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Case Report

Bilateral ureteral obstruction after open ureteral reimplantation in a 3-year-old patient with Williams Beuren syndrome ☆,☆☆

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ABSTRACT

Vesicoureteral reflux (VUR) is a common urological problem in the pediatric population and can be corrected by ureteral reimplantation in severe or persistent cases. This procedure is generally well tolerated, although complications, including ureteral obstruction, may occur in the postoperative period. We present a rare case of a 3-year-old with Williams Beuren syndrome who underwent bilateral ureteral reimplantation for VUR and subsequently developed bilateral ureteral obstruction with acute renal failure requiring nephrostomy tube placement within 48 hours of surgery.

Introduction

Vesicoureteral reflux (VUR) is frequently encountered in pediatric patients and may predispose to frequent urinary tract infections and subsequent renal scarring [1–3]. Definitive correction is surgical by ureteral reimplantation, either open, endoscopic, or robotic assisted laparoscopy [4,5]. Williams-Beuren syndrome (WS) is a rare genetic disease secondary to a chromosome 7 deletion resulting in distinctive cardiac, neurodevelopmental, connective tissue, and personality characteristics. Genitourinary abnormalities are also associated with WS, including bladder diverticula and detrusor overactivity in over 50% of cases [6]. While about 25% of patients with WS have a history of urinary tract infection (UTI) [7], the incidence of VUR is less well described, occurring in only 5 % of WS patients on a recent review [8]. We present a rare case of a 3-year-old female with WS and VUR who underwent bilateral ureteral reimplantation and subsequently developed postoperative ureteral obstruction with resulting obstructive uropathy and acute renal failure requiring nephrostomy tube placement within 48 hours of surgery.

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Case report

A 3-year-old female with a history of WS presented for elective open bilateral ureteral reimplantation due to a history of persistent bilateral VUR and breakthrough febrile UTI. She had a history of moderate pulmonary stenosis (peak gradient 45 mmHg), bilateral developmental hip dysplasia s/p surgical correction without any history of anesthesia complications, developmental delay, and hyperopia with astigmatism. Her bowel habits are normal and she has a history of increased urinary frequency and nocturnal enuresis. At 9 months of age, she was diagnosed with a febrile *E. Coli* urinary tract infection and underwent a renal ultrasound which noted left sided pelviectasis. A subsequent voiding cystourethrogram (VCUG) demonstrated grade IV right VUR and a normal appearing bladder without diverticula. She was managed with prophylactic trimethoprim-sulfamethoxazole which was changed to nitrofurantoin due to noted resistance on a urine culture obtained during a preoperative evaluation prior to open reduction of her left hip. A repeat VCUG demonstrated right grade I and left grade III VUR. Due to persistent VUR and a subsequent breakthrough febrile UTI, surgical correction was elected, and bilateral repair was planned to prevent development of high-grade VUR in the unrepaired ureter.

Patient presented for elective surgical correction. Preoperative complete blood count (CBC) and kidney-liver function tests were within reference ranges. She underwent bilateral open reimplantation via the intravesical cross-trigonal approach without any immediately apparent complications. She was observed overnight postoperatively with a urinary catheter in place for bladder decompression and was treated with intravenous fluids at her maintenance rate for weight. She had 600 cc total of urine output on postoperative day 1, which dropped to 100 cc total on postoperative day 2. A renal ultrasound was obtained on POD #2, which showed bilateral moderate pelvicalyceal dilation and a small amount of complex free fluid in the pelvis (Fig. 1). A cystogram was then performed which was negative for urine leak from the bladder. Later on POD #2, the patient had cessation of urine output into her foley catheter, and she became hypertensive, edematous, and developed vomiting. The foley catheter was flushed and then changed without noted urine output. Emergent workup was performed which revealed a creatinine of 2.83 (ref: 0.25-0.56 mg/dL), a BUN of 30 (ref: 5-25 mg/dL), a potassium of 7.5 (ref: 3.6-5.3 mmol/L), chloride of 109 (ref 95-108 mmol/L), a bicarb of 15 (ref: 20-28 mmol/L), and a urine pH of 5.0 (ref: 5.0-9.0). Albuterol was given, followed by a dextrose bolus and insulin dose for hyperkalemia, and she was transferred to the ICU. Due to presence of type 4 renal tubular acidosis, hypertension, and sudden oligo-anuria, there was concern for obstruction. A bilateral percutaneous antegrade pyelogram was performed and noted distal bilateral ureteral obstruction without flow of contrast into the bladder and no filling of the distal ureter (Fig. 2). Patient underwent emergent bilateral nephrostomy tube placement with 1.5 L of urine output immediately upon placement. Eight hours later, her BUN, creatinine, and potassium had normalized, and she had normal urine output via the nephrostomy tubes. A repeat bilateral nephrostagram was performed on POD #6 and was without obstruction and noted contrast filling the bladder. She was discharged home with nephrostomy tubes in place, which were removed 2 weeks later without complication. A follow-up ultrasound after nephrostomy tube removal showed no abnormal urinary tract dilation and she was able to void spontaneously with normal urine output.

