ISOLATED MALE EPISPADIAS WITH CONTINENCE IN A 40 YEAR OLD: A CASE REPORT

ABSTRACT

Epispadias is part of the bladderexstrophy-epispadias complex; it has a defective dorsal wall of the urethra with potential incompetence of urinary continence mechanism. Isolated male epispadias is due to failure of the urethral plate to tabularize on the dorsum of the penis. It is rare with an incidence rate of 1 in 117,000 live 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 of life. 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 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 but 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

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 theurinary 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 tabularize 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 in malesand 1 in 1,300,000 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 occur in association with bladder exstrophy are classified as non-syndromic. The etiology of thismalformation is still 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 diagnosedat birth in the male child, but in the females it may go unrecognized, until the girl experiences persistenturinary incontinence after toilet training or recalcitrant urinary tract infection.[1,7]Generally the phallus is broadand 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 cystourethrographis useful in evaluating the bladder capacity as well as presence of vesicoureteric reflux. [7]. Managing Epispadias poses a great challenge and it's 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 theirpsycho-sexualpotentials.[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 wide spread adoption for the repair of male epispadias. The modified cantwell-ransley repair technique which involves dissecting the urethral mucosa plate away from the corpora except for the distal-most 1 – 1.5cm so as to get the urethra deeper under the corpora at the glans level.[8] The other technique is thecomplete 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. Hehad modified the Cantwell-Ransley procedure, after thorough clinical and psychological evaluation with the aim of achieving the goals mentioned earlier.

PATIENT AND OBSERVATION

Patient Information

Mr Y.D. is a 40year old married man who presented to our clinic on account ofan 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 a dorsal and proximal point on his penis making his urine to splay. He was however, continent of urine and gets penile erections which curve upwards towards the anterior abdomen wall..He is married to his spouse for 20 years andeven thoughis consummated it is yet to result in any pregnancy since he is unable to deposit semen into her vaginaduring sexual intercourse. No gait abnormality was reported or observed in our patient.

Clinical Finding

On examination, we found an abnormally formed penis, with about 45 degreestwistto the left and a ventral hood and dorsal chordee.The urethral meatus opened dorsally close to the peno-pubic junctionand he had a supple and splayed urethral plate measuring4cm in width. Figures 1 and 2. Scrotal examination revealed normal testes bilaterally.

Timeline

There was no proper medical care duringhis 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. Followingthorough psychological and medical evaluation, he wascounselled and worked up for surgery.

Diagnostic Assessment

The diagnosis was made clinically with history and physical examination**.** Abdomino-pelvic sonography and pelvic X-rays conducted, revealed no abnormalityin his kidneys or anatomy of his pelvic bones. Figure 3. The complete blood count was normal with a pack cell volume of 47% and normal kidney functiontests.A micturating cystourethrogram revealed a good bladder capacity with no vesico-ureteric refluxseen.Figure 4

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 penis was then de-gloved down to the root and parallel incisions made on the sides of the urethral plate to mobilize it from the corpora cavernosa. Figure 5. The urethral plate was tubularized over a stent (size 16Fr silicon catheter) using 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. Figure 6. A W-flap was raised in the pubic region and the suspensory ligament was released to give length to the penile shaft. The penile skin was then apposed with vicryl 3/0 sutures and the skin flap closed. We left a supra-pubic catheter in place for temporary urinary diversion to rest the urethral repair.

Postoperatively he received intravenouslevofloxacine 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 medicationson the third post-operative day

namely; tolterodine 2mg twice daily, oral levofloxacine500mg daily and metronidazole 400mg thrice daily as well ashematinics. He was discharged home on the 9th post-operative day with the urethral catheter and suprapubic catheter in situ.

