Bilateral Ureteral Tapering and Secondary Ureteroneocystostomy for Late Stenosis in a Patient with Bladder Exstrophy

Emanuela Altobelli, Alfredo M. Bove, Federico Sergi, Maurizio Buscarini
Campus Biomedico, Urology Roma, Rome, Italy

Key Words
Bladder exstrophy • Hydronephrosis • Surgical technique

Abstract
Objective: To report a case of severe hydronephrosis 20 years after bladder exstrophy (BE) repair, managed by bilateral ureteral tapering and secondary ureteroneocystostomy.

Case Presentation: A 21-year-old woman with a history of BE and ureteral reimplantation, presented with hematuria-dysuria syndrome and recurrent febrile urinary tract infections. After counselling, she elected to undergo bilateral ureteral tapering and second ureteroneocystostomy. Conclusion: Hydronephrosis secondary to ureteralvesical stricture in BE patients can be successfully managed with ureteral reimplantation associated to ureteral tapering even after a prior reimplantation.

Introduction
Hydronephrosis following ureteral reimplantation (UR) is a common occurrence. In most cases, postoperative hydronephrosis is transient and not clinically significant, with a high incidence of complete resolution during the first 2 years. Ureteral obstruction occurs rarely following reimplantation. We report a case of severe hydronephrosis associated to febrile urinary tract infections (UTIs) 20 years after bladder exstrophy (BE) repair, managed successfully by bilateral ureteral tapering and secondary ureteroneocystostomy.

Case Report
A 21-year-old woman with a history of BE was referred to our department for hematuria-dysuria syndrome and recurrent febrile UTIs.

Her surgical history started on the fourth day of life. During the first procedure she underwent bladder closure without ostectomy in a staged reconstructive approach. At 6 years for low bladder capacity, a gastrocystoplasty was performed. At the age of 10, she underwent a bilateral UR secondary Cohen for hydronephrosis secondary to vesicoureteral reflux (VUR). The last procedure was performed at 19 years with the removal of umbilical endometriosis.

On physical examination she presented with lower midline and Pfannenstiel scars of previous procedures, pubis diastasis, bifid clitoris, stenotic and anteriorly displaced vagina and short perineum with the anus directly behind the urogenital diaphragm. No alterations on blood tests, with normal renal function (creatinine 0.7 mg/dl). Urine culture showed > 100,000 CFU of E. Coli and a specific antibiotic therapy was prescribed for 7 days. To treat the hematuria-dysuria syndrome a proton pump inhibitor therapy was started with success.

A severe bilateral hydronephrosis, greater on the left, associated to a compression of renal parenchyma, was highlighted at the ultrasounds and confirmed at the CT scan, with a diameter of 23 and 40 mm on right and left ureter, respectively (fig. 1). Right pelvis measured 33 mm and left pelvis 43 mm. A high post-void residual volume was evident.
Patient refused self clean intermittent catheterisation and urodynamic evaluation because of childhood catheter trauma. So to investigate the symptoms we decided to perform under general anesthesia a cystourethroscopy that showed a long and twisting urethra with a bladder full of mucus without alterations of urothelial and gastric mucosa. Cross trigonal ureteral meatus were stenotic bilaterally and retrograde stenting was not possible. During the same procedure, a retrograde cystourethrogram was executed without evidence of VUR. One month later she elected to undergo open surgery.

Surgical Technique
Because of previous surgical procedures, a laparoscopic approach was not indicated. An 18Ch transurethral Foley catheter was inserted. Patient was placed in supine position with 15° Trendelenburg tilt. After sterile draping a lower midline incision was performed. After peritoneal incision the exposure was obtained with a self-retaining retractor. The bulging and dilated ureters were identified extravesically and gently placed on traction with a vessel loop. Ureters were mobilized to the ureterovesical junction and a stiff and severe anastomotic stricture determining obstruction was verified bilaterally. Ureters were divided and hiatus oversewn. A 4-0 polyglactin stay suture was placed on the distal end of each ureter. Both ureters were fully mobilized to the extent of the exposure at the peritoneal reflection. Once the ureter was mobilized and decompressed, decision was made to proceed with extensive ureteral tailoring. To achieve a tunnel length to diameter ratio of at least 5:1, a 10Ch catheter was placed in the lumen of each ureter and these were tapered using Hendren clamps. Obviously only the intramural part of the ureter was tapered. The bladder was opened via a midline cystostomy and the ureter was reimplemented with an anti-reflux intravesical anastomosis according to Glenn-Anderson technique on left ureter. Right ureter was reimplemented with Lich-Gregoire extravesical approach. A mono-J ureteral catheter was retrograde positioned in both ureters. Oversewing detrusorrhaphy was performed for bladder closure. A perivesical Jackson-Pratt drain was left indwelling.

