**Case Report**

New Indian Journal of Surgery
Volume 6 Number 4, October - December 2015
DOI : https://dx.doi.org/10.21088/nijs.0976.4747.6415.8

# Bilateral Single System Ectopic Ureter with Bladder Agenesis Opening Intovaginalised Urogenital Sinus

**Shivam Priyadarshi**

*Professor, Dept. of Urology, SMS Medical College & Hospital, Jaipur

## Abstract

Single system ectopic ureters are rare and bilateral single system ectopic ureters (BSSEU) are even rarer. Common locations of openings of such ureters include vagina and vestibule in the females, and posterior urethra and seminal vesicles in the males. BSSEU opening into a vaginalized urogenital sinus with absence of urinary bladder is an extremely rare occurrence. Such a case is being reported and its management is discussed briefly.

**Keywords:** Single System Ectopic Ureters ; Vaginalized Urogenital Sinus; Bladder Agenesis; Continent Cutaneous Diversion.

## Introduction

Almost 80% of ectopic ureters are associated with duplicated system and 5-17% of them are bilateral [1]. Bilateral single system ectopic ureters (BSSEU) is a rare entity that may be associated with a hypoplastic bladder and bilateral renal abnormalities, typically dysplasia [2]. Vaginalized urogenital sinus is the term used when ureters drain into a well developed vagina that has a near normal vaginal introitus.3 We report a case of BSSEU opening into vaginalised urogenital sinus with bladder agenesis. Only four other similar cases have been reported so far in the literature [3,4,11,12].

## Case Report

A 5 year-old girl presented with continuous dribbling of urine without any voiding stream since birth. There was no history of any bowel complaints. Her mother denied use of any drugs or history of fever during pregnancy. Family history for congenital disorders was negative. On examination, the external genitalia were of female phenotype. On separating labia majora, a single opening with continuous urine leakage was seen (Figure 1). There was no skeletal defect or any other associated anomaly. Rest of the general physical examination, neurological examination and anal tone were normal. Routine blood and urine tests were all normal. On ultrasonography (USG) bilateral mild to moderate hydroureteronephrosis with good parenchymal thickness was noted. Intravenous urography (IVU) revealed bilateral mild to moderate hydroureteronephrosis with bilateral ectopic ureters

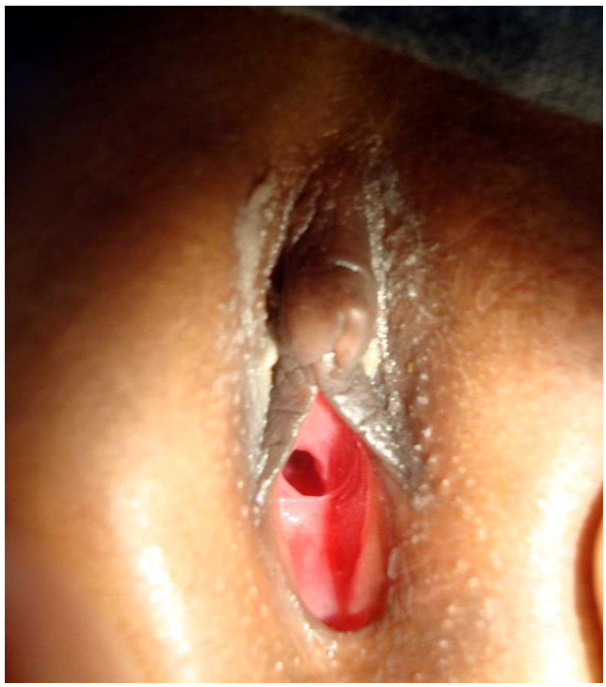

**Fig. 1:** Normal appearing external genitalia with a single opening

**Corresponding Author: Dr. Shivam Priyadarshi,** C-80, Gole Market, Jawahar Nagar, Jaipur – 302004 (Rajasthan).
E-mail: spriyadarshi64@gmail.com

