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Parent Perspectives of Health Related Quality of Life for Adolescents With Bladder Exstrophy-Epispadias as Measured by the Child Health Questionnaire-Parent Form 50™

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Abstract

Purpose—Few groups have examined health related quality of life for adolescents with bladder exstrophy-epispadias. We studied parent reported health related quality of life for adolescents with bladder exstrophy-epispadias using the Child Health Questionnaire-Parent Form 50.

Materials and Methods—We recruited 11 to 17-year-old participants with bladder exstrophy-epispadias and their parents. Parents served as proxy respondents for the adolescents by self-administering a validated generic health related quality of life instrument, the Child Health Questionnaire-Parent Form 50. We collected urinary incontinence, catheterization status, and medical and surgical history data. Mean questionnaire scores were compared to population based norms.

Results—Median age of the 55 patients was 14 years, 69% were male and 84% were white. Diagnoses included bladder exstrophy in 48 cases and epispadias in 7. Of the participants 29 (53%) reported urinary incontinence. The median number of lifetime surgeries was 9. Although physical and psychosocial summary measure scores were comparable to norms, the mean general health perception score was significantly worse than that of a population based sample (65.8 points, 95% CI 61.4–70.2 vs 73, 95% CI 71.3–74.7, $p = 0.004$). Mean family activity and parent emotional impact scores were also significantly worse than in a population based sample (83.6 points, 95% CI 79.3–88.0 vs 89.7, 95% CI 87.9–91.5, $p = 0.02$ and 67.7, 95% CI 61.9–73.6 vs 80.3, 95% CI 78.4–82.2, $p < 0.0001$, respectively). Comparison of incontinent to continent children revealed a lower mean score on the parent emotional impact scale (62.6 points, 95% CI 55.5–69.8 vs 73.4, 95% CI 63.9–82.9), which approached significance ($p = 0.06$).

Conclusions—Although overall adolescent quality of life was comparable to norms, parents reported significantly impaired adolescent general health and family activity as well as a negative parental emotional impact. Further research is needed to identify interventions that can decrease the adverse impact of bladder exstrophy-epispadias on family activity and parent emotional distress.

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Study received institutional review board approval.

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Keywords

urinary bladder; bladder exstrophy; quality of life; adolescent; congenital; hereditary and neonatal diseases and abnormalities

Bladder exstrophy-epispadias is a major congenital structural disorder in which the lower abdominal wall and bladder fail to form normally. Although this leads to urinary incontinence, the intellectual capacity of these children is not usually impaired. Surgical treatment for bladder exstrophy has evolved in the last 50 years, focusing mainly on providing urinary continence and improved cosmesis while maintaining kidney function, sexual function and fertility.¹ Surgical closure of bladder exstrophy is successful in many children but others require multiple surgeries, ranging from bladder re-closure to bladder neck procedures or continent catheterizable reservoirs.¹ While these interventions are implemented to improve the overall well-being of the child, there are few actual studies of HRQOL in this population and no agreed on measure of HRQOL.

Studies of long-term psychosocial outcomes in the bladder exstrophy population have yielded varied conclusions. Some groups found significant problems while others suggested that these children are surprisingly resilient.^{2–6} Using a clinician rated pediatric health status instrument Stjernqvist and Kockum noted lower self-care and general HRQOL in children with bladder exstrophy.⁷ However, there has been little study of self-reported or parent proxy reported HRQOL in these children. Measurement is needed to understand the effects of surgical and medical interventions from the patient perspective to compare different treatment strategies and provide counseling to patients and families about the future.

WHO defines health as a “state of complete mental, physical and social well-being and not merely the absence of disease or infirmity.”⁸ Since that time, the concept of HRQOL has developed into a set of tools for clinical research in clinical trials and in observational studies. HRQOL measures may be generic or disease specific. Generic instruments often benefit from validation in large, population based samples, and extensive research in reproducibility, reliability and responsiveness. They allow comparison of scores across different disease states, permitting greater usefulness in health services research. On the other hand, disease specific instruments are generally developed to assess the impact of the symptoms of a specific disease on HRQOL and, thus, they may be more sensitive to detect changes in disease impact with time. As a compromise, disease specific modules may be added to existing generic instruments to reap the benefits of each type of instrument.⁹

Measuring childhood HRQOL has challenges. Child self-reported HRQOL comes closest to measuring the experience of the child. However, children born with a disease or who grow up with one may have no reference for comparison. Because not all children can reliably self-report health status, parent reported instruments such as CHQ-PF50 have been developed. However, parent and child perspectives of HRQOL may differ and even be discordant, especially for internal feelings as opposed to observable behavior.¹⁰

We measured parent proxy reported HRQOL using CHQ-PF50 in adolescents with bladder exstrophy-epispadias and compared the scores to available population based norms.¹¹ We hypothesized that on CHQ-PF50 parents would report poorer HRQOL in their children than that in the general population and incontinence would predict poorer HRQOL.

