SAS Journal of Surgery
Abbreviated Key Title: SAS J Surg
ISSN 2454-5104
Journal homepage: https://www.saspublishers.com

OPEN ACCESS

Epispadias in a 32-Year-Old Patient with Congenital Malformatiom: Bladder-Epispadias Exstrophy at Birth

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DOI: 10.36347/sasjs.2021.v07i09.007 | **Received:** 05.08.2021 | **Accepted:** 09.09.2021 | **Published:** 11.09.2021

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Abstract **Review Article**

Bladder atrophy and / or epispadias This is an orphan genetic malformation affecting one child in 30 to 40 thousand births. This malformation requires immediate surgical management in a specialized center. Many interventions will be essential in order to resolve urinary and genital problems. Treatment protocols vary, but are still based on three essential steps: closing the bladder in the neonatal period; reconstruction of the urethra and penis in boys; bladder emptying and continence surgery. If the first two stages are well mastered, the last remains a challenge, because the techniques in force are unable to restore the active and complex mechanisms that manage bladder emptying and the maintenance of dryness between two bladder emptying. We now have to resolve to create passive subbladder resistance associated with intermittent multi-daily catheterization in order to preserve the upper urinary tract, which remains the first priority.

Keywords: Bladder exstrophy, Epispadias, Exstrophy, Congenital urinary incontinenE.

INTRODUCTION

Exstrophy-epispadias complex is the most severe form of midline abdominal malformation. [1]. EEC covers a spectrum with different levels of severity. The classic bladder exstrophy, the most common of this complex represents 60% of these lesions. The variants of epispadias represent 30%. The birth of a child with a variant of bladder exstrophy is exceptional. Its initial presentation can be confusing, which often delays treatment.

It is one of a wide range of midline defects, collectively called the exstrophy/epispadias complex.

OBSERVATION

The patient is 31 years old, native and resident in KALAA MGOUNA, single, without profession, male

ATCD

Patient followed in our training for bladder extrophy operated in 2010: cystectomy + replacement enterocystoplasty; Neo bladder pruning for intra bladder stone 15/02/2016; suture of anterior neo bladder breach 30/03/2016; disincrustation of distal end of bladder Foley catheter which is calcified 08/02/2017 and laparoscopic cholecystectomies on month 06/2019.

The patient was hospitalized for recovery of the penis; presented 4 days after hospitalization: a urinary tract infection.

PARACLINICAL

AUSP: Calcium tonalite opacity in the bladder air (Figure 1).

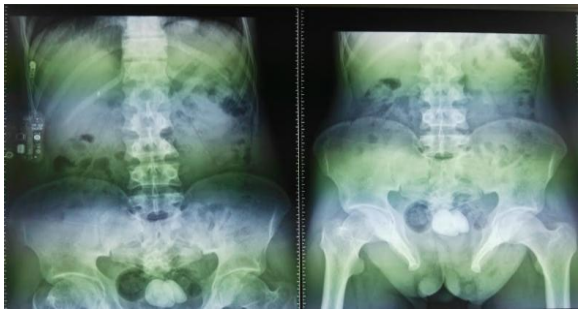

Figure 1: Calcium tonalite opacity in the bladder air

OPERATING TECHNIQUES

UROLOGY TEAM:

Patient in dorsal decubitus, under GA, right paramedian incision, musculo aponeurotic opening then

upward displacement of the intestinal anses, setting up of the GOSSET retractor, highlighting of the neo bladder, and Cystotomy, extraction of a bladder stone measuring 6 cm (Figure 4) and cystorraphy using vicryl 2.0 after setting up of a ch 14 silicon probe.

PLASTIC SURGERY TEAM

1st stage:

Infiltration with SS adrenalized 1/200000 opposite the pre-established tracing of the epispadias.

2nd step:

Incision according to the line with the cold blade n° 15 parallel to the haemostasis then detachment of the edges with the Metzenbaum scissors. (Figure 3).

Placement of a siliconized probe ch 14. Verification of direct closure of the freed mucosa, then closure by separate stitches with Vicryl 4/0, then with the surge passed to Vicryl R 3/0, solidification of the corpus cavernosum with Vicryl rapid 3/0 and closure of the skin fold by single stitch with Vicryl R 3/0 (Figure 5).