Follow up and Outcomes Measure

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

DISCUSSION

Isolated continent epispadias is a very rare congenital anomaly usually present at one end of thespectrum of bladder exstrophy epispadias complex (BEEC) andconstituting 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 other 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 presents as bifid clitoris and poorly developedlabia minora. [7]

The diagnosis is typically clinical and easy to make especially in the male child and further investigations are only required to exclude otherconcomitant 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 adultsjust like in the index patient.[6] Braga et al in their series reported a mean age at the time of surgical repair 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 anomalyis alsocharacterized by aplasia of the dorsal part of the urethra.[1,7,10]

Treatment of epispadiasis surgical and it is aimed at giving the patient normal urinary control, a straight, cosmetically and functionally acceptable penis. In literature various surgical procedures to treat this condition have been described and they are usually challenging, even in the hands of experts. [4,6,8]The twouniversally adoptedsurgical techniques in therepair of epispadias are; the Modified Cantwell-Ransley technique described by Gearhatand the Mitchell-Bagli penile disassembly techniquepopularized in 1996.[2,4,8]Mitchell-Bagli described 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 that was turntubularized 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 Africanpopulation as previously alluded to by the authors in an earlier

published case report.[6] This same reason for late presentation of epispadias is further corroborated by Gite et al intheir series in India.[8]

Isolated male epispadias with continence remains a rare findingand comprises less than10% 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 affecting the bladder neck and external sphincter leading to incontinence and stress urinary dribbling.[1,4,6] The index patient did not present with any of these associations and prior to his surgery he and his spouse were reviewed by the psychiatristdue to concerns about hispsycho-sexual potential.[8]He had a one-stage repair using the modified cantwell-ransley technique as described in 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 after the repair to mitigate the problem of bladder spasms.[8]Our patient developed surgical site infection which is not an uncommon complication following this surgery [8] We successfully managed the infectionconservatively 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 the management.

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.

Financial Support and Sponsorship - Nil.

Conflict of Interest - There are no conflicts of interest.

REFERENCES

1. Beaman GM, Cervellione RM, Keene D, Reutter H, Newman WG. The Genomic Architecture of Bladder Exstrophy Epispadias Complex. Genes (Basel). 2021;12(8):1149. doi: 10.3390/genes12081149. PMID: 34440323; PMCID: PMC8391660.
2. Thomas JS, Shenoy M, Mushtaq I,Wood D. Long-term outcomes in primary male epispadias. J Pediatr Urol. 2020;16(1):80.e1 -80e6 https://doi.org/10.1016/j.jpurol.2019.10.027
3. Reddy SS, Inouye BM, Anele UA, Abdelwahab, M., Le, B., Gearhart, J. P. Sexual Health Outcomes in Adults with Complete Male Epispadias. *J Urol*. 2015;194(4):1091-1095. doi:10.1016/j.juro.2015.04.082
4. Shehzad D, Shehzad M, Mutaal A. A Rare Case of Isolated Male Epispadias. Cureus. 2023;15(9):e46160. doi: 10.7759/cureus.46160. PMID: 37905290; PMCID: PMC10613130.
5. Braga LH, Lorenzo AJ, Bägli DJ, Khoury AE, Pippi Salle JL. Outcome analysis of isolated male epispadias: single center experience with 33 cases. J Urol.

2008;179(3):1107-12. doi: 10.1016/j.juro.2007.10.095. Epub 2008 Jan 18. PMID: 18206921.

6. Atim T, Mustapha A. Penopubic epispadias repair in adulthood: Surgical and functional outcome with modified Cantwell-Ransley procedure. Sub-Saharan Afr J Med. 2015;2:179-83. **DOI:** 10.4103/2384-5147.172448

7. Ali RN, Hussain K, Khalid H, Yasin Z, Attiq-ur-Rehman. A case report on continent isolated male epispadias. Professional Med J 2020; 27(6):1320-1322. DOI: 10.29309/TPMJ/2020.27.06.3899
8. Gite VA, Jain HM, Bote SM, Nikose JV. Modified Cantwell-Ransley repair for isolated continent epispadias in adult: Our experience. Indian J Plast Surg. 2017; 50 (1):68-73. doi:10.4103/ijps.IJPS_243_16.
9. Maitama HY, Ahmed M, Bello A, Mbibu HN. Epispadias with complete prepuce: A rare anomaly.Afr J Urol.2012; 18, 90–92
10. Othmane Y, Touzani A. Isolated male epispadias. Pan Afr Med J. 2017;28:202. doi: 10.11604/pamj.2017.28.202.14220
11. Hafez AT, Helmy T. Complete penile disassembly for epispadias repair in postpubertal patients. *Urology*. 2011; 78 (6):1407-1410.

