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# Congenital urinary incontinence in a 4-year old girl due to isolated epispadias: A case report and literature review

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**Abstract**
Epispadias is a congenital anomaly in which the urethral meatus opens dorsally on top of the penis, medial to the glands, or with absent or bifid clitoris in females. It often occurs as part of a complex anomaly called the bladder extrophy epispadias complex (BEEC) along with exposed bladder and bifid pelvic brim. The diagnosis of bladder extrophy epispadias complex is often clinical with identifications of the various component of the anomalies. Although this condition is rare it present with wide spectrum of deformities which ranged from mild to more complex anomalies, its management especially the complex variety has been a major challenge to pediatric surgeons worldwide. With a male to female ratio of 2-3:1, the occurrence of isolated epispadias in female patient is even rarer. Our patient is a 4-year old girl who presented with urinary incontinence since birth. She has never achieved urinary continence up to the age of 4-years, there is no history of any prior urethral procedures, no history of any trauma to the perineum, no passage of stone or blood in the urine. Child is the second of her mother's three children and her younger ones has achieved urinary continence. Pregnancy, delivery and puerperal history were essentially uneventful. On examination, we found a young girl, calm, but apprehensive otherwise stable, abdomen was full, moves with respiration, no area of tenderness, no palpable organomegaly, no intra-abdominal masses felt, there was distorted female genitalia with bifid clitoris and patulous urethral meatus. There was a normal vaginal orifice with intact hymen with poorly developed mons pubis and labia. A clinical diagnosis of isolated epispadias with urinary incontinence to rule out ectopic ureter in a 4-year old girl was made. She was worked up and prepared for surgical repair, and had a single stage procedure done. Post-operative cosmetic appearance of the genitalia was acceptable. Urethral catheter was removed on the postoperative day 10 and patient had achieved some continent of urine. She generally did well and has been on outpatient follow up visits at the pediatrics surgical outpatient clinic.

**Keywords:** Epispadias, single stage repair, incontinence, four years old

## Introduction
Epispadias is a rare congenital anomaly in which the urethra opens on the dorsal aspect of the penis or with bifid clitoris in females and other mal-development of the female external genitalia [1], it occurs as a result of failure of the urethral tube to tubularize on its dorsal aspect [1]. Epispadias is often seen as part of a component of a more complex anomaly called the bladder extrophy epispadias complex (BEEC) which despite being rare, poised a serious challenge to the pediatric surgeon. Isolated female epispadias is a very rare condition, it has an incidence of 1/480,000 female live birth [2]. The affected females present most commonly with urinary incontinence and abnormal appearance of the female external genitalia; bifid clitoris, ill developed mons pubis, patulous urethral meatus, widened bladder neck, small capacity bladder and an incompetent sphincteric mechanism [3]. The diagnosis of this condition is essentially clinical by observing the external genitalia and noting the various features and presentation of the condition, it is also easily missed diagnosis for an inexperience clinician. Ectopically located ureter into the neck of the bladder below the sphincter or into the vagina may be a differential. Radiological investigations can be done to evaluate other associated anomalies in the genitourinary system; voiding cystourethrogram, to check for bladder capacity which is reduced in this condition, vesicourethral reflux is also a common finding in epispadias as it is also a short urethra [4]. Investigations can also be done to establish the biochemical parameters of the patients, the hematological profile and to prepare the patient for operative management.

We shall discuss the single stage repair of this condition which is what we offer our patient and also look into the existing literature on other technique or methods for the management of this condition for a better enlightenment and to also spur the index of suspicion of this condition in children presenting with failure to achieve urinary continence in time in other to prevent delay in diagnosis and the deleterious impact of such on the psychosocial and psychosexual wellbeing of the girl child.

**Case report**

A four-year-old girl who was brought in by her parents with urinary incontinence since birth, she has failed to achieve urinary continence up to the age of 4-years. There was no history of any prior urethral procedures, no history of trauma to the perineum, no passage of stone or blood in the urine and no cloudiness of urine. Child is the second of her mother's 3 children and her younger ones had achieved urinary continence already. Pregnancy, delivery and puerperal history were essentially uneventful; mother has booked the pregnancy and taken her routine antenatal medications and pregnancy was taken to term, delivery was in a hospital with a good APGAR score and a birth weight of 3.4kg. No history of febrile illness by the mother during the pregnancy, no history of ingestion of un-prescribed medication during pregnancy, although patient has admitted to taken some traditional herbal concoctions during pregnancy. There is no family history of similar condition in any of the sibling or any family history of a congenital anomalies. Patient's developmental milestones were normal. They visited several peripheral hospitals and they attributed it to urinary tract infection and poor training, she was given several antibiotics but to no avail. When we examined her, we found a young girl, calm and stable but apprehensive, encompass by a stench of urine. Abdomen was full, moves with respiration, no area of tenderness, no palpable organomegaly, no intra-abdominal masses felt. There was distorted female genitalia with bifid clitoris and patulous urethral meatus, as well as normal vaginal orifice with intact hymenal rim with poorly developed mons pubis and labia Fig. 1. Her pulse rate was 92beats/min, blood pressure 120/70mmmHg, weight 30kg and height 112cm and a body mass index of 23.9kg/m$^2$. Retrograde urethrogram suggested an ectopic left ureter into the vagina because some contrast was found in the vagina. Contrast enhanced CT scan was then done and showed normally inserted both ureters into the bladder Fig. 2. We therefore did examination under anesthesia (EUA) which showed no urine coming from the vagina.

