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Isolated Male Epispadias with Continence in a 40 Year Old: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Epispadias is part of the bladder exstrophy-epispadias complex; it has a defective dorsal wall of the urethra with potential incompetence of the urinary continence mechanism. Isolated male epispadias is due to failure of the urethral plate to tubularize on the dorsum of the penis. It is rare with an incidence rate of 1 in 117,000 live births in males. The severity depends on the position of the urethral opening and ranges from peno-pubic to penile and glandular. Epispadias is usually repaired within the first year of life though some patients have presented in their second decade. Epispadias is classically associated with bladder exstrophy in over 90% of the cases while isolated epispadias with continence is very rare constituting less than 10% of cases. Separation of pelvic bones is seen in 70% of peno-pubic epispadias affecting the bladder neck and external sphincter leading to incontinence and stress urinary dribbling. The goals of repair include; achieving a cosmetically acceptable and functional penis, which is straight and adequate in length, enabling

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penetrative sexual intercourse and urinary continence. There have been reports of isolated epispadias in adults but none was as old as 40 years. This patient has been married for 20 years and has been unable to impregnate his spouse. He had modified Cantwell-Ransley procedure after a detailed clinical and psychological evaluation to achieve the goals of repair. Adults presenting with isolated continent peno-pubic epispadias are rare. Presenting at this age and after having been married for two decades could have a derogatory effect on body image, self-confidence, psycho-sexual and reproductive life. Surgical correction of this anomaly as we did has the potential to lead to acceptable outcomes as demonstrated in this index case.

Keywords: Urethra; continent; penile; exstrophy; isolated; anomaly.

1. INTRODUCTION

The Bladder Exstrophy Epispadias Complex (BEEC) is an abdominal midline malformation [1]. The severity of BEEC ranges from epispadias which is the simplest form to the more severe classic bladder exstrophy. Epispadias is part of the exstrophy-epispadias complex and it has a defective dorsal urethral wall with potential incompetence of the urinary continence mechanism [1,2,3]. Isolated male epispadias is characterized by the position of the urethral opening, from peno-pubic to penile and glandular [1,2]. It is due to failure of the urethral plate to tubularize on the dorsum of the penis during embryonic development thereby giving rise to a dorsally located ectopic urethral meatus [1,4]. Epispadias is rare with an incidence rate of 1 in 117,000 live births in males and 1 in 1,300,000 live births in females [1,2,5,6]. Epispadias is associated with many co-exisiting congenital conditions such as vesicoureteric reflux (VUR), bladder exstrophy, urinary incontinence and inguinal hernia [4,7]. The incidence of co-existing VUR is 30 – 75% and of inguinal hernia is 35% [7]. The vast majority of epispadias that occur in association with bladder exstrophy are classified as non-syndromic. The etiology of this malformation remains unknown,[1] however, it has been proposed that an abnormal overdevelopment of the cloacal membrane preventing medial migration of mesenchyme between the ectodermal and endodermal layers may be the basis for this anomaly [1,4,8].

Epispadias is typically diagnosed at birth in the male child, but in the female it may go unrecognized, until the girl experiences persistent urinary incontinence after toilet training or recalcitrant urinary tract infection [1,7]. Generally the phallus is broad and short with a dorsal chordee, an open urethral plate and absent dorsal foreskin in the male child, while in the female child, it presents as a bifid clitoris and labia minora. This is contrary to the case in hypospadias, where the meatus lies ventrally on the penile shaft [1,4,7].

The diagnosis of epispadias is clinical, but most of the time plain x –ray is required to assess the orientation of the pelvic bones, and ultrasonography to assess the upper urinary tract for concomitant renal anomalies. A micturating cystourethrogram is useful in evaluating the bladder capacity as well as the presence of vesicoureteric reflux [7]. Managing epispadias poses a great challenge and its treatment is mainly surgical, involving bladder closure, urethral reconstruction and osteotomies if pubic diastasis is present [4,7]. In adults who present late the challenges include long-standing changes to the exposed urethral mucosa and the surrounding tissues. These adult patients also have to battle with concerns about their psycho-sexual potential [8].

The traditional time of surgical repair of epispadias is usually within the first year of life, though some patients have presented in their second decade of life [2,3,6]. Achieving a cosmetically acceptable and functional penis, which is straight and adequate in length thus enabling penetrative sexual intercourse and urinary continence are the main goals of repair [2,4,6].

