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Bilateral single system ectopic ureters with bilateral urethral insertion and horseshoes kidneys in a female child

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ABSTRACT

A 14-months old female child was diagnosed with bilateral single system ectopic ureters opening into the urethra, with small bladder capacity, horseshoes kidneys, and bilateral hydronephrosis, presenting recurrent febrile UTI accompanied by continuous incontinence and elevated renal function. Early bilateral re-implantation of the ureters (modified Lich-Gregoir) was done in one setting, resulting jn no recurring febrile UTIs and continuous wetting, improving renal function parameter, competent bladder neck, and 10 folds increased in bladder capacity after 1-year follow up. We showed that earlier treatment enables patient to preserve both renal and bladder function without involving complex reconstructive surgery.

1. Introduction

Bilateral single-system ectopic ureter is a rare entity, posing a threat to renal function due to its association with ureteral reflux and/or obstruction. We present a case of a female child aged 14 months with bilateral single-system ectopic ureters opening into the urethra, with small bladder capacity, horseshoes kidneys, and bilateral hydronephrosis.

2. Case report

A 14-months female was consulted by pediatric department due to recurring fever, accompanied by cases of vomiting resulting in frequent dehydration. Patient had history of frequent hospital admission due to febrile urinary tract infection (UTI). Furthermore, reported cases of continuous incontinence were present even after insertion of catheter. Patient is an only child, with premature birth (36 weeks) per-vaginal labour by a midwife. Birthweight was 1900 g, birth-length was 48 cm, with normal APGAR score.

Renal function test was elevated with blood urea of 78.1 mg/dL and serum creatinine of 1.5 mg/dL. Urine analysis showed sterile pyuria with 6–10 RBC/HPF. Contrast Urography (CT) showed bilateral grade IV hydronephrosis and hydroureters. CT also reported unison of left and right kidneys (medial-inferior poles) at the level of L3-S1, suggesting horseshoe kidneys. Diethylenetriamine Pentaacetic Acid (DTPA) renal scintigraphy showed decreased in perfusion and intrarenal activities were shown to be greatly diminished. Right kidney showed vivid activity, while left kidney showed faint increase of activity (total GFR 41.18 ml/mnt).

Upon voiding cystourethrogram(VCUG) (Fig. 1), contrast filled the bladder with capacity of 10 ml. Reflux was reported in both ureters (grade V for right and II for left ureter). Patient was further examined for urethrocystoscopy and bilateral Retrograde pyelography(RPG) (Fig. 2). Cystoscopy showed normal bladder mucosa, no trabeculation nor diverticula, however, no ureteral orifices were identified within the bladder. As the sheath was pulled back about 5 mm from the bladder

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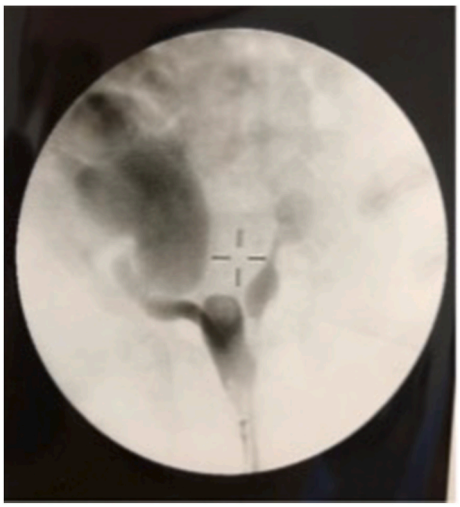

Fig. 1. VCUG showing 10 ml of contrast administration; functional capacity was 10 ml, reflux was reported on both ureters (grade V for the right dan grade II for the left kidney).

neck, a structure suspected as an ectopic left ureteral orifice on the left part of the urethra was identified. About 10 mm distal of the first orifice another structure was identified on the right side of the urethra, suspected as the right ureteral orifice. RPG showed megaureter in both ureters and contrast did not reach the Pelviocalyceal system.

Ureteral exploration was done, identifying both ureters at the anterior part of the urethra near the bladder neck. Bilateral ureteral reimplantation was done in one setting using modified dismembering Lich-Gregoir technique, with tunnelling (5:1 submucosal tunnel to ureteral diameter) done at the posterior lateral aspect of the bladder wall, with insertion of Double J stents. Patient tolerated the procedure with no immediate/long-term complications and was discharged four days after removing the Foley catheter. Ureteric stents were removed 12 weeks postoperatively. The patient had no recurrence of UTI ever since. Ultrasound showed residual bilateral hydronephrosis, with unremarkable residual urine post voiding. Renal function test were improved with blood urea of 79.2 mg/dl and creatinine of 0.7 mg/dl,.

