

# Exstrophy epispadias complex- Issues beyond the initial repair

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## ABSTRACT

Despite advances in the management of exstrophy epispadias complex (EEC), the quality of life of these patients is far from good. The post-operative period is complicated by numerous and variable events - infection, dehiscence, upper tract dilatation with deterioration, fistulas, stone formation and incontinence to name a few of the major complications. Redo surgery for bladder closure, bladder neck reconstruction, epispadias repair and closure of fistulas are frequently required. The current focus is on limiting the frequency and morbidity of the reconstructive procedures. A successful initial closure and early satisfactory cosmetic and functional results are gratifying for the family and the health care team, but this is only the beginning of the lifelong care necessary for bladder exstrophy (BE) patients. In this article, the long-term outcome of various treatment options and the continent procedures in BE has been reviewed, tracing the journey of these patients into adolescence and adulthood.

**Key words:** Exstrophy, epispadias, incontinence, bladder neck, ureterosigmoidostomy

## INTRODUCTION

Classic bladder exstrophy (BE) is reported to occur on an average in 1:10,000 to 1:50,000 live births. Although, the philosophy and technical background of almost all the operations had been described at least two centuries ago, the earliest form of modern staged reconstruction of exstrophy (MSRE) was developed in the 1970s only.<sup>[1]</sup> Subsequently, complete primary repair of exstrophy (CPRE) and radical mobilization of soft tissues (RMST) have also been described to treat BE.

Despite differences in the initial approach to bladder closure, the objectives of treatment have been to achieve a secure closure of the bladder, pelvis, and the

abdominal wall; preservation of renal function; provision of urinary continence; and creation of functional and cosmetically appealing genitalia.<sup>[2]</sup> Over the years, the management goals have shifted beyond survival towards the improvement of cosmetic and functional outcomes.

## BLADDER TEMPLATE, BONY PELVIS AND PELVIC MUSCLES

In addition to the exstrophic bladder plate and urethra, the EEC has a malformed bony pelvis and pelvic floor. A proper evaluation with examination under anaesthesia must be performed to assess the suitability of the bladder template for closure. Small templates may need to be delayed and given several months to grow.<sup>[3]</sup> Grossly, the pelvis in BE is laterally splayed leaving a large defect in the pelvic floor and no support for the genitourinary and gynaecologic organs. The pubic bones are shortened by approximately 30%.<sup>[4]</sup> These deformities lead to a waddling gait, premature hip osteoarthritis, and outward lower limb rotation and foot progression angle of 20-30°; the defect decreases rapidly during childhood and persists only slightly. Re-approximation of the bony pelvic ring has been demonstrated to be critical to the long-term successful reconstruction of BE.<sup>[5]</sup>

The chief component of active support of the pelvic floor is the pelvic diaphragm. In exstrophy patients, the pelvic floor is more flattened with anteriorly displaced anus and

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the anterior segment of the levator is shorter with a greater proportion of the puborectalis sling being posterior to the rectum. This represents a paucity of tissue available for the support of genitourinary and the gynaecologic pelvic organs. In addition, other pelvic muscle groups are also diverged laterally.<sup>[6,7]</sup>

## OUTCOME OF PRINCIPAL OPERATIVE PROCEDURES

1. Modern Staged repair of exstrophy (MSRE) is the most extensively performed and reported procedure, making it the standard with which all other repairs are compared. Timing of various stages of reconstruction of MSRE has been refined so as to achieve not only the goals of closure but also to decrease the technical difficulties.<sup>[3]</sup>

Continence is the single most relevant parameter when the long term outcome is assessed. However, the lack of standardized definitions for reporting continence makes the comparisons difficult. In a report on 108 patients treated with MRSE, overall 72% of patients were continent which was defined as being dry for greater than 3-hour intervals during the day, dry at night, able to voluntarily void without catheterization, and no need for augmentation. Interestingly, 6 of the girls (15%) became dry immediately after primary closure alone and 19 patients (20%) remained incontinent even after bladder neck reconstruction (BNR), with one-half requiring a continent diversion.<sup>[8,9]</sup>

