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Review Article

Classification and Surgical Management of Anorectal Malformations: A Systematic Review and Evidence-based Guideline From the APSA Outcomes and Evidence-based Practice Committee

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ABSTRACT

Objective: Treatment of neonates with anorectal malformations (ARMs) can be challenging due to variability in anatomic definitions, multiple approaches to surgical management, and heterogeneity of reported outcomes. The purpose of this systematic review is to summarize existing evidence, identify treatment controversies, and provide guidelines for perioperative care.

Methods: The American Pediatric Surgical Association Outcomes and Evidence Based Practice Committee (OEBP) drafted five consensus-based questions regarding management of children with ARMs. These questions were related to categorization of ARMs and optimal methods and timing of surgical management. A comprehensive search strategy was performed, and the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines were used to perform the systematic review to attempt to answer five questions related to surgical care of ARM.

Abbreviations: ARM, (anorectal malformation); VACTERL, (vertebral, anorectal, cardiac, tracheoesophageal fistula, renal, limb).

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Results: A total of 10,843 publications were reviewed, of which 90 were included in final recommendations, and some publications addressed more than one question (question: 1 n = 6, 2 n = 63, n = 15, 4 n = 44). Studies contained largely heterogenous groups of ARMs, making direct comparison for each subtype challenging and therefore, no specific recommendation for optimal surgical approach based on outcomes can be made. Both loop and divided colostomy may be acceptable methods of fecal diversion for patients with a diagnosis of anorectal malformation, however, loop colostomies have higher rates of prolapse in the literature reviewed. In terms of timing of repair, there did not appear to be significant differences in outcomes between early and late repair groups. Clear and uniform definitions are needed in order to ensure similar populations of patients are compared moving forward. Recommendations are provided based primarily on A-D levels of evidence.

Conclusions: Evidence-based best practices for ARMs are lacking for many aspects of care. Multi-institutional registries have made progress to address some of these gaps. Further prospective and comparative studies are needed to improve care and provide consensus guidelines for this complex patient population.

Level of Evidence: 3.

1. Introduction

Anorectal malformations (ARM) historically were viewed as an isolated surgical issue. Evidence over time has accumulated to reveal the true complexity of long-term medical management required to support these patients appropriately through childhood. The challenges of treating patients with these malformations have many considerations [1,2]. There are a wide range of anatomical subtypes of ARM, and their definitions have changed over time [1–5]. These patients have been shown to experience quality of life issues, even after appropriate initial surgical intervention [1,2]. Other facets of care have also evolved over time, including favored surgical approach [1–12]. Finally, many patients have involvement of other organ systems outside of the hindgut, even without a diagnosis of VACTERL association, adding additional complexity to their care.

The purpose of this systematic review from the American Pediatric Surgical Association (APSA) Outcomes and Evidence Based Practice (OEBP) Committee is to review the available literature surrounding the initial management of patients with ARM related to subtype definition, optimal surgical intervention approach, overall functional outcomes for these patients, and to provide recommendations based on the current evidence available.

2. Methods

2.1. Research questions

The membership of the APSA OEBP Committee formulated the following consensus-based questions a priori for this systematic review. This review was not registered and the standard review protocol used by the APSA Outcomes and Evidence Based Practice Committee was used. Five questions were developed, the first four of which will be reviewed in this manuscript:

1. What are the various types of ARM? How are they defined?
2. What is the appropriate initial surgical reconstruction for a neonate diagnosed with an ARM by subtype?
3. In patients for whom fecal diversion is indicated, what is the preferred type of colostomy that should be created? Where in the colon is the optimal location for colostomy?
4. What is the optimal timing of definitive repair in patients who are initially managed with fecal diversion? What is the optimal timing of definitive repair in patients who are initially managed with dilations?
5. What is the spectrum of reported outcome measures and do long-term outcomes differ by level of malformation, status of the sacrum and spine, and/or surgical timing? (Continence/soiling vs. constipation, sexual function, HRQOL/psychosocial measures)? What is the appropriate method for transitioning patients to adult care?

Question 5 will be reviewed in a separate subsequent publication.

2.2. Search strategy

A health sciences librarian developed a search strategy and conducted a comprehensive literature search using Medical Subject Headings (MeSH) through a combination of controlled vocabulary and keywords. The following databases and associated search dates were used for this review: Ovid Medline (1985 to July 19, 2021), EMBASE (1985 to July 19, 2021), Cochrane Database of Systematic Reviews (2005 to July 19, 2021, and Cochrane Controlled Trials (1997 to July 19, 2021). Due to the overall increased complexity and differential long-term care needs of patients with cloaca, this diagnosis was excluded from this systematic review.

2.3. Study selection

Our working group followed Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines during study screening and selection. We used a reference management software package (EndNote, Clarivate Analytics, New York, NY, USA) to generate a list of abstracts for review [13].

Animal studies, duplicates, non-English articles, studies in adult populations, case reports, and articles published prior to 1985 were excluded. Each article was screened via independent abstract review by at least two of the study authors using Rayyan (https://www.rayyan.ai) (CS, RR, AK, KR). We resolved conflicts via a second review by all authors to reach consensus. Studies included for full manuscript review were allocated to each appropriate research question. The level of evidence for each included study was assigned per the Oxford Centre for Evidence-Based Medicine (OCEBM) guidelines. Methodological index for non-randomized studies (MINORS) criteria were used to assess the methodological and reporting quality and risk of bias of appropriate articles.

3. Results

3.1. Search strategy

The search strategy revealed 1392 non-duplicated titles that met our criteria. Of these, 463 abstracts were assessed as possibly

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relevant and the full text was obtained for review. Three hundred-eleven records were excluded after full-text review. These were most commonly excluded due to the manuscript addressing the wrong outcome measure or evaluating the wrong population. There were 152 manuscripts included in the qualitative review and assigned to one or more of the pre-defined questions, 90 manuscripts were reviewed to attempt to answer the first four questions (Fig. 1). Twenty-two of these manuscripts evaluated more than one question. The remaining manuscripts will be reviewed in a subsequent manuscript reviewing the data available for question 5.

Question 1. What are the various types of anorectal malformations? How are they defined?

