Bilateral single-system ectopic ureter (BSSEU) is often associated with underdeveloped incompetent bladder neck; hence, to achieve continence, bladder neck reconstruction (BNR) is usually advocated with ureteric reimplantation. Presented here is a 14-year-old girl with BSSEU who achieved continence without BNR. An attempt is made to look at factors that could identify patients in whom BNR could be avoided.

**Keywords:** Ectopic ureter, incontinence, single system, ureteric reimplantation

**INTRODUCTION**

Bilateral single system ectopic ureters (BSSEU) are the rarest anomalies. These are usually associated with bladder neck incompetence as the bladder neck is not well formed. It is said that ureteric reimplantation alone is usually not sufficient. Many patients need some bladder neck procedure.

**CASE REPORT**

A 14-year-old girl (weight, 25 kg), born out of a third-degree consanguineous marriage, presented with total urinary incontinence since birth. Clinical examination was unremarkable with no signs of any neurologic deficit. Her biochemical workup was normal. Abdominal ultrasound and intravenous urography [Figure 1] revealed moderate right hydronephrosis with suspicion of right ureter opening in ectopic location in the vagina. Left kidney was normal, and the left ureter could not be traced. Micturating cystogram (MCU) showed approximately about 100 ml bladder capacity with no reflux on either side [Figure 2].

When examined under anesthesia, urethral and vaginal openings were normal, and she was continuously leaking urine from the urethral opening. No other abnormal openings were noted [Figure 3]. Cystoscopy showed a wide urethra and a bladder that could be distended to 100 ml. Both ureteric orifices were opening in the urethra, and trigon could not be identified; hence, a diagnosis of bilateral single-system ectopic ureter (BSSEU) was made [Figure 4].

After discussing various surgical options, a decision was taken to undertake bilateral ureteric reimplantation first, and surgery for continence if needed, at a later date. This decision was made based on two factors. The child did not have any malformations that could potentially impede continence, particularly cloacal or spinal anomalies where anatomical or neurological continence mechanisms are directly undermined. She had a 100 ml bladder capacity on MCU and cystoscopy. Bilateral ureteric reimplantation was performed by a combined intra–extravesical technique. Right ureteric tailoring was needed.

Postoperatively, recovery was uneventful. Although there was grade 1 stress incontinence and occasional nocturnal enuresis initially, she gradually learned to hold urine for a long time. One and half years postoperatively, she is continent with a dry period of 2 to 3 h. She has no nocturnal enuresis with occasional nighttime void.

**Address for correspondence:** Dr. Prashant Motiram Mulawkar, Tirthankar Superspeciality Hospital, Akola, Maharashtra, India. E-mail: pmulawkar@hotmail.com

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Bladder capacity has increased to 348 ml with complete resolution of hydronephrosis.

**DISCUSSION**

BSSEU is a rare congenital anomaly where the ureter opens caudal to the trigon in an unduplicated system.\[^1\] The ureter can open anywhere from the bladder neck to the perineum or into the vagina, uterus, or rectum. As the ureters do not open in the trigon, neither the trigon nor the bladder neck may be properly developed.\[^2\]

Management of BSSEU is challenging because of small bladder capacity and nonfunctional bladder neck sphincter. The aims of management are repositioning of the ureteric opening in the bladder, improving the bladder capacity, and correcting the urinary continence by creating urethral continence mechanism.\[^2\] Correction of reflux and preservation of renal function\[^3\] is also important.

Literature is not clear whether only ureteral reimplantation is adequate or if there is a need for bladder augmentation and bladder neck reconstruction (BNR). Continence rates in techniques without urinary diversion have been reported to be abysmally low at 20%.\[^4\] Jayanthi and colleagues\[^5\] reported a series of six girls and one boy who underwent many surgical procedures to achieve continence, but none worked. Authors felt that performing ureteric reimplantation would obviously not correct the urethral control mechanism. Moreover, doing bilateral reimplantation as the first stage would make subsequent surgery of BNR difficult due to the postoperative scarring. They proposed that the continence could only be achieved by closing the bladder neck and diverting urine through appendicovesicostomy.

