

ERN eUROGEN Guidelines on the Management of Anorectal Malformations, Part II: Treatment

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Abstract

Keywords

- ▶ ARM
- ▶ guidelines
- ▶ quality standard
- ▶ evidence-based
- ▶ consensus

Introduction Anorectal malformations (ARMs) are rare birth defects affecting the anorectum and oftentimes the genitourinary region. The management of ARM patients is complex and requires highly specialized surgical and medical care. The European Reference Network eUROGEN for rare and complex urogenital conditions aimed to develop comprehensive guidelines for the management of ARM applicable on a European level.

Methods The Dutch Quality Standard for ARM served as the basis for the development of guidelines. Literature was searched in Medline, Embase, and Cochrane. The ADAPTE method was utilized to incorporate the newest available evidence. A panel of 15 experts from seven European countries assessed currency, acceptability, and applicability of recommendations. Recommendations from the Dutch Quality Standard were adapted, adopted, or rejected and recommendations were formed considering the current evidence, expert opinion, and the European context.

Results Surgical and medical treatment of ARM, postoperative instructions, toilet training, and management of fecal and urinary incontinence were addressed. Seven

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new studies were identified. The panel adapted 23 recommendations, adopted 3, and developed 8 de novo. The overall level of newly found evidence was considered low.

Conclusion Treatment of ARM patients requires a multidisciplinary team and expertise about anatomical and surgical aspects of the disease, as well as long-term follow-up. This guideline offers recommendations for surgical and medical treatment of ARM and associated complications, according to the best available evidence and applicable on a European level.

Introduction

Anorectal malformations (ARMs) comprise a wide spectrum of congenital malformations of the anorectum and often including genitourinary region, and associated spinal, neurological, and orthopaedic anomalies.^{1,2} The management of ARM is complex and requires highly specialized surgical and medical care. While some ARMs are relatively easy to manage and have good functional outcomes (e.g., perineal fistula), others are challenging to treat, both in the short and long term (e.g., cloacal malformations).¹ Advances in our understanding of the pelvic anatomy and factors affecting long-term outcomes have led to a standardization of reconstructive procedures and postsurgical protocols since the first description of the posterior sagittal anorectoplasty (PSARP) by Peña and deVries in 1982 and the laparoscopically assisted anorectal pull-through (LAARP) by Georgeson et al in 2000.^{3,4} New techniques such as the anterior sagittal anorectoplasty (ASARP) were described and minimally invasive procedures were developed.⁵ Yet, uncertainties remain around the optimal treatment strategy of ARM and differences in management between countries persist.

In an effort to harmonize management strategies of ARM at the European level, the European Reference Network (ERN) eUROGEN for rare urogenital diseases and complex conditions assembled a panel of experts (the Adoption and Adaptation Working Group [AAWG]) with the aim to develop comprehensive guidelines for the management of ARM. The Dutch "Quality Standard for Anorectal Malformations" was used as a template for the adaptation process within the European context, which was performed according to the ADAPTE method.^{6,7} By adapting and updating the Dutch Quality Standard with the newest available evidence, the ERN eUROGEN guidelines aim to provide a patient-centered standard of care that bridges gaps and settles inconsistencies in treatment between European countries. The objective is to achieve a standardization in management of ARM and ultimately improve patient outcomes and quality of life.

The adaptation report comprises six categories: diagnostics, treatment, organization of care, collaboration, referral, and communication between providers, lifelong follow-up, and transition of care. Part two of the guidelines will focus on recommendations for surgical and medical treatment of ARM, postoperative instructions, toilet training, and management of fecal and urinary incontinence.

Methods

The methods for the adaptation of the Dutch Quality Standard for ARM has been reported previously (part I). Briefly, the Dutch Quality Standard was appraised according to the Appraisal of Guidelines for Research and Evaluation instrument and recommendations were adapted in adherence with the ADAPTE method (detailed methodology available in the Supplementary Material [online only]).^{7,8} A systematic review of the literature since 2017 was performed according to the PRISMA guidelines to identify new studies relevant to the treatment of ARM patients in the following databases: Medline, Embase, and Cochrane. The original search strategy as well as inclusion and exclusion criteria for studies is outlined in the Supplementary Material. Original recommendations and newly identified studies were assessed by the AAWG, a panel of 15 experts from seven European countries, including two patient representatives, within the ERN eUROGEN during a face-to-face meeting in Nijmegen (The Netherlands). Recommendations were either adopted, adapted, or newly developed based on the available evidence (→Table 1). To improve the rigor of development compared with the Dutch Quality Standard, a grade of recommendation was issued by the AAWG and methodologists (W.F.E.I., K.M.) for all recommendations (→Table 2). For some recommendations, it was unclear from the Dutch Quality Standard whether this recommendation was based on expert opinion only or if there was also supporting evidence. In this case, the recommendation was discussed by the AAWG, and a grade D recommendation ("expert opinion") was chosen if there was no supporting evidence.

