Anorectal malformations – surgical aspects and transition

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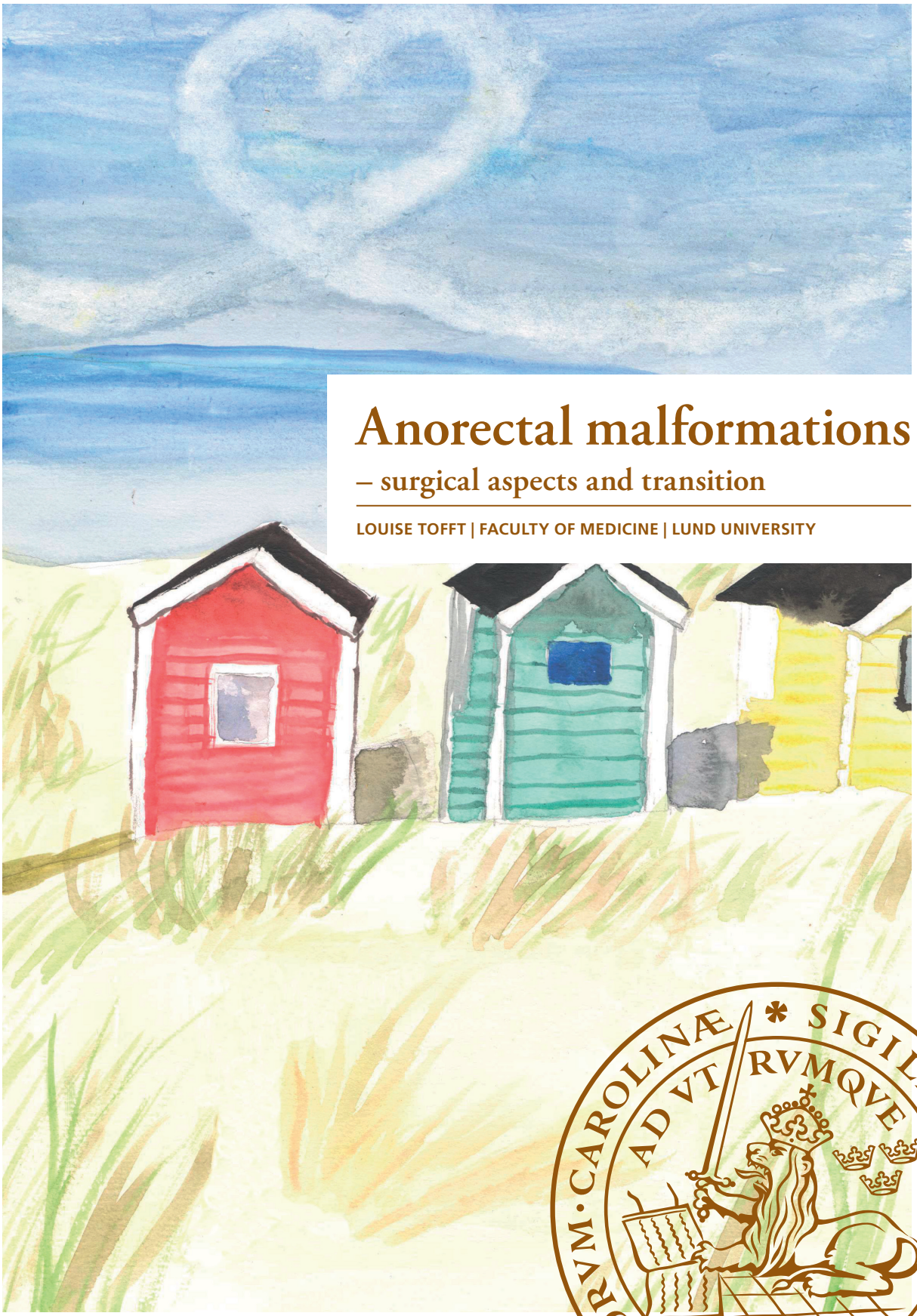

Anorectal malformations

– surgical aspects and transition

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LUND
UNIVERSITY

DOCTORAL DISSERTATION

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ANORECTAL MALFORMATIONS – SURGICAL ASPECTS AND TRANSITION

Abstract

Background: Anorectal malformations (ARM) occur in 1/5000 live births (1.2–1.6:1 male:female). Associated malformations are common, influencing, together with ARM-subtype complexity, long-term outcome, follow-up and need of transitional care. Reconstructive surgery, the posterior sagittal anorectoplasty (PSARP), is performed in infancy, either as a single-stage or multi-stage procedure with a colostomy.

Aims: 1. Assess frequency and risk factors of post-PSARP wound dehiscence. 2. Explore patient-expressed needs and expectations of transitional care. 3. Assess accuracy of pre-PSARP fistula diagnostics, and 4. significance of postoperative abdominal scarring and propose a scar treatment approach.

Methods: 1. Retrospective study of all ARM infants treated in Lund between 2001–2016 evaluating PSARP wound complications and management including multivariate logistic regression analysis of potential risk factors. 2. Focus group study of ARM adults with qualitative content analysis. 3. Retrospective study of male ARM infants treated in Lund 2001–2020 defining pre-PSARP fistula diagnostic accuracy of radiological- and endoscopical modalities compared to intra-PSARP subtyping. Diagnostic superiority receiver-operating characteristic curve (ROC) analysis. 4. Patient- and observer-reported cross-sectional study of previously multi-stage PSARP treated ARM children (>5y) and adults, evaluating abdominal scar symptoms through the validated Patient and Observer Scar Assessment Scale (POSAS), including Pearson's *r* for correlation with age, and pictorial evaluation of possibility of scar treatment by a plastic surgeon. Ethical approval was obtained.

Results: 1. 90 infants (41% female) were included. Colostomy protected against wound dehiscence; 11(22%) versus 17(43%) p=0.04. No risk factor was identified. 2. 16 adults (63% female) were included. Identified key elements of adequate transition: improved knowledge of ARM among patients and adult care givers, support with coping strategies, structured communication between patient, paediatric- and adult care, and easy access to specialised adult care through patient navigators. 3. 38 male infants were included. Cystoscopy and high-pressure colostogram had the highest diagnostic accuracy (70% and 66%, respectively). No diagnostic superiority was identified. 4. 27 children and adults (48% female) were included. 6(22%) reported scar pain, 5(19%) pruritus and 9(33%) altered behaviour. Worse POSAS score and increasing age correlated, r=0.4 p=0.04. 21(78%) were technically suitable for plastic surgery, among whom 8(30%) requested treatment. No gender differences were identified.

Conclusions: Colostomy seems to protect against post-PSARP wound dehiscence. Patients identify knowledge, structured communication and easy access to specialised adult care as key factors for adequate transition. Current pre-PSARP fistula diagnostics only reach a maximum accuracy of 70%. Postoperative scar morbidity should be addressed in follow-up, and plastic surgery considered.

Key words Anorectal malformations, posterior sagittal anorectoplasty, wound dehiscence, transition, fistula, colostogram, cystoscopy, voiding cystourethrogram, endoscopy, post-operative scarring, scar treatment

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To the late Ingmar Grybäck.

Former Chief of Surgery
at Centralsjukhuset in Kristianstad,
my first mentor in Paediatric Surgery,
and in Signe's words: 'Farfar-doktorn!'.
In memoriam.

Table of Contents

List of papers

This thesis is based upon following original papers, hence referred to as Paper I–IV:

I. Tofft L, Salö M, Arnbjörnsson E, Stenström P. Wound dehiscence after posterior sagittal anorectoplasty in children with anorectal malformations. Biomed Research International. Volume 2018; Article ID 2930783 https://doi.org/10.1155/2018/2930783/

II. Tofft L, Hoel A, Håkansson C, Zawadzki A, Gjone H, Øresland T, Bjørnland K, Stenström P. Key components of successful transition for adolescents born with anorectal malformations – a Nordic focus group study. International Journal of Adolescent Medicine and Health. 2020; 20200052 https://doi.org/10.1515/ijamh-2020-0052/

III. Tofft L, Salö M, Arnbjörnsson E, Stenström P. Accuracy of pre-operative fistula diagnostics in anorectal malformations. BMC Pediatrics. 2021; 21:283 https://doi.org/10.1186/s12887-021-02761-6/

IV. Tofft L, Klasson S, Salö M, Hambraeus M, Arnbjörnsson E, Stenström P. Patient-reported physical and psychosocial significance of abdominal scarring in anorectal malformations. (Submitted for publication)

Abbreviations

ARM	Anorectal malformations
AUC	Area under the curve
BFS	Bowel Function Score
CI	Confidence interval
CT	Computed tomography
GI-tract	Gastro-intestinal tract
MRI	Magnetic resonance imaging
OR	Odds ratio
PedsQL	Pediatric Quality of Life Inventory™
POSAS	Patient and Observer Scar Assessment Scale
PSARP	Posterior sagittal anorectoplasty
PSARVUP	Posterior sagittal anorectal vaginal urethroplasty
ROC	Receiver-operating characteristic curve
SGA	Small for gestational age
UTI	Urinary tract infection
VAC	Vacuum-assisted closure
VACTERL	Vertebral-, Anorectal-, Cardiac-, Tracheal-, Oesophageal-, Renal- and Limb malformations
VCUG	Voiding cystourethrogram

Preface

A historical centralisation of care in Sweden regarding some paediatric surgical diagnoses including anorectal malformations (ARM) began in 2016, aiming to improve quality of care. The final decision was preceded by a thorough application process, in which different aspects of highly specialised paediatric surgical care were assessed by the Swedish National Board of Health and Welfare.

In June 2018, highly specialised care in paediatric surgery for ARM, Hirschsprung's disease, vaginal aplasia, bladder exstrophy, oesophageal atresia and congenital diaphragmatic hernia was centralised in Sweden, from four tertiary paediatric surgery departments originally, to two national highly specialised care centres today: Skåne University Hospital in Lund and Karolinska University Hospital in Stockholm.

For me, as a fairly new resident in paediatric surgery in 2016 at the Department of Paediatric Surgery in Lund, the department's application process to become a national highly specialised care centre prompted the beginning of my clinical thesis in ARM. I was given the task of contributing to the accounting of surgical outcomes in the application by compiling data from our clinical registers and evaluating surgical outcome and complications in ARM over the last 15 years. It gave me a complete overview of all ARM patients treated in Lund since 2001 when our current digital medical record system was launched.

The application process raised a very important question for us all: What aspects are important in achieving high-quality care for ARM patients?

This question led to the writing of my first paper in this thesis regarding frequency and risk factors for wound dehiscence, a known complication after reconstructive surgery for ARM.

Surgeons usually avoid talking about the complications that they have encountered during their operations, but transparency and honesty are vital for improving surgical outcome. By investigating how, when and hopefully why surgical complications occur, countermeasures can be effectuated and evaluated, and the frequency of complications may potentially decrease. This is a quite hands-on way of promoting high-quality care.

As time goes by, your tiny infant patient grows up. One day, a tall and somewhat surly teenage patient stands before you, demanding a completely different

competence from you than he or she did as an infant or their guardians did. When working with patients born with ARM, it soon becomes apparent that ARM is a chronic condition, despite reconstructive surgery in infancy. Patients often live with a hidden handicap of, for example, severe constipation and/or faecal- and urinary leakage, affecting all aspects in their life including intimate relationships. ARM therefore requires potential life-long follow-up and management. A well-functioning transition from paediatric- to adult care is requested, but sadly reported to be inadequate by our patients. Exploring patients' self-expressed needs and expectations of transitional care in ARM became my second paper in this thesis. Hopefully this will be one step further towards high-quality care in ARM throughout the patient's life.