METHODS

Study Population

We obtained institutional review board approval for the study. Child assent and parent consent were obtained from all participants. Participants were recruited through the tertiary referral pediatric urology clinic at our institution and an institutional approved bladder exstrophy database. Patients were identified as potentially eligible for study when they had a diagnosis of bladder exstrophy-epispadias and age between 11 and 17 years. Recruitment consisted of a mailed invitation, in person or telephone eligibility screening and a consent process. Study inclusion criteria were age 11 to 17 years, a bladder exstrophy or epispadias diagnosis and the ability to read English at least at a fourth grade level. Foster children or wards of the state were excluded due to institutional policy. A gift card was given to each participant after completing the questionnaire. Children with other diagnoses, including spina bifida and cloacal exstrophy, were excluded from analysis.

Data Collection

Baseline demographics and medical history were obtained by structured interview with the parent and a medical record review. Child reported incontinence, catheterization status and bother level were obtained by structured interview with the adolescent, consisting of a standard set of questions asked of each participant. CHQ-PF50, a generic HRQOL parent proxy instrument, was given to the parents of participants.¹¹ CHQ-PF50 was self-administered at the clinic or completed at home and mailed to the study coordinator.

Child Health Questionnaire-Parent Form 50

CHQ is a set of generic quality of life instruments designed for 5 to 18-year-old children.¹¹ CHQ measures 11 multidimensional scales and 2 summary measures.^{11,12} The parent form is available in 2 lengths (50 or 28 items) and a youth version (87 items) is also available for adolescents 10 to 17 years old. We used the parent form with 50 items, which requires approximately 20 minutes to complete. Scores can be analyzed separately, and CHQ scale scores are combined for an overall physical and psychosocial summary measure score. The scales are scored from 0 to 100 with higher scores indicating better quality of life. The 2 summary measures are transformed so that they may be compared to the mean $\pm$ SD of 50 ± 10 in the general American population.¹¹ A clinically meaningful difference is considered to be 5 points.¹¹

Normative values are available for the scale and summary measure scores, and benchmarks for the parent reported CHQ versions are also available for some conditions.¹¹ Previous groups tested construct validity, convergent validity and sensitivity to change, and internal consistency was 0.62 to 0.94.¹³ CHQ focuses on functioning and in this way it is similar to SF-36®.

Statistical Analysis

Stata® 10 statistical software was used for all analyses. Baseline characteristics were determined for the study sample. Outcome variables were defined as the mean of each CHQ-PF50 summary score and scale. Mean summary and scale scores were determined and 95% CIs were constructed. We compared mean summary measure scores and scale scores between study participants and an available published representative population sample¹¹ using the t test. Power calculations were based on the measurement characteristics of CHQ-PF50. Assuming a SD of 10, a mean of 50 and a 2-sided α of 0.05 we had approximately 94% power to detect a 5-point difference in summary measure means between study participants and the norm.

As a secondary analysis, we compared mean CHQ-PF50 scores between adolescents who did and did not report any urine leakage. However, in the comparison of incontinent vs continent children in our sample of adolescents with bladder exstrophy-epispadias we had only 46% power to detect a 5-point difference. Although the study was underpowered to compare subgroups, we explored associations of covariates, including incontinence status, with CHQ-PF50 scores.

RESULTS

Study Sample

We identified potentially eligible patients between January 2006 and August 2009 from pediatric urology clinic visit lists and an institutional bladder exstrophy database. They were invited to undergo formal eligibility screening and recruitment by letter or in person. Of invited patients 84 underwent formal eligibility screening and were eligible for the study, including 69 who completed the consent process, of whom 55 returned the CHQ-PF50 questionnaire. Nonrespondents were defined as those who refused to participate, did not complete the consent process or failed to return CHQ-PF50 after consenting to the study. When we compared the characteristics of the 29 nonrespondents and the 55 respondents, there were no significant differences in age or number of lifetime surgeries. There was a slightly larger proportion of female nonrespondents than female respondents (41% vs 31%) but this was not statistically significant. There was also a larger proportion of nonrespondents who reported urinary leakage compared to respondents (65% vs 53%) but it was not statistically significant.

