Pediatric Surgery International (2024) 40:179
https://doi.org/10.1007/s00383-024-05766-1

ORIGINAL ARTICLE

Urological outcomes in adult females born with anorectal malformation or Hirschsprung disease

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Accepted: 29 June 2024
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Abstract

Introduction Women born with anorectal malformation (ARM) or Hirschsprung disease (HD) may have impaired urologic function resulting in sequelae in adulthood. This study assessed and compared self-reported urinary outcomes in adult females born with ARM or HD to a reference population.

Methods This was an IRB approved, cross-sectional study of female-born patients with ARM or HD, who completed surveys between November 2021 and August 2022. Female patients between the ages of 18 and 80 years were included. Lower Urinary Tract Symptom Questionnaires were administered through REDCap and the responses were compared to a reference population using Chi-squared or Fisher's exact tests.

Results Sixty-six born female patients answered the questionnaires, two of them identified as non-binary. The response rate was 76%. Median age was 31.6 years. The majority were born with cloaca (56.3%), followed by other type of ARMs (28.1%), complex malformation (9.4%), and HD (6.3%). A history of bladder reconstruction was present for 26.6%. Catheterization through a channel or native urethra was present in 18.8%. Two had ureterostomies and were excluded from the analysis. Seven had chronic kidney disease or end-stage renal disease, three with a history of kidney transplantation.

Patients with cloaca had significantly higher rates of urinary incontinence, urinary tract infection, and social problems due to impaired urological functioning, when compared to an age-matched reference population (Table 3).

Conclusion This study emphasizes the need for a multi-disciplinary team that includes urology and nephrology following patients with ARM long term, especially within the subgroup of cloaca.

Level of Evidence III.

Keywords Anorectal malformation · Cloaca · Hirschsprung disease · Urinary function · Urological outcome · Urinary incontinence

Abbreviations

ARM	Anorectal malformation
HD	Hirschsprung disease
UI	Urinary incontinence
CKD	Chronic kidney disease
ESRD	End-stage renal disease
UTI	Urinary tract infection
UDS	Urodynamic study
PSARP	Posterior sagittal anorectoplasty

Introduction

The diagnosis of an anorectal malformation (ARM) is associated with urogenital anomalies with a reported prevalence ranging from 18.4—85% [1–4], with a higher reported prevalence depending on type and severity of the ARM. In patients with Hirschsprung disease, the reported incidence ranges from 5.1 to 25% [5–8] Urinary and renal outcomes of adult ARM and HD patients are currently sparse with recent publications emphasizing the importance of transitional care with particular interest in urogenital health [1, 9, 10]. In addition, standardization of screening methods in ARM as

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Published online: 06 July 2024

well as HD for urinary tract anomalies and dysfunction has been suggested by several authors [1, 2, 5–7, 11]. The aim of this study is to evaluate self-reported urinary and renal outcomes in female-born patients with ARM or HD in comparison to a reference population.

Methods

This was a cross-sectional study of female-born patients born with either ARM or HD. Patients between the ages of 18 and 80 years of age were included. Patient self-reported demographic data were linked to surgical records to ensure accurate colorectal diagnoses and anatomy, in the case of ARM. Patients were asked to provide basic demographic information and complete surveys through REDCap which included self-reported urological function outcomes.

To further evaluate and compare the study participants to a healthy reference population, Lower Urinary Tract Symptom Questionnaires were administered. Only patients who answered those questionnaires between November 2021 and August 2022 were included. Lower Urinary Tract Symptoms Questionnaire responses were compared between the study sample and a reference population using Chi-squared or Fisher's exact tests. For the reference population, the females aged 18–26 years were chosen, since this is most similar to the study sample population [12]. However, for some questions, Kyrklund et al. did not report statistics for this reference population. In these cases, another similar reference population was chosen from the reported results by Kyrklund. The specific reference population used for each Lower Urinary Tract Symptoms Questionnaire question is noted in Table 3.