The definitions for specific subtypes of ARM are varied in the literature and have changed over time. Initial descriptions of low and high groups of ARMs were described by Stephens based on his work defining the pubococcygeal line [14–16]. This initial classification of these anomalies was expanded in Wingspread, Wisconsin in 1984 [1]. While the Wingspread low, intermediate, and high classification is often still referred to in the literature, more anatomic based definitions have been used since the Peña classification was created in 1995 [2,5]. The Peña classification is based on the fistula location and divided by anatomic and phenotypic sex. Subsequently in 2005, the International Conference for the Development of Standards for the Treatment of Anorectal Malformations was organized at Krickenbeck Castle, Germany [4]. This workshop brought together 26 international authorities on congenital malformations of the pelvis and perineum where Peña's classification was modified, with rare variants included [4]. The Krickenbeck classification primarily identified major clinical groups and variants of anorectal malformations, with the major goal of the meeting being the development of a system that would be comparable for follow-up studies. Outside of these publications, there has been relatively sparse literature related to how the specific ARMs are defined.

All articles were reviewed to evaluate for common nomenclature. The 152 articles were reviewed and categorized by the terminology or definition schema used to describe their patient population. There were 105 (69%) papers that used some type of anatomic language to describe their cohort of anorectal malformations, either in combination with Wingspread and Krickenbeck classification or as isolated anatomic language. The anorectal malformation subtype was identified by the location of the fistula in the majority of these publications. Thirty-two (21%) manuscripts used Wingspread classification with no other descriptive anatomic language. The papers using Wingspread classification were published between the years 1985–2021, with this terminology being

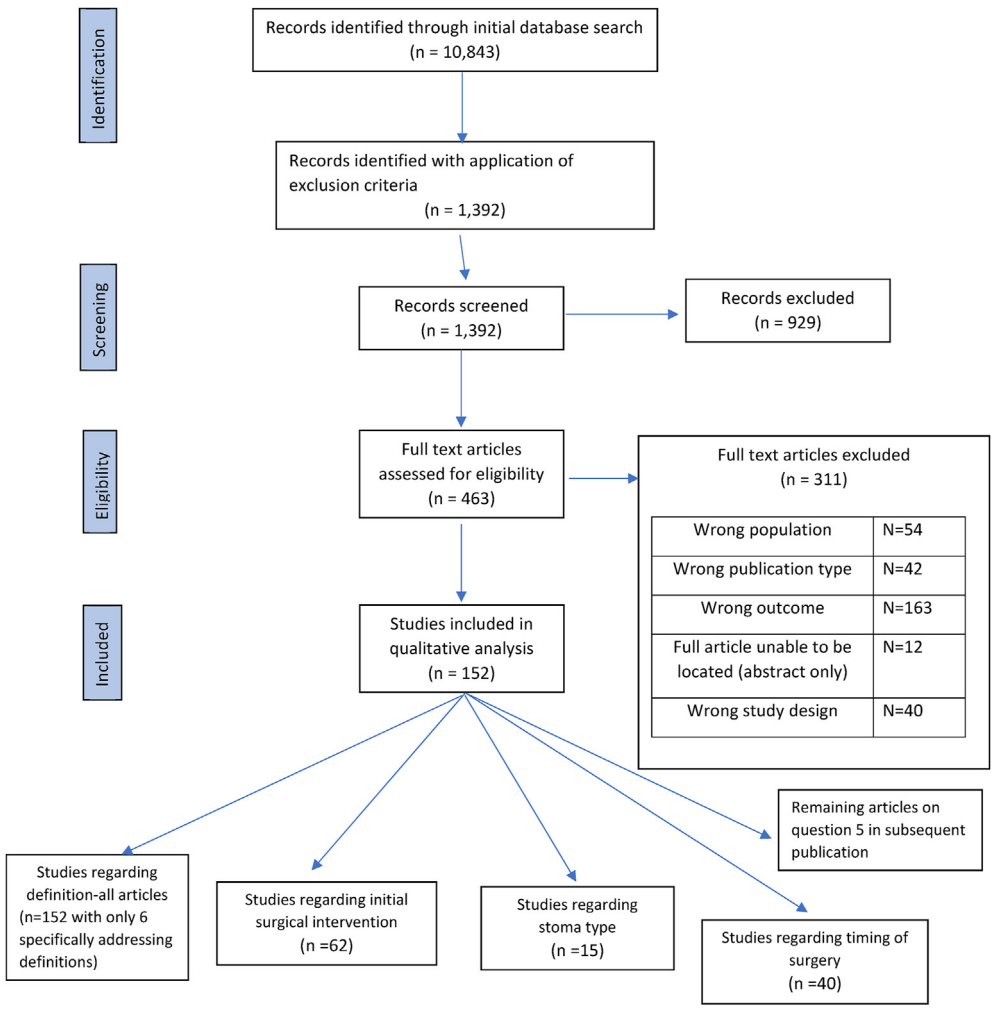

Fig. 1. Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) methodology. 10,843 records were identified with the initial database search. 1392 records remained following application of exclusion criteria. 463 full-text articles were assessed for inclusion and 152 articles were included in the qualitative analysis. All 152 manuscripts were reviewed to provide background for question 1 regarding classification. 101 of these manuscripts were reviewed in this manuscript reflecting the evidence available for questions 2–4. The remaining articles will be reviewed in the subsequent article reviewing question 5. Some articles addressed more than one of the pre-identified questions.

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used more frequently and in isolation in earlier time points in the period of literature evaluated. Twelve (8%) publications specifically noted the Krickenbeck classification for anorectal malformations with the majority additionally characterizing the patients with an anatomic description. Fifteen papers (10 %) did not specify the type of anorectal malformations represented by the patients included in their analysis. Most of these papers with no anatomic specification evaluated older age patient groups or quality of life, and the specific anorectal malformation diagnosis was not defined (Fig. 2).

Outside of the three landmark manuscripts that classified anorectal malformations in the time period of reviewed manuscripts, only a few publications addressed the specific definitions of various anorectal malformations in their methodology further. Most of these papers described rare variants, rather than the more common subtypes. A single paper used specific language to describe the anatomic anatomy of perineal fistulas and vestibular fistulas in the manuscript [3]. In terms of more common subtypes, Halleran et al. described a method for classification of the specific type of rectourethral fistulas in male patients using a distal colostogram [11]. For these malformations, in addition to defining each anatomic subtype, they suggested that the relationship of the distal rectum to the pubococcygeal line (above or below) is important both for classification and management. This specific anatomic description of the fistula, if used uniformly, was suggested to allow for clearer more defined subgoups of ARMs as well as allow for benchmarking of outcomes across centers and more specific study of each malformation and surgical approach.

4. Recommendations and observations

To appreciate the impact of various types of anorectal malformations on functional and surgical outcomes better, anatomic nomenclature for fistula location and classification should be used. Our recommendation includes documenting the specific anatomic fistula location in the operative report to classify the anorectal malformation and improve the ability to compare outcomes for specific subgroups. While classifications into different groups for ease of assessment has been proposed with updates over the years by experts, the language currently in the literature is varied and challenging to compare. While outside the search interval for this manuscript, recently, the Pediatric Colorectal and Pelvic Learning Consortium (PCPLC) published standard definitions for each category of anorectal malformation [12]. Based on the prior expert nomenclature developed, studies described above, and the recent consensus publication by the PCPLC, we make the following recommendation to adopt the standard nomenclature in Table 1.