Another aspect of surgical outcome in ARM, important for any responsible surgeon and for promoting high-quality care, is adequate surgical work-up regarding the correct preoperative anatomical imaging of the patient's malformation. Surgeons need to be prepared and have their proposed surgical approach ready in order to carry out the operation to the highest level of expertise. No surgeons want any anatomical surprises in the operating room. In my third paper of this thesis I evaluated the accuracy of preoperative fistula diagnostics in ARM by current radiological- and endoscopical modalities. Improvements are within reach.

A previously scientifically neglected aspect of surgical outcome in ARM, brought to our attention by our patients at follow-up, is postoperative abdominal scarring after previous diverted colostomies and other associated surgical interventions in childhood. These visible and public signs of a congenital condition are seldom desired by patients: to some patients they cause much distress. Defining physical and psychosocial significance of abdominal scarring after surgery performed in infancy for patients born with ARM and proposing a scar treatment approach constituted my fourth paper in this thesis. Maybe this aspect of quality of care is transferable to other paediatric surgical patient groups?

To conclude, in order to define and promote necessary elements for high-quality care in ARM, the overall purpose of this clinical thesis was to provide new knowledge and novel aspects on challenges for further improving surgical outcome and follow-up treatment in patients with ARM, from the perspectives of both the patient and the paediatric surgeon.

Background

Anorectal malformations

Anorectal malformations (ARM) constitute a spectrum of congenital malformations of the anorectum and pelvic floor [1,2]. The malformation is also referred to as 'anal atresia' or 'imperforate anus', illustrating the typical status when a child is born without an anal opening or with a misplaced one.

The incidence of 1/5000 live borns with ARM is fairly consistent world-wide [3,4]. This translates to 40–50 children born every year with ARM in Sweden [5].

ARM are more common in boys (1.2–1.6:1) [4,6,7] and are associated strongly with other congenital anomalies: 30–70% of ARM patients are reported to have at least another concomitant malformation [3,6]. Types and reported prevalence of associated malformations are shown in **Table 1** [3,7–13].

Table 1. Associated concomitant malformations in patients with ARM.

Malformations	**Reported prevalence** (%)
Vertebral	4–22
Cardiac	17–36
Tracheal and oesophageal	4–22
Gastrointestinal	5–15
Urogenital	26–70
Skeletal	12–44
VACTERL-association*	11–22

*≥ 3 of concomitant vertebral-, anorectal-, cardiac-, tracheal-, oesophageal-, renal- or limb malformations; ARM: anorectal malformations.

Associated syndromes are reported in 9–19% of cases, both non-chromosomal, such as Townes–Brocks syndrome and Currarino syndrome, and chromosomal, such as Trisomy 21 [8,11,14].

The genesis of ARM and associated anomalies such as the VACTERL-association has still not been clarified, but is considered multi-factorial on both a genetic and an epigenetic level. Genetic mapping is on-going with some observed family clusters. *De novo* mutations are considered to predominate [6,15,16]. Associated risk factors such as *in-vitro* fertilisation and maternal obesity have been observed [14,17]. ARM

occur early, around gestation weeks 4–8, through a not fully clarified disturbance of the foetal development of the gut and urogenitalia [15].

Subtypes

The following photographs and illustrations (**Figures 1–6**) show the most common subtypes of ARM in both genders with rectal fistulas at different levels [18]. ARM can also present without fistula.

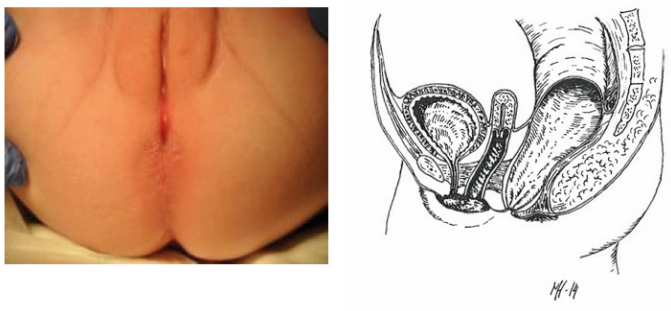


Figure 1. Female with a perineal fistula.

Figure 2. Female with a vestibular fistula.

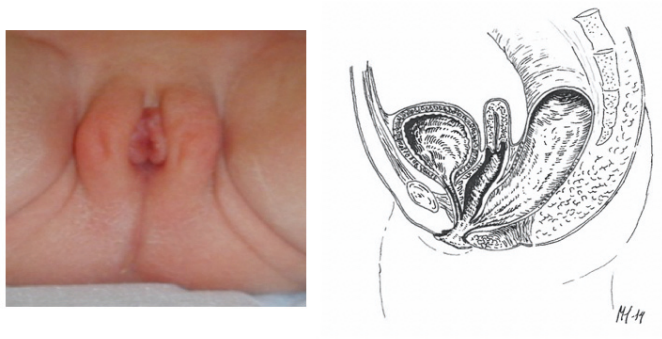


Figure 3. Female with a cloaca malformation.

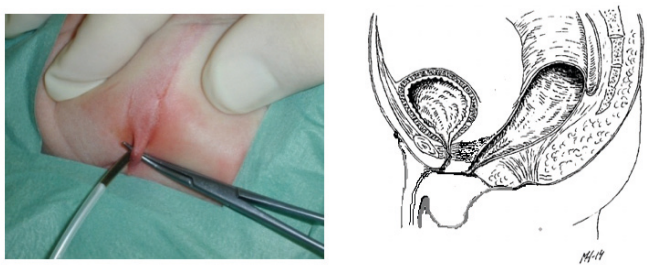


Figure 4. Male with a perineal fistula.

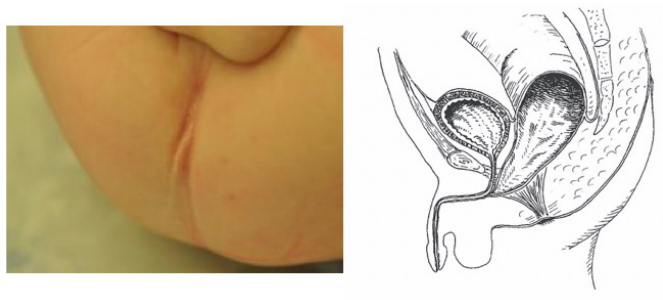


Figure 5. Male with either a recto-urethral fistula…

Figure 6. … or with a recto-bladder neck fistula.

The location of the rectal fistula partly determines whether or not the new-born child needs an acute diverting colostomy or can proceed directly to primary anorectal reconstruction surgery [19,20]. If a cutaneous anal opening is present, allowing adequate bowel emptying, and the infant has no other life-threating anomaly or medical condition, the need for an acute colostomy decreases.

Table 2. Reported ARM subtype prevalence in both genders [3,10].

Subtypes of ARM	**Females** (%)	**Males** (%)
Perineal fistula	21–41	41–43
Vestibular fistula	28–60	-
Cloaca	8	-
Recto-urethral fistula*	-	29–38
Recto-bladder neck fistula	-	6
No fistula	4–5	10–15
Others	6–19**	5–7***

*Further subdivided into recto-prostatic fistula and recto-bulbar fistula; **Anal stenosis, recto-vaginal fistula, H-fistula, rectal atresia, rectal stenosis, cloacal extrophy; ***Anal stenosis, pouch colon, rectal atresia, rectal stenosis, cloacal extrophy; ARM: anorectal malformations.

Surgery

Patients with ARM are treated surgically in infancy, either through a single-stage procedure, in which the infant is referred directly to primary reconstructive surgery of the anorectum and pelvic floor within a few weeks, or through a three-step procedure, in which the infant receives a diverting colostomy within a few days' post-partum, and then undergoes reconstructive surgery within a couple of months, and finally has the colostomy reversed when the anorectoplasty has healed [4,20].

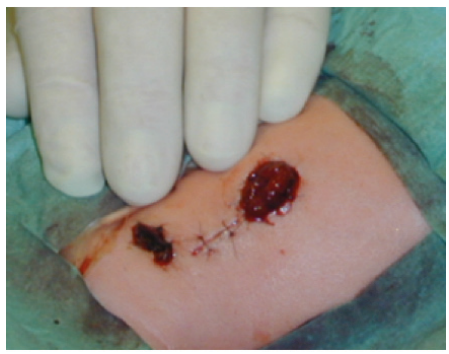


Figure 7. Divided colostomy.

Colostomy

A divided proximal sigmoidostomy has traditionally been the colostomy of choice in patients with ARM, allowing a complete diversion of the faeces and enough distal bowel length for the forthcoming reconstructive operation [4,21]. The classical procedure involves a partial abdominal paramedian incision not respecting skin Langer lines (**Figure 7**) [20], which might negatively influence aesthetically favourable wound healing after the colostomy is closed [22]. A loop sigmoidostomy is a less extensive procedure and preferred by some centres [21]. Whether or not the loop sigmoidostomy implies a higher risk of ostomy-related complications, e.g. distal stool impaction, urinary tract infection (UTI) or stoma prolapse, than the divided sigmoidostomy is under debate [23–26].

Preoperative malformation imaging

An infant born with ARM should be screened radiologically with plain X-ray, ultrasonography and sometimes with magnetic resonance imaging (MRI) for associated malformations, with special focus on cardiac-, sacral-, urinary tract- and genital anomalies plus tethered spinal cord since these associated malformations influence long-term outcome in patients with ARM [19,20,27–29].

If a recto-perineal fistula is not visible, preoperative fistula diagnostics are also undertaken in order to plan and perform the reconstructive surgery safely [30–34]. There are several radiological operator-dependent methods available in fistula diagnostics, of which high-pressure colostogram by passive colostomy (**Figure 8**) and voiding cystourethrogram (VCUG) are used most commonly. These have a diagnostic accuracy of 52–100% in the literature [31,35–39].

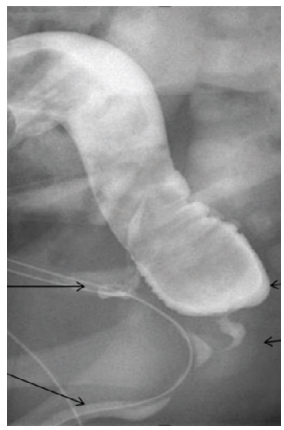


Figure 8. Colostogram.

Other radiological modalities in preoperative fistula- and pelvic diagnostics are upcoming, such as computed tomography (CT), high-frequency ultrasonography and MRI. Ultrasonography and MRI have the advantage of no radiation and the latter offers a non-operator dependent, potentially complete pre-operative anatomy imaging of the pelvic area [32,39–42]. MRI is therefore an appealing method but, to date, its use is limited due to current poor imaging resolution in infants, lack of studies on the impact of high-resolution MRI in infants plus the need for general anaesthesia during examination. Perioperative endoscopical methods for fistula diagnostics include cystourethroscopy and endoscopy of the passive colostomy [20,43].

Posterior sagittal anorectoplasty

Patients with ARM have undergone surgical reconstruction through posterior sagittal anorectoplasty (PSARP) since the 1980s, a method developed by Pieter A. Devries and Alberto Peña [44].

