Bilateral single system ectopic ureters: Case report with literature review

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ABSTRACT

Bilateral single system ureteral ectopia (BSSEU) is one of the rarest entities in urology, with less than 80 cases reported so far. Incontinence resulting from the underlying anomaly can be devastating to the child. It is generally agreed that suitable urinary continence and long dry intervals are seldom obtainable because of poorly developed trigone and bladder neck area. We herein report a case of BSSEU managed by bilateral ureteric reimplantation, achieving satisfactory continence and bladder capacity without the need for urinary diversion or bladder neck reconstruction.

Key words: Ectopic ureter, incontinence, megaureter

INTRODUCTION

An ectopic ureter opens at a site other than the posterio-lateral aspect of the trigone.[1] The embryological basis of single system ureteral ectopia is the cranial origin of the ureteral bud from the mesonephric duct. This results in a delay in its incorporation into the urogenital sinus, which limits the ingrowth of the mesenchyme necessary for the formation of the trigone and the vesical neck.[2] Therefore, an absent trigone or a poorly developed bladder neck is invariably encountered in patients with bilateral single system ureteral ectopia (BSSEU). Urinary incontinence is frequently noted, especially in females.[2] In children with small capacity bladder, the bladder may or may not regain its normal size and function. Thus, this anomaly possesses a major challenge to a paediatric surgeon or a paediatric urologist.

Despite this embryological absence of bladder neck and trigone, some patients attain continence with ureter implantation alone without bladder neck reconstruction or urinary diversion. We are presenting a case of bilateral single system ectopic ureter in which the patient became continent with bilateral ureteric reimplantation only.

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CASE REPORT

A three-year-old female child presented with incontinence, recurrent urinary tract infection and fever since birth. In spite of total incontinence, she had a normal voiding pattern after toilet training. On examination, her general condition was poor and she had subnormal growth. A per abdomen examination revealed bilateral flank fullness with palpable lump in the lumbar region extending to the corresponding iliac regions. Her undergarments were wet and had a uriniferous odour. All the routine investigations were normal including kidney function tests.

Ultrasonography revealed bilateral gross hydro-uretero-nephrosis. Intravenous urogram [Figure 1] revealed right hydro-uretero-nephrosis with nonvisualized left kidney. Voiding cystogram [Figure 2] revealed adequate bladder capacity and right vesico-ureteric reflux in voiding phase. Left ante grade pyelouretogram [Figure 3] revealed grossly dilated left pyelocalyceal system and ureter till its lower end. Cystoscopy and vaginoscopy were carried out under general anaesthesia. Right ureteric orifice was found just distal to the bladder neck and left orifice just proximal to the bladder neck. The bladder neck was developed with an ill-defined trigone. Bladder capacity was approximately 100 ml. A DTPA renal scan revealed 70 and 30% relative function of right and left kidney with bilateral hydro-uretero-nephrosis. The GFR was 56 and 24 ml/min/1.73 m^2 in right and left kidney respectively. Definitive diagnosis of bilateral single system ectopic ureter was made. Bilateral tapering and reimplantation of ureters into bladder were performed. Postoperative recovery was uneventful. In the immediate postoperative period, the patient was totally continent but had to void frequently, which improved later. Renal function of both kidneys improved, but substantially in the right kidney. At 1



Figure 1: Intravenous urogram showing right hydro-uretero-nephrosis with nonvisualized left kidney



Figure 2: MCU showing adequate bladder capacity and right vesicoureteric reflux



Figure 3: Left antegrade pyeloureterogram showing grossly dilated left pyelocalyceal system with grossly dilated ureter till lower end

year after surgery, the GFR was 88 and 28 ml/min/1.73 m^2 and the differential function was 76 and 24% in right and left kidney respectively. Intravenous-urogram and cystogram revealed bilaterally functioning kidneys with adequate bladder capacity.

DISCUSSION

Nearly 80% of the ectopic ureters are associated with a duplicated system. While single system ectopias are less common (10–20%),[1] BSSEUs are even rarer.[2] Attainment of continence is the prime issue in the management of BSSEU. Bilateral unduplicated ectopic ureters and unilateral single ectopic ureter draining a solitary kidney causes unusual problems in management due to the small bladder and nonfunctional bladder neck sphincter.[1] The controversy remained whether to perform only ureteral reimplantation or ureteral reimplantation with augmentation and bladder neck reconstruction or bladder neck closure with continent urinary diversion. In a study by Kesavan *et al*.,[3] the bladder neck and trigone were maldeveloped in 75% of bilateral and 54% of unilateral ectopic ureter. Heuser *et al*.[4] reported that only ureteral reimplantation may not solve the problem of incontinence due to insufficient development of the trigone and bladder neck. Hence, subsequent surgical procedures have to be kept in mind while deciding the surgery in children with bilateral single system ectopic ureter. Various procedures has been described in literature, including Young–Dees–Leadbetter, Kropp, artificial sphincter and pubo vaginal sling to increase the bladder neck resistance. Jayanthi *et al*.[5] proposed that total day and night time continence was only achieved by bladder neck closure, appendico-vesicostomy and augmentation.

However, Podesta[6] reported that a bladder with BSSEU is not necessarily useless and that a majority of patients can achieve normal bladder function and capacity along with satisfactory continence. William *et al*.[7] found that there was a spontaneous increase in bladder capacity with time and that intestinal augmentation may not be necessary in all patients. We performed bilateral ureteral reimplantation and the patient was fully continent during follow-up. Thus, the major reconstructive surgery can be avoided in some patients with BSSEU by performing ureteral reimplantation alone. Rarely, the bladder may fail to develop adequate storage capacity, which may interfere with reconstructive efforts and continence. Augmentation is thence necessary by either colo-cystoplasty or caeco-cystoplasty to enlarge bladder capacity.

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Source of Support: Nil, **Conflict of Interest:** None declared.