans from infancy through puberty, addressing bowel and bladder function, vaginal patency, and reproductive health.

**CONCLUSION:** Congenital cloacal malformation requires a structured, lifelong, multidisciplinary management strategy. Our data demonstrates the complexity of the condition and the need for individualized surgical planning based on precise anatomical delineation. Long-term follow-up is essential to optimize functional outcomes and quality of life into adulthood.

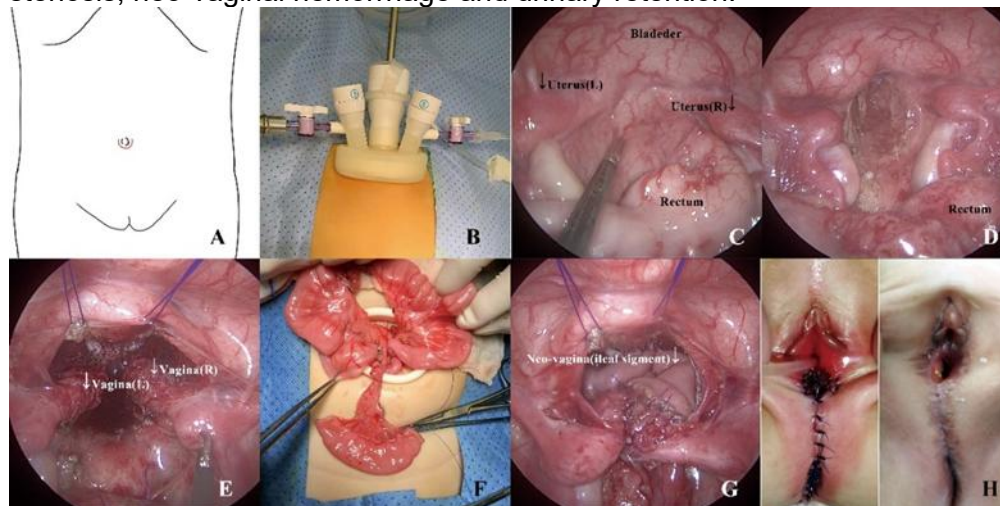
## SINGLE-PORT LAPAROSCOPIC ASSISTED VAGINAL RECONSTRUCTION FOR THE MANAGEMENT OF CLOACA: A NOVEL APPROACH

Z. Wang, J.-K. Yu, W. Zhong, H. Xia  
(Guangzhou, China)

**PURPOSE:** Reconstructing a cloaca accompanied by a short urethra presents significant challenges. Traditionally performed via laparotomy, we present an initial series of single-port laparoscopic-assisted vaginal reconstruction (SPLAVR) and report the preliminary results.

**METHODS:** Patients who underwent SPLAVR between November 1, 2021, and August 31, 2025, were retrospectively reviewed. Data, including the demographic characteristics and clinical presentations of all 11 patients, were reviewed retrospectively. Patients were clinically followed up for at least 6 months, and outcomes, including length of stay (LOS), operation time, amount of blood loss, and postoperative complications, were documented.

**RESULTS:** Eleven female patients meeting the surgical indications underwent SPLAVR between November 1, 2021, and August 31, 2025. Eight patients underwent ileal vaginoplasty, two had colon vaginoplasty, and one had vaginal switch. The age at the time of surgery ranged from 4 to 12 months, and the body weight ranged from 7.0 to 10.2 kg. The sacral ratio varied from 0.28 to 0.89. The operations lasted from 430 to 660 min, with blood loss ranging from 10 to 30 ml, and length of stay varying from 10 to 22 days. Post-operative complications were neo-vaginal stenosis, neo-vaginal hemorrhage and urinary retention.



**CONCLUSION:** Single-Port Laparoscopic Assisted vaginal reconstruction offers surgeons a cosmetically appealing and minimally invasive approach to address this challenging condition.

**MID-TERM FUNCTIONAL OUTCOMES AFTER SURGICAL REPAIR OF CLOACAL MALFORMATION: EXPERIENCE FROM A TERTIARY CENTER IN CHINA**

J. Wang, M. Zhang  
(Shanghai, China)

**PURPOSE:** To evaluate mid-term bowel and urinary function, quality of life and associated risk factors in patients following the surgical repair of cloacal malformations.

**METHODS:** We retrospectively reviewed female patients who underwent surgery for cloacal malformation at a tertiary center in China between June 2006 and June 2024. Patients were stratified by common channel length into Group A ( $\geq 3$  cm) and Group B ( $< 3$  cm). Postoperative bowel function, urinary functional outcomes and Pediatric Quality of Life (PedsQL) were analyzed.

**RESULTS:** Thirty-one patients were included (Group A,  $n=7$ ; Group B,  $n=24$ ). Associated anomalies involved the reproductive (45.2%), urinary (38.7%), spinal (35.5%), and gastrointestinal (22.6%) systems. The median age at surgery was 11 months (range, 6–15 months), with a median follow-up of 8.6 years (range, 4–19 years). Three patients were lost to follow-up, and one died of renal failure, 27 patients were available for functional evaluation. Rectal mucosal prolapse occurred in 9 patients (33.3%). Constipation was reported in 11/27 (40.7%) and fecal incontinence in 6/27 (22.2%). Multivariable analysis showed that a longer common channel length independently increased the risk of fecal incontinence. Group B had a significantly higher Bowel Function Score than Group A ( $15.73 \pm 2.28$  vs  $12.75 \pm 1.50$ ,  $P=0.027$ ). Daytime urinary incontinence ( $n=4$ ), nocturnal enuresis ( $n=5$ ), and voiding difficulty ( $n=3$ ) showed no significant between-group differences. Urinary tract infection (UTI) occurred in 9/27 (33.3%), with two patients requiring prophylactic antibiotics and three requiring clean intermittent catheterization. UTI was significantly more frequent in Group A than Group B. PedsQL scores (median [IQR]) were significantly lower in Group A for physical functioning (87.14 [81.25–91.96] vs 100.00 [90.00–100.00],  $P=0.009$ ), social functioning (85.00 [85.00–91.67] vs 100.00 [91.67–100.00],  $P=0.012$ ), and total score (87.06 [85.31–88.95] vs 93.75 [87.50–98.75],  $P=0.035$ ). Emotional ( $P=0.215$ ) and school functioning ( $P=0.064$ ) did not differ significantly.

**CONCLUSION:** In this mid-term follow-up cohort, outcomes after cloacal repair varied substantially. Common channel length was a key determinant of bowel outcomes, UTI risk, and PedsQL performance. Accurate preoperative assessment and intensified follow-up with targeted interventions for patients with a long common channel may optimize functional prognosis and quality of life.

## SOCIODEMOGRAPHIC AND CLINICAL CHARACTERISTICS OF YOUTH LOST TO FOLLOW-UP AFTER PARTICIPATION IN PEDIATRIC BOWEL MANAGEMENT

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**PURPOSE:** Participation in a bowel management program can significantly improve patient bowel functioning. Patients should be evaluated at least annually to assess their current regimen and make any necessary adjustments. This study describes the medical and sociodemographic characteristics of patients who did not participate in a follow-up visit within one year after successful completion of a bowel management program.

**METHODS:** The patient database was reviewed to identify patients who participated in a full bowel management week program but did not return for follow-up with the colorectal providers from 2016-2025. Demographic variables were examined including diagnosis, insurance type, geographic location, and Area Deprivation Index (ADI).

**RESULTS:** Among 102 patients who did not attend follow-up within one year of completing the pediatric bowel management program, mean age was  $17.2 \pm 10.2$  years (median 14; range 4–52), and 50.5% were male. The most common diagnoses were anorectal malformation (51.5%) and idiopathic constipation (31.1%), followed by Hirschsprung disease (8.7%) and spinal etiologies (7.8%). Most patients were privately insured (66.0%), while 26.2% had public insurance. The cohort was predominantly White/Caucasian (70.9%) and non-Hispanic/Latino (68.9%); 18.4% identified as Hispanic or Latino. English was the preferred language for 90.3% of patients. Thirty-seven percent resided within Colorado, 56% resided in the United States, and 7% resided outside of the US. Area Deprivation Index data were available for 92 patients, with a mean ADI of  $30.6 \pm 22.8$  (median 28.5; Range 1-94), reflecting wide socioeconomic variability.

**CONCLUSION:** While follow-up visits are essential for ongoing evaluation and maintenance of bowel management, a substantial number of patients did not return for follow-up within one year of program completion. Most of these patients reside outside of the state where the program is based and have private insurance. These results emphasize the importance of expanding provider and institutional capacity to deliver telehealth services across state lines, and they highlight the need to more thoroughly identify structural and logistical barriers to follow up in order to develop strategies that support sustained, long term patient engagement.

## THE STRAIN OF CONSTIPATION: A RETROSPECTIVE STUDY AT A SOUTH AFRICAN TERTIARY CHILDREN'S HOSPITAL

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(Cape Town, South Africa)

**PURPOSE:** The burden of functional constipation on tertiary healthcare, particularly in resource-constrained settings, remains under-characterised. This study aims to describe the clinical characteristics, management approaches, and outcomes of children presenting with functional constipation at a tertiary-level paediatric hospital in South Africa.

**METHODS:** A retrospective chart review was conducted at Red Cross War Memorial Children's Hospital (RCWMCH) between 1 March 2019 and 30 May 2019. Children aged 0-13 years who presented to paediatric surgical and medical outpatient departments with a diagnosis of constipation were included. Children with an organic cause for their constipation were excluded. Data was extracted from patient medical records according to diagnostic codes. Descriptive statistics were used to summarise findings.

**RESULTS:** A total of 210 patients were included. Ninety-nine patients (47%) were aged 1–5 years and sixty-two patients (30%) were aged 5–10 years. The median symptom duration was 24 weeks; sixty-three patients (30%) had symptoms persisting for over one year. First time attendees made up seventy percent (70%) of the cohort which equated to 186 patients. Multiple healthcare provider involvement was common, and 37 patients (17%) required hospitalisation (median duration of admission was 2 days). While 106 patients (50%) were referred to a dietitian, only thirteen of sixty-five patients (20%) of those with psychosocial stressors were seen by a social worker. Clinical improvement was reported by patient or caregiver in 128 cases (61%) of cases.

**CONCLUSION:** Functional constipation in children imposes a substantial burden on tertiary services. Its management is fraught with gaps in continuity of care and lacks cohesive multidisciplinary involvement.

## THE UTILITY OF TRANSABDOMINAL RECTAL ULTRASOUND IN PAEDIATRIC BOWEL MANAGEMENT PROGRAM

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**PURPOSE:** We investigate the utility of clinician performed transabdominal rectal ultrasound (TRUS) in a paediatric bowel management program (BMP). We aim to correlate the measurement of the transrectal diameter (TRD) with a standardized symptoms list and clinical symptoms of stool retention in paediatric patients with Anorectal Malformation (ARM), Hirschsprung's Disease (HD) and Functional Constipation (FC).

**METHODS:** Prospective cohort study including patients in the paediatric colorectal outpatient clinic at a tertiary paediatric colorectal center. Clinician bedside measurement of the transrectal diameter with our institutional ultrasound protocol. We included 380 patients. Sex-ratio was adjusted in control group for HD (CG1), while sex was balanced for ARM and FC (CG2). ARM and HD were categorized into subgroups (a-/symptomatic at the different follow up visits) according to the list of symptoms and in FC following ROM-IV criteria. ARM and HD patients included after successful surgery and  $\geq 1$  year old, while total aganglionosis excluded.

**RESULTS:** All symptomatic subgroups had significantly higher TRD than control groups ( $p < 0,001$ ) and reduced after successful F/P ( $p < 0,015$ ). Cut-offs calculated for each cohort and collective cut-off was 2,985 cm (Sensitivity = 81,92 %, Specificity = 97,73 %, Area under curve = 0,91 (0,87 - 0,94), Accuracy 88,67 %). Correlation of TRD and age for CG2 was  $r = 0,188$  with a positive correlation turnover at the age of 5 years.

**CONCLUSION:** Final results demonstrate non-inferiority of TRUS to abdominal X-ray (AXR) in paediatric BMP and reproduce results of prior studies. The calculated cut-offs exceed most of the Receiver Operating Characteristics for AXR.

## OUTCOMES AND REVISION RATES AFTER ANTEGRADE CONTINENCE ENEMA APPENDICOSTOMY IN A SINGLE-CENTER PAEDIATRIC COHORT IN A LOW- AND MIDDLE-INCOME COUNTRY

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**PURPOSE:** Antegrade continence enema (ACE) appendicostomy is widely used for children with complex colorectal and neurogenic bowel conditions, yet outcome data from low- and middle-income countries (LMICs) are limited. We aimed to describe indications and revision rates after ACE in a single-center LMIC paediatric cohort.

**METHODS:** A retrospective review was performed of all ACE appendicostomies undertaken at a tertiary paediatric surgical unit between 2012 and 2025. Theatre and clinic records were used to obtain demographic data, indication for ACE, need for revision, and age at index surgery and revision, and descriptive statistics summarised indications and revision rates by diagnostic group.

**RESULTS:** Forty-six children underwent ACE: 37 (80%) were male. The most common indication was anorectal malformation (ARM) in 32/46 patients, followed by spina bifida/myelomeningocele (MMC) in 8, Hirschsprung disease (HD) in 2, functional constipation in 1, perineal trauma in 1, and 2 with unknown indication. In 7 patients the ACE was combined with a Mitrofanoff (4 MMCs and 3 ARMs). Sixteen patients (35%) required at least one surgical revision of the ACE; in 2, the ACE was replaced with a cecostomy, both having a split appendix for ACE and Mitrofanoff. Revisions were required in 9/32 (28%) ARM patients, 3/8 (38%) spina bifida/MMC patients, 2/2 (100%) HD patients, and 2/2 (100%) patients with unknown indications. Age at ACE surgery was available for 34 patients, with a median of 100 months (8.3 years), and age at revision was available for 8/16 (50%) patients, with a median interval of 5 months from index surgery (range 3–50).

**CONCLUSION:** In this LMIC paediatric cohort, ACE appendicostomy was used predominantly for ARM and spina bifida/MMC, with an overall revision rate of 35% and most revisions occurring within the first six months. This rate appears slightly higher than reports from high-income settings (approximately 10–33%), likely reflecting resource constraints such as limited availability of dedicated ACE stoppers and delayed access to hospital when stoppers or catheters are dislodged. ACE remains a valuable option in this context, and our findings highlight opportunities to optimise local protocols and supply chains to improve outcomes in this cohort.

## **MALONE PROCEDURE FOR THE MANAGEMENT OF FECAL INCONTINENCE IN PEDIATRIC PATIENTS AT A TERTIARY REFERRAL CENTER IN MÉXICO: A COMPARISON OF TWO SURGICAL TECHNIQUES**

M.P. Resendiz-Vélez, A. Costá-Roig, E.J. Fernández-Portilla, R. Dávila-Pérez, A. Domínguez-Muñoz  
(Mexico City, Mexico)

**PURPOSE:** To evaluate the outcomes of the Malone procedure in pediatric patients with fecal incontinence and to compare results between the open and laparoscopic techniques.

**METHODS:** A comparative analysis of demographic and surgical variables was conducted in pediatric patients with fecal incontinence who underwent a Malone procedure (open or laparoscopic) between 2022 and 2025. A total of 16 patients were included.

**RESULTS:** Operative time was shorter in the laparoscopic group (128 vs. 162 minutes in the open group); however, this difference did not reach statistical significance ( $p = 0.41$ ). Similarly, mean length of hospital stay was reduced in laparoscopic patients (3.2 vs. 4.4 days;  $p = 0.13$ ). Complication and reoperation rates were comparable between groups, with two cases of wound infection in each group, both associated with patients presenting higher body mass index (BMI). This difference was not statistically significant ( $p = 0.19$ ). Correlation analysis revealed a weak trend toward longer operative times with increasing patient age ( $r = 0.43$ ;  $p \approx 0.06$ ). No significant associations were observed between operative time and length of stay, nor between age and hospital stay.

**CONCLUSION:** Although no statistically significant differences were identified between the open and laparoscopic approaches to the Malone procedure, the laparoscopic technique was associated with shorter operative times and reduced hospital stay, which may represent clinically relevant advantages. Both techniques demonstrated comparable safety profiles and similar complication rates. Larger studies are needed to confirm these trends.

## WHY STANDARDIZATION MATTERS: A PATIENT ORGANISATION'S PERSPECTIVE ON LIFELONG CARE

M. Mohideen, M. Neumayr  
(Vienna, Austria)

**PURPOSE:** Anorectal Malformations (ARM) and Hirschsprung Disease (HD) are rare congenital conditions requiring complex surgical treatment and long-term multidisciplinary care. SoMA Austria, founded in 2020, represents affected individuals and families in Austria and advocates for improved long-term outcomes, coordinated care and patient support. This presentation highlights why surgical standardization and structured lifelong care are essential from a patient organisation's perspective.

**METHODS:** Drawing on patient advocacy experience, peer support activities and collaboration with paediatric surgical centres, this presentation examines how healthcare structures, surgical standardization and long-term follow-up influence patient outcomes and quality of life.

**RESULTS:** Access to structured, guideline-based care in designated centers of expertise is a key determinant of outcomes. Standardized terminology and clear, consensus-based descriptions of surgical procedures are not only matters of scientific precision. They ensure that clinicians are truly talking about the same procedures, applying comparable standards, and evaluating outcomes in a consistent way across centers.

Surgical quality has lasting consequences for functional outcomes, particularly continence. These outcomes shape everyday life, influencing physical health, psychological well-being, and social participation. Difficulties may affect self-esteem, relationships, family planning, and career opportunities.

Because of this, clear terminology and shared standards in specialized centers are essential for improving care. They allow results to be compared, practices to be refined, and—most importantly—help ensure that patients receive the best possible treatment.

**CONCLUSION:** From a patient advocacy perspective, standardization in terminology, surgical techniques and care pathways is fundamental for equitable and high-quality care. Centralised expertise, specialised surgical training and structured interdisciplinary follow-up into adulthood are essential to improve long-term outcomes and support affected individuals in leading self-determined and socially integrated lives.

**MOSAIC PROMOTER DELETIONS REVEAL A COMMON SPORADIC HIRSCHSPRUNG DISEASE SUBTYPE AND ENABLE BLOOD-BASED MOLECULAR STRATIFICATION**

H. Xia, Y. Zhu, X. Zuo, Y. Zhang  
(Guangzhou, China)

**PURPOSE:** Hirschsprung disease is a congenital enteric neuropathy with distal aganglionosis and dysmotility. Germline mutations explain many familial cases, but most sporadic cases lack a clear molecular explanation. We aimed to identify a previously unrecognized mechanism for sporadic disease and develop a blood-accessible biomarker for molecular stratification.

**METHODS:** We performed RNA sequencing on paired aganglionic and ganglionic colon biopsies from 103 sporadic Hirschsprung disease patients, with control colons (n=22). Blood whole-genome sequencing was available for 41 profiled patients. Candidate regulators were assessed in patient tissues, functionally perturbed in zebrafish and mice, and mechanistically profiled in knockout induced pluripotent stem cell-derived enteric neural crest cells. Mosaic promoter deletions were called from colon whole-genome sequencing (n=30; matched blood n=27) and summarized as a blood mosaic promoter deletion score, evaluated in blood sequencing cohorts (discovery: 89 patients/43 controls; validation: 165 patients/42 controls), alone and combined with polygenic risk score.

**RESULTS:** A predominant subgroup (82/103, 79.6%) showed coordinated repression of enteric neurogenesis programs. Promoter motif enrichment implicated PRDM9, which localized to the normal enteric nervous system but was downregulated in aganglionic tissue with promoter hypermethylation. Loss of this factor reduced enteric neuron output and impaired gut motility in zebrafish and mice. In enteric neural crest cells, knockout redistributed genome-wide double-strand breaks toward promoters and enhancers, generated mosaic promoter deletions at neurogenesis loci, and correlated with transcriptional repression and impaired neuronal differentiation. The blood mosaic promoter deletion score discriminated cases from controls (area under the receiver operating characteristic curve 0.78 discovery; 0.82 validation) and improved with polygenic risk score (0.89 and 0.91, respectively).

**CONCLUSION:** These data link ectopic genome breaks to mosaic promoter deletions and repression of enteric neurogenesis in a prevalent sporadic Hirschsprung disease subtype, and support complementary, noninvasive molecular stratification using a blood-derived score.

## NEURONAL MARKER EXPRESSION PROFILES IN GANGLION CELLS OF THE ENTERIC NERVOUS SYSTEM IN HIRSCHSPRUNGS DISEASE

C. Nebeling, M. Lee-Theilen, E. Gradhand, F. Friedmacher, U. Rolle, Y. Braun  
(Frankfurt, Germany)

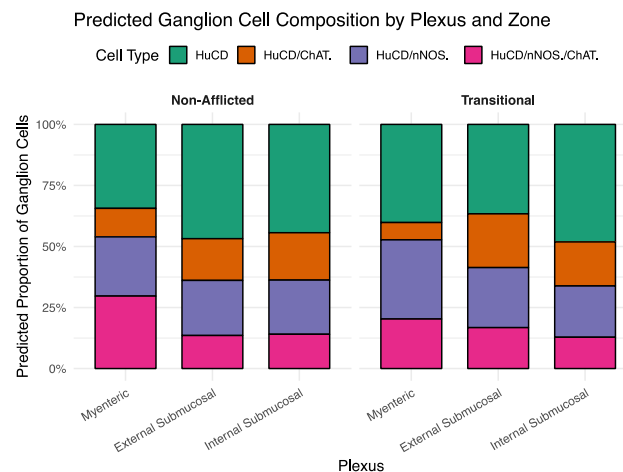
**PURPOSE:** This study aimed to characterize the expression of neuronal markers in the enteric nervous system (ENS) of patients with Hirschsprung's disease (HD). Specifically, it investigated marker expression across different bowel regions (aganglionic, transitional zone, and non-afflicted) and ENS components (myenteric, external and internal submucosal plexuses) to determine whether expression profiles correlate with specific areas.

**METHODS:** Eight patients who underwent transanal endorectal pull-through for histologically confirmed HD between 2022 and 2024 were included. Full-length antimesenteric sections were stained immunohistochemically for HuC/D, p75, VIP, ChAT, and nNOS. Combined immunofluorescent staining for DAPI, HuC/D, nNOS and ChAT was additionally performed. Slides were evaluated by digital pathology and marker profile expression was analyzed using a Bayesian multinomial regression model.

**RESULTS:** All neuronal markers were detected in the analyzed specimens. Nerve fibers and ganglionic cells consistently stained positive for p75, VIP, and nNOS across all regions. HuC/D and ChAT expression was restricted to ganglionic cells. Although ganglion cell marker expression did not differ between the transitional zone and unaffected bowel, the distribution of ganglion cells between the myenteric and submucosal plexuses varied, with a higher number of HuCD<sup>+</sup>/ChAT<sup>+</sup>/nNOS<sup>+</sup> cells observed in the myenteric plexus (**Fig. 1**).

**CONCLUSION:** While neuronal marker expression profiles differ between plexuses they are consistent between ganglionic cells of the transitional zone and non-afflicted bowel. These findings suggest that marker expression is uniform across these regions, irrespective of the bowel area affected by HD.

**Fig. 1.** Stacked bar plots depicting the estimated distribution of neuronal marker expression in ganglion cells by plexus and zone using a Bayesian Dirichlet–multinomial regression model.



## TRANSCRIPTOMIC CHARACTERIZATION OF HIRSCHSPRUNG DISEASE AND THE SCREENING AND VALIDATION OF DIAGNOSTIC MOLECULAR BIOMARKERS

D.B. Li, S. Sun, S. Zheng, C. Shen  
(Shanghai, China)

**PURPOSE:** This study aimed to screen objective and quantifiable molecular biomarkers based on transcriptomic profiles to aid recognition of HSCR aganglionic segments and to provide evidence for intraoperative evaluation of resection margins.