In PSARP, the patient is placed in the prone position, the pelvic floor is divided in the sagittal plane, the rectum is mobilised to skin level, the rectal fistula is located and divided, the neo-anus is centred in the external sphincter complex with the assistance of electro-myostimulation and is sutured to skin level, and the pelvic floor including the perineal body is restored (**Figure 9**).

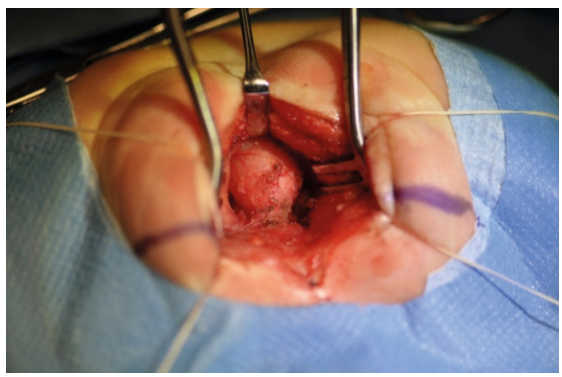


Figure 9. Posterior sagittal anorectoplasty (PSARP).

The mobilisation of the rectum and the division of a high fistula, such as the bladder neck fistula, can be assisted by a laparotomy or, in recent years, at most tertiary centres, by laparoscopy [45,46].

The most complicated ARM subtype in females, the cloaca malformation, is reconstructed through the posterior sagittal anorectal vaginal urethroplasty (PSARVUP), with an additional total urogenital mobilisation and, sometimes,

depending on the length of the common channel, assisted laparotomy with bowel–vaginal replacement [47].

The least complicated ARM subtype in both genders, the perineal fistula, is reconstructed through a limited PSARP, with less extensive pelvic floor dissection, commonly in a one-stage procedure without a colostomy [20].

Postoperative wound complications

Postoperative wound infection and dehiscence are well-known complications of the PSARP procedure, previously reported in 0–40% of single-staged PSARP [48–54], and are considered to influence surgical outcome in terms of misplaced neo-anus, increased patient suffering and consumption of healthcare resources by a prolonged in-hospital stay [20,21].

The pathogenesis of post-PSARP wound dehiscence is not understood fully or established scientifically. Different explanatory models have been presented, such as bacterial infection and inadequate tissue perfusion [2,26,55,56].

A three-step procedure is considered to be the safer alternative in preventing wound complications post-PSARP, bearing in mind that colostomy-related complications are reported to occur in 8–33% of cases [24,26].

In order to decrease the risk of wound dehiscence in single-stage PSARP, empiric prolonged intravenous antibiotic prophylaxis, topical antibiotics, preoperative bowel irrigation, postoperative fasting and loperamide are used [21]. There is, at present, one published case-control study in favour of vacuum-assisted closure (VAC) post-PSARP [57].

Postoperative scarring

The three-step reconstructive procedure plus all other surgical interventions, e.g. urinary tract operations, performed during childhood and adolescence in patients born with ARM [20,21], leave postoperative abdominal- and thoracic scarring: visible traces of the past on the patient's body.

Scarring after surgical procedures performed in infancy and childhood grows with the child [58,59]. Not respecting Langer lines when making surgical incisions, not following proper incision-closure techniques or not adhering to modern postoperative wound management strategies, predispose to a less aesthetically-favourable outcome with broad secondary healed and/or depressed scars [22,60,61].

The physical and psychosocial impact of scarring has been described previously in other patient groups, but not specifically in patients with ARM [59,62,63]. Follow-

up of scar morbidity, such as pain, pruritus and less aesthetically favourable outcomes, is not included in current ARM follow-up programmes [64,65].

Follow-up

ARM are considered to be a more or less chronic condition of varying degrees depending on the ARM subtype, associated anomalies and sometimes surgical iatrogenic injury [29,66]. Reported mortality is 3–16%, higher in extremely premature children and in patients with severe concomitant malformations [3,6].

The life-long morbidity, despite reconstructive surgery in infancy, lies in impaired bowel-, urinary tract- and sexual function, reported in up to 76% of adult patients, with e.g. bloating symptoms, chronic constipation and faecal- and urinary leakage. These symptoms require treatment in the form of laxatives, enemas and intermittent bladder catheterisation in order to maintain a social bowel- and bladder function [27–29,66–69]. Psychological problems, such as depression related to the somatic symptoms, have been reported [70,71]. ARM therefore demand potential life-long multi-disciplinary follow-up and treatment [27–29,65].

Transition

The need for a transition from paediatric- to adult care for patients with chronic ARM sequalae is undisputed among patient associations and paediatric surgeons [64,65,72–74].

Unfortunately, this transition is reported to be inadequate and is sometimes completely missing [28,29,68]. This has left adolescent and adult patients feeling alone and abandoned with severe, untreated bowel- and bladder symptoms, which affect their everyday life.

The main challenge of transitional care in patients with ARM, an experience shared by other patient groups in need of transition, is the actual interface towards adult healthcare. The sender, the paediatric surgery department, is well defined, but the intended recipient in adult healthcare is not. Knowledge and former clinical experience of rare congenital conditions, such as ARM may vary among adult healthcare providers [64,72,73,75–78], and may prevent an adequate transition. In addition, person-centred care is desirable in the entire healthcare system [79,80], but the manner in which adolescents born with ARM wish to design their transfer to adult care has yet to be explored and implemented.

Aims

Paper I

Assess the frequency of wound dehiscence after PSARP.

Identify possible risk- and protective factors for wound dehiscence after single-stage PSARP.

Paper II

Explore needs and expectations of transitional- and adult healthcare among adolescents and adults born with ARM.

Paper III

Evaluate the diagnostic accuracy of preoperative clinical symptoms, VCUG, high-pressure colostogram, and endoscopy of the urinary tract and diverted ostomy, regarding presence and location of fistulae compared to perioperative findings in male neonates born with ARM.

Paper IV

Assess the significance of postoperative abdominal scarring in children, adolescents and adults born with ARM, who have been reconstructed through a three-step procedure with a colostomy in infancy.

Identify any possible gender differences regarding the significance of postoperative abdominal scarring in ARM.

Propose a scar treatment approach in children, adolescents and adults born with ARM, who have been reconstructed through a three-step procedure in infancy.

Methods

Setting

The studies were conducted between 2016–2021 at the Department of Paediatric Surgery in Lund, Sweden.

The Department of Paediatric Surgery in Lund was, until June 2018, a low-volume tertiary paediatric surgical centre for ARM by international standards, serving 2 million residents. After the Swedish centralisation of paediatric surgical care in 2018, Lund became a high-volume centre for ARM, now serving 5 million residents. According to current national birth rates [81], Lund is now expected to treat 20–30 new-born children with ARM every year.

Reconstructive surgery in patients with ARM through the PSARP procedure was implemented at the Department of Paediatric Surgery in Lund in 1989 and, during the first 10 years, it was performed solely by one single senior consultant in paediatric surgery. During the years 2000–2018, reconstructive surgery was performed by a total of 10 paediatric surgeons. The present colorectal team, now performing all PSARP-procedures, includes two senior and two junior consultants in paediatric surgery. Stoma surgery is performed by all, at present 15, consultants at the department.

Study design and cohort

Children, adolescents and adults born with ARM and treated at the Department of Paediatric Surgery in Lund were the study participants of Papers I–IV. In Paper II, adult ARM patients treated at the Department of Paediatric Surgery in Oslo, Norway, were included additionally.

Papers I and III were retrospective medical record studies.

Paper II was a Nordic multi-centre qualitative focus group study from the Departments of Paediatric Surgery in Oslo and Lund and the Pelvic Floor Centres at the Departments of Surgery in Akershus, Norway and Malmö, Sweden.

Paper IV was a patient- and observer-reported cross-sectional study from the Department of Paediatric Surgery in Lund and the Department of Plastic Surgery in Malmö.

Inclusion and exclusion criteria

Paper I

Patients born with ARM who underwent either a single-stage or three-step reconstructive procedure through PSARP, limited PSARP or PSARVUP during 2001–2016 were identified in hospital registers. Patients who died before reconstruction, had their reconstructive procedure performed elsewhere, had not yet undergone their reconstructive procedure at the time of the study, or had anal stenosis that only required dilatations were excluded.

Paper II

Patients over 18 years of age and born with ARM were identified in hospital registers and invited to participate. Patients with cognitive disabilities or living more than a 3-hour car drive away from either Lund or Oslo were excluded.

Paper III

Male neonates born with ARM without a perineal fistula, reconstructed with a diverted colostomy and submitted to fistula diagnostics prior to PSARP during 2001–2020 were identified in hospital registers. Patients who had a single-staged PSARP without a diverted colostomy or a PSARP performed elsewhere were excluded.

Paper IV

Patients over 5 years of age, including adults, born with ARM and reconstructed with a diverted colostomy with a minimum of 4 years' follow-up after stoma closure were identified in hospital registers and invited to participate. Patients with cognitive disabilities or any previous surgical scar treatment were excluded. During the study period a few intended participants were excluded due to the institution's Covid 19-related patient contact restrictions for study purposes.

Outcome

Paper I

Primary outcome was wound dehiscence, defined as superficial (only skin) or deep (subdermal structures involved) and graded according to the Clavien Dindo classification [82], within 30 days after PSARP, limited PSARP or PSARVUP.

Analysed independent variables were gender, birth weight, cardiac malformation, VACTERL-association, colostomy, weight at surgery, postoperative antibiotics < 1 day (= one prophylactic dose), and postoperative fasting 0–3 days.

Paper II

Primary outcome was patient-expressed experiences of transitional- and adult healthcare in ARM.

Secondary outcomes were patient-expressed experiences of living with ARM and suggestions for improvements in transitional care.

Paper III

Primary outcomes were fistula presence and location.

Paper IV

Primary outcomes were physical symptoms of abdominal scarring, defined as pain, pruritus and visual appearance, and psychosocial effects, defined as patient-expressed feelings towards, and behaviours related to, their abdominal scarring.

Secondary outcomes were patient request for scar treatment and technical possibility of scar treatment.

Analysed independent variables were gender, age, Patient and Observer Scar Assessment Scale (POSAS) score [83], Bowel Function Score (BFS) [84] and Pediatric Quality of Life Inventory (PedsQL)^{TM} score [85].

Data collection

Paper I

Medical records were reviewed regarding:

- Background;
- Surgical procedures performed;
- Postoperative management after reconstructive surgery (antibiotic prophylaxis, fasting and wound management);
- Postoperative complications within 30 days after reconstructive- and stoma surgery (wound dehiscence, wound infection, sepsis, urinary tract infection and stoma complications);
- Postoperative complication management (antibiotic treatment, fasting, ostomy placement, re-operation).

Paper II

Data were collected through gender-divided focus group discussions [86,87] with adult ARM patients in Oslo and Lund. Eligible study participants were informed and invited by postal invitations.

A discussion moderator at each centre (with documented proficiency in group communication) initiated, enhanced and followed through the discussion topics among focus group participants with semi-structured open-ended questions and fictional case stories [88]. The moderator also ensured that every focus group participant was considered in the discussions. One or two other researchers at each centre acted as silent, note-taking observers during focus group discussions.

Topics discussed were previously known issues and challenges throughout life that might arise for people born with ARM: bowel- and urinary function, sexual function and intimate relationships, psychological issues, social aspects, school- and working life, and transition from paediatric- to adult care [27–29,68,89].