Table 1 shows the characteristics of the 55 children in the study sample. In children median age was 14 years (range 11 to 18), 38 (69.1%) were male and 46 (83.6%) were white. The underlying diagnosis was bladder exstrophy in 48 children (87.3%) and epispadias in 7 (12.7%). One patient had a bladder exstrophy variant. The median number of lifetime surgeries was 9 (range 3 to 26) and 45.5% of patients underwent between 8 and 12 surgeries. Of the children 29 (52.7%) reported that they currently leaked urine, 32 (58.2%) performed intermittent catheterization and 23 (41.8%) irrigated the bladder.

Table 1 also lists the characteristics of the 55 responding parents. Median age was 45 years (range 34 to 57), 50 (91%) were female, 52 (94.6%) were white and all were the biological parent of the child. Of the responding parents 41 (74.6%) had at least a college degree, indicating the generally high socioeconomic status of the study population, 83.6% reported working full or part time and 83.6% were married.

CHQ-PF50 vs Population Based Univariate Comparison

Table 2 shows CHQ-PF50 summary measure and scale score means and 95% CIs in the study sample and in a published population sample of 391 American children.¹¹ Physical and psychosocial summary measures did not differ significantly from the norms. The mean scale scores on physical functioning, bodily pain, self-esteem, mental health and family cohesion were also comparable to the norms. The child behavior scale was significantly better than the norm (82.0 points, 95% CI 78.6–85.5 vs 75.6, 73.9–77.3, $p = 0.006$). Despite good scores on those scales the general health perception score was significantly worse than the norm (65.8 points, 95% CI 61.4–70.2 vs 73.0, 95% CI 71.3–74.7, $p = 0.004$). The family activity scale was significantly lower than in the population based sample (83.6 points, 95% CI 79.3–88.0 vs 89.7, 95% CI 87.9–91.5, $p = 0.02$). The parent emotional impact scale was also significantly lower than in the population based sample (67.7 points, 95% CI 61.9–73.6 vs 80.3, 95% CI 78.4–82.2, $p < 0.0001$).

We explored the association of incontinence status with the different CHQ-PF50 summary scores and scales. There were no statistically significant differences between continent and incontinent participants on any scale. However, there was a lower mean score on the parent emotional impact scale in the incontinent group than in the continent group (62.6 points, 95% CI 55.5–69.8 vs 73.4, 95% CI 63.9–82.9, $p = 0.06$).

DISCUSSION

Compared to available, population based CHQ-PF50 norms the parents of adolescents with bladder exstrophy-epispadias reported good quality of life for children but also substantial negative effects on the family life and parent emotional impact domains. There was no significant difference between the study sample and the reference population in overall physical and psychosocial quality of life, as measured by CHQ-PF50. Although parents regarded the general health of the child as worse than that of healthy peers, they reported that the children functioned similarly to peers in physical and psychosocial domains. These findings are consistent with our previous results using a different child reported generic HRQOL instrument, the Child Health and Illness Profile-Adolescent Edition.¹⁴ Similarities to norms suggest that some health outcomes of bladder exstrophy-epispadias are quite good.

In contrast to the positive report of child quality of life, parents self-reported that they experienced emotional distress and family conflicts as a result of the illness of the child. In the subscale measuring emotional impact on the parent the sample mean was 12.6 points lower than that in the population based sample, indicating a statistically significant, clinically meaningful difference. This subscale measures the severity of suffering and worry experienced by the parent due to the health, emotional, behavior or learning problems of the child and it is our most striking finding. The subscale measuring family activity, which deals with the impact of health or behavior problems of the child on family activity and conflict or arguments in the home, was similarly affected since it was an average of 6.1 points lower than the norm.

These findings corroborate the previous study by Mednick et al, who evaluated stress and coping mechanisms in the parents of children younger than 10 years diagnosed with bladder exstrophy.¹⁵ The most common parent stressors in this group were worry about the long-term impact of the illness and helping the child with hygiene needs. In a previous study we also found that parents of adolescents with chronic kidney disease due to urological disorders reported lower CHQ-PF50 physical summary measure scores as well as a negative effect on family activity and parent emotional impact compared to those in a population based reference sample.¹² Many studies show decreased quality of life and psychological suffering in the parents of chronically ill children.^{16,17}