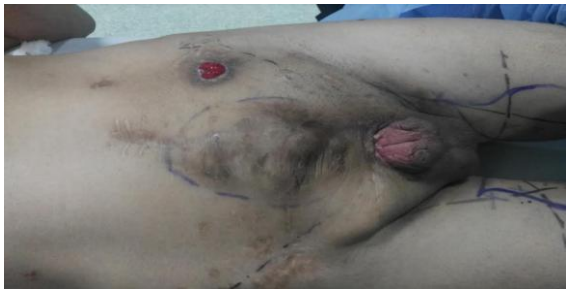

Figure 2: Epispadias in A Young Patient Operated on For Extrophy of the Bladder

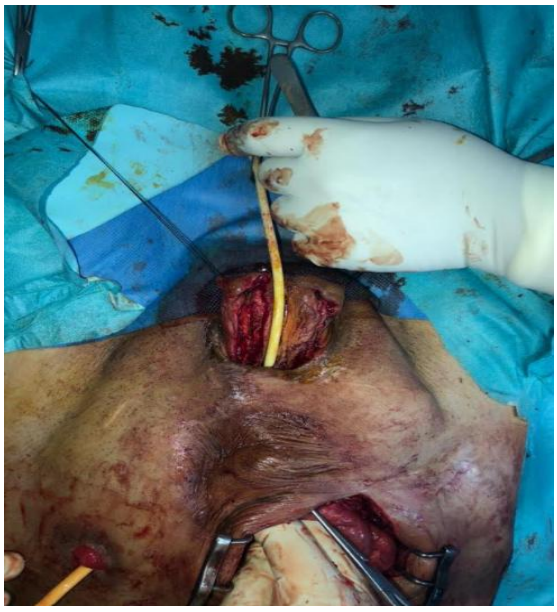

Figure 3: Incision With The Metzenbaum Chisel

Figure 4: Extraction of a 6 Cm Stone

Figure 5: Covering of An Epispadias

POST OPERATIVE

Post-operative evolution: It was simple, without short-term complications.

DISCUSSION

AGE:

The reported incidence of exstrophy-epispadias complex ranges from 2.1 to 4.0 per 100,000 live births [2-4]... Classic bladder exstrophy appears to occur more frequently in white infants and the incidence varies by geographic region and socioeconomic and insurance status [5].

SEX:

Bladder exstrophy is more common in boys with a preponderance of 3/1 and is not usually associated with abnormalities. Variants are more

frequent in girls and seem to be associated with various malformations.

ATCD:

In both sexes, urologic malformations (e.g., ureteropelvic junction obstruction, ectopic pelvic kidney, horseshoe kidney, renal hyo- or agenesis, megaureter, ureteral ectopia, and ureterocele) are present in approximately one-third of all cases of exstrophy-epispadias complex, predominantly in the cloacal exstrophy population [1]. However, a 100% prevalence of bilateral vesicoureteral reflux due to failure of ureterovesical junction development across the exstrophy-epispadias complex spectrum warrants an antireflux procedure with every bladder neck plasty.

Spinal and orthopedic anomalies

The incidence of spinal anomalies varies considerably across the spectrum of exstrophy-epispadias complex. In children born with classic bladder exstrophy, spinal abnormalities occur in approximately 7% of cases, whereas a heterogeneous group of congenital spinal abnormalities resulting from defective neural tube closure early in fetal life and abnormal development of the caudal cell mass can be confirmed by magnetic resonance imaging (MRI) in nearly 100% of patients with cloacal exstrophy. Therefore, EC neonatal patients should undergo ultrasound and spinal radiographs to define individual spinal abnormalities ranging from hemivertebrae to myelomeningocele.

Gastrointestinal Abnormalities

Gastrointestinal tract anomalies are primarily associated with cloacal exstrophy and will rarely be present in classic bladder exstrophy. In addition to a common hindgut residue of variable size, omphaloceles are found in 88-100% of cases in CE. Gastrointestinal malrotation or duplication.

CLINICAL:

The diagnosis of EEC is usually made clinically by inspection after birth.

PARACLINICAL:

Imaging studies

Ultrasound as the main study

After birth, a basic ultrasound examination of the kidneys is mandatory for all patients with exstrophy-epispadias complex. Later, regardless of the method of reconstruction, renal ultrasound is a perfect screening method to distinguish upper urinary tract changes during follow-up.