The focus groups were gender-divided due to the potentially sensitive topics discussed.

Discussions were audio recorded and transcribed verbatim with removal of any personal data.

Paper III

Medical records were reviewed regarding:

- Background;
- Preoperative symptomatology;
- Preoperative X-ray reports (high-pressure colostogram and VCUG);
- Perioperative examinations findings (cystoscopy and endoscopy of the atretic rectum);
- Final ARM-subtype classification during PSARP.

VCUGs and high-pressure colostograms were performed according to standard clinical practice [38,90] by five paediatric radiology specialists. No secondary reviews of X-ray reports were performed for the study.

Cystoscopies and endoscopies of diverted ostomies including fistula catheterisation with a guide wire from the atretic rectum to the urinary tract [43] were performed during PSARP anaesthesia by five paediatric surgeons.

Paper IV

The study was conducted in person at the Department of Paediatric Surgery in Lund. Eligible study participants or minors' guardians were contacted initially by phone and subsequently presented with written study information and invitation by email.

Children under 13 years of age participated with guardian assistance. Participants with several abdominal scars after surgical procedures related to the anorectal- or other concomitant malformations were asked to personally select and assess a maximum of three scars. The assessed scars were measured and photographed.

Scar assessment

The validated quantitative scar assessment instrument Patient and Observer Scar Assessment Scale (POSAS) [83,91] was used (**Figure 10**). It consists of a patient scar scale and an observer scar scale, with a final POSAS score of minimum 11 (no scar symptoms, just as normal skin) up to a maximum of 110 (severest scar symptoms, worst scar imaginable). A cut-off was set to 55 or higher for moderate to severe physical scar symptoms.

Participants answered additional questions regarding psychosocial aspects of abdominal scarring, any scar treatment requests and appropriate age for any scar treatment.

A senior consultant in paediatric surgery (with documented proficiency in plastic surgery) assessed each participant-selected scar clinically and completed the

observer scar scale. Neither the observer nor the participant (or guardian) was allowed to be involved in each other's score.

Observer Scar Assessment Scale

normal skin										*worst scar imaginable*
Vascularisation	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**
Pigmentation	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**
Thickness	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**
Relief	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**
Pliability	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**

Total score Observer Scar Scale:

Patient Scar Assessment Scale

No, no complaints										*Yes, worst imaginable*
Is the scar painful?	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**
Is the scar itching?	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**
No, as normal skin										*Yes, very different*
Is the colour of the scar different?	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**
Is the scar more stiff?	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**
Is the thickness of the scar different?	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**
Is the scar irregular?	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**

Total score Patient Scar Scale:

Figure 10. Patient and Observer Scar Assessment Scale (POSAS).

Scar treatment assessment

Afterwards, a senior consultant in plastic surgery conducted a pictorial scar treatment assessment regarding the possibility of scar treatment and improvement, potential surgical-, medical- and anaesthesiological methods and treatment timing. The plastic surgeon had all patient data available, except information on gender and any scar treatment requests enabling an objective assessment.

Bowel- and urinary function, and quality of life assessment

The established quantitative bowel function assessment instrument BFS [84] was used. The BFS maps bowel symptoms and bowel management, e.g. constipation, faecal soiling and use of enemas, with a minimum score of 1 (worst) and a maximum of 20 (best). A cut-off value of 17 or lower was considered as impaired bowel

function [84]. Participants also answered additional questions regarding the overall impact of bowel function on everyday life.

Urinary function was assessed by a questionnaire mapping different urinary tract symptoms and urinary management, e.g. incontinence, urinary tract infection (UTI), use of intermittent catheterisation and the overall impact of urinary function on everyday life [92].

The validated and age-adapted instrument PedsQL^{TM} [85] was used for participants under 19 years of age regarding overall physical and psychosocial functioning. PedsQL^{TM} contains 21–23 questions depending on age, generating a mean score from 0 (worst) to 100 (best). A mean score of 83 has been seen in healthy individuals and 77 in chronically ill individuals [85].

Statistical analyses

In group comparisons for dichotomous data, Fisher's exact test was used while the Mann-Whitney U-test was used for continuous parameters. Continuous data were not distributed normally and were therefore presented as median (min–max), and categoric data as absolute numbers and percentages, *n* (%) [93].

In Paper I, to evaluate risk factors for wound dehiscence, multivariate logistic regression analysis was used and presented as odds ratios (OR) with a 95% confidence interval (CI) [93].

In Paper III, to evaluate diagnostic accuracy of preoperative symptoms and examination modalities regarding fistula presence and location, contingency tables of true outcome (final ARM-subtype classification during PSARP) and findings of symptoms and examinations were devised. Diagnostic accuracy (%) was calculated by (true positive + true negative cases) / all evaluated cases.

To compare diagnostic ability regarding fistula presence, a receiver-operating characteristic (ROC) curve analysis was used, with calculation of the area under the curve (AUC) and its 95% CI [93].

In Paper IV, to evaluate linear correlations between the single highest (worst) POSAS-scored scar per participant, age, BFS and PedsQL^{TM} score, Pearson's correlation analysis was used [93].

A p-value < 0.05 was considered to be statistically significant.

Qualitative content analysis

In Paper II, transcribed data from focus group discussions were analysed through qualitative content analysis. The analysis was made on both a manifest level; of what was literally described and said by participants, and on a latent level; the underlying meaning perceived by analysing researchers [94,95].

The content analysis followed standardised steps, as shown in **Figure 11**.

The analysing process involved continuous comparison with raw data of the transcribed text, discussion and categorisation adjustments before analysing researchers could agree on the final result.

The analysing process was exemplified in the audit trail, where concrete examples of meaning units, condensed meaning units, sub-categories and categories were shown.

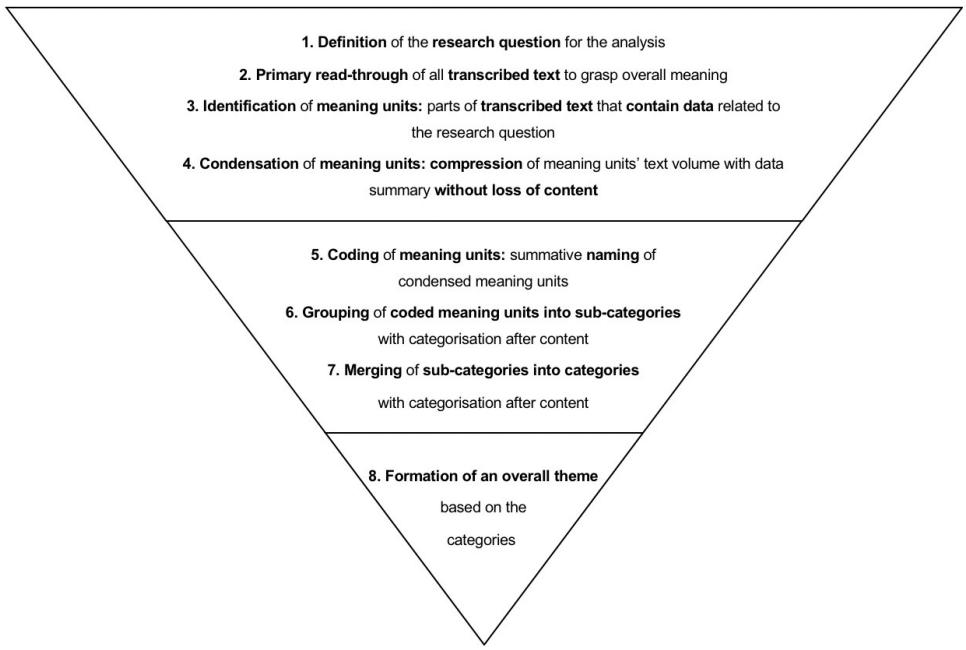


Figure 11. Qualitative content analysis.

Trustworthiness in qualitative research

Standard qualitative research criteria for reaching trustworthiness [96] were applied and ensured in Paper II by:

1) Credibility:
    - Data collection from multiple heterogeneous focus groups;
    - Request of participants' immediate oral feed-back and clarification during focus group discussions and their written anonymous feed-back afterwards;
    - Involvement of several researchers in the analysing process.
2) Transferability:
    - Thorough description of the strategic sampling method;
    - Thorough description of the results.
3) Dependability:
    - Constant comparison and connection to raw data (transcribed text) during analysis.
4) Confirmability:
    - Continuous discussion between researchers during data collection and analysis;
    - Transparent description of the analysis steps in the audit trail.

Ethical considerations

All studies of this thesis were carried out in accordance with relevant guidelines and regulations [97,98].

Paper I

The study was approved by the Regional Ethics Review Board, Southern Region, Sweden (DNR 2017/191) with waiver of informed consent.

Paper II

The study was approved by the Regional Ethics Review Boards at involved institutions, Sweden and Norway (DNR 2017/867, DNR 2017/1895).

Eligible patients received written study information. Participants received additional oral information and gave written informed consent.

Participants were informed of the possibility of withdrawal from the study at any time without stated cause or impact on future healthcare. Participants were offered optional individual counselling outside the study.

Paper III

The study was approved by the Regional Ethics Review Board, Southern Region, Sweden (DNR 2017/191) with waiver of informed consent.

Paper IV

The study was approved by the National Ethics Review Authority in Sweden (DNR 2020-00529).

Eligible patients received oral and written study information. Participants, or minors' guardians, gave written informed consent. Participants, or minors' guardians, whose scar photos were selected for anonymous publication, gave written publication consent.

Participants, or minors' guardians, were informed of the possibility of withdrawal from the study at any time without stated cause or impact on future healthcare. Participants were offered optional individual counselling outside the study.

Results

Paper I

Ninety neonates (41% female) were included (**Table 3**).

Table 3. Patients born with ARM and reconstructed through PSARP, limited PSARP or PSARVUP.

	All patients	Three-step procedure	Single-stage procedure	*p*-value*
Total	90 (100)	50 (56)	40 (44)	
Gender female	37 (41)	19 (38)	18 (45)	0.53
ARM subtype				
Perineal (M/F)	24 (45) / 17 (46)	3 (6)	38 (94)	
Vestibular	15 (41% of F)	14 (28)	1 (3)	
Recto-urethral	20 (38% of M)	19 (38)	1 (3)	
Recto- bladder neck	4 (8% of M)	4 (8)	0	
Cloaca	4 (11% of F)	4 (8)	0	
No fistula (M/F)	5 (9) / 1 (2)	6 (12)	0	
Prematurity[a]	15 (19)	11 (24)	4 (13)	0.25
SGA[b]	23 (29)	16 (33)	7 (22)	0.32
Concomitant malformations				
Total (at least one)	63 (70)	44 (88)	19 (48)	*< 0.001*
Cardiac[c]	28 (33)	18 (38)	10 (36)	0.48
Urinary tract[d]	25 (29)	20 (42)	5 (14)	*0.008*
Other skeletal	19 (21)	14 (28)	5 (13)	0.12
Vertebral[e]	30 (50)	25 (64)	5 (24)	*0.003*
Spinal cord[f]	14 (30)	10 (29)	4 (31)	0.93
Genital	17 (19)	14 (28)	3 (8)	*0.015*
GI-tract	12 (13)	9 (18)	3 (8)	0.21
VACTERL	24 (27)	19 (38)	5 (13)	*0.008*
Concomitant syndromes				
Total	15 (17)	10 (20)	5 (13)	0.40
Trisomy 21	6 (7)	5 (10)	1 (3)	0.22
Antibiotic prophylaxis > 1 day	33 (37)	19 (38)	14 (35)	0.83

Values presented as the absolute number and percentage of patients, *n* (%); Number of patients with available data: [a] = 77, [b] = 80, [c] = 84, [d] = 85, [e] = 60, [f] = 47; ARM: anorectal malformations, PSARP: posterior sagittal anorectoplasty, PSARVUP: posterior sagittal anorectal vaginal urethroplasty; M: male, F: female; Prematurity: GW< 37; SGA: small for gestational age; GI-tract: gastro-intestinal tract; VACTERL: ≥ 3 of concomitant vertebral-, anorectal-, cardiac-, tracheal-, oesophageal-, renal- or limb malformations; *Fisher's exact two-tailed test.