In addition, rates of urinary incontinence were compared between the study sample and a reference population of 30–39 years old US women using Chi-squared or Fisher's exact tests [13]. For our study sample, the following question was used to measure urinary incontinence: "Does urine ever leak without physical activity or need to urinate?". Comparisons between the study sample and reference populations were performed for the full study sample, by diagnosis, and by common channel length for cloaca patients.

The patient population was assessed and compared to healthy references twice, once including all patients, and once excluding patients above age 40 years, due to concerns that urinary functioning and incontinence worsens with age.

A p value <0.05 was considered significant. Descriptive statistics are summarized as mean (SD) and median (Q1, Q3) or frequency (%). All analyses were performed in R (version 4.1.2).

This study was approved by the Colorado Multiple Institutional Review Board (COMIRB) (APP001-1).

Results

Demographics and medical conditions

A total of 66 female-born patients participated and answered all questions, with two identifying as non-binary (Table 1), two were excluded due to having ureterostomies. The response rate was 76%. Median age was 31.6 years with an age range from 22 to 80 years. Patients were born with cloaca (56.3%), other types of ARM (28.1%), complex malformation (9.4%), and HD (6.3%). Twenty of the patients with cloaca had a common channel ≤ 3 cm (55.6%), thirteen (30.5%) had a common channel longer than 3 cm, and

Table 1 Overall demographics and surgical history of study population

	Overall ($N=64$)
Age (years) at time of survey completion	
Mean (SD)	33.5 (9.81)
Median (Q1, Q3)	31.6 (27.0, 36.6)
Gender identity	
Female	62 (96.9%)
Non-binary or transgender	2 (3.1%)
Diagnosis	
ARM	18 (28.1%)
Cloaca	36 (56.3%)
Complex malformation	6 (9.4%)
Hirschsprung disease	4 (6.3%)
In cloaca, common channel length ($N=36$)	
Less or equal 3 cm	20 (55.6%)
Longer than 3 cm	11 (30.5%)
Unknown	5 (13.9%)
Antegrade enema procedure	
Yes	17 (26.6%)
No	47 (73.4%)
Sacral Ratio	
Normal >0.7	24 (36.4%)
Intermediate 0.4–0.7	17 (25.8%)
Poor <0.4	3 (4.5%)
Unknown	22 (33.3%)
Tethered cord release	
Yes	8 (12%)
No	58 (88%)
Bladder neck reconstruction/augmentation	
Yes	17 (26.6%)
No	47 (73.4%)
Ileostomy/colostomy	
Yes	5 (7.8%)
No	59 (92.2%)

in five (13.9%), the length was unknown. Seven patients underwent a total urogenital mobilization. Complex malformation patients had a diagnosis of cloacal exstrophy, covered cloacal exstrophy, posterior cloaca, or other complex malformations not otherwise specified (three patients). A third (36.4%) had a normal sacral ratio (SR) with 0.7 or higher, 25.8% had a SR between 0.4–0.7, 4.5% had a poor SR of <0.4 and the SR was unknown in 33%.

Patients' surgical history included bladder reconstruction or augmentation in 26.6%. Four of them had a diagnosis of a complex malformation, the remaining patients were born with a cloaca. Three cloaca patients had a long common channel. Eight patients had a tethered cord release.

Seven reported chronic kidney disease (CKD) or end-stage renal disease (ESRD), three had CKD stage III and higher, one had ureterostomies and was on hemodialysis, and three had a kidney transplant. The patients with CKD III and higher all were born with a single kidney, except for two (one with cloacal exstrophy with ectopic ureters and one had an unrepaired UG sinus for 10 years). The three that had a kidney transplant were born with a cloaca and a single kidney, as well as a poor sacral ratio (<0.4) and/or long common channel or renal insufficiency at birth. High blood pressure requiring antihypertensive medication was reported by 28.7%, the majority had a cloaca (73.7%). Two patients had diabetes mellitus (one with cloaca and one with complex malformation).

