



Sexual function and fertility of adult males with anorectal malformations or Hirschsprung disease

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Abstract

Purpose The long-term impact of anorectal malformations (ARM) and Hirschsprung disease (HD) on sexual function is well recognized but understudied. This study evaluated self-reported sexual and fertility outcomes in adult males with ARM and HD.

Methods This was an IRB approved, prospective study of males in the Adult Colorectal Research Registry who completed surveys between October 2019 and March 2022. Electronic surveys were administered after consenting to being contacted for research. Patients completed the International Index of Erectile Function (IIEF) questionnaire and provided information on fertility outcomes.

Results Sixty-five patients completed outcome questionnaires: 11 (16.9%) had HD and 54 (83.1%) had an ARM. Nineteen patients reported some degree of erectile dysfunction per IIEF criteria, a greater proportion of whom have ARM ($p=0.046$). Twenty (30.7%) have reported having children; there were no differences in rates between HD and ARM patients. Most patients had not attempted to conceive, but eight patients, all of whom have ARM, have pursued fertility investigation or treatments.

Conclusion More patients with ARM reported some degree of erectile dysfunction compared to those with HD. Additionally, some have required fertility treatments. Further investigation is warranted to ensure true low rates of sexual and fertility dysfunction in patients with HD.

Keywords Hirschsprung disease · Anorectal malformation · Sexual function · Fertility · Erectile function

Introduction

Hirschsprung disease (HD) and anorectal malformations (ARM) are rare conditions that are often surgically treated early in a patient's life. As the majority of these patients now survive to adulthood, there is renewed focus on quality of life [1, 2]. However, they are known to experience long-term

sequelae that may impact their health and well-being. Due to their surgical history and congenital conditions, they are at risk for sexual and fertility impairment for both anatomical and psychosocial reasons [1, 3, 4].

Efforts to categorize the fertility outcomes of patients with ARM and HD have been made, but there remains a paucity of data on long-term sexual and fertility outcomes for these patients. Males with ARM have been shown to have preserved erectile and orgasmic function following posterior sagittal anorectoplasty (PSARP) repair, but fertility may be impaired in those with recto-urethral fistulas [5]. There is even less data on these outcomes in males with HD [6]. Erectile dysfunction and infertility have been reported previously in patients with HD, but the rate of these are unclear, and recent data have suggested they have similar sexual outcomes and satisfaction compared to healthy controls [7].

As most patients with ARM and HD reach adulthood, an understanding of their expected sexual and fertility

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outcomes is important, to allow for counseling of parents, and eventually patients, as they age. This study examines the self-reported sexual function and fertility outcomes of a cohort of male patients with ARM and HD.

Methods

This was a prospective study of male patients born with congenital colorectal conditions. Patients greater than 18 years of age were identified in the Colorectal Database, which has been prospectively maintained since 1980 by the senior authors and includes patients from all over the world. Inclusion criteria were a diagnosis of anorectal malformation or Hirschsprung Disease, these patients were contacted by mail to provide consent for enrollment in the Adult Colorectal Research Registry and allow for contact for future research purposes. Patients who live in the European Economic Area were excluded due to inability to meet local research compliance requirements. This study was approved by the Colorado Multiple Institutional Review Board (COMIRB).

Patients were sent electronic surveys through REDCap, a secure, web-based application to capture data for clinical research [8]. Enrollment occurred between October 2019 and March 2022. Patients were asked to provide basic demographic information and complete multiple surveys examining self-reported physical and mental health, information on quality of life, bowel function, urinary function, sexual function, and fertility outcomes. Patients received financial incentives for completing surveys, though this was only offered to patients within the United States due to COMIRB restrictions. Data collected are part of a large-scale long-term outcomes study, but only those who completed the sexual function and fertility outcomes surveys were included in this study.

The International Index of Erectile Function (IIEF) survey was administered to obtain information on sexual function. It is a self-administered 15-item questionnaire, which examines four main domains of sexual function: erectile function, orgasmic function, sexual desire, and intercourse satisfaction. It has been shown to be a well-validated measure of erectile function [9]. Severity of erectile function was defined as severe (score: 6–10), moderate (11–16), mild to moderate (17–21), mild (22–25) and no erectile dysfunction (26–30) as previously reported [10]. An additional question regarding erectile hardness score (EHS) was asked based on prior studies [11]. Questions about relationships and fertility were asked, to ascertain whether they had tried to have children, number of children fathered, and if they required any fertility treatments [5].