Fifty patients (56%) had a three-step reconstructive procedure with a diverted colostomy. These patients had more complicated ARM subtypes and more frequently associated malformations, particularly of the urogenital tract and vertebrae. There was no difference regarding prolonged antibiotic prophylaxis (> 1 day) between the single-stage and three-step procedure groups

Wound dehiscence occurred in 28 patients (31%), with a superficial dehiscence (only skin) in 19 patients (21%) and a deep dehiscence in nine patients (10%). There was a statistically significant higher frequency of wound dehiscence in the single-stage procedure group compared to the three-step procedure group; 17 patients (17/40 43%) versus 11 patients (11/50 22%), *p*-value=0.04.

Having a colostomy was the only statistically significant proved difference between the wound dehiscence- and the non-wound dehiscence group (**Table 4**).

Further multivariate analysis of the single-stage procedure group could not identify any risk factor for wound dehiscence (**Table 5**).

Table 4. Wound dehiscence in 90 patients born with ARM and reconstructed through PSARP, limited PSARP or PSARVUP.

	Wound dehiscence	No wound dehiscence	*p*-value*
Total	28	62	
Gender female	11 (39)	26 (42)	1
SGA[a]	7 (28)	16 (29)	0.92
Cardiac malformation[b]	9 (33)	19 (33)	1
VACTERL	6 (21)	18 (29)	0.61
Weight at surgery[c] (g)	3800 (2200–9400)	5000 (1600–14000)	0.33**
Colostomy	11 (39)	39 (63)	*0.04*

Values presented as the absolute number and percentage of patients, *n* (%), and as median (min–max); Number of patients with available data: [a] = 80, [b] = 84, [c] = 62; ARM: anorectal malformations, PSARP: posterior sagittal anorectoplasty, PSARVUP: posterior sagittal anorectal vaginal urethroplasty; SGA: small for gestational age; VACTERL: ≥ 3 of concomitant vertebral-, anorectal-, cardiac-, tracheal-, oesophageal-, renal- or limb malformations; *Fisher's exact two-tailed test, **Mann-Whitney U-test, two tailed.

Table 5. Multivariate logistic regression analysis of risk factors for wound dehiscence in 40 patients reconstructed with single-staged PSARP or limited PSARP.

	Single-stage procedure	Wound dehiscence	OR (95% CI)	*p*-value
Total	40	17 (43)		
Gender female	18 (45)	6 (35)	0.50 (0.14–1.81)	0.35
Cardiac malformation	10 (25)	7 (41)	3.73 (0.78–17.88)	0.07
Low weight at surgery (< 3500 g)	15 (38)	5 (29)	1.56 (0.36–6.69)	0.72
Antibiotics < 1 day	26 (65)	10 (59)	1.60 (0.43–5.94)	0.52
Fasting 0–3 days	34 (85)	16 (94)	4.44 (0.47–42.18)	0.22

Values presented as the absolute number and percentage of patients, *n* (%); PSARP: posterior sagittal anorectal plasty; OR: odds ratio, CI: confidence interval.

Paper II

Sixteen participants (63% female), with a median age of 24 (19–47) years, were included in a total of four gender-divided focus groups, with one female (n = 5) and one male (n = 3) focus group at each centre. Focus group discussions lasted between 60–120 min each.

The overall theme that emerged from the content analysis of the focus group discussions regarding participants' needs and expectations of transitional- and adult healthcare, was their desired feeling of normality. The formed preceding sub-categories and categories of the content analysis, and the identified key components of successful transition for adolescents born with ARM, are displayed in **Figure 12**.

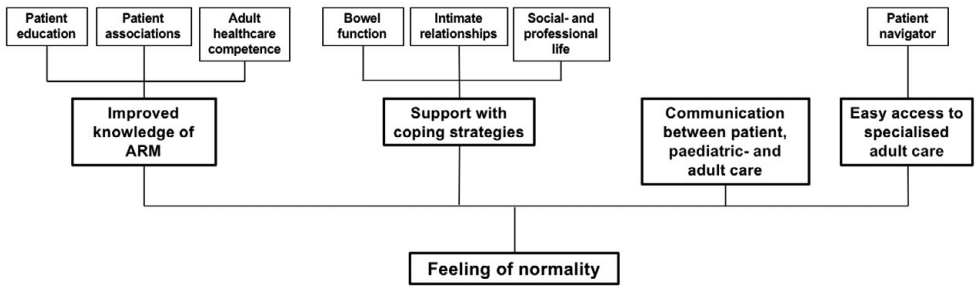


Figure 12. Key components of successful transition for adolescents born with anrectal malformations.

Examples of meaning units (quotes from the transcriptions) and condensed meaning units leading up to this categorisation are displayed in the audit trail (**Table 6**).

Participants expressed a need for a pedagogical and age-adapted patient education from an early age at the paediatric surgery department to strengthen patients' condition literacy as a way to self-empowerment.

Many participants seemed to have, over time as young adults, accepted and adapted to their bowel- and bladder symptoms affecting their everyday life. They emphasised the importance of support with coping strategies to manage a chronic condition. They suggested peer-to-peer mentoring; younger patients matched with older patients, and strengthening of patient associations for support and information.

Participants described a general low level of knowledge regarding ARM among adult healthcare providers. Participants described a powerlessness in trying to obtain adequate help as adolescents and adults. They had failed many times due to adult healthcare's perceived incompetence and ignorance, and also elusive organisational structures making it impossible to know where or whom to turn to.

Participants identified a non-existing communication between paediatric- and adult healthcare, preventing adequate transition. They wanted a structured, individually

adapted transition with no fixed transitional age, led by the responsible physicians from both sides, passing their clinical information on to adult care. Participants wanted medical help 'on demand' as adults through an appointed patient navigator, facilitating contact with adult care providers. Participants recommended the pelvic floor centres to be transitional referral centres of choice.

Table 6. Audit trail: examples from the content analysis process regarding participants' needs and expectations of transitional- and adult healthcare.

Meaning unit (quotes from the transcription)	**Condensed meaning unit**	**Sub-category**	**Category**
'You know that something's wrong but you don't know what it is.'	Participants lacked knowledge of their condition	Patient education	Improved knowledge of ARM
'It's great to be at such meetings [patient association gatherings]. You meet people with the same challenges as yourself.'	Patient associations were valuable sources of support and information	Patient associations	Improved knowledge of ARM
'You are at the hospital, and you try to explain: "I have anal atresia", and they have no idea what it is … somewhere along the way I've just given up.'	Adult healthcare lacked knowledge of the condition	Adult healthcare competence	Improved knowledge of ARM
'You constantly ask yourself: Where is the nearest bathroom?'	Need for a restroom close by restricted and challenged daily life	Bowel function	Support with coping strategies
'It has affected my intimate life. It took years before I dared involve myself intimately.'	The condition affected intimate relationships negatively	Intimate relationships	Support with coping strategies
'You can always give your employer another explanation, maybe say that my back is better if I stand up working.'	The condition affected and challenged working life	Social- and professional life	Support with coping strategies
'You have to conduct this [transitional] meeting when the person is ready for it … I think you should have this talk later at the pelvic floor centre. The paediatric surgeon could come there.'	Transitional meetings with paediatric- and adult care givers were warranted		Communication between patient, paediatric- and adult care
'It should be some kind of a coordinator who knows what it is about.'	Coordinating nurses were warranted for facilitating contact	Patient navigator	Easy access to specialised adult care

Paper III

Thirty-eight male neonates were included. Thirty-one (82%) had a recto-urinary tract fistula (recto-bulbar fistula *n* = 8, recto-prostatic fistula *n* = 17, and recto-bladder neck fistula *n* = 6) and seven (18%) had no fistula at final intraoperative classification during PSARP.

Preoperative diagnostic accuracy of fistula *presence* reached a maximum of 92% through ostomy endoscopy (**Table 7**).

Table 7. Diagnostic accuracy of pre- and perioperative examinations for fistula presence in male neonates born with ARM with final classification during PSARP, *n* = numbers, (%) = percent.

	Fistulae* *n* = 31	**No fistula** *n* = 7	**Diagnosic accuracy****
Symptoms	14 (45)	0	21/38 (55)
Stool-coloured urine	8 (26)	-	
UTI	5 (16)	-	
Urine in colostomy	5 (16)	-	
VCUG	29	7	23/36 (64)
Visible fistula	16 (55)	0	
Colostogram	28	7	25/35 (71)
Visible fistula	18 (64)	0	
Cystoscopy	19	1	14/20 (70)
Visible fistula	13 (68)	0	
Ostomy endoscopy	19	5	22/24 (92)
Visible fistula	17 (89)	0	
Guide wire used	11 (58)	-	

Values presented as the absolute number and percentage of patients, *n* (%); ARM: anorectal malformations, PSARP: posterior sagittal anorectoplasty, UTI: urinary tract infection, VCUG: voiding cystourethrogram; *Recto-bulbar fistula *n* = 8, recto-prostatic fistula *n* = 17, and recto-bladder neck fistula *n* = 6; **Diagnostic accuracy (%) = (true positive cases + true negative cases) / all evaluated cases.

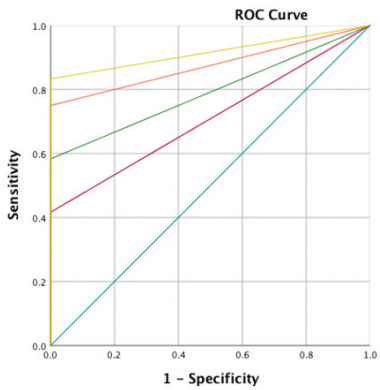
Preoperative diagnostic accuracy of fistula *location* reached a maximum of 70% through cystoscopy, closely followed by high-pressure colostogram (66%) (**Table 8**).

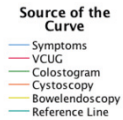
Diagnostic ability did not differ statistically between modalities (**Figure 13**).

Table 8. Diagnostic accuracy of pre- and perioperative examinations for fistula location in male neonates born with ARM with final classification during PSARP, *n* = numbers, (%) = percent.

	Fistulae *n* = 31			No fistula *n* = 7	Diagnosic accuracy*
	Recto-bulbar *n* = 8	Recto-prostatic *n* = 17	Bladder neck *n* = 6		
VCUG	8	15	6	7	21/36 (58)
Correct fistula location	5 (63)	8 (53)	1 (17)	7 (100)	
Colostogram	8	15	5	7	23/35 (66)
Correct fistula location	5 (63)	8 (53)	3 (60)	7 (100)	
Cystoscopy	4	12	3	1	14/20 (70)
Correct fistula location	2 (50)	9 (75)	2 (67)	1 (100)	

Values presented as the absolute number and percentage of patients, *n* (%); ARM: anorectal malformations, PSARP: posterior sagittal anorectoplasty, VCUG: voiding cystourethrogram; *Diagnostic accuracy (%) = (true positive cases + true negative cases) / all evaluated cases.





