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The urologist and child hydronephrosis caused by ureteral anomalies

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Abstract

Congenital hydronephrosis caused by ureteral anomalies, like ureteral duplicity, megaureter, ureteral ectopy and ureterocele, must be differentiated from ureteropelvic junction obstruction (UJO) hydronephrosis and from the hydronephrosis caused by vesicoureteral reflux. These represent a differentiated branch of congenital abnormalities in children even if not so common, but this fact should not be disconsidered. Over a five years period, from 111 operated children in our Clinic, we performed 13 interventions for congenital hydronephrosis, 11 (84.61%) being caused by ureteral abnormalities. Here, there were described particular cases, with diagnosis steps and treatment decisions. Ureteral ectopy can be manifested by loss of urine drops in cases where ureteral holes are located in the vagina, septum or urethra, inferior to the sphincter mechanism. Incontinence in boys never occurs because the ectopic ureter never opens under the sphincter mechanism. If the ureter opens in the genital tract, patients may clinically present with the epididymitis symptom. From autopsy statistics in the US, the incidence of ureteral duplex is estimated to be less than 1%. When the duplex is associated with urinary infection, the incidence of ureteral duplex increases up to 8%.

Keywords: hydronephrosis, ureteral, abnormalities, congenital, megaureter.

Introduction

Hydronephrosis is the one of the most common congenital abnormalities of the urinary tract. The frequency of congenital hydronephrosis diagnosed through prenatal ultrasound is about 0.6–5.4% of the newborns [1–3].

The left kidney is more frequently affected than the right side and is more common in males [4]. Congenital hydronephrosis caused by ureteral anomalies, like ureteral duplicity, megaureter, ureteral ectopy and ureterocele, must be differentiated from ureteropelvic junction obstruction (UJO) hydronephrosis and from the hydronephrosis due to vesicoureteral reflux (VUR). These entities represent a differentiated branch of congenital abnormalities in children even if not so common but that should not be disconsidered, because they can cause many complications of kidney functions [5–8].

Ureteral ectopy can be manifested by loss of urine drops in cases where ureteral holes are located in the vagina, septum or urethra, inferior to the sphincter mechanism [9]. Incontinence in boys never occurs because the ectopic ureter never opens under the sphincter mechanism. If the ureter opens in the genital tract, patients may clinically present with the epididymitis symptom.

Aim

The aim of the study was to evaluate the causes and to treat congenital hydronephrosis due to ureteral anomalies like ureteral duplicity, megaureter, ureteral ectopy and ureterocele that must be differentiated from UJO hydronephrosis and from the hydronephrosis due to VUR.

Patients, Materials and Methods

Between 2011 and 2015, there were operated 111 children with a diverse urological pathology, with an average age of 11.57 years old. During these five years, 13 patients with congenital hydronephrosis were operated, of whom 11 patients with ureteral abnormalities, one patient with posterior valve and reflux, and another patient with congenital phimosis and VUR. The dominant symptomatology was a persistent or recurrent urinary tract infection, the most common etiology being *Escherichia coli*, but also permanent urinary incontinence.

The ultrasound examination was performed in all patients. Echography is the simplest and least harmful exploration that can identify hydronephrosis both prenatally and immediately postnatally. Nearly all of these patients also have a ureteral dilation that suggests a ureteral abnormality from the beginning.

Urography has also been performed, which provides both anatomical data and renal function data, although it has been thought to be overstretched lately. No imaging complements, such as magnetic resonance or computed tomography (CT) were required. The investigation that can differentiate between dilation and obstruction, and especially the functional difference between the two kidneys, is the diuretic renoscintigram that we have called for selectively. A functional difference of less than 40% recommends corrective intervention and the function less than 10% recommends nephrectomy.

The exploration we performed in all patients with

hydronephrosis was the retrograde and mictional cystography that gives us data about the posterior urethra and the confirmation or refutation of VUR of its degree as well as data on bladder and bladder anatomy (information about the tract, size and dilation of the ureter, important data for the surgical strategy).