- MINORS criteria scores for the reviewed manuscripts ranged from 4 to 18.
- Level of Evidence 5; grade of Recommendation D and E.

Question 2. What is the appropriate initial surgical reconstruction for a neonate diagnosed with an anorectal malformation by subtype?

The question of most appropriate initial surgical intervention by ARM subtype remains difficult to answer. The most common options described include primary neonatal repair of the ARM, dilation with delayed surgical repair of the ARM, or initial diverting colostomy followed by ARM repair. The type of initial management and selection of operative approach is typically based on multiple patient and anatomical factors, as well as surgeon preference. Additionally, multiple operations have been described for the management of ARM including the posterior sagittal anorectoplasty (PSARP), anterior sagittal anorectoplasty (ASARP), cutback operation, V–Y plasty, anal transposition, internal sphincter sparing anoplasty (ISSA), and laparoscopic assisted anorectoplasty (LAARP). Definitions of operations performed for repair of ARM are summarized Table 2. Due to the limited data available, for this review we focused on the literature available for types of surgical approach related subtype of ARM. Sixty-three studies were identified that included a description of anorectal malformation type in male and female patients and also detailed the operative technique [6–10] [17–74]. Fifty-nine studies were primarily retrospective cohort studies; three were prospective cohort studies; and one was a randomized controlled study [6,8–10] [17–74].

The most common operations for ARM described were the PSARP, ASARP, and LAARP. The articles evaluating surgical technique were organized into six types of comparisons:

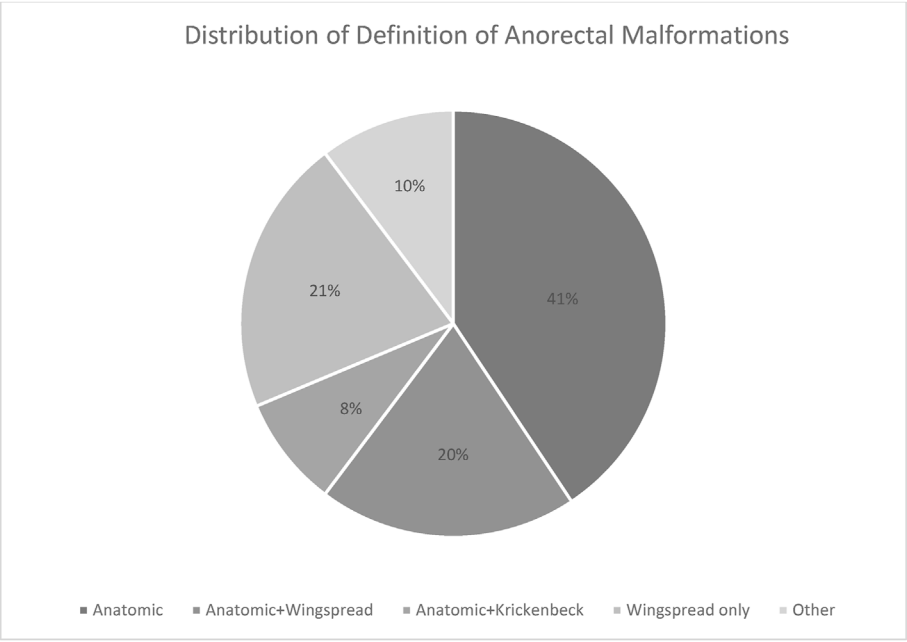

Fig. 2. Distribution of definition of anorectal malformations.

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Table 1
Recommended anorectal malformation nomenclature.

Nomenclature	Definition
Anterior ectopic anus	A normal anus, with a normal size, dentate line, and anal canal, that appears to be in a slightly anterior position (ie short perineal body) but is correctly centered within the sphincter complex. *This term should be abandoned despite its use in medical coding as well as prior literature.*
Perineal fistula in a male	The distal rectum inserts into the perineal skin anterior to the perfect center of the sphincter complex, is measured as smaller than appropriate for the child's age, and/or creates a sub epithelial tract
Perineal fistula in a female	The distal rectum inserts into the perineal skin (not mucosalized tissue) anterior to the perfect center of the sphincter complex, is measured as smaller than appropriate for the child's age, and/or creates a sub epithelial tract
Anal stenosis	A perfectly centered anal orifice within the sphincter complex that is measured as too small for the child's age, with or without a skin-lined funnel anal canal
Rectobulbar urethral fistula	The distal rectum inserts at the "elbow" of the urethra or more distal on the urethra as seen on an imaging study (ie distal colostogram or other) [7]
Rectoprostatic urethral fistula	The distal rectum inserts proximal to the "elbow" (at the triceps) of the urethra but still distal to the bladder neck on an imaging study (ie distal colostogram or other) [7]
Recto bladder neck fistula	The distal rectum inserts at the bladder neck (above the urinary sphincter) on imaging study (ie distal colostogram or other)
Rectovaginal fistula	The distal rectum inserts into the posterior wall of the vagina proximally to the hymen, with a typical urethral orifice.
H-type rectovaginal fistula (congenital)	A normal vagina and normal anus with a fistula connecting the two anatomic structures
Vestibular fistula	The distal rectum inserts into the vestibule distal to the hymen and within mucosalized tissue, with a typical urethral orifice
Rectal atresia	An anal canal perfectly centered within the sphincter complex and a rectal atresia proximally.
Rectal stenosis	An anal canal perfectly centered within the sphincter complex with a rectal stenosis proximally.
ARM with no fistula	A blind ending distal rectum with no communication to the urinary tract or vagina without a normal anal canal

- Evaluation of PSARP/ASARP technique only [6–8,10,21,24,27,31,33,38,39,42,48,51,55–57,60,61,66,67,69–71].
- ARM repair of patients with and without a stoma [17,41,43,47,53,63,64,74].
- Timing (early vs. delayed) of PSARP [28,30,35,54,62].
- LAARP repair only [9,20,25,26,29,40,49,52,58,59].
- LAARP vs. PSARP technique [18,19,22,32,34,37,44–46].
- LAARP with and without a stoma [23,50,68,72,73].

Studies often contained heterogenous types of malformations but reported complications as a cohort. Additionally, age and weight at operation, length of stay, associated anomalies including

Table 2
Definition of type of Anorectal Malformation surgical repair and stages of surgical approach.