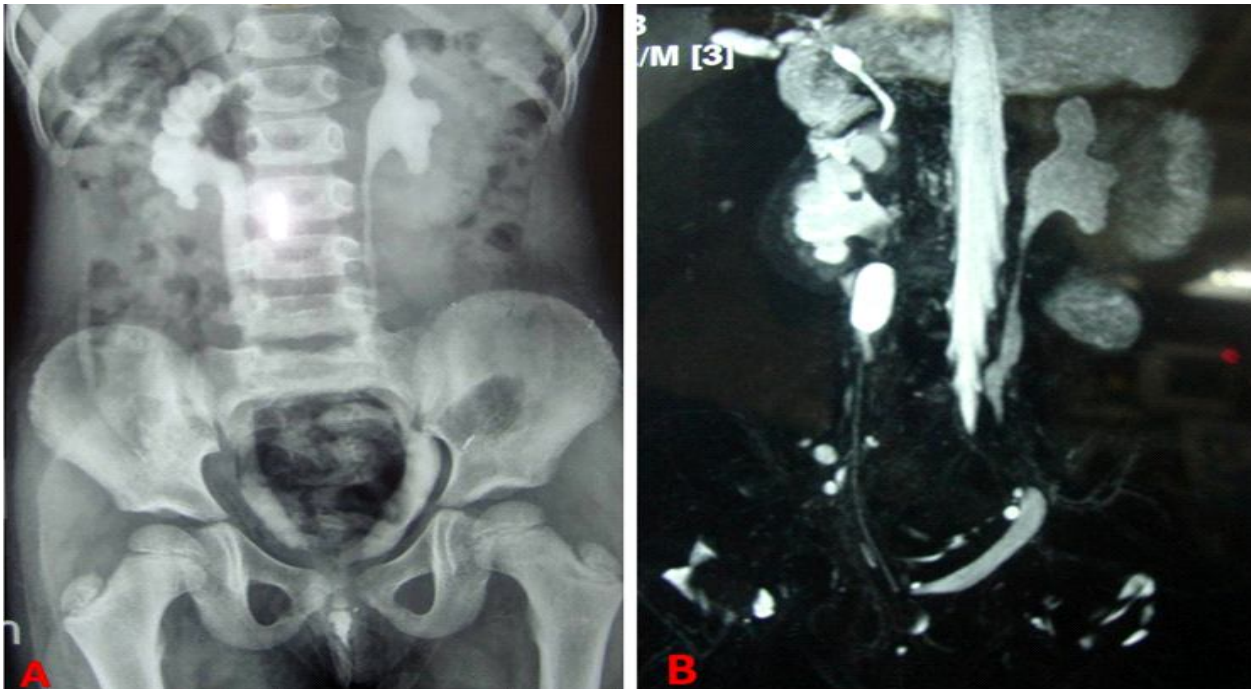


**Fig. 2A:** Intravenous urography (IVU) film showing bilateral mild to moderate hydroureteronephrosis with bilateral ectopic ureters. **Fig. 2B:** Magnetic resonance imaging (MRI) film showing bilateral ureters opening into the vagina

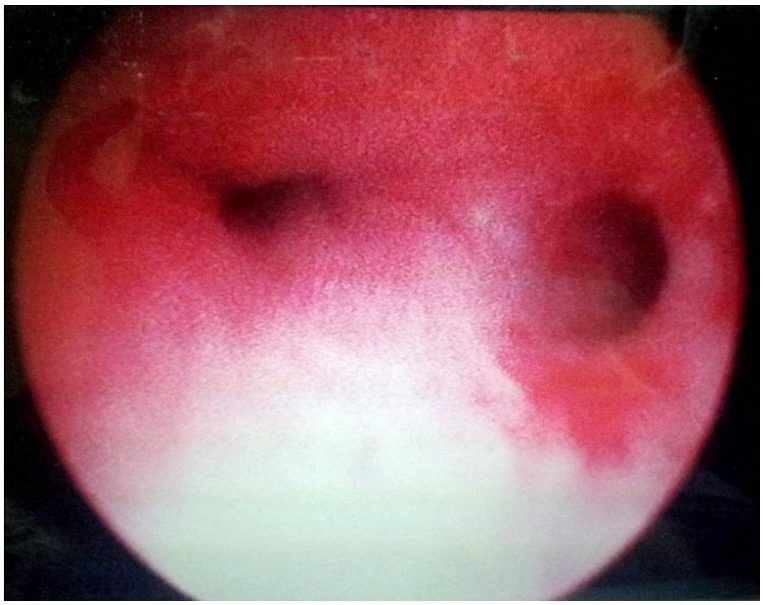

**Fig. 3:** Endoscopic examination through vaginal opening revealed bilateral dilated ureteral orifices opening into a vaginaliged urogenital sinus without any identifiable trigone and bladder neck

(Figure 2A). However, no contrast was seen in the urinary bladder. Rather, small amount of contrast was seen in the vagina. Magnetic resonance imaging (MRI) with urography was performed which revealed absence of the urinary bladder, with bilateral ureters opening into the vagina (Figure 2B). Uterus and ovaries were normal. Karyotyping revealed normal 46XX genotype. On endoscopic examination under anesthesia, bilateral dilated ureteral orifices were seen opening into vaginaliged urogenital sinus without any identifiable trigone and bladder neck (Figure 3).