MRI imaging in CEE

a) Pelvis and pelvic floor in exstrophy-epispadias complex

MRI has characterized the specific defect of the exstrophy-epispadias complex of the pelvis with an anterior outward pelvic rotation of 18 °, a posterior outward pelvic rotation of 12° and a shortening of the pubic branches of about 30% [5, 6]. In infancy, external rotation of the foot of approximately 30° is evident, although this rotation abnormality improves with age. A flatter and less conical lifting sling, a significantly larger coronal levator hiatus diameter, and a normal double lifting angle have been demonstrated after pelvic reconstruction and osteotomy in young children with the exstrophy-epispadias complex compared to normal anatomy [5, 6]... In addition, the classic bladder exstrophy, the anus elevator muscle is distributed more posteriorly with a shortened anterior segment in the coronal plane [5, 6]... This specific abnormality of the pelvic floor can determine rectal prolapse, occasional anal incontinence and predispose women to uterine prolapse [7].

b) Genital anatomy in exstrophy-epispadias complex

Pelvic MRI provides adequate information about the internal genitalia before and after surgery. Therefore, MRI is mandatory before uterine prolapse repair and in case of complex penile surgery. In the literature, MRI has clarified the complex genital anatomy of the exstrophy-epispadias complex in both sexes. In adult males the exstrophy-epispadias complex, Silver *et al*., documented an inherent body deficiency with a shorter anterior body length. This deficiency contributes, in addition, to the penile cord or bilateral attachment of the penis to the ascending pubic branches and to the specific appearance of the short and curved penis in the exstrophy-epispadias complex [8, 9]. The development of the bulbospongiosus muscle in the exstrophy-epispadias complex remains unclear, as only the rudiments are found on the base of the penis. On MRI, the prostate had a nearly normal volume, but even after reconstruction of the bladder neck, it did not extend circumferentially around the urethra [8]. The slightly smaller seminal vesicles had a regular shape and location; however, fructose levels were normal in more than 50% of the patients with exstrophy-epispadias complex after functional reconstruction, indicating the fact that surgery cannot severely alter seminal vesicle function. Despite the complex abnormality of the penis and bladder, male patients with exstrophy-epispadias complex usually have normal sperm production and transport from the testes to the verumontanum [1].

SURGICAL TECHNIQUES

Functional reconstruction of the bladder

The concept of reconstruction is based on the fundamental agreement that the exstrophic bladder has the potential to acquire normal bladder function as a low-pressure reservoir for urine storage, active voiding, and protection of the upper urinary tract after functional reconstruction. In addition to clinical observation, urodynamic studies have reported a normal filling and emptying pattern, but impaired compliance and stability, primarily after Young-Dees-Leadbetter bladder neck reconstruction. This bladder neck

reconstruction is now considered a non-nerve-sparing technique, maintaining normal detrusor function in only about 25% of cases [1]. In addition, it is a published statement that bladder neck reconstruction has the capacity, although a passive mechanism of increasing subvesical resistance, to result in all cases in complicated bladder emptying [11]. Most other authors emphasize terminal continence involving a possibility of active, non-scarring, unobstructed emptying of the bladder neck region [1].

On the basis of retrospective studies, the main successful operative attempt with the bladder model is considered the main predictor of success. To date, the quality and size of the bladder plate and its actual influence on the outcome of a functional reconstruction are not predictable.

In conclusion of previous reconstruction failures, a staged concept was derived from the studies of Jeffs and Cendron in the early 1970s. The traditional staged reconstruction popularized by Jeffs and Gearhart has been a standard approach for many years. As a modification, the so-called "modern stepwise approach" is currently advocated by John Gearhart [1]. He has made this three-step concept - beginning with closure of the bladder, posterior urethra and abdominal wall: after adaptation of the pelvic ring within the first 48 hours of life - popular with many other experts around the world [1].

Male Genital Reconstruction

Mportant for penile reconstruction in exstrophy-epispadias complex is the unique presence of two completely separate corpora cavernosa without any vascular anastomosis and completely isolated neurovascular bundles functioning on the body exterior. Regardless of the type of epispadia repair, the following four key issues must be addressed to ensure a functional and cosmetically acceptable penis:

1. Dorsal cord correction
2. Urethral reconstruction for micturition and sperm transport
3. Glandular reconstruction
4. Closing of the penile skin.

To deflect the curved epispadic penis, Phillip Ransley introduced the concept of dorsal cord release by incision and dorsomedial anastomosis of the corpora cavernosa above the urethra. As an advancement, the Cantwell-Ransley technique completely detached the urethral plate from the corpora allowing for more effective urethral transposition under the corpora, and thus more effective correction of the dorsal curvature by lateral rotation of the corpora. The hallmark of Mitchell penile disassembly is the complete detachment of the urethral plate from the corpora cavernosa and glans. This technique utilizes the advantage of constant blood supply to the corpora cavernosa, urethra, and glans with paired dorsal arteries and neurovascular bundles, deep cavernous arteries to the corpora and cancellous tissue to the proximal urethral plate [1,12]. After complete separation of the corpora, the glans is divided and the urethra is placed ventrally, often resulting in a hypospadic meatus requiring additional repair of the penile tip [1, 12]. In addition, it has been questioned whether complete penile disassembly gains as much penile length as possible complications may arise. The surgical advantages of the Cantwell-Ransley and Mitchell techniques are the more anatomical reconstruction with only slight deviation of the penis and a low rate of fistula due to coverage of the neourethra through the corpora cavernosa. If mobilization of the urethral plate from the corpora is sufficiently radical, the corpora can most likely be joined without tension, and without corporotomy and the need for complete mobilization of the neurovascular bundles. However, the scarring and brevity of the neurovascular bundles may later cause severe, often incorrigible penile deviation.