Patient demographic data were linked to surgical records to ensure accurate colorectal diagnoses and anatomy, in the case of ARM. Data were reported as mean (standard deviation [SD]) or as frequencies depending on distribution. A subset analyses was performed comparing outcomes between patients with ARM and HD. Among patients with ARM, subset analyses comparing outcomes based on type of malformation and sacral ratio (SR) were performed. Sacral ratio data was provided by the Colorectal Database and was defined as normal (≥ 0.7), intermediate (0.4–0.69), or abnormal (< 0.4). Patients missing data on exact type of malformation, with missing sacral ratio data or who did not complete the sexual function survey, were excluded from the analysis. Differences were tested using *t* test or ANOVA for continuous variables and Chi-squared or Fisher's exact tests for categorical variables with significance set at $p < 0.05$. All analyses were performed using SPSS Statistics 28 (Armonk, NY: IBM Corp).

Results

Overall population

Sixty-five male patients completed the sexual function or fertility outcomes questionnaires and were included in analysis (response rate: 56.5%). Two patients with ARM completed the fertility outcomes questionnaires but did not complete the sexual function questionnaires. The mean age was 31.6 (9.3) years. The majority were of White race (87.7%, $n=57$) and Not Hispanic or Latino ethnicity (90.8%, $n=59$). There were 11 (16.9%) patients with HD and 54 (83.1%) with ARM. The most common type of ARM malformation was recto-perineal fistula (26.0%, $n=14$). All diagnoses and full demographic data are provided in Table 1.

Sexual function outcomes

There were 63 individuals who completed the sexual function surveys. The average (SD) erectile function domain score was 23.5 (9.6) for the overall population, which is categorized as mild erectile dysfunction. Though patients with HD had a higher average score compared to patients with ARM [27.0 (7.7) vs 22.7 (9.9)], there was no statistically significant difference ($p=0.181$). In fact, in all domains, patients with HD had higher average scores than patients with ARM. Differences in sexual desire were higher for patients with HD [8.6 (2.3) vs 7.4 (2.1)] and approached significance ($p=0.090$). When categorizing by severity of erectile dysfunction, a greater proportion of patients with HD had no degree of erectile dysfunction (90.9 vs 65.3%, $p=0.046$). Full data are reported in Table 2.

Table 1 Overall demographics

	Overall Population (<i>n</i> = 65)
Age, year [mean (SD)]	31.6 (9.3)
Race	
White	57 (87.7%)
Black or African-American	2 (3.1%)
Asian	1 (1.5%)
Hispanic/Puerto Rican	1 (1.5%)
More than one race	2 (3.1%)
Unknown	2 (3.1%)
Ethnicity	
Not Hispanic or Latino	59 (90.8%)
Hispanic/Latino	4 (6.2%)
Unknown	2 (3.1%)
Diagnosis	
Hirschsprung's	11 (16.9%)
Cloacal Exstrophy	1 (1.5%)
ARM, unknown type	6 (9.2%)
ARM, recto-urethral bulbar fistula	13 (20.0%)
ARM, recto-urethral prostatic fistula	13 (20.0%)
ARM, recto-bladder neck fistula	3 (4.6%)
ARM, recto-perineal	15 (23.0%)
ARM, no fistula	3 (4.6%)

When stratifying by type of anorectal malformation, there were 46 patients included. Scores for each domain are reported in Table 3. There were no statistically significant differences. Interestingly, based on categorization of severity of erectile function, the average erectile function domain scores for all types of malformation, except cloacal exstrophy and recto-perineal fistula, meet criteria for mild or mild to moderate erectile dysfunction (EF 22–25 or 17–21).