**METHODS:** Ten stenotic-segment samples from HSCR patients were included as the Aganglionic group. Nine proximal ganglionic-segment samples from HSCR patients and four colonic samples from non-HSCR controls were included as the Ganglionic group. Bulk RNA sequencing and differential expression analysis were performed to identify upregulated/downregulated genes and potential biomarkers. The discriminatory performance of candidate biomarkers was evaluated using receiver operating characteristic (ROC) curves. Key biomarkers were further validated by two-dimensional immunofluorescence staining and three-dimensional tissue-clearing imaging.

**RESULTS:** Compared with the Ganglionic group, 449 downregulated and 172 upregulated genes were identified in the Aganglionic group. Downregulated genes were mainly enriched in pathways related to synaptic structure and function, neurotransmitter signaling, and ion channels, whereas upregulated genes were primarily associated with protein assembly and steroid/lipid biosynthesis. Multiple genes showed strong discriminatory ability for identifying aganglionic segments. Representative downregulated biomarkers included VIP, PIRT, PHOX2B, CPLX2, SULT4A1, NEFL, NSG2, CACNA1B, KIF5A, and ELAVL4; representative upregulated biomarkers included MIR3648-1, AC087477.5, MSMO1, SQLE, MIR3648-2, RNA5-8SN2, RCC2P6, RUSC1-AS1, AC025918.1, and ALDH1A2. In validation using an HSCR mouse model and clinical tissues, VIP clearly revealed differences in the distribution of inhibitory neural networks within the intestinal wall in both 2D and 3D imaging, effectively distinguishing aganglionic segments from control segments.

**CONCLUSION:** This study systematically identified a panel of candidate molecular biomarkers with high diagnostic discriminatory power based on HSCR transcriptomic profiles, and specifically validated the potential utility of VIP for recognizing aganglionic segments. These findings provide new molecular evidence supporting objective adjunctive diagnosis and resection-margin assessment in HSCR.

## THE NERVE TRUNKS IN RECTOSIGMOID HIRSCHSPRUNG DISEASE: IS 40 MICRONS A RELIABLE PARAMETER FOR DEFINING THE TRANSITION ZONE?

S. Montedonico, L. Navarrete, J. Bracchiglione, P. Ferrer-Rosende, P. Poggi  
(Viña del Mar, Chile)

**PURPOSE:** A complete resection of the transition zone in Hirschsprung disease (HD) is critical for obtaining optimal clinical outcomes. A number of authors have described the presence of submucosal hypertrophic nerves >40 microns as indicative of transition zone. The aim of our study was to analyze the number of nerve trunks over 40 microns on the three histological zones in HD.

**METHODS:** Ten rectosigmoid HD were operated on with the endorectal Soave technique. Resected specimens were processed using the Swiss roll technique, enabling detailed morphometric histological analysis throughout the entire length of the resected intestine. For each millimeter (mm) of the samples, we measured the maximum transverse diameter of each nerve trunk. We applied a nonlinear mixed-effects sigmoidal model to represent the gradual change in this histological parameter along the specimen. We measured the median total number of nerve trunks over 40 microns and the median number of nerve trunks over 40 microns per mm in the aganglionic zone, the transition zone and the ganglionic zone.

**RESULTS:** The median total number of nerve trunks over 40 microns were 33 (16.0-54.8) in the submucous and 17 (2.0-31.8) in the myenteric plexus of the entire aganglionic zone; 6.5 (6.0-15.2) in the submucous and 7.0 (5.2-10.5) in the myenteric plexus of the transition zone and zero trunks in the proximal ganglionic zone. The median number of nerve trunks over 40 microns per mm were: 0.98 (0.85-1.22) in the submucous and 1.26 (0.83-1.48) in the myenteric plexus of the aganglionic zone; 0.08 (0.08-0.25) in the submucous and 0.24 (0.18-0.42) in the myenteric plexus of the transition zone and zero trunks in the proximal ganglionic zone.

**CONCLUSION:** Hypertrophic nerve trunks over 40 microns are absent in the ganglionic bowel and are extremely infrequent in the transition zone of rectosigmoid HD.

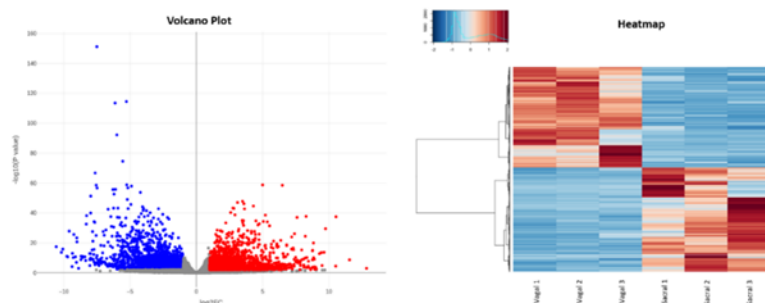
## TRANSCRIPTOMIC SIGNATURES OF VAGAL AND SACRAL ENTERIC NEURAL CREST CELLS IN MICE

N. Nakazawa-Tanaka, N. Fujiwara, K. Miyahara, R. Sueyoshi, T. Okazaki, G. Miyano  
(Chiba, Japan)

**PURPOSE:** The enteric nervous system (ENS) is primarily derived from vagal enteric neural crest cells (ENCCs), while sacral ENCCs contribute only slightly to the distal hindgut. In Hirschsprung disease, vagal ENCCs fail to complete their caudal migration, resulting in distal intestinal aganglionosis. Although sacral-derived nerve fibers increase within the aganglionic segment, sacral ENCC-derived ganglion cells are not observed. The molecular differences between vagal and sacral ENCCs that may underlie these contrasting behaviors remain poorly understood. Therefore, this study aimed to characterize the transcriptomic differences between vagal and sacral ENCCs in a mouse model.

**METHODS:** Sox10-Venus transgenic mice, in which ENCCs are fluorescently labeled, were harvested at embryonic day 18.5. Vagal ENCCs were isolated from the small intestine, and sacral ENCCs were obtained from the pelvic plexus (n = 3 embryos per group). Dissociated cells were cultured to generate neurospheres. Total RNA was extracted and subjected to bulk RNA sequencing using the Illumina NovaSeq X Plus platform. Differential gene expression analysis was performed using DESeq2, followed by Gene Ontology (GO) enrichment analysis.

**RESULTS:** RNA sequencing identified 3,913 differentially expressed genes between vagal and sacral ENCCs, including 1,971 genes upregulated and 1,942 downregulated in sacral ENCCs compared with vagal ENCCs (**Figure**). GO enrichment analysis demonstrated that sacral ENCCs showed significant enrichment of pathways related to extracellular matrix organization and cell-matrix adhesion, suggesting a phenotype strongly influenced by the surrounding microenvironment. In contrast, gene categories associated with neuronal differentiation, synaptic function, and axon development were less represented in sacral ENCCs.



**CONCLUSION:** This study demonstrates distinct molecular signatures between vagal and sacral ENCC populations. Sacral ENCCs appear to exhibit stronger extracellular matrix interactions and reduced neuronal differentiation compared with vagal ENCCs. These findings provide new molecular insights into the biological differences between these neural crest populations and may help explain why sacral ENCCs fail to compensate for the absence of vagal ENCCs in Hirschsprung disease.

## IMPAIRED FETAL ENTERIC NEURODEVELOPMENT IN A MOUSE MODEL OF INTESTINAL NEURONAL DYSPLASIA

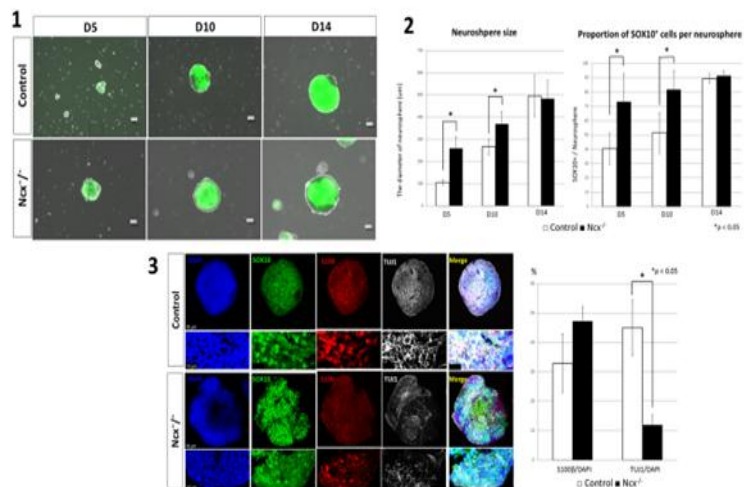
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(Tokyo, Japan)

**PURPOSE:** Intestinal Neuronal Dysplasia (IND) is an enteric neuropathy associated with severe bowel dysmotility and symptoms that clinically resemble those of Hirschsprung's disease, but the developmental timing of the underlying abnormality remains unclear. To investigate whether such abnormalities emerge during fetal life, we established a novel Sox10-*VENUS*-positive, *Ncx*-knockout mouse model that enables fluorescent identification of enteric neural crest-derived cells (ENCCs) and used it to examine early neuronal differentiation and enteric neurodevelopment.

**METHODS:** ENCCs were isolated from embryonic day 13.5 gut of knockout mice and wild-type littermates (n=6/group) and cultured under non-adherent conditions for 14 days to generate neurospheres. Neurosphere diameter and the proportion of SOX10-positive cells were quantified on days 5, 10, and 14 to evaluate early progenitor expansion and maintenance. On day 14, neuronal and glial differentiation was assessed by immunofluorescence staining for TUJ1 and S100 $\beta$ , and overall neurosphere morphology was compared between groups. Differences were considered statistically significant at p<0.05.

**RESULTS:** Knockout neurospheres were significantly larger than controls on day 5 (259.6 $\pm$ 52.0  $\mu$ m vs 105.4 $\pm$ 13.1  $\mu$ m, p=0.0103) and day 10 (369.6 $\pm$ 56.4  $\mu$ m vs 267.0 $\pm$ 34.9  $\mu$ m, p=0.0315), but not on day 14 (**Figure 1**). The proportion of SOX10-positive cells was also higher in knockout cultures on day 5 (73.2 $\pm$ 19.8% vs 40.5 $\pm$ 11.0%, p=0.0201) and day 10 (81.8 $\pm$ 13.1% vs 51.5 $\pm$ 14.2%, p=0.0202) (**Figure 2**), suggesting expansion or persistence of immature neural crest-derived cells. By day 14, knockout neurospheres showed significantly fewer TUJ1-positive cells than controls (11.9 $\pm$ 3.4% vs 45.0 $\pm$ 9.5%, p=0.029), whereas S100 $\beta$ -positive cell proportions were not significantly different. In addition, knockout neurospheres lost normal spheroid architecture, appeared structurally disorganized, and showed heterogeneous neuroglial distribution (**Figure 3**), supporting delayed or defective neuronal differentiation rather than a simple increase in cell numbers.

**CONCLUSION:** These findings indicate that *Ncx* deficiency disrupts fetal enteric neurodevelopment, resulting in a persistent precursor cell-dominant state, impaired neuronal differentiation, and compromised three-dimensional spheroid organization, which likely underlie the postnatal ganglion hyperplasia and dysmotility observed in this model.



## DEVELOPMENT OF THE JPCC BARRIERS TO PEDIATRIC COLOSTOMY CARE SCORING SYSTEM A MODIFIED DELPHI STUDY

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**PURPOSE:** In low- and middle-income countries, children with colorectal conditions frequently require colostomies and face substantial clinical, social, and financial barriers to stoma care. This study aimed to identify key barriers to pediatric colostomy care from the perspectives of caregivers and health care professionals in Southern Africa and to develop a scoring system to quantify these barriers.

**METHODS:** A modified Delphi study was conducted including caregivers of children with colostomies attending the Johannesburg Pediatric Colorectal Clinic (JPCC) and health care professionals managing pediatric stomas in South Africa, Namibia, and Zimbabwe. Purposive sampling was used to invite 20 caregivers and 20 healthcare professionals; participants completed two REDCap survey rounds, followed by a virtual expert panel meeting. In Round 1, participants listed at least six barriers to colostomy care, which three authors independently thematically grouped into barrier statements. In Round 2, participants rated each statement as: “relevant”, “relevant but needs editing”, “not relevant”, with consensus defined as a rate of agreement of at least 75. An expert panel of eight healthcare professionals then refined wording, finalized domains, and agreed on response options.

**RESULTS:** Of 40 invited individuals, 23 (57.5%) participated in Round 1 (12 healthcare professionals: 9 doctors, 2 nurses, 1 unknown profession; and 11 caregivers), generating 134 individual barriers that were consolidated into 35 statements across five domains (stoma products, stoma care, social challenges, stoma care training, and financial challenges). Sixteen participants completed Round 2, and all 35 statements reached the predefined consensus, with adjusted relevant ratings ranging from 75 to 100. During Round 3, the expert panel reviewed and reworded statements to improve clarity and neutrality, resulting in a final 36-item JPCC barriers to colostomy care scoring system. The panel agreed on a three-point Likert response scale (1 “not a challenge”, 2 “a moderate challenge”, 3 “a significant challenge”), with a “not applicable” option where appropriate.

**CONCLUSION:** This study presents the first scoring system specifically designed to capture and quantify barriers to pediatric colostomy care in a limited-resource context. It offers a practical framework for clinical assessment, research, and advocacy. Multicenter validation and cross-cultural adaptation are now needed.

## "SOAVE TO SWENSON" PULL-THROUGH FOR HIRSCHSPRUNG DISEASE IN INFANTS: POSTOPERATIVE OUTCOMES IN A RETROSPECTIVE COHORT

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**PURPOSE:** Hirschsprung disease (HSCR) requires pull-through surgery, yet postoperative Hirschsprung-associated enterocolitis (HAEC) and functional morbidity remain major concerns. We developed a hybrid "Soave to Swenson" (StS) technique designed to enable complete aganglionic resection while minimizing pelvic nerve/urethral injury and avoiding a long residual rectal muscular cuff.

**METHODS:** We performed a single-center retrospective cohort study of infants (1–12 months) undergoing one-stage pull-through for short- or long-segment HSCR from January 2017 to December 2024. Patients receiving traditional Soave pull-through (2017–2020) were compared with those receiving StS pull-through (2021–2024). The primary outcome was first HAEC within 24 months. Kaplan–Meier analysis with log-rank testing and Cox regression (adjusted for segment length, age at surgery, and sex) were used. Secondary outcomes included perioperative recovery metrics, early complications, and bowel/urinary function at 3–4 years (BFS, POFC, DVSS). Continuous variables were analyzed with Student's t test or the Mann–Whitney U test, as appropriate, and categorical variables were compared using the chi-square test or Fisher's exact test.

**RESULTS:** A total of 334 infants were included (Soave, n=184; StS, n=150). StS was associated with a lower 24-month cumulative incidence of first HAEC (9.71% vs 18.87%; log-rank P=0.020) and reduced risk on adjusted analysis (aHR=0.45, 95% CI 0.24–0.84; P=0.013). Operative time and length of stay were similar, but StS patients started oral feeding earlier (1.37±0.74 vs 1.67±0.90 days; P<0.001), had shorter urinary catheter duration (2.99±1.64 vs 4.35±1.63 days; P<0.001), and required a shorter duration of postoperative anal dilation (3.29±0.57 vs 4.05±0.99 months; P<0.001). Early complications were comparable. At 3–4 years (n=176), StS showed higher continence-related outcomes (POFC=10.85±1.66 vs 10.24±1.53; P=0.018) with similar DVSS.

**CONCLUSION:** In infants with HSCR, the StS pull-through was associated with a lower risk of HAEC within 24 months and signals of improved continence without increased perioperative morbidity or urinary dysfunction. These findings support prospective, multicenter validation with longer-term functional and quality-of-life follow-up.

## INFLAMMATORY BOWEL DISEASE IN THE FIRST TEN YEARS OF LIFE: THE SURGICAL REALITY

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(London, UK)

**PURPOSE:** Very Early-onset (VEO) and Early-onset (EO) of inflammatory bowel disease (IBD) are distinct clinical entities. Surgical indications and outcomes are poorly understood. Clinical features and interventions at a tertiary center are compared.

**METHODS:** Retrospective review of children < 10yrs with IBD (2004-2025). Patients were divided into Group 1=VEO-IBD (<6 years) and group 2=EO-IBD (6–9 years) as per Paris classification. Demographics and clinical features were collected. Fishers exact test was used for statistical analysis and data presented as median and interquartile (IQR) ranges.

**RESULTS:** 91 patients (48M:43F). Group 1=42 (46%), median age at diagnosis was 3.85 (2.6-4.9) years. Group 2=49 (54%), median age at diagnosis was 8 (7-9) years. Male predominance was observed overall, more in group 1. Most patients were white British, reflecting demographic distribution of referral population. Atypical features noted in group 2 that included oral ulcers, cheek and jaw swelling and severe anemia. Five (4F:1M) required surgery for perianal Crohn's. Group 1[(n= 3)(6 years at surgery)]; two had seton placements, one had a colostomy. Group 2 [(n= 2)(11 years)]; both required ileostomies. Only one had a family history of IBD. There is no significant association between gender and surgery requirement (p=0.09). Family history and age at diagnosis were not surgical predictors either. The mean follow up for the surgical patients in group 1 was 11 (4-11) years and the median in group 2 was 6.5 years.

	Group 1	Group 2	P value
Number (%)	47%(51)	53%(40)	
Age at diagnosis-years (IQR)	3.85%(2.6-4.9)	8 %(7-9)	
Male proportion %	68%(32)	56%(22)	
Crohn's (%)	43%(22)	52%(20)	
IBD unclassified (%)	19%(9)	24%(9)	
Positive family history (%)	15%(7)	16%(6)	1.0
Associated syndromes (%)	11%(5)	12%(4)	1.0
Rectal bleeding/diarrhea (%)	57%(29)	72%(28)	0.4

### CONCLUSION:

1. Perianal phenotype in Crohn's was associated with higher risk of surgical intervention.
2. Atypical presentation and unclear clinical predictors underscores the importance of early surgical input and careful selection for surgery.

## **THE BOTTOM LINE: DOES ALL POO BELONG IN THE LOO?**

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**PURPOSE:** Neurodiversity is well known to be associated with significantly high rates of gastrointestinal problems, including idiopathic chronic constipation (CCFI) with or without faecal incontinence (1). These patients are often challenging when deciding on treatment modalities. We aim to ask the question: Is it beneficial for these complex neurodiversity patients to poo out of their bottom?

**METHODS:** Retrospective data for all our patients from January 2023 to January 2025 were included. 18 had CCFI with neurodiversity: male predominance (50%) and median age 11 years (range: 4-18 years). Our multi-disciplinary team consists of: paediatric surgeons, gastroenterologist, and clinical nurse specialist, and clinical scientist, psychologist, safeguarding team, health play specialist and radiologist. The team manages these patients and decisions are made with a multi-professional approach.

**RESULTS:** Patients challenges prior to having a stoma included: persistent symptoms and failed conservative (laxatives, enemas) and invasive (anal irrigation, transcutaneous electrical stimulation, antegrade continence enema) treatments, interoception difficulties, trouble expressing the need to defaecate (understanding rectal sensation), poor compliance or cognitive understanding of toileting regimes and debilitating impact on quality of life.

**CONCLUSION:** From our specialised service we found that:

1. Passing a bowel motion in the toilet is not for everyone.
2. Tailor patient care to the individual.
3. Diagnostic tools (such as anorectal physiology, transit x-rays, neurodiversity assessment) are useful to guide management.
4. Multi-professional approach to these patients is vital, due to the multifactorial nature of their condition.
5. Improved quality of life for patient and their family.

## WHY USE HIGH RESOLUTION ANORECTAL MANOMETRY (HRAM) AND ENDOANAL ULTRASOUND IN CHILDREN?

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(London, UK)

**PURPOSE:** The pathophysiology of childhood defaecation disorders (constipation, faecal incontinence, Hirschsprung disease, anorectal malformation, other [tumour, trauma]) remains unclear. Improved physiological evaluation offers the ability to describe abnormal findings in greater detail. Clinical history and comprehensive examination remains the mainstay of clinical diagnosis in these children. Although HRAM is slowly becoming recognised (1), there are currently no agreed guidelines and thus a risk that will lead to diversity in practice. The British Society of Paediatric Gastroenterology, Hepatology and Nutrition (BSPGHAN)—Motility Working Group (MWG) has published guidance on the use of HRAM in children with defaecation disorders (2). Our aim was to present the reasons why HRAM is useful.

**METHODS:** Our specialised service opened in 2016. Since then, our service has provided a unique, comprehensive, scientific and holistic approach to our patients. As a multi-disciplinary team, we have conducted 10 reasons why doing HRAM, is more than just a useful tool.

**RESULTS:** The 10 reasons:

### 10 reasons

1. Standard practice in adults in colorectal disease
2. Key physiological and structural information
3. Provides scientific and objective evidence in a condition that depends on parental reporting.
4. Continues to yield novel scientific insights
5. It's safe and well tolerated  
Objective assessment of dyssynergic defecation
6. HRAM allows to see emerging phenotypes
7. It provides reassurance for management decisions are correct (in an evidence vacuum).
8. Provides realistic expectations
9. Allows excellent outcomes

**CONCLUSION:** HRAM is becoming the investigation of choice for understanding the pathophysiology in children with defaecation disorders. It is clear, that conducting HRAM, in selective cases, enables us to gain information in a safe, scientific and beneficial manner. This tool is more than just a toy, but rather provides the missing piece the puzzle for the clinician, patient and their families.

## DOWN SYNDROME AND VACTERL ASSOCIATION IN ANORECTAL MALFORMATION

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**PURPOSE:** Down syndrome (DS) and VACTERL association are commonly associated with anorectal malformation (ARM). DS is a chromosomal disorder whereas the aetiology of VACTERL remains largely unknown. Despite overlapping organ and system involvement, no study has systematically compared these two conditions. We aimed to characterise the phenotypical difference between ARM patients with DS and VACTERL.

**METHODS:** Retrospective review of ARM database from a tertiary centre was carried out to identify all follow-up patients born between 2007 and 2024, to study all aspects of VACTERL association (defined as 3 or more features present). ARM with DS was compared with ARM with VACTERL for sex, ARM anomaly and systems involvement.

**RESULTS:** Among 92 ARM patients, 10 had DS (11%) and 24 had VACTERL association (26%). Only one patient had both DS and VACTERL. All DS had ARM with no fistula. Sex distribution of both DS and VACTERL were identical, and nearly half of both groups had cardiac abnormalities. However, there were significantly higher proportions of vertebral (46% vs. 0,  $P=0.014$ ), spinal (50% vs. 10%,  $P=0.032$ ) and renal anomalies (83% vs. 40%,  $P=0.034$ ) present in the cohort of ARM with VACTERL than DS. The numbers of system involvement was also higher in VACTERL than DS (3 vs. 2,  $P<0.0005$ ) (**Table**).

Associated anomalies	ARM + DS (N=10)	ARM + VACTERL (N=24)	P-value
Male sex	6 (60%)	14 (58%)	1
ARM with no fistula	10 (100%)	0	<b>&lt;0.001</b>
Vertebral	0	11 (46%)	<b>0.014</b>
Spinal	1 (10%)	12 (50%)	<b>0.032</b>
Anorectal	10 (100%)	24 (100%)	1
Cardiac	5 (50%)	13 (54%)	1
Tracheal	0	1 (4%)	0.706
Oesophageal	0	0	-
Renal	4 (40%)	20 (83%)	<b>0.034</b>
Limb	1 (10%)	6 (25%)	0.644
Median number of VACTERL features (range)	2 (2-3)	3 (3-5)	<b>&lt;0.0005</b>

**CONCLUSIONS:** The incidence of VACTERL in ARM was nearly twice as high as DS. Despite both were multisystem conditions with high predisposition for cardiac abnormalities, VACTERL has significantly higher vertebral, spinal and renal anomalies compared to DS. The differences in phenotype appear to be distinctly variable rather than simple overlap. Future research on the underlying divergent embryological mechanisms is warranted.

