



REVIEW ARTICLE

Bladder Exstrophy Epispadias Complex: Meeting the Challenge!

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Born with normal upper urinary tracts, children with BEEC have continued to test the ingenuity of Paediatric Surgeons.

The upper tracts are almost always normal in the neonates with classic BE

before abdominal wall, posterior urethral, and bladder closure. However, the upper tract changes or deterioration of renal function almost always occurs after reconstructive surgery. (Fig. 1)

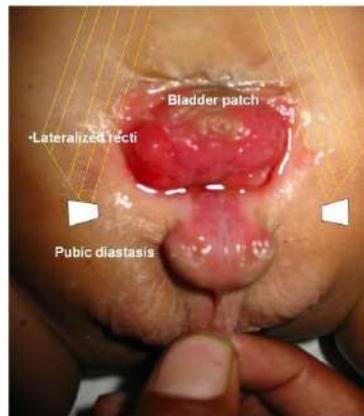


Fig. 1. Bladder Exstrophy - Epispadias Complex: Poor quality detrusor; lack of bladder neck sphincter; Short stubby penis

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The objectives of treatment are to achieve a secure closure of the bladder, pelvis, and the abdominal wall; preservation of renal function; urinary continence and creation of functional and cosmetically appealing genitalia.

Since the first description by Young *et al.* the goal of repair of BEEC is to create a safe reservoir for upper tracts, while providing socially acceptable voluntary voiding, has alluded the surgeons universally [1].

Overtime various procedures have been described in different permutations & combinations. These are:

- i) Jeffs & Cendron in 1970- staged repair: bladder closure followed

by BNR & Epispadias repair, then

- ii) Modern Staged Repair of Exstrophy (MSRE) wherein, Epispadias repair precedes BNR &
- iii) Complete primary repair (CPRE) which include bladder closure, partial BNR

Epispadias repair in one stage. CPRE was performed on the premise that closure of bladder, bladder neck & the urethra together will stimulate early bladder cycling & induce voluntary control on micturition [2] (Fig. 2).



Fig. 2. Complete primary repair of bladder exstrophy-epispadias complex in a neonate

With CPRE: Up to, 70% of renal units develop (HN); pyelonephritis is seen in up to 28% of patients, 20-24% develop cortical renal scarring, and 10% nephrolithiasis [3-6].

Detrusor under activity has also been reported by this technique [7]. Overall, long-term renal outcomes are similar to those of other techniques presented in the literature. Ureteric

reimplantation (UR) needed adequate bladder capacity. Different perceptions exist for capacity. Therefore, ureteric reimplantation simultaneous to bladder closure has been considered an over-do. (Ellison).⁶ As a result a more conservative approach has been adopted to VUR & a very small number subjected to reimplantation.

Growth of the bladder is fundamental to achieve these goals. Impetus to growth has been expected by increasing outlet resistance by BNR – Jeff & Gearhart; MSRE; CPRE & recently by Bladder Neck Injection [8-9].

While BNR was expected to provide impetus for bladder growth, it was soon realized, that, increasing outlet resistance leads to upper-urinary-tract damage and impairment of bladder function.¹⁰

BNR has also been recognized to impair bladder development but generates high intravesical pressures which were detrimental to the upper tracts [11].

Different aspects of outcomes from each of these approaches have been studied at varied intervals with wide ranging results. Renal damage rates has been reported to be 13% to 20% [12,13].

In another series 14.7% had renal scarring & 4.5 % had renal insufficiency [14].

Continence depends upon technically creating an adequate balance between

- Urine storage (which implies high outlet resistance and low storage pressure) and
- Complete bladder emptying (which implies low outlet resistance and a transient increase in bladder pressure);
- Surgery cannot achieve 'continence' (which implies active mechanisms) .Continence rates of only 10% to 30% were reported with this approach [15-16] (Fig. 3).

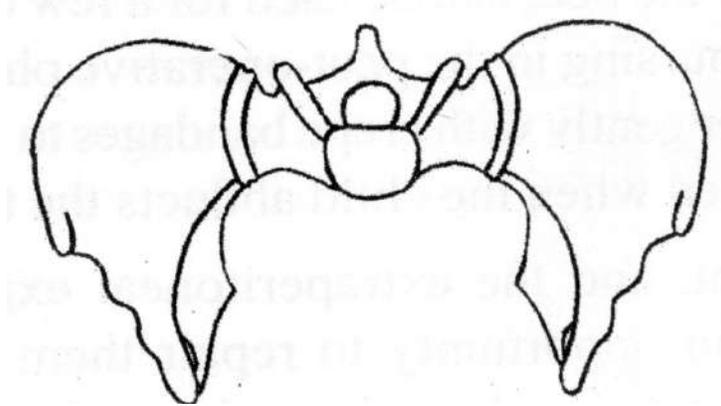


Fig. 3. Pubic diastasis in bladder exstrophy-epispadias complex (diagramatic).

Assessment of continence is often difficult, as data given by families are not always accurate and because the criteria used by various authors vary, even within the same institution. The fact that surgery cannot produce "continence" (which implies active mechanisms) emphasises the difficulty in finding an adequate balance between urine storage (which implies high outlet resistance and low

storage pressure) and full bladder emptying (which implies low outlet resistance and a temporary increase in bladder pressure).

Augmentation helps but all segments of native bladder malignancy.

The concept of gastric neo-bladder: In BEEC the bladder is represented by a small patch of urinary bladder with its exposed mucosa

herniating from the lower abdomen. attempts to preserve the native bladder aimed at providing a receptacle for urine which would provide normal urinary control, store urine under pressures which are safe for upper tracts. Another alternative is to divert urine to the bowel.

After a ureterosigmoidostomy, bladder and bowel adenocarcinoma: Smulders and Woodhouse's outstanding series of 103 neoplasms demonstrated that this patient population has a 700-fold higher probability of developing cancer than the age-matched general population. Lack of growth of the bladder in exstrophy has been attributed to intrinsic abnormalities. Following reconstructive surgery, some of these abnormal morphometric patterns return to normal, while others do not.

This arises the fundamental question of whether the exstrophic bladder cannot heal after reconstructive surgery or whether the operation was ineffective. Did the surgical reconstruction aid in bladder

repair or did it worsen an already damaged bladder?

If the underlying morphometric and neurological abnormalities cannot be completely rectified by surgery, these issues are far from moot. Our reconstructive efforts leave behind irreversible renal injury & a childhood with morbidity of urinary incontinence with a bladder forever risk for malignancy.

These concerns cannot currently be answered, but they need to be solved in order to guide future reconstructive attempts to treat bladder exstrophy. Till then, we are offering these children with a Gastric neo-bladder after initial CPRE. Native bladder is replaced on attaining 5 years of age. The procedure involves harvesting the gastric bladder laparoscopically and lowering it to the pelvis. The remaining repair involves ureteric reimplantation and creating the Mitrofanoff channel.

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