When analyzing by sacral ratio, there were data available for 50 patients with ARM. Most patients had normal SR (62%, *n* = 31), and 19 (38%) had intermediate SR. No

patients in this series had an abnormal SR. When comparing domain scores from the IIEF, there were no statistically significant differences between groups. The greatest difference was in erectile function domain score, though likely not clinically meaningful. Patients with normal SR had an average score of 21.8 (11.1) and patients with an intermediate SR had an average score of 21.2 (11.2) (*p* = 0.982). Full data are reported in Table 4.

Fertility outcomes

There were 65 men who completed the fertility outcomes survey. Overall, 41 (63.1%) reported being in stable relationships. Twenty (30.8%) individuals reported having children and 17 (26.1%) reported having children with their current partner. Eight (12.3%) reported retrograde ejaculation; 7 with ARM and one with HD. Eight (12.3%) individuals reported requiring fertility investigation or treatment. All eight patients have ARM. Two patients reported using sperm donors to conceive children and one patient required testicular sperm extraction due to retrograde ejaculation to conceive. All data are reported in Table 5. There were no statistically significant differences between those with ARM and HD.

On analysis based on type of ARM, patients with all ARM anatomy, except cloacal exstrophy, reported having children. The anatomy of patients requiring fertility interventions or assistance include: recto-urethral bulbar fistula (2), recto-urethral prostatic fistula (2), recto-perineal fistula (1), recto-bladder neck fistula (1), and unknown ARM anatomy (2).

Discussion

This study reports the sexual and fertility outcomes in adult males with anorectal malformations and Hirschsprung disease. In general, we found that patients with ARM had higher rates of erectile dysfunction than those with HD.

Table 2 Sexual function (IIEF Score), overall population and by congenital colorectal condition

	HD (<i>n</i> = 11)	ARM (<i>n</i> = 52)	<i>p</i> value	Overall study cohort (<i>n</i> = 63)	Control values*
Erectile function domain score	27 (7.7)	22.7 (9.9)	0.181	23.5 (9.6)	25.8
Orgasmic function domain score	8.7 (2.9)	7.9 (3.2)	0.448	8.1 (3.2)	9.8
Sexual desire domain score	8.6 (2.3)	7.4 (2.1)	0.090	7.6 (2.2)	7.0
Intercourse satisfaction domain score	12.5 (4.5)	9.6 (6.2)	0.147	10.1 (6.0)	10.6
Overall satisfaction domain score	8.6 (2.1)	7.5 (2.4)	0.164	7.6 (2.4)	8.6
Erectile hardness score	4.0 (0.3)	3.7 (0.8)	0.208	3.7 (0.7)	–

*Values reported by age-matched controls in Rosen et al. initial study using IIEF [9]. Values provided strictly for comparative reference; no statistical analysis performed

Table 3 Sexual function (IIEF Scores), by anorectal malformation

	ARM, no fistula (<i>n</i> = 3)	ARM, recto- perineal fistula (<i>n</i> = 14)	ARM, recto-bladder neck fistula (<i>n</i> = 3)	ARM, recto-ure- thral bulbar fistula (<i>n</i> = 12)	ARM, recto- urethral prostatic fistula (<i>n</i> = 13)	Cloacal exstrophy (<i>n</i> = 1)	<i>p</i> value
Erectile function domain score	20.7 (16.1)	25.6 (8.0)	22.3 (7.0)	22.0 (11.1)	21.4 (10.8)	30.0	0.846
Orgasmic function domain score	6.7 (5.8)	8.1 (7.2)	8.0 (2.6)	7.1 (3.7)	8.0 (3.7)	8.0	0.969
Sexual desire domain score	7.7 (4.0)	7.2 (2.1)	8.7 (0.58)	7.1 (3.4)	7.4 (1.9)	9.0	0.863
Intercourse satisfaction domain score	9.7 (8.4)	12.1 (4.6)	8.7 (7.7)	8.9 (6.8)	8.2 (6.9)	15.0	0.597
Overall satisfaction domain score	9.3 (1.2)	7.7 (2.3)	8.3 (1.5)	8.2 (2.5)	6.5 (3.1)	10.0	0.391