Surgery	Definition
Posterior Sagittal Anorectoplasty (PSARP)	With the child in the prone position, electrical stimulation to define the anterior and posterior sphincter limits, midline incision in the posterior sagittal plane, followed by definition of the posterior and lateral planes around the fistula, and lastly, separating the anterior rectal wall from the posterior vaginal wall or urethra. This separation is then followed by gaining of rectal length, perineal body repair, tacking the posterior rectal wall to the sphincter, closure of the posterior sagittal wound, and anoplasty. While the posterior sagittal incision historically extended from the coccyx anteriorly to the fistula, minimization of the PSARP incision has become more favored in recent times especially for paitents with perineal and vestibular fistulas.
Anterior Sagittal Anorectoplasty (ASARP)	With the child in supine position, a posterior sagittal incision is created from the posterior margin of the fistula to the posterior limit of the proposed anoplasty site, followed by dissection through the perineal muscles, identification of the rectum, separation of the rectum from the anterior structures (typically the vaginal wall), gaining of rectal length, rectal pull-through, and reconstruction of the sphincter complex. Commonly performed for vestibular fistulas in patients who are genetically born female.
Sphincter Sparing PSARP	Typically described as dissection of the fistula while leaving the perineal body intact. Once mobilized, the rectum is passed underneath the perineal body into the sphincter followed by anoplasty.
Cutback Anoplasty Operation	A procedure that consists of creating a deep posterior sagittal incision in the posterior wall of the rectum and the sphincter. The rectal wall is then sutured to the posterior skin. This is typically performed in the case of a perineal fistula with sphincter around the majority of the anal opening or a perineal fistula at the anterior aspect of the anal sphincter.
V–Y Plasty	Creation of a triangular flap of tissue (making a V-shaped incision from the edges of the anus/anoplasty) and repositioning it to increase the anoplasty diameter.
Potts Anoplasty (anal transposition)	The fistula and rectum are mobilized and transposed through the muscle complex to the site of new anus via an incision in the center of the external sphincter as determined by muscle stimulator.
Anal Transposition	A term that is often used as part of other surgical approaches such as Potts anoplasty, trans-sphincteric ano-rectoplasty (TSARP), trans-fistula ano-rectoplasty (TFARP) and sphincter-saving ano-rectoplasty (SSARP)
Internal Sphincter Sparing Anoplasty (ISSA)	A subcategory of Sphincter Sparing PSARP (and similar to the concept of anal transposition). Creation of an incision in the anal sphincter complex, followed by the repair of the anorectal malformation, aimed at preserving the internal anal sphincter muscle and avoiding damage to the sphincter complex (i.e. extending the incision beyond or through the sphincter). The end of the fistula is preserved in this approach to preserve where the internal anal sphincter is thought to reside.
Laparoscopic Assisted Anorectoplasty (LAARP)	Use of laparoscopic instruments to facilitate the dissection and identification of the rectum and rectourethral or rectovaginal fistula if applicable, followed by rectal pull-through and reconstruction of the sphincter complex.
Repair stages	**Description**
Single stage repair	A single-stage repair involves a single surgery to reconstruct the anus and rectum in one procedure.
Two stage repair	A two stage repair involves two separate surgeries, typically performed several months apart. In the first surgery, the surgeon reconstructs the anus and creates a temporary colostomy to allow the reconstructed area to heal. In the second surgery, the colostomy is closed.
Three stage repair	A three-stage repair involves three separate surgeries, typically performed over several months or years. In the first stage, the surgeon creates a colostomy to divert stool away from the anorectal area. In the second stage, the anus is reconstructed. In the third stage, the colostomy is closed.

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spinal anomalies, length of follow-up and proportion of patients who reported outcomes including continence were not consistently reported across publications.

5. Male and female perineal fistula anorectal malformations

5.1. Perineal fistula

Twelve studies described female perineal fistula repair in a total of 1078 patients, and 9 studies described male perineal fistula repair in 298 patients. The most common surgical approaches for repair in these studies were ASARP and PSARP techniques [6,21,24,27,31,32,35,38,39,42,48,54,60,63,64,67,69,71]. In all of these manuscripts, it is unclear what proportion of these patients had an ostomy created prior to ARM repair. The reason for this ambiguity is that while these studies reviewed the outcomes for perineal fistula repair, they often included mixed populations of malformation types. While reporting on the presence of a stoma prior to ARM repair was not standardized between publications, 13.8% of females (N = 149/1078) and 9.7% of males (N = 29/298) with perineal fistulas had a stoma in place at the time of their ARM repair. Primary complications reported following ARM repair included rectal prolapse, anal stricture, and wound infection (5–20% for each complication). Three papers specifically discussed perineal fistula repair with and without colostomy creation, but in the data reported the stoma formation was not assigned specifically to the ARM malformation reported thereby making comparison for wound complications between patients with diverted and undiverted perineal fistulas remaining unable to be assessed [47,63,64].

6. Other female anorectal malformations

6.1. Rectovestibular fistula

A total of 27 studies evaluated surgical management specific to rectovestibular fistula in 2200 female patients [6,17,21,24,27,28,31–33,35,38,39,41–43,47,48,51,53,54,60,61,63,64,69,70,74]. Nine studies evaluated performing a PSARP/ASARP with and without a colostomy [17,28,41,43,47,53,63,64,74]. The most common surgical approach described was the PSARP or ASARP, and sphincter sparing techniques were also commonly described [51,70]. Other surgical techniques described included cutback anoplasty, anal transposition, internal sphincter sparing, and sacroperineal approaches. For those undergoing a PSARP/ASARP the most common complications cited were wound infection (4.8%; N = 71/1494), anal stenosis (4.4%; N = 65/1494), and rectal prolapse (4%; N = 61/1494). A stoma was present prior to surgery in 18.3% (273/1494). Again, the data presented in these manuscripts was unclear in regard to the timing of the stoma placement before or after ARM repair. A smaller number of studies did compare patient outcomes with (N = 114) and without (N = 215) stomas at the time of definitive repair of rectovestibular fistula. In these series wound infections were similar between single stage without a stoma (19.8%; N = 54/273) and three-stage repairs (18.4%; N = 21/114).

Four studies reported laparoscopic approaches (LAARP) for rectovestibular fistula in a total of 52 patients [25,29,34,49]. A colostomy was created for all of these patients prior to repair. Rectal prolapse (17.3%; N = 9/52), wound infection (5.8%; N = 3/52) and anal stricture were the most common complications following laparoscopic repair of rectovestibular fistula (28.8%; N = 15/52).