The majority (89.4%) of patients had a history of urinary tract infection (UTI), and in 66%, the last infection was less than 12 months ago. All patients with cloaca except one had a history of UTIs. Nine of them had more than six UTIs within the last year. At the time of the study, eight took oral or intravesical antibiotics to either treat or prevent a UTI. Five patients were treated for overactive bladder with Oxybutynin, all were born with a cloaca.

Urinary leakage without any physical activity or need to urinate was reported by 37.5% (excluding patients with ureterostomy) (Table 2). Forty-seven percent of the patients with cloaca had urinary incontinence. The highest rate of urinary incontinence was found within the complex malformation group, but this group was small with only six patients in total. None of the HD patients had urinary incontinence. Patients with cloaca had a significantly higher rate of urinary incontinence compared to the reference population (p value <0.001), but the rates were similar by channel length (42.1% for ≤ 3 cm, 40% for >3 cm).

A quarter (16 patients) reported a history of pregnancy (16 patients), all of them gave birth through a C-section, four had urinary incontinence, three had a cloaca. Those without urinary incontinence, a third (12 patients) had a history of pregnancy and birth via C-section, the majority were patients with ARM (5 patients), followed by cloaca (4 patients).

Lower Urinary Tract Symptoms Questionnaire

A total of 64 patients were included for further comparison to a reference population, 2 patients with ureterostomies were excluded. Overall, most patients had a history of UTI's and 37.5% of them suffered of frequent urge to urinate (Table 2). Urge incontinence was reported by 22%, stress incontinence by 23%, and 16% had unconscious urine leakage. Most patients did not have any night-time wetting (82%) or social problems due to urinary incontinence (87.5%).

The subpopulations were then compared to the age-matched reference population with the identical 9-item questionnaire previously published by Kyrklund [12]. Since the surgical approach has significantly changed with implementation of the PSARP operation, patients older than 40 years of age were excluded. In addition, because urinary incontinence rate increases with age, patients older than 40 years of age were excluded for a more homogenous study population for comparison. Nine patients were older than 40 years of age (43–80 years). The statistical analysis did not change if patients older than 40 years of age were included or excluded. Patients with ARM showed significantly higher rates of urinary urge, urge incontinence, and urine incontinence (incontinence without physical activity or need to urinate) (Table 3). Patients born with cloaca reported significant impairment in all areas except for the frequency of urination. Interestingly, although statistically significant when compared to the healthy reference group, 85% of patients with cloaca did not report any social problems, even though 47.5% of patients with cloaca suffered of urinary incontinence.

Patients with complex malformations had similar results to cloaca patients of urinary incontinence and social problems due to incontinence but had higher reported rates of night-time bedwetting. HD patients had similar results as the healthy references, except for the symptom of urinary urge.

Overall rates of urinary incontinence were compared between the study sample and a similar age-ranged reference population of US women between the age of 30–39 years, since this was closest to the median age of our study population (Table 4) [13]. Both for patients ≥ 40 years and the entire study cohort, those with cloaca had significantly higher rates of urinary incontinence compared to the reference population (both p value <0.001). Patients with ARM had slightly lower rates of urinary incontinence compared to the reference population (22.2% in all patients, 21.4% in patients ≥ 40 years compared to 28% of the reference population), but this difference was insignificant (p value $=0.77$ and 1, respectively).