Discussion

Hydronephrosis is a relatively common finding postoperatively after ureteral reimplantation [9]. Postoperative ureteral obstruction is a noted but rare complication after ureteral reimplantation and has been reported more frequently after bilateral extravesical ureteral reimplantation. With endoscopic approach, a retrospective review noted postoperative ureteral obstruction was less than 1% of treated patients [10]. Postoperative ureteral obstruction is also rare post open intravesical ureteral reimplantation for VUR, occurring in

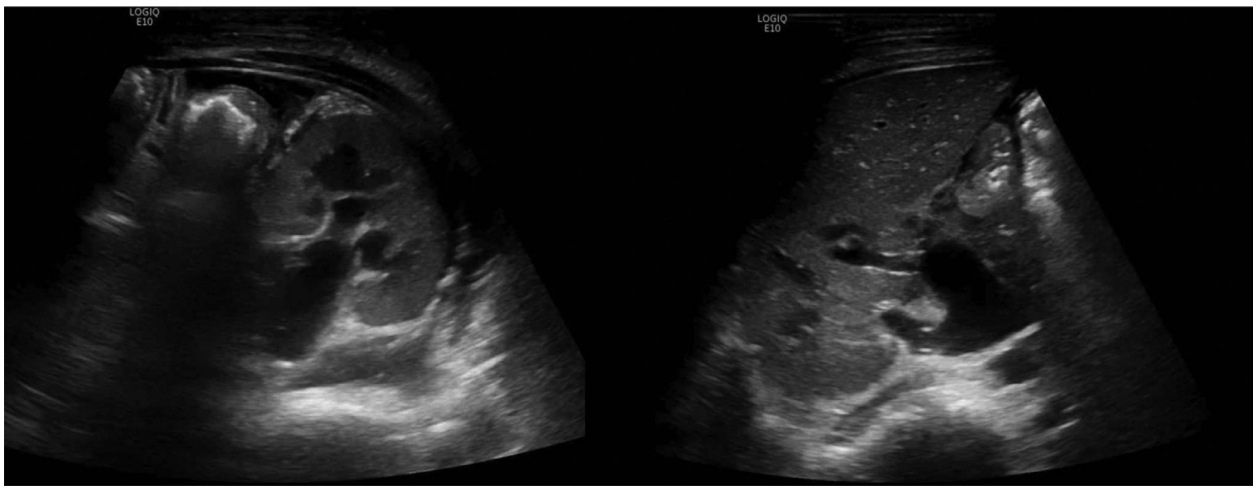

Fig. 1 – Renal ultrasound on POD #2 demonstrating bilateral pelvicalyceal dilatation.