## **AUTISM SPECTRUM DISORDER IN ANORECTAL MALFORMATIONS AND HIRSCHSPRUNG'S DISEASE: SHOULD WE BE WORRIED?**

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**PURPOSE:** It has been proposed that children with congenital gastrointestinal malformations, including anorectal malformations (ARMs), might be at higher risk of neurodevelopmental disorders such as autism (ASD) and attention deficit hyperactivity disorder (ADHD). As current evidence is limited, we aimed to assess the incidence of ASD and ADHD among patients with ARM and Hirschsprung's Disease (HSCR) at our institution.

**METHODS:** We retrospectively reviewed all patients managed in our pediatric colorectal clinic with either ARM or HSCR in the period 2012-2025. Patients with known genetic syndromes causing neurodevelopmental delay and patients under 18 months of age were excluded.

The remaining cohort was evaluated for the presence of ASD or ADHD. Clinical, surgical and psychosocial characteristics were evaluated.

**RESULTS:** Following exclusion criteria, 79 patients (48 ARMs; 31 HSCR) were identified. The mean age was 7.2 years, with a male predominance (56%). ASD or ADHD was diagnosed in 7 patients (4 ARMs; 3 HSCR), resulting in an overall incidence of 8.8%. All but one of these patients were male. Three of the four ASD cases were diagnosed before age 3, suggesting an almost four-fold increase in early infantile autism compared to Austria's general population rate of 1%. The three ADHD cases were identified in school-aged children. All patients are engaged in individualized developmental and behavioral therapies.

**CONCLUSION:** Patients with ARM and HSCR appear to be at higher risk of ASD beginning in early infancy. As evidence suggests that early intervention can improve prognosis, awareness regarding early signs of ASD and ADHD should be raised amongst managing pediatric surgeons.

**PREDICTORS OF HIRSCHSPRUNG'S DISEASE IN CHILDREN UNDERGOING RECTAL BIOPSY: A RETROSPECTIVE COHORT STUDY**

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**PURPOSE:** Hirschsprung's disease (HD) is a congenital disorder defined by the absence of ganglion cells in the distal bowel. Because its clinical presentation often overlaps with functional constipation, rectal biopsy is required for definitive diagnosis. This study aimed to identify clinical and radiological factors positively or negatively associated with HD to improve diagnostic stratification.

**METHODS:** We performed a retrospective cohort study of all children who underwent full-thickness rectal biopsy at our tertiary pediatric surgery centre between 2010 and 2025. Demographic data, clinical presentation, perinatal history, and radiological findings were collected. Patients diagnosed with HD were compared with those with functional constipation. Penalized logistic regression was used to identify independent predictors of biopsy-proven HD. A predefined subgroup analysis was conducted for children older than six months.

**RESULTS:** A total of 108 patients were included; 60 (55.6%) were diagnosed with HD. A suspicious contrast enema was strongly associated with HD ( $p = 0.005$ ), whereas prematurity was independently associated with the absence of HD ( $p = 0.001$ ). In the subgroup of children older than six months ( $n = 47$ ), HD was confirmed in 20 patients (42.6%). In this subgroup, a suspicious contrast enema remained the only significant positive predictor of HD ( $p = 0.042$ ).

**CONCLUSION:** A suspicious contrast enema is a strong and independent predictor of HD in children undergoing rectal biopsy and the sole predictor in patients older than six months. Prematurity is independently associated with a lower likelihood of HD. These findings may support more targeted diagnostic pathways and reduce unnecessary invasive procedures.

## HIRSCHSPRUNG DISEASE IN LATIN AMERICA: A MULTINATIONAL SURVEY OF SURGICAL STRATEGIES AND FOLLOW-UP PRACTICES

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**PURPOSE:** Hirschsprung's disease (HD) is a congenital bowel disorder with a low prevalence. Current practice shows substantial heterogeneity in surgical approaches and long-term functional follow-up. In Latin America there are a lack of information of the treatment of this disease. Describing real-world practice may inform standardization and quality-improvement priorities. Aim of this study was to characterize surgical strategies, intraoperative resources, and follow-up practices for rectosigmoid HD and total colonic aganglionosis (TCA) across Latin-American centers.

**METHODS:** Cross-sectional study using a standardized electronic survey. Variables included country, specialty, and annual case volume; technique and approach for rectosigmoid HD; technique, reconstructive strategy, and timing for TCA; resources for anal exposure and vascular control; availability of pediatric anesthesia and use of neuraxial blockade; and follow-up practices.

**RESULTS:** 88 responses were analyzed, including surgeons from all Latin-American countries. Most respondents were pediatric surgeons (77%) and pediatric colorectal surgeons (18%). Reported annual volume was <5 cases/year in ~49%, 5–10 cases/year in ~38% and >10 cases/year 13%. For rectosigmoid HD, the reported techniques were Swenson (45%), Soave (33%), and Duhamel (22%).

Regarding the surgical approach for rectosigmoid disease, the most used were transanal (43%), open abdominoperineal laparotomy (29,5%), and laparoscopic assisted (26%). For TCA, respondents reported Swenson (47.7%), Duhamel (34%), and Soave (18,3%); the predominant reconstruction was straight ileorectal pull-through, but 11% perform a J-pouch. The most common timing for ileal pull-through was after achieving urinary continence (43.8%). Concerning follow-up, 74% reported >10 outpatient visits; 74% performed routine anal dilations with Hegar dilators. Most surgeons reported availability of pediatric anesthesia, although access to advanced intraoperative resources varied among centers

**CONCLUSION:** Despite the shared Spanish language in most of Latin-America pediatric surgeons and face similar financial hardship in their healthcare systems, current practice in HD in Latin-America shows marked variability in technique, approach, and follow-up practices, including key decisions for TCA (reconstructive strategy and timing). These findings support the need for regional consensus, minimum resource standards, and multidisciplinary functional follow-up models focused on long-term outcomes.

## TOTAL COLONIC AGANGLIONOSIS IN CHILDREN: LONG-TERM SURGICAL OUTCOMES AND QUALITY OF LIFE

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(New Delhi, India)

**PURPOSE:** Management of total colonic aganglionosis (TCA) remains challenging with limited data on long-term outcomes. This study aimed to evaluate surgical management, complications, and long-term functional outcomes in patients with TCA.

**METHODS:** A retrospective follow-up study was conducted in children with TCA. Surgical procedures performed, postoperative complications, stoma-related issues, and reinterventions were analyzed. Long-term outcomes included fecal continence, growth, anemia, and quality of life as perceived by patients and parents.

**RESULTS:** Nine children, median age at definitive surgery of 16 (9–96) months) were evaluated: two were female. Eight underwent total colectomy. Martin's modification of Boley-Scott pull-through was done in eight, while one underwent laparoscopy-assisted resection with Martin's modification of Duhamel ileal pull-through. Two patients underwent reversed isolated ileal segment anastomosis with the rectal stump, and one had a left colon patch followed by ileoanal anastomosis. Temporary ileostomy closure was achieved in eight patients after a median of 6 months (3–12 months). One child with Bardet–Biedl syndrome developed torrential bleeding following ileostomy closure, necessitating refashioning. The ileostomy closure was repeated but was followed by obstructive symptoms due to dysmotility and adhesive obstruction and had to be refashioned. At a median follow-up of 68 months (24–394 months), all patients had normal hemoglobin levels. One patient required re-ileostomy for adhesive obstruction, and another experienced prolonged anemia secondary to recurrent enterocolitis. Six of eight patients achieved social continence after stoma closure. Despite frequent hospitalizations and early failure to thrive, overall quality of life was satisfactory.

**CONCLUSION:** Although TCA is associated with significant early morbidity, sustained multidisciplinary care enables children and families to adapt, ultimately achieving acceptable continence and quality of life.

## CONGENITAL HYPOGANGLIONOSIS: PHENOTYPE-DEPENDENT OUTCOMES AND TEMPORAL EVOLUTION OF DIAGNOSTIC AND SURGICAL STRATEGIES: A SYSTEMATIC REVIEW

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**PURPOSE:** Congenital hypoganglionosis (CH) is a rare enteric neuropathy characterized by reduced and small ganglion cells in the myenteric plexus. Since the systematic review by Puri et al. in 2010, advances in diagnostic techniques have improved the management of CH. We aimed to systematically review the clinical outcomes and diagnostic evolution of CH in children.

**METHODS:** A systematic review was conducted in accordance with PRISMA guidelines using PubMed and Web of Science (1978–2025). Of 396 identified records, 24 duplicates were removed. After screening 372 records, 43 full-text articles were assessed for eligibility, and 32 studies including 298 pediatric patients were analyzed. Disease extent was strictly classified as diffuse/panintestinal (clearly stated), colon-limited, segmental, or unspecified. Extracted variables included age at presentation, staining methods (e.g., calretinin, S-100, quantitative HuC/D), surgical strategy, and mortality.

**RESULTS:** Among 298 patients, 144 (48%) were classified as diffuse/panintestinal, 25 (8%) as colon-limited, 22 (7%) as segmental, and 107 (36%) remained unspecified (**Table 1**). In studies reporting mortality for diffuse/panintestinal disease (n = 105), 28 deaths (26.7%) were documented. In contrast, no deaths were reported in colon-limited series (0/24) (**Table 2**). Mortality was mainly seen in the diffuse phenotype. Most diffuse cases presented in the neonatal period. Calretinin staining was mainly reported between 2010 and 2019 (14 patients). Quantitative HuC/D staining was reported only after 2020 (42 patients). This reflects increasing use of standardized diagnostic criteria. Surgical management also changed over time. Before 2009, colectomy was commonly performed. After 2010, treatment became more phenotype-based, with increased use of stoma formation and intestinal rehabilitation strategies.

**Table 1. Extent-Based Phenotype Distribution**

Extent	Number of Patients
Diffuse/Panintestinal	144
Colon-limited	25
Segmental	22
Unspecified	107

**Table 2. Mortality by Extent (Reported Only)**

Extent	Deaths / Reported	Mortality
Diffuse	28 / 105	26.7%
Colon-limited	0 / 24	0%

**CONCLUSION:** Diffuse or panintestinal neonatal forms are associated with high mortality, whereas colon-limited disease has good survival. Recent advances in quantitative diagnostic techniques, especially Hu C/D staining, have improved diagnostic accuracy and supported phenotype-based treatment. Future management should include extent-based stratification to improve outcomes in severe neonatal cases.

## DIAGNOSTIC AND THERAPEUTIC EXPERIENCE IN CHILDREN WITH ALLIED HD

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(Beijing, China)

**PURPOSE:** To summarize the clinical characteristics, diagnostic and therapeutic approaches of allied HD in children.

**METHODS:** A retrospective analysis was conducted on the data of 17 pediatric patients with allied HD who underwent surgical treatment in the Department of General Surgery of our hospital from January 2020 to June 2025. The clinical characteristics, diagnostic methods, surgical approaches, and prognosis were summarized.

**RESULTS:** Among the 17 cases, there were 6 males and 11 females, with onset ages ranging from 0 to 173 months. Three cases had confirmed delayed meconium passage, and clinical manifestations included constipation, abdominal pain, abdominal distension, vomiting, fever, etc. All patients complained constipation, and 9 school-aged cases suffered abdominal pain. Eight patients had undergone surgical treatments at other hospitals, including laparotomy exploration, enterostomy, colostomy closure, and radical surgery for HD. However, postoperative symptoms often showed no significant improvement. After admission, all patients underwent lower gastrointestinal contrast imaging and anorectal manometry. Lower gastrointestinal contrast imaging revealed abnormal colonic morphology, with significant prolapse of the transverse colon in the school-aged group, while some patients exhibited rigidity and stenosis in the left colon, with the distal rectum remaining normal. Anorectal manometry indicated the presence of anorectal inhibitory reflex but elevated resting anal pressure. Before radical surgery, rectal mucosal biopsy, full-thickness biopsy of colon, and ileostomy were performed. Radical surgery was conducted more than six months after the ileostomy, with the surgical approach determined based on biopsy pathology results and intraoperative colonic morphology, including partial colectomy, subtotal colectomy, total colectomy, or the Soave operation. Postoperative histopathological examination of resected colon revealed significantly reduced ganglion cells in the affected segments, small ganglion cell size, and hypoplasia of some ganglion cell. Follow-up periods ranged from 3 to 67 months, with 17 cases showing resolution of postoperative constipation and 8 cases experiencing relief from abdominal pain, along with improved nutritional status. Among them, 2 cases were prone to pseudo-intestinal obstruction, which could be alleviated with conservative treatment.

**CONCLUSION:** The diagnosis of allied HD in children requires a comprehensive evaluation of medical history, lower gastrointestinal contrast imaging, rectal mucosal biopsy, and histopathological examination of full-thickness colon biopsies. The extent of surgical resection should be determined based on intraoperative findings of colonic morphology and histopathological results. While bowel function improvement is satisfactory, vigilance is required for signs of pseudo-intestinal obstruction.

## THE FIRST PULL-THROUGH PROCEDURE IN 19TH CENTURY VIENNA

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**PURPOSE:** To examine the first described pull-through procedure in colorectal surgery within the context of nineteenth-century Viennese surgical practice.

**METHODS:** A review of published surgical literature was conducted alongside analysis of previously unreported archival documents relating to the earliest pull-through procedure performed in Vienna, Austria, during the late nineteenth century.

**RESULTS:** Analysis of original publications, operative reports, and contemporaneous surgical texts demonstrates that Julius von Hochenegg (1859–1940) performed and described the first documented pull-through procedure. His technique involved mobilisation of the bowel with preservation of the anal sphincter complex, anticipating later standardised approaches. Hochenegg's operative concepts prefigured key principles of modern pull-through surgery, including tension-free anastomosis and preservation of continence. Despite clear documentation of these innovations, subsequent surgical literature largely overlooked his contributions, with priority frequently attributed to later surgeons. This reassessment confirms Hochenegg's precedence in both the technical execution and conceptual development of sphincter-preserving colorectal surgery.

**CONCLUSION:** Julius von Hochenegg should be recognised as a foundational figure in the history of the pull-through procedure. His work in nineteenth-century Vienna established surgical principles that remain central to the management of Hirschsprung disease and modern colorectal surgery. Restoring his contributions helps correct the historical record and highlights the importance of Viennese surgery in the evolution of paediatric and colorectal surgical practice.

## COLOSTOMY WITH TRANSUMBILICAL APPROACH AS AN ALTERNATIVE IN PATIENTS WITH ANORECTAL MALFORMATIONS

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**PURPOSE:** When performing a conventional colostomy in patients with anorectal malformations (ARM), we observed postoperative complications involving the incision between the colostomy and the distal fistula, which represented a significant expense for families and the hospital. Our objective is to present our experience with an alternative to the conventional colostomy performed in patients with ARM to avoid these complications.

**METHODS:** Medical records of children with ARM treated at our hospital between 2019 and 2025 were retrospectively reviewed. We described postoperative complications between conventionally performed colostomies (oblique incision in the left flank of the abdomen) and those performed with a transumbilical approach with no incision between the stomas.

**RESULTS:** We studied 28 children, 12 girls and 16 boys. Twenty-three conventional colostomies (group 1) and five transumbilical colostomies (group 2) were performed. Postoperative complications were present in 8 patients (34.7%), including stoma stenosis (50%), skin dehiscence between the stomas (37.5%), surgical site infection (12.5%), stoma necrosis (12.5%), and aponeurosis dehiscence between the stomas (12.5%). In the second group, the complication rate was 0%. When performing transumbilical colostomy in girls, the internal genitalia were always checked without difficulty in visualizing them.

**CONCLUSION:** To date, performing transumbilical colostomies has been the best alternative for us, as it reduces complications and lowers costs. Unfortunately, the limitations we faced were the limited number of patients and the inability to access all medical records.

## PREOPERATIVE SERUM ALBUMIN AS A PREDICTOR OF WOUND HEALING IN PEDIATRIC ANORECTAL MALFORMATION REPAIR: A SINGLE-INSTITUTION STUDY

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**PURPOSE:** Serum albumin is a recognized predictor of surgical wound healing and success. Its role in the postoperative course following the surgical correction of anorectal malformations (ARM) has not yet been explored. We investigated whether preoperative albumin predicts wound healing disorders in children undergoing different operative stages of ARM repair.

**METHODS:** Preoperative serum albumin levels and postoperative wound complications were retrospectively reviewed for all patients who underwent correction of congenital ARM at our institution from January 2020 to December 2025. Patients were stratified into four groups based on the surgical procedure: (1) pre-colostomy, (2) pre-PSARP, (3) pre-intestinal reconstruction, and (4) pre-cloacal repair. Preoperative serum albumin (g/dL) was recorded within 7 days before the procedure. Descriptive statistics and a one-way ANOVA were performed to compare the groups.

**RESULTS:** Twenty-two patients (m: f = 14:8) were included, encompassing 49 total procedures. The mean albumin level across all stages was 3.89 g/dL. ANOVA analysis ( $F = 1.49$ ,  $p = 0.23$ ) showed no statistically significant difference between the mean albumin levels in the four groups: pre-colostomy ( $3.76 \pm 0.75$  g/dL), pre-PSARP ( $3.79 \pm 0.57$  g/dL), pre-intestinal reconstruction ( $4.20 \pm 0.48$  g/dL), and pre-cloacal repair ( $4.01 \pm 0.72$  g/dL). Wound complications occurred in 18.18% of patients ( $n=4$ , f:m = 2:2). These included one perineal breakdown after rectovestibular fistula and two surgical site infections occurring after cloacal repair and colostomy closure. Although mean albumin levels were slightly lower in patients with complications ( $3.95 \pm 0.39$  g/dL) compared to those without ( $4.03 \pm 0.57$  g/dL), there was no statistically significant difference in albumin levels between the two groups.

**CONCLUSION:** In our cohort, preoperative serum albumin levels were not a significant predictor of wound healing disorders in patients undergoing ARM repair. While the limited sample size precludes the definitive exclusion of a correlation between low preoperative albumin and impaired healing, the current findings do not justify routine preoperative albumin substitution for elective ARM repair.

## **BOWEL AND URINARY OUTCOMES IN SPINAL ANOMALIES ASSOCIATED WITH ANORECTAL MALFORMATION: INSIGHTS FROM 18 YEARS OF EXPERIENCE**

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**PURPOSE:** This study compared the bowel and urinary outcomes in spinal anomalies associated with anorectal malformation (ARM) managed either conservatively or with proactive spinal surgery.

**METHODS:** All patients with ARM born in 2007-2024 in a tertiary centre were retrospectively reviewed. Spinal anomalies were identified through VACTERL screening and categorised according to treatment: conservative management or spinal untethering surgery. Bowel outcomes (Krickenbeck bowel function score) and urinary outcomes were analysed. Continuous variables were reported as median (range) and analysed using non-parametric methods.

**RESULTS:** Fourteen of 92 ARM had associated spinal anomalies (15%). Half underwent spinal surgery, while the remainder were managed conservatively (n=7 each). Surgical interventions were performed proactively at 19 months (6-76) despite paucity of neurological symptoms or signs, based on key radiological findings on MRI, including syrinx, lipoma and fatty filum associated with a low-lying cord. Patients managed conservatively demonstrated radiological features suggestive of a low-lying cord but lacked clinical or radiological signs of tethered cord syndrome. Median follow-up ages for the surgical and conservative cohorts were 49 (8-129) and 62 (24-166) months, respectively (P=0.535).

High-type ARM was the most common anomaly (surgical 57% vs. conservative 71%, P=0.80). All patients achieved voluntary bowel movement (100%). A higher proportion of the conservative cohort experienced soiling (43% vs. 0, P=0.148), although not statistically significant. Constipation occurred in three conservatively managed patients and four surgical patients (P=0.788) with equal proportions requiring enemas (28%).

Renal anomalies were comparable between the conservative and surgical cohorts (71% vs 57%, P=0.577). Urinary outcomes were also similar: lower urinary tract syndrome (28% vs. 43%, P=0.577), urinary tract infection (14% each), urinary incontinence (0 vs. 29%, P=0.127), requirement for clean intermittent catheterisation (29% each) and urinary tract dilation (29% vs. 57%, P=0.280).

**CONCLUSION:** Half of the patients with spinal anomalies associated with ARM underwent spinal surgery. A proactive surgical approach for tethered cord release was not associated with procedural complications. Bowel and urinary outcomes were comparable between patients managed conservatively and managed with proactive surgery. Careful identification of key radiological feature on spinal MRI in asymptomatic patients, combined with appropriate caregiver counselling, is essential to guide evidence-based management.

## LONG-TERM BOWEL FUNCTION AFTER SURGERY FOR HIRSCHSPRUNG'S DISEASE: COMPARATIVE EVALUATION OF ANORECTAL MANOMETRY AND KRICKENBECK FUNCTIONAL OUTCOMES

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**PURPOSE:** Despite definitive surgery for Hirschsprung's disease (HD), a proportion of patients continue to experience persistent bowel dysfunction, including constipation and fecal incontinence. This study aimed to evaluate long-term postoperative bowel function using anorectal manometry (AM) and the Krickenbeck criteria, and to compare functional outcomes between surgical approaches.

**METHODS:** A cross-sectional study was conducted in children who underwent definitive surgery for HD between 1 January 2014 and 31 December 2018. Postoperative bowel function was evaluated using anorectal manometry parameters and the Krickenbeck criteria for fecal continence. Functional outcomes were compared between operative techniques and with a control group.

**RESULTS:** Sixteen postoperative HD patients (15 males) and 18 controls (16 males) were included. Among the HD cohort, 7/16 (43.8%) underwent transanal endorectal pull-through (TERPT) and 9/16 (56.2%) underwent an abdomino-perineal (Scott–Boley) pull-through. Primary pull-through without stoma diversion was performed in 13/16 (81.25%) patients, whereas 3/16 (18.75%) underwent staged repair.

The urge to defecate was present in all patients, while the capacity to verbalize the need to defecate and the ability to voluntarily hold bowel movements were present in 15/16 (93.75%) and 12/16 (75%) patients, respectively. At a median follow-up of 40 months, the relative risk of soiling following TERPT was 1.5 (95% CI 0.945–2.38) compared with the Scott–Boley procedure.

Anorectal manometry demonstrated significantly lower mean resting anal pressure ( $p = 0.001$ ) and mean maximum squeeze pressure ( $p = 0.013$ ) in HD patients compared with controls. However, there was no statistically significant difference between TERPT and Scott–Boley procedures in mean resting anal pressure ( $p = 0.785$ ) or mean maximum squeeze pressure ( $p = 0.489$ ).

**CONCLUSION:** This study highlights persistent anorectal physiological abnormalities in postoperative Hirschsprung's disease despite satisfactory clinical outcomes. Soiling was more frequent following TERPT, whereas constipation was more common after the Scott–Boley procedure. The findings suggest that postoperative bowel dysfunction patterns differ between TERPT and Scott–Boley procedures, while anorectal manometric parameters remain largely independent of the surgical approach.

## MANOMETRY AFTER ANORECTAL MALFORMATION SURGERY: PREDICTING OUTCOMES AND GUIDING MANAGEMENT

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**PURPOSE:** Long term follow up after surgery for anorectal malformations (ARM) is essential in pediatric colorectal care. Anorectal manometry provides objective assessment of sphincter configuration and function, particularly resting and squeeze pressures and sphincter symmetry. This study evaluated manometric parameters and their association with clinical characteristics and functional outcomes.

