

Genital reconstruction in exstrophy patients

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ABSTRACT

Introduction: Surgery for bladder exstrophy has been evolving over the last four to five decades. Because survival has become almost universal, the focus has changed in the exstrophy-epispadias complex to improving quality of life. The most prevalent problem in the long-term function of exstrophy patients is the sexual activity of the adolescent and adult males. The penis in exstrophy patients appears short because of marked congenital deficiency of anterior corporal tissue. Many patients approach for genital reconstruction to improve cosmesis as well as to correct chordee. We report our series of male patients seeking genital reconstruction following exstrophy repair in the past.

Materials and Methods: Fourteen adolescent/adult male patients attended urology services during the period January 2000-December 2009 seeking genital reconstruction following exstrophy repair in the past.

Results: Three patients underwent epispadias repair, four patients had chordee correction with cosmetic excision of skin tags and seven patients underwent chordee correction with penile lengthening. All patients reported satisfaction in the answered questionnaire. Patients undergoing penile lengthening by partial corporal dissection achieved a mean increase in length of 1.614 ± 0.279 cm dorsally and 1.543 ± 0.230 cm ventrally. The satisfactory rate assessed by the Short Form-36 (SF-36) showed that irrespective of the different genital reconstructive procedures done, the patients were satisfied with cosmetic and functional outcome.

Conclusions: Surgical procedures have transformed the management in these patients with bladder exstrophy. Bladders can be safely placed within the pelvis, with most patients achieving urinary continence and cosmetically acceptable external genitalia. Genital reconstruction in the form of correction of chordee, excision of ugly skin tags and lengthening of penis can be performed to give the patients a satisfactory cosmetic and functional system.

Key words: Exstrophy, function, genital reconstruction, outcome

INTRODUCTION

The exstrophy-epispadias complex of genitourinary malformations can be as simple as a glandular epispadias or an overwhelming multisystem defect such as cloacal exstrophy. The male genital defect is severe and is probably the most troublesome aspect of the surgical reconstruction, independent of the decision whether to treat with modern staged closure,

combined closure, or a form of urinary diversion. It was believed that the individual corpora cavernosa were of normal caliber but appeared shorter because of the wide separation of the crural attachments, the prominent dorsal chordee, and the shortened urethral groove. However, Silver and colleagues^[1] described the genital defect for the first time in bladder exstrophy in greater detail. Magnetic resonance imaging (MRI) was used in adult men with bladder exstrophy and compared with results for age- and race-matched controls. They found that the anterior corporal length of male patients with bladder exstrophy was almost 50% shorter than that of normal controls. Although the posterior length of the corporal body was the same as in age-matched controls, the diameter of the posterior corporal segment was greater than in normal controls. It was also found on MRI that the diastasis of the symphysis pubis increased the intrasymphyseal and intercorporal distances but the angle between the corpora cavernosa was unchanged because the corporal bodies were separated in a parallel fashion. Therefore, the penis appears short not only because of the diastasis of the pubic symphysis, as thought in the past, but also because of marked congenital deficiency of anterior corporal tissue.^[1,2] Because survival

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has become almost universal, the focus has changed in the exstrophy-epispadias complex to improving quality of life. Improvement in functional outcomes has become the mainstay of improving the quality of life. It is well recognized that the most prevalent problem in the long-term function of exstrophy patients is the sexual activity of the adolescent and adult males,^[3,4] but several studies^[5-7] show that despite a significant functional handicap an acceptable cosmetic appearance and adequate potential for sexual activity can be achieved in most patients, with preserved fertility, despite psychosexual development being inevitably affected. We report our series of male patients seeking genital reconstruction following exstrophy repair in the past.