IIEF control values are provided in Table 1 for comparative reference

Table 4 Sexual function (IIEF Scores), by sacral ratio

	SR ≥ 0.7 (<i>n</i> = 31)	SR 0.40–0.69 (<i>n</i> = 19)	<i>p</i> value
Erectile function domain score	21.8 (11.1)	21.2 (11.2)	0.982
Orgasmic function domain score	7.2 (3.6)	7.5 (4.1)	0.765
Sexual desire domain score	6.9 (2.9)	7.1 (2.7)	0.672
Intercourse satisfaction domain score	9.1 (6.4)	9.5 (6.8)	0.835
Overall satisfaction domain score	7.2 (3.1)	6.9 (3.0)	0.685

Table 5 Relationship and fertility outcomes, overall population and by congenital colorectal condition

	Overall Population (<i>n</i> = 65)	HD (<i>n</i> = 11)	ARM (<i>n</i> = 54)	<i>p</i> value
Are you currently in a stable relationship?				0.158
Yes	41 (63.1%)	9 (81.8%)	32 (58.2%)	
No	24 (36.9%)	2 (18.1%)	22 (40.0%)	
Have you had sexual intercourse?				0.379
Yes	53 (81.5%)	10 (90.9%)	43 (79.6%)	
No	12 (18.4%)	1 (9.1%)	11 (20.4%)	
Have you tried to have children with your partner?				0.110
Yes—we have conceived	17 (26.1%)	5 (45.5%)	12 (22.2%)	
Yes—we have not yet conceived	4 (6.2%)	0	4 (7.4%)	
No	44 (67.7%)	6 (54.5%)	38 (70.4%)	
Do you have biological children?				0.247
Yes	20 (30.8%)	5 (45.5%)	15 (27.8%)	
No	45 (69.2%)	6 (54.5%)	39 (72.2%)	
Have you needed any fertility investigations or treatments?				0.245
No—I/We have not needed them (no problems conceiving)	24 (36.9%)	5 (45.5%)	18 (33.3%)	
No—I/We have not tried to have a baby	33 (50.8%)	6 (54.5%)	28 (51.9%)	
Yes	8 (12.3%)	0	8 (14.8%)	

Additionally, though males with either HD or ARM are able to naturally father children, only patients with ARM reported requiring fertility investigations or treatment in this series.

Patients in this series answered the IIEF questionnaire, which examines the four domains of male sexual function [9]. The use of IIEF in patients with HD and ARM has

been reported previously in males: 4.2% of HD patients experienced some degree of erectile dysfunction compared to 13.6% of the ARM population [12]. In contrast, our reported rates were higher; though our HD sample size was smaller than in prior reports, our ARM population size was greater. These numbers contrast to a rate of 5.6% for erectile dysfunction for all males and rates less than 2% for males younger than 39 years of age [13]. Furthermore, it is important to note that classical repairs, such as abdominoperineal or sacroabdominoperineal repairs, have been shown to be associated with erectile and ejaculatory dysfunction and sexual distress [12, 14–16]. Though these repairs are not the surgical approach used by the senior authors, some patients in our series had their initial repair at other institutions, and it is known such operations can impact sexual function and fertility. When comparing ARM and HD patients, our study did not find statistically significant differences in average IIEF scores but did find that differences in sexual desire approached significance. The implication for this is patients with reduced sexual desire should undergo testing for androgen and prolactin levels [8].

Use of the IIEF questionnaire is advantageous as it is well-validated and has been utilized in multiple different patient populations. IIEF scores have been studied extensively in the inflammatory bowel disease (IBD) population; despite obvious differences between these patients and those with HD or ARM, these patients similarly may require multiple colorectal surgeries with pelvic dissection and have chronic medical conditions that can impact multiple domains. Multiple prior studies have shown that IBD patients have lower average IIEF scores compared to healthy controls, with disease activity and depression influencing subscores [17, 18]. Our study showed patients with ARM reported lower average scores in the domains of erectile function, orgasmic function, intercourse satisfaction, and overall satisfaction compared to reported control data. Patients with HD only reported lower average scores in the orgasmic function domain [8]. Though we were unable to detect differences between these cohorts, these data suggest further investigation and follow-up is indicated as patients, regardless of congenital colorectal condition, may have impaired sexual function compared to the unaffected population.