**METHODS:** We performed a retrospective review of all ARM patients who underwent water perfused anorectal manometry between 01/2009 and 12/2021. Parameters included resting/mixed and squeeze pressures, sphincter asymmetry, and rectal balloon volume at first sensation. These were correlated with ARM type, comorbidities, and functional outcome at last follow up using the Bowel Function Score (BFS). Statistical analyses included Spearman's rank correlation, ROC analysis, logistic and multiple regression, Chi square testing, and calculation of predictive values. Ethical approval was obtained.

**RESULTS:** Seventy-five patients (118 manometries) were analyzed. Mean resting or mixed pressures at pull through maneuver correlated with ARM severity ( $r = -0.25$ ,  $p = .011$ ) and BFS ( $r = 0.32$ ,  $p = .004$ ) and moderately predicted acceptable outcomes (AUC 0.60; OR 8; sensitivity 92.3%; specificity 40%). Rectal balloon volume correlated with BFS at the time of measurement ( $r = -0.28$ ,  $p = .047$ ). In children under four, inter quadrant asymmetry correlated with later BFS ( $r = -0.37$ ,  $p = .036$ ) but lacked predictive strength. Multiple regression identified resting pressure, ARM severity, neurodevelopmental delay, and spinal abnormalities—but not asymmetry—as independent correlates of BFS ( $R^2 = 0.61$ ,  $p < .001$ ). Other asymmetry measures and squeeze pressures showed associations with age but not with outcomes.

**CONCLUSION:** Anorectal manometry is a valuable adjunct in ARM follow up. Resting pressure and rectal filling sensation are meaningful indicators of long term bowel function, whereas asymmetry and squeeze pressures have limited prognostic relevance.

## CHILDHOOD DYSSYNERGIA: A SYSTEMATIC REVIEW

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**PURPOSE:** Paediatric dyssynergic defecation (DD) is an under-recognised cause of chronic, refractory constipation resulting from impaired coordination of recto-anal and pelvic floor muscles during defecation. Although adult DD is well described, paediatric-specific evidence remains limited, heterogeneous, and poorly standardised. Our aim was to systematically review existing literature on paediatric DD, describing its epidemiology, diagnostic approaches, therapeutic interventions, and research gaps.

**METHODS:** A systematic search of MEDLINE and EMBASE was conducted from inception to May 2024 using predefined terms covering dyssynergia and paediatric constipation. Eligible studies included children  $\leq 18$  years assessed for dyssynergic defecation using high-resolution anorectal manometry (HRAM) or conventional anorectal manometry (ARM), defecography, or electromyography (EMG). Case studies, non-English articles, and adult-only cohorts were excluded. Twenty-three studies met inclusion criteria.

**RESULTS:** The twenty-three studies met inclusion criteria. Those included originated from Europe (n=9), North America (n=8), Asia (n=4), Middle East/ North Africa (n=1), and Oceania (n=1). HRAM and ARM consistently identified dyssynergic patterns among children with chronic constipation, with reported prevalence ranging from 35–72% of referred patients. Biofeedback therapy, pelvic floor physiotherapy, and interferential current therapy showed improvements in stool frequency, defecatory coordination, and continence. However, diagnostic criteria, manometric definitions, and treatment protocols varied substantially between studies.

**CONCLUSION:** Paediatric DD remains under-recognised, with significant heterogeneity in diagnostic standards and therapeutic approaches. High-quality multicentre trials, paediatric normative HRAM values, and long-term outcome studies are urgently needed to inform evidence-based management.

## INVESTIGATION OF THE CORRELATION BETWEEN ANAL CANAL PRESSURE AND ELECTROMYOGRAPHY IN PEDIATRIC PATIENTS AFTER SURGERY FOR ANORECTAL MALFORMATION

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**PURPOSE:** Biofeedback (BF) therapy for fecal incontinence involves training anal contraction and relaxation while visualizing changes in measurements from an anorectal manometry or electromyography (EMG) device. Its effectiveness for fecal incontinence following surgery for anorectal malformation (ARM) has been reported. However, there is no consensus on whether manometry or EMG is the superior modality for measurement, and evidence regarding the correlation between these two indices remains scarce. Therefore, we investigated the correlation between anal canal pressure and EMG potential in this patient population.

**METHODS:** Pediatric patients with postoperative ARM were enrolled. EMG using a pediatric-specific sensor and anorectal manometry were performed, and defecatory function scores were collected. Data are presented as median (interquartile range [IQR], range). The relationship between anal pressure and EMG potential was analyzed using Spearman's rank correlation coefficient.

**RESULTS:** Nine patients (8 males, 1 female; aged 7–13 years) were analyzed, excluding one who could not perform voluntary contraction. Anal pressure was 30.6 mmHg (IQR: 24-49; range:20-72) at rest and 74.7 mmHg (IQR: 53.4-124.6; range: 38-226.7) during squeeze. EMG potential was 68.8  $\mu$ V (IQR: 61.3-106.3; range: 28.75-116) at rest and 607.5  $\mu$ V (IQR: 387.5-1412.25; range: 125-2018) during squeeze. The correlation coefficient was  $r = 0.30$  ( $p = 0.42$ ) at rest and  $r = 0.69$  ( $p = 0.04$ ) during voluntary contraction, indicating a moderate-to-strong correlation during squeeze. Conversely, two patients with similar squeeze pressures (49 and 52 mmHg) exhibited markedly different EMG potentials (488  $\mu$ V and 1838  $\mu$ V, respectively).

**CONCLUSION:** A strong correlation was confirmed between anal pressure and EMG potential during voluntary contraction. However, as these results were obtained from measurements taken at different times, further validation through simultaneous measurement during the same contraction is necessary to evaluate the physiological relationship between these two indices with higher precision.

## **EFFICACY OF PELVIC FLOOR REHABILITATION IN THE TREATMENT OF FECAL INCONTINENCE IN PATIENTS WITH ANORECTAL MALFORMATIONS AND HIRSCHSPRUNG'S DISEASE: A PRELIMINARY PROSPECTIVE STUDY**

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**PURPOSE:** Fecal incontinence is a frequent and highly debilitating complication in pediatric patients undergoing surgery for anorectal malformations (ARM) and Hirschsprung's disease (HD). Pelvic floor rehabilitation represents a potential therapeutic option; however, evidence regarding its efficacy in the pediatric population remains limited. This study evaluated the efficacy of pelvic floor physical therapy in improving fecal incontinence in pediatric patients surgically treated for ARM or HD.

**METHODS:** A prospective study was conducted on 11 pediatric patients suffering from post-surgical fecal incontinence, aged between 6 and 17 years (7 with ARM and 4 with HD), to be included in the pelvic floor rehabilitation program. Assessment was performed by telephone questionnaires administered to parents: this consist of an objective section based on the Krickenbeck classification and a subjective section regarding the efficacy of the treatment.

**RESULTS:** The collected data show a reduction in the use of medications and bowel management, along with an overall improvement in fecal incontinence in terms of frequency and volume of leakage. An improvement in the perception of the urge to defecate was observed in most patients. The clinical benefits were less pronounced in patients with ARM compared to those with HD. Almost all parents reported satisfaction with the rehabilitation process.

**CONCLUSION:** Pelvic floor rehabilitation appears to be a valuable supportive treatment for fecal incontinence in pediatric patients operated on for ARM and HD. The main limitation of this study is the small sample size; further prospective studies on larger cohorts are required.

**BENCHMARKING LONG-TERM BOWEL OUTCOMES AFTER REPAIR OF ANORECTAL MALFORMATIONS: A SYSTEMATIC REVIEW AND PROPORTIONAL META-ANALYSIS**

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**PURPOSE:** Anorectal malformations (ARM) are complex congenital anomalies frequently associated with persistent long-term bowel dysfunction despite advances in surgical reconstruction. Reported functional outcomes vary substantially across institutions, limiting meaningful benchmarking and informed patient counselling. We performed a systematic review and proportional meta-analysis to establish pooled international estimates of long-term bowel function following ARM repair.

**METHODS:** This systematic review was conducted in accordance with PRISMA guidelines. PubMed/MEDLINE, Scopus, Embase and the Cochrane Library were searched for original studies reporting postoperative bowel functional outcomes in patients with ARM. Studies utilising validated scoring systems (Krackenbeck classification, Rintala score, or Bowel Function Score) were included. Primary outcomes were voluntary bowel movements, constipation, moderate-to-severe soiling (Grade II–III), and requirement for structured bowel management. Random-effects meta-analysis using inverse variance methods with Freeman–Tukey transformation was performed. Statistical heterogeneity was assessed using  $I^2$  statistics and prediction intervals.

**RESULTS:** Thirty-six studies comprising 3,460 patients met inclusion criteria; sex distribution was reported in 3,295 patients (1,150 males and 2,145 females). Functional bowel outcomes were available for 2,908 patients. The pooled prevalence of voluntary bowel movements was 71% (95% CI 61–81%;  $I^2 = 96%$ ) based on 25 studies ( $n=2,112$ ). Constipation occurred in 34% (95% CI 25–45%;  $I^2 = 96%$ ) across 28 studies ( $n=2,375$ ). Moderate-to-severe soiling was reported in 27% (95% CI 17–39%;  $I^2 = 97%$ ) from 30 studies ( $n=2,512$ ). Additionally, 35% (95% CI 23–47%;  $I^2 = 93%$ ) required structured bowel management (15 studies,  $n=969$ ). Substantial heterogeneity was observed across all outcomes.

**CONCLUSION:** Although most patients achieve voluntary bowel control after ARM repair, clinically significant long-term morbidity remains common. Approximately one-third require ongoing bowel management and approximately one-quarter experience moderate-to-severe fecal incontinence. These pooled data help to establish an evidence-based international benchmark and reinforce the need for standardized reporting and structured long-term functional surveillance to optimise patient-centred outcomes in ARM.

## **AN INTERIM ANALYSIS OF THE STANDARD CARE (STOMA BAGS) AND DOUBLE DIAPERS IN PEDIATRIC PATIENTS WITH STOMAS AT A TERTIARY HOSPITAL IN SOUTH AFRICA**

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**PURPOSE:** In resource-limited settings, commercial stoma bags are often unavailable for pediatric colostomy patients, necessitating affordable and accessible alternatives for colostomy care. This is an interim analysis of a study comparing standard of care (stoma bags (SB)) with double diapers (DD) in patients with colostomies at the Johannesburg Pediatric Colorectal Clinic (JPCC) at Chris Hani Baragwanath Academic Hospital, South Africa.

**METHODS:** A prospective, randomized descriptive study enrolled children <3 years with newly created colostomies whose caregivers consented; participants were randomized via REDCap to SB or DD and followed up at routine visits until colostomy closure.

**RESULTS:** Of 43 screened patients, 39 were enrolled; 21 (55%) were female, with a mean (SD) age of 113.37 (154.45) days and weight of 4.64 (1.52) kg. Most had anorectal malformations (30.79%), end colostomies (22, 58%), and 28 (74%) had sigmoid colostomies. At analysis, 23 (58.97%) underwent closure. At enrolment, 18 (45%) were randomized to SB and 21 (55%) to DD; 28 (71.79%) remained in their assigned group, while 11 (28.20%) crossed over due to guardian preference (5, 29.41%) or healthcare provider recommendation (12, 70.59%).

Baseline mean (SD) peristomal skin scores (using the OST scoring system) were higher (worse) 1.75 (3.24) for the DD group compared to 1.28 (2.32) for the SB group ( $p=0.472$ ). Follow-up scores among those remaining in their groups were: SB group 2.08 (2.56) and DD group 0.38 (1.39) ( $p=0.55$ ). Patients who crossed over had mean (SD) scores of 2.16 (2.64) at enrollment and 2.21 (2.68) at follow-up. Mean (SD) clinic visits were 2.73 (2.20) for the SB group, 5.73 (3.50) for the DD group, and 6.00 (3.03) for the mixed group ( $p=0.02$ ). When supplies were depleted, 12 (32.43%) returned for more, while 25 (67.57%) purchased diapers. Out-of-pocket expenditure was lower in SB 1 (6.7%) and 5 (29.4%) in DN group ( $p=0.01$ ). Patients who remained in the SB group reported satisfaction with bag quality 10 (66.7%), compared to those who crossed over 4(26.7%) ( $p=0.02$ ).

**CONCLUSION:** Diapers are an accessible, safe alternative to commercial stoma bags for patients with colostomies.

## A SIX-YEAR INTERNATIONAL IBERO-AMERICAN COLLABORATIVE NETWORK IN PEDIATRIC COLORECTAL SURGERY

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**PURPOSE:** Rare congenital colorectal disorders such as Hirschsprung disease (HD) and complex anorectal malformations pose significant surgical and long-term functional challenges. The limited number of cases at individual institutions and the technical complexity involved highlight the importance of structured inter-institutional collaboration. Furthermore, many pediatric surgeons in Ibero-America work within healthcare systems that face similar economic challenges and resource shortages. The shared Spanish language and comparable clinical and socioeconomic settings helped create a collaborative academic group for exchanging experiences and jointly addressing complex colorectal cases. We describe the development, structure, and academic impact of an Ibero-American pediatric colorectal collaborative forum.

**METHODS:** We conducted a descriptive study of the Ibero-American Pediatric Colorectal Club's academic activities from April 2020 to February 2026. The club organizes structured monthly virtual meetings in Spanish dedicated to discussing complex pediatric colorectal cases. Most participants work in Latin America within similar healthcare systems, cultural environments, and economic constraints, which encourages open discussion of both clinical and practical management challenges. The data collected included the number of sessions, the number of cases presented, the participating countries, and the thematic distribution of cases.

**RESULTS:** During the study period, 62 meetings were held, with 137 clinical cases presented (an average of 2.2 cases per session). Participation included specialists from 12 countries across Ibero-America and the United States. Cases discussed included HD (including complex cases and redo operations), anorectal malformations, cloacal anomalies, fistulas, complex intestinal reconstructions, inflammatory and oncologic colorectal conditions, and ethical considerations in severe congenital anomalies. The initiative demonstrated sustained continuity over six years, stable international participation, and consistent engagement of high-volume referral centers. The structured, case-based format facilitated peer-to-peer exchange, exposure to diverse surgical strategies, and shared reflection on decision-making in complex scenarios, particularly in resource-limited environments.

**CONCLUSION:** This six-year experience demonstrates that structured transnational collaboration is feasible and can support academic integration and shared learning in rare and complex pediatric surgical conditions. Common language and similar healthcare environments across Ibero-American countries appear to facilitate the development of sustainable collaborative academic networks.

## FROM SILOS TO SYSTEM: A TRANSDISCIPLINARY PATHWAY FOR PEDIATRIC COLORECTAL CARE IN A MIDDLE-INCOME COUNTRY

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**PURPOSE:** Children with colorectal conditions in low-middle-income countries (LMICs) face delayed presentation, fragmented follow-up, and preventable complications. High-income centers have demonstrated that multidisciplinary colorectal programs integrating colorectal surgery, urology, gynecology, radiology, and nursing reduce morbidity and enhance patient satisfaction. However, such models are rarely implemented in LMICs, where care often remains siloed.

**METHODS:** Between 2019 and 2026, a tertiary Egyptian hospital transitioned from parallel, uncoordinated pediatric surgery and urology services to a transdisciplinary pediatric colorectal pathway. Core adaptations from high-income models included: (1) weekly joint colorectal–urology clinics with single, shared consultation; (2) prospective registry capturing diagnoses, operations, and functional outcomes for >300 patients with anorectal malformations (ARM), Hirschsprung disease (HD), and functional constipation (FC); (3) nurse-led bowel management and telehealth using locally sourced supplies; and (4) integration of pelvic floor physical therapists and adult gastroenterologists providing pediatric anorectal manometry, with cross-learning through paired clinics and procedural proctoring to address limited pediatric expertise. Regular teaching sessions for residents and fellows reinforced protocol adoption.

**RESULTS:** Consolidation into joint clinics increased the proportion of ARM/HD/FC patients receiving same-day colorectal–urology assessment to 88%, reducing multiple hospital visits and conflicting treatment plans. Registry coverage reached >80% of eligible patients with documented 6-month follow-up data. Nurse-led coordination and telehealth reduced loss-to-follow-up from historically >50% to 22% in the registered cohort, while maintaining high adherence to bowel management protocols using affordable, locally repurposed supplies. Early incorporation of pelvic floor physiotherapy and anorectal manometry enabled protocolized evaluation and rehabilitation for children with persistent dysfunction despite limited local pediatric experience in these modalities.

**CONCLUSION:** Establishing a pediatric colorectal service in LMICs requires dismantling of specialty silos and translating multidisciplinary, data-driven models into low-cost, learning systems. Transdisciplinary clinics, nurse-led continuity, shared training, and strategic use of adult expertise offer a feasible framework for sustainable colorectal care in resource-constrained settings.

## LEONA: LOGISTICAL, INSTITUTIONAL, AND SOCIOCULTURAL ASPECTS OF PAEDIATRIC BOWEL MANAGEMENT PROGRAMS: GLOBAL PRACTICE, IMAGING, IRRIGATION PRACTICES, AND COMMUNITY SUPPORT

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**PURPOSE:** To compare the factors facilitating bowel management according to World Bank income classification, including imaging practices, irrigation fluid choices, community nursing support, and treatment adherence for bowel management programs (BMP).

**METHODS:** An online survey was distributed between January-February 2026 via the snowball method. Responses were stratified by World Bank income classification: High-Income (HIC), Upper-Middle-Income (UMIC), Lower-Middle-Income (LMIC), and Low-Income Countries (LIC). Chi-square and Fisher's exact tests with Bonferroni-corrected pairwise comparisons were used. (Ethics approval available)

**RESULTS:** 160 responses received from 55 countries (HIC n=63 (39%), UMIC n=41 (26%), LMIC n=45 (28%), LIC n=11 (7%)), predominantly from tertiary centres (n=147). Colorectal fellowship training was more commonly available and completed in HICs. Prior to initiating BMP, non-HIC centres mainly relied on abdominal X-ray, whereas HIC centres more frequently used ultrasound, typically performed by clinicians rather than radiologists. In HICs, tap water was commonly used for home irrigation. Community nursing support and social acceptance of TAI were higher in HICs, with significantly better adherence to TAI (**Table 1**).

**Table 1.** Key findings comparing HIC vs non-HIC settings

Variable	HIC	Non-HIC	p-value
Colorectal fellowship trained	28.6%	7.2%	0.0003
Abdominal X-ray before BMP	61.9%	87.6%	0.0001
Ultrasound before BMP	44.4%	11.3%	<0.0001
US performed by clinician	62.8%	7.1%	<0.0001
Tap water for home irrigation	68.3%	17.2%	<0.0001
Community nursing support	43.1%	7.4%	<0.0001
TAI adherence >50%	81.7%	44.0%	<0.0001
TAI social acceptance	77.8%	36.6%	<0.0001

**CONCLUSION:** These findings highlight gaps that likely contribute to lower adherence and social acceptance challenges in non-HIC settings. Addressing these through fellowship programs, point-of-care ultrasound training, and strengthening community nursing may improve outcomes in resource-limited environments.

## MAGNESIUM-RELEASING MICROSPHERES ENHANCE ENTERIC NEURAL CREST CELL MIGRATION AND LACTYLATION-DRIVEN NEUROGENESIS FOR HIRSCHSPRUNG DISEASE

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**PURPOSE:** Hirschsprung disease (HSCR) involves distal intestinal aganglionosis caused by impaired migration and differentiation of enteric neural crest cells (ENCCs). While surgery relieves obstruction, it does not restore intrinsic enteric innervation. We investigated whether magnesium-releasing microspheres (MgRMs) provide sustained magnesium signaling to enhance ENCC migration and neuronal differentiation via metabolic and epigenetic reprogramming, thereby promoting functional enteric nervous system regeneration.

**METHODS:** Primary mouse ENCCs were cultured under MgRMs, soluble magnesium, or control conditions. Migration was assessed by live-cell imaging and Transwell assays. Neuronal differentiation was evaluated using qPCR, Western blotting, and immunofluorescence for  $\beta$ III-tubulin and HuC/D. Metabolic reprogramming was analyzed by extracellular flux assays, intracellular lactate measurement, glycolytic enzyme profiling, and  $^{13}\text{C}$ -glucose tracing to determine glycolytic flux and lactate production. Histone lactylation was examined by Western blot and ChIP-qPCR at neuronal gene promoters. Mechanistic roles were tested using focal adhesion kinase (FAK) and lactate dehydrogenase A (LDHA) inhibitors. For in vivo validation, ENCCs combined with MgRMs or soluble magnesium were transplanted into a murine aganglionosis model, followed by histological analysis of ganglion density and intestinal motility.

**RESULTS:** MgRMs provided sustained magnesium exposure and exerted stronger biological effects than soluble magnesium. ENCC migration velocity increased significantly ( $32.4 \pm 4.1 \mu\text{m/h}$  vs  $18.7 \pm 3.5 \mu\text{m/h}$  in controls,  $p < 0.001$ ), with a 2.1-fold increase in Transwell migration. Neuronal differentiation was enhanced, as  $\beta$ III-tubulin-positive cells increased to  $46.8\% \pm 4.5\%$  ( $p < 0.001$ ). MgRMs elevated extracellular acidification rate (1.8-fold) and intracellular lactate levels (2.3-fold,  $p < 0.01$ ), while  $^{13}\text{C}$ -glucose tracing confirmed increased glycolytic flux toward lactate. H3K18 lactylation increased 2.5-fold and was enriched at neuronal gene promoters. Inhibition of FAK or LDHA abolished MgRM-induced neurogenesis. In vivo, MgRMs significantly increased ganglion density (1.9-fold vs cells alone,  $p < 0.01$ ) and reduced intestinal transit time by 38% ( $p < 0.05$ ), outperforming soluble magnesium.

**CONCLUSION:** MgRMs enhance ENCC migration and neuronal differentiation by linking sustained magnesium delivery to glycolytic reprogramming and lactylation-driven epigenetic activation, offering a promising biomaterial-based regenerative therapy for HSCR.

**ENDOGENOUS PEPTIDE PDLMNA ALLEVIATES ENTERIC NEURAL CREST CELL MIGRATION DEFECTS BY COMPETITIVELY BINDING ALKBH5 WITH LMNA**

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**PURPOSE:** Hirschsprung's disease (HSCR) is a common congenital intestinal malformation caused by the absence of ganglion cells in the distal intestine due to impaired migration of enteric neural crest cells (ENCCs). The etiology of HSCR remains unclear to date, and previous studies have indicated that endogenous peptides may be involved in its pathogenesis.

**METHODS:** Single-nucleus RNA sequencing and liquid chromatography-mass spectrometry (LC-MS) were used to identify the key genes and endogenous peptides associated with the development of HSCR. Immunohistochemistry and enzyme-linked immunosorbent assay (ELISA) were performed to detect their expression levels in the intestinal tissues of HSCR patients. Cell experiments were conducted to verify their effects on ENCCs, and *Ednrb*<sup>-/-</sup> knockout mouse models and zebrafish models were used to confirm their critical roles in the pathogenesis of HSCR and their potential value in its treatment.

**RESULTS:** Single-nucleus RNA sequencing revealed a significant upregulation of the LMNA gene in neurons and intrinsic glial cells of HSCR patients. Notably, our research team identified a peptide derived from Lamin A/C (encoded by the LMNA gene) in previous endogenous peptidomic studies on HSCR, which was named PDLMNA (peptide derived from Lamin A/C). Further cell and animal experiments confirmed that LMNA could specifically bind to the ALKBH5 protein, thereby significantly inhibiting the migration and proliferation of ENCCs; in contrast, the endogenous peptide PDLMNA could competitively bind to ALKBH5, effectively reversing the inhibitory effect of LMNA on ENCC migration.