MATERIALS AND METHODS

Fourteen adolescent/adult male patients attended urology services during the period January 2000–December 2009 seeking genital reconstruction following exstrophy repair in the past. A detailed history of previous treatment was noted. Information pertaining to the timing and method of bladder closure, outcome of initial surgery, subsequent operations, other hospital in-patient episodes and eventual status of the urogenital system were recorded from previous hospital records. All the patients were interviewed after obtaining a written informed consent, with an understanding that privacy and confidentiality issues would be maintained. The interview consisted of a pre-constructed semi-structured questionnaire covering demographic data, previous medical and surgical data, childhood/school/other educational information, employment information, relationship history, assessment of physical and sexual function, and where appropriate a clinical examination. Each interviewed patient was asked to complete an Short Form-36 (SF-36)(v2) health and general well-being assessment questionnaire.

The patients were examined in detail and assessed for urinary continence, erectile function and ejaculation as and when felt necessary or demanded by the patients themselves. All patients underwent routine laboratory tests which included urinary examination and culture sensitivity. Routine ultrasound imaging of the urinary tract was carried out in all. Computed Tomography (CT) and/or MR imaging [Figure 1a, b] was carried out whenever felt necessary. Urinary continence/incontinence was assessed by assessing voiding diary, post-void residual volume, uroflowmetry and cystometry. Erectile function was assessed by doing the provocative rigiscan, using visual stimulus.

All patients were counseled for erectile function, ejaculation and fertility. Genital reconstructive surgery was planned after detailed discussion with the patients. Patients were counseled regarding the results of penile lengthening and individual expectations. In patients needing correction of epispadias, the patients underwent a modified Cantwell-Ransley procedure. Skin chordee was corrected by single

or double Z plasty [Figure.2a, b]. Wherever scar tissue was the cause of the chordee, scar tissue around the urethra and corporal bodies was cleared to release the chordee [Figure 2c]. Penile lengthening was carried out by partial dissection of the corporal bodies up to the inferior border of the pubic rami and approximation of the corporal bodies in the midline [Figure 3a, b]. Following surgery the patients were followed up, reassessed and interviewed in relation to general health.

RESULTS

During the 10-year study period, 14 patients aged between 18 and 27 (mean age 23 years) approached the urological services for genital reconstruction. Nine of these had undergone treatment at our centre previously. Six of these had undergone staged exstrophy repair in childhood and the remaining three had presented in adulthood with exstrophy and had undergone cystectomy with catheterizable modified Mainz pouch. The remaining 5/14 patients had undergone staged exstrophy repair at other centers.

Eleven out of 14 patients who had undergone staged

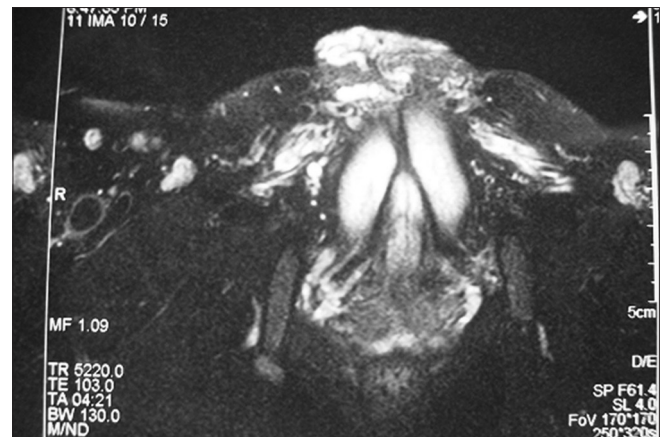


Figure 1a: MRI imaging of corporal bodies

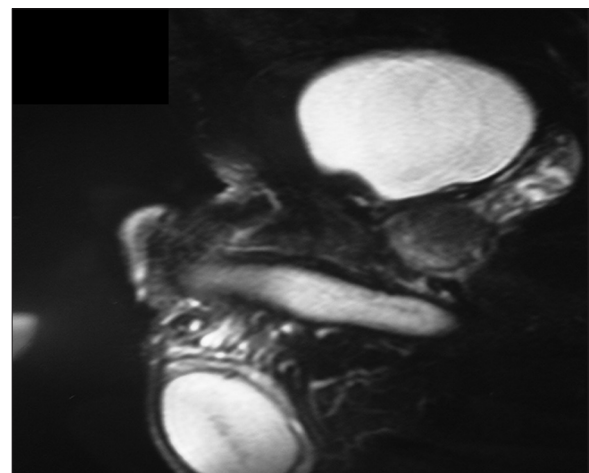


Figure 1b: Anterior segment of the corporal body is short in comparison with the posterior segment