Diagnostic modalities	AUC (95% CI)	P value
Bowel endoscopy	0.92 (0.74–1.00)	0.18
Cystoscopy	0.88 (0.64–1.00)	0.23
Colostogram	0.79 (0.47–1.00)	0.35
VCUG	0.71 (0.30–1.00)	0.50
Symptoms	0.71 (0.30–1.00)	0.50

AUC: Area under the curve, CI: Confidence interval; Null hypothesis: true area=0.5

Figure 13. Comparison of diagnostic ability of pre- and perioperative examinations of fistula presence in male neonates born with anorectal malformations with final classification during posterior sagittal anorectoplasty.

Paper IV

Twenty-seven participants (48% female), with a median age of 12 (5–24) years, were included. Participants had a median of 2 (1–8) scars after previous abdominal surgeries.

Six participants (22%) reported recurrent scar pain and five (19%) scar pruritus. The median POSAS score for the highest (worst) scored scar was 47 (18–78). Scars after previous colostomies and associated laparotomies scored the highest. There was a correlation between higher POSAS score and increasing age (**Figure 14**).

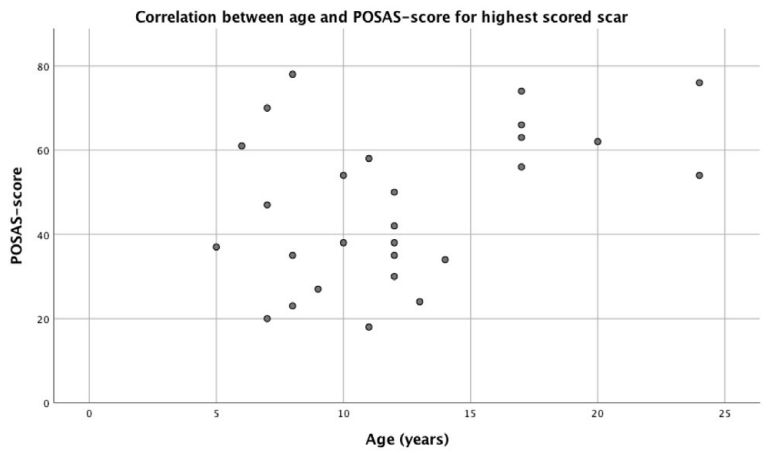


Figure 14. Correlation between age and POSAS score for highest (worst) scored scar (11=as normal skin, 110=as worst scar imaginable). Pearson's *r*=0.40 *p*-value 0.04.

Scar photographs including corresponding POSAS scores show different aesthetic outcomes in **Figures 15–17**.

Nine participants (33%) reported altered behaviour due to abdominal scarring, e.g. always wearing full-covering clothes. Twelve participants (44%) were proud of their scars. Five participants (19%) regarded their scars with dismay and discomfort.

Eight participants (38% of females and 21% of males), all 12 years old or older, requested scar treatment, mainly for aesthetic reasons.

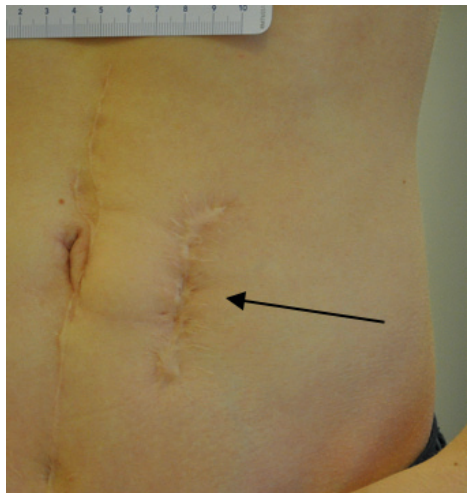


Figure 15. A broad secondary-healed and partially depressed abdominal scar after a left paramedian incision with a former divided sigmoidostomy (arrow) in a male participant, here 16 years old, with POSAS score of 74.

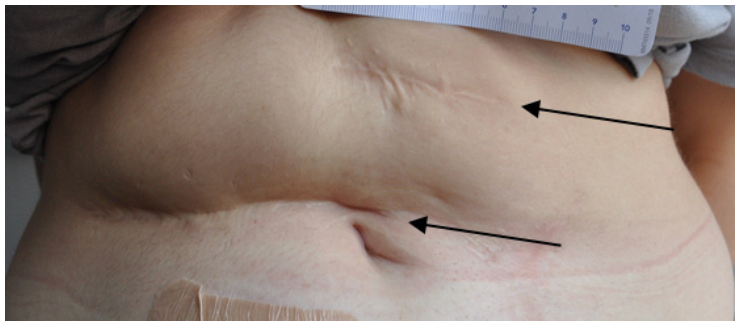


Figure 16. Depressed and partially secondary-healed abdominal scars after an upper right transverse incision (lower arrow) and a former transversostomy (upper arrow) in a female participant, here 23 years old, POSAS scores of 76 (lower arrow) and 74 (upper arrow).

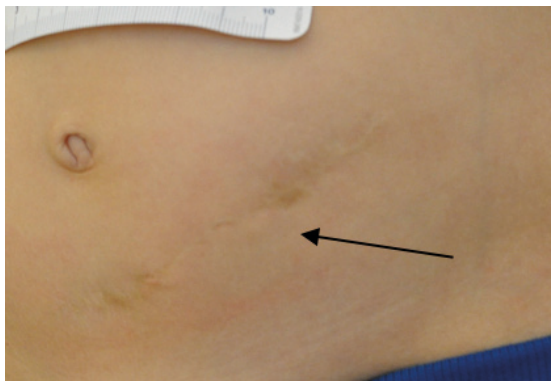


Figure 17. A flat and thin abdominal scar after a lower left transverse incision with a former divided sigmoidostomy (arrow) in male participant, here 8 years old, with POSAS score of 35.

No statistically significant gender differences were identified between POSAS-, BFS- and PedsQL^{TM}-scores or scar treatment requests (**Table 9**).

Table 9. Gender comparisons regarding POSAS- (11=best, 110=worst), BFS- (1=worst, 20=best), and PedsQL^{TM}-scores (0=worst, 100=best) and scar treatment requests by participants.

	Females *n* = 13	**Males** *n* = 14	***p*-value***
POSAS score of highest (worst) scored scar	50 (18–78)	38 (20–74)	0.42
BFS score	13 (9–20)	14 (8–20)	0.49
PedsQL^{TM} score	79 (52–96)	82 (52–99)	0.53
Scar treatment request	5 (38)	8 (57)	0.45**

Values presented as the absolute number and percentage *n* (%), and as median (min–max); POSAS: Patient and Observer Scar Assessment Scale; BFS: Bowel Function Score; PedsQL^{TM}: Pediatric Quality of Life Inventory^{TM}; *Mann-Whitney U-test, two tailed; **Fisher's exact two-tailed test.

Scars in 21 participants (78%), including all eight participants requesting scar treatment, were technically suited for plastic surgical scar correction for aesthetic reasons according to the plastic surgeon's pictorial assessment. Postsurgical scar pain relief could not be guaranteed.

In five presumptive cases (24%) the plastic surgeon requested co-surgery with a paediatric surgeon because of possible interference with the bowel or bladder during abdominal wall dissection.

Surgical scar correction in children under the age of 7 years was not considered favourable due to expected body growth and scar maturation.

Discussion

The following new aspects, with clinical relevance in promoting high-quality care in patients with ARM, are presented in this thesis:

1. Colostomy seems to protect against, but does not eliminate the risk of, wound dehiscence after PSARP, the pathogenetic mechanisms of which remain unclear.
2. Enhanced early patient education to promote a desired condition literacy is needed, according to adult ARM patients.
3. Low level of knowledge of ARM among adult healthcare personnel and a non-reachable adult healthcare system seem to obstruct adequate transitional care, according to adult ARM patients.
4. Standard fistula diagnostic modalities only reached a maximum of 70% diagnostic accuracy in our series regarding correct preoperative ARM subtyping.
5. Postoperative abdominal scarring may generate scar-related morbidity, e.g. pain and poor aesthetic outcome, and should be inquired about in the clinical follow-up of ARM patients.
6. Scar-corrective plastic surgery should be considered in selected ARM patients.

What promotes uncomplicated wound healing after PSARP?

The common notion [24,26] that a colostomy protects against wound dehiscence after PSARP was confirmed in our study but, according to our results, a colostomy does not eliminate the risk of wound dehiscence. Our hypothesis that low weight at surgery or concomitant cardiac malformation would influence wound healing was not confirmed, which might, of course, relate to the retrospective study design with established quite large data fall-out regarding weight at surgery and with potential observational bias, plus our small study sample with secondary low power to reach any statistical significances.

Our study leaves us with one major question of clinical importance unanswered: what are the contributing factors to uncomplicated wound healing after PSARP?

These factors have still not been clarified, although different theories have been postulated mainly regarding associated bacterial wound infection and the colostomy's role in reducing perineal bacterial contamination [56]. One study has investigated the possible pathogenetic role of perioperative perineal tissue tension with subsequent reduction of tissue perfusion [55]. One case-control study has shown that vacuum-assisted closure (VAC) seems to reduce the risk of wound dehiscence after PSARP by increasing perineal tissue perfusion and reducing wound oedema [57]. Further studies are needed in this field.

The practical, clinical consequence in our department of our study on wound dehiscence is that our awareness of this complication has been raised among all staff members. It has been shown previously that an important factor of reducing post-surgical morbidity and mortality is the whole team's ability to recognise a patient's deviation from the standard postoperative course early on and to take appropriate rescuing countermeasures [99]. Our care after PSARP includes regular, meticulous wound inspection and wound cleaning and other continuous clinical assessments regarding, for example, observations of increasing pain or body temperature, and to be prepared to perform bacterial cultivation and change antibiotics or prolong fasting. We also have established standard homogenous regimens of postoperative antibiotic prophylaxis, fasting and wound management for both single-stage and three-step procedures. We believe that we can see that our wound dehiscence rates are decreasing, but that statement must, of course, be proved in a future clinical study.

Condition literacy among patients

Patient education and improved condition literacy as a way to self-empowerment in chronic conditions are previous identified important aspects of transition from paediatric to adult care [64,76,78,100–103] and were confirmed in our focus group study. The results indicated a need for an increased pedagogical effort from the paediatric surgery department regarding early, age-adapted patient information and education in ARM. Participants presented several suggestions for improving patient education, i.e. the development of hospitals' digital information sites to modern standards according to patients' and their families' needs in all stages of life, facilitation of patient group meetings, peer-to-peer mentoring and formation of patient associations.

In our department we have started to implement these excellent improvement ideas by updating written patient- and parent-information materials, organising patient family gatherings and webinars, and continuing in engaging in different patient association meetings as lecturers and professional partners, which have been held online since the Covid pandemic. One difference between Sweden and Norway is that Norway has a specific patient association for ARM which Sweden does not currently have. If an official ARM patient association was to be founded in Sweden,

as desired by Swedish participants in our study, we would, as a national highly specialised ARM care centre, gratefully gain a powerful ally in promoting high-quality care in ARM patients.