Patient in lithotomy position under general anesthesia with endotracheal tube in situ, a vertical incision was done on either side of the urethral plate and carefully dissected down to the bladder neck Fig. 3. The urethra was mobilized from both sides with the urethral plate in the middle, a size 8 silicone urethral catheter was inserted and the urethra was reconstructed over it. Clitoral reconstruction was done. Patient tolerated the procedure very well and had an uneventful post-operative period. The catheter was removed after ten days and there was satisfactory cosmetic appearance of the external genitalia Fig. 4. Patient achieved continence both day and night with an initial dryness of up to 2 hours with very few nights wet for which she underwent rehabilitation and conservative management with pelvic floor exercise. Two months after the surgery, she had achieved complete dryness both day and night and had resumed school.

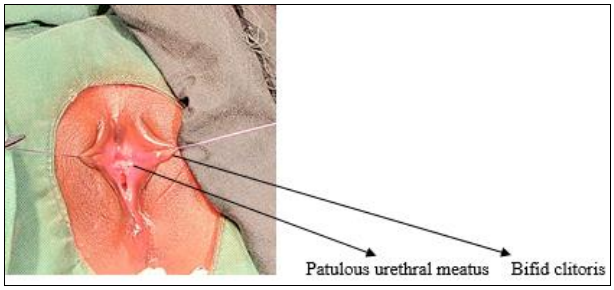


**Fig 1:** Female epispadias with bifid clitoris, patulous urethral meatus and ill developed labia.

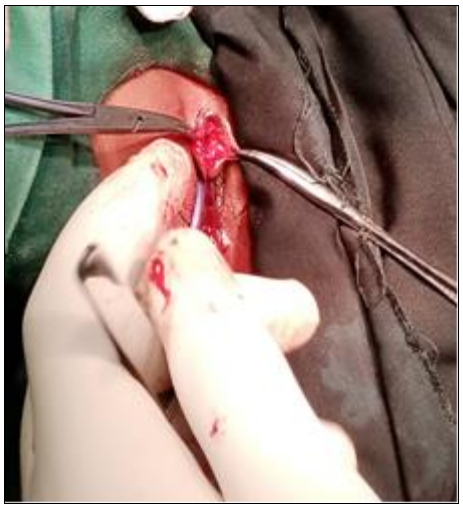

**Fig 2:** Incision around the urethral plate before it was tubularize over a stent/catheter

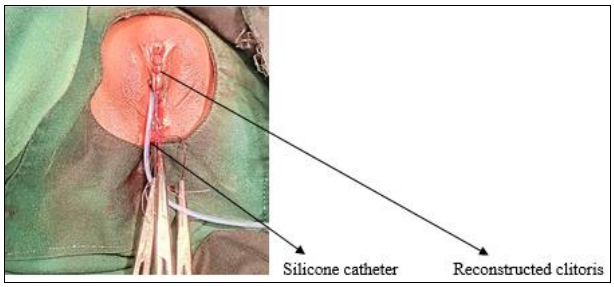


**Fig 3:** Post-operative picture with the silicone catheter in-situ

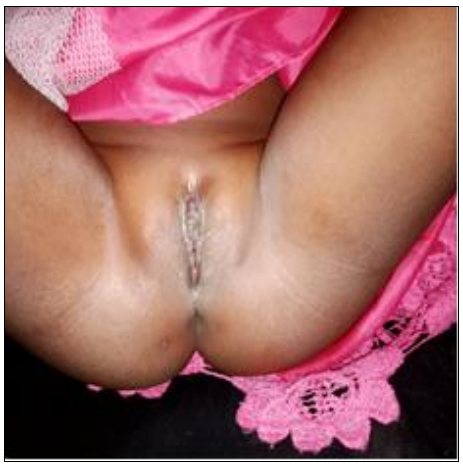

**Fig 4:** External genitalia after repair and removal of urethral catheter

## Discussion

Isolated female epispadias is an extremely rare congenital anomaly of the lower genitourinary system in girls with an incidence of one in 484,000 female children [2], epispadias often occurs as part of a complex of anomalies called the bladder extrophy epispadias complex [2, 4], which although rare poised a serious challenge to the pediatric surgeon. Attempt has also been made by Davis to classify the isolated female epispadias into lesser degree with patulous urethral orifice, to intermediate degree which has its urethral plate partially split along its length but with normal bladder neck. Such patients present with occasional incontinence to the worst form which has the entire length of the urethra involved and the bladder neck thus rendering the sphincteric mechanism incompetent, such patient as in our case report, presented with complete incontinence of urine since birth, with marked reduction in bladder capacity due to under development of the bladder as a result of non-retention of urine[5] other features include bifid or absence of clitoris, underdeveloped mons pubis, poorly develop labia. This diagnosis can be easily missed if the external genitalia are not carefully examined. High index of suspicion is also needed in a female child who has failed to achieve continence since birth with abnormal appearance of the external genitalia. This diagnosis is mostly clinical with the history of incontinence since birth and demonstration of the characteristic appearance of the external genitalia. Failure to make appropriate diagnosis as seen in this case when they visited several hospitals before presenting to us, can lead to a vicious cycle of cascading psychosocial and psychosexual problems