**CONCLUSION:** LMNA is involved in the pathogenesis of HSCR by specifically binding to the ALKBH5 protein and inhibiting the migration and proliferation of ENCCs, while its derived endogenous peptide PDLMNA can ameliorate the above pathological defects through competitive binding to ALKBH5. This study clarifies a novel pathogenic mechanism of HSCR and provides a potential therapeutic target for the disease.

## CHRONIC WIRELESS VAGUS NERVE STIMULATION ALLEVIATES PEDIATRIC INFLAMMATORY BOWEL DISEASE THROUGH REGULATION OF TRYPTOPHAN METABOLIC REPROGRAMMING AND CD4<sup>+</sup> T CELL IMMUNE BALANCE

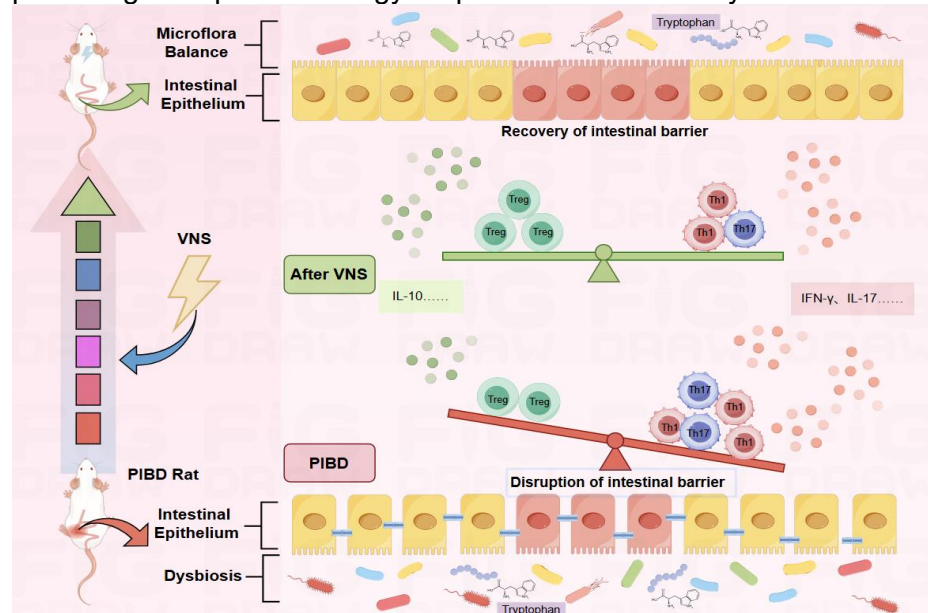
Q. Wang, Q. Liu, W. Liu, Z. Luo, J. Feng  
(Wuhan, China)

**PURPOSE:** Pediatric inflammatory bowel disease (PIBD) is increasingly prevalent and significantly impairs growth and quality of life in children. Current therapies often show limited efficacy and may require surgical intervention in severe cases. Vagus nerve stimulation (VNS) has emerged as a promising neuromodulatory strategy for controlling inflammation. This study investigated the therapeutic potential and underlying mechanisms of chronic VNS in PIBD.

**METHODS:** A PIBD model was established in 3-week-old female Sprague–Dawley rats using oxazolone enemas (10 mg/kg). A flexible, biodegradable, and biocompatible wireless VNS device suitable for long-term implantation in juvenile rats was developed. The safety profile and optimal stimulation parameters were determined by evaluating systemic inflammatory responses and organ integrity. Using the optimized parameters, rats received VNS treatment for 32 days. Multi-omics and multi-level analyses were performed to identify key regulatory pathways involved in the therapeutic effects.

**RESULTS:** Oxazolone-treated rats developed typical PIBD manifestations, including weight loss, diarrhea, hematochezia, lethargy, and severe intestinal injury, which were exacerbated following vagotomy. These rats exhibited reduced vagal tone and impaired vagal function. The developed biodegradable wireless VNS device demonstrated stable long-term performance and excellent biocompatibility. VNS stimulation for 20 minutes (0.5 mA, 2 ms, 1 Hz) significantly attenuated lipopolysaccharide-induced systemic inflammation. One month of VNS treatment markedly improved clinical symptoms and colon pathology in PIBD rats. Multi-omics analyses revealed that VNS restored the balance between pro-inflammatory Th1 and Th17 cells and anti-inflammatory Treg cells by regulating tryptophan metabolic reprogramming, thereby interrupting downstream immune cascades and reducing tissue injury.

**CONCLUSION:** Chronic wireless VNS delivered via a biodegradable neurostimulator effectively alleviates intestinal inflammation in PIBD. The therapeutic effect is mediated through modulation of immune cell function and tryptophan metabolic reprogramming, resulting in improved intestinal recovery and growth. These findings suggest that bioelectronic neuromodulation represents a promising therapeutic strategy for pediatric inflammatory bowel disease.



## INTERVAL APPENDECTOMY AFTER NON-OPERATIVE MANAGEMENT FOR PEDIATRIC UNCOMPLICATED APPENDICITIS: FEASIBILITY OF SAME-DAY DISCHARGE

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**PURPOSE:** Pediatric uncomplicated acute appendicitis (UCAA) is managed either by emergency appendectomy or antibiotic non-operative management (ANOM). Although interval appendectomy (IA) is not routinely performed after successful ANOM, reliable predictors of recurrence are lacking. As a result, patients and families must live with ongoing uncertainty and anxiety regarding if and when recurrence may occur. We evaluated IA after successful ANOM as a patient-centered strategy to reduce recurrence-related uncertainty, with particular emphasis on the feasibility and safety of same-day discharge (SDD).

**METHODS:** We retrospectively reviewed children aged ≤18 years who underwent laparoscopic IA for UCAA after ANOM at two centers between 2017 and 2025. UCAA was defined as the absence of signs of perforation or abscess. Borderline cases between UCAA and complicated appendicitis were excluded. Patient demographics, perioperative outcomes (including discharge status and postoperative complications), appendiceal histopathology, and satisfaction with ANOM followed by IA (patients/families, anesthesiologists, and nursing staff) were analyzed.

**RESULTS:** IA was performed due to family/patient preference or recurrence after ANOM at another institution. Two patients declined IA; therefore, 120 patients were analyzed (SDD, n=40; inpatient management, n=80) (**Table 1**). Median age was 13.0 years and median body mass index was 18.3 kg/m<sup>2</sup>; an appendiceal fecalith was present in 22 patients (18.3%). The median interval from IA scheduling to surgery was 48.5 days and median operative time was 38.0 minutes. Postoperative complications occurred in 3 patients (2.5%): port-site bleeding (n=1) and appendiceal stump abscess (n=2). No unplanned visits or readmissions occurred. SDD was successful in 39/40 (97.5%); one patient required overnight observation due to port-site bleeding. Recurrence while awaiting scheduled IA occurred in 5 patients (4.2%); all improved with repeat ANOM and underwent elective IA without emergency surgery. Histopathology demonstrated inflammatory changes in all specimens, and no normal appendix was identified (**Figure 1**). Patients/families satisfaction was 83.3%, and staff satisfaction was 89.2%.

Table 1. Outcomes of interval appendectomy (IA) following antibiotic non-operative management (ANOM) for uncomplicated acute appendicitis (UCAA) in children

	Overall (n=120)	SDD (n=40)	IM (n=80)	P (SDD vs IM)
Age (years)	13.0 (11.0–14.3)	15.0 (11.0–17.0)	12.0 (10.0–14.0)	< 0.01
Gender (male)	81 (67.5%)	34 (85.0%)	47 (58.8%)	< 0.01
BMI (body mass index)	18.3 (17.2–20.4)	19.4 (17.6–21.8)	18.1 (16.4–20.0)	0.06
Number of prior appendicitis episodes	1.0 (1.0–2.0)	1.0 (1.0–2.0)	1.0 (1.0–2.0)	0.29
Interval from first appendicitis episode to IA (days)	90.5 (48.4–204.8)	137.0 (55.0–358.0)	85.0 (46.5–147.5)	0.06
Recurrence while awaiting scheduled IA	5 (4.2%)	1 (2.5%)	4 (5.0%)	0.66
Interval from IA scheduling to surgery (days)	48.5 (24.0–76.0)	35.0 (20.8–53.8)	56.0 (31.8–79.5)	0.02
Presence of appendiceal fecalith	22 (18.3%)	7 (17.5%)	15 (18.8%)	1.00
Operative time (minutes)	38.0 (32.0–62.0)	32.5 (28.0–37.3)	62.0 (58.0–66.0)	< 0.01
Postoperative complications	3 (2.5%)	1 (2.5%)*	2 (2.5%)**	1.00
SDD success	NA	39/40 (97.5%)	NA	NA

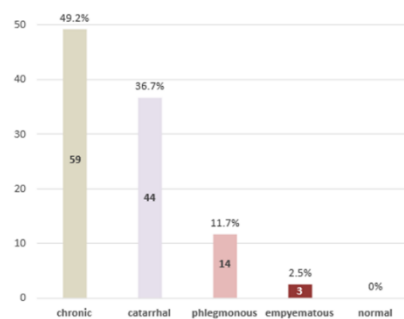
Note: Values are presented as medians with interquartile ranges (IQR) or as n (%).

SDD, Same-day discharge. IM, Inpatient management.

\*Postoperative complications in SDD: trocar site bleeding (n=1).

\*\*Postoperative complications in IM: Abscess at the appendiceal stump (n=2).

Figure 1. Histopathological findings of the appendix after interval appendectomy (n=120)



**CONCLUSION:** IA after successful ANOM for pediatric UCAA was associated with low morbidity and high feasibility of SDD. The universal presence of inflammatory changes supports IA as a reasonable option to reduce uncertainty related to recurrence.

## DOES RADIOLOGICAL ASSESSMENT OF RECONSTRUCTED ANORECTUM CORRECTLY MIRROR THE POSTOPERATIVE FUNCTIONAL OUTCOME IN CHILDREN WITH ANORECTAL MALFORMATION?

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(Kolkata, India)

**PURPOSE:** Long-term functional outcome after repair of Anorectal Malformation (ARM) remains considerably poor despite advancements in anatomical knowledge and surgical techniques. We wanted to study the relationship of morphology of reconstructed anorectum with functional outcome.

**METHODS:** We studied 58 operated patients of ARM (excluding perineal fistula, anal stenosis, persistent cloaca, pouch colon, associated spinal anomaly) over an 18-month period in a tertiary-care hospital in Eastern India. The patients having undergone staged repair, were assessed  $\geq 6$  months after stoma reversal. Age, sex and type of anomaly were recorded. Physical examination and Rintala and Lindahl continence scoring (RLS) were used for functional assessment. MRI pelvis and trans-perineal ultrasonography (TPUS) were used to assess thickness of internal (IAST) and external (EAST) anal sphincters and position of neorectum in relation to sphincter complex (SC). Single trained radiologist performed both tests after blinding the MRI plates to reduce diagnostic bias. Appropriate statistical tests were used and Spearman Rho Correlation Coefficient ( $\rho$ ) was calculated.

**RESULTS:** 37 boys [11 high ARM, 26 intermediate ARM] and 21 girls [19 vestibular anus, 2 intermediate ARM]; ranging from 3.5 to 11 years. RLS: 17.2% EXCELLENT ( $\geq 18$ ), 53.4% GOOD ( $17 \geq 12$ ), 25.9% FAIR ( $11 \geq 7$ ), 3.4% POOR ( $\leq 6$ ). Patients with vestibular anus had highest RLS, followed by intermediate and then high ARM. IAST and EAST:  $1.24(\pm 0.32)$  mm and  $1.82(\pm 0.39)$  mm respectively on TPUS;  $1.47(\pm 0.33)$  mm and  $2.17(\pm 0.44)$  mm respectively on MRI. On TPUS and MRI respectively, 4 and 9 patients had neorectum eccentrically located within SC. No significant difference in sphincter thickness (ST) was present among different ARM types on either modality. ST on TPUS and MRI were comparable ( $p$ -value $<0.001$ ;  $\rho$ -0.756). Those with abnormally located neorectum had lower RLS ( $p$ -value:0.012). Weak but significant correlation ( $p$ -value:0.039;  $\rho$ -0.271) was present between RLS and EAST on MRI.

**CONCLUSION:** Morphological assessment of neorectum and functional outcome can be correctly correlated in postoperative patients of ARM. TPUS and MRI pelvis are interoperable and comparable for this purpose. TPUS may prove to be a valuable tool for this purpose in resource-poor areas. Knowledge of anatomy of neorectum may help plan management to improve the quality of life.

## REHABILITATION STRATEGY FOR CHILDREN WITH IMPAIRED COLON AND PELVIC FLOOR FUNCTION: FROM “NORMAL VALUES” TO COMPENSATORY CAPACITY ASSESSMENT

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(Moscow, Russia)

**PURPOSE:** To present a structured rehabilitation program for children with impaired colon and pelvic floor function after anorectal surgery, based on compensatory capacity assessment.

**METHODS:** Retrospective summary of 120 observations: 78 after primary repair of anorectal malformations (ARM) in our center, 6 after redo reconstructions, 11 undergoing rehabilitation after surgery in other hospitals, 18 with functional anorectal disorders without congenital malformations or anatomical defects, and 7 with anatomical defects after primary ARM repair not yet reoperated at the time of analysis.

Rehabilitation started with staged neoanus dilatation without forced diameter increase, with progression according to tissue adaptation. Stool control was maintained even during dilatation in children <1 year; if no stool >24 hours, a cleansing enema was used. Scar assessment was performed with Hegar dilators; a dense scar tightly gripping Hegar #12 triggered proteolytic therapy (1-month course, repeat after 2 months if needed) and physiotherapy when indicated. At 1 year, perineal ultrasound and digital rectal examination were performed. Voluntary external anal sphincter (EAS) contraction is limited before 3–4 years; an abnormal pelvic movement pattern during voluntary straining (anterior shift when “holding”, posterior shift when straining) was considered a marker of predominant puborectal mechanism. Biofeedback therapy was initiated from 4 years in children with abnormal patterns, and from 5 years in those without voluntary EAS contraction, with reassessment ≥6 months later.

**RESULTS:** Functional adaptation disorders predominated after primary reconstructions, particularly after stoma closure when a new reservoir function develops. In high ARM variants, absent spontaneous evacuation was managed by regular cleansing enemas with dynamic regimen adjustment until defecation control matured. Scar monitoring helped prevent progression of cicatricial complications; in 2 girls a dense scar regressed by 1 year following proteolytic therapy. Biofeedback contributed to development of a voluntary continence component and reduction of anal insufficiency severity in children with preserved rehabilitation potential.

**CONCLUSION:** Rehabilitation after ARM repair should be approached as a long-term, controlled adaptation process to anatomy and medical interventions. A compensatory-capacity framework supports individualized strategies combining staged dilatation, stool control, scar management, and biofeedback to improve bowel management and the likelihood of social continence.

## RETHINKING INITIAL DIVERSION IN TYPE-1 CONGENITAL POUCH COLON: LOOP ILEOSTOMY VERSUS WINDOW COLOSTOMY

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(Lucknow, India)

**PURPOSE:** Congenital Pouch Colon (CPC) represents an uncommon and severe variant of Anorectal Malformation (ARM), characterized by a markedly shortened and dilated colon that poses considerable surgical challenges. Traditionally, management involves staged procedures beginning with a window colostomy, followed by coloplasty with proximal ileostomy, and eventual closure of the stoma. However, window colostomy is frequently associated with complications such as stomal stenosis and prolapse of the pouch. This study aimed to compare the short-term outcomes of window colostomy and loop ileostomy when used as the primary diversion in neonates with ARM associated with Type-1 CPC, in order to determine the safer and more effective initial surgical strategy.

**METHODS:** A total of 72 neonates (male-to-female ratio 7.1:1) diagnosed with ARM and Type-1 CPC and managed with staged surgical repair over a ten-year period were included in this study. The diagnosis was established by abdominal radiography demonstrating a characteristic pouch-like dilatation of the colon in a neonate with ARM. Based on the type of initial diversion, patients were divided into two groups: Group A (n = 34) who underwent window colostomy and Group B (n = 38) who underwent loop ileostomy. Stoma-related complications—including stomal stenosis, obstruction, prolapse, and peristomal skin excoriation—were recorded and compared between the two groups.

**RESULTS:** In Group A, 13 patients developed stomal stenosis or obstruction. Of these, four improved with serial dilatation, four required surgical revision, and five ultimately required conversion to loop ileostomy. Additionally, complete prolapse of the pouch occurred in five patients, necessitating reduction and creation of a loop ileostomy. In contrast, no cases of stomal stenosis were observed in Group B, although four patients developed mild stomal prolapse. The incidence of peristomal skin excoriation was comparable in both groups.

**CONCLUSION:** Loop ileostomy as the initial diversion procedure in neonates with Type-1 CPC appears to result in fewer and less severe complications compared with window colostomy. Therefore, loop ileostomy may be considered a safer and more effective primary surgical option in the initial management of this condition.

## CHARACTERISTICS OF GANGLIONIC COLONIC LESIONS IN PATIENTS WITH ANORECTAL MALFORMATION

N.G. Kolesnikova, V.G. Svarich  
(Saint Petersburg, Russia)

**PURPOSE:** To study the morphology of neurons in the colon in patients with anorectal malformation.

**METHODS:** 36 patients with anorectal malformation are included in the work. Patients are divided into 2 groups. The first group consisted of 24 patients who took a biopsy from the second distal centimeter of the descending intestine during the primary correction of anorectal malformation. The second group includes 12 patients after correction of anorectal malformation with constipation. In children of this group, the biopsy was taken from the ascending, sigmoid, lowered intestine.

**RESULTS:** In the first group of patients in the second centimeter of the lowered intestine, agangliosis of the muscle layer was detected in 87% of cases, but not everyone had constipation after the correction of the defect. Patients of the second group with constipation always had a pathology of the intestine brought to the anus (in 25% it was hypogangliosis, in 75% - its agangliosis). Hypogangliosis of the sigmoid colon was noted in 91% of patients, and in the ascending intestine hypogangliosis was detected in 58% of patients.

**CONCLUSION:** In most patients, during the primary correction of anorectal malformation, agangliosis of the distal part of the intestine reduced to the anus was detected. In patients with constipation after correction of anorectal malformation, there was always a pathology of the number of neurons brought to the anus of the intestine, and in most cases there was hypoangliosis of the sigmoid colon. In a patient with no constipation after correction of anorectal malformation, the lowered, sigmoid and ascending intestine did not have ganglion apparatus pathology.

## **SINGLE-PORT ROBOTIC-ASSISTED SOAVE PROCEDURE FOR HIRSCHSPRUNG'S DISEASE BY DA VINCI XI SYSTEM**

J.L. Chen, Y.R. Zhang, S. Lin, D. Xu  
(Fuzhou, China)

**PURPOSE:** To evaluate the safety and feasibility of single-port robotic-assisted Soave procedure for Hirschsprung's disease (HD) in children using the da Vinci Xi system.

**METHODS:** A retrospective analysis was conducted on 10 pediatric patients who underwent robotic-assisted Soave procedure for HD, including 8 males and 2 females, with a median surgical age of 61.5 months (range: 8–141 months). Among them, 8 cases were short-segment type and 2 were long-segment type. We describe the detailed steps for performing a single-incision-assisted Soave procedure in children with Hirschsprung's disease using the da Vinci Xi system. Intraoperative and postoperative recovery indicators were recorded.

**RESULTS:** All 10 cases were successfully completed without conversion to open surgery or intraoperative blood transfusion. The mean operative time was  $238.7 \pm 30.9$  minutes, with an estimated blood loss of  $6.7 \pm 2.8$  mL. Postoperative gastrointestinal function recovered well, with first flatus occurring at  $25.8 \pm 5.9$  hours and first defecation at  $42.6 \pm 9.5$  hours. The average postoperative hospital stay was  $11.2 \pm 3.3$  days. Two cases developed mild complications (colitis and perianal dermatitis), which resolved after symptomatic treatment.

**CONCLUSION:** The single-port da Vinci robotic-assisted Soave procedure for pediatric HD is safe and feasible, offering advantages such as minimal bleeding, rapid recovery, and low complication rates, with satisfactory short-term outcomes. Additionally, the robot's stable operating system and high-magnification 3D stereoscopic vision facilitate the identification and protection of pelvic nerves.

## CLINICAL MANIFESTATIONS AND MANAGEMENT OF ANASTOMOTIC LEAKAGE AFTER THE SOAVE OPERATION

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(Beijing, China)

**PURPOSE:** Anastomotic leakage is one of the most common complications that can occur following the Soave operation. The purpose of this study was to document our experience with diagnosis and management of anastomotic leakage following the Soave surgery to reduce the incidence and enhance therapeutic effect.

**METHODS:** From January 2020 to December 2025, a retrospective study was conducted on 30 children who admitted to our department for anastomotic leakage following the Soave operation. Medical records were analyzed with focus on the clinical manifestations, initial diagnosis, imaging examination, management and outcomes of the patients. The Rintala score was used to assess postoperative bowel function.

**RESULTS:** The Soave procedure was used as the primary surgical method in all cases. HD was the most common primary disease (14/30, 46.7%), and HD allied disease was also possible cause (4/30, 13.3%), while constipation was the cause of other cases. Fever and abdominal pain were the most common early symptom (21/30, 70.0%), and perianal or pelvic abscess was typical in late diagnosed cases (4/30, 13.3%). Ultrasonography, lower GI series, CT and MRI were used in the imaging examination. Enterostomy (29/30, 96.7%) was performed when anastomotic leakage was diagnosed. Repair of the leakage in situ was conducted in six patients, and redo pull-through was performed in 18 cases. Other six patients underwent fistulectomy. Follow-up was conducted in 18 cases who had closed the stoma. Rintala score was  $16.21 \pm 2.75$ .

**CONCLUSION:** Typical clinical presentation and necessary imaging examination were essential for the diagnosis of anastomotic leakage after the Soave operation. Enterostomy should be performed when anastomotic leakage occurs. Repair of the leakage in situ, redo pull-through and fistulectomy could be used in the management.

## RECURRENT HIRSCHSPRUNG-ASSOCIATED ENTEROCOLITIS

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**PURPOSE:** Hirschsprung-associated enterocolitis is among the most dangerous complications of Hirschsprung's disease. Cases of recurrent enterocolitis after surgery occur with different surgical treatment methods and at any patient age. The complexity of treating Hirschsprung-associated enterocolitis formed the basis for conducting this study and for identifying approaches to address the problem, including the development of therapeutic and diagnostic measures for recurrent enterocolitis in patients with Hirschsprung's disease after transanal endorectal pull-through surgery.

**METHODS:** From 2010 to 2024, 112 patients under 3 years of age with Hirschsprung's disease were operated on in the Republican Scientific and Practical Center for Pediatric Surgery using the transanal endorectal pull-through surgery. During this period, we observed 25 cases of postoperative enterocolitis. Of these, 16 patients had recurrent postoperative enterocolitis.

**RESULTS:** An algorithm was developed to identify the causes of recurrent Hirschsprung-associated enterocolitis, as well as a set of therapeutic measures to be implemented when they are detected. In patients with recurrent Hirschsprung-associated enterocolitis, autoimmune processes associated with inflammation are triggered in the intestinal wall. Preventive treatment for postoperative enterocolitis significantly reduces the risk of its development. The use of the posterior muscle cuff excision method during transanal endorectal pull-through surgery (in 2015) led to a decrease in the number of postoperative enterocolitis associated with colostasis (the total number of enterocolitis decreased from 34% in the period 2010-2014, to 16,7% in the period 2015-2019, and 14.3% in the period 2020-2024;  $p=0,032$ ;  $\chi^2=4,69$ ).