Our study has some limitations, including the relatively small sample size. Although it was adequately powered to compare the study group to the population based mean, power to compare continent vs incontinent patients or adjust for multiple variables was limited. Another limitation is the observational, cross-sectional nature of the study design, which limited our ability to draw causal inferences. Although approximately half of the participants reported some urine leakage, results suggest that the health outcomes of bladder exstrophy-epispadias are quite good with overall HRQOL scores similar to norms. Additional possible explanations for this observation are the resilience and adaptability of children who grow up with a major congenital birth defect. It may reflect in part the generally high socioeconomic status of our population. Also, this generic instrument may not be sensitive enough to detect the influence of urological symptoms such as incontinence or catheterization bother on HRQOL. Another limitation was the measurement of incontinence, defined as any urine leakage as reported by the adolescent. Future studies could use validated patient reported measures of incontinence or develop new measures of bladder symptom severity in the exstrophy population.¹⁸

Despite these limitations our study provides evidence that parents of children with bladder exstrophy-epispadias experience significant negative emotional impact due to the health, emotional, behavior or learning problems of the child. Future research could help elucidate the major reasons for and triggers of this emotional distress in parents. An implication of our findings is the potential to help parents by providing support, dialogue and potentially counseling to help mitigate the emotional burden and stress on families. Further research is needed to identify interventions that can decrease the adverse impact of bladder exstrophy-epispadias on family activity and the emotional distress of parents of affected children.

CONCLUSIONS

In this study of HRQOL in families with an adolescent with bladder exstrophy-epispadias parents reported that children had significantly impaired general health and experienced family activity limitations. They also reported a negative emotional impact on the parent. Nonetheless, overall CHQ-PF50 physical and psychosocial summary scores were comparable to those in an available, population based sample. Further research is needed to understand the child perspective of the effects of bladder exstrophy-epispadias associated urological symptoms on HRQOL.

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Abbreviations and Acronyms

CHQ	Child Health Questionnaire
CHQ-PF50	CHQ-Parent Form 50
HRQOL	health related quality of life

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Table 1

Characteristics in 55 study participants

Characteristics	No. Respondents (%)
<i>Child</i>	
Racial background:	
White	46 (83.6)
Mixed race	7 (12.7)
Black	1 (1.8)
Unknown	1 (1.8)
Hispanic ethnic background	6 (10.9)
No. lifetime surgeries:	
3–7	17 (30.9)
8–12	25 (45.5)
Greater than 12	13 (23.6)
<i>Parent</i>	
Racial/ethnic background:	
White	52 (94.6)
Hispanic	2 (3.6)
Black	1 (1.8)
Education:	
High school/General Educational Development diploma	8 (14.6)
Vocational school or some college	6 (10.9)
College degree	25 (45.5)
Professional or graduate degree	16 (29.1)
Marital status:	
Married	46 (83.6)
Separated	2 (3.6)
Divorced	7 (12.7)
Work status: *	
Working full or part time (outside or home based)	46 (83.6)
Not working due to child health	2 (3.6)
Not working due to other reasons	2 (3.6)
Looking for work outside home	0
Homemaker	9 (16.4)

* Does not total 55 respondents due to option, "select as many as apply."

Table 2

CHQ-PF50 summary measures and scales in 55 study patients with bladder exstrophy-epispadias vs 391 respondents in representative American population¹¹

CHQ-PF50 Summary Measures + Scales	Mean Study Sample (95% CI)	Mean Representative Sample (95% CI)	p Value (t test)
Physical summary measure	50.7 (47.8–53.5)	53.0 (52.1–53.9)	0.0740
Psychosocial summary measure	51.1 (48.8–53.4)	51.2 (50.3–52.1)	0.9501
Physical functioning	94.8 (90.6–99.1)	96.1 (94.7–97.5)	0.5389
Role physical	92.7 (87.0–98.5)	93.6 (91.8–95.4)	0.7491
General health perceptions	65.8 (61.4–70.2)	73.0 (71.3–74.7)	0.0038
Bodily pain	79.1 (72.9–85.3)	81.7 (79.8–83.6)	0.3542
Family activity	83.6 (79.3–88.0)	89.7 (87.9–91.5)	0.0219
Role emotional/behavior	96.2 (92.8–99.5)	92.5 (90.7–94.3)	0.1574
Parent impact–time	90.1 (86.1–94.1)	87.8 (85.8–89.8)	0.4097
Parent impact–emotional	67.7 (61.9–73.6)	80.3 (78.4–82.2)	0.0000
Self-esteem	76.5 (71.3–81.8)	79.8 (78.1–81.5)	0.1991
Mental health	75.1 (71.0–79.3)	78.5 (77.2–79.8)	0.0840
Behavior	82.0 (78.6–85.5)	75.6 (73.9–77.3)	0.0063
Family Cohesion	77.5 (72.1–82.8)	72.3 (70.2–74.4)	0.0947