Results
Postoperatively, orals were allowed on day 1. Drain and Foley catheter were removed on day 3 and 6, respectively. Cystoscopy and cystourethrogram were performed as an outpatient 4 weeks postoperatively to remove ureteral stents and transurethral catheter. Urine culture 1 month after surgery was positive for *E. Coli* growth and antibiotic therapy was prescribed based on the antibiogram.
A postoperative abdominal ultrasound performed 3 months after surgery showed left hydronephrosis, confirmed at the MRI, showing left hydroureteronephrosis (33 mm of diameter) with anastomotic stenosis and a regular right ureter (fig. 2). Three-month urine culture was negative for bacterial growth. Patient was asymptomatic and her quality of life was improved.

**Discussion**

After initial closure, all extrophy patients have VUR. In BE, the ureters dip deeply into the pelvis before entering the bladder in a nearly cephalad direction, emerging through the bladder muscle with a minimal submucosal tunnel [1]. This reflux is usually managed with antimicrobial prophylaxis, surveillance, and ureteral reimplantation concurrent with bladder neck plasty [2–5]. Our patient developed recurrent UTIs and worsening hydronephrosis, and at the age of 10 underwent first bilateral UR according to Cohen’s technique. During the procedure bladder neck plasty was not performed.

However UR is effective in reducing postoperative pyelonephritis [6], and UTI is the most common complication. There is surprisingly little literature describing the long-term clinical outcomes after UR. The International Reflux Study in Children prospectively compared the effects of successful surgery and effective medical management on the incidence of UTIs. During a follow-up of 5 years that study reported an incidence of 38% of infections of the lower urinary tract among conservatively treated patients, compared with 39% among those treated by surgery. The incidence of pyelonephritis was 21 and 10%, respectively, and was statistically significantly different [7]. In another long-term, retrospective study of patients 10 years after UR, Beetz et al. [8] found that, among the 83.5% of patients they could contact, 17% had experienced postoperative febrile UTI. In contrast to these high rates of postoperative pyelonephritis, Whittam et al. [9] reported in a recent series that of 395 patients who underwent UR febrile UTI was diagnosed in just 4.6%, although the follow-up time was relatively short at a mean of 15 months. Accordingly, Cooper et al. [10] reported an incidence of 37.6% of UTIs among 96 women who had UR in childhood and who were followed for 16–25 years afterward. As emphasized by Mor et al. [11] there is the need to establish a protocol for the long-term follow-up of patients who have had UR during childhood. Even patients who were managed successfully by surgery are prone to recurrent UTIs, progressive renal scarring, hypertension and complications during pregnancy. Our patient present only recurrent febrile UTIs associated to severe hydronephrosis. The primary complication that is assessed by postoperative imaging is obstruction, and presents clinically in addition to ultrasonographically [12–14]. Ellsworth et al. [15] reported on 3 cases of ureteral obstruction during the first week after surgery. All of these patients were symptomatic and were treated with the placement of a double-J stent. Androulakakis et al. [16] also reported 3 incidences of obstruction, all of which presented symptomatically, and only one of which required reoperation. Additionally, temporary obstructive situations may occur following surgery, including ureteral edema, intramural hematomas applying pressure on the ureter, temporary lack of peristalsis in the operated ureter, and kinking of the ureter at the hiatus as the bladder fills [17]. These obstructions can be partial and self limiting following surgery, are varied between each patient, and contribute to the length of time it takes for postoperative hydronephrosis to resolve. They do not represent clinically important obstruction, however [18–20]. All this study evaluates only a short-term follow-up. There are no studies about worsening hydronephrosis 20 years after UR.

**Conclusions**

Even if there are some studies about the postoperative UTIs and hydronephrosis, there are no data about reinvention 20 years after primary UR. Our case demonstrates the success of UR associated to ureteral tapering even after a prior reimplantation in BE patient.
References