The Indiana group reported on its exstrophy series defining continence as dry interval greater than 3 hours, no stress incontinence, and dry at night. They found that 4 of 48 patients (8%) before bladder neck reconstruction, 7/48 (15%) after bladder neck repair, and 30/48 (63%) after bladder neck reconstruction and bladder augmentation were continent. The overall 85% continence rate achieved in this series relied quite heavily on augmentation (69%) and clean intermittent catheterization (67%).<sup>[10]</sup>

Evolution in the surgical management of exstrophy bladder is reflected in a report by Mouriquand *et al.* who achieved overall continence rate of 89% after procedures such as bladder augmentation, Mitrofanoff conduits, urethral slings and artificial urinary sphincters when the conventional BNR did not yield the desired results.<sup>[11]</sup>

2. Complete primary repair of exstrophy (CPRE) consisted of bladder closure with a penile disassembly epispadias repair. BNR is performed in only those CPRE patients who require such a repair. In one series, it has been shown that BNR was necessary in 80% of male and 57% of female patients after CPRE.<sup>[12]</sup> In this report of 39

patients, 74% of the toilet trained patients (17/23) had achieved daytime continent intervals of 2 or more hours. Patients attained continence after CPRE alone (22%) or required an additional procedure such as Mitchell bladder neck repair (17%), bladder neck injection (13%) or both bladder neck repair and injection (22%). Only one of the children required bladder augmentation, whereas 4 used intermittent catheterization with a Mitrofanoff conduit.<sup>[12]</sup>

In a review of 32 exstrophy closures performed in Boston Children's Hospital using CPRE, only 19% (4/21) of children older than 4 years of age were dry after CPRE alone and 4 required BNR. One of those 4 developed dry intervals greater than 3 hours with the rest experiencing dry intervals of 2–3 hours.<sup>[13]</sup>

3. The Kelly's repair involves radical mobilization of the internal and external sphincter muscles of the bladder and urethra to provide a continence mechanism.<sup>[14]</sup> The data on 31 patients with Kelly's repair from Melbourne showed 6 patients to be completely continent. Four more patients became dry with augmentation and intermittent catheterization and with the inclusion another 11 partially continent patients, the continence rate of 70% was reported. The pelvic organ prolapse may also decrease in women with BE after the Kelly's technique.<sup>[14]</sup> The Great Ormond street Hospital reported their initial outcomes with Kelly's repair. Day and night time continence for patients was reported as 73% and 25%, respectively. The continence improved greatly with duration of the follow up suggesting that normal cycling of the bladder has an advantageous effect on ultimate bladder function.<sup>[15]</sup>
4. Many patients with EEC continue to experience urinary incontinence even after BNR, augmentation and intermittent catheterization with or without closure of the bladder neck. In such children, Mainz II ureterosigmoidostomy pouch has been used taking advantage of the good rectal continence in these children. Pouch is a diversion procedure and normal per urethral voiding is not considered a goal. There were no major peri-operative complications reported, but the overall late complication rate was 25%, including bowel obstruction, urinary calculi, and incisional hernias.<sup>[16]</sup> Advantages of this approach are very high continence rates, lower reoperation rates and technical ease. However, the frequency of stooling can be detrimental to the lifestyle and the metabolic dysfunction is very likely unless lifetime replacement therapy is provided. In a study of 52 individuals, who had undergone ureterosigmoidostomy before 1990, a significant risk of urofecal incontinence and pelvic organ prolapse was identified.<sup>[17]</sup> Although risk of tumors has not been a problem in the most recent literature, it needs long-term studies in children. It may not be the primary option but does have a place in children born with very small bladders, exstrophy

cripples, and those with limited access to professional medical resources.