Table 1 Definition of adopted, adapted, and new recommendations

Adopted	Recommendation is endorsed by the AAWG without changes
Adapted	Recommendation was modified and adapted to the needs and context of the ERN
New	Recommendation was created based on the included new evidence or expert opinion

Abbreviations: AAWG, Adoption and Adaptation Working Group; ERN, European Reference Network.

Table 2 Correspondence between grades of recommendations and study level of evidence

A	Based on consistent level 1 studies
B	Based on consistent level 2 or 3 studies or extrapolations from level 1 studies
C	Based on level 4 studies or extrapolations from level 2 or 3 studies
D	Based on expert opinion or inconclusive/inconsistent studies of any level.

Results

The following sub-questions were re-assessed by the AAWG:

- Which treatments are suitable for patients with ARM? Which reconstructive operations are performed, and which treatments are still needed afterwards?
- What instructions are given to parents for actions after the reconstructive operations? Which health care provider gives the instruction and when is this instruction given?
- What is the work-up for the treatment of urinary and fecal continence problems and any associated complications? What individually adaptable protocol can be followed for toilet training and at what age is this offered?
- What is good prevention and treatment for diaper dermatitis?

- Which aids are available (incontinence materials, rinse aids)? Which care provider can advise on this?
- How is/are the parent(s) of the patient with ARM supported in self-management?

A total of seven new studies addressing the treatment of ARM were identified and considered by the panel in their recommendations.^{9–15} A detailed summary of all newly included studies for each sub-question is provided in the Supplementary Material. The overall level of newly found evidence was considered to be low. Overall, 23 recommendations were adapted from the Dutch “Quality Standard for Anorectal Malformations,” 3 were adopted, and 8 were developed de novo. Key recommendations of the panel according to the available evidence are summarized in **Fig. 1**.

Recommendations for Treatment Modalities

- Which treatments are suitable for patients with ARM? Which reconstructive operations are performed, and which treatments are still needed afterwards?

Patients with ARM should be referred to a pediatric surgical department as soon as the diagnosis is made (**Table 3**). Within the first 24 hours, the patient must be thoroughly examined and meconium discharge as well as type of ARM assessed. Additional comorbidities and potential lethal pathologies, especially congenital heart diseases, should be identified. In case of bowel obstruction, the patient should