Continent cutaneous diversion using a segment of sigmoid colon and Mitrofanoff's principle with bilateral ureteral reimplantation was performed. Clean intermittent catheterization of cutaneous stoma was started 3 weeks after the procedure. At 9 months of follow up, patient is doing well, has capacious neobladder (Figure 4A and 4B) and continence for 4-6 h of dry intervals. She has preserved renal function without any metabolic abnormality.

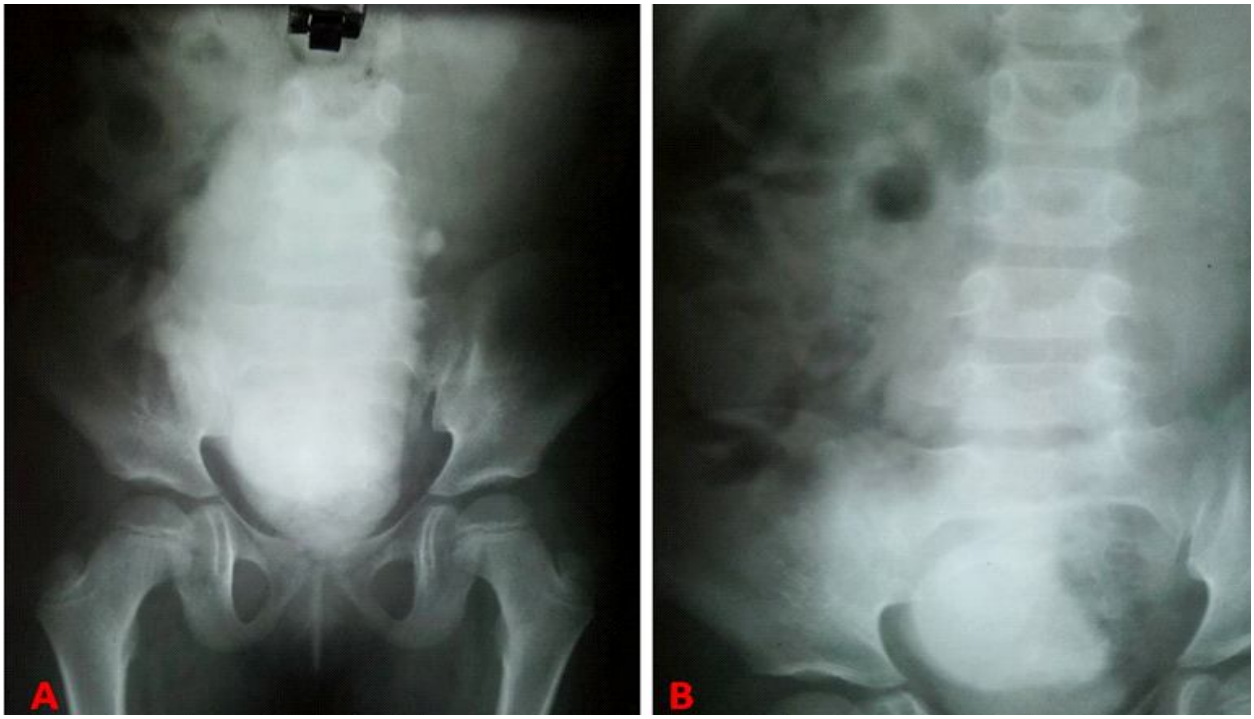


**Fig. 4A:** Follow up imaging showing prevoid capacious neobladder. **Fig. 4B:** Postvoid film showing adequate emptying

## Discussion

Ectopic ureter is the term used to denote any ureter whose orifice terminates anywhere (inside or outside urinary tract) other than the normal trigonal location [5]. Approximately, 80% of ectopic ureters are associated with duplicated pelvicalyceal system. Single system ectopic ureters are rare as do bilateral ectopic ureters (5-17%) [1]. BSSEU is even a rarer entity that and may be associated with hypoplasia of the urinary bladder and bilateral renal abnormalities, typically dysplasia [2]. The embryological basis of single system ectopic ureter is the cranial origin of ureteral bud from the mesonephric duct, which results in its delayed incorporation into the urogenital sinus, and thus abnormally limited ingrowth of the mesenchyme required for normal development of the trigone and bladder neck [6]. In patients with bladder agenesis and without any hind gut abnormalities like in our case, it is proposed that the division of cloaca is normal but there is either a primary developmental failure or secondary atrophy of the urogenital sinus. The latter phenomenon is due to failure to incorporate the mesonephric ducts and ureter into the trigone leading to lack of distension with urine and consequent atrophy [7].