Multiple similar reports in favor of bladder neck repair are available in the literature. Heuser\[^6\] suggested that only ureteral reimplantation is not adequate as the incontinence was thought due to maldeveloped trigon and bladder neck. The problem of incontinence was also related to the short urethra and ill-formed bladder neck;\[^2\] hence, Young’s operation for epispadias was performed in some of these patients with varying success. In the case series reported by Wakhlu,\[^7\] there were five girls with BSSEU. Four underwent bilateral ureteric reimplantation but were incontinent. Both the patients by Heuser *et al.*\[^6\] who underwent ureteric reimplantation and were incontinent. The older one of these needed implantation of artificial urethral sphincter.

Johnin *et al.*\[^3\] went a step further to question the need for abandoning the bladder and bladder neck closure. He proposed that every attempt should be made to achieve continence through the urethra in these children. In contrast, Podestà *et al.*\[^8\] proposed a conservative approach (ureteric reimplantation alone without BNR) in almost all cases. Such approach is especially useful when at least one ureter is above the level of urogenital diaphragm. Ciongradi\[^9\] reported a case of USSEU with absent contralateral kidney managed by ureteric
reimplantation alone. One girl with BSSEU who underwent bilateral ureteric reimplantation and became continent was reported by Ahmed.[10]

In a very interesting review by Stavrinides et al.,[11] they studied eight children with BSSEU and found the continence to be linked to the preoperative bladder capacity. They found that if the preoperative investigations showed an empty bladder on serial ultrasound and/or a wide-open BN with small or even moderate bladder capacity at cystoscopy, bladder neck repair is surely needed. In children with bladder filling to at least 30% of expected bladder capacity for age on preoperative ultrasound or apposition of the BN at cystoscopy, satisfactory continence could be achieved after ureteric reimplantation alone. Podestà et al.[8] also had similar observations. Our patient did not have any neurological co-abnormalities and had shown more than 100 ml capacity on MCU and cystoscopy so the decision of avoiding BN repair was justified.

Although ureteric reimplantation is the primary treatment, there is no unanimity about the technique of ureteric reimplantation. Podestà et al.[8] has recommended intravesical reimplantation. The authors feel that detachment near the bladder neck (by extravesical approach) can cause damage to the bladder neck and local innervation. In contrast, the proponents of extravesical reimplantation feel that the procedure of separating the ureter from the bladder neck or urethra may damage the bladder neck which is already incompetent.[12]

On the basis of available literature, it seems only bilateral ureteric reimplantation is likely to succeed if both the ureters are opening in the urethra and at least one ureter is above the level of the urogenital diaphragm. As the urine from the ureter enters the urinary tract below the internal sphincter,[13] some urine goes retrograde in the bladder while some leaks out. This probably explains the 100 ml bladder capacity seen on MCU in our patient. Our patient had an acceptable bladder capacity of 100 ml, and both the ureters were opening in the urethra, so a simple procedure such as ureteric reimplantation gave good results.

It can be argued that the increase in the bladder capacity from around 100 to around 350 postsurgery may not be according to the presumed bladder capacity growth of around 30 ml/year. However, the rate of bladder growth of around 30 ml/year has been found only till the age of 9 years.[14] The initial assessment in our case was approximate on cystoscopy and MCU. Further, the functional bladder capacity depends on many factors.[15] It has been correlated to other child factors like in one study, they calculated FBC as FBC = −268.4 + 7.38 × flow time + 3.15 × suprasternal height + 1.25 × thickness of fat fold. Moreover, unexpectedly larger bladder growth after bilateral ureteric reimplantation has also been demonstrated in children with complete primary repair of bladder exstrophy.[16] The formulae for expected bladder capacity may not be universally applicable. A systematic review[17] has suggested that such formulae for expected bladder capacity should be abandoned.

In females, mechanism of urinary incontinence in BSSEU is manifold. It is well understood that the female urethra is not just a urinary conduit but a functional unit that plays a major role in continence. Our patient was older, without neurologic abnormalities and with 100 ml bladder capacity; hence avoiding BN repair was justified in this child. Although no decisions can be made on a single case report, based on our experience and based on the review of the literature, we propose looking at these factors before a decision of surgical plan is made. Availability of data and material

All the data pertaining to the case are presented herewith. No additional data are available.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published, and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.
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Conflicts of interest

There are no conflicts of interest.

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