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Original Articles

Outcome in anorectal malformation type rectovesical fistula: a nationwide cohort study in The Netherlands

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ABSTRACT

Purpose: Outcomes of patients with an ARM-type rectovesical fistula are scarcely reported in medical literature. This study evaluates associated congenital anomalies and long-term colorectal and urological outcome in this group of ARM-patients.
Methods: A retrospective Dutch cohort study on patients treated between 1983 and 2014 was performed. Associated congenital anomalies were documented, and colorectal and urological outcome recorded at five and ten years of follow-up.
Results: Eighteen patients were included, with a mean follow-up of 10.8 years. Associated congenital anomalies were observed in 89% of the patients, 61% considered a VACTERL-association. Total sacral agenesis was present in 17% of our patients. At five and ten years follow-up voluntary bowel movements were described in 80% and 50%, constipation in 80% and 87%, and soiling in 42% and 63% of the patients, respectively. Bowel management was needed in 90% and one patient had a definitive colostomy. PSARP was the surgical reconstructive procedure in 83%. Urological outcome showed 14 patients (81%) to be continent. No kidney transplantations were needed.
Conclusion: In our national cohort of ARM-patients type rectovesical fistula that included a significant proportion of patients with major sacral anomalies, the vast majority remained reliant on bowel management to be clean after ten years follow-up, despite "modern" PSARP-repair. Continence for urine is achieved in the majority of patients, and end-stage kidney failure is rare.

Anorectal malformations (ARM) represent a complex group of congenital anatomical anomalies of the anorectum, characterized by the absence of a normal anus at its anatomic position central in the sphincter complex [1,2]. The estimated prevalence ranges between 1:2000–5000 live births [2,3]. There is a variety of clinical presentations, ranging from mild to complex ARM, and associated congenital anomalies are present in 40–70% [4–6]. Most anomalies involved are those that are also part of the VACTERL-association (vertebral, cardiac, tracheo-esophageal, renal and limb). Although syndromes are encountered in approximately 10% of the patients (e.g. Townes-Brocks, Currarino syndrome, trisomy 21), the majority of ARM is non-syndromic [2,7].

An anorectal malformation type rectovesical or bladder neck fistula is characterized by a fistulous termination of the rectum to the bladder, typically the bladder neck. Rudimentary elements of the internal anal sphincter are present in this fistulous termination. When preserved, they might improve continence in ARM-patients, although data are contradictory [8,9]. This type of anorectal malformation is a severe type of ARM occurring exclusively in males, in approximately 10% of the patients [10]. It is further associated with hypoplasia of the buttocks and external sphincter complex, contributing to the poor functional outcome for these patients. Data on patients with a rectovesical fistula (RVF) are typically reported as part of larger cohorts of the full spectrum, or of 'high' malformations, which also include recto-urethral-prostatic fistulas [2,11–13]. Until now, ARM-patients with a rectovesical fistula have not been described as a single cohort. Therefore, a retrospective nationwide study was conducted to evaluate associated anomalies and long-term functional colorectal and urological outcome for this rare group of patients.

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1. Patients and methods

A retrospective cohort study was performed in the six pediatric surgical centers in The Netherlands. Charts of all patients with an ARM type rectovesical fistula treated between 1983 and 2014 were reviewed and data on associated congenital anomalies, type of surgery and its timing and complications were collected. Concerning the work-up on associated (VACTERL) anomalies, all patients in The Netherlands are screened for the presence of tethered cord by ultrasound in the first week of life [14]. In case of presence of a tethered cord or doubt on its presence, a spinal MRI is nowadays performed within the first year of life.

For the patients with a minimum follow up period of 48 months, the long-term outcome was assessed, considering the fact that in the general Dutch population children are supposed to be toilet trained by the age of four [15].

The functional colorectal outcome (voluntary bowel movement, constipation, soiling and continence) was documented based on chart notes by the treating surgeon at 4–5 years and 10 years of follow up, respectively, and was graded on the Krickenbeck scoring scale for obstipation and soiling [16]. Voluntary bowel movement is defined by the ability to feel urge with the capacity to verbalize and hold. Data on longer follow-up was scarce, partly because of loss to follow-up or discharge from follow-up, and therefore not analyzed.

The urological and renal function outcomes (spontaneous voiding, continence, clean intermittent catheterization, urinary diversions or other urological surgery, recurrent urinary tract infections, end-stage kidney failure) after 5 years were also documented in this study.