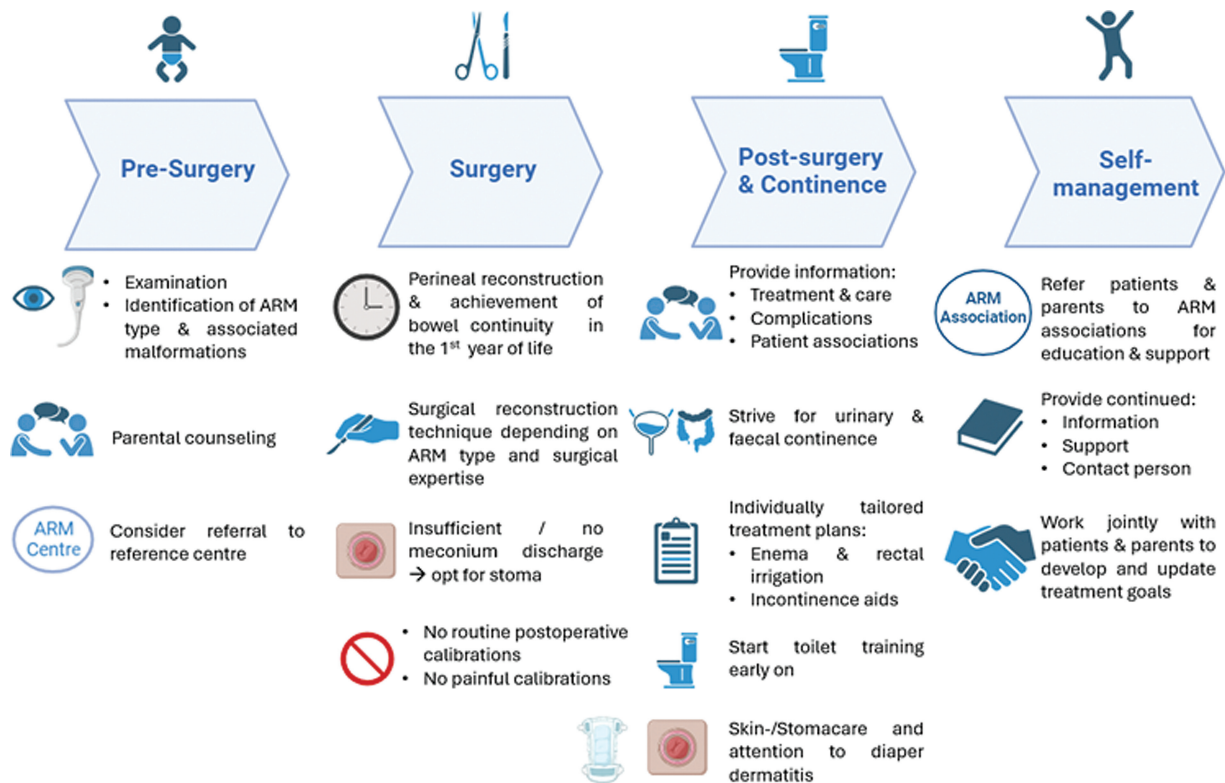


Fig. 1 Key recommendations for surgical and medical treatment strategies of children with ARM from the prenatal period to adulthood (made with Biorender).

Table 3 Recommendations for treatment modalities

Treatment modalities		Grade of recommendation
1	Refer patients with an ARM diagnosis to a pediatric surgery department when available. The following measures are recommended: <ul style="list-style-type: none"> • Patient examination in the first 24 hours on whether there is meconium discharge and which type of ARM is involved. • Exclusion of other potentially lethal pathology, especially congenital heart disorder. • Intravenous fluid management in case of bowel obstruction or enteral nutrition if defecation is possible. • Parental counselling on the condition. [adapted]	D
2	Patients with cloaca and cloacal exstrophy should be referred to a reference center with experience in the multidisciplinary treatment of these patients and long-term follow-up. [new]	D ⁹
3	In patients with insufficient or absent meconium discharge, establish a venous access and insert a nasogastric tube. Provide IV antibiotics. Inform the parents not to feed the baby or child. [adapted]	D
4	For forms of ARM in which there is no meconium discharge after birth, surgery is performed. Often, three surgeries are required: <ul style="list-style-type: none"> • Creating a colostomy • The reconstruction surgery • Reversal/closure of the stoma [adapted]	D
5	If there is a phenotype of ARM in which (sufficient) meconium is discharged (e.g., ARM with perineal or vestibular fistula or anal/rectal stenosis), the anus should be calibrated and/or painlessly dilated until the daily evacuation of meconium is possible. If there is no sufficient meconium evacuation, a stoma should be considered. [adapted]	D ²
6	Choose the timing of reconstructive surgery (either in the neonatal period or during infancy) based on the height of the rectum and the presence of a connection to the urinary tract. [adapted]	D
7	Primary reconstruction and restoring bowel continuity should be done within the first year of life, unless there are contraindications for surgery such as life-threatening comorbidities (e.g., cardiac). [new]	C ¹⁰
8	In certain cases of perineal fistula, nonoperative treatment might be considered. [new]	B ¹¹
9	Type of colostomy: no evidence for superiority between either loop versus split. Place: descending sigmoid stoma is preferred but, in some cases (cloacal malformations or associated gastrointestinal anomalies), a transverse stoma might be an option. [new]	D
10	The chosen surgical technique depends on the abnormalities found and the surgeons' expertise. The most common surgical techniques are the formation of an anoplasty, ASARP, PSARP, LAARP (ASARP: anterior sagittal anorectoplasty; PSARP: posterior sagittal anorectoplasty; LAARP: laparoscopically assisted anorectoplasty), or a combination of the different techniques. [adapted]	C ¹²
11	In case of anal stenosis at the skin level during follow-up after anorectoplasty, anal dilatation under general anesthesia should be considered, otherwise stricturoplasty. [new]	B ¹³
12	Postoperative routine calibrations at home may be considered but are not mandatory. Painful calibrations must by all means be avoided. If a calibration program is started, it should not be longer than 3 to 4 months, and an intensive follow-up is needed. [new]	D

Abbreviation: ARM, anorectal malformation.

be managed with intravenous (IV) fluids, otherwise enteral nutrition is permitted. Patients with complex ARM such as cloacal malformation or cloacal exstrophy ought to be referred to a reference center with experience in multidisciplinary treatment and long-term follow-up.⁹ For all types of ARM, parental counselling on the condition is essential from an early stage on.

