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REVIEW ARTICLE

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Post-operative colonic manometry in children with anorectal malformations: A systematic review

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Abstract

Background: Children with anorectal malformations may experience constipation and fecal incontinence following repair. The contribution of altered anorectal function to these persistent symptoms is relatively intuitive; however, colonic motility in this cohort is less well understood. Manometry may be used to directly assess colonic motility.

Purpose: The purpose of this systematic review was to synthesize the available evidence regarding post-operative colonic motility in children with anorectal malformations and evaluate the reported equipment and protocols used to perform colonic manometry in this cohort. This systematic review was conducted in compliance with Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA). We conducted a systematic review of four databases: Embase, MEDLINE, PubMed, and the Cochrane Library (1st January 1985–22nd July 2021). Studies reporting colonic manometry performed in children following anorectal malformation repair were assessed for eligibility. Data were extracted independently by two authors. Four studies were eligible for inclusion. Of the combined total cohort of 151 children, post-operative colonic manometry was conducted in 35. Insufficient reporting of medical characteristics, bowel function, and manometric outcomes restricted comparison between studies, and limited clinical applicability. No results from high-resolution colonic manometry were identified. Despite the prevalence of post-operative bowel dysfunction in children with repaired anorectal malformations, this systematic review highlighted the markedly limited evidence regarding post-operative colonic motility. This cohort may benefit from assessment with high-resolution techniques; however, future work must emphasize adherence to standardized manometry protocols, and include robust reporting of surgical characteristics, bowel function, and manometric outcomes.

KEYWORDS
anorectal malformation, colon, gastrointestinal motility, manometry

Abbreviations: –, not reported; ARM, anorectal malformation; CM, colonic manometry; EAS, external anal sphincter; FI, fecal incontinence; IAS, internal anal sphincter; MWS, Modified Wingfield Score; NOS, Newcastle-Ottawa Scale; PRISMA, Preferred Reporting Items for Systematic Reviews and Meta-Analyses; PROSPERO, International Prospective Register of Systematic Reviews; PSARP, posterior sagittal anorectoplasty; QoL, quality of life; WP, water-perfused.

1 | INTRODUCTION

Anorectal malformations (ARM) result from abnormal development of the hindgut. Affecting one in 3000–5000 children, they exist on a spectrum, ranging from those requiring limited operative intervention, to more complex anomalies necessitating multi-system repair. The overarching principles of surgical management are to disconnect the fistulous connection between the rectum, urinary, and/or genital tract (if present), and establish an anal opening within the sphincter complex, to maximize subsequent fecal continence.

Early approaches to surgical repair of ARM resulted in significant mortality or morbidity, with reduced quality of life.[1,2] Operative management was revolutionized by the advent of the posterior sagittal anorectoplasty (PSARP).[3] This approach enabled more complete visualization of the anorectal region, facilitating enhanced anatomic reconstruction.[3] Long-term data are increasingly available to assess functional outcomes in adulthood for patients managed in the PSARP era. Although available studies suggest greatly improved outcomes compared with those achieved following classical pull-through procedures, persistent bowel dysfunction affects a significant proportion of ARM patients into adulthood.[4,5]

The processes regulating defecation and fecal continence are multifactorial. They rely on the coordinated interplay of sensory and motor functions of the colon, rectum, and anus.[6–8] Abnormal anorectal function in the setting of an ARM has long been recognized as contributing to fecal incontinence.[9] Characteristic defects in the anal canal and sphincter complex may be intuitively associated with impaired continence, despite meticulous anatomic reconstruction.[10] The contribution of colonic motility to the functional outcomes of this cohort is less well defined.

Manometry may be used to investigate gastrointestinal motility. In the colon, alterations in motility are implicated in symptoms of bowel dysfunction.[11] Emerging insights from high-resolution techniques are contributing to our interpretation of normal colonic motility,[11] and may enhance our understanding of pathological variations in dysfunctional states.[11,12]

The objective of this systematic review was to summarize the methodology and outcomes of colonic manometry performed in children with a repaired ARM. We aimed to synthesize the existing literature, appraise the current understanding of colonic motility in this population, and identify directions for future inquiry.

Key Points

- Persistent bowel dysfunction affects a significant proportion of children with a repaired anorectal malformation. Manometry may be used to evaluate colonic motility in this cohort.
- Evidence regarding colonic motility patterns in this cohort is exceedingly limited. Although there is a suggestion that altered colonic motility may be implicated in the bowel dysfunction described, no commonly reported conclusions were identified.
- Standardized cohort and outcome reporting frameworks would enhance the utility of future work.

2 | METHODS

We conducted this systematic review according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) protocol (Table S2).[13] The methodology was published prospectively (PROSPERO registration: CRD42020177344). The search was conducted in MEDLINE, Embase, PubMed, and the Cochrane Library in March 2020 and repeated in July 2021. It was restricted to human studies published since January 1, 1985. The search strategy is summarized in Appendix S1.