Two surgical techniques have gained widespread adoption for the repair of male epispadias. The modified cantwell-ransley repair technique involves dissecting the urethral mucosa plate away from the corpora except for the distal-most 1 – 1.5cm to get the urethra deeper under the corpora at the glans level [8]. The other technique is the complete penile disassembly which was described by Mitchel and Bagli in 1996 [8].

The choice between the techniques depends on the surgeon's expertise and the specific case [4,6,8]. Even though, there have been reports of

isolated male epispadias in adults, none was as old as 40 years of age and married. The index patient was married for 20 years at the time of presentation, but had been unable to impregnate his spouse. He had modified Cantwell-Ransley procedure, after thorough clinical and psychological evaluation to achieve the goals mentioned earlier.

2. CASE PESENTATION

2.1 Patient Information

Mr Y.D. is a 40year old married man who presented to our clinic on account of an abnormally formed penis since birth. Growing up he noticed his penis looked different from that of his friends and he urinated with an upward stream from an abnormal dorsal opening making his urine to splay. Fig. 1. He was, however, continent of urine and got penile erections which curved upwards towards the anterior abdominal wall. He is married to his wife of 20 years and even though their marriage is consummated, it is yet to result in pregnancy since he is unable to deposit semen in the vagina during coitus. No gait abnormality was reported or observed in our patient.

2.2 Clinical Finding

On examination, we found an abnormally looking penis, with about 45 degrees twist to the left, a ventral hood and dorsal chordee. The urethral meatus opened dorsally close to the peno-pubic junction and he had a supple and splayed out urethral plate measuring 4cm in width. Figs.1 and 2. Scrotal examination revealed no abnormality.

2.3 Timeline

There was no proper medical care during his childhood as he grew up an orphan with his older sibling being the caregiver. It was only after being married for 20 years that his wife convinced him to seek proper medical evaluation and treatment. Following thorough psychological and medical evaluation, he was counseled and worked up for surgery.

2.4 Diagnostic Assessment

The diagnosis of epispadias was made clinically with history and physical examination**.** Abdominopelvic sonography and pelvic X-rays conducted revealed no abnormality in his kidneys or pelvic bone anatomy. Fig. 3. The complete blood count was normal with a packed cell volume of 47% and normal kidney function tests. A micturating cystourethrogram revealed good bladder capacity with no vesico-ureteric reflux seen Fig. 4.

2.5 Therapeutic Intervention

He had modified Cantwell-Ransley repair under combined spinal and epidural anesthesia. At surgery, we did meatal advancement and glanuloplasty using the Heineke-Mikulicz technique. The penile skin was degloved to the root of the penis and parallel incisions made on the urethral plate to mobilize it from the corpora cavernosa. Fig. 5. The mobilized urethral plate was tubularized over a stent (size 16Fr silicon catheter) by continuous submucosal suturing using monocryl 4/0. Caverno-cavernostomy was done on both sides of the corpora to allow the tubularized urethral to be repositioned ventrally before suturing the cavernosa together above the urethra. A W-flap was raised at the pubic region and the suspensory ligament released to give length to the penile shaft. The penile skin was then apposed with vicryl 3/0 sutures and the skin flap closed. Fig 6. We left a supra-pubic catheter in place for temporary urinary diversion to rest the urethral repair.

Postoperatively he received intravenous levofloxacin 500mg once daily, metronidazole 500mg thrice daily and penthidine 50mg 6 hourly injections for 48hours. He commenced oral feeds same day of surgery and oral medications on the third post-operative day namely; tolterodine 2mg twice daily, oral levofloxacin 500mg daily and metronidazole 400mg thrice daily as well as hematinics. He was discharged home on the 9th post-operative day with the urethral and suprapubic catheters in-situ.

2.6 Follow up and Outcomes Measure

Eight days following discharge from the hospital, he developed a superficial surgical site infection which we treated successfully with povidone iodine daily dressings. The suprapubic and urethral catheters were removed by the 4th post-operative week. He voided satisfactorily and is happy with the functional and cosmetic outcome. Fig. 7.