Table 2 Lower Urinary Tract Symptoms Questionnaire by diagnosis, age 22–80 years

	ARM (*N* = 18)	Cloaca (*N* = 36)	CC ≤ 3 (*N* = 18)	CC > 3 (*N* = 10)	Complex (*N* = 6)	HD (*N* = 4)
Have you had a urinary tract infection?						
Yes	14 (77.8%)	35 (97.2%)	17 (94.4%)	10 (100%)	6 (100%)	2 (50.0%)
No	4 (22.2%)	1 (2.8%)	1 (5.6%)	0 (0%)	0 (0%)	2 (50.0%)
How many times do you pass urine each day?						
1–3 times	3 (16.7%)	4 (11.1%)	3 (16.7%)	1 (10.0%)	0 (0%)	1 (25.0%)
4–8 times	11 (61.1%)	25 (69.4%)	12 (66.7%)	7 (70.0%)	5 (83.3%)	3 (75.0%)
More than 8 times	4 (22.2%)	7 (19.4%)	3 (16.7%)	2 (20.0%)	1 (16.7%)	0 (0%)
Do you need to strain to start/continue urination?						
Never	15 (83.3%)	21 (58.3%)	12 (66.7%)	7 (70.0%)	3 (50.0%)	2 (50.0%)
Seldom	3 (16.7%)	6 (16.7%)	3 (16.7%)	2 (20.0%)	3 (50.0%)	2 (50.0%)
Often	0 (0%)	5 (13.9%)	2 (11.1%)	0 (0%)	0 (0%)	0 (0%)
Always	0 (0%)	4 (11.1%)	1 (5.6%)	1 (10.0%)	0 (0%)	0 (0%)
Do you get a sudden urge to pass urine?						
Never	8 (44.4%)	6 (16.7%)	3 (16.7%)	2 (20.0%)	2 (33.3%)	3 (75.0%)
Seldom	7 (38.9%)	14 (38.9%)	8 (44.4%)	3 (30.0%)	0 (0%)	0 (0%)
Often	3 (16.7%)	8 (22.2%)	1 (5.6%)	4 (40.0%)	4 (66.7%)	0 (0%)
Always	0 (0%)	8 (22.2%)	6 (33.3%)	1 (10.0%)	0 (0%)	1 (25.0%)
Is the urge so strong that urine escapes before reaching the toilet?						
Never	11 (61.1%)	14 (38.9%)	9 (50.0%)	4 (40.0%)	4 (66.7%)	4 (100%)
Seldom	5 (27.8%)	12 (33.3%)	4 (22.2%)	4 (40.0%)	0 (0%)	0 (0%)
Often	2 (11.1%)	8 (22.2%)	4 (22.2%)	2 (20.0%)	2 (33.3%)	0 (0%)
Always	0 (0%)	2 (5.6%)	1 (5.6%)	0 (0%)	0 (0%)	0 (0%)
Does urine ever leak upon straining (e.g., laughing, sneezing, or coughing)?						
Never	10 (55.6%)	16 (44.4%)	8 (44.4%)	5 (50.0%)	2 (33.3%)	2 (50.0%)
Seldom	5 (27.8%)	9 (25.0%)	5 (27.8%)	3 (30.0%)	3 (50.0%)	2 (50.0%)
Often	2 (11.1%)	5 (13.9%)	3 (16.7%)	1 (10.0%)	1 (16.7%)	0 (0%)
Always	1 (5.6%)	6 (16.7%)	2 (11.1%)	1 (10.0%)	0 (0%)	0 (0%)
Does urine ever leak without physical activity or need to urinate?						
Never	14 (77.8%)	19 (52.8%)	10 (55.6%)	6 (60.0%)	3 (50.0%)	4 (100%)
Seldom	2 (11.1%)	9 (25.0%)	5 (27.8%)	2 (20.0%)	3 (50.0%)	0 (0%)
Often	2 (11.1%)	6 (16.7%)	3 (16.7%)	2 (20.0%)	0 (0%)	0 (0%)
Always	0 (0%)	2 (5.6%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)
Do you have night-time wetting (bedwetting)?						
Never	18 (100%)	28 (77.8%)	15 (83.3%)	9 (90.0%)	3 (50.0%)	4 (100%)
Less than once a week	0 (0%)	8 (22.2%)	3 (16.7%)	1 (10.0%)	3 (50.0%)	0 (0%)
More often than once a week	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)
Every night	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)
Do you have social problems due to urinary incontinence?						
No	17 (94.4%)	31 (86.1%)	15 (83.3%)	9 (90.0%)	4 (66.7%)	4 (100%)
Yes—due to daytime urinary incontinence only	1 (5.6%)	1 (2.8%)	0 (0%)	0 (0%)	1 (16.7%)	0 (0%)
Yes—due to night-time urinary incontinence only	0 (0%)	2 (5.6%)	2 (11.1%)	0 (0%)	0 (0%)	0 (0%)
Yes—due to daytime and night-time urinary incontinence	0 (0%)	2 (5.6%)	1 (5.6%)	1 (10.0%)	1 (16.7%)	0 (0%)

