



# Persistent fecal incontinence into adulthood after repair of anorectal malformations

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## Abstract

**Purpose** Persistent fecal incontinence beyond childhood is common in ARM patients. The aim of this study was to analyze a consecutive series of adult patients with persistent incontinence, establish the causes, and evaluate whether further treatment could be offered.

**Methods** Forty-four adult ARM patients with reported incontinence were invited. Eighteen patients (11 males, median age 40.5 years, range 18–50 years) accepted and underwent clinical examination, rectoscopy, and 3D-ultrasound. Five had previously been treated with secondary surgery to improve continence.

**Results** Seventeen of the 18 patients had abnormal findings at examination. Eight patients had obstruction of the reconstructed anus. Eleven patients had sacral deformities. Nine patients had a defect in the external anal sphincter and nine patients could not contract the sphincter on demand. Five patients had significant prolapse of mucosa. In one patient, the neoanus was totally misplaced, one patient had a rectovaginal fistula, and one patient had short bowel syndrome due to several small bowel resections. Ten patients were offered conservative and five surgical treatment.

**Conclusions** This case series of adults shows that a majority of the patients can be offered further treatment. This indicates a need for structured follow-up of ARM patients into adulthood.

**Keywords** ARM · Fecal incontinence · Rectoscopy · Adult outcome

## Introduction

Persistent fecal incontinence into adulthood is a common long-term complication after repair of anorectal malformation (ARM), which in severe cases might require formation of a permanent colostomy. There are nine publications that report the long-term term outcome of complex ARM into adulthood

[1–9]. Earlier studies showed that 21–27% of the patients had a poor outcome. More recently, 85% of girls with rectovestibular or perineal fistulas and 76% of boys with recto-urethral fistulas were shown to be socially continent. In these studies, only 6% of the girls and 9% of the boys had a poor outcome. One problem with these studies is that they have been based on questionnaires, which makes it more or less impossible to discern the underlying mechanisms of incontinence.

The aim of the present report was to clinically examine adult ARM patients that reported persistent fecal incontinence, and to define the underlying causes of incontinence and analyze possible treatment options.

## Materials and methods

### Patients

All patients with ARM diagnosed at the Department of Pediatric Surgery, Akademiska sjukhuset, Uppsala, Sweden,

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from 1961 to 1993 were recently reviewed [9]. The cohort consisted of 136 patients. We included patients who reported a Miller's incontinence score [10] above 5 (meaning that they at least had leakage of loose stool once a week) and who reported that their incontinence had a negative effect on their daily quality of life. Patients who had a stoma were excluded. Forty-four patients fulfilled the criteria and were invited to our outpatient clinic for further clinical evaluation. Eighteen patients (41%) accepted the invitation and were included in the study. The characteristics of both the included patients and the patients that declined the invitation were similar except that all females with perineal fistulae declined the invitation. The included patients had mean age at follow-up of 50.5 years (range 26.5–51.0), 11 were male (ARM type: 8 with rectobulbar, 2 with rectoprostatic and one without a fistula) and seven were female (ARM type, 5 vestibular and 2 without fistula).

### Examinations and recommendation of treatments

All included patients underwent a semi-structured interview of approximately 30 min (range 10–49 min) followed by clinical examination of the abdomen and anal region. At the examination, the patients were asked to contract the anal sphincter. A rectoscopy was performed with the patient in the left lateral position. An adult rectoscope was used as standard but a pediatric rectoscope was available if the neoanus was too narrow. Thereafter, anal endosonography was performed as described by Gustafsson et al. [11], except that 3D equipment was used (Ultrasound scanner 2050, 12–16 MHz, B-K Medical, Mileparken 34, Herlev, Denmark). The external and internal sphincters were evaluated. Written interpretation of radiographs of the sacrum were available in all patients. We did not have access to all radiographs and could therefore not calculate sacral ratio.

The examinations were completed in all except two patients. In one patient, it was not possible to complete the rectoscopy/anal endosonography due to significant stenosis of the anal canal and pain at examination. One patient could not cooperate enough to undergo the rectoscopy and was examined clinically and with endosonography. All patients were interviewed and examined by the first author.

After the consultation, further treatment options were discussed and recommended according to the algorithm presented in Table 1.

### Ethical and statistical considerations

The study was approved by the Regional Ethics Review Board, Uppsala, Sweden. All patients provided written informed consent. No statistical analysis was done as this was an observational study.

## Results and discussion

### History

One patient, who had been operated several times for small bowel obstruction, had a malabsorption syndrome due to several small bowel resections. One patient had a clear history of a rectovaginal fistula with vaginal leakage of flatus and feces.

At the consultation, only three patients reported that they had seen a surgeon specifically for their anal incontinence in adult life.