Figure 2a: Dorsal chordee with the skin band

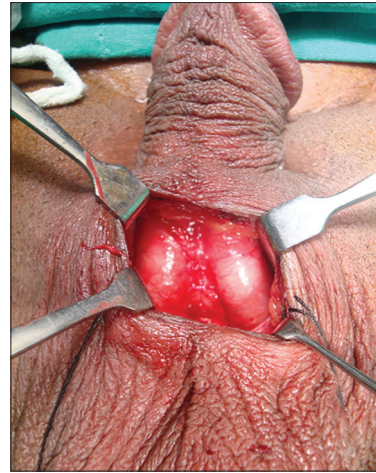


Figure 3a: Dissection of the corporal bodies



Figure 2b: Double Z-plasty

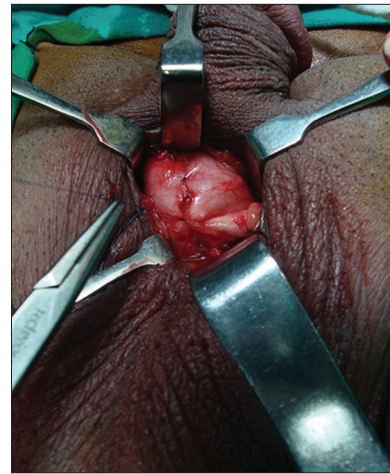


Figure 3b: Midline approximation of the partially dissected corporal bodies



Figure 2c: Postoperative straightening of the penis

exstrophy repair in childhood, had each undergone the repair in 3.9 ± 0.73 stages (range 3–5). Seven of these 11 patients presented to us for chordee correction with penile lengthening. All these patients had dorsal chordee and short broad penis. The remaining four patients presented to

us for chordee correction and excision of skin tags for a good cosmetic appearance. All these 11 patients were dry and continent during daytime, with a bladder capacity of 200 ml. Most of them experienced occasional night-time enuresis. None of these patients wanted any further treatment for their Grade I stress incontinence and were satisfied with the outcome of previous surgery. These patients were experiencing good erections and sought treatment as they were interested in marriage and sexual relations.

Three out of 14 patients who had undergone cystectomy with continent diversion presented to us for epispadias repair, lengthening of penis and chordee correction. All these three patients had the Mainz pouch with a capacity of 350–450 cc and needed to catheterize once in 4–5 h. All patients remained dry during daytime and only one of these patients complained of occasional night-time urinary leak. All these three patients had improved self-esteem, confidence and social interaction. All these three patients were experiencing good erections and were interested in getting married and having sexual relations.

Three patients underwent epispadias repair, four patients underwent chordee correction with excision of skin tags and seven patients underwent chordee correction with penile lengthening procedures. The results of the same are shown in Table 1. Patients undergoing penile lengthening by partial corporal dissection achieved a mean increase in length of 1.614 ± 0.279 cm dorsally and 1.543 ± 0.230 cm ventrally. There was no significant increase in penile length in those patients who underwent epispadias repair and those who underwent chordee correction alone. The satisfactory rate assessed by the SF-36 showed that irrespective of the different genital reconstructive procedures done, the patients were satisfied with cosmetic and functional outcome. Patients who were interested in having a longer penis were advised to use vacuum erection device for passive stretching of corpora. The results following the use of these devices have not been assessed.