6.2. Rectovaginal fistula

Eight studies evaluated a total of 47 patients with rectovaginal fistula utilizing PSARP or ASARP only ARM repair (N = 23), or as part of a comparison of ARM repair with and without stoma (N = 24) [6,8,24,27,42,47,53,56,63]. The surgical technique used in studies comparing patients with and without stoma was also PSARP or ASARP approach.

Five studies reviewed laparoscopic repair of rectovaginal fistula in a total of ten patients, often in comparison to PSARP alone without laparoscopy [18,22,26,34,46]. A colostomy was created prior to repair for all patients. Overall, complication rates were difficult to establish as again, each? series contained multiple ARM subtypes and complications were recorded together. However, rectal prolapse, anal stenosis and wound infections occurred in decreasing order. The distance between the fistula insertion into the vagina and the perineum was not reported in any of these studies.

7. Other male anorectal malformations

7.1. Rectobulbar urethral fistula

Ten papers specifically discussed repair for ARM of the rectobulbar fistula subtype [8,30,39,42,53,57,63,64,74]. Five articles described PSARP in a total of 42 patients, the majority of which received a colostomy prior to surgery [8,39,42,55,57]. Complications included rectal prolapse (33.3%; N = 14), anal stenosis (14.3%; N = 6), and wound infection (9.5%; N = 4). Three studies compared single stage primary PSARP vs. three-stage repair (N = 47 single vs. N = 60 three-stage). Increased risk of wound infection was reported in the single stage repairs, but the exact increase could not be quantified due to the manner in which the data were reported [53,64,74]. A single institution study described timing of PSARP in males with colostomy but did not report any post operative outcomes [30].

LAARP approach for rectobulbar urethral fistula was evaluated in 11 studies [22,25,34,36,40,46,49,52,58,59,72]. Most studies evaluating LAARP performed the surgery in three stages (N = 164) (Table X) [25,40,49,52,58,59]. Rectal prolapse and anal stenosis were the most frequently reported complications. Four studies compared LAARP vs. PSARP repair (N = 31 LAARP vs. N = 22 PSARP) [22,34,36,46]. Rectal prolapse was seen in both groups (41.9%; N = 13/31 LAARP vs. 59%; N = 13/22 PSARP), and wound infection occurred more frequently in those with a PSARP approach (0%; N = 0/31 LAARP vs. 13.6%; N = 3 of 22 PSARP).

7.2. Rectoprostatic urethral fistula

Rectoprostatic fistula is reported in the reviewed literature to be repaired most commonly via LAARP surgical approach. Eight studies evaluated LAARP in a total of 195 patients, all of whom had a colostomy created prior to definitive repair [20,25,26,29,40,49,58,59]. The most common complication was rectal prolapse (11.3%; N = 22) followed by anal stenosis (4.6%; N = 9). Five studies evaluated patients who underwent PSARP vs. LAARP (N = 56 PSARP vs. N = 71 LAARP) [19,22,34,38,44]. Similar rates of rectal prolapse (41.1%; N = 23 vs. 49%; N = 35) and anal stricture (7.1%; N = 4 vs. 7%; N = 5) were observed, but the PSARP group had a higher rate of wound infection (21%; N = 12 vs. 0%; N = 0). The last group of 5 studies evaluated repair of rectoprostatic fistula using LAARP approach but grouped different numbers of stages of repair together including primary repair without a colostomy, two-stage repair, and single vs. three-stage repair [23,50,68,72,73]. In these comparative studies, rectal prolapse was the most common complication (one and two stage 11.3%; N = 6 of 53 vs. three stage 20.0%; N = 6/30), followed by wound infection (one and two stage 3.8%; N = 2 of 53 vs. three stage 10.0%; N = 3/30) and stricture (one and two stage 3.8%; N = 2 of 53 vs. three stage 0%; N = 0/30).

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PSARP was described as the method of repair in six studies [8,10,30,42,53,63]. Two studies evaluating only 7 patients with PSARP approach did not utilize a colostomy and anal stenosis was the only complication reported [8,10]. Three studies using PSARP approach evaluated groups with and without colostomy and timing of repair without recording complications in either group. A variety of surgical techniques were utilized to repair the ARM in these papers [30,53,63]. Mention of a remnant of the original fistula (ROOF) was inconsistently described as a complication.

7.3. Recto-bladder neck fistula

Since 1985, LAARP has been reported more commonly for patients with ARM of the bladder neck subtype [9,19,20,22,23,25,34,40,44–46,49,50,59,68,73]. For patients with bladder neck fistula and LAARP repair, a colostomy was created prior to repair in about half of the patients (N = 63 of 122; 51.6%). The most common complications noted following repair for these patients included rectal prolapse (N = 35, 29%) and anal stenosis (N = 11, 9%). Six studies were identified comparing LAARP vs. PSARP for recto bladder neck fistula (N = 22 LAARP vs. N = 21 PSARP) [19,22,34,44–46]. A colostomy was created primarily in all patients in these series. Those who underwent PSARP had a higher rate of rectal prolapse (PSARP 14.1%; N = 20 vs. LAARP 8.5%; N = 12) and anal stenosis (PSARP 4%; N = 6 vs. LAARP 0%; N = 0), and similar rate of wound infection (PSARP 7%; N = 10 vs. LAARP 8.5%; N = 12). Finally, those patients with bladder neck fistula who underwent LAARP primarily or with two-stage repair as reported in 4 studies, complications occurred in 4–26% [23,50,68,73]. Complications in these patients included rectal prolapse (13.6%; N = 3, vs. 25%; N = 13), anal stenosis (4.6%; N = 1 vs. 26.9%, N = 14), wound infection (4.5%; N = 1 vs. 3.9%; N = 2), and indication for eventual colostomy creation (N = 1).

While bladder neck fistula is repaired most often via LAARP approach, 5 studies described PSARP as a combined abdominal with perineal approach in a total of 7 patients. Two series compared single vs. three-stage approach (N = 2 single stage vs. N = 5 three-stage) [10,39,42,53,64]. Most of these patients had a colostomy created at birth. Primary complications again included rectal prolapse (N = 14, 88%), anal stenosis (N = 8, 50%), and wound infections (N = 2, 12.5%).

8. Recommendations and observations

Studies contained largely heterogenous groups of ARMs subtypes, making direct comparison for each surgical technique challenging. In genetically female patients, repair is typically performed via a PSARP/ASARP for rectoperineal, vestibular, and rectovaginal fistulas. Rectovaginal fistula is also reported to be repaired via LAARP, but the distance between the rectum and the perineum or other anatomical characteristics that made this approach favorable were not reviewed in any of the studies.