2.1 | Study selection

Following removal of duplicate studies, article titles, and abstracts were independently evaluated for eligibility by two authors (H.E.B and M.Y.T.). Observational studies—including cohort, longitudinal, cross-sectional, and case-control studies—and clinical trials were included for review. Animal studies, conference abstracts, systematic reviews, and meta-analyses were excluded.

The inclusion criteria utilized for screening were as follows: post-operative colonic manometry performed in children following surgical correction of an ARM; pediatric cohort (aged 0–18 years); and English language. Studies reporting manometry outcomes in cohorts with multiple conditions (ARM and other diagnoses) were included, provided results from children with ARM were described separately in the final analysis.

2.2 | Data extraction

Data were extracted independently by two authors (H.E.B. and M.Y.T.). With respect to cohort characteristics, extracted data points included: patient age (at primary repair and at time of manometry); sex; reported ARM type; associated anomalies; operative repair type; and evaluation of post-operative bowel function, including assessment instruments and outcomes. Manometric characteristics evaluated included: type of manometry; protocol (including preparation and sedation); parameters assessed (including evaluation criteria); catheter type and sensor spacing; and reported motility outcomes, including correlation with post-operative bowel function.

2.3 | Quality assessment

Quality appraisal of non-randomized study methodology was performed according to the Newcastle-Ottawa Scale (NOS).[14] It is an eight-item scale, which applies criteria in three groups: selection, comparability, and exposure. Higher scores reflect greater methodological quality; highest quality studies evaluated using NOS may receive a total maximum of nine.

3 | RESULTS

3.1 | Search results

Following removal of duplicates, a total of 256 unique records were identified. Of these, four reported post-operative colonic manometry and were suitable for analysis. Study selection is described in Figure 1.

3.2 | Study characteristics

Four studies reported colonic manometry results in children with repaired ARM.[15–18] Colonic manometry was utilized to investigate the pathophysiology of post-operative bowel dysfunction[15,16,18] and evaluated as a tool to predict the outcome of subsequent surgical intervention (cecostomy placement).[17] Study characteristics are summarized in Table 1.

3.3 | Cohort characteristics

From the combined total cohort of 151 children, 35 (21 male, age range <2–18 years) patients with ARM underwent post-operative colonic manometry. The median manometry cohort size was 10 patients with ARM (range 2–13). Cohort characteristics are provided in Table 1

Anorectal malformation type was reported for 25 children in the combined colonic manometry cohort. Malformation types were variably classified as high (n = 9); low (n = 4); cloacal malformation (n = 1); and by fistula location as: anovestibular (n = 1), perineal (n = 4), rectourethral (n = 2), rectovaginal (n = 1), rectovesical (n = 1), and vestibular (n = 2). Associated anomalies were reported by two studies: five children had sacro-/spinal anomalies.[17,18] Neither reported estimated severity. Operative repair type was described by only one of four included studies.[18] Clinical characteristics are summarized in Table 1.

3.4 | Quality appraisal

Quality evaluation of non-randomized studies, performed in accordance with the NOS, identified poor quality of all eligible studies. Quality appraisals are presented in Table S1.

3.5 | Colonic manometry characteristics

3.5.1 | Equipment

Three studies performed colonic manometry using water-perfused catheters, with four to eight sensors spaced at 10–15 cm intervals.[15–17] Burjonrappa et al.[18] did not report the colonic manometry equipment utilized. Colonic manometry specifications are summarized in Table 2.

3.5.2 | Bowel preparation and sedation regimen

Three studies described the bowel preparation regimen utilized.[15–17] In two, colonic lavage was achieved using either sodium phosphate or polyethylene glycol,[15] or polyethylene glycol alone,[17] the day prior to the procedure. Heikenen et al.[16] described an overnight fast prior to colonoscopy; however, did not report utilizing any additional medications to facilitate bowel preparation.

With respect to sedation, three studies reported use of procedural sedation alone,[16] or either sedation or general anesthesia.[15,17] Burjonrappa et al.[18] did not describe the sedation protocol employed. Preparation and sedation regimens are summarized in Table 2.

3.5.3 | Catheter placement and securement

Two studies reported colonoscopic placement of the manometry catheter, using a guidewire technique.[15,16] Van den Berg et al.[17] similarly utilized colonoscopic placement for the majority of patients (30 children); however, fluoroscopic guidance was used in addition to endoscopy. In two patients, interventional radiology alone was used to achieve catheter placement.[17] Placement approach was not described by diagnostic sub-group.

No study described catheter securement. Van den Berg et al.[17] referenced a previously published protocol, which described external taping of the manometry catheter to the patient's thigh.[19] Notably, no study reported maneuvers to secure the manometry catheter tip intraluminally—such as tethering the catheter to the colonic mucosa—to minimize catheter migration. Catheter placement and securement techniques are summarized in Table 2.