Two of these 14 patients are married and have normal erections and sexual function. Three others are in a relationship and

are having normal erections and sexual function. Four of these patients are experiencing proper ejaculation. Till last follow-up, no pregnancy has been reported.

DISCUSSION

The anatomy of the exstrophy pelvis has been investigated clinically, by cavernosography, CT, MRI, by experimental models and by dissection.^[8] The pelvic ring is open anteriorly. The pubic bone is poorly formed and 30% shorter than normal.^[9] The two halves of the pelvis are rotated downwards by 12° so that the inferior pubic ramus (to which the corpora are attached) is nearly parallel with the floor when the patient is standing upright. Neonatal osteotomy does change this orientation to a certain degree, even though the symphysis is not completely closed. However, the pelvic floor remains flatter than in normal babies in the coronal plane (103 vs. 80 degrees), whilst being the same when seen in the sagittal plane.^[9]

Table 1: Patient Characteristics.

No	Age	No of previous surgeries	Genital Reconstruction	Dorsal Penile length		Ventral Penile length		Increase in length (Dorsal)	Increase in length (Ventral)	P value	SF-36 Score
				Preop	Postop	Preop	Postop				
1	18	3	Epispadias repair	4.0	4.1	6.0	6.1			NS	2740
2	20	5	Epispadias repair	3.5	3.7	5.0	5.2			NS	2780
3	22	3	Epispadias repair	3.8	4.0	5.4	5.6			NS	2800
mean 2773.3 ± 30.55											
4	24	5	Chordee correction, scar excision	4.0	5.0	5.4	6.4				2840
5	26	3	Chordee correction, scar excision	3.9	4.9	5.5	6.5			NS	2780
6	26	5	Chordee correction, scar excision	5.0	6.0	6.8	7.7			0.0342 0.1018	2740
7	26	3	Chordee correction, scar excision	4.6	5.6	6.6	7.6				2780
mean 2785 ± 41.23											
8	19	4	Chordee repair, Penile Len	4.0	5.2	6.0	7.2	1.2	1.2		2760
9	21	4	Chordee repair, Penile Len	3.9	5.8	5.8	7.6	1.9	1.8		2760
10	23	4	Chordee repair, Penile Len	4.2	5.7	6.0	7.5	1.5	1.5		2790
11	25	4	Chordee repair, Penile Len	4.8	6.4	6.0	7.8	1.6	1.8	0.0001	2820
12	27	4	Chordee repair, Penile Len	4.5	5.9	6.3	8.0	1.4	1.7		2800
13	25	4	Chordee repair, Penile Len	4.3	6.0	5.9	7.3	1.7	1.4		2760
14	27	4	Chordee repair, Penile len	4.8	6.8	6.4	7.8	2.0	1.4		2740
								1.6 ± 0.27	1.5 ± 0.23		mean 2775.7 ± 27.18

Stec and colleagues^[10] using three-dimensional (3-D) models created from CT scans of children with classic bladder exstrophy and normal age-matched controls, found that the puborectal slings were supporting two times more body cavity area than normal. The levator ani group is positioned more posteriorly in exstrophy patients, with 68% located posterior to the rectum and 32% anterior (versus 52% posterior and 48% anterior in healthy controls). The levators are also rotated outward 15.5 degrees, and in the coronal aspect the levators are 31.7 degrees more flattened than normal. This deviation from normal makes the exstrophy puborectal sling more flattened than its normal conical shape. There was no significant difference in the length or thickness of these muscles between patients with exstrophy and normal controls.^[10]

