

Minimal invasive management of complete renoureteral duplication with upper pole megaureter in children

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ABSTRACT

Renal malformations represent a large part of the pathology referred to the pediatric surgeon. Most of them are diagnosed during pregnancy. We present one case of complete ureteral duplex with upper pole obstructive megaureter managed in a private hospital using minimally invasive methods for both diagnosis and treatment due to good teamwork. We do not want to demonstrate that other investigations are not leading to the same diagnose, we want to highlight an alternative to the classic approach. We hope that our experience is a starting point for further research and that in future radiation and general anesthesia in children will be avoided as much as possible for renal investigations.

Keywords: renal malformations, ureteral duplex, minimal invasive

INTRODUCTION

Congenital anomalies are now easily identified using prenatal ultrasound. Renal and urinary tract malformations represent about 30 % of all anomalies identified during pregnancy (1). Because they play a causative role in about half of cases of chronic kidney disease requiring dialysis or renal transplant in children (2), it is important to have a complete and correct diagnosis of these anomalies, select the cases where treatment is necessary as soon as possible, in order to preserve renal function, to minimize renal damage and prevent or delay the onset of end-stage kidney disease.

Renoureteral duplications are not as rare as initially thought. With an incidence of approximately 1% in the general population, duplex kidneys

may be uni- or bilateral, complete or incomplete. It is known that most duplex kidneys do not become diseased, but when associated with upper pole ureteral ectopia, obstruction, ureteroceles or lower pole vesico-ureteral reflux can generate important morbidity with urinary flow obstruction, urinary tract infection, sepsis and/or renal function loss (3).

International protocols for managing these cases and decision making algorithms (Pediatric Urology web Book published by The European Society of Pediatric Urology or The Joint Clinical Guideline for: Management of Antenatally Diagnosed Hydro-nephrosis published by NHS UK) are now available and give clear pathways for investigating these children. The tendency is to reduce invasive procedures in children and procedures like intravenous urography are now rarely recommended due to high

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doses of radiation and unprecise information provided (4,5,6,7).

We report one case of complete renal duplication with upper pole obstructive megaureter that was managed in a private hospital. The aim of this paper is to show that this cases can be managed using minimal invasive methods available while keeping up with current European guidelines.

We obtained the consent of the parents for this case presentation.

CASE PRESENTATION

Presenting concerns

T.E.M is a 2 months old girl who presented to our clinic with fever and moderately altered general estate. At the time of presentation, physical examination revealed a normally developed for age female infant, with low fever and low degree of dehydration, with normal looking genitals. Laboratory findings were consistent with the diagnosis of urinary tract infection (urine analysis – bacteriuria, leukocyturia, nitrits positive). Oral antibiotic treatment was initiated and further investigations were performed. History revealed that the infant had a normal intrauterine development but right ureterohydronephrosis (grade II) had been detected at 23 weeks of gestation.

Clinical findings

Serial ultrasound were performed in order to follow up the hydronephrosis and ureteral dilatation (table 1).

Renal ultrasound examination was performed at birth and showed progression of ureteral dilatation, and again at first presentation in our Department: upper pole grade IV hydronephrosis, very thin renal parenchyma of the upper pole, greatly dilated and tortuous upper pole ureter, no ureterocele, right lower pole, left kidney and urinary bladder with normal appearance (figure 1).

TABLE 1. Progression of right renal dilatation as seen on prenatal and at birth ultrasonography

Gestational age (weeks)	Right kidney sizes	Ureteral dilatation
23 w	3.3/2.04/2.62 cm	1.70 cm
28 w	5/2.76/4.03 cm	Grade II hydronephrosys
31 w		1.20 / 3.39 cm - size of the lower right ureter grade I-II
35 w	5.24/3.60/3.67 cm	1.2/3.66 cm - size of the intrarenal urinary tracts
At birth		1.4 cm retro-vesical ureter

When antibiotic treatment was completed (7 days), urine examination showed no infection and low dose antibiotic urinary tract infection prophylaxis was started.