For genetically male patients, perineal fistulas were repaired most commonly via PSARP/ASARP, either with or without a colostomy. Rectobulbar fistulas were repaired most commonly via PSARP approach, and less frequently with LAARP. Prostatic and bladder neck fistulas were repaired most frequently with LAARP. A PSARP approach for rectoperineal fistula without a colostomy appeared to have a similar proportion of wound infections compared to those repaired with a colostomy created prior to definitive repair. These data, however, were not reported in a manner that allows for determination of total complication rate or direct comparison. Complications reported for patients with ARM repaired via LAARP approach occurred in the following order of decreasing frequency: rectal prolapse, anal stricture, wound infection, and persistent remnant of the original fistula.

Given the current absence of data demonstrating any single approach more appropriate than another for any particular ARM subtype, at this time management is best left to the discretion of the operating surgeon based on their specific expertise and the unique characteristics of the patient, including comorbid conditions. Based on the data available, PSARP/ASARP, and LAARP are acceptable methods of surgical repair for patients with ARM. In terms of specific surgical approach, timing of repair, repair with or without a colostomy, and indication for use of laparoscopy, the type of malformation and location of fistula anatomically does help to dictate the most ideal technique of intervention. Sphincter mapping techniques should be used to avoid mislocation of the anoplasty. Laparoscopic approaches have been favored for male rectourethral fistulas with the inferior most rectum above the pubococcygeal line while PSARP type approaches have been favored as more ideal for male fistulas with the inferior most rectum below the pubococcygeal line [7]. Futhermore, it has been suggested that if the rectum is the first structure encountered from the posterior sagittal approach, success can be achieved utilizing the posterior-only PSARP approach. This has not been studied, however, in a systematic manner and variation in practice pattern definitely exists amoung colorectal surgical experts. Lacking this ideal study setup in current studies makes it difficult for more specific comparisons and recommendations to be made.

Common short-term complications seen for all surgical repairs include wound infection, prolapse, and stricture. Specific complications appear to be more common based on surgical technique performed with wound infection and dehiscence being potentially more common in primary PSARP repair and rectal prolapse and concern for remnant of original fistula being more common in laparoscopic techniques. Long-term complications were not able to be assed due to heterogeneity of the data and length of follow up. Please see supplemental Tables 1 and 2 for summary of article data.

- MINORS criteria scores for the reviewed manuscripts ranged from 4 to 21.
- Level of Evidence 3–4; grade of Recommendation C.

Question 3. In patients for whom fecal diversion is indicated, what is the preferred type of colostomy that should be created? Where in the colon is the optimal location for colostomy?

Two main types of colostomy have been reported for fecal diversion in the setting of anorectal malformations, including loop and divided. The ideal segment of colon to be utilized has been debated, but most studies focus on the transverse, descending, or sigmoid colon when creating a colostomy. Twenty articles were identified through the initial search and after review, 15 were applicable to the above question [39,73,75–87].

Peña et al. published a review of 1470 patients who had undergone colostomy, 1420 of which were from a referring facility [84]. The 50 performed at their institution were created as a divided colostomy in the descending colon. Four hundred and sixty-four patients from referring hospitals had an ostomy complication (33%), compared to 8% (n = 4) of the 50 patients with stoma placement by Pena et al. Of this 8%, all complications occurred after stoma closure. The most common complications (N = 616) associated with stoma placement noted by the authors on initial evaluation included mislocation (defined as either stomas located too close together, too distal, or too high on the abdominal wall) (46%, n = 282) and prolapse (19%, n = 119). Stoma revision was required in 16% of patients (n = 230/1420), most commonly due to mislocation (60%, n = 137).

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8.1. Type of colostomy: Loop vs divided

In a study of 473 patients, including patients with Hirschsprung and/or anorectal malformations, loop colostomies (N = 364) had more associated complications overall than divided colostomies (N = 109) (88%, n = 316 vs 61%, n = 65) [75]. This study did not stratify populations and complications by disease type. Other smaller studies (N = 157 and N = 171 patients, respectively) found no significant difference in total complications between divided and loop colostomies [76,81]. A study of 144 patients did find a statistically significant difference (p = 0.031) in the rate of total complications, with a rate of 31.5% (23/73) in loop colostomies versus 15.5% (11/71) in divided colostomies [83]. Gardikis et al. reviewed the course of 68 patients, and also noted significantly higher overall complications in loop colostomies (p = 0.001) [77]. A study of 182 children with ARM found a statistically significant higher rate of complications in divided colostomies (44% (4/9) vs 12% (22/171)) [82]. Two studies found no difference in rates of stoma revision between divided and loop techniques [81,83].

The rate of prolapse for loop colostomies in these series was reported between 7.7% and 23.4% [75–77,81,82] compared to 0%–11% for divided colostomies [75,77,81–83,87]. In four studies, this difference was statistically significant, and in two it was not [75,77,81–83,87]. The rate of stomal obstruction or stenosis was similar between loop and divided colostomies, occurring in less than 6% of the patients [75,81,83]. There were no differences identified in wound infection rates or rates of stoma necrosis [81]. Two studies found no difference between type of stoma and retraction rates [81,83]. No studies directly compared the rate of wound dehiscence. One study found a dehiscence rate of 3% (2/58) in divided colostomies, and while a subsequent series identified a rate of 7.7% (2/26) of dehiscence for loop colostomies [76,77].

A noted concern related to loop colostomies is the inability to separate the proximal and distal ends of the bowel, thus allowing stool to potentially spill into the urinary tract in those patients with a rectourethral fistula and result in an increased risk of urinary tract infections (UTIs). In a study of 171 patients examining loop vs. divided colostomies, UTI risk was not increased in those who underwent a loop colostomy (44% vs 39%, NS (p = 0.64)) [81]. Mullassery et al. and Oda et al. reviewed 182 and 144 patients, respectively, and showed no statistically significant difference in UTI rates for patients with divided vs. loop colostomies [82,83].

The divided sigmoid colostomy has been safely performed laparoscopically with minimal complications [78,86]. One advantage of this technique is the added ability to examine the internal gynecologic structures in female patients. In a single series evaluating laparoscopic colostomy, a single reported complication of mucus fistula prolapse was noted [86]. Another method of colostomy creation involves a skin and muscle bridge between a sigmoid loop colostomy, which may allow for more complete diversion and prevent retraction [80]. Iqbal et al. reviewed a series of 60 infants who underwent this procedure, 34 (57%) of which had an ARM diagnosis. Ninty-three percent of these stomas were created in the sigmoid colon. The most common post-operative complication noted was parastomal excoriation (28%, 17/60). Other complications included stoma retraction (3%, 2/60), necrosis (3%, 2/60), prolapse (5%, 3/60), and stenosis (8%, 5/60).