The cavernosograms and CT imaging in exstrophy patients have revealed the reason for the shortness of the penis in comparison with those of normal men. It is not, as might be believed that the open symphysis is the reason for the penis to be buried in the perineum, but the corpora are actually deficient in length, though of normal caliber. Silver and colleagues^[1] used MRI in adult men with bladder exstrophy and compared with results for age- and race-matched controls. They found that the anterior corporal length of male patients with bladder exstrophy was almost 50% shorter than that of normal controls. However, although the posterior length of the corporal body was the same as in age-matched controls, the diameter of the posterior corporal segment was greater than in normal controls. It was also found on MRI that the diastasis of the symphysis pubis increased the intrasymphyseal and intercorporal distances but the angle between the corpora cavernosa was unchanged because the corporal bodies were separated in a parallel fashion. Therefore, the penis appears short not only because of the diastasis of the pubic symphysis, as thought in the past, but also because of marked congenital deficiency of the anterior corporal tissue.^[1]

It is obvious from our study and other studies^[11] that exstrophy/epispadias patients seek medical help to improve the appearance and functional aspect of the penis. The most common deficiencies of the penis that the patients complain of are dorsal chordee or bending and the short appearance of the penis. These deficiencies have been well-recognized and surgical remedies are emerging to be accepted as standard. In some patients the corpora may have been surrounded or involved in contraction and scarring of the urethral plate and pericorporeal tissue. Lengthening of the penis can be achieved by clearing all the scar tissue away and cleaning the corpora.^[12] This dissection is the first step in chordee correction, however, in our experience this step has not allowed any penile lengthening. The inferior pubic rami in these patients lie in a horizontal plane preventing lengthening of the penis by partial detachment of the corpora. Some length can be gained by dissection from the body of the pubis, however it may not be useful to continue

dissection posterior to the junction with the inferior pubic rami. Kelley^[13] achieved good lengthening in 9/11 children with complete detachment of the corpora from the inferior pubic rami, however, follow-up was not reported. Such a procedure could lead to damage to the blood supply reaching the corpora through Alcock's canal. Patients in our series also who underwent chordee correction and penile lengthening achieved adequate lengthening. Lue and El-Sakka^[14] evaluated the results of chronic intermittent stretching with a vacuum erection device in four patients with penile shortening from severe Peyronie's disease.

At the six-month follow-up three patients had each gained 2 inches but had decreased erectile rigidity. All patients were satisfied. Whether vacuum erection device would help patients with exstrophy, one will have to await trials, however Kazem *et al.*,^[15] reported that the vacuum erection device was not an effective method for penile elongation.

It is well recognized that the most important problem in the long-term function of exstrophy patients is the sexual activity of the adolescent/adult males.^[3,4] With or without correction, patients have a normal libido. If the penis is straight enough, most of these individuals are able to have penetrative sexual relations. Mesrobian *et al.*,^[6] reported that 83% of men with reconstruction in infancy had satisfactory intercourse. Exstrophy/epispadias patients are known to have fewer casual sexual partners than would be expected and are known to form very stable partnerships with normal women and have a normal family life.

Patients are known to show interest and feelings towards long-term relationships, marriage and family, however, the physical aspects of their exstrophy condition seemed to pose some restriction on their relationships with the opposite gender.^[11] Men are known to have achieved paternity with their partners, with no need for assisted fertility techniques.^[11] In a review of 44 patients with exstrophy, Woodhouse^[16] reported that seven had initiated one or two pregnancies from which there were five children. Similarly Mesrobian *et al.*,^[6] reported that 12 of the 53 patients were married, five of whom had fathered children. The main cause of infertility appears to be repeated prostatic and bladder infections.

Several studies^[5-7] have shown that despite a significant functional handicap, one can achieve good acceptable cosmetic appearance along with good and adequate potential for sexual activity. The patients in our study were satisfied with the appearance of their penis and genitalia and reported satisfactory erectile function. Today surgical techniques have transformed the management of exstrophy in children, from an era of urinary diversion to the present day wherein lower urinary tract reconstruction allows good quality bladders to be safely placed deep into the pelvis. Urinary continence is achieved in most patients and cosmetically acceptable genitalia can be expected to allow normal sexual function.

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