3.6 | Protocols and motility parameters

The approach to colonic motility assessment was relatively conserved between studies; however, differences were identified with respect to the criteria used to define motility events, normal colonic motility, and features of dysmotility. Three studies reported colonic motility assessment during fasting, following ingestion of a meal, and after administration of Bisacodyl.[15–17] High-amplitude propagated contractions (HAPCs) were the most commonly assessed motility

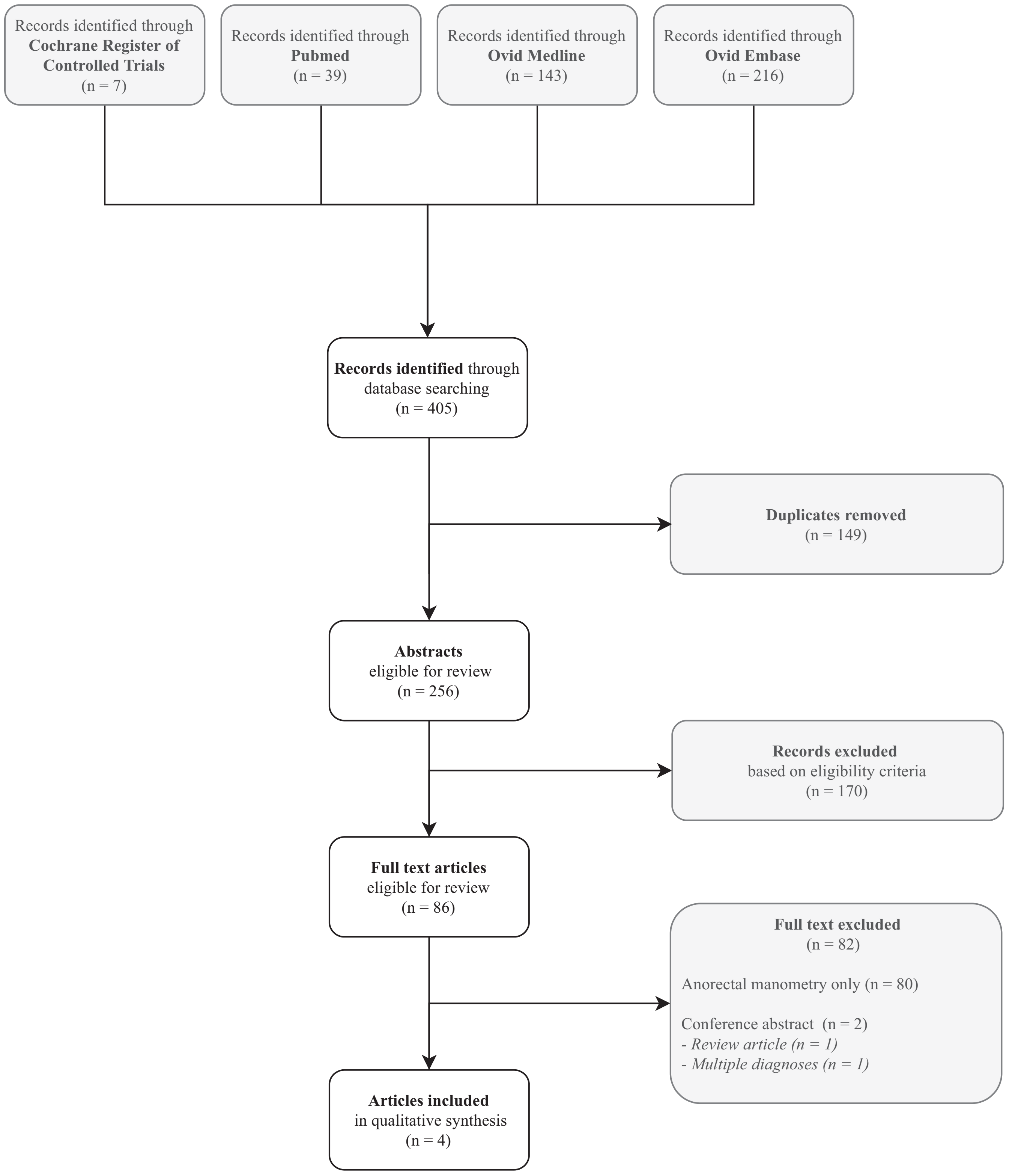

FIGURE 1 Flow diagram of study identification and selection

parameter; however, no two studies used the same criteria to define them. Only one study defined the evaluation of meal response utilized.[15] Burjonrappa et al.[18] did not describe their approach to assessment. Parameters, including motility assessment criteria, are summarized in Table 3.