**CONCLUSION:** A decrease in the number of enterocolitis can be achieved by using modern methods of surgical treatment of Hirschsprung's disease, timely detection of the anatomical cause (stenosis or residual aganglionosis) and its elimination, adequate postoperative treatment, the initiation of preventive therapy for Hirschsprung-associated enterocolitis, and long-term follow-up of patients operated on for Hirschsprung's disease

## MANAGEMENT OF TOTAL COLON AGANGLIONOSIS IN DEVELOPING COUNTRIES

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**PURPOSE:** To present the problems in the management of 25 children with total colon aganglionosis (TCA). The clinical features, diagnosis & diagnostic difficulties are discussed.

**METHODS:** From Jan 1991 to Dec 2023, 25 children with TCA were treated. There were 19 males & 6 females. Two sets of twins – in one set both had TCA. In the other set, one had TCA. Ethnic ratio: Chinese 16, Malay 4 & Indians 5. Associated malformations were: Covered anus -2, Exomphalos with cluster of malformation, Ileal atresia in 1, Ileal stenosis -2. Family history in 3, polydactyly -1, None had Trisomy 21 Syndrome. 18 presented in neonatal period, 5 in less than 3 months & 2 at 6 months. Plain X-ray abdomen & suction rectal biopsy is needed for all cases. Laparoscopy & biopsy in 5 cases. Diagnosis was delayed in 6 cases due to multiple procedures. Contrast enema is characteristic in these cases. The extent of aganglionosis: Mid ileum -3, 65 cm from DJ in 2. Extent of small bowel leads to delayed adaptation. Severe enterocolitis in defunctioned small bowel, needed emergency resection at 3 & 6 months. Failed Duhamel pull-through of caecum & ascending colon in two cases needed further management. Swenson's was done in -1, Low oblique ileorectal anastomosis was performed below the peritoneal reflection in 18 cases. Martin's procedure was done in one. Postoperative anal dilatation was performed after 10 days. All children needed nutritional support, enteral & parenteral nutrition needed for 6 to 12 months. Home parenteral nutrition was given to 4. Three children had enterocolitis & needed care. Four had constipation due to growth & needed ano-rectal myectomy (Lynn's myectomy. Excoriation was treated with Tapioca powder, zinc oxide & miconazole cream).

**RESULTS:** Length of aganglionic small bowel varied from 10 to 20 cms from ileo-caecal valve to 60 cms. These children take time to get better. 21 are alive, 1 lost to follow up. Follow up is from 2 to 42 years.

**CONCLUSION:** Proper neonatal care, nutritional support & continuous long-term care is essential for satisfactory results. We prefer the low oblique ileo-rectal anastomosis & it gives good long-term results. Post-op regular anal dilatation is needed in the first 2-3 years.

## THE CONDITION OF THE GANGLION APPARATUS IN THE SEGMENT CLOSEST TO THE AGANGLIONIC SEGMENT AND IN THE ASCENDING INTESTINE IN HIRSCHSPRUNG'S DISEASE

N.G. Kolesnikova, V.G. Svarich  
(Saint Petersburg, Russia)

**PURPOSE:** It was decided to study the number of neurons and Kahal cells in the intestinal wall above the agangliosis zone and in the ascending intestine in the rectosigmoid and subtotal forms of Hirschsprung's disease.

**METHODS:** 18 patients are included in the work, including 11 patients with a rectosigmoid form of Hirschsprung's disease and 7 children with a subtotal form. Biopsies were examined in the intestine closest to the aganglionic segment and in the ascending intestine.

**RESULTS:** Neuronal pathology in the intestine closest to the aganglion segment was detected only in 66% of patients (12 out of 18). Among them there were 3 variants of histological pathology: hypogangliosis (83% of cases), absence of Kahal cells in 16% of patients (in 2 out of 12), and neuronal dystrophy was detected in 1% of patients.

Pathology of neurons in the ascending intestine was detected in 55% of cases (10 out of 18). 2 variants of histological pathology were identified: hypogangliosis (80% of cases), absence of Kahal cells in 20% of patients (in 2 out of 10).

**CONCLUSION:** The number of neurons in the segment closest to the aganglionic segment was reduced in 66% of cases. The number of neurons in the ascending intestine in the rectosigmoid and subtotal form of Hirschsprung's disease was reduced in 55% of cases. Kahal cell pathology was detected in 11% of cases. Hypogangliosis of the ascending intestine did not affect the outcome of treatment in the rectosigmoid form of Hirschsprung's disease with lowering of the normoganglionic intestine.

## **CHRONIC INTESTINAL PSEUDO-OBSTRUCTION IN PEDIATRIC MELAS: SMOOTH MUSCLE PATHOLOGY**

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(Athens, Greece)

**PURPOSE:** To investigate the pathological alterations of intestinal smooth muscle in children with chronic intestinal pseudo-obstruction (CIPO) associated with mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes (MELAS), aiming to clarify mechanisms beyond enteric neuropathy.

**METHODS:** A systematic review of PubMed, Scopus, and Cochrane Library was performed. Studies reporting histopathological findings, muscle biopsies, and clinical correlations in paediatric patients with MELAS-associated CIPO were included, encompassing both case series and relevant reviews.

**RESULTS:** CIPO affects up to 40% of children with MELAS. Although enteric nervous system abnormalities are described, 15–38% of patients show normal intestinal histology, indicating additional pathogenic mechanisms. Evidence suggests mitochondrial dysfunction in intestinal smooth muscle contributes to impaired motility. Histopathological findings include mitochondrial proliferation, structural abnormalities of smooth muscle fibres, and decreased oxidative enzyme activity, despite preserved neural architecture. These findings support a myopathic mechanism in a subset of paediatric MELAS cases, explaining clinical pseudo-obstruction despite normal neural histology.

**CONCLUSION:** In paediatric MELAS-associated CIPO, smooth muscle pathology due to mitochondrial dysfunction significantly contributes to intestinal dysmotility. Recognition of myopathic involvement is crucial for understanding disease mechanisms, guiding management, and informing future research on targeted therapies. Further studies are needed to correlate mitochondrial defects with functional motility assessments and clinical outcomes in children.

## **DESCENDING PERINEUM IS THE MOST FREQUENTLY DETECTED DEFECOGRAPHY INDICATOR IN CONSTIPATION IN CHILDREN AND THE EFFECTIVENESS OF CONSERVATIVE TREATMENT FOR IT**

N.G. Kolesnikova, V.G. Svarich, E.V. Yakusheva  
(Saint Petersburg, Russia)

**PURPOSE:** To show using defecography that descending perineum is often detected in children with constipation and to evaluate the effectiveness of conservative treatment of constipation with it.

**METHODS:** The work included 87 children with functional constipation. Children with functional constipation who underwent defecoscopy and colodynamic examination were selected as inclusion criteria. Video defecoscopy was performed by filling the rectum with a contrast mixture (30% barium sulfate solution) according to the original method. The obtained parameters of video defecoscopy were compared with its normal values. The effectiveness of 3 courses of complex conservative treatment of constipation accompanied by descending perineum for 6 months was assessed.

**RESULTS:** In 78.2% of cases, descending perineum was detected in children with "functional" constipation during defecography. In most cases, the anal canal did not open (in 95.4% of cases, which was combined with a pathological rectoanal reflex during colodynamic examination). The effectiveness of complex conservative treatment of constipation with descending perineum according to Rome criteria - 4 is low - constipation regressed in only 24.1%.

**CONCLUSION:** Descending perineum is a very frequently detected pathological indicator of defecography in constipation in children (in 78.2% of patients), in most cases it is combined with obstruction in the anal canal and internal sphincter of the anus and is extremely poorly amenable to conservative treatment for six months (only in 24.1% of observations).

## IS THE PRESENCE OF A PSOAS ABSCESS AN OBLIGATORY PREDICTOR OF STOMA FORMATION DURING ILEOCECAL RESECTION IN PEDIATRIC PATIENTS WITH CROHN'S DISEASE?

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(Saint Petersburg, Russia)

**PURPOSE:** To evaluate the treatment outcomes after ileocecal resection in patients with Crohn's disease and the presence of a psoas abscess.

**METHODS:** The study was retrospective. From 2016 to 2026, 85 ileocecal resections were performed in children under 18 years of age with complicated forms of Crohn's disease, of which 10 (11,8 %) patients had psoas abscess.

**RESULTS:** The median age at surgery was 15 years. Bio-naive were 7 patients (70%). In 8 of 10 patients, there was no systemic corticosteroid therapy before the ileocecal resection. Antibacterial therapy with broad-spectrum drugs was performed in 8 of 10 patients (80%). In 3 patients (30%) abscess was detected intraoperatively. The operation time of ileocecal resection was  $272,20 \pm 74,08$  min. After the surgery, all children received anti-relapse treatment including anti-TNF drugs. In 75% of patients (8/10) a stoma was not performed during the ileocecal resection. The level of infectious complications in 4 of 10 patients (40%) was superficial and did not exceed 1 on the Clavien - Dindo scale. There were no cases of anastomotic leakage. In one case required ureteral stenting due to cicatricial changes in the abscess area.

**CONCLUSION:** The presence of a psoas abscess is one of the factors for possible intestinal anastomosis failure, but is not a reliable predictor of an unfavorable outcome of surgical intervention.

## CLINICAL CHARACTERISTICS AND SURGICAL TREATMENT OF CONGENITAL GLUTEAL DERMAL SINUS TRACT IN CHILDREN: A 15-YEAR SINGLE-CENTER CLINICAL EXPERIENCE

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(Shanghai, China)

**PURPOSE:** To summarize the clinical features, diagnosis, treatment, and prognosis of congenital gluteal dermal sinus tracts in children, providing insights for better clinical management.

**METHODS:** A retrospective analysis was conducted on 20 patients diagnosed with congenital gluteal dermal sinus tracts at the Children's Hospital of Fudan University over 15 years. The analysis focused on age, gender, diagnostic delay, diagnostic methods, course and opening of the sinus tract, and postoperative follow-up.

**RESULTS:** The study included 6 males and 14 females, with symptom onset ranging from birth to 12 years (median: 5 months). Initial symptoms included abnormal gluteal depression with recurrent infections (n=8), unexplained recurrent infections (n=6), and asymptomatic gluteal depression or small holes (n=6). The time from symptom onset to diagnosis ranged from 1 to 102 months (median: 21 months). MRI was positive in 87.5%, identifying the tract in 81.25%. CT scans were positive in all cases, but only 50% identified the tract. Sinus tractography successfully identified the tract in 7 of 10 patients. The sinus tracts were classified into three types based on the opening location. All sinus tracts were excised during surgery, with an average length of 4.95 cm. Two patients experienced recurrence and underwent reoperation, while the remaining 15 had no recurrence.

**CONCLUSION:** Congenital gluteal dermal sinus tracts are more common in females and often misdiagnosed. CT is more sensitive but less specific than MRI. Sinus tractography helps determine the tract's path. Prognosis is generally good after complete excision, with recurrence being the main postoperative complication.

**ASSOCIATION BETWEEN APPENDECTOMY, TONSILLECTOMY, SURGICAL STRESS, AND CROHN'S DISEASE: A RETROSPECTIVE CASE-CONTROL STUDY**

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**PURPOSE:** This study aimed to evaluate whether appendectomy, tonsillectomy, and/or surgical stress are associated with Crohn's disease (CD) and to assess the time interval between these interventions and CD diagnosis.

**METHODS:** This retrospective case-control study included patients diagnosed with CD at Muğla Training and Research Hospital between 2012 and 2024. The control group consisted of individuals without inflammatory bowel disease (IBD), retrospectively selected from pediatric surgery and adult gastroenterology outpatient clinics at a minimum ratio of 2:1. Groups were matched for age, sex, smoking status, and vitamin D deficiency. Histories of appendectomy, tonsillectomy, and surgical procedures performed under general anesthesia were recorded. In patients with CD, the interval between surgical procedures and CD diagnosis was calculated. Statistical analyses were performed using Jamovi version 2.6, with p values < 0.05 considered statistically significant.

**RESULTS:** A total of 237 individuals were included (112 CD patients and 125 controls). Age and sex distributions were similar between groups. Smoking was significantly more frequent in the CD group than in controls (54.5% vs. 37.6%,  $p = 0.009$ ). Vitamin D deficiency did not differ significantly between groups. Appendectomy was more common among CD patients (17.9% vs. 9.6%); however, the difference was not statistically significant ( $p = 0.096$ ). Tonsillectomy and surgery performed under general anesthesia were not associated with CD. The median interval between appendectomy and CD diagnosis was seven years, while the median interval between surgery under general anesthesia and CD diagnosis was four years.

**CONCLUSION:** Appendectomy, tonsillectomy, and surgical stress were not identified as independent risk factors for Crohn's disease in this cohort. Smoking was the only factor significantly associated with CD.

## IS PERITONEAL DRAINAGE NECESSARY AFTER LAPAROSCOPIC APPENDECTOMY FOR COMPLICATED ACUTE APPENDICITIS IN CHILDREN

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(Tanta, Egypt)

**PURPOSE:** The role of routine peritoneal drainage following laparoscopic appendectomy for complicated acute appendicitis in children remains controversial. This study aimed to evaluate whether peritoneal drainage improves postoperative infectious and recovery outcomes compared with no drainage.

**METHODS:** A prospective randomized controlled trial was conducted at a tertiary pediatric surgical center. A total of 210 children ( $\leq 18$  years) with complicated appendicitis (appendicular mass, abscess, gangrene, perforation, or free peritoneal pus) undergoing laparoscopic appendectomy were randomized into two equal groups: drainage (Group A, n=105) and no drainage (Group B, n=105). Standardized operative and postoperative protocols were applied. The primary outcome was overall surgical site infection (SSI) within 30 days. Secondary outcomes included intra-abdominal infection (IAI), postoperative ileus, time to return of bowel function, duration of antibiotic therapy, length of hospital stay, and hospital costs.

**RESULTS:** Baseline demographic and clinical characteristics were comparable between groups. Overall SSI did not differ significantly between Group A and Group B (20.0% vs. 27.6%,  $p=0.312$ ). However, Group A demonstrated a significantly lower rate of IAI (6.67% vs. 15.24%,  $p=0.046$ ) and postoperative ileus (4.76% vs. 13.33%,  $p=0.050$ ). Time to return of bowel function was shorter in the drainage group (median 13.9 vs. 17.3 hours,  $p<0.001$ ), with reduced antibiotic duration (median 5.8 vs. 7.4 days,  $p<0.001$ ) and shorter hospital stay (median 1.7 vs. 3.6 days,  $p<0.001$ ). Adjusted analyses confirmed peritoneal drainage as an independent protective factor against IAI (adjusted OR 0.38, 95% CI 0.15–0.95,  $p=0.039$ ). Mean 30-day hospital costs were significantly lower in Group A ( $p<0.001$ ).

**CONCLUSION:** Routine peritoneal drainage did not significantly reduce overall SSI after laparoscopic appendectomy for complicated appendicitis in children. However, drainage was associated with fewer intra-abdominal infections, faster recovery, and reduced hospital stay and costs. Selective use of peritoneal drainage may be beneficial and warrants further multicenter pediatric studies.

## REDO POSTERIOR SAGITTAL ANORECTOPLASTY (REDO-PSARP) AT AN UNCONVENTIONAL AGE IN A PREVIOUSLY REPAIRED ANORECTAL MALFORMATION (ARM): A CASE REPORT AND REVIEW OF THE LITERATURE

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(Milan, Italy)

**PURPOSE:** Redo-PSARP is typically indicated for persistence or development of anatomical defects after repair of ARM. The procedure poses unique challenges due to prior surgery, and published series mostly cluster around toddlers and preschool-age children. Intervening at older age entails additional difficulties, including increased technical complexity, higher complication rates, less predictable outcome and heightened psychological burden. We present a late redo-PSARP case, successfully performed from both surgical and patient outcome perspective.

**METHODS:** Description of a late redo-PSARP case and literature review.

**RESULTS:** A 20-year-old female, previously treated elsewhere with primary repair of an isolated rectovestibular fistula, was referred to our institution aged 6 due to fecal incontinence.

The neoanus was completely anteriorly displaced. Multiple bowel management optimization strategies were implemented, but fecal continence was never achieved. Owing to parental limitations, viability of surgical intervention was deferred until adulthood when at 19 years of age, the patient elected to undergo redo-PSARP. A multidisciplinary transition of care program was effectively implemented. Anatomical correction was attained, with meaningful functional improvement reaching spontaneous continence and reflecting in strong patient endorsement. The patient was discharged after 9 days on oral macrogol. Follow-up showed regular bowel movements with no soiling or interval incontinence.

Neoanal stenosis was noted on postoperative day 42, successfully treated with anal dilations. Pelvic floor rehabilitation was initiated 2.5 months postoperatively.

To our knowledge, 17 redo PSARP in late adolescence and adulthood are reported. Fecal incontinence was the indication in 16/17, and continence was reached in 12; complications were reported in 3 patients.

**CONCLUSION:** Current literature on late redo-PSARP remains limited, as most adult series describe primary repair of previously untreated ARM rather than revision. This case, expanding on existing literature, highlights the feasibility of late redo-PSARP at experienced colorectal centers. In carefully selected patients, with correctable anatomical defects and good potential for bowel control, this intervention can substantially improve both function and quality of life. Beyond surgical act, a positive outcome requires a well-structured follow-up along with an effective transition from pediatric to adult care. Moreover, realistic-outcome counselling, compliance and psychological readiness of the patient are crucial to minimize complications while optimizing outcomes.

## A DECADE OF SACRAL NEUROMODULATION IN PEDIATRIC URINARY AND FECAL INCONTINENCE: LONG-TERM EFFICACY AND QUALITY OF LIFE OUTCOMES

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**PURPOSE:** Sacral neuromodulation (SNM) has emerged as an effective therapy for pediatric patients with refractory urinary incontinence (UI) and, more recently, fecal incontinence (FI). However, long-term outcomes and quality of life (QoL) data in this population remain limited. This study aims to assess the impact of SNM on QoL in patients with medically refractory FI, with a 10-year follow-up.

**METHODS:** We conducted a retrospective review of pediatric patients treated with SNM between 2015 and 2025. The implantation was performed in two stages (testing and permanent phase). Clinical data included diagnosis, prior therapies, Cleveland Clinic Incontinence Scale (CCIS) scores, complications, and postoperative QoL assessed by the CCVIA questionnaire (lifestyle, behavior, self-perception/depression, and embarrassment domains).

**RESULTS:** 12 patients (mean age 15 years) were included, with a median follow-up of 7 years. Diagnoses included anorectal malformation (4), Hirschsprung disease (3), myelomeningocele (1), lipomeningocele (1), agenesis of the corpus callosum (1), and Prune Belly syndrome (1) and Angelman syndrome (1). 2 patients were excluded due to non-response and device-related complications. All patients had undergone previous biofeedback and tibial nerve stimulation. Mean preoperative CCIS score was 18, with a mean reduction of 66% ( $p < 0.01$ ). Complications occurred in 22% (two cases of fever, back pain, and device extrusion), all resolved after reimplantation. Postoperatively, 88% of patients reported improved QoL, with mean CCVIA (Children's Continence and Quality of Life Assessment) scores  $> 3/5$  in all domains.

**CONCLUSION:** Sacral neuromodulation is a safe and effective long-term therapy for refractory fecal incontinence in pediatric patients. It significantly improves both continence and quality of life, with low complication rates when appropriate patient selection is applied.

**BOWEL AND URINARY FUNCTIONAL OUTCOMES IN SACROCOCCYGEAL TERATOMA**

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**PURPOSE:** Sacrococcygeal teratoma (SCT) is a rare congenital condition, with an estimated incidence of up to 1 in 40,000. Standard treatment consists of complete resection including coccygectomy. Long-term outcomes are variable due to the risk of urinary and bowel functional sequelae after resection. International reports describe long-term urinary and/or bowel functional sequelae in approximately 22–24% of patients. Aim of this study was to describe urinary and bowel functional outcomes in patients who underwent surgery for SCT at a single institution.

**METHODS:** We identified patients who underwent surgery for SCT between 2011 and 2025. Patients younger than 4 years were excluded. We recorded age at surgery, Altman classification, histopathology, management, age at last follow-up, and current urinary and bowel symptoms. Bowel function was assessed using the St Mark's Incontinence Score in patients with fecal incontinence and the Wexner Constipation Score in patients with constipation. Urinary symptoms were evaluated using the Dysfunctional Voiding Symptom Score (DVSS).

**RESULTS:** Nine patients were identified; seven could be contacted and were included in the analysis. Five patients (71.4%) were female. Six (85.7%) had a prenatal diagnosis. According to the Altman classification, three patients (42.9%) were stage I, three (42.9%) stage II, one (14.3%) stage III, and none were stage IV. Histopathology showed mature teratoma in six patients (85.7%) and immature teratoma in one (14.3%); no malignant tumors were identified. No patient reported fecal incontinence. Wexner Constipation Scores were 0 in one patient (14.3%), 1 in two (28.6%), 2 in two (28.6%), and 4 in two (28.6%). Five of seven patients experienced SCT recurrence, managed with surgery with or without chemotherapy. DVSS scores were 0 in three patients (42.9%), 1 in two (28.6%), 3 in one (14.3%), and 4 in one (14.3%).

**CONCLUSION:** In this series, none of the assessed patients met thresholds for clinically significant fecal incontinence (St Mark's  $\geq 8$ ), constipation (Wexner  $\geq 5$ ) or voiding dysfunction (DVSS  $\geq 6$ ). Our findings suggest favorable functional outcomes. Results might not be generalizable, because of relatively good baseline prognosis of cases, the small sample size, and single-center design.

## **AN EASY AND LOW-COST OPTION FOR NON-COMPLICATED EXTERNAL HEMORRHOIDS**

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(Hermosillo, Mexico)

**PURPOSE:** We aimed to present the results of sclerotherapy with hypertonic saline solution at 17.7% (17.7% NaCl) as a low-cost option treatment for non-complicated external hemorrhoids.

**METHODS:** Seven patients were included: five boys and two girls, with a history of hemorrhoids ranging from 6 months to 3 years, with no family history or history of constipation. Presenting symptoms included foreign body sensation, pruritus, pain, and fecal tenesmus. All patients were previously treated by different physicians with laxatives, topical lotions, and/or herbal remedies. Each procedure was performed in the operating room under sedation, with the patient in the lithotomy position, without prior bowel preparation, and with hypertonic saline at 17.7%. A Lone Star separator was used and 0.5 ml of the solution was injected into each hemorrhoidal packet. After the procedure, each patient was discharged from the hospital and began treatment with sennosides.

**RESULTS:** The first patient who was treated, required three treatment sessions, each one spaced two weeks apart, as the hemorrhoids did not regress adequately. However, the other six patients achieved complete resolution of their hemorrhoids with just one injection session. Neither patient experienced any intra- or postoperative complications. Currently, there has been a follow-up period of 3 months to 4 years without hemorrhoid recurrence.

**CONCLUSION:** Treatment for uncomplicated external hemorrhoids with 17.7% saline-based sclerotherapy is a simple, safe, and cost-effective option with successful results and a very low complication rate.