The immediate and long-term management of patients with ARM depends on the presence or absence of sufficient meconium discharge during the first day of life. In patients

with insufficient or absent meconium discharge, first steps include establishing a venous access, insertion of a nasogastric tube, initiation of a nothing-by-mouth protocol, and IV antibiotics. Then, a three-tiered surgical approach consisting of creation of colostomy, reconstructive surgery, and stoma reversal is generally required. Concerning the type of colostomy, either a loop or split stoma can be created with no clear evidence favoring one technique over the other, although loop colostomies may be prone to stoma prolapse.¹⁶ A descending colostomy is the preferred method, but in

some cases (e.g., cloacal malformation) a transverse stoma may be considered.^{1,17} In ARM forms, where sufficient meconium is discharged (e.g., ARM with perineal fistula), the anus should be calibrated and/or painlessly dilated until daily evacuation of stool is possible.²

Timing of reconstructive surgery is dependent on many factors including the height of the rectum and the presence of a connection to the urinary tract, but should be performed within the first year of life, provided there are no contraindications to surgery.¹⁰ The surgical technique for reconstruction depends on the type of ARM and surgical expertise. Established techniques include anoplasty, ASARP, PSARP, LAARP, or a combination of different techniques.^{3–5,12} Clear superiority of one technique over the other has not been demonstrated and rather depends on the local surgical expertise and patient-specific context. Additionally, whether a single-stage or multi-stage surgery is more suitable is a matter of ongoing debate.¹² However, in case of extended or destructive wound infection, a colostomy might be required. For selected cases of perineal fistula, where the anus is surrounded by at least 80% of sphincter musculature and the size is sufficient, a nonoperative approach may be considered.¹¹ In any case, pros and cons of each approach need to be outlined and discussed with parents (or guardians) to make an informed decision.

If during follow-up, an anal stenosis at the skin level develops, anal dilatation under general anesthesia may be considered, but stricturoplasty represents a valuable alternative.¹³ Postoperative routine calibrations are not mandatory but can be considered.¹³ It is important to note that studies suggest remarkable distress of nurses involved in performing dilatations and psychosocial trauma of parents and patients alike.^{18–20} Additionally, anal dilatations have been reported to be painful in more than two-thirds of cases, thus, it is essential to avoid painful calibrations by all means.²¹ Furthermore, calibrations should not be performed for longer than 3 to 4 months and require intensive follow-up.

Recommendations for Postoperative Instructions for Parents

- What instructions are given to parents for actions after the reconstructive operations? Which health care provider gives the instruction and when is this instruction given?

Comprehensive information about the condition, treatment, (postoperative) care, possible complications, and follow-up should be given to parents of ARM patients by both pediatric surgeons and nurses from the beginning according to the AAWG (→Table 4). This includes information about constipation and risk of incontinence, as well as the possibility of multiple surgeries even at an older age. Information should be offered repeatedly, and health care professionals need to assure themselves that it was understood by parents. Additionally, easily understandable resources, such as leaflets, brochures, Web sites, or webinars, as well as contact information of patient organizations, need to be made available. If no ARM-specific patient organizations exist within their own country, information about international patient organizations for education and support should be given. In case a child receives a stoma, a specialized nurse should be involved to provide dedicated stoma care and educate parents on stoma management.

Recommendations for Toilet Training and Treatment of Fecal and Urinary Incontinence

- What is the work-up for the treatment of urinary and fecal continence problems and any associated complications? What individually adaptable protocol can be followed for toilet training and at what age is this offered?

Achieving urinary and fecal continence should be the goal for all ARM patients. If continence cannot be achieved, all involved parties should aim for the highest possible quality of life, possibly by implementing bowel management to attain pseudo-continence (→Table 5). An individually tailored protocol considering patient-specific requirements and the child's maturity should be developed with patients and parents. Toilet training can be initiated between 2 to 5 years of age. Special attention should be paid to the care of the perineal skin, proper posture, and seating (e.g., toilet seat reducer), as well as regular toilet times. If problems arise during toilet training, a multidisciplinary team, consisting of pediatric surgeons, nurses, physical therapist, and possibly psychologists should be involved.