Diagnostic focus and assessment

In order to determine the cause of the ureteral dilatation and make a treatment plan we needed to perform a voiding cystourethrogram and a diuretic isotopic renogram.

We performed a renal ultrasound using SonoVue contrast solution (figure 2). Very good images were obtained and the diagnosis of complete right renal duplex system with upper pole obstructive megaurether was established. There was no ureterocele image, there was no lower pole or left side vesicoureteral reflux.

Diuretic isotopic renogram was the next step, in order to evaluate renal function. We performed Tc-DTPA renal scan and global normal function was observed with total filtration rate of 215 ml/min, almost even distribution of renal function among the two kidneys right/left = 44/56% The right upper pole had no detectable function and persistent upper pole ureterohydronephrosis while the lower pole had transitory hydronephrosis, moderately increased residual activity and nonobstructive diuretic response (figure 3).



FIGURE 1. Renal ultrasound at firts presentation showing: a) grade IV hydronephrosis with very dilated pelvis and very thin parenchyma of the rihgt upper pole b) toruous and dilated ureter measuring 14 mm in the retrovesical area c) no ureterocele image

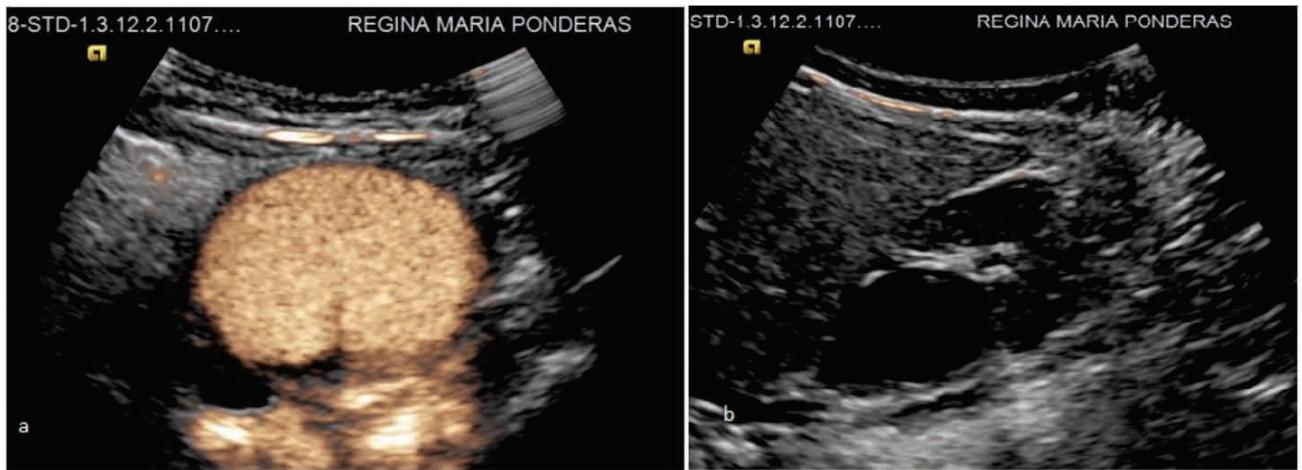


FIGURE 2. SonoVue ultrasound - no progression of the contrast in the upper urinary tract, no ureterocele a) bladder b) right kidney

Because of the clear date provided by the ultrasound and renal nuclear scan, we did not consider necessary to perform an MRI scan or CT scan, which at this age would involve general anesthesia.