3.6.1 | Features of normal and abnormal colonic motility

Study criteria relating to normal and abnormal motility are summarized in Table 4. Features of "normal" colonic motility were

TABLE 1 Clinical characteristics of included studies

First author	Year	Cohort (Total [Male])		Study population summary	Age at time of manometry	Reported anorectal malformation type	Associated anomalies	Operative repair type
		Total[a]	Manometry[b]					
Burjonrappa[18]	2010	86 (53) Developed megarectum: 14 (8)	2 (0)	Patients with megarectum after surgery for anorectal malformation	Not reported.	High: 1 (rectovaginal fistula) Low: 1 (anovestibular fistula)	**Sacral anomalies**: 13 / 86 Manometry cohort: 1 (sacral hemivertebrae)	Laparoscopic pull-through with anal transposition at three months, after initial dilatation: 1 Anal transposition at three months,[c] after initial dilatation: 1
Demirogullari[15]	2010	20 (11) Anorectal malformation: 10 (6)	*Group 1*[d]: 10 (6)	Children with repaired anorectal malformations compared with children with motility disorders.	Group 1 (anorectal malformations): mean 9.6 years (range 4–18) Group 2 (motility disorders): mean 12.1 (range 6–19)	Fistula: Rectovesical: 1/10 Rectourethral: 2/10 Vestibular: 2/10 Perineal: 4/10		
Heikenen[16]	1999	13 (6)	13 (6) ARM: 10 (4)	Children with fecal incontinence refractory to standard medical therapy post-repair of anorectal malformation	Mean 8.6 years (range 5–13)	High: 9/13 Low: 4/13		
van den Berg[17]	2006	Anorectal malformation: 10 (9)	10 (9)	Children undergoing colonic manometry prior to cecostomy placement for severe defecation disorders, including children with repaired anorectal malformations	Not reported. Mean age at cecostomy placement: 6 years (2–11) *Occurred median 2 months post-manometry (range 0–15 months).*	-	Spinal abnormalities: 4	

Abbreviations: –, not reported; ARM, anorectal malformation; CM, colonic manometry.

[a]Total cohort with anorectal malformation.

[b]Refers to cohort of children undergoing *post-operative* colonic manometry following anorectal malformation repair.

[c]Complicated by rectovaginal fistula requiring colostomy formation and subsequent vaginoplasty.

[d]Group 1: children with anorectal malformations

TABLE 2 Colonic manometry equipment and protocols reported by included studies

First author	Year	Catheter Type	Sensor spacing (cm)	Sensors (no.)	Preparation	Sedation/anesthetic	Catheter placement	Catheter tip location	Catheter securement
Burjonrappa[18]	2010	-	-	-	-	-	-	-	-
Demirogullari[15]	2010	WP	10–15 cm	8	Oral sodium phosphate (Fleet) or polyethylene glycol, completed day prior.	Sedation or general anesthesia (not further specified).	Colonoscopy, using a guidewire technique.	Caecum: 4 Transverse colon: 1 Hepatic flexure: 3 Splenic flexure: 2	Not reported.
Heikenen[16]	1999	WP	10–15 cm	4–8	Overnight fast.	Procedural sedation (not further specified).	Colonoscopy, using a guidewire technique.	Proximal catheter port positioned in the right colon, with the distal port in the neorectum. Verified radiologically.	Not reported.
van den Berg[17]	2006	WP	10–15 cm	8	Polyethylene glycol colonic lavage via nasogastric tube, completed day prior.	Sedation or general anesthesia (not further specified).	Colonoscopy with fluoroscopic guidance (n = 30) Interventional radiology alone (n = 2)	Proximal (caecum to splenic flexure): 26 Distal (descending to sigmoid colon)[a]: 6	Not reported. Methods reported to comply with a published protocol, which describes taping the manometry catheter to the thigh.[19]

Abbreviations: (–), not reported; WP: water-perfused.

[a]Technical difficulties prevented placement of the manometry catheter in the "proximal colon" (caecum to splenic flexure) in six of 32 patients; not described by diagnostic sub-group.

TABLE 3 Colonic manometry parameters: reported definitions and criteria

First author	Parameters assessed	HAPC criteria	Assessment of meal response	Pharmacologic stimulation	Definitions of other included parameters
Burjonrappa[18]	-	-	-	-	Hyperperistalsis: not further defined.
Demirogullari[15]	Propagated contractions, HAPCs, LAPCs. Motility recorded: -Fasting: 60 minutes -Post-meal: 180 minutes -Post-Bisacodyl: 40 minutes	Propagated contractions with an amplitude of at least 60 mmHg.	Gastrocolonic response. Propagated contractions after the meal were considered a normal gastrocolonic response.	Bisacodyl 0.2 mg/kg	Propagated contractions: pressure waves migrating over 3 or more adjacent recording channels at a velocity of 0.2 – 12 cm/s. LAPCs: propagated contractions with an amplitude less than 60 mmHg.
Heikenen[16]	HAPCs (including % propagating to the neorectum). Motility recorded: -Fasting: 30–120 minutes -Post-meal[a]: 60–120 minutes -Post-Bisacodyl: 20–60 minutes	Contractions: - with an amplitude of at least 80 mmHg - lasting at least 10s - propagating at least 30 cm	-	Bisacodyl 0.2–0.4 mg/kg[b]	-
van den Berg[17]	HAPCs. Motility recorded:-Fasting: 60 min -Post-meal: 60 min -Post-Bisacodyl: 40 min or until the catheter was expelled	Contractions: - with an amplitude of at least 60 mmHg - lasting at least 10s - propagating at least 30 cm	-	Bisacodyl 0.2 mg/kg	Colonic segments subdivided into proximal (cecum to splenic flexure) and distal (descending and sigmoid colon); patterns of colonic contractions determined by qualitative analysis

Abbreviations: (–), not reported; HAC, high-amplitude contraction; HAPC, high-amplitude propagated contraction; LAPC, low-amplitude propagated contraction; WP, water-perfused.