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

Figure 2. Plain Pelvic X- ray showing normal anatomy

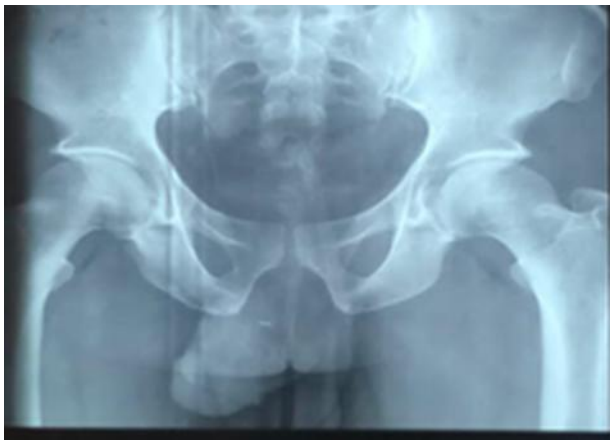

Figure 3. Clinical picture showing degree of chordee in epispadias

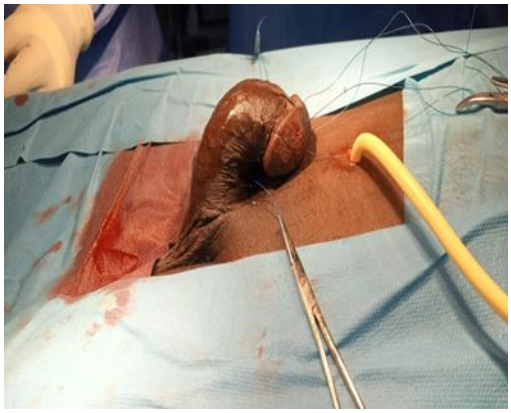

Figure 4. Micturating cysto-urethrogram showing normal bladder capacity and no vesico-ureteric reflux

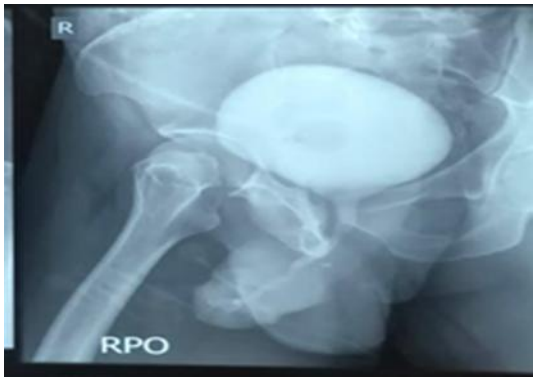

Figure 5. Separation of the urethral plate from the cavernosa

Figure 6. Immediate post-operative clinical picture

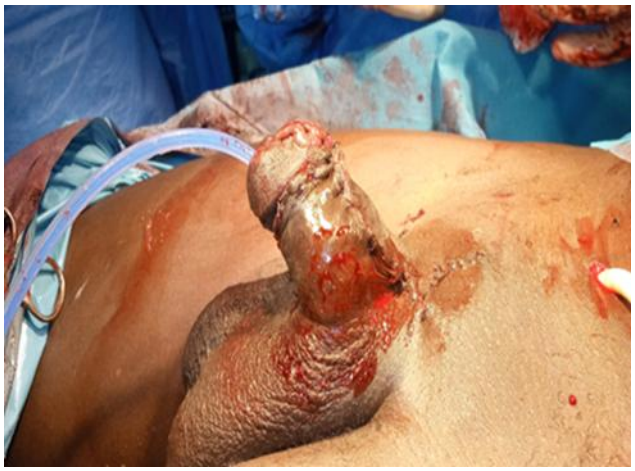

Figure 7. Clinical picture 2 months after repair