No incontinence was reported upon follow-up. VCUG examination done 12 months post-surgery (Fig. 3) showed competent bladder neck, with a capacity of 100 ml (within the normal range limit by age). There was only a grade I reflux on the left ureter in 80 ml of contract.

3. Discussion

Ectopic ureter occurs only 1:2000 in newborns, about 20% of them present as a single system, and rarely manifest bilaterally. Urinary incontinence and recurrent UTIs are the commonly reported main complaints. The hardest challenge for these cases is how to preserve bladder size and function development as the outcome of the operation whilst preserving renal function.[1] We decided to perform a bilateral ureteral re-implantation using modified Lich-Gregoir technique. We found that surgical treatment given as early as 14 months old would benefit the patient in terms of preserving renal function. Immediate and long-term follow-up resulted in improvement, and no recurring febrile UTIs were reported.

Due to its pathogenesis, the treatment of choice for bilateral single-system ectopic ureters remains a controversy. As reported in previous studies, it is suggested that the abnormal uretero-bladder connection in patients with bilateral single-system ectopic ureters is associated with the frequent occurrences of hypoplastic bladder and poorly developed trigone and bladder neck.[2–5] Continence becomes one main issue, whether the surgery outcome may or may not result in adequate voluntary control of micturition for the patient; should the surgeon perform only ureteral reimplantation, ureteral reimplantation with augmentation and bladder neck reconstruction, or bladder neck closure with continent urinary diversion?[2]

Ectopic ureter is closely related to maldevelopment of the ureterotrigonal system. The bladder and ureters are supported by contractable muscular membrane forming an anti-reflux mechanism. When ureter orifice does not insert within the trigone of the bladder, proper muscle fibers may seize to develop, therefore resulting in functional abnormality. The anatomical anomaly may result in retention/obstruction while the anti-reflux failure may develop as a vesicoureteral reflux.[3] The CT-scan for this patient showed that both kidneys were developed (albeit with horseshoes kidneys), and dismembering Lich-gregoir approach may correct the anatomical anomaly and provide an anti-reflux system using the tunnelling technique. Furthermore, by performing the surgery early, we aimed to preserve the bladder function by letting the bladder develop through stimulation by physiological micturition. This study resulted in an increased of bladder capacity (10–100 ml), achieving normal capacity by age (90–120 ml). Incontinence was also resolved, and competent bladder was achieved. As this is the first reported case in literature, the result may differ in other cases depending on the preexisting abnormalities of the bladder. Bladder cycling in future practice may provide benefits in examining the bladder functional ability to void, before deciding the approach or technique for correction.

Not many publications have explained explain the co-occurrences of horseshoes kidneys and ectopic ureters. However, five known cases of co-existing entities have been published, all treated with bilateral ureteral reimplantation. All five have reported good outcomes in preserving renal functions.[3–5]

