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Cloacal malformation patients report similar quality of life as female patients with less complex anorectal malformations

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ABSTRACT

Purpose: Cloacal malformations are the most complex type of anorectal malformation in females. This study aimed to report quality of life (QoL) of patients with a cloacal malformation for the first time in literature.
Materials and Methods: Female patients with an anorectal malformation participating in a follow-up program for congenital malformation survivors aged 5 or older were eligible for this study. QoL was assessed with the PedsQL™ 4.0 inventory. Scores of patients with a cloacal malformation (CM) were compared with those of female patients with rectoperineal or rectovestibular fistulas (RP/RV) and with reference data.
Results: A total of 59 patients (67% response rate; 13 patients with cloacal malformation) were included, QoL was assessed by patient self-report at median age of 12 years (8–13), and by parent proxy-report at median age of 8 years (5–12). There were no differences between groups regarding the presence of associated anomalies, with also no differences regarding anomalies in the urinary tract (CM vs. RP/RV = 31% vs. 15%, $p = 0.237$). Scores of the cloacal malformations group were similar to those of the comparison group, except the proxy-reported scores on school functioning (60.0 vs. 80.0, $p = 0.003$). Proxy-reported scores of cloacal malformation patients were significantly lower than reference values on total QoL-score, psychosocial health, and emotional and school performance. Patients (irrespective of type of ARM) who suffered from fecal soiling reported significantly lower scores with regard to psychosocial health (71.7 vs. 81.7, $p = 0.034$) and its subscale school performance (65.0 vs. 80.0, $p < 0.001$). QoL-scores reported by cloacal malformation patients did not differ significantly from the reference values of the healthy population. Parents of cloacal malformation patients reported significantly lower total QoL, emotional and school performances, as well as a lower general psychosocial health for their children relative to reference data of healthy children.
Conclusion: Patients with cloacal malformations and females with less complex anorectal malformations report similar QoL. Parents of cloacal malformation patients report more problems on several psychosocial domains relative to the healthy reference group. To monitor these matters, long-term follow-up protocols should contain multidisciplinary treatment including periodical assessment of psychosocial well-being.

In patients born with a cloacal malformation the anus is absent and the rectum, vagina and urethra confluence in one common channel. Cloacal malformations are considered more complex than other types of anorectal malformations (ARM) in females, such as rectoperineal or rectovestibular fistulas [1]. Most ARM patients will undergo multiple surgical procedures in the first years of life, which aim to achieve the best possible bowel function, urological function, and gynecological function. However, the majority of cloacal malformation patients will still suffer from long-term impairments in one or more of these functions [2]. These impairments may have an impact on QoL. On the other hand, a study in patients with ARM – but not including patients with cloacal malformations – found that impairments such as fecal incontinence or constipation had almost no effect on QoL and that psychosocial functioning was more important [3]. In another study by Hartman et al., however, poorer QoL was reported for female patients and for patients suffering from associated congenital anomalies [4]. Thus, as a cloacal malformation obviously occurs in females only, and considering that in up to 88% of cases it is associated with other congenital anomalies [5], there is reason to expect that QoL in the latter patients will be lower than that in female patients with other ARM. We are not aware of studies that have addressed this assumption. With this study, therefore, we aimed to report QoL in patients with cloacal malformations during childhood and adolescence for the first time in literature and to

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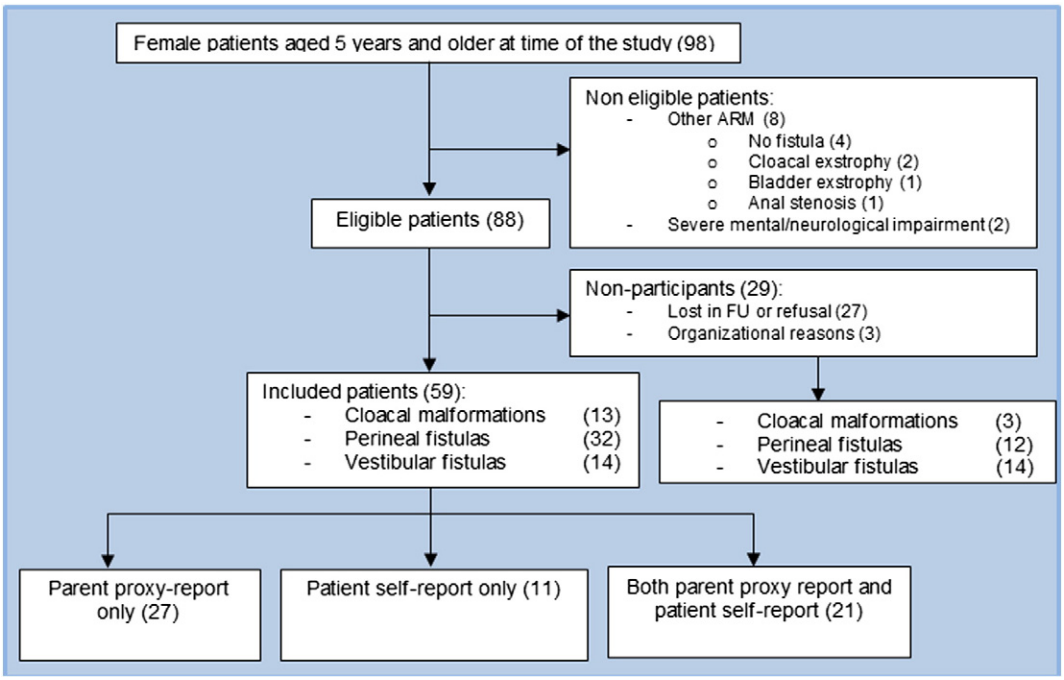