Relationship and fertility outcomes for patients with HD have been reported previously. Our sample size is small with only 11 respondents, but the majority of individuals reported being in a stable relationship and having sexual intercourse. Prior studies have shown an increased risk of sexual issues in adults who have undergone surgery for HD, but that there are no differences in relationship stability for males compared to controls [7, 19]. We also noted that the five patients with HD in our series who wanted to have children were able to do so naturally. There has been

even less data published on fertility outcomes in those with HD, though one prior study reported 5% have experienced fertility issues [20]. It is encouraging no patients in our cohort reported requiring fertility investigations or treatments, but our overall number of HD patients is low and only five have attempted to conceive. One patient in our series with HD reported retrograde ejaculation, but also reported having biological children, indicating his fertility is not fully impaired. Further research to truly understand the fertility outcome and potential in males with HD is warranted.

There exists more published data on the sexual outcomes of males with ARM. Over 90% of patients with ARM have reported normal erections [5]. In our series, the average EHS was 3.7; 84% reported an EHS of 4 and 11% reported an EHS of 3, which is still sufficient for penetration [11]. Therefore, about 5% of our patients reported inadequate erections for penetration. When analyzing by type of ARM, we found no statistically significant differences in sexual function domain scores. Prior studies have shown patients with ARM do not have impaired sexual function compared to controls [5]. Our patients reported values less than controls in most IIEF domains. To our knowledge, this is the first study that investigated the relationship between SR and sexual function outcomes. Our cohort did not include any patients with an abnormal SR, but we found no differences among those with intermediate and normal SR. Prior studies have shown presence of sacral defects does not impact sexual dysfunction and erectile dysfunction in patients with ARM, but these studies did not incorporate SR [21]. It is known that sacral anomalies increase the risk of neurological dysfunction, which can impact sexual functioning [22, 23]. Given that our study does not include those with abnormal SR, we cannot conclusively say SR does not impact sexual function in patients with ARM.

Over half of our patients with ARM reported being in stable relationships and 27% reported having biological children. Prior studies on fertility in patients with ARM have largely focused on female fertility [5, 24]. No differences in fertility rates based on complexity of ARM in males have been reported, with an average childbirth rate of 15% [24]. In our series, eight men required fertility assessment or intervention and two ultimately required donor sperm. There are multiple reasons males with ARM may have impaired fertility, including endocrine abnormalities, increased rates of cryptorchidism, which can cause azoospermia, and potential iatrogenic damage [22–25]. Males with ARM have been shown to have impaired semen parameters including, sperm count, motility and testicular volume compared to healthy controls [22]. Within our series, the two males who required donor sperm had recto-urethral bulbar and recto-urethral prostatic fistulas. It is unclear if there is any association between specific ARM and fertility outcomes or if

it is secondary to iatrogenic causes. For example, during reconstruction for recto-urethral bulbar and recto-urethral prostatic fistulas, the rectum is mobilized from the urinary tract and injury to important structures may not be recognized. Regardless of the cause, as these patients continue to age, further understanding of their hormone levels as well as anatomy will be important for appropriate counseling and care.

Limitations to this study include small sample size, though it is one of the largest studies of self-reported sexual and fertility conditions in patients with congenital colorectal conditions. When trying to stratify the study population based on type of malformation and quality of the sacrum, our study became underpowered and may not have detected differences due to small absolute numbers. Additionally, these data are all self-reported and we recognize there may be responder bias. Finally, because it is self-reported, our data lack objective measures that may better inform fertility potential in these patients.

Conclusion

This study provides self-reported sexual function and fertility outcomes in adult male patients with HD and ARM. Though patients with HD reported lower rates of erectile dysfunction and need for fertility investigations as well as higher levels of satisfaction in sexual domains compared to ARM, our absolute number of patients with HD is small and warrants further study. Patients with ARM reported higher rates of some degree of erectile dysfunction and multiple patients required fertility investigations. There are a variety of factors that may contribute to these challenges. Continued follow-up and study are indicated for both populations to ensure optimal sexual health and fertility outcomes.

Author contributions MLR, AB, and DW were responsible for study design. MLR and JK collected data; MLR performed data analysis. MLR, AB, and DW wrote the main manuscript text. All authors critically reviewed the manuscript.

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Declarations

Conflict of interest The authors declare no competing interests.

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