8.2. Umbilical colostomies

Three papers described the technique of an umbilical colostomy [73,79,85]. Hamada et al. performed seven temporary umbilical loop colostomies in patients with anorectal malformations (rectobulbar and rectovesibular fistulas) [79]. Three of these umbilical colostomies were in the sigmoid colon and four were placed in the transverse colon. A single complication of mucosal prolapse was identified. Five stomas were closed electively after completion of the anoplasty. Yang et al. created temporary umbilical colotomies in 20 patients with prostatic urethral and bladder neck fistula subtypes of anorectal malformations [73]. Fifteen of these ostomies were located in the transverse colon and five were in the sigmoid colon. Several months later, these stomas were closed and LAARP was performed. These authors reported no complications, including parastomal hernia, obstruction, wound infection, or urinary tract infections. Sakaguchi et al. compared temporary umbilical loop colostomies to traditional abdominal colostomies and found no difference in complication rates [85].

8.3. Location of colostomy in colon

Colorectal experts have recommended using the distal descending or proximal sigmoid colon at the location where it separates from the retroperitoneum as an ideal location for a colostomy due to its fixation at this point which may reduce the incidence of prolapse [88]. The transverse colon has fewer attachments, and transverse colostomies, therefore, may have a reported higher rate of prolapse [84]. One study that included patients with ARMs and other diagnoses found no difference in complication rates based on the segment of colon utilized in creation of the colostomy [75]. Demirogullari et al. reviewed 157 patients with ARMs and noted that descending colostomies had a higher complication rate compared to sigmoid or transverse colostomies (28% (11/38) compared to 10% (3/28) and 7.1% (6/84), respectively) [76]. Oda et al. found a higher rate of complications with transverse colostomies compared to those in the sigmoid colon (OR 4.33), particularly in regards to rates of prolapse [83].

In a study of Hirschsprung and ARM patients, the rate of prolapse was 23.1% (79/341) for transverse colostomies, vs. 13.6% (18/132) for sigmoid colostomies, which was statistically significant [75]. Mullassery et al. also found that transverse colostomies had a significantly increased rate of prolapse when compared to sigmoid colostomies [82]. Wilkins et al. evaluated 272 patients with colostomies and found no significant difference in prolapse rates between transverse and sigmoid colostomies [87]. Rates of stomal obstruction, stenosis, stricture, or revision did not differ between the groups [75,82].

9. Recommendations and observations

Both loop and divided colostomy may be acceptable methods of fecal diversion for patients with a diagnosis of anorectal malformation. Loop colostomies appear to have higher rates of prolapse in the literature reviewed. There are no identifiable differences, however, in rates of wound complication or urinary tract infection. In terms of the segment of colon used in creation of a colostomy, transverse colostomies also appear to have higher rates of prolapse, but there was no difference seen for rates of other complications.

- MINORS criteria scores for the reviewed manuscripts ranged from 3 to 20.
- Level of Evidence 3–4; grade of Recommendation C.

Question 4. What is the optimal timing of definitive repair in patients who are initially managed with fecal diversion? What is the optimal timing of definitive repair in patients who are initially managed with dilations?

Forty publications were determined to have enough data regarding timing of the operation. Three papers included a comparison of outcomes by age at operation; the remaining reported age at operation without relation to other findings [30,35,54].

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Twenty-seven publications included details regarding the timing of repair for patients initially managed with fecal diversion [17–20,25,28–31,36,37,41,43,45,50,53,56,58,64,72–74,77,89–92]. Eighteen publications included details about the timing of repair for patients initially managed with dilations [6,17,28,31,35,38,41,43,48,51,54,60–62,66,70,71,93].

Figure 3 shows the median or mean age at operation for patients initially managed with fecal diversion. Five papers provided only a median or mean age with no range [17,19,30,37,45]. Harumatsu et al. compared intermediate- and high-type ARM male patients operated on before five months of age (early group) to those repaired after five months of age (late group), included separately in the figure [30]. Those who underwent an early operation were described to have "better bowel function" but this is not well defined and therefore hard to interpret when concluded in these papers. Three papers reported patients grouped into a range for age of surgical repair: two to 12 months, three to six months, and six to 24 months [29,77,89]. The initial operation occurred before one year of age in 20 publications [17,20,25,30,31,36,37,41,43,45,50,53,56,58,64,72–74,91,92]. The median or mean age was within three to six months in nine publications [29,50,53,58,64,72–74,91].

Figure 4 depicts the median or mean age of operation for patients initially managed with dilations. One publication had only a mean age reported (3 months) [38]. Two publications reported patients grouped into a range for age of surgical repair: four to six months, and zero to 60 months [17,48]. Nine publications performed the anoplasty prior to six months of age [35,38,51,54,60,61,66,70,93]. Five publications performed the operation between six and 12 months of age [6,31,41,43,62,71]. Irfan et al. compared early repair (<7 days old) to delayed repair (8 days old to 6 weeks of age) for babies with perineal and rectovestibular fistula, and found no differences in wound and non-wound complications including reoperations, and readmissions [35]. A study using NSQIP-P database (National Surgical Quality Improvement Program – Pediatric) evaluated those with perineal fistula and rectovestibular fistula repaired early (<7 days of life) vs. delayed (6 weeks–8 months) [35]. A study using the PCPLC consortium database also evaluated timing of repair as early <14 days vs. delayed >14 days after birth [54]. Both of these studies demonstrated equivalent rates of complications (18.2%; 12 of 66 early vs. 12.1%; 28 of 231 late and 12.9%; 4 of 31 early vs. 15.0%; 20 of 133 late) and wound infections (13.6%; 9 of 66 early vs. 10.4%; 24 of 231 late and 6.5%; 2 of 31 patients in early vs. 5.3%; 7 of 133 late).

10. Recommendations and observations

Only three studies directly compared surgical outcomes based on age at repair; two found no difference in post operative complications. While a wide range of patient ages were often grouped together for analysis, the majority of publications reported patients initially managed with colostomy underwent definitive ARM operation before one year of age. Similarly, for patients managed with initial dilations, the majority of authors (16/18) performed an anoplasty before one year of age, with a little over half of those occurring in the first 6 months of life. While both early and delayed repair of ARM appears safe from a post operative complication standpoint, the optimal timing for definitive repair in patients initially managed with either fecal diversion or dilations could not be determined from this review. The timing of surgery should be based on surgeon preference, and patient characteristics, with an understanding that most surgeons perform definitive ARM repair before age 6 months for those undergoing dilations, and before one year in those that have undergone diversion.