In addition, I am now trying in every consultation with a young person born with ARM to include the child attentively and discuss these important issues of condition literacy in an age-adapted way, as our study participants advocated, and shift focus away from the parents and their needs. Furthermore, following our study participants' suggestions as well as recommendations in the literature, my goal in consultations with teenagers is that the parent remains a silent, supportive bystander throughout the consultation, and that parts of the consultation take place without the parent in the room. This enables the adolescent to hopefully express their views, feel able to talk in confidence about sensitive issues, such as sexual function and intimate relationships, take control over their condition and practise personal responsibility towards their upcoming transition to adult care [74].

Structural differences between paediatric- and adult healthcare – transitional care's biggest challenge?

The general lack of experienced knowledge of ARM among adult healthcare workers and a non-reachable adult healthcare system was a major finding in our focus group study and considered by the adult participants to be a significant influencing factor of poor transition to adult care. Participants had been told to seek primary care for their bowel and bladder problems or had been forced to attend the emergency care department for their worsened, chronic symptoms. Understandably, according to the participants, both primary- and emergency care lacked competence and resources for specialised ARM care. In a rare and often complex condition, such as ARM, one could argue that the patient should be transferred from a specialised, high-volume paediatric centre to a corresponding specialised, high-volume adult one [74]. Adult ARM care should not be the primary- or emergency care's responsibility according to our results. Participants instead advocated adult specialised pelvic floor centres to be transitional referral centres of choice, having the necessary surgical- urological- and gynaecological competence and care under one organisational umbrella.

Every process, including the transitional care process, must have a leader taking charge and will move things forward. Regarding transitional care in ARM, and other congenital paediatric surgical conditions, this leader must be the responsible paediatric surgeon as pointed out in previous studies as well as our own [64]. But, every sender needs a defined receiver. For us at the Department of Paediatric Surgery in Lund, our natural adult transitional care partner for many years for patients with complex ARM has been the Pelvic Floor Centre at the Department of Surgery in Malmö. This cooperation is built on, and demands, personal contact

amongst staff members enabling multilevel, bilateral competence transmission. Its realisation is challenged by shortage of resources and the necessary concurrent responsibilities of the Pelvic Floor Centre. Every paediatric surgery department treating patients with ARM must develop their own, site-adapted, transitional system. This work must progress continuously, according to our results, to promote and defend adult high-quality care in ARM.

Improvement of preoperative fistula diagnostics

Our study revealed an individual diagnostic accuracy of a maximum 70% regarding correct preoperative radiological- or endoscopical fistula localisation, which is in line with previous studies [31,35–39]. The results indicate that preoperative fistula diagnostics and malformation imaging require technical development and refinement to improve diagnostic accuracy and provide the paediatric surgeon with a more precise and adequate pre-PSARP surgical work-up in order to reduce the risk of perioperative iatrogenic injury [30].

These necessary diagnostic improvements could happen on multiple levels. Firstly, further diagnostic sharpening of current modalities at paediatric radiology departments is already ongoing. Paediatric radiologists in our department report a synergetic effect when performing high-pressure colostogram and VCUG simultaneously that seems to improve fistula diagnostic accuracy. This technique has only been practised in our department for a couple of years and was, therefore, not analysed specifically in our present study.

Secondly, further development of upcoming radiological modalities and techniques such as high-frequency ultrasonography, high-resolution MRI and 3D printing of individual anatomic models are anticipated. Both ultrasonography and MRI have the advantage of no radiation, with a current reported fistula localisation diagnostic accuracy of 61–84%, and the potential to offer high-resolution complete malformation imaging [40,41]. Apparent disadvantages include ultrasonography being operator-dependent and MRI requiring general anaesthesia.

Scar morbidity – the surgeon who did it should fix it!

Abdominal scarring might contribute to postoperative physical- and psychosocial morbidity in ARM according to our results, where moderate-to-severe scar-related symptoms, such as recurrent scar pain and poor visual outcome, were present in 29% of participants. Scar-related morbidity has been acknowledged previously in other comparable patient groups [59,62,63]. Scar symptom severity and increasing age correlated in our series, which might be a result of a change in surgical ostomy placement technique over time at our department, and/or the maturation of the

patient's body with more subcutaneous fat tissue, and/or increasing self-awareness as the patient ages.

In our study, the plastic surgeon declared the vast majority of postoperative scars to be eligible for corrective plastic surgery due to poor aesthetic outcome, if the patient themself expressed a wish for that treatment to occur. Thirty percent of study participants expressed such treatment wishes.

Therefore, symptom evaluation of postoperative scarring should preferably be included in ARM follow-up programmes with subsequent referral for plastic surgical evaluation if needed. We believe that a similar scar follow-up is applicable for other non-ARM patient groups where surgery is performed in childhood.

Additionally, it is important to remember that the most efficient way of preventing scar-related morbidity is to avoid scar formation in the first place. Surgeons should therefore avoid unnecessary incisions that are too wide and remember that scars grow proportionally with the child [58,59]. A laparoscopic approach offers an advantage in reducing incision length compared to a laparotomy [45]. Surgeons should also try to respect Langer lines and practice modern incision closure techniques, preferably intracutaneous suturing in infants, and modern postoperative wound management to promote favourable surgical wound healing [22,60,61]. Some study participants exhibited, for example, less aesthetic favourable dot scarring lateral of the linear incision scar after a transcutaneous single suture closure technique that might have had a better visual outcome if intracutaneous sutures had been used instead.

Challenges in ARM research

Research in rare diseases, such as ARM, share many study design challenges [104]. Foremost, it is difficult, or at least it takes a very long time, for almost any centre to include enough study participants to achieve statistical power enabling robust study conclusions. Adult study participants in Paper II expressed wishes for further research in long-term outcome regarding sexual function and fertility in ARM patients, and genetics, to understand the genesis of ARM, all of which demand large sample sizes in order to be able to draw statistically significant conclusions. The need of larger multi-center trials was also pointed out in a recent systematic literature review regarding long-term functional urinary and sexual outcomes in patients with ARM [105].

The European ARM-net consortium [106] is an example of an international association where, since 2010, researchers have joined forces to perform larger, multi-national clinical-, epidemiological- and genetic studies in patients born with ARM. The Department of Paediatric Surgery in Lund is an ARM-net member and contributor to this group.

However, despite that, it is still possible to produce clinically significant research on small sample sizes, as long as the research question is valid, sampling criteria translucent, outcome variables that are well-defined and conclusions sound, based on proper study design and statistical methods [104]. Many clinically important research questions, often with the words 'why?' or 'how?' in them, demand a qualitative research approach, where credibility does not depend on sample size [96].

One specific challenge in ARM research is the heterogenous nature of ARM: patients are a group of many different subtypes, all with reported different outcomes depending on subtype complexity and associated malformations [27–29,66–68]. It is important to not start comparing particular ARM patients with others with the condition, or draw general conclusions based on selection bias. One aspect might perhaps be most relevant and true for people born with one particular subtype, while being completely irrelevant for others. However, if one sample subtype is selected, it is even more difficult and challenging to include enough participants…

Study limitations

Apart from general small sample sizes hampering statistical power with the risk of a type II error in Papers I, III and IV, both studies I and III suffered from the retrospective design with quite a large proportion of missing data and risk of observational bias with incorrect data.

Paper III could, in addition, have been strengthened with a secondary review of X-ray reports. Paper IV suffered from a low inclusion rate of invited, eligible study participants, possibly explained by a time-consuming study design and lack of financial compensation, imposing a risk of selection bias with a possibly skewed group of participants.

Paper IV could have been strengthened if several plastic surgeons had performed the scar treatment assessments independently.

Conclusions

Paper I

Wound dehiscence is a common complication after PSARP, in this study occurring in 31% of all procedures, including 22% of three-step procedures with a divided colostomy and 43% of single-stage procedures. A divided colostomy was the only identified protective factor for wound dehiscence. No risk factor for wound dehiscence in single-stage PSARP was identified.

Paper II

Adults born with ARM identified improved knowledge of ARM (among patients as well as adult healthcare personnel), support and strengthening of coping strategies, structured communication between patient, paediatric- and adult care providers, and easy access to specialised adult care facilitated by appointed patient navigators as key elements of adequate transitional care.

Paper III

Cystoscopy and high-pressure colostogram had the highest diagnostic accuracy of preoperative fistula localisation in ARM, reaching a maximum of 70% regarding correct preoperative ARM subtyping.

Paper IV

Postoperative abdominal scarring in ARM patients after surgery performed in childhood may result in scar-related morbidity in terms of pain, pruritus and less favourable aesthetic outcome with effects on social behaviour. Postoperative scarring should, therefore, require attention in clinical follow-up programmes of ARM patients, including potential corrective plastic surgery in selected cases.

Populärvetenskaplig sammanfattning

Varje år föds ca 110 000 barn i Sverige, och 40–50 av dem föds helt utan ändtarmsöppning eller har öppningen på fel plats. Tillståndet kallas anorektala missbildningar (ARM) eller analatresi. Denna avhandling bygger på fyra olika studier om ARM. Studierna handlar om hur vården för personer födda med ARM kan förbättras.

Vi tar avstamp hos det lilla spädbarnet som föds utan ändtarmsöppning och tittar på hur man bättre kan förbereda, genomföra och följa upp barnets kirurgiska behandling. Vi tar även del av gruppdiskussioner mellan vuxna personer födda med ARM. De pratar om hur det är att växa upp och leva sitt fortsatta liv med en medfödd missbildning. De ger oss även riktlinjer för hur en bra övergång mellan barn- och vuxensjukvård skulle kunna organiseras.

Vad är ARM?

ARM innebär att barnet antingen föds helt utan ändtarmsöppning eller att en liten öppning sitter på fel plats i underlivet. Ofta kan barnet även ha missbildningar i hjärtat, ryggkotorna, ryggmärgen, njurarna eller könsorganen. Mindre ofta kan barnet även ha missbildningar i mat- och luftstrupen, andra delar av tarmkanalen samt i armarnas och benens skelettstrukturer. Barnet kan även födas med ett syndrom, t.ex. trisomi 21 (Downs syndrom).

Vi vet ännu inte varför barnet får ARM eller de andra missbildningarna, men vi tror att svaret ligger i barnets gener, att något hände när barnet blev till. Det är ingen förälders fel att man får ett barn med ARM. Missbildningen syns inte på ultraljud före födseln. Det är vanligare att pojkar föds med ARM än flickor, varför vet vi inte. Det är mycket ovanligt i Sverige att barn dör p.g.a. ARM eftersom tillståndet kan behandlas.

Kirurgisk behandling vid ARM

Ett barn som föds utan synlig ändtarmsöppning är livshotande sjukt. Avföringen behöver ju komma ut! Därför måste det nyfödda barnet flyttas till en barnkirurgisk klinik för akut operation. Barnet får då en tillfällig tarmstomi, då tjocktarmen öppnas och sys ut på magen, så att avföringen kan komma ut i en liten plastpåse som kan

bytas på magen. Efter stomioperationen kan barnet börja äta och må bra inför kommande missbildningsoperation av ändtarmen.

Om barnet föds med en liten öppning på fel plats, behöver barnet ofta ingen tarmstomi men däremot regelbundna lavemang för att kunna tömma tarmen tillräckligt bra och må bra inför kommande missbildningsoperation.