It is essential that parents are educated to recognize constipation and subsequent overflow incontinence. Bowel incontinence may be managed by enema, rectal irrigation, or oral laxatives (when associated with constipation). If rectal irrigation is not possible or inefficient, antegrade irrigation via a Malone stoma can be considered. In case of

Table 4 Recommendations for postoperative instructions for parents

Instructions to parents		Grade of recommendation
1	Pediatric surgeons and nurses must inform parents on available ARM resources such leaflets, brochures, Web site, webinars on the (neonatal) aspects of ARM in an understandable way (i.e., lay language), etc. as well as patient associations. [adapted]	D
2	The pediatric surgeon should provide relevant information to parents whose children undergo ARM surgery about conditions, treatment, care after surgery, and possible complications. [adopted]	D
3	A specialized nurse in ARM should always be involved after the operations of a child receiving a stoma. [adapted]	D

Abbreviation: ARM, anorectal malformation.

Table 5 Recommendations for toilet training and treatment of fecal and urinary incontinence

Toilet training		Grade of recommendation
1	It is crucial to strive for urinary and fecal continence. If continence cannot be achieved, the highest possible quality of life should be aimed (e.g., by facilitating pseudo-continence with bowel management). [adapted]	D
2	It is important to keep in mind at all times of the patient's course, the association between ARM and urological malformations, and to act accordingly, involving the relevant health care professionals. [adapted]	D
3	The approach to toilet training in children with ARM, depending on complexity, comorbidity, and psychosocial context, can be initiated at the age of 2 to 5 years (as with all children). [adapted]	D
4	An individually tailored protocol according to the children's ARM sub-type and form is recommended for toilet training. This is, on the one hand, to be done in connection with the different degrees and forms of ARM and comorbidity and on the other hand in connection with the differences per child in maturity and development phase. [adapted]	D
5	In case of problems with toilet training (unsuccessful management, whether or not related to complex problems), it is recommended that the toilet training problem should be discussed and treated within a multidisciplinary team. [adopted]	D
6	It is recommended during fecal toilet training to pay attention to <ul style="list-style-type: none"> Care of the perineal skin. Toilet posture, toilet seat reducer, relaxation of the child during toilet use, using fixed times on the toilet. Pediatric pelvic floor physical therapy. Optional supervision by a psychologist/remedial educationalist. [adapted]	D
7	Patient-parents should be educated to recognize constipation and subsequent overflow incontinence. [adapted]	D
8	Bowel incontinence may require an enema, rectal irrigation, or laxatives (when associated with constipation), which could be temporary. [adapted]	D
9	If rectal irrigation is not working or not possible, e.g., rectal trauma, physical inability, antegrade irrigation should be considered. [adapted]	D
10	In case of persistent and specific problems of fecal incontinence, re-do surgery may be considered, in coordination with an experienced surgeon or center. If redo surgery is not recommended, definite stoma may be considered as a last resort. [adapted]	C ¹⁵

symptomatic patients with mislocated anus or stenosis, re-do surgery in concertation with an experienced surgeon or center may be contemplated at any stage.¹⁵ As a last resort, a definite stoma can be created. Furthermore, associations between ARM and urological malformations should be kept in mind and if necessary, pediatric urologists should be consulted. Lastly, it is important to note that recommendations on management of incontinence are mainly based on expert opinion and that evidence originating from studies is lacking.

Recommendations for Treatment of Diaper Dermatitis

- What is good prevention and treatment for diaper dermatitis?

Proper care of wound, stoma, and perianal region needs to be ensured and should be provided on a regular basis by specialized skin and/or stoma nurses in and outside the hospital (→ **Table 6**). Especially the perianal region needs to be monitored, as recurrent or extensive wound infections can have a negative impact on long-term functional outcomes. Parents can perform sitz baths or apply ointments (e.g., zinc ointment) to relieve irritations and use hydrophilic

barrier creams to protect the perineal skin from stool and urine. Similarly to the previous section about treatment of incontinence, recommendations for prevention and treatment of diaper dermatitis are based on experience and consensus among the AAWG, rather than robust evidence.

Recommendations for Products Aiding in the Management of Incontinence

- Which aids are available (incontinence materials, rinse aids)? Which care provider can advise on this?