In Table 1, we present the main malformations of the urinary system treated by us:

Table 1 – ***Urological pathology and surgical treatment of 13 patients with congenital hydronephrosis***

No. of patients	Congenital pathology	Intervention
1	Myelomeningocele Right megaureter Neurological bladder Bilateral ureteral reflux Right pyonephrosis	*First session*: Bladder augmentation from pelvic right megaureter; left ureter antireflux reimplantation. *Second session*: Right percutaneous nephrostomy; right nephrectomy after seven years. *Third session*: Left percutaneous nephrostomy and finally definitive cutaneous left ureterostomy.
1	Iliac right renal ectopy with right ureter in uterus	Right nephrectomy and left ureter antireflux reimplantation.
1	Right obstructive megaureter	Left ureteral modeling and Lich–Gregoir technique reimplantation on the left.
1	Bilateral duplex kidney, bilateral VUR	Bilateral antireflux implantation on left and right unilateral ureter reimplantation.
1	Congenital phimosis, bilateral VUR	Prepuce dorsal incision, adesiolysis.
1	Third type urethral posterior valve	Endoscopic valve incision.
7	Renal duplicity, ureteral ectopy	Superior left renal pole resection and partial ureterectomy.

VUR: Vesicoureteral reflux.

Here are some of the particular aspects of the cases we dealt with:

Case No. 1

We present a 6-month-old patient with complete pyeloureteral duplicity on the right, with reflux in the lower renal pelvis ureter and incomplete left ureteral duplication (ureteral juxtaposition), with reflux in the lower renal pelvis ureter and obstruction on the superior ureter (Figure 1).

By Pfannenstiel incision, in general anesthesia, there was performed left-hand re-implantation in the rifle tube (common sheath) and the re-implantation of the right-hand antireflux of the lower renal pelvis ureter.

The reimplantation complied with the ratio of 5/1 between the length of the submucosal tunnel and the diameter of the ureter, imitating somewhat the Waldeyer sheath and the tract of the intramural ureter. Ureteral reimplantation was performed under the protection of two plastic tubes of 6 Charrière (Ch), which were removed transvesicocutaneous after 12 days. Submucosal vesical suture was protected by a Foley 8 Ch catheter, which has been maintained for 14 days. At eight months after surgery, there is also bilateral renal dilation grade 1/2, and after two years – the kidney is normal (Figure 2), has a good overall condition, has smooth micturition and good residual flow.

Case No. 2

A 17-year-old female patient treated for urinary infections for years. The clinical examination diagnosed a permanent urinary incontinence but with preserved micturitions. The ultrasound showed a left-sided pyelo-ureteral duplication. At this point and taking into account permanent urinary incontinence, we suspected an ectopic ureter whose endocaptic ureteral opening was confirmed endoscopically (Figures 3 and 4). Retrograde ureteropyelography performed preoperatively reveals the duplicity of the collector system.

Case No. 3

A 3-year-old child whose imagistic and endoscopic explorations reveal an obstructive megaureter; ureteral modeling and antireflux bladder reimplantation are practiced by extravesical approach (Lich–Grégoire method) (Figure 5).

The preoperative biological balance was normal, including urinalysis that was sterile. The incision allowed a good approach to the ureter. It is very important to preserve the ureteral vascular system knowing that the ureter is vascularized by branches of the aorta, the renal artery, the gonadal artery (medial) and form the internal iliac artery (lateral). The loss of intraoperative blood was minimal; the patient did not require transfusion. Reduction of the ureteral lumen can be done by excision (Hendren) or reduction of the lumen through the envelope (Kalicinski). To avoid delayed stenosis of the ureter, lumen reduction should not be less than 10 Ch. The antireflux reimplantation mechanism respected the classical ratio of 5/1 between the submucosal tunnel of the intravesical ureter and the diameter of the lumen (Figures 6 and 7). Two-month ultrasound scanning showed a normal kidney (Figure 8).

Case No. 4

We present a 6-month-old patient with posterior valves suspected on retrograde urethrocystography (Figure 9) that was confirmed in endoscopic evaluation. After their hot cut, the bilateral reflux resolved almost completely five months after the intervention. The intervention was performed in general anesthesia under antibiotic protection. Sectioning of the valves was performed under direct visual control with the resectoscope using an endoscopic anvil.

The intervention was completed by fitting an 8 Ch catheter maintained for seven days.

In some patients, the urethra may be too small for urethrocystoscopy. When this occurs and the situation imposes it, the vesicostomy can be performed. The

vesicostomy was done in one case using just a bladder wall and should not be confused with the bladder suprapubic drainage with a plastic tube or a Foley catheter. The use of tubes for suprapubic cystostomy is plagued by numerous complications such as infection, bladder inflammation and accidental removal. By the presence of vesicostomy, the bladder maintains its micronial cycle at low pressures, eliminating urine through the stoma opening. Urologists will take up these patients with urethral valves because they require long-term management to avoid progressive alterations in bladder function, upper and lower urinary system.