on the girl child with increasing level of hopelessness as the urinary leakage persisted. Radiological investigations are also necessary to rule out other causes of incontinence and or vesicoureteral reflux which could be associated with the condition in up to 75% of the time[6]. Cystourethroscopy can be done to access the bladder capacity of the patient and delineate the position of the ureteric orifices and also done for follow up to show improvement in bladder capacity of the patient post operatively. We did not do Cystourethroscopy due to lack of pediatrics scopes in our institution. Intravenous urography (IVU) was done and it shows both the right and left ureters normally situated, the bladder and the urethra with wide bladder neck Fig. 5.

The aim of treatment of this patient is to achieve urinary continence, reconstruction of a functional and cosmetically appealing and acceptable external genitalia for the girl child and preservation of the upper tract and kidneys from recurrent infection and insults. This can be achieved by surgical reconstruction of the bladder neck, urethra and the external genitalia in a single stage procedure [7, 8, 9]. Staged procedures are less advocated for now as those involves multiple sessions of anesthesia and surgery resulting in higher morbidities with no any demonstrable superior outcome [10] We thus advocate for the single stage technique for the repair of isolated female epispadias for effective continence restoration, acceptable cosmetics and genital functions, however a more longer follow up is needed and more cases are needed to solidify the superiority of this technique over the staged approach.

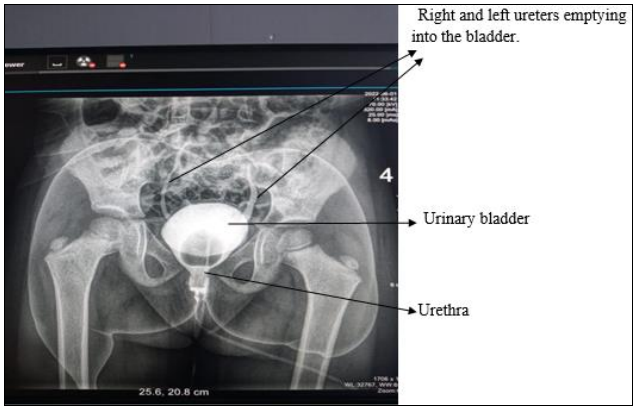


**Fig 5:** Intravenous urography showing the ureters emptying into the bladder, with no ectopic ureters or abnormal positioning. Notice also the wide urethra and bladder neck

## Reference

1. Anand S, Lotfollahzadeh S. Epispadias. 2022 Jun 3. In: Stat Pearls[internet]. Treasure Island (FL): Stat Pearls Pubishing; c2022 Jan. PMID: 33085327.
2. Allen L, Rodjani A, Kelly M, Inoune M, Hutson JM. female epispadias: are we missing the diagnosis? BJU int. 2004 Sep 1;94(4):613-615.
3. Atilgan D, Uluocak N, Erdemir F, Parklaktas BS. Female epispadias: A case report and review of literature. Kaohsiung J Med. Sci. 2009 Nov 1;25(11):613-616.
4. Ali S, Moosa H, Khan SA. Isolated female epispadias with urinary incontinence. J Coll. Phys. Surg. Pak. 2020 Apr;30(4):438-439.
5. Gearhart JP, Peppas DS, Jeffs RD. Complete genitourinary reconstruction in female epispadias. J Urol. 1993;149:1110.
6. Kramer SA, Kelalis PP. Surgical correction of female epispadias. Eur Urol. 1982;8:321-4.
7. Young HH. A new operation for cure of incontinence associated with epispadias. J Urol. 1922;7:1-32.
8. Ahmed S, Gough DCS. Reconstruction of the mons venereum using omentum in female patients with epispadias/extrophy. Br J Urol. 1997;79:987-8.11
9. Bhat AL, Bhat M, Sharma R, *et al*. Single-stage perineal urethroplasty for continence in female epispadias: A preliminary report. Urology. 2008;72:300-3.10.
10. Mollard P, Basset T, Mure PY, Female epispadias. J Urol. 1997;158:1543-6.