## ROLE OF OSTEOTOMY

Recreating the bony pelvic ring and mobilizing the pelvic floor to place the closed vesicourethral unit deeply in the levator hammock are the critical steps during repair of BE. The main role of osteotomy appears to be to relax tension on the repaired bladder and abdominal wall during the process of wound-healing.<sup>[18]</sup> Current data suggest that although osteotomy may promote continence by restoring the sling of the pelvic floor muscles around the urethra, the position of the pubic rami does not contribute greatly to the achievement of continence. Osteotomy does have an associated complication rate of 4% with one-half of these being neurological complications.<sup>[19,20]</sup> Therefore, it is advisable to refrain from performing osteotomy unless it is necessary for the successful closure of bladder and abdominal wall. Osteotomy is recommended in patients beyond the neonatal age because the procedure is associated with an improvement in post-operative continence as compared to soft tissue closure alone. This appears to be due to a better restoration of the pelvic floor muscles and not just the approximation of the symphysis pubis alone.<sup>[19]</sup>

## INDIAN SCENARIO

There have been a very few reports of long-term follow up on BE from India.<sup>[21-24]</sup> There have been excellent contributions describing newer concepts in embryology, better repair of abdominal wall, scrotal skin transfer for improved perineum, umbilicoplasty, use of muscle stimulators, role of ureterosigmoidostomies, three-loop technique for anterior pubic fixation and application of urodynamics in exstrophy management.<sup>[25]</sup> In a recent report of 210 exstrophy patients from 1984-2010, 105 patients had socially acceptable continence: 43%(57/132) from the bladder neck reconstruction group and 85%(48/56) from the bladder augmentation group. Fifteen patients underwent permanent urinary diversion by either ureterosigmoidostomy or colon conduit. The authors concluded that it is possible to provide all the patients with socially acceptable continence with bladder neck division and catheterizable continent stoma as the last resort. Anticholinergic medication with imipramine or oxybutinin is a useful adjunct in the overall management.<sup>[24]</sup>

In two other reports, consisting of 12 patients after primary turn in of bladder and 31 patients after modified BNR respectively, persistent bladder abnormalities were detected on urodynamic assessment and were thought to be the primary reasons for the failure of the bladder to increase in size and persistence of incontinence.<sup>[25,26]</sup>

## ONGOING PROBLEMS

The need for surgical correction in this condition often continues through adolescence and into adulthood.<sup>[27]</sup> The plight of these patients forces them to visit hospitals repeatedly and they are likely to have long-term medical and psychosexual dysfunction. Urinary continence with ability to void spontaneously and efficiently directly determines the quality of life (QOL) of these patients. All other parameters of QOL are, in turn, affected by it. In addition to social continence, anxiety about the genital appearance and performance, and psychosocial isolation are the ongoing issues of concern affecting meaningful integration of these children into the society.

## CONTINUING INCONTINENCE

Achieving urinary continence is now possible in most children. Persistent incontinence secondary to inadequate bladder outlet resistance in BE has been traditionally corrected with BNR. However, there was an inability to maintain the initial level of continence achieved after BNR. Decompensation of the bladder wall due to increased bladder-outlet resistance has been thought to be responsible for this phenomenon.<sup>[28]</sup>

Further strategies for the management of ongoing incontinence include bladder neck closure and continent catheterizable abdominal channel with bladder augmentation, intestinal neo-bladder formation with continent catheterizable channel, intestinal cutaneous urinary diversion, or continent rectal pouch/ureterosigmoidostomy. In a series of 32 exstrophy patients undergoing salvage continence surgery, the reasons for failure of initial reconstruction were poor bladder capacity or failed BNR, and upper urinary tract deterioration. With careful patient selection, 29 patients could achieve continence with a variety of salvage surgical techniques.<sup>[29]</sup>

It has been suggested that the onset of puberty may improve continence secondary to the prostatic growth and many tend to defer definitive treatment in those failing initial reconstruction. However, evaluation of the prostatic size and configuration by magnetic resonance imaging does not support an anatomic contribution to the outlet obstruction by post pubertal prostate enlargement.<sup>[30]</sup> Anecdotally, meaningful continence is unlikely to develop if periods of 3-hour dry intervals are not achieved within 1-2 years after BNR.