Fig. 1. Clinical picture of peno-pubic epispadias before surgery

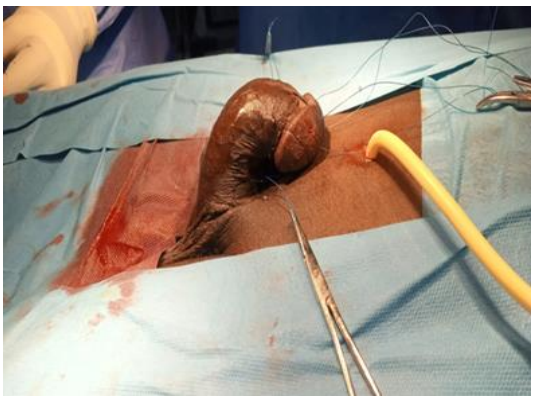

Fig. 2. Clinical picture showing degree of chordee in epispadias

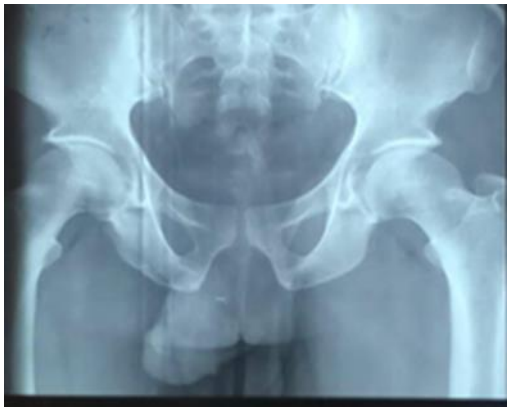

Fig. 3. Plain Pelvic X- ray showing normal anatomy

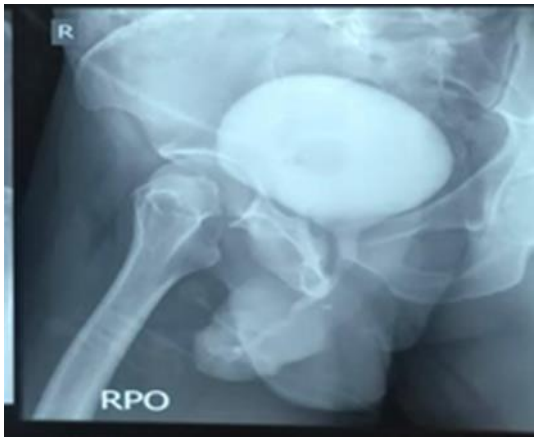

Fig. 4. Micturating cysto-urethrogram showing normal bladder capacity and no vesico-ureteric reflux

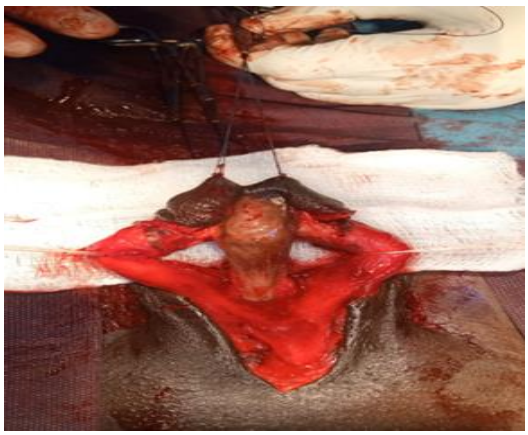

Fig. 5. Separation of the urethral plate from the cavernosa

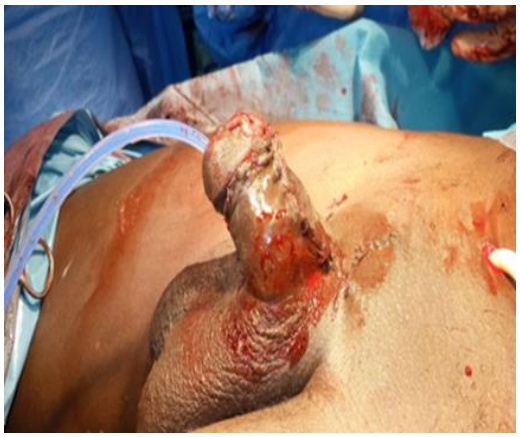

Fig. 6. Immediate post-operative clinical picture

Fig. 7. Clinical picture 2 months after repair

3. DISCUSSION

Isolated continent epispadias is a very rare congenital anomaly usually present at one end of the spectrum of bladder exstrophy epispadias complex (BEEC) and constituting less than 10% of cases of epispadias [1,7,8]. It is classically associated with bladder exstrophy in over 90% of cases [1,9]. Its presentation varies from an isolated urethral defect to severe bladder exstrophy with concomitant malformations involving the kidney, groin and colorectal region [7]. In the male child common examination findings are displacement of the meatus or an open urethral plate. In the female child, epispadias present as bifid clitoris and poorly developed labia minora [7].