[a]Age-appropriate meal": not further specified with respect to caloric or macronutrient content.

[b]Bisacodyl administered into the right colon only if no HAPCs were identified during fasting or postprandial periods.

TABLE 4 Colonic motility criteria and key findings of included studies

First author	Criteria: normal motility classifications	Criteria: abnormal motility classifications	Key findings	Limitations
Burjonrappa[18]	Not specified.	Hyperperistalsis: not further defined	• Hyperperistalsis reported in both patients with megarectum who underwent colonic manometry.	• No description of colonic manometry assessment (equipment, protocol, parameters) or interpretation. • Limited reporting of colonic manometry outcomes: description of results limited to "hyperperistalsis", which is not further defined.
Demirogullari[15]	Not specified.	Abnormal motility: absence of propagated contractions during all phases of the study or a segmental absence of propagated contractions.	• Colonic response to intraluminal Bisacodyl stimulation was stronger in children with anorectal malformation than children with severe colonic dysmotility. • HAPCs identified in 60% and PCs identified in all children with repaired anorectal malformation. • PCs found to reach rectosigmoid region in only 4/10 patients.	• Symptom characterization not provided. • Emphasis on propagated contractions rather than HAPCs may limit comparability of findings.
Heikenen[16]	HAPCs are a feature of normal colonic motility; normally only 14% of HAPCs propagate to the rectum. Normal motility not further specified.	Not defined.	• All patients demonstrated HAPCs; 81% of HAPCs propagated to the neorectum. • FI multifactorial, including propagation of excessive numbers of HAPCs into the neorectum, IAS dysfunction, and inability of neorectum to sufficiently accommodate stool.	• Limited by manometry techniques available. • Limited reporting of colonic manometry outcomes. • Symptom characterization not provided (pre- or post-treatment).
van den Berg[17]	Normal motility: HAPCs occurring from proximal to distal colon post-meal and/or post-Bisacodyl administration.	Proximal: HAPCs in the proximal colon only Distal: HAPCs in the distal colon only Total: absence of HAPCs in the entire colon -Persistently low-amplitude contractions: <60 mmHg -Colonic inertia: complete absence of contractions Simultaneous contractions: not further defined	• Anorectal malformation sub-group. • Normal motility[a]: 4/9; proximal HAPCs only[a]: 1/9; total absence of HAPCs[a]: 3/9; absence of HAPCs in distal colon[b]: 4/7; Bisacodyl response: 6/10. • Colonic manometry helpful in predicting cecostomy response: HAPCs after Bisacodyl administration predictive of success, including those with proximal HAPCs only; absence of HAPCs in the entire colon significantly associated with poor response.	• Small anorectal malformation sub-group (n = 10) with associated spinal anomalies in 40%. • Limited demographic data or medical characteristics provided. • Limited by manometry techniques available.

Abbreviations: FI, fecal incontinence; HAPC, high-amplitude propagated contraction; IAS, internal anal sphincter; PC, propagated contraction.

[a]Excluding those with missing proximal colonic data.

[b]Including those with missing proximal colon data; excluding patients with total colonic absence of HAPCs.

reported by two studies.[16,17] Although both referred to the presence of HAPCs as necessary for a motility pattern to be considered normal, van den Berg et al. also required the observed HAPCs to propagate from the proximal to distal colon.[17] Heikenen et al.[16] alternatively proposed, "normally, only 14% of HAPCs propagate through to the rectum," but did not specify a required length, duration or anatomical site of propagation. Demirogullari et al.[15] did not provide the definition of normal colonic motility applied; however, the presence of propagated contractions (either HAPCs or low-amplitude propagated contractions [LAPCs]) following meal ingestion was considered indicative of a normal gastrocolonic response.

With respect to "abnormal" motility patterns, both Demirogullari et al. and van den Berg et al. considered the absence of propagated contractions to be abnormal.[15,17] Van den Berg et al.[17] further subclassified patients as demonstrating either, "persistently low amplitude contractions" (<60 mmHg) or "colonic inertia" (defined as the complete absence of contractions). In addition, the segmental absence of HAPCs was considered to reflect abnormal motility. This pattern was designated according to anatomical location (of the observed HAPCs) as either proximal or distal.[17]

Burjonrappa et al.[18] did not report features of normal or abnormal motility. Both patients in that study were reported to demonstrate "hyperperistalsis." Although this pattern was not further defined, it was inferred to represent an abnormal motility pattern.

3.7 | Colonic manometry outcomes

Key manometry findings are summarized in Table 4. Heikenen et al.[16] assessed the role of colonic motility in the pathophysiology of post-operative fecal incontinence. All children with repaired ARM demonstrated HAPCs, 81% of which propagated to the neorectum (normal defined as 14% of HAPCs reaching the rectum). The increased proportion of HAPCs propagating to the neorectum was suggested to contribute to fecal incontinence in these children.