Further, vaginalized urogenital sinus is the term used to describe when BSSEU are inserted into a well developed vagina that has a normal or near normal introitus [3].

Kesavan et al, showed that the bladder neck and trigone is maldeveloped in 75% of bilateral and 54% of unilateral ectopic ureters [8]. The ectopic ureter itself is also abnormal, usually to a greater degree in the single system than in the duplicated system. It is variably dilated and drainage is impaired [9]. Muscle cells may show severe alterations on ultra-structure studies. Whether these changes are developmental or acquired is not yet known [10].

Present case of BSSEU with openings into vaginalized urogenital sinus associated with absence of bladder and urethra is extremely rare. Only four such cases are on record so far in the literature [3,4,11,12]. Sheldon and Welch4 reported first such case in which the urogenital anomalies were more severe with a rudimentary vagina and bilateral dysplastic kidneys, resulting in end stage renal failure. In the second case BSSEU were opening into a vaginalized urogenital sinus with capacity of 20 ml and poorly developed trigone and bladder neck [3]. Nazimet al [11] reported another case of BSSEU draining into the urogenital sinus with right non-functioning kidney. Mandalet al [12] reported another similar case in which BSSEU were draining into dilated vagina with bladder being absent.

Our case was more typical. Bladder and urethra were absent. BSSEU were opening into vaginalized urogenital sinus with no identifiable trigone or bladder neck. There were no other associated abnormalities making the management relatively

easier. Attainment of continence with preservation of renal function is the primary issue in these cases. This re-emphasizes the need for continent diversion as the primary procedure in complex cases of BSSEU with absent or poorly developed trigone and bladder neck. So in our case continent cutaneous diversion with bilateral ureteric reimplantation was the viable option. Although bladder preservation and bladder neck reconstruction have been advocated by many in case of BSSEU or solitary ectopic ureter, in these cases ectopic ureter opened either into a normal urethra or vagina with a normal urethra.

Our case report adds to the spectrum of literature on BSSEU. In our opinion, continent cutaneous diversion should be adopted as a single stage definitive procedure in such rare and difficult cases to avoid continuing disabilities and poor outcomes in terms of upper and lower tract function. Further, management modalities should be tailored according to the anomalies in urinary tract as well as the reproductive tract in an individual patient.

## Conclusion

A rare case of bilateral single system ectopic ureters with agenesis of bladder and ureters opening into a vaginalized urogenital sinus is reported. Continent cutaneous diversion as a single stage definitive procedure is a viable treatment option to avoid disabilities and deterioration of upper and lower tract function in such rare and complex cases.

## References

1. Ahmed S, Barker A. Single-system ectopic ureters: A review of 12 cases. J PediatrSurg 1992; 27: 491-6.
2. Johnin K, Narita M, Kim CJ. Bilateral single system ectopic ureters with hypoplastic bladder: how we treat these challenging entities? J Pediatr Urol. 2007; 3: 243-6.
3. Singh BP, Pathak HR, Andankar AG. Bilateral single system ectopic ureters opening into vaginalized urogenital sinus. Indian J Urol 2010; 26: 126-8.
4. Sheldon CA, Welch TR. Total anatomic urinary replacement and renal transplantation: A surgical strategy to correct severe genitourinary anomalies.JPediatrSurg. 1998; 33: 635-8.
5. Glassberg KI, Braren V, Duckett JW, Jacobs EC, King LR, Lebowitz RL, et al. Suggested terminology for duplex systems, ectopic ureters and ureteroceles. J Urol 1984; 132: 1153-4.
6. Redman JF, Lightfoot ML, Reddy PP. Bilateral single ureteral ectopia in a boy. Urology 2002; 60: 514.
7. Krull CL, Heyns CF, de Klerk DP. Agenesis of the bladder and urethra: a case report. J Urol. 1988; 140: 793-4.