



Outcome in adults with anorectal malformations in relation to modern classification – Which patients do we need to follow beyond childhood?



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ABSTRACT

Background/purpose: Knowledge about the functional outcome in adults with anorectal malformations is essential to organize structured transition to adult care for this patient group. The aim of this study was to investigate the functional outcome and quality of life in adults with anorectal malformations characterized according to the Krickbeck classification.

Methods: Of 256 patients diagnosed with anorectal malformations at our institution in 1961–1993, 203 patients could be traced and were invited to participate in the study. One hundred and thirty-six patients replied (67%) and were compared with one hundred and thirty-six population based sex and age-matched controls. Patients and controls were evaluated with both a validated questionnaire as well as a study-specific questionnaire to assess bowel function. SF-36 was used for quality of life. Outcome in nine incontinence-related parameters, 10 constipation-related, 6 urogenital function-related, and 13 quality of life parameters were assessed in the patients and compared to the outcome of controls as well as to the type of anorectal malformations according to the Krickbeck classification.

Results: The ARM-patients had an inferior outcome ($P < 0.05$) for all incontinence parameters, 8 of 10 parameters for constipation, 2 of 6 for urogenital function and 7 of 13 quality of life parameters. Patients with rectobulbar and vestibular fistulas had the worst statistical outcome but patients with cloaca and rectoprostatic/bladder-neck fistula had worse outcome in absolute numbers. Forty-four patients (32%) reported incontinence of stool at least once a week and 16 (12%) had a permanent colostomy.

Conclusions: The functional outcome and quality of life in adults with anorectal malformations are closely related to the type of malformation. A large proportion of the patients have persistent fecal incontinence, constipation and sexual problems that have a negative influence on their quality of life. Structured multidisciplinary follow-up of adults with anorectal malformations by pediatric and colorectal surgeons, as well as urologists and gynecologists is therefore advocated.

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Anorectal malformations (ARMs) are relatively common with an incidence of 1:2500 to 1:5000 live births [1,2]. The clinical presentation is highly variable; from mild forms managed with minor surgical procedures to complex malformations that require multistaged surgery. ARMs are somewhat more common in males than in females with 56% of the cases presenting in males [3].

Improved perioperative care and management of associated anomalies have resulted in a larger proportion of survivors among children with ARM. Consequently, colorectal surgeons, gastroenterologists and family doctors are faced with an increasing number of young adults

with impaired bowel dysfunction owing to ARM. The severity of dysfunction, in relation to type of ARM and previous surgical procedure, is important to understand in order to meet the requirements of these patients and organize transition to adult care.

Most physicians who encounter adult patients with ARM are faced with the older classifications systems from 1970 (the “International classification”) and 1984 (the “Wingspread classification” [4]). These classifications roughly divided the ARMs into low, intermediate and high anomalies. This classification was a good way of determining how the anomalies should be approached surgically (from the perineum or the abdomen) but did not correlate well with the prognosis.

There are a few studies published on the long-term outcome of low anomalies in adult patients, reporting good continence according to scoring systems, but between 14% and 25% of the patients have soiling and up to 49% have fecal smearing [5–11].

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Studies conducted on adult ARM patients with high or intermediate malformations are scarce [12–17]. Most are of older date, only including patients operated with abdominoperineal or abdominosacroperineal procedures. The outcome in these studies was disappointing with 0% to 15% of the patients having normal bowel control and 21% to 27% having a poor outcome. During the last decades most patients have been operated with posterior sagittal anorectoplasty (PSARP [18]). The bowel function of these patients seems superior compared with that of patients operated with older techniques [19,20].

Since the older classification systems were found to be insufficient the Krickenbeck classification (Fig. 1) was developed in 2005 [21]. It was designed to facilitate evaluations of treatment outcomes and is today the predominant classification system used by pediatric surgeons. It has been shown that the outcome in pediatric patients is related to the type of anomaly according to this classification [22]. Up to recently no such studies on adult patients had been published. During the last year papers [23–26] on mixed pediatric and adult materials utilizing the Krickenbeck classification have been published. The results from these papers imply that the classification can be used to predict adult outcome.

The aim of this study was to evaluate the functional outcome and quality of life (QoL) of all patients older than 18 years treated for ARM at our Department and compare with healthy controls. The secondary aim was to relate the outcome to the Krickenbeck classification.