ARM anorectal malformation, *CC* common channel length, *CC ≤ 3* common channel length equal or less than 3 cm. *CC > 3* common channel length longer than 3 cm, *Complex* complex malformation, *HD* Hirschsprung disease

Table 3 Comparison of questionnaire answers by diagnosis to reference population [12]

Question	Reference population+	ARM $N=18$	Cloaca $N=36$	CC ≤ 3 cm $N=18$	CC > 3 $N=10$	Complex $N=6$	HD $N=4$
Have you had a urinary tract infection?	Females aged 18–26	0.047	< 0.001	< 0.001	0.002	0.032	NS
How many times do you pass urine each day?	Males and females aged 18–26	NS	NS	NS	NS	NS	NS
Do you need to strain to start/continue urination?	Males and females aged 18–26	< 0.001	< 0.001	0.002	< 0.001	0.008	0.04
Do you get a sudden urge to pass urine?	Males and females aged 18–26	NS	< 0.001	< 0.001	< 0.001	< 0.001	0.002
Is the urge so strong that urine escapes before reaching the toilet?	Males and females aged 18–26	< 0.001	< 0.001	< 0.001	< 0.001	0.001	NS
Does urine ever leak upon straining (e.g., laughing, sneezing or coughing)?	Females aged 18–26	NS	< 0.001	NS	0.034	NS	NS
Does urine ever leak without physical activity or need to urinate?	Males and females aged 8–26	0.007	< 0.001	< 0.001	< 0.001	0.001	NS
Do you have night-time wetting (bedwetting)?	Females aged 18–26	NS	< 0.001	0.006	NS	< 0.001	NS
Do you have social problems due to urinary incontinence?	Males and females aged 13–26	NS	< 0.001	< 0.001	0.032	< 0.001	NS

ARM anorectal malformation, *CC* common channel length, *CC ≤ 3* common channel length equal or less than 3 cm *CC > 3* common channel length longer than 3 cm, *Complex* complex malformation, HD: Hirschsprung disease

+ Kyrklund K, Taskinen S, Rintala R, Pakarinen M:Lower Urinary Tract Symptoms from Childhood to Adulthood: A Population Based Study of 594 Finnish Individuals 4 to 26 Years Old. J Uro Vol 188, 588–93, 2012. https://doi.org/10.1016/j.juro.2012.04.016

Table 4 Comparison of urinary incontinence (UI) rates to reference population rate of 28% [13]. *p* value did not change within significance range if patients older than 40 years of age were excluded

Sample population	Rate of UI %	*p* value
Full sample population		
All patients ($N=64$)	37.5	0.002
Patients ≤ 40 years old ($N=54$)	35.7	0.010
By diagnosis ($N=64$)		
ARM ($N=18$)	22.2	0.772
Cloaca ($N=36$)	47.2	< 0.001
Complex malformation ($N=6$)	50	0.104
Hirschsprung disease ($N=4$)	0	0.589

Discussion

Urological tract involvement associated with ARM occurs in up to 50% of patients and poses an increased risk of secondary damage to the kidneys and urinary tract [1–4, 10]. CKD Stage II or higher in patients with ARM was found to be as high as 28% in literature which included males and females. As previously published by several authors and confirmed with this study, patients with cloaca are at highest risk of developing chronic kidney disease and renal failure within female patients with ARM [4, 14]. Six percent had ESRD with hemodialysis or kidney transplantation, all of them born with cloaca and a single kidney, which is very similar to what was previously reported by Warne et al. (6.4%) [15]. Besides long-term renal outcome, patients with cloaca suffered of significantly higher rates of UTIs, urinary incontinence, and lower urinary tract symptoms compared to a healthy, age-matched reference population.