Demirogullari et al.[15] compared colonic motor activity between children with repaired ARM and those with severe colonic dysmotility. Despite a lower amplitude criterion for HAPCs than Heikenen et al.[16] (60 mmHg vs. 80 mmHg), HAPCs were identified in only 60% of children with repaired ARM. Similarly, propagated contractions (encompassing both LAPCs and HAPCs) reaching the neorectum were identified in a smaller proportion of their cohort (40%).[15] More children with severe colonic dysmotility demonstrated a meal response. However, a greater proportion of children with repaired ARM demonstrated HAPCs following intraluminal Bisacodyl stimulation (70% vs. 40%; $p = 0.019$). These differences were suggested to indicate that a different etiology may underlie the bowel dysfunction symptoms experienced by children with repaired ARM compared with those with colonic dysmotility.

Van den Berg et al.[17] identified HAPCs following Bisacodyl administration in a comparable 60% of children with a repaired ARM. Bisacodyl response was found to be predictive of a successful outcome following cecostomy formation, irrespective of whether the HAPCs propagated throughout the colon or were confined to the proximal segment (cecum to splenic flexure). Notably, four of nine children (with proximal colonic data) demonstrated "normal" colonic motility. Total colonic absence of HAPCs was associated with poor cecostomy response.

TABLE 5 Post-operative bowel function assessment instruments and reported outcomes

First author	Year	Assessment	Post-operative bowel function
Burjonrappa[18]	2010	Modified Wingfield Score (MWS) for fecal continence: normal function = 0, constipation/FI = 1, intermittent FI = 2, continuous FI = 3.	Megarectum cohort prior to excision: MWS 3 Post-op: low malformations: normal (0) 6/8; (1) 1/8; 2 1/8; high malformations: normal (0) 4/6; (1) 2/6. Post-operative function of colonic manometry cohort not specifically defined.
Demirogullari[15]	2010	Symptom criteria and assessment not reported.	Voluntary bowel movement without soiling: 2/10; without soiling for 24 hours: 4/10; awaiting operative intervention: 4/10.
Heikenen[16]	1999	Symptom criteria and assessment not reported.	All had FI refractory to medical therapy; however, bowel function not further defined.
van den Berg[17]	2006	Parent-reported bowel function assessed using a questionnaire. Bowel movement frequency: <5/ week; 5/week – 3/day; >3/day. Fecal incontinence: none, occasional, few episodes per week, few episodes per week to daily, constant. Successful cecostomy outcome: normal frequency (5/week – 3/day) and less than occasional FI.	ARM sub-group (*n* [%]). Bowel movement frequency <5/week: 6 (60%) Fecal incontinence occurring more than occasionally: 9 (90%).

Abbreviations: FI, fecal incontinence; MWS, Modified Wingfield Score.

3.8 | Post-operative outcome assessment

Instruments and outcomes of post-operative bowel function assessment are summarized in Table 5. Burjonrappa et al.[18] was the only study to utilize an established measure of bowel function (Modified Wingfield Score to assess fecal incontinence). From a pre-operative cohort score of three (continuous fecal incontinence), there was a trend toward improved function post-operatively: 75% and 67% of the low and high malformation groups, respectively, reported a score of zero ("normal" function). Van den Berg et al.[17] utilized a study-specific questionnaire, which included parent-reported assessment of bowel movement and fecal incontinence frequency. Approach to bowel function assessment was not reported by either of the other two studies.[15,16]

4 | DISCUSSION

Anorectal malformations represent a complex pathological spectrum. Despite surgical correction, a significant proportion of affected children will experience post-operative constipation and/or fecal incontinence. While characteristic defects in the anal canal and sphincter mechanism are well recognized as contributing to the bowel dysfunction experienced, the impact of colonic motility in this cohort is less well understood. To establish if colonic motor abnormalities also exist in these patients, there have been several papers published, which detail the results of colonic manometry studies performed in this cohort. To our knowledge, this is the first review to systematically evaluate these.

What became immediately apparent was that there were very few studies reporting such findings to include in this review. Only four studies met the inclusion criteria, indicating that there is a lack of data evaluating colonic function in children following repair of ARM. Furthermore, due to a lack of protocol standardization and technical constraints, the clinical relevance of these studies was difficult to determine. Consequently, knowledge of colonic motility in this cohort remains exceedingly limited.

Children clearly benefit from surgical repair of ARM. However, persistent bowel dysfunction continues to impact a significant proportion of patients.[4,5] Bowel continence and defecation involve coordination of motor patterns in the colon, rectum, and anus. The perseverance of symptoms after anorectal repair in these children may indicate coexistent abnormalities of colonic motility. That so few studies have even attempted to evaluate colonic motor patterns in these children suggests that either the colon is not considered part of the potential pathophysiology, or that the existing studies give no clear indication that a problem exists. Based upon this review, the latter could certainly be the case. Even though only four studies were identified for review, each utilized different equipment and protocols. These differences will be discussed below.