The diagnosis is typically clinical and easy to make especially in the male child and further investigations are only required to exclude concomitant abnormalities.[4] Based on the degree of involvement of the urinary sphincter, urinary incontinence could be a presenting complaint [1].

It is very unusual for patients with isolated epispadias to present for the first time as adults just like in the index case [6]. Braga et al in their series reported a mean age at the time of surgical repair of epispadias to be 16.8 months [5]. The site of the urethral opening varies depending on the severity ranging from peno-pubic to penile and glandular [1,2,4]. This anomaly is also characterized by aplasia of the dorsal part of the urethra [1,7,10].

Treatment of epispadias is surgical and it is aimed at giving the patient normal urinary control, and a straight, cosmetically and functionally acceptable penis. In the literature, various surgical procedures to treat epispadias have been described and they are usually challenging, even in the hands of experts [4,6,8]. The two universally adopted surgical techniques in the repair of epispadias are; the Modified Cantwell-Ransley technique described by Gearhat and the Mitchell-Bagli penile disassembly technique popularized in 1996 [2,4,8]. Mitchell-Bagli described the complete splitting of the corporal bodies and hemi-glans into separate halves with total dissection of the urethral plate off the corpora. This makes for proper ventralization of the tubularized urethra [2,4,8]. Cantwell, on the other hand, proposed a technique based on the complete mobilization of the urethral plate which was in turn tubularized and transplanted ventrally between the corpora. It was Young who modified the classical Cantwell repair by avoiding complete urethral plate mobilization for better preservation of its vascularity thus decreasing the risk of urethral fistula formation [4,8]. Our patient, who was married for 20 years, presented to us at the age of 40 years following persuasion from his wife. There was no record of medical consultation during his young age as he began as an orphan very early in life and the caregiver did not make sufficient effort toward having the abnormality treated [11]. Delayed presentation in our patient may not be unconnected to poverty and ignorance which is prevalent in our sub-saharan African population as previously alluded to by the authors in an earlier published case report [6]. The same reason given for late presentation of epispadias in our patient is further corroborated by Gite et al in their series in India [8].

Isolated male epispadias with continence remains a rare finding and comprises less than 10% of all epispadias cases [1,7,8]. It is

commoner for epispadias to present with bladder exstrophy as seen in over 90% of the cases [1,9]. Separation of pelvic bones is seen in 70% of peno-pubic epispadias and this affects the bladder neck and external sphincter leading to incontinence and stress urinary dribbling. [1,4,6]. The index patient had normal pelvic bone anatomy and did not present with incontinence. Before his surgery, he and his spouse were reviewed by the psychiatrist due to concerns about his psycho-sexual potential [8]. He eventually had a one-stage repair using the modified cantwell-ransley technique as described in the literature [4,5,6,8]. We routinely insert a suprapubic cystostomy to serve as a proximal diversion and improve the outcome of the surgery [6,8]. The idea of leaving in place a suprapubic diversion is also supported by Gite et al [8]. We administered oral anti-cholinergics for the period the indwelling catheter was in place to mitigate the problem of bladder spasms [8]. Our patient developed a surgical site infection which is not an uncommon complication following this surgery [8]. We successfully managed the infection conservatively with povidone iodine dressings. There were no other complications noted and the patient was followed up for 6 months and reported satisfaction with the final outcome of his management.

4. CONCLUSION

Adults presenting with isolated continent peno-pubic epispadias are rare. Presenting at this age and after having been married for two decades could have a derogatory effect on body image, self-confidence, psycho-sexual and reproductive life. Surgical correction of this anomaly as we did has the potential to lead to acceptable outcomes as demonstrated in this index case.

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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