Guiliani postulated three types of etiopathology leading to CKD in patients with ARM: developmental (born with an insufficient nephron mass), infectious (recurrent UTIs due to genito-urinary anomalies), and a combination of both [16]. Since the nephron mass can only be preserved and not increased postnatally, prevention and preservation of renal function is of utmost interest. Up to this date, there is no published standardized follow-up protocol for patients born with ARM. Our data showed that patients with cloaca have the highest incidence of ESRD, with impairment of urinary incontinence and lower urinary tract symptoms. Based on this data, we strongly suggest a standardized follow-up protocol (including clinical, laboratory, and radiological studies) with a collaborative approach including involvement of urology and nephrology early in life. At our center, patients born with a cloaca will be initially assessed during the postnatal period with a renal and spinal ultrasound and urology consultation. At the time of surgical correction, a cysto- and vaginoscopy, including measurement of the common channel length is performed. This will be repeated at the time of colostomy closure (usually 3 months postoperatively). At the age of 1 year, further evaluation of the urinary tract with a urodynamic study (UDS) will be obtained. In addition, besides the follow-up renal ultrasound (US) at 3 months and 1 year, laboratory markers, BMP, and Cystatin C will be drawn and repeated every year. If the patient has UTI's, an abnormal upper urinary tract on US, abnormal UDS,

remains incontinent by the age of 5 years, has CKD stage II or higher, the UDS will be repeated.

Interestingly, although almost half (47.5%) of the patients with cloaca suffered of urinary incontinence, the majority of them did not report any impact on their social life. This could be explained by the fact that urinary incontinence is less stigmatized within the society and urinary incontinence pads and other hygienic products are readily available in stores.

Urinary incontinence can occur and may worsen in healthy females during or after pregnancy with an incidence as high as 40% with vaginal birth delivery being a risk factor [17, 18]. In our study, a quarter reported successful pregnancy and delivery via C-section only. Of those who were (urinary) continent, the majority were ARM patients, compared to those with urinary incontinence; these were cloaca patients. Patients with recto-perineal and recto-vestibular fistulas are expected to have similar fertility rates as the healthy population [19, 20]. As previously published, fertility concerns are highest among cloaca patients with a third of them having fertility problems [21].

This study has some weaknesses, one of which is the comparison to a healthy, mixed gender reference group in some of the assessed items of the urinary tract symptom questionnaire. Therefore, additional evaluation with comparison of the study population to age-matched US females in regards of urinary incontinence was performed. No change of the significance was seen.

The nature of this retrospective study entails that some screening methods as well as the PSARP technique, that are now standard in ARM patients, were not implemented at the time of birth in patients born earlier than 1982. To diminish this impact, statistical analysis was performed twice with and without patients age > 40 years of age, and no difference within the statistical results was seen. For the subgroup of complex malformations and HD, statistical analysis was performed but due to the small patient population subgroup, the results should be read with caution.

In addition, patients who are catheterizing through a native urethra or underwent bladder augmentation and are catheterizing through a channel (Mitrofanoff or Monti), their symptoms are not well assessed with this questionnaire and further specific evaluation with creation of a validated questionnaire for patients who catheterize is planned for future studies.

We encourage the implementation of follow-up protocols for patients with ARM to detect, prevent damage, and preserve renal and urinary tract function, since chronic kidney disease is the highest risk factor likely to decrease life expectancy [1]. In addition, raising awareness and education to enhance nephroprotective measurements should be provided to caregivers and patients to minimize risk factors for future renal deterioration.

Conclusion

This study emphasizes the necessity of a multi-disciplinary team, including urology and nephrology, that follows patients with ARM long term, especially within the subgroup of cloaca.

Author contributions LAW wrote the manuscript. LAW, EMG, AB, EHC and JMR analysed the data and EHC and JMR prepared figures 1 - 4. All authors carefully reviewed the manuscript.

Data Availability No datasets were generated or analysed during the current study.

Declarations

Competing interests The authors declare no competing interests.

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