1.1. Statistics

Frequencies of nominal variables are presented including percentages. Continuous variables with a normal distribution are presented with a mean and standard deviation. In some instances also a range is shown for extra information. For continuous variables with a skewed distribution the median and range are provided. Numbers are too small to perform statistical significant tests.

2. Results

Eighteen patients were identified and included. Mean follow up was 10.8 years (SD: 5 years, range: 2–19 years). Table 1 shows the associated congenital anomalies observed in 16 patients (89%). Eleven patients met the criteria of a VACTERL-association (61%). Most prevalent associated anomalies were sacral vertebral in 12 patients (67%), being hemivertebrae, sacral fusion, partial sacral agenesis and coccygeal agenesis. Three patients had a total sacral agenesis. No patient was diagnosed with Currarino syndrome. Anatomical kidney anomalies were present in 11 patients (61%). Of this group there were three patients with a single kidney, three presented with a horseshoe kidney, two had ureteropelvic junction (UPJ) stenosis, one dysplastic kidney, another a dysfunctional kidney and one had multicystic kidney disease.

Table 1
Associated congenital anomalies in 18 patients with ARM and a rectovesical fistula.

Type of anomaly	N (%)
Cardiac	5 (28)
Oesophageal	1 (6)
Spinal cord	4 (22)
Sacral	12 (67)
Vertebral*	2 (11)
Renal	11 (61)
Limb	2 (11)

* 1 missing value.

All patients needed a colostomy in the neonatal period, with a complication rate of 17% (3 of 18 patients, in one patient data were missing). The colostomy was closed at a mean age of 11 months (SD: 5 months, range: 3–24 months). One patient had a definitive colostomy at the age of 5, after multiple laparotomies and ischemic complications.

In the majority of patients (83%), reconstructive surgery was performed using a posterior sagittal anorectoplasty (PSARP). For the abdominal phase a laparotomy was performed in 10 and a laparoscopy in 6 patients (62% and 19% respectively). Of these 6 laparoscopy-assisted procedures, 3 were combined with a PSARP and 3 had a laparoscopy-assisted pull through procedure. One of these last patients eventually had a definitive colostomy. In two patients the data on the surgical treatment were missing. The median age at reconstructive surgery was six months (range: 2–26 months). To prevent anal stenosis, anal dilatations according to the Peña guidelines were standard postoperative care in all pediatric surgical centers [2].

Table 2 shows the number of complications (12) after reconstructive surgery, which occurred in 59% of the patients. They are divided in anorectal (10) and urological (2). Of the anorectal complications, two were considered major (anal stenosis and megacolon), as they have lead to major redo-surgery. All other anorectal complications can be considered mild, although the consequences were multiple additional surgical procedures in three patients. Data on possible reoperations were missing in the other patients. The urological complications, being urinary tract infection and a urinary retention, were considered mild. A mean of 5.9 surgical procedures per patient (SD: 2.8, range: 2–12), being reconstructive and/or urological, was needed for definitive repair.

Table 3 shows functional colorectal outcome analyses after five and ten years according to the Krickenbeck criteria in 15 and 10 patients, respectively. Twelve patients (80%) had voluntary bowel movements after five years and five patients (50%) after ten years.

Constipation was documented in 12 out of 15 patients after five years, and 7 out of 8 at ten years follow-up (80 and 87%, respectively). Soiling was reported in 42% of 12 patients after five years, and 63% of eight patients after ten years follow-up. Grade 3 soiling, defined as socially disabling, was present in 17% and 13% of the patients after five and ten years respectively.

After ten years follow-up, bowel management was needed in 90% of the patients (9 out of 10) for the treatment of incontinence and constipation. This was mostly done by administering rectal enemas without the need for an ACE-conduit [17]. There were no reliable data on the timing of commencement of bowel management. Fifty percent of the patients used laxatives, initiated by the individual surgeon not based on any predefined criteria, and occasionally on request by the patient and/or his parents. One patient (10%) had a definitive colostomy.

Urological outcome on 16 patients could only be evaluated at 5 years follow-up (Table 4). Seven of the included 18 patients (39%) had vesico-

Table 2
Complications after reconstructive surgery.

Complication*	N = 17**	Surgical intervention
None	7 (41%)	
Anorectal	10	
Anal stenosis	4	1 redo-PSARP, 3 repetitive anal dilatations under anesthesia
Anal prolaps	2	Unknown
Mechanical bowel obstruction	2	Unknown
Megacolon	1	Twice plication in a single patient
Stenosis of colostomy	1	Unknown
urologic	2	
Urinary tract infection	1	
Urinary retention	1	
Total complications	12	

* Patients can have multiple complications.
** N is number of patients.