Many different types of incontinence materials exist and can help in managing fecal incontinence. Depending on the national context, availability of materials, reimbursements options, and possibility of support by qualified specialized nurses in implementing measures at home can differ vastly. Patients and parents should be provided with comprehensible information about available options, ideally in collaboration with specialized nurses and physicians or surgeons (→ **Table 7**). Incontinence aids include manual or automated enema/irrigation systems with a variety of nozzles, incontinence briefs, or protective padding. Patients and parents

Table 6 Recommendations for treatment of diaper dermatitis

	Diaper dermatitis	Grade of recommendation
1	Wound, stoma, and perianal care should be monitored by a skin and/or stoma nurse in or out of the hospital on regular basis. [adapted]	D

should be encouraged to contact patient associations for further support and exchange with other affected parties.

Recommendations for Supporting Parents and Patients in Self-Management

- How is/are the parent(s) of the patient with ARM supported in self-management?

Empowering patients and parents to manage themselves is key for a successful treatment and patient (or parent)/health care professional relationship.^{22,23} Patients should be referred to patient associations for support and education about self-management of their disease and the opportunity to exchange with similarly affected individuals (► Table 8). A contact point or person available 24/7 should be provided to patients. Health care professionals and patients and/or their parents should work together to create an individually tailored care plan to reach personal treatment goals. Jointly formed treatment goals should be documented, periodically evaluated, and adjusted if needed.

Evidence Gaps and Future Research

While this document addresses a wide range of aspects crucial for comprehensive patient care and provides recommendations for different treatment modalities, postoperative care, management of incontinence, and self-management, future research is needed to optimize treatment of patients with ARM. Even though advancements have been made in the past decades, there is still a lack of studies with high level of evidence and quality. In fact, many recommendations, especially on postoperative care, are based on expert opinion rather than robust evidence, and the field would greatly benefit from conducting randomized controlled trials, possibly in a multi-site setting and high-quality prospective registries. Combined with clearly specified PICO (patient/population, intervention, comparison and outcomes)-type questions issued for the revision of this guideline, higher grades of recommendations may be feasible for some topics. Type of surgical reconstruction is mainly based on local surgical expertise and there is little evidence supporting one type of reconstructive surgery or colostomy over the other, or a single versus multi-stage approach. Recommendations for postsurgical care and especially use of postoperative calibrations vary between centers, but there is strong consensus that postoperative calibrations must be painless. Lastly, future investigations on how supporting patients and parents in self-management ameliorates treatment outcomes and perceived quality of life may further enhance management of ARM patients.

Table 7 Recommendations for products aiding in the management of incontinence

Available aids		Grade of recommendation
1	Patients and parents should be provided with good information about incontinence materials and rectal irrigation products, ideally in collaboration between a specialized ARM nurse and a physician or surgeon. [adapted]	D
2	Patients and parents should be informed of additional aids for incontinence, together with their limitations. [adapted]	D
3	It is recommended that parents and/or patients contact their ARM patient associations for further support. [new]	D

Abbreviation: ARM, anorectal malformation.

Table 8 Recommendations for supporting parents and patients in self-management

Self-management support of parents and patients		Grade of recommendation
1	Refer patients with ARM to patient associations for support and education about self-management of the disease, or to other patients for peer learning. [adapted]	D
2	Include in the patient information material an up-to-date point of contact or a named person to whom patients can reach out to when needed, for information and/or support in 24/7. [adapted]	D
3	It is recommended to work jointly with (the parents of) a patient with ARM to develop a care plan to reach personal treatment goals. [adapted]	D
4	Health care professionals should document information about agreed treatment goals in the medical records and/or individual care plans. [adopted]	D
5	It is recommended to review the treatment goals and care plan periodically. [new]	D

Abbreviation: ARM, anorectal malformation.

Conclusion

Treatment of ARM patients requires a multidisciplinary team and expertise in anatomical and surgical aspects of the disease as well as long-term follow-up. Part two of the eUROGEN guidelines, developed by an expert panel from major European centers, offers recommendations for surgical and medical treatment of ARM and associated complications according to the best available evidence and applicable on a European level. By fostering a unified evidence-based approach for ARM treatment across Europe, this guideline aims to provide a benchmark for management of ARM and enhance patient care.

Collaboration

This work was performed in collaboration with experts involved in ERN ERNICA, EUPSA, and Qualicura.

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Conflict of Interest

The authors would like to disclose that two of the authors of this adaptation report (I.D.B., C.S.) were also authors of the Dutch Quality Standard. However, it is important to note that neither I.D.B. nor C.S. has any conflict of interest to declare and they did not advocate for any particular viewpoints or positions in relation to the content of the Dutch Quality Standard. No conflicting interests were declared from the other authors.

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