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Quality of life in female patients with bladder exstrophy-epispadias complex: Long-term follow-up

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Summary

Introduction
Bladder exstrophy-epispadias complex (BEEC) is a congenital malformation that requires multiple surgeries during childhood and life-long follow-up. It often presents with conditions that have the potential to impact quality-of-life (QoL) and psychosocial functioning of affected patients, such as incontinence and sexual dysfunction. The aim of this study is to examine the QoL, urinary continence, sexual function, and overall health in a long-term series of female patients with BEEC.

Method
A retrospective review was performed of female patients with BEEC born between 1964 and 1996. Thirty-three patients were asked to complete four validated questionnaires to evaluate their QoL regarding urinary continence and sexual activity (ICIQ, Potenziani-14, and PISQ-12 questionnaires). Nineteen patients completed and returned the questionnaires. The overall QoL was assessed with the SF-36 questionnaire, and demographics were evaluated. Statistical analysis was performed to compare the general QoL with that of the general population.

Results
The median age of the patients was 26 years (range 18–50) (Table). A low to moderate impact of urinary incontinence on QoL was reported by 30% of patients in the ICIQ. Also as a result of urinary incontinence, 84% of patients reported a moderate to severe impact on their sexual lives. Twelve patients got married with eight gestations and five births. SF-36 reported general QoL comparable with that of the general population in five out of eight items. Differences were seen in the mental health, emotional role, and physical functioning items ($p < 0.001$). The main factors for the differences were poor body image, anxiety, and urinary incontinence. A satisfactory social life was reported by 70% of patients.

Conclusion
Questionnaire studies on BEEC consistently report a high rate of patients not answering, 43% in the present study. The rarity of the disease determines a small sample size, which diminishes statistical power and could potentially conceal small differences with controls. Despite these limitations, the present findings are consistent with previous studies on BEEC with validated QoL questionnaires: adult women with BEEC suffer psychosocial impact mainly from incontinence, and also from gynecological complications, but their resilience and coping mechanisms allow them to achieve a quasi-normal QoL. Female patients with BEEC reported a normal QoL in five of eight items in the SF-36 questionnaire. Urinary incontinence was the main factor for the moderately decreased QoL according to specific questionnaires.

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Mean (range) age, years		26 (18–50)
Previous history	Total number of surgeries, mean (range)	13 (3–16)
Initial surgery, *n*	Primary closure	16
	Cystectomy + ureterosigmoidostomy	2
	Cystectomy + cutaneous ureterostomy	1
	Osteotomy	8
Bladder enlargement or reservoir	Ileal reservoir	33%
	Colonic reservoir	67%
	Mitrofanoff conduit	67%
Lithiasis, *n*	Renal	2
	Bladder	4
Vesicoureteral reflux		36%
Chronic renal failure		6%
Genital prolapse		31%
Rectal prolapse		15%
Continence status, *n* (%)		
Normal continence		12 (63)
Occasional leak		7 (37)
	Mild incontinence, little QoL impact, *n*	5
	Moderate incontinence, big QoL impact, *n*	2
Continence management, *n*	Young-Dees cervicourethroplasty	7
	Silimed perimitrofanoff constrictor	3
	Bulking agents	4
	Closed bladder neck	7
	Autocatheterization	14
	Incontinent	6
Sexual function and fertility	Sexually active	84%
	Not in a relationship	10%
	Dyspareunia	42%
	C-section, *n*	5
	Abortions, *n*	3
Education level, *n*	High school	11
	Higher education	8
Occupational status, *n*	Active workers	12
	Unemployed	5
	Invalidity status	2

Introduction

Bladder exstrophy-epispadias complex (BEEC) is a congenital malformation presenting with pubic diastasis, anterior displacement of internal genitals and labia majora, and protrusion of the bladder through a defect of the lower abdominal wall [1]. It is one of the most severe and challenging conditions in pediatric urology. Most of the affected patients require multiple surgeries during childhood, and life-long follow-up by a specialized multidisciplinary care team. This congenital defect can affect patients' quality of life (QoL) both in the short and long term, and its psychosocial impact is not well understood [2].

Literature evaluating the effect of BEEC on QoL is still very scarce: a meta-analysis by Diseth *et al.* [3] showed that only 0.8% of studies on BEEC look into the psychosocial or QoL implications of the disease. Several studies found BEEC to have a significant impact on mental health and QoL, particularly regarding sexual function and anxiety disorders [2], with patients scoring lower than the general population on questionnaires assessing general health perception and physical activity limitations [4].

Over the last decade, research on BEEC has led to better care management and reconstructive techniques, ultimately improving the morbidity and mortality of BEEC. Patients now reach adulthood aiming for a good QoL, including normal sexual function and acceptable urinary continence.

We designed a retrospective study to evaluate the QoL of adult females with BEEC, including parameters of urinary continence, sexual function, and general health. Our aim is to quantify the overall result of treatment of BEEC and follow-up into functional adulthood, not the result of any particular surgical technique.

Material and methods

The study population was selected from the patients followed at the pediatric unit of a third-level urology-specialized center. Selection criteria included diagnosis of BEEC, female gender, and age >18 years. The study was