Case No. 5

An 8-year-old patient with myelomeningocele and subarachnoid-peritoneal shunt with neurological bladder and urinary incontinence. Clinical examination, retrograde cystography, renal ultrasonography, urography and tomography scan reveal bilateral grade 4 reflux on the right and grade 3 on the left (Figure 10).

Surgical strategy included preparation of a flap from the dilated juxtaposed ureter, which will enlarge the bladder. Reimplantation of the remaining left ureter is done in the antireflux manner spliced on plastic 6 Ch tubes to the skin (Figures 11 and 12).

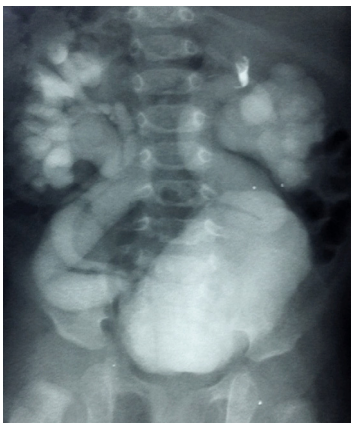

Figure 1 – ***Urography showing complete right pyeloureteral duplication and incomplete left ureteral duplication (ureteral juxtaposition); grade III–IV ureterohydronephrosis.***

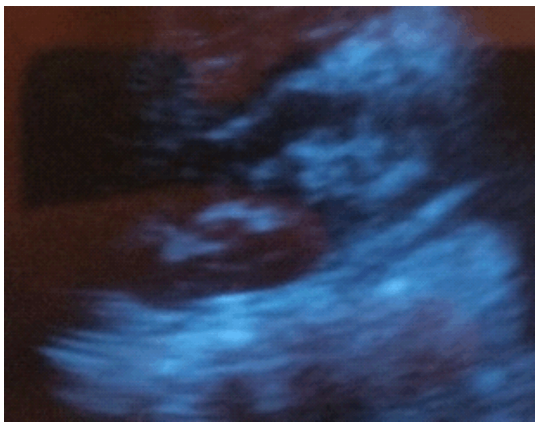

Figure 2 – ***Ultrasound aspect after two years post-surgery (the kidney is normal).***

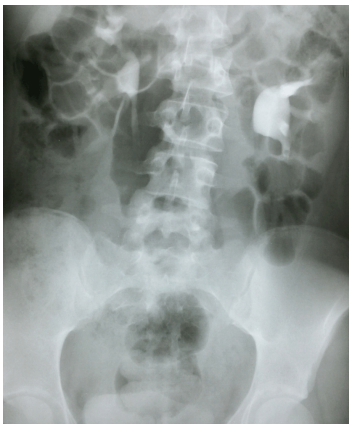

Figure 3 – ***Urography showing bilateral renal function and amputation of the superior pyelocaliceal system.***