- MINORS criteria scores for the reviewed manuscripts ranged from 7 to 21.
- Level of Evidence 2–4; grade of Recommendation D.

11. Discussion

This review summarized the heterogeneous literature available related to initial surgical management of ARMs. The overall scientific quality of the literature was poor with predominantly Level of Evidence 3–4 and Grade of Recommendations C-D. This systematic review highlights the need for prospective, multicenter studies to understand best practices for ARM patients better.

Uniform definitions and precise anatomic descriptions of the location of fistula should be utilized for the reporting of type and classification of ARMs in order to better compare patient groups.

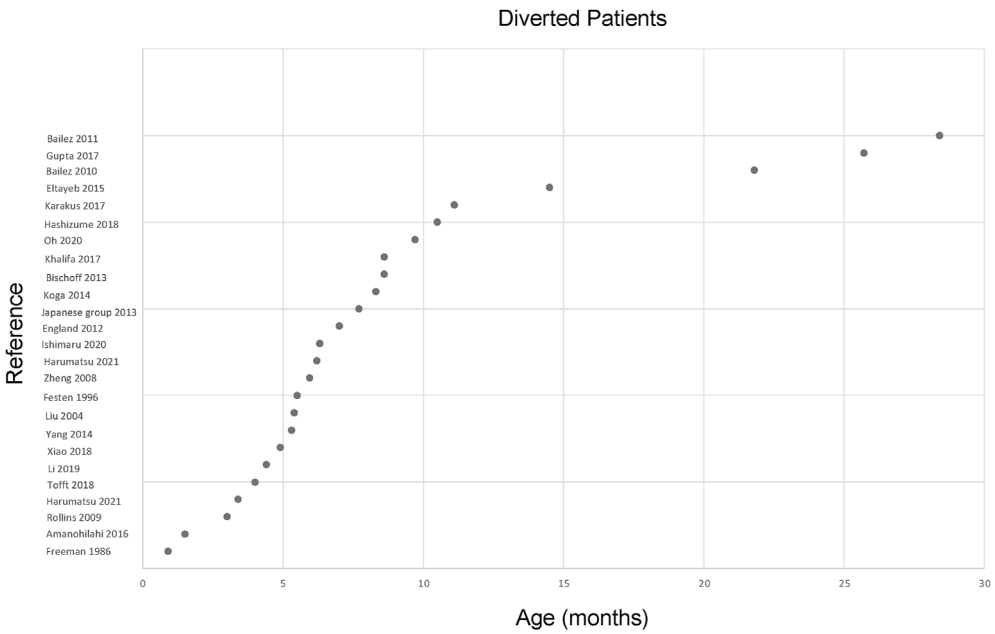

Fig. 3. Age per reference of surgical repair in patients diverted with an ostomy.

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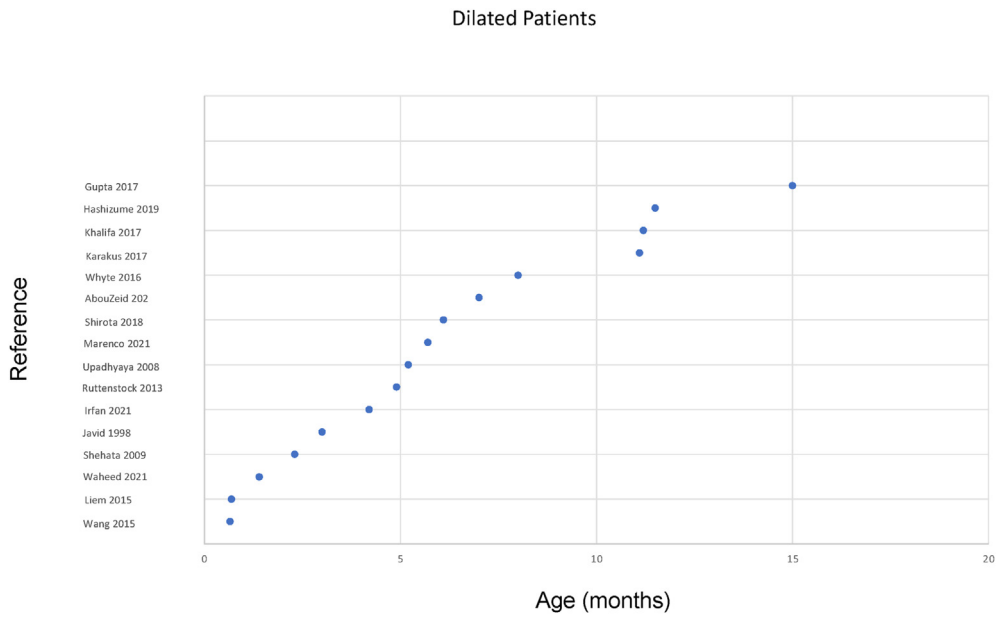

Fig. 4. Age per reference of surgical repair in patients managed initially with dilations.

Efforts through national and international consortia to streamline and unify definitions and improve diagnostic clarity of ARMs are underway [12]. The authors recommend using anatomic descriptions based on fistula location for research efforts as opposed to grouping ARM patients into subgroups in order to improve our ultimate understanding of their outcomes and allow for more specific short and long term data acquisition [11,12].

While multiple operations are used in practice to repair ARMs, the current literature historically has compared heterogeneous groups of patients and operations, making definitive recommendations challenging. In these cases, while we cannot definitively say the ideal approach for surgical intervention based on the available evidence, expert involvement and potential referral should be pursued early in care if there are any questions about repair approach, as many technical pitfalls exist for these operations. Both loop and divided colostomy may be acceptable methods of fecal diversion for patients with a diagnosis of anorectal malformation, however, loop colostomies do appear to have higher rates of prolapse in the literature reviewed in this series. The optimal timing for definitive repair in patients initially managed with either fecal diversion or dilations could also not be determined from the review. However, between early and late ARM group repairs there did not appear to be a significant difference in short term post operative complications. This literature review demonstrates the need for high quality, multicenter studies through consortiums in order to better answer these questions. Ultimately, ARMs are a rare and heterogeneous conditions with a high rate of comorbid conditions that greatly impact outcomes and management. World wide efforts are ongoing through the Pediatric Colorectal and Pelvic Learning Consortium, European Anorectal Malformations-Net Reigistry (ARMNet), and other consorita to help answer the remaining questions outlined by this review and many more to continue efforts to improve the outcomes and quality of life for these patients.

Conflict of interest

None of the authors have a financial or other conflicts of interest related to the issues addressed in this manuscript.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jpedsurg.2024.06.007.

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