4.1 | Approach to colonic manometry: equipment, protocols, and assessment parameters

Three of four studies utilized low-resolution, water-perfused manometry catheters, with sensors spaced at 10–15 cm intervals.[15–17] Low-resolution manometry provides a very incomplete picture of colonic motility, with many motor patterns either missed or misinterpreted.[20] These findings prompted a consensus recommendation for the mandatory use of high-resolution manometry in the colon; where possible, a catheter inter-sensor interval of no more than 2 cm is now recommended.[20,21] The low-resolution catheters utilized by studies included in this review likely reflect best-practice technology available at the time. However, application of high-resolution techniques will offer greater insights into the colonic motility of this cohort.

Procedural limitations were also evident, such as the approach to catheter tip securement to minimize intraluminal migration. This issue was alluded to by van den Berg et al., who recorded for 40 min post-Bisacodyl or "until the catheter was expelled," presumably due to its propulsion through the colon secondary to peristalsis.[17] Notably, this study emphasized segmental assessment of colonic motility (including proximal, distal, and total colonic absence of HAPCs). It is unclear how potential catheter migration was accounted for in the determination of anatomic location of HAPCs reported in the analysis. No included study described the approach to catheter securement. One well-established practice is to secure the catheter tip to the colonic mucosa using an endoclip, which may minimize potential movement and/or expulsion during assessment.[22,23]

4.2 | Manometry outcomes

4.2.1 | Manometry outcomes in relation to surgical characteristics

Post-operative continence may be affected by a variety of factors, including ARM type, associated sacral and/or spinal anomalies, and operative repair (including complications). For 10 of the 35 children in this review, their ARM type was not documented; the presence of spinal anomalies not reported in 23; and the primary operative repair unknown for the majority (33 children, 95%). Accurate understanding of ARM type is essential not only to facilitate precise operative repair, but also to prognosticate continence potential.[7,10] The ARM Index may be used to predict continence in patients with an ARM. It incorporates assessment of the quality of the spine and sacrum, in addition to ARM type.[24] More complex ARM (such as cloacal exstrophy and rectobladder neck fistula) are considered to present the greatest challenge to attaining fecal continence.[24] Conversely, children with ARM with a good prognosis for fecal continence are at greater risk for developing severe constipation.[6] At a minimum, studies with these children should incorporate assessment of these principal determinants of continence.

Similarly, the approach to surgical repair may have important implications for subsequent bowel function. This is reflected in the improved continence reported following widespread adoption of the PSARP for primary repair.[4,5] Likewise, post-operative complications and the need for redo procedures may be associated with poorer outcomes[10,25]; although re-operation may achieve anatomic reconstruction, it may be unable to restore the continence potential the patient held at birth.[26] The exceedingly limited detail provided regarding primary repair type, complications, and re-operation is a major limitation of the existing literature.

4.2.2 | Colonic manometry outcomes in relation to bowel function

Fecal incontinence following ARM repair is commonly considered to result from either severe constipation with overflow soiling, or "true" incontinence, when the patient's anatomic features are considered insufficient to support fecal control.[7,24] Heikenen et al.[16] focused on colonic motility in children with fecal incontinence refractory to medical management following ARM repair. High-amplitude propagated contractions were identified in all subjects, 81% of which propagated to the neorectum. While no control group was used, the authors reported that the proportion of HAPCs propagating to the neorectum was greater than the proposed norm of 14%, and this increase may have contributed to fecal incontinence in these children.[16] The two patients assessed by Burjonrappa et al. had a pre-intervention Modified Wingfield Score of three, similarly reflective of continuous fecal incontinence.[18] Both children were reported to demonstrate an undefined "hyperperistalsis." The lack of definition makes the motility patterns in these children difficult to compare to data presented by Heikenen et al.[16]

In contrast to the study by Heikenen et al., Demirogullari et al. applied less restrictive criteria for both HAPCs and assessment of motility events reaching the neorectum (HAPCs vs. HAPCs and LAPCs).[15] Despite this, only six of 10 children assessed by Demirogullari et al. demonstrated HAPCs, and PCs (encompassing HAPCs and LAPCs) reached the rectosigmoid region in only four. It is likely that these differences reflect the heterogeneity of the patient populations assessed by each study. Heikenen et al.[16] included only children with refractory fecal incontinence; however, the symptom profile of children assessed by Demirogullari et al. was not reported, preventing comparison.[15]

In children with Hirschsprung disease, HAPCs propagating to the neorectum have been associated with fecal incontinence.[27] Additionally, a greater number of HAPCs—potentially resulting from colonic transection during surgical repair[28]—has been identified in those with multiple episodes of soiling daily (termed "colonic hyperactivity").[29] The number of HAPCs was quantified by only one study; bowel function, including presence and frequency of incontinence episodes, was not reported.[15] Whether a greater total number of HAPCs overall, in addition to a greater proportion of HAPCs propagating to the neorectum, contributed to fecal incontinence cannot be ascertained.