CONCLUSION

Bladder extrophy and/or epispadias This is an orphan genetic malformation affecting one child in 30 to 40 thousand births.

This malformation requires immediate surgical care in a specialized center. Numerous interventions will be essential in order to solve the urinary and genital problems.

The goal is to improve bladder emptying and to obtain times without urine leakage (called "dry time") long enough to allow a social life as normal as possible. Surgery does not make it possible to completely reconstitute what nature has failed to do.

In addition, we often have to combine an enlargement of the bladder with a bowel loop to these different treatments. This step-by-step surgery has variations depending on surgical habits.

Conflicts of Interest: The authors declare no conflicts of interest.

Authors' Contributions

All authors contributed to the conduct of this work. All authors also declare that they have read and approved the final version of the manuscript.

RÉFÉRENCES

1. Gearhart, J. P. (2001). Le complexe exstrophie vésicale-épispadias-exstrophie cloacale. Urologie pédiatrique. Edité par: Gearhart JP, Rink RC, Mouriquand PDE. Philadelphie: WB Saunders Co, chapitre, 32, 511-546.
2. Anonyme. (1987). Epidémiologie de l'exstrophie de la vessie et des épispadies: une communication de l'International Clearinghouse for Birth Defects Monitoring Systems. *Tératologie*, 36, 221-227.

3. Yang, P., Khoury, M. J., Stewart, W. F., Beaty, T. H., Chee, E., Beatty, J. C., ... & Gordis, L. (1994). Comparative epidemiology of selected midline congenital abnormalities. *Genetic epidemiology*, *11*(2), 141-154.
4. Caton, A. R., Bloom, A., Druschel, C. M., & Kirby, R. S. (2007). Epidemiology of bladder and cloacal exstrophies in New York State, 1983–1999. *Birth Defects Research Part A: Clinical and Molecular Teratology*, *79*(11), 781-787.
5. Stec, A. A., Pannu, H. K., Tadros, Y. E., Sponseller, P. D., Fishman, E. K., & Gearhart, J. P. (2001). Anatomie du plancher pelvien dans l'exstrophie de la vessie classique à l'aide de la tomographie informatisée 3D: aperçus initiaux. *J Urol*, 166, 1444-1449.
6. Sponseller, P. D., Bisson, L. J., Gearhart, J. P., Jeffs, R., Magid, D., & Fishman, E. (1995). L'anatomie du bassin dans le complexe d'exstrophie. *J Chirurgie osseuse et articulaire*, 77(2), 177-189.
7. Woodhouse, C. R. J., & Hinsch, R. (1997). L'anatomie et la reconstruction des organes génitaux féminins adultes dans l'exstrophie classique. *BJU*, 79, 618-622.
8. Gearhart, J. P., Yang, A., Leonard, M. P., Jeffs, R., & Zerhouni, E. A. (1993). Taille et configuration de la prostate chez les adultes souffrant d'exstrophie de la vessie. *J Urol*, 149, 308-310.
9. Silver, R. I., Yang, A., Ben-Chaim, J., Jeffs, R., & Gearhart, J. P. Longueur du pénis à l'âge adulte après reconstruction d'exstrophie. *J Urol*, 157(3), 999-1003.
10. Ebert, A. K., Bals-Pratsch, M., Seifert, B., Reutter, H., & Rösch, W. H. (2008). Fonction génitale et reproductrice chez l'homme après reconstruction fonctionnelle du complexe Exstrophy-Epispadias - résultats à long terme, *Urologie*, 72(3), 566-569.
11. Mouriquand, P. D. E., Bubanj, T., Feyaerts, A., Jandric, M., Timsit, M., Mollard, M., Mure, P. Y., & Basset, T. (2003). Résultats à long terme de la reconstruction du col vésical pour l'incontinence chez les enfants présentant une exstrophie vésicale classique ou des épispadies incontinentes. *BJU*, 92, 997-1002.
12. Mitchell, M. E., & Bagli, D. J. (1996). Démontage complet du pénis pour la réparation des épispadias: la technique Mitchell. *J Urol*, 155, 300-303.