The study did not aim to investigate how the type of surgery performed influences the long-term outcome. A separate paper, based on the same patient material, has been published on that issue [20].

1. Methods

1.1. Patients and controls

The case records and operative registry at the Department of Pediatric Surgery, University hospital, Uppsala, Sweden, were reviewed for all patients diagnosed with ARM from 1961 to 1993 and clinical data were extracted. Out of a total of 256 operated, 22 could not be traced and 32 were dead (31 in infancy and one in adulthood), leaving a cohort of 202 patients. Invitations to participate and questionnaires were sent to these 202 patients. One hundred thirty-six patients (67%) responded and were included in the study (Fig. 2).

The type of ARM was reclassified according to the Krickenbeck classification based on preoperative charts, drawings of the malformation, x-ray reports, radiographs and the surgical reports. Owing to difficulty to adequately distinguish prostatic from vesical fistulas, these patients were included in one group.

Five age- and sex-matched control subjects for every patient were randomly selected from the National Swedish Population Register (The National Swedish Population Register includes all Swedish citizens and all individuals are identifiable by a unique code which also gives

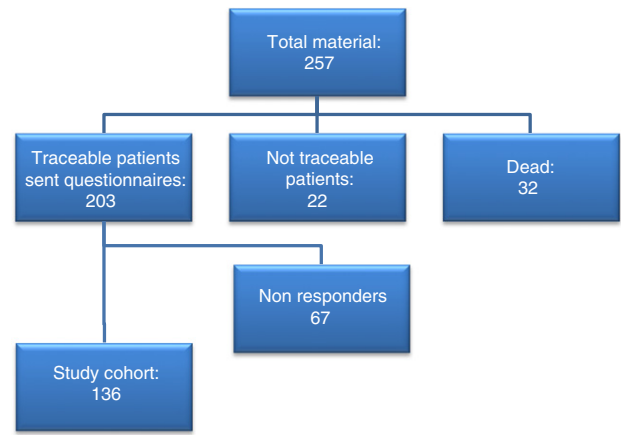


Fig. 2. Flow chart of the inclusion into the study.

information about birth date and gender). They received the same questionnaires as the patients and an invitation to participate in the study. The response rate of control subjects was 24% and one age- and sex-matched control could be assigned to each patient. The controls possible comorbidities were checked from the questionnaires. No control patient had any important comorbidities.

The characteristics of the included patients and the controls as well as for the patients that did not respond or could not traced/deceased are presented in Table 1. The reported incidence of major synchronous malformations is presented in Table 2.

The study was approved by the Regional Ethical Review Board (Dnr 2007/066). All patients and controls provided written informed consent.

1.2. Assessment of patients and controls

The assessments included a validated bowel function questionnaire [27], the Swedish version of SF-36, as well as a designated survey with general questions with relevance for ARM. The bowel function questionnaire consists of 49 questions relating to fecal incontinence, constipation and general bowel function symptoms, allowing calculation of Miller's incontinence score. This score is based on the type and frequency of incontinence episodes; 0 represents total continence and 18 represents total incontinence [28]. The questionnaire also gives information of type of incontinence (classified as soiling, urge, nonurge or combination incontinence), medication, anal sensibility, deferring time and whether the anal continence affects social function in different ways.

SF-36 is a general QoL instrument not specifically designed to evaluate patients with colorectal disease. The Swedish SF-36 form is validated for the Swedish population [29].

<p>Male</p> <p>Perineal (cutaneous fistula)</p> <p>Recto-urethral fistula:</p> <p style="padding-left: 20px;">Bulbar</p> <p style="padding-left: 20px;">Prostatic</p> <p>Recto-vesical fistula</p> <p>No fistula</p> <p>Anal stenosis</p>	<p>Female groups</p> <p>Perineal (cutaneous) fistula</p> <p>Vestibular fistula</p> <p>Cloaca</p> <p>No fistula</p> <p>Anal stenosis</p>
<p>Rare/regional variants</p> <p>Pouch Colon</p> <p>Rectal atresia/stenosis</p> <p>Rectovaginal fistula</p> <p>H-type fistula</p> <p>Others</p>	

Fig. 1. Categories of anorectal malformations according to the Krickenbeck classification [20].

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