## CAN NEOVAGINA PROLAPSE BE PREVENTED IN PATIENTS WITH ANORECTAL MALFORMATIONS?

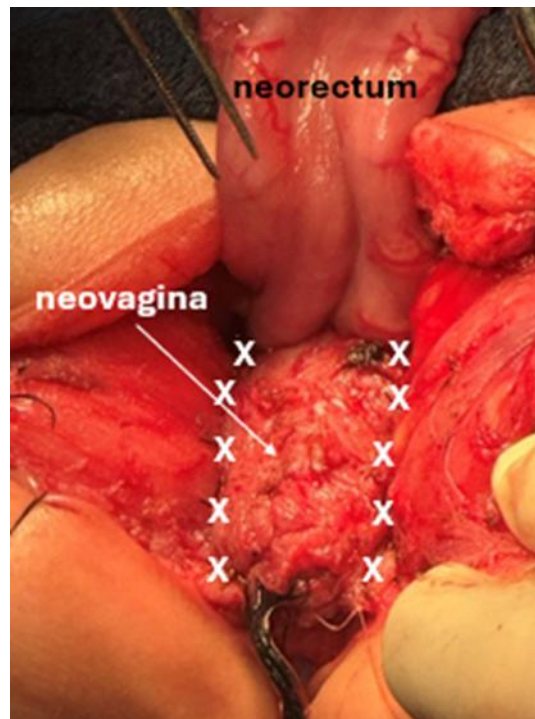
A. Bischoff, L. Sucre, D. Doster, L. de La Torre, J. Ketzer, A. Pena, V.I. Alaniz  
(Aurora, USA)

**PURPOSE:** Patients with anorectal and urogenital malformation may require bowel vaginal replacement. Although uncommon, neo-vagina prolapse is a challenging complication that seems to be more common in patients with poor sacrum and poor suspensory pelvic mechanisms (pelvic musculature). No preventative strategies have been described in the literature for this patient population. This report presents a surgical technique developed in an attempt to prevent neo-vagina prolapse.

**CASE PRESENTATION AND SURGICAL TECHNIQUE:** A seventeen-month-old female patient with a late diagnosis of rectoperineal fistula, poor sacral ratio (0.3), and absent uterus and vagina underwent a posterior sagittal approach with mobilization of the rectum. Mullerian agenesis was confirmed intra-operatively. Since the patient had a poor functional prognosis for bowel control, the rectum was used to replace the vagina, preserving the mesorectum as blood supply. To prevent future neo-vagina prolapse, the lateral walls were anchored to the surrounding musculature with multiple interrupted 5-0 non-absorbable sutures. The upper rectum was pulled down to create a neo-anus. The immediate postoperative course was uneventful with appropriate healing during early follow-up.

**CONCLUSION:** Fixation of the lateral walls of the neo-vagina to the pelvic muscles may decrease the incidence of post-operative neo-vagina prolapse. Long-term follow-up is needed to confirm the efficacy of the proposed technique.

**Figure.** Neovagina with "X" demonstrating fixation sutures of the lateral walls to the surrounding musculature.



## RECTOVESTIBULAR FISTULA WITH ABSENT VAGINA: TECHNICAL NUANCES AND CLINICAL CONSIDERATIONS

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**PURPOSE:** Rectovestibular fistula (RVF) with vaginal agenesis is a rare, frequently misdiagnosed variant of anorectal malformations presenting with only two perineal openings (urethra and fistula) and absent vaginal introitus. Reconstruction options include separating the fistula for neo-anus creation with vaginal replacement or preserving the fistula as neovagina with rectosigmoid pull-through. Long-term outcome data, particularly regarding bowel and menstrual functions, remain limited. We report our institutional experience with the latter approach.

**METHODS:** We retrospectively reviewed girls with RVF and vaginal agenesis managed at a single tertiary center (2016–2026). Diagnosis was established by perineal inspection, distal colostogram, and cystoscopy. All patients underwent initial diverting colostomy. Definitive repair preserved the RVF and distal rectum as neovagina. Transabdominally, the proximal rectosigmoid was anastomosed to Müllerian structures, followed by mobilization and pullthrough of the proximal colon through the sphincter complex as neo-anus. Müllerian anatomy was classified per ASRM criteria. Bowel function was graded by the Krickenbeck score.

**RESULTS:** Six patients underwent repair at a median age of 3.2 years (range 2–5). Anatomic spectrum included distal vaginal atresia with normal upper vagina/cervix/uterus (n=4) and cervicovaginal agenesis with functional uterine corpus (n=2). Uterine anomalies comprised didelphys (n=2) and bicornuate (n=1) uteri; all had patent endometrial cavities. All patients underwent RVF preservation as neovagina and utero-neovaginal anastomosis with proximal colon pull-through at the time of repair. No perioperative complications were recorded. At a median follow-up period of 6.2 years (range 1-10 years), all patients demonstrated voluntary bowel movements. Soiling occurred in three patients (Grade 1, n=1; Grade 2, n=2). Constipation was managed conservatively (Grade 1, n=3; Grade 2, n=3). Neovaginal caliber was clinically adequate (>6 cm) in all. Among the four post-pubertal patients, all experienced regular menstrual egress through the neovagina.

**CONCLUSION:** High clinical suspicion is mandatory to diagnose vaginal agenesis in RVF. Preserving the native RVF as a neovagina with proximal colon pull-through is a reproducible, low-morbidity strategy that yields favorable bowel function and permits normal menstruation in patients with functional uterine tissue. Long-term follow-up into adulthood is required to clarify sexual function and fertility potential.

## OVERLOOKED IMPERFORATE ANUS AND VAGINAL AGENESIS IN EARLY INFANCY: DIAGNOSTIC CHALLENGES AND SURGICAL MANAGEMENT

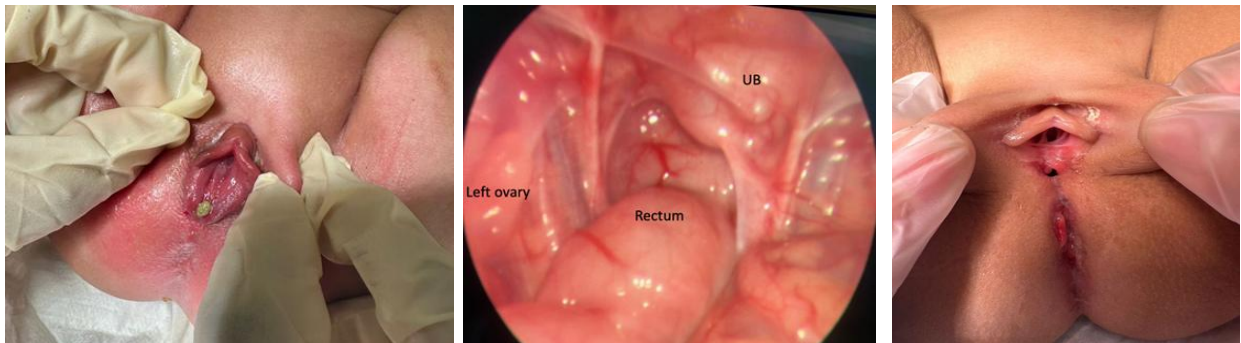
A. Hakeem, J. Almmary  
(Riyadh, Saudi Arabia)

**PURPOSE:** To emphasize the importance of careful neonatal perineal examination, early detection of anorectal and genital anomalies and timely referral to specialized center for optimal management.

**CASE PRESENTATION:** We report the case of a female infant in whom both imperforate anus and vagina agenesis were overlooked during the neonatal period. A 22-month-old girl was first referred to the Pediatric Surgery Department at three months of age after her mother noted the absence of an anal opening. She was diagnosed with an anorectal malformation (ARM), rectovestibular fistula type. A comprehensive evaluation for VACTERAL association revealed small atrial septal defect, normal sacrum, normal spine with the conus terminating at L1, a small filar cyst, and a solitary right ectopic kidney.

Examination Under Anesthesia (EUA) demonstrated a single anterior urethral opening, imperforate anus with rectovestibular fistula and no identifiable vaginal opening (**Figure 1**). Diagnostic laparoscopy revealed absence of the uterus, right ovary, and both fallopian tubes with presence of only a left ovary (**Figure 2**). A diverting sigmoid colostomy was performed.

At time of definitive reconstruction, repeat laparoscopy confirmed complete absence of the vagina, uterus and both fallopian tubes, with both ovaries present. Definitive reconstruction was achieved via an abdominoperineal approach, consisting of posterior sagittal anorectovaginoplasty. The rectum was preserved, and a stoma pull-through was performed with total vaginal replacement using the left descending colon. Follow up EUA demonstrated three distinct perineal openings: urethral meatus, colon neovagina, and neoanus (**Figure 3**). The neovagina measured 9 cm in length, the vaginal introitus accommodated a size 5 Hegar, and the neoanus was appropriately located within the muscle complex, accommodating size 13 Hegar dilator.



**CONCLUSION:** Imperforate anus with concomitant vaginal agenesis is a rare congenital anomaly that maybe overlooked at birth, particularly when perineal examination is incomplete or findings are subtle. Early recognition and comprehensive multidisciplinary evaluation are critical to avoid delayed diagnosis and to enable optimal staged reconstructive management.

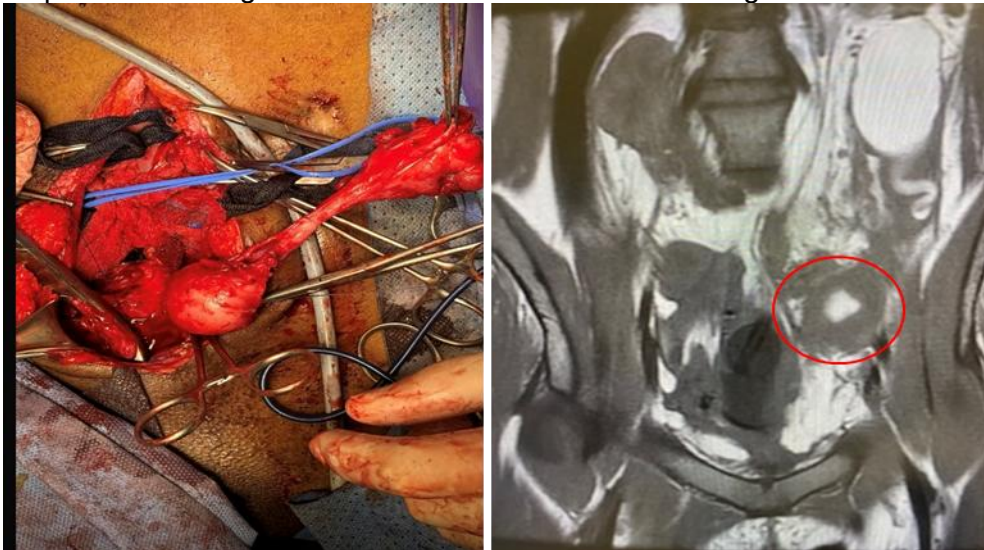
## LATE PRESENTING OBSTRUCTED MÜLLERIAN ANOMALIES FOLLOWING CLOACAL REPAIR: TWO ADOLESCENT CASE REPORTS

F. Abdulfattah, J. Almaary  
(Riyadh, Saudi Arabia)

**PURPOSE:** Non-communicating obstructed hemivagina is a rare Müllerian duct anomaly that may remain undiagnosed until adolescence, often due to subtle or absent symptoms in early life. We report the cases of two adolescent girls with cloaca anomalies, in whom uterus didelphys with a non-communicating obstructed hemivagina was initially missed, highlighting delayed presentation and diagnostic challenges associated with previously successful cloacal anomaly repairs.

**CASE PRESENTATION:** We report two adolescent girls with cloacal anomalies and normal menstruation who presented with recurrent lower abdominal pain despite normal menstrual cycle. The first patient, a 17-year-old girl with a history of cloaca repair, solitary kidney, neurogenic bladder, end stage renal disease, and renal transplantation, developed cyclic pain at 13 years of age. Pelvic MRI revealed a right unicornuate uterus connect to a proximal distended fluid filled vagina, and a left cavitated non-communicating rudimentary horn with hydrosalpinx. Surgical excision of the left Müllerian structure resulted in complete symptom resolution (**Figure 1**).

The second patient, a 13- year- old with cloacal anomaly managed early in life with colostomy and vesicostomy, single ectopic kidney, and chronic kidney disease on hemodialysis. After optimizing her renal condition, posterior sagittal anorecovaginoplasty with primary vaginal pull-through was performed. She presented with cyclic pain at 12 years of age. Pelvic MRI demonstrated a right elongated unicornuate uterus connected to a stretched vagina, and a contralateral non-communicating rudimentary horn containing functional endometrium associated with left hematosalpinx (**Figure 2**). She was initially managed with hormonal suppression therapy and planned for surgical excision of the non-communicating Müllerian structure.



**CONCLUSION:** Non-communicating obstructed hemivagina associated with cloaca anomalies may present late in adolescence, even after successful cloacal reconstruction. A high index of clinical suspicion is required in patients presenting with cyclical abdominal pain despite normal menstruation. Detailed anatomical evaluation using pelvic MRI is essential in providing diagnostic and surgical roadmap.

## RECTAL STAB INJURIES IN CHILDREN: A SINGLE-CENTRE EXPERIENCE

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(Prague, Czech Republic)

**PURPOSE:** Penetrating anorectal injuries in children are rare. Despite the historical importance of colostomy in preventing infectious complications, primary repair is often feasible. This study aimed to review our experience in the management of rectal stab injuries.

**METHODS:** We performed a retrospective study of patients who underwent surgery for penetrating rectal injury at our institution between 2010 and 2025. Medical records and operative findings were analysed. Long-term bowel and bladder function were also evaluated.

**RESULTS:** Three patients were identified (two boys and one girl), aged 5–14 years. In all cases, the mechanism of injury was impalement by a metal object (a toy car axle and a rebar rod). The leading presenting symptoms were rectal bleeding and perineal pain, followed later by abdominal pain. One injury was extraperitoneal, and two were intraperitoneal. The extraperitoneal injury was managed with a diverting colostomy, which was closed after six weeks. The two patients with intraperitoneal injuries underwent laparoscopic exploration of the abdominal cavity. In both cases, the transmural rectal injury was treated with primary repair. One patient had an associated bladder wall laceration and in one case, an associated ileal injury required resection with primary anastomosis. No postoperative complications occurred. Follow-up period ranged from 5 months to 12 years. Urinary and faecal continence were preserved in all patients during follow-up.

**CONCLUSION:** Primary repair of transmural rectal injuries in children appears to be a safe and feasible option. However, in cases of delayed diagnosis or significant bacterial contamination, faecal diversion remains an appropriate therapeutic approach in the paediatric population.

## REPAIR OF ACQUIRED ANOVESTIBULAR FISTULAE

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(Cape Town, South Africa)

**PURPOSE:** Paediatric acquired anovestibular fistula (aAVF), without prior perineal trauma or surgery, is a classic presentation of HIV-associated “perineal disintegration syndrome”. Surgical morbidity and recurrence rates remain high, with few described approaches. Perirectal fat-pad interposition with rectal advancement flaps reduce recurrence but compromise continence. We describe a case series of patients treated with a continence-preserving approach.

**METHODS:** A retrospective medical record review of paediatric aAVF was performed at our centre for 2019-2026.

**RESULTS:** Three girls with aAVF presented at a mean age of 7.6 months (2 – 18 months). All had a history of stool passage through the vaginal vestibulum, loose stools and an active (67%) or history of (33%) severe fungal napkin dermatitis. Pain during defecation (67%) was associated with severe perineal dermatitis, treated with topical and oral antifungals. All patients had a normally-sited anus with simple anterior fistula (1-5 mm diameter) at anal crypt-level and no associated anomalies. Two were maternally HIV-exposed (66%) with one HIV-positive (33%). One patient (33%) had active cytomegalovirus infection treated with ganciclovir. Tuberculosis testing was negative. All failed a 6-month-minimum trial of conservative management, despite nutritional and immunological optimization. Median age at surgery was 10 months (range 2–60 months). One patient had a covering colostomy; the others had preoperative bowel preparation with oral antibiotics. All patients underwent fistulectomy and repair using fat-pad interposition. A vertical elliptical incision was made around the fistula orifice on the anorectal side and a transverse incision on the vestibular side. Excision was in close proximity to the tract to preserve anal tissue and sphincter fibres and the proximal vagina was mobilized slightly off the rectum. Pedicled perirectal fat was interposed from the vestibular side. All aAVF tract histology showed non-specific inflammation. Post-operative course was uneventful and the eldest patient has successfully potty trained (follow-up range 1–24 months).

**CONCLUSION:** Fistulectomy with vertical anal wall and tangential vestibular mucosal closure around fat-pad interposition is a safe and effective technique for aAVF management, avoiding disruption of the sphincter muscle and dentate line.

## **BENIGN INFANTILE PROCTOCOLITIS: AN IMPORTANT DIFFERENTIAL FOR RECTAL BLEEDING IN INFANCY**

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(New Delhi, India)

**PURPOSE:** Rectal bleeding in infancy often causes significant concern for caregivers and clinicians, frequently prompting extensive investigations. Food Protein–Induced Allergic Proctocolitis (FPIAP), also referred to as benign infantile proctocolitis, is a common but under-recognized cause of rectal bleeding in otherwise healthy infants. It represents a non–IgE-mediated hypersensitivity reaction to dietary proteins, most commonly cow’s milk protein, transmitted either through infant formula or maternal breast milk. The condition usually presents during the first few months of life with blood-streaked stools, mucus, or mild diarrhea and typically resolves spontaneously by the end of the first year. We present two cases that highlight the clinical course and diagnostic challenges associated with this condition.

**METHODS:** Two infants presenting with persistent rectal bleeding were evaluated and managed over 6 months. Clinical evaluation, stool studies, and imaging were performed to exclude infectious and structural causes.

**RESULTS:** The first case involved a 6-week-old male infant receiving mixed feeding who developed diarrhea followed by blood and mucus in stools. Stool cultures were negative and a Meckel’s scan was normal. Despite maternal dietary elimination of dairy, eggs, nuts, soy, fish, and wheat, bleeding persisted for several weeks. Following discontinuation of cow’s milk-based formula and substitution with an alternative milk preparation, the bleeding resolved within 48 hours.

The second case involved a 3-month-old female infant on mixed feeds who presented with progressively increasing rectal bleeding. Stool cultures were negative. Colonoscopy demonstrated minor mucosal ulcerations with features of sigmoiditis and proctitis. Persistent bleeding for nearly two months resolved rapidly after elimination of cow’s milk-based formula and dietary modification. At a follow-up of 12-16 months, both babies are doing well.

**CONCLUSION:** FPIAP should be considered in infants presenting with rectal bleeding despite otherwise normal clinical status. Early recognition can prevent unnecessary invasive investigations and anxiety. Giving a trial of extensively hydrolysed formula may be prudent before undertaking extensive investigations. Management consists of reassurance, elimination of offending dietary proteins, and gradual dietary reintroduction as tolerance develops. Awareness of this benign condition is essential for appropriate and timely management.

## TOTAL COLONIC DUPLICATION IN INFANCY: A CASE REPORT AND FOCUSED REVIEW OF OPERATIVE MANAGEMENT

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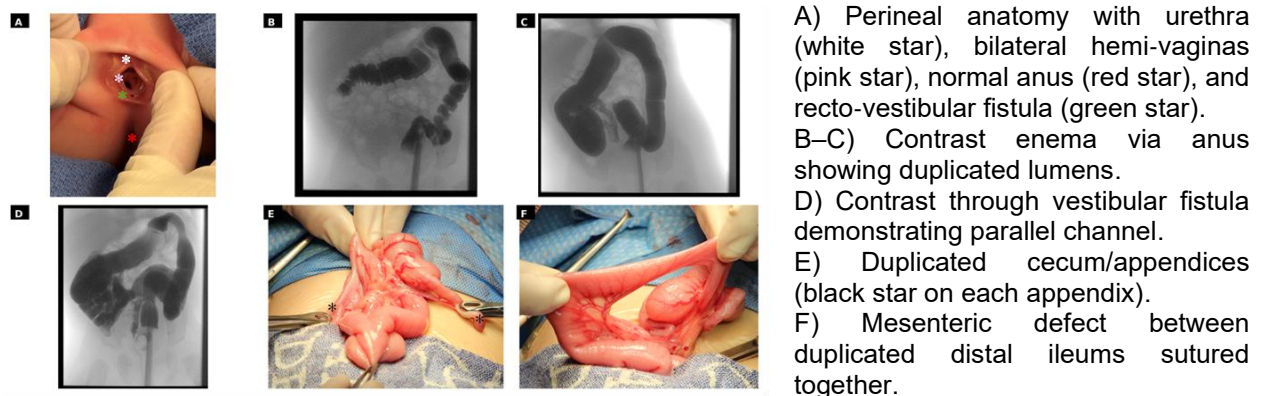
**PURPOSE:** Total colonic duplication (TCD) is an exceptionally rare congenital anomaly frequently associated with anorectal and genitourinary malformations. Presentation may mimic an H-type rectovestibular fistula in the presence of a normal orthotopic anus, leading to diagnostic uncertainty. Operative management is further complicated by shared mesenteric vasculature and complex distal anatomy. We present a case initially suspected to be an H-type fistula that was intraoperatively identified as TCD, emphasizing the role of the posterior sagittal anorectoplasty (PSARP) approach in definitive management.

**METHODS:** We report the case of an otherwise healthy 5-month-old girl presenting with stool passage per vagina, ultimately diagnosed with total colonic duplication (TCD). She underwent posterior sagittal excision of a rectovestibular fistula with colo-colonic anastomosis (lumen unification). Postoperative recovery was uneventful. A review of primary research studies describing TCD and its surgical management was performed. Reports limited to partial or segmental colonic duplications were excluded.

**RESULTS:** The patient initially presented at 3 months with stool passage per vagina and rectum. Examination under anesthesia revealed a rectovestibular fistula, normal anus, and vaginal septum with two cervixes. During planned PSARP at 5 months, two parallel, non-communicating colons sharing a common mesentery were identified, consistent with TCD. The posterior sagittal approach allowed precise fistula ligation under direct visualization with preservation of the sphincter complex. The duplicated distal segments were mobilized within the shared mesentery without vascular compromise. Lumen unification was achieved via side-to-side colo-colonic anastomosis, avoiding colectomy. The patient was discharged on postoperative day six without complication. Literature review demonstrates heterogeneous operative strategies, including common-wall division, mucosectomy, colectomy, and staged diversion, guided primarily by mesenteric vascular anatomy and distal configuration.

**CONCLUSION:** Careful preoperative and intraoperative anatomical evaluation is essential in TCD. In cases with shared mesentery, lumen unification rather than colectomy reduces ischemic risk while preserving bowel length and continence potential. Concurrent correction of associated anorectal and mesenteric anomalies can be performed safely in experienced colorectal centers.

**Figure.** Patient perineal and colonic anatomy.



## DUPLICATION OF THE RECTUM PRESENTING AS A PROLAPSING RECTAL MASS IN INFANTS

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**PURPOSE:** To present our experience with two cases of rectal duplications presenting as prolapsing rectal mass.

**CASE PRESENTATION:** 18-month-old female child presented with a large prolapsing rectal mass. She had difficulty in passing urine & straining to pass stools. The mass was palpable in the suprapubic region. CT scan & MRI showed a cystic duplication attached to rectum & compressing the rectum laterally. A satellite cyst was seen posteriorly. The L2 to L5 vertebral bodies were split. The lateral view showed an intraspinal cyst. At laparotomy, a trap door incision, a seromuscular flap was raised & the mucous cyst was gradually enucleated. The cyst was adherent to the lower end & ruptured. The collapsed cyst wall was retrieved through the rectum & excised completely. The mucosal & muscular defect repaired. The sero-muscular flap was sutured. The satellite cyst removed. The intraspinal (dermoid) cyst was removed later. A 2-year-old male child presented with difficulty in passing stool, a prolapsing rectal mass when straining, abdominal distension & subacute bowel obstruction. A mass was palpable in the supra pubic region. He developed acute bowel obstruction. At laparotomy, a tubular duplication was noted from the splenic flexure down to the rectum. It was on the anti-mesenteric side of the normal bowel. The duplicated segment of the descending colon down to the mid rectum excised. The residual rectal duplication's mucosa was removed & rectal wall was repaired. Bowel continuity restored. Both children recovered well.