Bladder augmentation and continent outlet construction are the pillars of optimal success in patients who have failed BNR or whose bladder is not adequate for reconstruction.<sup>[31]</sup> Frimberger and investigators reported 19 patients with failed prior continence diversions. The commonest

reason identified for failure of these techniques was de-intussusception and ongoing bladder neck incompetence. In most of the cases, bladder neck closure and creation of a catheterizable abdominal continent channel led to reliable continence.<sup>[32]</sup>

Permanent urinary diversion dramatically changes the quality of life of many children. It remains a therapeutic option when incontinence interferes with social life or the renal function is compromised. Stein *et al.* demonstrated continence in 95% of a group of 128 exstrophy-epispadias patients who had undergone Mainz uretero-sigmoidostomy. During a 10-year follow-up, the renal function was preserved and no malignancies were reported.<sup>[33]</sup> In a series of 13 patients of ureterosigmoidostomy from Switzerland, majority of the patients had normal or mildly impaired renal function and a well functioning ureterosigmoidostomy after a mean follow up period of 50 years.<sup>[34]</sup> The long-term cancer risk of ureterosigmoidostomy in young patient population is feared to be high. A report consisting of series of 103 neoplasms showed that the risk of malignancy in this group of patients is 700 times greater than an age-matched general population.<sup>[35]</sup> Surveillance is critical for the patients undergoing this procedure.

## GENITAL APPEARANCE AND SEXUAL FUNCTION

In patients with EEC, the issues such as sexuality, sexual function and fertility gain more importance once these patients advance to puberty and adulthood. The aesthetic aspects of the genitals acquire greater significance with age and may affect the self-esteem.

Penile appearance and organ length are of big concern, especially after puberty. There is a question whether, the incomplete distal disassembly of the modified Cantwell Ransley's repair prevents corporal lengthening to their optimal size.<sup>[36]</sup> However, the undersized phallus and the chordee are caused by the anatomical deficiencies of corporal length and the corporeal disproportion.<sup>[37]</sup> Also, ventral plication is an option to correct chordee and will always lead to penile shortening because of corporal reduction. Therefore, it appears that proximal mobilization together with corporal grafting is potentially the most effective on penile lengthening. If the symphysis pubis is too wide, approximation with the help of osteotomies will bring the corpora more medial and advance the penis out of the pelvis, however, the length of neurovascular bundles is the limiting factor.<sup>[38]</sup>

Excellent aesthetic outcomes can be achieved by clitoroplasty in EEC female patients by using a variety of surgical techniques. In a study of 26 female patients; 3 isolated epispadias and 23 with bladder exstrophy, a total of 33 clitoroplasties were performed. Twenty-three of the 26 patients (88%) had a satisfactory or excellent aesthetic outcome following clitoroplasty. Authors recommended a

'second look' reassessment near puberty to identify poor cosmesis and offer secondary clitoroplasty to improve aesthetic outcome.<sup>[39]</sup>

Heterosexuality is usually expressed in both the sexes and most of them have adequate sexual function. Urinary diversion in some series seems to result in better ejaculatory hence, fertility outcome in male patients. Recent series have shown equally good results with primary reconstruction. Most of the female patients have normal fertility while male patients have significantly low fertility.<sup>[40]</sup> Nineteen men born with classic BE were evaluated, out of which 11 (58%) presented with erectile dysfunction as compared to 9 (23%) age matched controls. Erectile dysfunction was more common in patients with bladder exstrophy who underwent multiple continence surgeries. Orgasmic function was also significantly lower in patients with bladder exstrophy than in controls. No difference was observed between the groups in the sexual desire, sexual satisfaction and overall satisfaction domains.<sup>[41]</sup>

In a review of 21 adult male exstrophy patients, most of whom had been managed with single stage exstrophy closure in childhood, Ebert and associates reported good erectile function in all and normal ejaculation in 19 patients. The authors suggested that single-stage reconstruction yields better ejaculatory function. Successful fatherhood was however, not reported.<sup>[42]</sup> Mathews *et al.* reported on urogynaecological and obstetrical issues in 83 adult female exstrophy-epispadias patients. Sixteen were sexually active and 8 had managed 13 successful pregnancies in total. Five women reported that they had occasional, restricted sexual activity due to dissatisfaction with their genital appearance.<sup>[43]</sup>