### Findings at clinical examination

Seventeen out of the 18 patients had at least one abnormal finding at examination. Eight patients (44%) had an obstruction of the reconstructed anus (defined as inability to examine the patient with an adult 20-mm rectoscope). Five patients (28%) had significant mucosal prolapse. Nine patients (50%) could not contract the sphincter on demand despite several attempts. In one patient, the neoanus was totally misplaced. The presence of a rectovaginal fistula could not be confirmed during clinical examination of the patient with a history of vaginal discharge and vaginal flatulence.

### Ultrasonographic and radiological findings

Nine patients (50%) had a defect in the external anal sphincter. The rectovaginal fistula could not be identified. Eleven patients had sacral deformities on plain radiographs performed during childhood.

### Further therapy

**Changes in conservative therapy** Eleven patients (61%) were found to have either none or inefficient oral medication. They received optimization of medical treatment (six with macrogol, four with loperamide, and one with cholestyramine). Five of these patients were also referred to biofeedback.

**Further surgical interventions** Five patients were offered further surgical interventions. Antegrade continence enema was recommended for two patients with obstruction who were using daily enemas and had problems taking them in a retrograde fashion. Excision of mucosal prolapse was recommended for two patients, who had significant problems with soiling of mucus. One patient was scheduled for sacral nerve modulation. In addition three patients were offered to be included in a prospective study of injectable bulking therapy.

The patients with total neoanal misplacement and rectovaginal fistula had both lived with their problems for more than 40 years. They were not interested in major surgical intervention.

**Table 1** Algorithm of recommended therapy in relation to clinical findings

Clinical finding	Recommended therapy
1. Medical treatment absent or inappropriate	Optimization
2. Intact sphincter on ultrasound and inability to contract sphincter on demand	Referral to bio-feedback
3. Significant mucosal prolapse and soiling with mucus	Surgical excision
4. Dependent on retrograde enemas and having problems with administration	Antegrade continence enema with Chait button.
5. No contraindications for injectable bulking therapy	Invited to participate in a prospective study of this therapy on ARM patients.
6. Contraindication for injectable bulking therapy and no sacral deformity	Sacral nerve modulation

One patient had a severe stenosis of the anal canal and rectum and it was discussed whether or not he should receive an antegrade continence enema or a permanent colostomy. This was decided against due to severe obesity, multiple abdominal operations and the patient's poor experience of having a stoma during childhood.

## Discussion

Our results indicate that the major anatomical causes of persistent fecal incontinence in adults with repaired ARM are obstruction, mucosal prolapse, sacral deformity, and defects of the external sphincter. Physiologically, the lack of innervation/function or ability to “find” the external sphincter is a major problem. These findings were not unexpected. However, the observation that more than half of the patients had an insufficient conservative treatment and that five patients could be offered further surgical treatment surprised us. Few patients in our series had ever seen a doctor for their incontinence problems as adults. Why had these patients been left with their problems without any attempt of help from the health care? In our opinion, there are two answers to this question: firstly, when the patients leave pediatric surgical care, they have been left with the impression of that “this is as good as it gets”, and secondly: the patients has not had anywhere to seek advice. The pediatric surgeon has ended the contact and to date very few clinics have experience of the care of ARM in adulthood. In Sweden today, there exists only one clinic that specifically specializes in these group of patients. Out of the five patients that were offered further invasive treatment, three were offered treatments that were not available when they left pediatric surgical care. This strongly implies that patients have not had the opportunity to get the benefit of recent medical advances.

One can note that the patients with perineal fistula all declined the invitation. If this was because of that they had less problems or due to other reasons is unknown. Quite few patients in our material had rectoprostatic/vesicular fistulae or cloaca. One of the reasons for this is that many of these patients had a colostomy and were therefore not included in this study.

This small cohort study has several limitations. First: 26/44 subjects declined participation and although clinical characteristics seemed fairly similar between participants and non-participants; we actually do not know which findings would be disclosed among non-participants. Neither did we have the possibility to let the patients undergo MRI of the spinal canal, EMG, anorectal manometry, or measurement of colonic transit time. Second: We do not know how the patients with Miller incontinence score < 5 look in a clinical situation. They may or may not present the same clinical findings as patients that report incontinence.

## Conclusions

The results of this study strongly indicate that there is need for a structured follow-up of ARM patients into adulthood. This follow-up needs to be done at specialized clinics so that the patients can get adequate structured follow-up, medical advice, and state of the art treatment not only during childhood but throughout life.

## Compliance with ethical standards

The study was approved by the Regional Ethics Review Board, Uppsala, Sweden. All patients provided written informed consent. No statistical analysis was done as this was an observational study.

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