Of note, van den Berg et al. identified post-Bisacodyl HAPCs in 60% of subjects with severe defecation disorders evaluated prior to cecostomy formation.[17] This finding was comparable to that of Demirogullari et al.[15] Approximately 90% of the children assessed by van den Berg et al. experienced "more than occasional" fecal incontinence (few episodes per week—constant incontinence); however, this was associated with reduced bowel movement frequency in 60%, suggesting a degree of overflow incontinence. These discrepancies highlight the importance of robust symptom assessment to facilitate identification of manometry-symptoms correlates: the fecal incontinence experienced by children assessed by van den Berg et al. may represent a different entity to those evaluated by Heikenen et al.[16,17]

It is clear from the papers included in this review that HAPCs were the main motor patterns identified and discussed. While these motor patterns play an important role in normal defecation, colonic motility may also play a role in helping to maintain bowel continence. High-resolution manometry studies have identified a predominantly retrograde propagating cyclic motor pattern, originating at the rectosigmoid junction. It has been proposed that this motor pattern acts as a rectosigmoid brake, controlling rectal filling.[30–33] The presence or absence of cyclic motor activity in the distal colon of the children included in this review was not mentioned. In part, this was because it has not routinely formed part of colonic manometry studies. In addition, the wide sensor spacing of the catheters used in these studies would have been unable to detect the propagating nature of the cyclic motor pattern.[20] However, given fecal incontinence plays a major role following ARM repair, it is a manometric correlate that should be included with the introduction of high-resolution manometry studies.

The variable approaches used by these studies to define motility events are reflective of the broader paucity of data guiding our understanding of "normal" colonic manometry. There are no colonic manometry data from healthy pediatric cohorts; with the invasive nature of current manometry techniques, this is unlikely to change.[12] Furthermore, while an attempt has been made to form a consensus of both terminology and understanding of what motor patterns form part of "normal" motility,[11] there remains a lack of knowledge as to true normal ranges, in either children or adults. As demonstrated in this review, definitions of "normal" have typically been developed independently by each laboratory. Emerging insights from adult high-resolution colonic manometry have offered a more nuanced understanding of colonic motility and highlighted the limitations of simplistic criteria currently used in children.[11] Utilization of high-resolution techniques in pediatric cohorts, in combination with an increasingly detailed approach to analysis,[34–36] may lead to a similar evolution in our understanding of childhood motility.

Finally, underpinning any meaningful assessment of manometry-symptom correlates is the accurate, replicable, and comparable evaluation of bowel function. Only one study utilized a previously established measure of bowel function,[18] while the remainder either developed a study-specific questionnaire[17] or did not report their approach to assessment.[15,16] For example, all patients assessed by Heikenen et al. had "fecal incontinence refractory to medical therapy".[16] Demirogullari et al.[15] evaluated patients prior to surgical interventions, presumably instituted to manage bowel dysfunction (antegrade continence enema procedure, sigmoid resection). However, as symptom criteria and severity were not reported, opportunities to elicit useful comparisons between cohorts were limited.

4.3 | Limitations

Our study was limited to children aged less than 18 years. Although this enables assessment of exclusively pediatric data, results from mixed pediatric and adult populations may have been excluded, if the results for participants aged less than 18 years were not described separately. Similarly, children with ARM included in cohorts with mixed diagnoses may have been excluded, if the results were not reported separately. Our search was restricted to English language articles; findings may be subject to language bias.

5 | CONCLUSION

This review highlights how few studies have assessed colonic function in children with ARM. Clearly, if colonic motility disorders exist in this cohort, very few groups are trying to define them. Of the four studies included in the review, there were no commonly reported conclusions on which to base our understanding of post-operative colonic motility in children with previous repair of an ARM. In addition, there are no clear data detailing how colonic manometry findings may relate to ARM subtypes, operative intervention, and clinical assessments of bowel function. Although there is a suggestion that altered colonic motility may be implicated in the post-operative bowel dysfunction experienced by this cohort—particularly in the setting of fecal incontinence—studies are limited by heterogeneous cohorts, insufficient reporting of key operative characteristics, and variable descriptions of motility findings. Standardized cohort and outcome reporting frameworks would enhance the utility of future research. The advent of high-resolution colonic manometry offers the promise of enhanced understanding of colonic motility in health and disease; however, a concerted effort is required to ensure future work avoids the pitfalls prevalent throughout much of the existing literature.

AUTHOR CONTRIBUTIONS

Dr Evans-Barns and Ms Tien performed the literature search, article screening, and data extraction. Dr Evans-Barns drafted the initial manuscript. Dr Trajanovska, Dr Safe, A/Prof. Dinning, and Prof. Hutson critically reviewed and revised the manuscript for intellectual content. A/Prof. King conceptualized and supervised the execution of the systematic review. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

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CONFLICT OF INTEREST

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DATA AVAILABILITY STATEMENT

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

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SUPPORTING INFORMATION

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