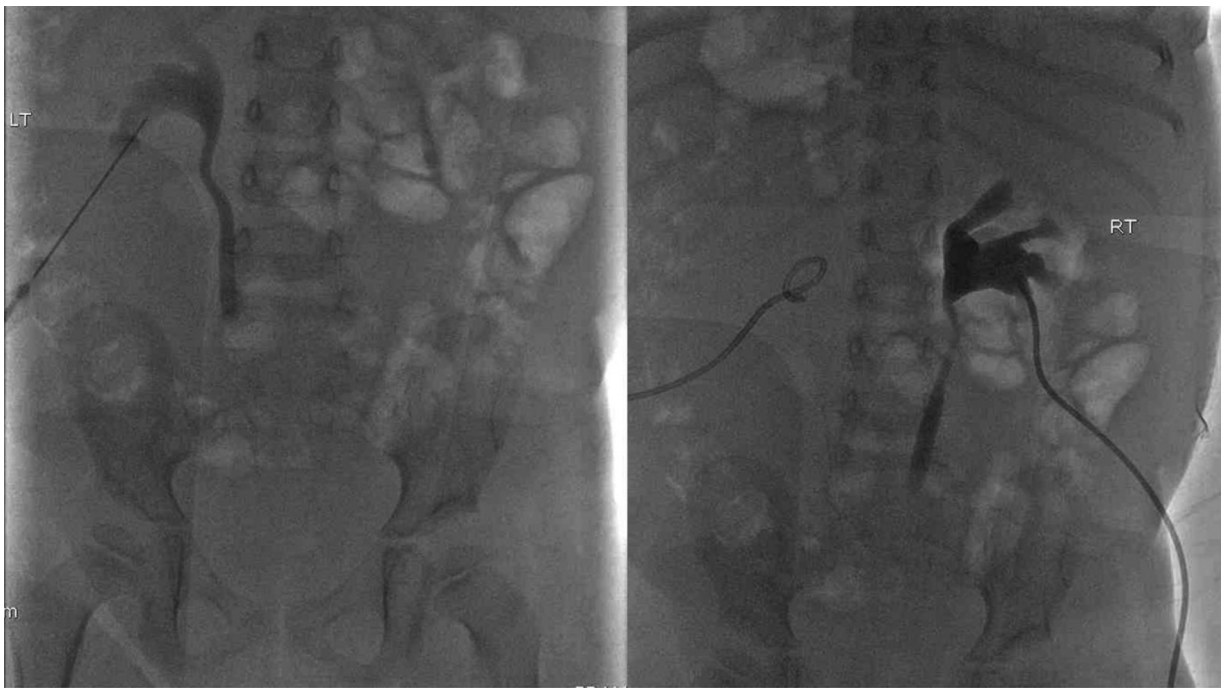

Fig. 2 – Percutaneous antegrade pyelogram showing concern for distal bilateral ureteral obstruction without flow of contrast into the bladder.

approximately 2.8% of patients [11]. Mechanisms for obstruction include edema, hematoma, and intraoperative kinking of the ureter during reimplantation [12], although it is difficult to determine the presence of obstruction based on hydronephrosis alone, which may be present both pre and post operatively due to high grade VUR [13].

We present a case of a 3-year-old patient with WS who developed post-operative bilateral ureteral obstruction with subsequent acute renal failure after undergoing bilateral open intravesical ureteral reimplantation. There was no noted kinking, hematoma, nor edema noted intraoperatively that would explain her obstructive findings postoperatively. Decreased peristalsis in the ureters following operative manipulation has been reported, although this is almost always temporary and self-limiting [9]. WS is a genetic condition involving deletions on chromosome 7 which typically involve the elastin gene, which is responsible for the characteristic cardiac and connective tissue disorders noted in this syndrome. Decreased elastin protein production results in decreased numbers of elastin fibers and thus decreased tissue recoil, including in the ureter. Ureter recoil is facilitated by elastin fibers within the urothelium, lamina propria, and smooth muscle [14].

In our case, our hypothesis was that the patient developed postoperative ureteral obstruction secondary to her underlying WS syndrome related elastin deficiency, exacerbated by intraoperative ureteral manipulation and subsequent decreased peristalsis. This was suspected after the patient produced over 1 L of urine output via nephrostomy tubes when the obstruction was relieved. It was likely that she did not experience expected ureteral recoil after manipulation due to her elastin deficiency secondary to WS. This finding resolved with time and she was able to produce normal urine output within 2 weeks after her surgery.

To our knowledge, there have been no other described cases of postoperative ureteral obstruction resulting in acute renal failure in patients with WS and VUR undergoing ureteral reimplantation. Clinicians should remain aware that patients with WS may have underlying bladder diverticula and develop VUR, and that they are high risk for postoperative complications due to elastin deficiency resulting in decreased ureteral recoil and subsequent obstruction of urine flow. We recommend considering ureteral stent placement in WS patients who undergo bilateral ureteral reimplantation.

Conclusion

Postoperative ureteral obstruction after open intravesical ureteral reimplantation is exceedingly rare. Obstruction may be secondary to intraoperative complications, although certain patients with underlying genetic abnormalities may be at higher risk, such as those with underlying WS and elastin deficiency. Clinicians should remain aware of this when pursuing surgical correction of VUR in patients with WS and can consider ureteral stent placement in WS patients undergoing bilateral ureteral reimplantation.

Ethical approval

None to declare.

Patient consent

Written, informed consent was obtained from the patient's guardian for publication of this case.

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