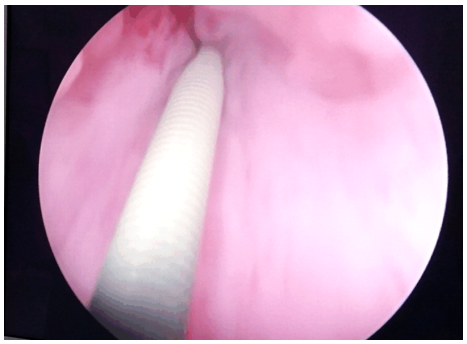

Figure 4 – ***Ureteroscopy with ectopic ureteral opening cauterized.***

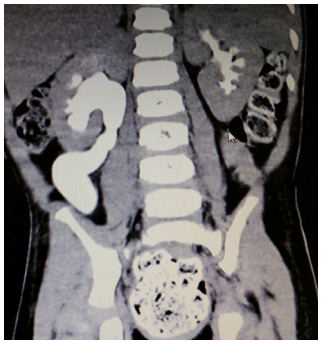

Figure 5 – ***Tomography scan aspect of obstructive megaureter.***

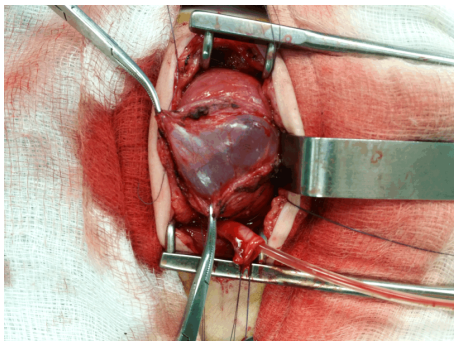

Figure 6 – ***Bladder detrusor mucosa dissection.***

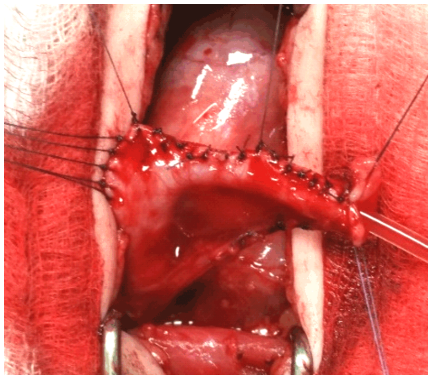

Figure 7 – ***Ureteral aspect after modeling.***

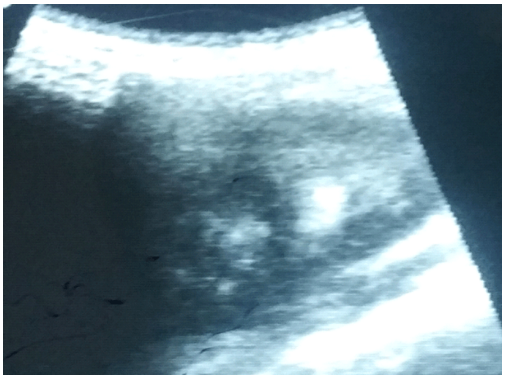

Figure 8 – ***Ultrasound aspect of the kidney two months after surgery.***

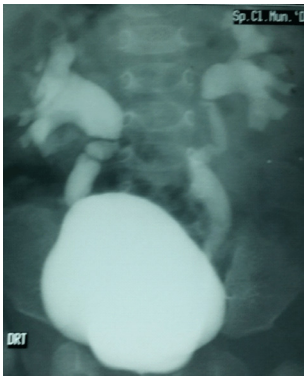

Figure 9 – ***Retrograde urethrocystography showing bilateral reflux.***

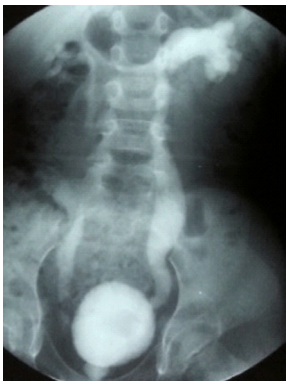

Figure 10 – ***Tomography scan aspect showing bilateral reflux.***

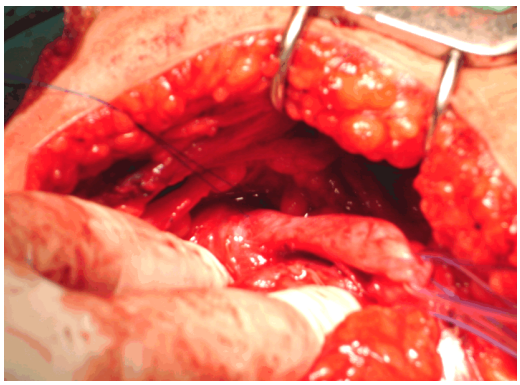

Figure 11 – ***Juxtavesical ureter used for the enlargement of the bladder.***

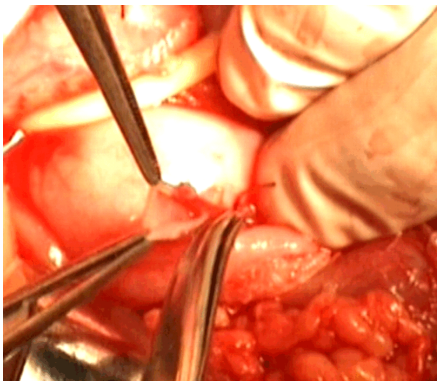

Figure 12 – ***Reimplantation of the remaining left ureter is done in the antireflux manner.***

The postoperative progression was favorable, with no notable incidence. After 14 days since surgery, the Foley bladder catheter is extracted, after which the patient resumes the micturitions with acceptable jet and with minimal residue. He returns to the Clinic after seven years (he was monitored all this time) with prolonged febrile syndrome. Imaging explorations reveals a right hydropyonephrosis.