**CONCLUSION:** Rectal duplications are rare & accounts for 1 to 8% of all duplications. Kraft in 1962 reviewed the literature & found 38 cases of tubular hindgut duplications and 32 were cystic lesions. The epithelial lining is usually colonic as in our cases, cystic duplications are best excised completely. Duplications can be removed by a trans-anal or modified posterior sagittal approach. Care should be taken not to compromise the normal rectal lumen. An abdominal approach is needed to excise lesions that extend proximally. Association with Split-Notochord syndrome (SNS) is very rare. A high index of suspicion is needed to diagnose these uncommon lesions.

## RARE COLORECTAL MANIFESTATIONS OF PEDIATRIC GASTROINTESTINAL LYMPHOMA: CLINICAL SPECTRUM AND SURGICAL IMPLICATIONS

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(Kochi, India)

**PURPOSE:** Pediatric colorectal malignancies are rare, with lymphomas- predominantly non-Hodgkin lymphoma (NHL) representing the most common type, accounting to 0.5-2 % of large bowel malignancies. Colorectal NHL can be primary or secondary. This study analyzes the rare manifestations of pediatric colorectal lymphoma and examines their diagnostic challenges and surgical implications.

**METHODS:** Pubmed search was done with search words "lymphoma of colon pediatric" with filter of last 5 years on 12 Feb 2026. The search comprised of 47 articles, among which 10 relevant articles were selected. After going through the articles, one article was excluded. 9 articles were finally included in the study.

**RESULTS:** Colorectal lymphoma can be a primary malignancy or it can arise in the background of inflammatory bowel disease. It can also be a secondary extension from bulky intra- abdominal lymphoma. Clinical manifestations can present either as flare up of inflammatory bowel disease or colorectal carcinoma or bowel obstruction or acute abdomen. This can lead to potential diagnostic delay and inappropriate escalation of medical therapy, unless early biopsy is performed. The routine use of broad-spectrum anti-infectives in lymphoma decreased the gastrointestinal integrity and increased opportunistic infections in the gut, complicating the clinical assessment. Imaging aids in differentiation from epithelial malignancies. It also guides in staging, treatment and operative planning. Rare presentations of colonic lymphoma were its appearance in a child (below five years) on medical management for ulcerative colitis, and in another patient on Vedolizumab for Crohn's disease. Another rare presentation recorded occurrence of primary rectal lymphoma in an eleven-year-old child. Early presentation (below 10 years) with synchronous colorectal carcinoma and family history of malignancy, suggested an underlying genetic cancer predisposition syndrome. Surgery plays a crucial role in managing complications like perforation, obstruction and intussusception when lymphoma is the lead point. Laparoscopy assisted en-bloc resection is feasible in selected cases.

**CONCLUSION:** Pediatric colorectal lymphomas represent a clinically significant and occasionally misleading entity. While systemic chemotherapy remains the cornerstone of treatment, surgical intervention has a defined role in tissue diagnosis and management of complications. Early recognition and coordinated multidisciplinary care are essential to optimize outcomes.

**COLO-RECTAL CARCINOMAS IN CHILDREN**

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(Kuala Lumpur, Malaysia)

**PURPOSE:** To present our experience with two cases of Colorectal carcinomas, one arising from the Colon and another from a rectal duplication.

**CASE PRESENTATION:** A 13-year-old female child presented 6 months after appendectomy, with a history of fever and diarrhea. Rectal examination revealed a large pelvic mass, and ultrasound showed a cystic mass. At laparotomy, 50ml of purulent mucoid fluid was aspirated & a drain was left. Streptococcus faecalis was grown and treated with antibiotics. She was afebrile and was discharged home. Two months later, she presented with weight loss and recurrence of symptoms. Rectal examination revealed a large, indurated mass anteriorly. Ultrasound and CT scan showed a well-defined solid & cystic mass. Trucut biopsy showed mucin-producing adenocarcinoma, probably of bowel origin. Carcinoembryonic antigen was elevated, 398 ng/mL (normal range: 0-5 ng/mL). At re-laparotomy, the pelvic mass was exposed & it resembled the bowel wall & was attached to the rectum. It was invading the surrounding structures. A diversion colostomy was performed. She was started on 5-Fluorouracil and advised to have radiotherapy. Parents refused further treatment. A 11-year-old male child presented with acute bowel obstruction, 3 weeks after appendectomy. Laparotomy showed a mass in the descending colon causing obstruction. A transverse colostomy was performed. Colonoscopy and biopsy showed adenocarcinoma. Subtotal colectomy & lymph node clearance was performed. Histopathology confirmed a mucinous adenocarcinoma, with carcinoid-like features in some areas. Lymph nodes were positive for tumor. He was given FLEX chemotherapy & recovered well. Bowel continuity was restored. He was alive and well for 2 years & developed secondaries in liver & abdomen. He died a month later.

**CONCLUSION:** Colorectal carcinomas in children, below the age of 15 years, are rare. Its occurrence is much rarer in a rectal duplication. Review of the literature shows, the diagnosis is often delayed due to its rarity, and the outcome is poor. Lack of suspicion results in an advanced stage (T3) at presentation. Less than 10 cases of adenocarcinoma & carcinoids are reported in Colorectal duplications in children & mostly in females. A high index of suspicion is essential for early diagnosis of colorectal carcinoma in children.

## PAEDIATRIC AFRICAN DEGENERATIVE LEIOMYOPATHY: A RARE BUT DEVASTATING DISORDER

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**PURPOSE:** African Degenerative Leiomyopathy (ADL) is a region-specific myopathic form of paediatric intestinal pseudo-obstruction (PIPO) in African children and is not compatible with long-term survival, with most deaths in the second decade of life from bowel complications. It presents with progressive abdominal distension due to intestinal smooth-muscle degeneration, often with urinary and cardiac anomalies, and is diagnosed histologically by characteristic “tiger stripes” in the muscularis propria. We describe our institutional experience with paediatric ADL at a tertiary hospital in South Africa.

**METHODS:** A retrospective review of paediatric patients with ADL in our institutional database was performed. Data on presentation, diagnostic investigations, surgical interventions, complications, and outcomes were extracted.

**RESULTS:** Twenty-three patients were included. Mean age at presentation was  $9.87 \pm 3.18$  years, with a female-to-male ratio of 11:12. The most common presenting symptoms were abdominal distension (15/23 - 65%), constipation (13/23 - 57%), and bowel obstruction (5 - 22%). Urinary or cardiac information was available for 9 patients (5 normal, 3 renal and 1 cardiac anomalies).

Family history data were available in 9 patients: 2 had an affected sibling, and 3 had a parent from the Eastern Cape, the region with the highest reported incidence of ADL.

Diagnosis was confirmed histologically in 19 patients (full-thickness rectal biopsy in 10 and mapping biopsies in 9). Four patients were included based on typical clinical presentation or family history.

Rectal irrigations and flatus tube decompression were initiated in all patients. Palliative surgery was performed in 10/23 patients (43.5%), including venting colostomy with Mic-key button in 4 (17.4%), colectomy in 6 (26.1%). Post-operative complications occurred in 8 patients [5/8 (62.5%) bowel obstruction, 1/8 (12.5%) leaking Mic-key button, 1/8 (12.5%) Mic-key button dislocation, and 1/8 (12.5%) abdominal compartment syndrome).

Seven patients died during the study period (mean age  $12.7 \pm 3.95$ ), although mortality may be underestimated as only 9 remain in active follow-up.

**CONCLUSION:** ADL is a rare but severe gastrointestinal disease associated with high morbidity and mortality. Conservative management remains the main approach due to the high rate of surgical complications. Early diagnosis and prompt symptom management are essential to improve quality of life and delay complications in resource-limited settings.

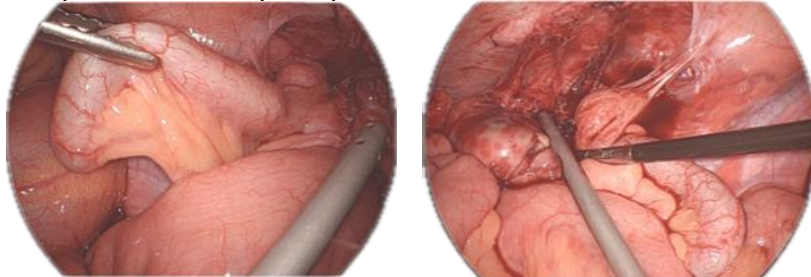
## PEDIATRIC CECAL DIVERTICULITIS: CLINICAL PRESENTATION, MANAGEMENT, AND REVIEW OF THE LITERATURE

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(St. Louis, USA)

**PURPOSE:** Acute diverticulitis is extremely uncommon in the pediatric population and often presents with symptoms indistinguishable from acute appendicitis. We report a case of cecal diverticulitis in an adolescent and present a structured review of the literature to characterize clinical features, diagnostic pitfalls, and management in children.

**METHODS:** Clinical data for the index patient were retrospectively reviewed from medical records, and parental consent was obtained. A structured literature search was performed using PubMed, Scopus, and Embase with predefined keywords. English language studies reporting patients younger than 18 years were included. Eligible publications consisted of case reports and case series.

**RESULTS:** A previously healthy 16-year-old male presented with three days of right lower quadrant abdominal pain. Laboratory evaluation showed a WBC of  $11.5 \times 10^3/\mu\text{L}$  and a CRP of 53.9 mg/L. Computed tomography demonstrated findings suspicious for cecal diverticulitis. Initial conservative management with oral antibiotics was attempted, but worsening pain prompted diagnostic laparoscopy. During laparoscopy an inflammatory cecal mass was identified. The appendix appeared normal and was adjacent to a single non perforated cecal diverticulum. Because of surrounding inflammation appendectomy was deferred to avoid potential stump complications. The postoperative course was uneventful.



The literature search identified 27 studies including three case series and 24 case reports comprising 101 pediatric patients. Most cases (77%) originated from Asia. Median age at presentation was 11 years (IQR 9-15) with no sex predominance (male 51%). The ascending colon, predominantly the cecum, was involved in 81% and a solitary diverticulum was present in 83%. Abdominal pain occurred in 100% of patients. Other common symptoms were fever (34%), nausea or vomiting (34%), anorexia (32%) and constipation (9%). CT was performed in 34% of cases and correctly suggested diverticulitis in 62%. Complicated diverticulitis was reported in 34% of the cases (perforation 23%, abscess 9%, fistulas 6%). Preoperative misdiagnosis, mostly appendicitis, occurred in 44% of patients and diverticulitis was correctly identified intraoperatively one third of the times.

**CONCLUSION:** In children and adolescents diverticulitis most often arises from a solitary cecal diverticulum and closely mimics appendicitis. Awareness may improve diagnosis, and uncomplicated cases can be treated conservatively with careful follow-up and appropriate clinical monitoring.

## MEGAPOUCH AFTER ILEAL POUCH ANAL ANASTOMOSIS FOR ULCERATIVE COLITIS IN PEDIATRIC PATIENTS: TWO LONG TERM CASES

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(Pavia, Italy)

**PURPOSE:** Long term complications of ileal pouch anal anastomosis (IPAA) performed for ulcerative colitis (UC) during childhood remain poorly described. Recent studies suggest that patients undergoing colectomy and IPAA at younger ages may have higher rates of pouch related complications, including pouchitis, pouch failure, and Crohn like disease (CLDI) of the pouch. Some adult IBD experts have suggested delaying pouch reconstruction until after puberty in order to reduce late complications. We report two patients who developed megapouch after IPAA performed during prepubertal age for severe UC.

**CASE 1:** Female, severe UC, with massive persistent bleeding, no-responsive to medical therapy; PUCAI > 65; at 11 years she underwent to colectomy and terminal ileostomy. IPPA with 7 cm hand-sewn pouch was performed 6 months later. After being lost to follow up, she represented at the age of 31 years with chronic pouchitis. Magnetic resonance imaging demonstrated a large megapouch with suspected CLDI disease of the pouch. She is currently receiving medical treatment with ustekinumab, with partial clinical improvement, although surgical pouch revision may be required.



Fig. 1. MR study of the megapouch

**CASE 2:** Male, UC patient submitted at the age of 10 years to total colectomy and ileostomy. During follow-up he developed symptomatic pouch complications consisting of megapouch associated with pre-pouch ileal stenosis and intermittent volvulus. Redo surgery with pouch reconfiguration and temporary ileostomy was performed at the age of 21 years. Postoperative recovery was uneventful.



Fig. 2. Rectal enema:  
Pre-pouch ileal stenosis

Fig. 3. Reconfiguration of megapouch

**CONCLUSION:** Megapouch may represent a late complication of IPAA performed in childhood. Possible contributing factors include pouch growth during puberty, increased risk of pouchitis, and development of Crohn like disease of the pouch. These observations support careful consideration of the timing of pouch reconstruction after colectomy and highlight the importance of long term multidisciplinary follow up involving both pediatric and adult IBD surgeons.

## **INTRACORPOREAL MAPPING OF THE SPHINCTER COMPLEX IN LAARP USING A STANDARD TRANSCUTANEOUS STIMULATOR: A VIDEO PRESENTATION OF A LOW-COST TECHNIQUE**

I. Elhalaby, O. Koura, M. Eid, E. Elhalaby  
(Tanta, Egypt)

**PURPOSE:** Optimal continence after laparoscopic-assisted anorectoplasty (LAARP) for high anorectal malformations depends on accurate centering of the neo-rectum within the sphincter–levator complex. Conventional perineal mapping with external stimulators provides only an indirect approximation of the underlying muscle anatomy and may result in off-center tracts. Dedicated laparoscopic stimulators enable direct intracorporeal mapping but are expensive and not widely available. This video presentation aims to demonstrate a simple, low-cost technique for intracorporeal sphincter complex mapping in LAARP by repurposing a standard transcutaneous muscle stimulator and routine laparoscopic instruments.

**METHODS:** A 5-month-old male infant with a rectoprostatic fistula underwent LAARP. Following laparoscopic rectal mobilization and fistula division, the levator hiatus and sphincter complex were exposed. The output lead of a conventional transcutaneous muscle stimulator was connected to the proximal metal portion of a 5 mm laparoscopic instrument, using the insulated shaft to conduct current to the distal tip. Under laparoscopic visualization, the tip was applied sequentially to the levator hiatus. Intracorporeal contraction of the sphincter–levator complex was observed and simultaneously correlated with perineal skin response. The pull-through tract was created through the point of maximal functional contraction via precise muscle splitting, followed by serial dilatation, rectal pull-through, and anocutaneous anastomosis.

**RESULTS:** The video provides step-by-step visualization of the setup and application of intracorporeal stimulation, the resulting localized contraction of the sphincter–levator complex, and real-time concordance with external perineal mapping. In this index case, the technique allowed clear identification of the functional center of the sphincter complex and a midline, sphincter-centered pull-through without the need for additional specialized equipment. The maneuver was easily integrated into the standard LAARP workflow and added minimal operative time.

**CONCLUSION:** Repurposing a standard transcutaneous muscle stimulator through insulated laparoscopic instruments effectively creates a laparoscopic muscle stimulator from existing resources. This low-cost, reproducible technique provides direct intracorporeal functional mapping of the sphincter–levator complex and may assist colorectal surgeons in improving anatomic accuracy and potentially continence outcomes in LAARP and related procedures.

## RECTO-VESTIBULAR FISTULA WITH NORMAL ANUS IN A FEMALE INFANT

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(Taipei, Taiwan)

**PURPOSE:** H-type recto-vaginal and recto-vestibular fistulas represent a rare subset of anorectal malformations (ARMs), characterized by an abnormal communication between a normally situated anus and the genital tract. While these malformations are documented globally, they exhibit a significantly higher incidence in Asia compared to Western nations. Diagnosis is frequently delayed, as standard imaging often fails to delineate the fistula tract; direct inspection under anesthesia remains the diagnostic gold standard. Several surgical approaches have been reported with varying advantages and disadvantages, and postoperative recurrence rates range from 5% to 30%.

**METHODS:** We present a case of a female infant with an H-type recto-vestibular fistula and provide a comprehensive literature review.

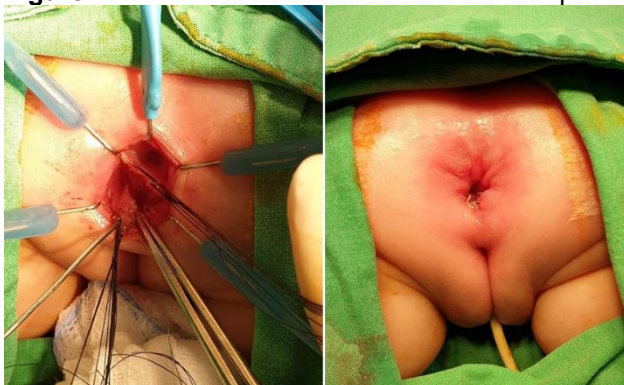
**RESULTS:** A two-month-old highland indigenous female infant presented with recurrent vulvar abscesses that were non-responsive to antibiotic therapy. Detailed physical examination and careful probing under direct inspection in the outpatient clinic revealed a suspected recto-vestibular fistula. Following surgical repair, there was no recurrence over a six-year follow-up period. Reported surgical approaches primarily include transanal mobilization, endo-rectal mucosal advancement flap, anterior sagittal anorectoplasty, and simple resection. We opted for the endo-rectal mucosal advancement flap due to its advantages in preserving the perineal body, achieving excellent cosmetic outcomes, and obviating the need for a protective colostomy.

**CONCLUSION:** Although surgical approaches vary, postoperative outcomes remain comparable. Clinical suspicion and meticulous examination are vital for achieving an early diagnosis and ensuring a favorable prognosis.

**Figure 1.** Passage of stool via vestibular area and probe used to identified recto-vestibular fistula.



**Figure 2.** Endorectal mucosal advancement flap.



## UNUSUAL ASSOCIATION OF CLASSIC BLADDER EXSTROPHY WITH ANAL STENOSIS: A CASE REPORT

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(New Delhi)

**PURPOSE:** Classic bladder exstrophy is a rare congenital anomaly usually associated with a rectal prolapse. We report an unusual case of classic bladder exstrophy associated with anal stenosis in an infant.

**METHODS:** We report an unusual case of bladder exstrophy associated with anal stenosis

**RESULTS:** A male infant presented at 6 months of age with an exposed bladder plate following two failed bladder exstrophy repairs performed elsewhere at approximately one month of age. The first repair dehiscenced on postoperative day five, and a redo exstrophy–epispadias repair failed within three days, although the epispadias repair remained intact. On examination at presentation, the child had classic bladder exstrophy with a protruding bladder plate, left undescended testis, and marked anal stenosis associated with chronic constipation. Digital rectal examination was not possible due to narrowing of the anal canal that could only be negotiated with an infant feeding tube size of 6 french. The anal opening was dilated to 8 and 10 french feeding tube. The anal dilatation was repeated under general anesthesia to negotiate a size 8/10/12 hegar dilator and little finger. The exstrophy repair was then done. The baby recovered well and the wound healed well. The baby has a dry interval of about 20 mins in the day time and almost dry in night time.

**CONCLUSION:** Recognition and management of the associated anorectal pathology played a crucial role in achieving successful redo closure. This case highlights the importance of routine anorectal evaluation in patients with bladder exstrophy, particularly in redo cases, and contributes to the limited literature on colorectal associations in classic bladder exstrophy.

## **A RARE CASE REPORT OF HIRSCHSPRUNG DISEASE IN 11-MONTH-OLD MALE CHILD OPERATED VIA SOAVE TECHNIQUE FOLLOWED BY RESECTION OF 57 CM OF THE AGANGLIONIC COLONIC SEGMENT**

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(Skopje, North Macedonia)

**PURPOSE:** Hirschsprung's disease represents a neurocristopathy, caused by defective migration, proliferation, differentiation and survival of neural crest cells, ultimately leading to gut aganglionosis. While the disease typically involves shorter colonic segments, extensive aganglionosis is rare and presents significant diagnostic and therapeutic challenges. Our purpose is to report a challenging case report of Hirschsprung's disease in 11-month-old male child.

**METHODS:** Presentation of an extremely challenging case report study of Hirschsprung's disease in 11-month-old male child.

**RESULTS:** We report the case of an 11-month-old male child with chief medical concerns of chronic constipation, abdominal distension, and failure to thrive since birth. Irigography revealed a markedly dilated proximal colon with a transition zone in the recto-sigmoid colon. Definitive diagnosis was confirmed by colon biopsy, followed by Malone-Appendicostomy. The patient undergone irrigation 3 times per week during three weeks, then, the patient underwent surgical management using the Soave endorectal pull through technique. Intraoperative findings revealed an unusually long aganglionic segment, necessitating resection of 57 cm of colon. Postoperative recovery was uneventful, with gradual improvement in bowel function and nutritional status. At follow up, the child demonstrated satisfactory growth and complete resolution.

**CONCLUSION:** Our case emphasizes the significance of early recognition of atypical presentations of Hirschsprung's disease, as delayed diagnosis may result in severe complications such as enterocolitis and growth retardation. Additionally, it underscores the adaptability of the Soave technique in managing extensive aganglionosis, offering favorable outcomes even in rare and complex cases. Reporting such unusual presentations contributes to the existing literature and aids clinicians in anticipating surgical challenges, tailoring operative strategies, and optimizing patient care in pediatric colorectal surgery.

## **INTESTINAL POUCH RESERVOIR FORMATION IN TOTAL COLONIC AGANGLIOSIS: A CASE STUDY ON RE-OPERATIVE MANAGEMENT IN A 2-YEAR-OLD PATIENT**

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(Saint Petersburg, Russia)

**PURPOSE:** The pathogenetic basis of surgical treatment for Hirschsprung disease is the resection of the aganglionic zone. Aim was to highlight the potential complications of reservoir techniques in patients with total colonic aganglionosis (TCA).

**CASE PRESENTATION:** In 2025, a 2-year-old boy was admitted to the Clinic of Saint Petersburg State Pediatric Medical University following surgical treatment for TCA at another healthcare institution. Upon admission, the patient presented with complaints of chronic constipation (absence of spontaneous bowel movements), dependency on daily enemas, abdominal distension, and foul-smelling loose stools. Additionally, the child exhibited periodic episodes of dehydration and anemia.

**Medical History:** From the age of 8 months, the patient experienced bowel movement delays, decreased weight gain, and abdominal distension. At 13 months, he underwent surgery consisting of a laparotomy, colonic revision, resection of the ileocecal angle, ileostomy, and colostomy. The histological findings confirmed TCA. At 18 months, a subtotal colectomy was performed, and a tunnel was created along the posterior surface of the rectum. Through this tunnel, a stapled anastomosis was established between the small intestine and the rectum. Postoperatively, the child experienced a month-long fever, high inflammatory marker activity, and a lack of spontaneous stools.

Upon admission to the Pediatric University endoscopic examination revealed signs of anastomosis, and granulations in the pseudodiverticulum area. In the first stage, a protective enterostoma was created. Two weeks later, following a reduction in stoma output, definitive surgery was performed: a laparotomy with resection of the reservoir and aganglionic rectum, followed by a Swenson ileoanal anastomosis.

During the postoperative period, a profuse discharge of clear fluid from the anus was observed; this was interpreted as a reactive response of the bowel to prolonged obstruction and chronic inflammation. Two months later, once the stool had thickened to an 'applesauce' consistency, the ileostomy was closed. At the 12-month follow-up, the patient had up to four spontaneous bowel movements per day, with only episodic nighttime fecal soiling.

**CONCLUSION:** Pouch procedures performed in the presence of residual aganglionic segments in TCA fail to enhance quality of life and reoperations associated with a possible complication rate.