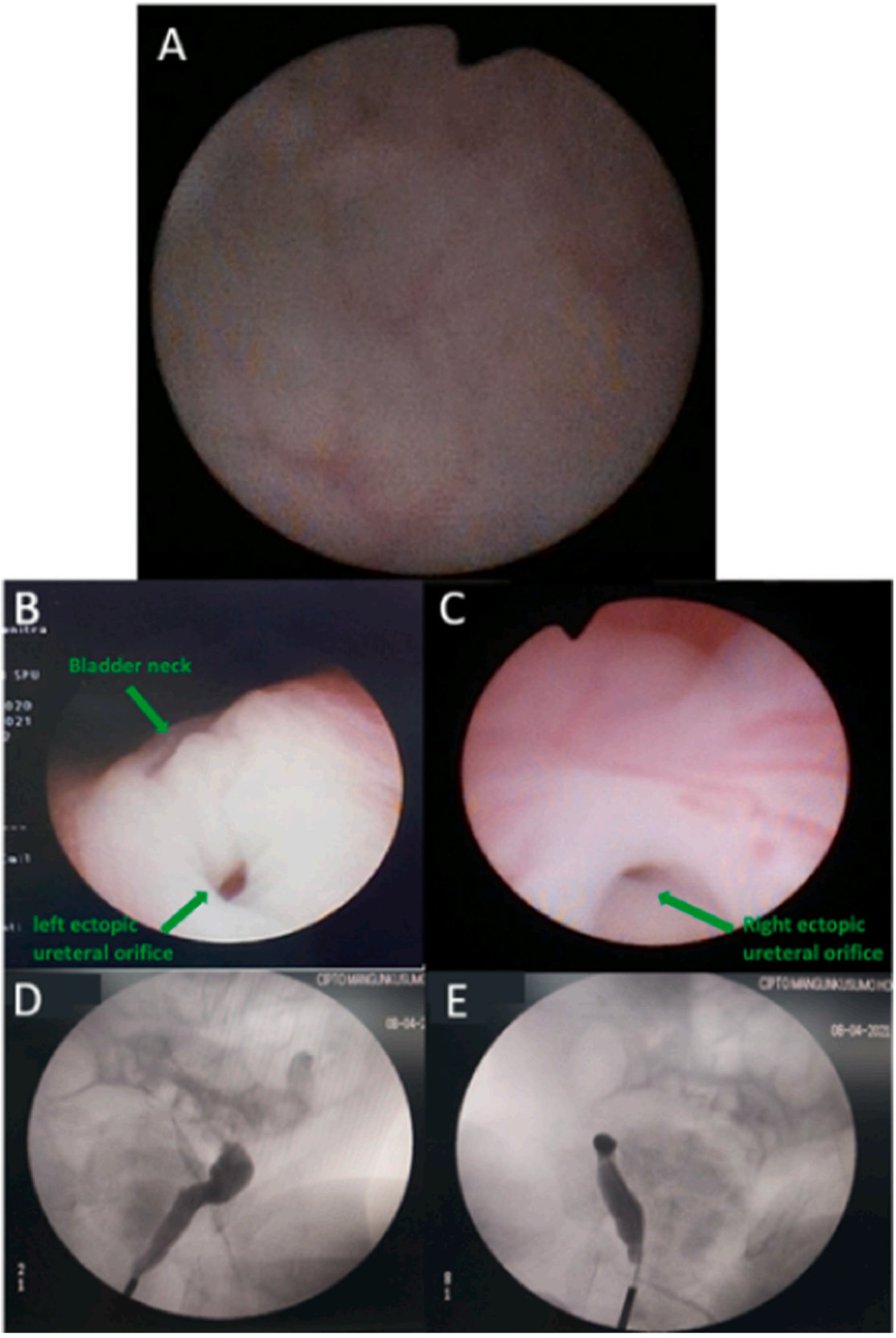

Fig. 2. A) Bladder of the 14 months old female patient, no ureteral orifice was found. B) A structure suspected as the left ectopic ureteral orifice; 5 mm distal from the bladder neck. C) A structure suspected as the right ectopic ureteral orifice; 10 mm distal from the first orifice. D) left RPG using a ureteral catheter, a megaureter was found and contrast did not reach the Pelviocalyceal system. E) Right RPG was done using ureteral catheter, a megaureter was found and contrast did not reach the Pelviocalyceal system.

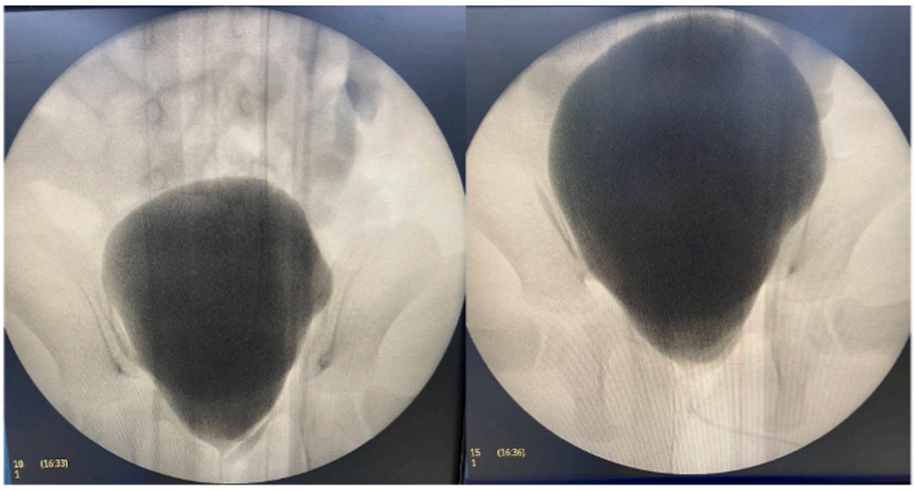

Fig. 3. VCUG taken 12 months after bilateral re-implantation of the ureter (*Lich-Gregoir* technique) showing 100 ml of contrast administration; bladder neck was competent, functional capacity was 100 ml, reflux was only reported on 80 ml contrast on left ureter as gr I reflux.

4. Conclusion

We have performed bilateral ureteral reimplantation in one setting at an early age of 14 months old resulting in improvement of renal function, bladder capacity, and continence upon follow-up. The result suggests that earlier treatment may attain goals of treatment without involving complex reconstructive surgery.

Declaration of competing interest

There is no financial conflict of interest.

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Abbreviations

CT: Contrast Urography
DTPA: Diethylenetriamine Pentaacetic Acid
VCUG: voiding cystourethrogram
RPG: Retrograde pyelography