I Sverige opereras barn födda med ARM sedan 2018 endast på två barnkirurgiska kliniker; vid Skånes Universitetssjukhus i Lund och vid Karolinska Universitetssjukhuset i Solna.

Barnet genomgår en grundlig utredning med olika röntgen- och ultraljudsundersökningar där man letar efter andra missbildningar och försöker bilda sig en uppfattning om hur den anorektala missbildningen ser ut inför kommande missbildningsoperation.

Missbildningsoperationen, när barnet får en ändtarmsöppning på rätt plats i centrum av ringmuskeln, kallas posterior sagittal anorektal plastik, förkortat PSARP. PSARP-operationen görs inom ett par månader efter barnets födelse. När detta operationsområde i underlivet vid den nya ändtarmsöppningen har läkt opererar man ned en eventuell tarmstomi för att koppla ihop tarmsystemet igen. Barnet har då avföring genom sin nya ändtarmsöppning.

Trots PSARP-operationen kan barnet, ibland genom hela livet, behöva mer eller mindre hjälp med avföringen, t.ex. med lavemang och mediciner mot hård avföring, och ibland även hjälp med urintömningen. Detta beror på att ARM, och troligtvis även PSARP-operationen i sig, ofta påverkar hela bäckenområdet och nerverna från ryggslutet till tarmen och blåsan som styr tömningen och känsligheten. Den som är född med ARM behöver därför en överlämning från barn- till vuxensjukvården för att kunna leva ett så bra och normalt liv som möjligt.

Första studien

Här ville vi titta närmare på såruppsprickning, en vanlig komplikation till PSARP-operationen. Det innebär att stygnen i underlivet släpper och såret glipar. Man vet inte varför detta händer ibland, men misstänker att bakterier eller nedsatt genomblödning i såret skulle kunna vara orsaken.

Vi kontrollerade i patientjournalerna hur många PSARP-opererade barn i Lund 2001–2016 som hade råkat ut för såruppsprickning och om vi kunde hitta någon ytterligare riskfaktor för detta, t.ex. lägre vikt vid PSARP-operationen eller samtidig hjärtmissbildning.

Studien på totalt 90 barn, varav 37 flickor och 53 pojkar, visade att såruppsprickning hade skett hos 28 barn (31%). De 50 barn som hade en tarmstomi, d.v.s. inte hade avföring direkt vid sin nya ändtarmsöppning, skyddades något men inte helt mot

såruppsprickning efter PSARP-operationen. Vi kunde inte hitta någon annan skyddande faktor eller någon riskfaktor.

Andra studien

Då tarm- och urinproblem av ARM kan förekomma genom hela livet behöver en del tonåringar överföras från barn- till vuxensjukvården. Vuxna kan även ha problem med t.ex. sexualfunktion och fertilitet, något som en barnkirurg inte har kompetens att behandla. Tyvärr upplever många vuxna patienter att denna överföring fungerar dåligt och att de inte får hjälp för sina problem kopplade till ARM.

Därför bad vi vuxna personer födda med ARM att klargöra vad som är viktigt vid en överföring och hur den på bästa sätt bör gå till. Denna studie gjordes via ett nordiskt samarbete mellan de barnkirurgiska klinikerna i Lund och Oslo, och bäckenbottencentra för vuxna i Akershus utanför Oslo och i Malmö.

Vi bjöd in vuxna födda med ARM till så kallade fokusgrupper i Lund och Oslo, där deltagarna satt ned och samtalade med varandra och en diskussionsledare om problem och utmaningar med att leva med ARM och om övergången till vuxensjukvården. De könsuppdelade samtalen spelades in och transkriberades. Texternas innehåll analyserades därefter med en så kallad kvalitativ innehållsanalys där man grupperar, beskriver och tolkar informationen från texten.

Totalt 16 vuxna deltog: 5 kvinnor och 3 män i varsin fokusgrupp i både Sverige och Norge. Deltagarna lyfte fram ökad kunskap om ARM, både hos patienten och hos vuxenvårdgivarna, strukturerad kommunikation mellan patienten, barn- och vuxenvårdgivarna samt lättåtkomlig specialiserad vuxensjukvård, som särskilt viktiga delar för en fungerande övergångsvård. Deltagarna efterlyste tidiga, gärna digitala, utbildningsinsatser för patienterna redan på barnkirurgen, men också stödjande gruppträffar och mentorskap mellan yngre- och äldre patienter, samt att barn- och vuxenvårdgivarna tog ansvar för sin kunskapsöverföring, så att patienten slapp undervisa vuxenvården. Deltagarna ville snabbt kunna kontakta rätt vuxenvårdgivare när de hade problem och de efterlyste utsedda patientlotsar för att underlätta kontakten med vården. Deltagarna ansåg att specialiserade bäckenbottenmottagningar, i exempelvis Malmö och Akershus, med kirurger, urologer och gynekologer var bäst lämpade i vuxenvården att ta över ARM-patienter.

Tredje studien

Som barnkirurg är det väldigt viktigt att veta exakt hur missbildningen som ska opereras ser ut för att kunna planera PSARP-kirurgin och undvika onödiga överraskningar under operationen. Vi vet att topp-kirurgi behövs för att resultatet ska bli så bra som möjligt.

Därför ville vi utvärdera de nuvarande röntgen- och undersökningsmetoderna, t.ex. röntgen med kontrast i tarmen eller fiberoptikundersökningar, som vi använder för att kartlägga ARM hos pojkar inför PSARP-operationen. Flickor med ARM föds oftast med en liten synlig ändtarmsöppning på fel ställe och behöver därför inte genomgå just dessa röntgenundersökningar.

Vi kontrollerade i patientjournalerna hur ’rätt’ de olika undersökningsmetoderna hade jämfört med det ’facit’ vi fick under PSARP-operationerna av pojkar födda med ARM 2001–2020 och om någon metod var bättre än någon annan.

Studien på 38 pojkar visade att undersökningsmetoderna hade rätt i ca 70% av fallen. Ingen av metoderna var statistiskt sätt bättre än någon annan.

Fjärde studien

Både ARM och andra missbildningar gör att vissa barn opereras i buken många gånger under uppväxten. All kirurgi lämnar ärr. Och ärren växer i takt med barnet. I avhandlingens tidigare studie med vuxna i fokusgrupper diskuterades ärr efter kirurgi där man menade att ärren spelar roll.

Vi ville därför i den sista studien undersöka ärrens betydelse för patienterna, om ärren gav några kroppsliga och själsliga symptom, om patienterna var intresserade av plastikkirurgisk ärrbehandling och om en plastikkirurg ansåg att deras ärr kunde behandlas och i så fall hur.

Barn över 5 år och unga vuxna födda med ARM tillfrågades om de ville komma till oss för ärrundersökning, ärrfotografering och ifyllande av olika enkäter om ärr, tarm- och urinfunktion och livskvalitet. Barn deltog tillsammans med en vårdnadshavare. En plastikkirurg bedömde sedan ärrfotografierna avseende möjlighet till ärrbehandling.

Tjugosju personer, 5–24 år gamla, varav hälften flickor/kvinnor, med vardera 1–8 stycken ärr på magen, deltog i studien. Sex deltagare (22%) hade återkommande smärta i ärren, fem (19%) återkommande klåda och nio (33%) alltid heltäckande kläder på sig eller undvek offentliga badplatser då de inte ville visa sina ärr för andra människor. Vissa av deltagarna kände stolthet över sina ärr. Vissa avskydde sina ärr. De flesta hade erfarenheter av andra människors oartiga stirrande och ovälkomna frågor om ärren. Ju äldre deltagarna var desto värre var deras ärrsymptom och ärrutseende med breda och insjunkna ärr. Åtta deltagare (30%), alla över 12 års ålder, önskade plastikkirurgisk ärrbehandling. Det fanns ingen könsskillnad i ärrsymtom eller önskemål om ärrbehandling. Enligt plastikkirurgen var ärren hos 21 (78%) av deltagarna möjliga att göra finare med plastikkirurgi om patienten önskade det.

Studiernas betydelse och vägen framåt

Efter första studien har vi läkare, sjuksköterskor och undersköterskor på barnkirurgen i Lund försökt bli bättre på att hålla utkik efter och känna igen tecken på såruppsprickning. Alla barn får nu samma typ av förebyggande antibiotika. Om de inte har en tarmstomi får de fasta med näringsdropp, för att undvika avföring mot den nya ändtarmsöppningen. Operationssåret rengörs försiktigt flera gånger per dag. Vi tycker nu att färre av våra patienter får såruppsprickning, men denna hypotes behöver prövas med en ny studie. Vi behöver också bedriva mer forskning för att klargöra varför såruppsprickning efter PSARP-operationen inträffar.

I enlighet med den andra studiens resultat vill vi på barnkirurgen i Lund fortsätta att utveckla vårt tidigare påbörjade samarbete med Bäckenbottencentrum kirurgi vid Skånes Universitetssjukhus i Malmö. Vi arbetar med information och utbildning för patienterna, deras anhöriga och andra vårdgivare genom bl.a. webbinarier. Vi försöker strukturera och individanpassa övergången för varje behövande tonåring. Vi försöker alltid vara tillgängliga för våra gamla patienter och hjälpa till och förmedla kontakt med lämpliga vuxenvårdgivare.

I fokusgrupperna lyfte deltagarna även sexualfunktion, fertilitet och genetik inom ARM som viktiga, framtida forskningsområden från patienternas perspektiv.

Tredje studiens resultat, att undersökningsmetoderna hade som mest 70% 'rätt', är inte tillräckligt bra tycker vi. Därför behöver vi förbättra våra befintliga undersökningsmetoder tillsammans med röntgenläkarna för att bättre avbilda och förstå barnets ARM innan PSARP-operationen. Vi behöver även vidareutveckla andra metoder, t.ex. detaljerad ultraljudsteknik och magnetkameraundersökningar.

Fjärde studien visade att ärr efter bukkirurgi i barndomen kan ge symptom som bör föranleda ställningstagande till plastikkirurgisk ärrbehandling om patienten så önskar. Tidigare tillägnades inte ärren någon betydelse vid vår barnkirurgiska uppföljning. Vi bör tvärtom uppmärksamma ärrproblematik precis som alla andra ovälkomna symptom och skicka patienten för plastikkirurgisk bedömning om patienten så vill. Vi barnkirurger behöver även tänka till lite extra och inte skapa större ärr än vad som precis behövs när vi opererar det lilla barnet. För även om ärret inte mäter så långt på den nyfödda, så kommer det att växa med patienten och bli mycket större.

Sammanfattningsvis har studierna i avhandlingen visat att

1. Tarmstomi skyddar något mot såruppsprickning efter PSARP-operationen men vi förstår fortfarande inte mekanismerna bakom.
2. Övergången mellan barn- och vuxenvården skulle kunna förbättras genom högre kunskapsnivå om ARM hos patienter och vuxenvårdgivare, bättre kommunikation mellan patient, barn- och vuxenvårdgivare samt mer lättåtkomlig specialiserad vuxenvård.

3. Metoderna för missbildningskartläggning innan PSARP-operationen har i nuläget som mest 70% rätt enligt vår studie och behöver förbättras.
4. Ärr efter bukkirurgi i barndomen kan ge problem för patienten. Ärrproblematik bör uppmärksammas och plastikkirurgisk ärrbehandling övervägas.

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