In a long-term retrospective study of 100 patients with EEC (76 boys and 24 girls), genital satisfaction and genital touching were rated low, and avoidance of nudity in public areas was common. It is noteworthy that 93.9% expressed an interest in psychological assistance.<sup>[44]</sup>

## PSYCHOSOCIAL PROBLEMS

It is impossible to quantify the potential psychological effects of multiple surgical procedures on a child, particularly considering the high incidence of complications that accompany each stage of reconstruction. In 1996, in a long-term follow-up study, it was seen that the children with exstrophy do not have clinical psychopathic disturbance although alterations in adaptive and acting-out skills exist.<sup>[45]</sup> However, Reiner *et al.* recognized that children with exstrophy have more noticeable behavioural and developmental problems as compared with their peers and the psychosexual dysfunction and anxiety were universal in adolescent boys. The boys with genital anomalies should undergo an assessment for psychosexual development impairment.<sup>[46]</sup>

The main predictors of mental health were parental warmth and patient genital appraisal in the 11 to 14-year age group, and parental warmth and urinary continence function in the 15 to 20-year age group. Out of a total of 22 adolescents, 19 with bladder exstrophy and 3 with epispadias, 9 (41%) were urinary incontinent, 6 (27%) had persistent fecal staining. Ten (59%) were dissatisfied with the penile appearance and 11 (50%) met the criteria for psychiatric diagnoses.<sup>[47]</sup> The continence issues and dissatisfaction about genital appearance had a negative impact on the mental health and psychosocial functioning in exstrophy patients.<sup>[45]</sup> Wilson and co-workers reported that adolescents with exstrophy have strong personality traits and developed a range of creative coping strategies to meet the demands of living with this chronic health condition.<sup>[48]</sup>

The exstrophy patients, when integrated into the mainstream, are robust, healthy, and well adjusted, functioning well in the society. Baird *et al.* reported on a long-term outcome study in 15 subjects born with exstrophy. All of those interviewed had attended mainstream school, and 13 achieved examination success. Eleven worked full time. Five of the group were married, and 5 were in long-term relationships.<sup>[49]</sup> However, in classic bladder exstrophy, clinically significant tendencies to develop suicidal behaviour have been found in males in all age groups and screening for such tendencies in the at-risk groups is recommended. On the other hand, affected females have more close friendships and fewer disadvantages in relation to the healthy female peers.<sup>[50,51]</sup>

Only a few groups have examined the health related quality of life for adolescents with bladder exstrophy-epispadias. The mean general health perception score, mean family activity and parent emotional impact scores were significantly worse than in a population based sample.<sup>[52]</sup> Though, further studies are required to elucidate the interventions needed to decrease the adverse impact, the functional results seem to be the most likely predictive factor of health-related QOL score.<sup>[53]</sup>

## ORTHOPEDIC PROBLEMS

Gait abnormalities are seen in exstrophy children as a consequence of bony defects. Many of these children learn to walk with waddling gait initially which resolves as the children grow. In a study of 14 adult exstrophy patients, pelvic radiographs and a validated hip joint rating were used for biomechanical analysis to determine hip joint forces and stresses and their clinical effect. A few develop gait problems, hip dysplasia and back pain.<sup>[54]</sup> Early osteotomy has long term effects on patients' instinctive walking pattern and neutralises some of the effects of bladder exstrophy.<sup>[55]</sup>

## CONCLUSIONS

The adolescents with bladder exstrophy and epispadias had significant physical and mental problems. Genital

malformation and urinary and fecal incontinence may have a negative impact on mental health and psychosocial functioning. Cosmetic appearance and functionality of the genitalia should be reassessed at or near puberty along with the status of social continence. There is a need to include psychosocial experts on health care teams to reveal the amount of distress caused by these anomalies and to offer the psychosocial support. Recent long-term outcome studies of bladder exstrophy patients treated with primary reconstruction and QOL thereof, may warrant reassessing the place of alternative management options.

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