A straight right nephrostomy is drawn under fluoroscopic control, which drains the pus after which the clinical state improves without the kidney restoring its function.

There was performed right side nephrectomy. It is surgically cured externally with minimal micturition, with minimal residual (urodynamic balance) and with good left kidney unit, with minimal hydronephrosis.

The patient comes back to the clinic for febrile syndrome and left grade 3 hydronephrosis. We mounted a nephrostomy after which the symptoms changed. At six months after the left nephrostomy, he again returns for grade 3 hydronephrosis and 200 mL postmictional residue (urodinamic tests). A definitive left cutaneous ureterostomy was decided to be performed.

Discussions

The renal duplex system involves the presence of two ureters. It is believed that renal duplication is a dominant autosomal disease [10] and has an increased prevalence in Caucasian women [11, 12]. A patient with a duplex abnormality may have bifid ureters (incomplete duplication) or two ureters that are separated into the bladder (full duplication) [13]. The upper ureter is more likely to be associated with an ectopic insertion, the affected pole being hypoplastic or dysplastic [14] (Figure 1). The most common anomaly associated with an ectopic ureter is renal hypoplasia or dysplasia in patients with a single collecting system, and hypoplasia/dysplasia of the affected pole of the kidney in patients with a duplex system [15]. In one series, 25 of 33 ectopic ureters were associated with a poorly functioning or nonfunctional renal moiety [16]. The presence of a poorly functioning renal moiety may influence the surgical decisions [17].

The duplex kidney in children is more susceptible to reflux than is the non-duplex kidney, and this leads to both ureteric and pelvicalyceal dilation, and to chronic pyelonephritis in the duplex side in those children who develop urinary tract infections. Chronic pyelonephritis was found in 22% of patients less than 15 years old, significantly more often than in adults (p<0.001), although the incidence of duplication was unchanged [18, 19].

The megaureter is defined as a ureter over 7 mm in diameter. Megaureter can be classified into four categories: obstructive megaureter, refluxing megaureter, non-obstructive and non-refluxing megaureter. Each group can be divided into two subgroups: primary megaureter; secondary megaureter. Additionally, the massively dilated ureter may be decompressed with ureterostomy, pyelostomy, or nephrostomy drainage, which often allows a substantial decrease in ureteral size and greatly reduces ureteral bulk during both tailoring and reimplantation [20].

In average cases of obstructive megaureter, surgery may not be necessary, these patients should be followed up, clinically and radiologically monitored periodically, and clinically treated with antibiotics. Sometimes, it requires urinary drainage for infections that do not respond to antibiotics. Ureteral decompression of the dilated ureter requires antireflux ureteroneocystostomy, pyelostomy, nephrostomy, ureterostomy, which allows a reduction in ureteral dimensions and a reduced need for remodeling and reimplantation.

VUR is the retrograde passage from bladder in the ureter and/or the kidneys (Figures 4–6). Reflux-induced kidney damage is actually the result of association with urinary tract infection (UTI) [21]. Leonardo da Vinci and Galen were the first to recognize the importance of the ureterovesical junction (UVJ) and the link between it and the VUR.

Reflux associated with urethral valves is secondary to high bladder pressure and is associated with renal lesions [22]. These valves described by Young are endoscopically observed as a membrane in the posterior urethra. The prevalence of VUR is significantly higher in the presence of genital abnormalities such as renal duplex (46%) [23].

The work-up of a febrile urinary tract infection is generally performed to detect VUR and its possible complications. The imaging modalities most commonly used for this purpose are renal–bladder ultrasound, voiding cystourethrogram and dimercapto-succinic acid scan. These studies each contribute valuable information, but carry individual benefits and limitations that may influence

their efficacy. Biochemical markers are not commonly used in pediatric urology to diagnose or differentiate high-risk disease, but this is the emerging frontier, which will hopefully change our approach to VUR in the future.

Conclusions

The cases presented above underline the importance of using the correct exploration techniques for final diagnose and correct surgical approach (ultrasound, urography, retrograde cystography, retrograde ureteropyelography, endoscopy, tomography scan). An important exploration that can differentiate between dilation and obstruction, and especially the functional difference between the two kidneys, is the diuretic renoscintigram that we have called for selectively. A functional difference of less than 40% recommends corrective intervention and the function less than 10% recommends nephrectomy. Our study underlines the complexity of the urological malformative pathologies.

Conflict of interests

The authors declare that they have no conflict of interests.

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