Fig. 1. Flowchart of patient inclusion.

compare outcomes with those in female patients born with a rectoperineal or a rectovestibular fistula. We furthermore compared them with reference values of the general Dutch population.

1. Methods and materials

All ARM patients treated in our institution (Erasmus MC-Sophia Children's Hospital) are invited to join a prospective, structured follow-up program for congenital malformation survivors [6,7]. The assessment protocol for this program is standard of care at our institution. The Erasmus MC Medical Ethical Review Board (IRB) ruled that the "Medical Research in Human Subjects Act" does not apply to this study, since subjects are not being submitted to any handling, nor are there rules of human behavior being imposed. Therefore, IRB approval was waived. All parents were informed about the study and provided permission to use the data for research purposes. Included in the program is a periodical assessment of QoL using proxy-reports from the age of 5 years and additionally self-reports from 8 years onwards. All female ARM patients with a cloacal malformation, or a rectoperineal or rectovestibular fistula aged 5 years and older whose QoL was assessed were eligible. Severe mental or neurological impairment was an exclusion criterion (Fig. 1).

QoL was assessed with the PedsQL™ 4.0 inventory [8,9], which was constructed to test health-related QoL in children and adolescents. Validated, age-appropriate versions are available for children aged 5 to 18 years as a parent proxy-report and children/adolescents aged 5 to 25 as patient self-report. The questionnaire assesses both physical health (8 items) and psychosocial health, divided in emotional, social, and school performance (5 items each). All 23 items scored will generate a total QoL-score with a maximum possible score of 100. Parent proxy-reports were completed by the parents at home, and self-reports were completed with the help of a psychologist at the outpatient clinic (without parents).

If a patient (and/or her parents) had completed the questionnaire more than once, only the latest self- and proxy-reports were used for this study. Data regarding associated anomalies, surgical procedures, and functional outcome such as colorectal function and urinary continence were obtained in the same outpatient clinic with the use of structured interviews. Colorectal function was classified according to the Krickenbeck criteria [10]. Dysfunctional voiding was defined as either urinary incontinence or need for intermittent catheterization.

Data were stored and analyzed using SPSS version 21 (SPSS Inc, Chicago, Ill). QoL-scores are expressed as median (IQR) unless otherwise stated. Mann–Whitney U nonparametric tests and one-sample Wilcoxon signed rank tests were used to compare QoL-scores of different types of ARM and to compare these scores with Dutch reference data previously obtained in 74 children aged 7.4–18.2 years [11].

2. Results

2.1. Patient characteristics

Since the introduction of the follow-up program in 1999, 88 female patients with ARM aged five or older were seen at outpatient appointments and met all inclusion criteria. Fifty-nine (67%) of them, and/or

Table 1A
Clinical characteristics: patient self-report group ($n = 32$).

	CM (11)	RP/RV (21)	Total (32)	*p*-Value
Median age (years)	14 (12–19)	12 (8–13)	12 (8–13)	**0.009**
Associated anomalies	6 (55%)	12 (57%)	18 (56%)	1.000
Voluntary bowel movements	4 (36%)	18 (86%)	22 (69%)	**0.013**
Soiling	6 (55%)	7 (33%)	13 (41%)	0.283
Grade 1	3 (27%)	1 (5%)	4 (13%)	
Grade 2	3 (27%)	5 (23%)	8 (25%)	
Grade 3	0 (–)	1 (5%)	1 (3%)	
Constipation	5 (45%)	11 (52%)	16 (50%)	1.000
Grade 1	0 (–)	1 (5%)	1 (3%)	
Grade 2	0 (–)	4 (19%)	4 (13%)	
Grade 3	5 (45%)	6 (28%)	11 (34%)	
Colostomy	3 (27%)	1 (5%)	4 (13%)	0.106
Urological outcome				
Spontaneous voiding	6 (55%)	15/20 (75%)	21/31 (68%)	0.423
Clean intermittent catheterization	2 (18%)	3/20 (15%)	5/31 (16%)	
Incontinence	2 (18%)	2/20 (10%)	4/31 (13%)	
Diversion	1 (9%)	0 (–)	1/31 (3%)	

Bold/italic font indicates statistical significance.
CM *cloacal malformation group*; RP/RV *rectoperineal/rectovestibular group.*