Therapeutic focus and assessment

Taking into account all these data, we concluded that the complete diagnosis was: total right renal

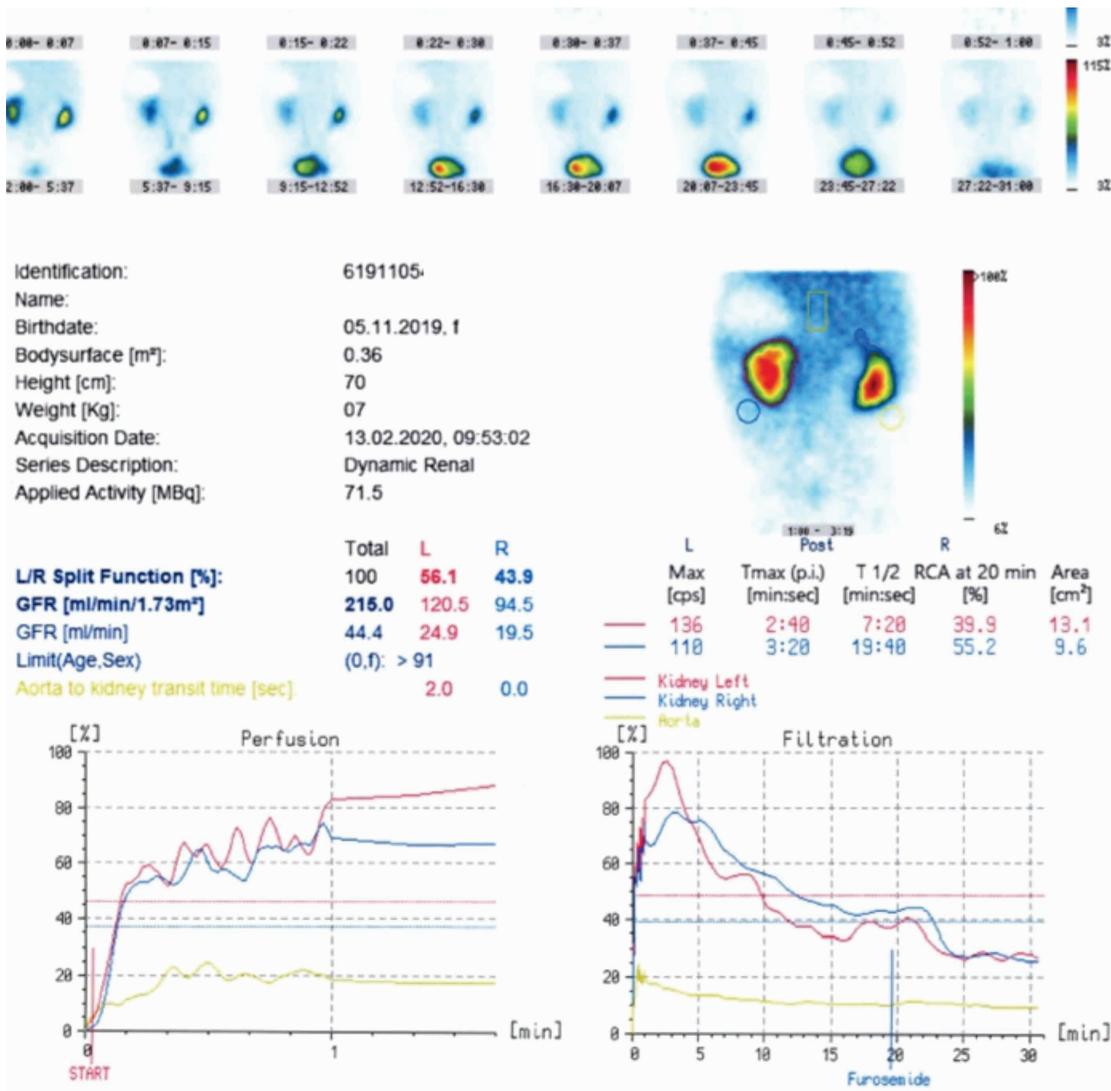


FIGURE 3. Tc 99 DTPA renal nuclear scan

duplex with upper pole obstructive megaureter and no upper pole renal function and we considered that upper pole nephroureterectomy is the best option.

Retroperitoneal, laparoscopic and open approach were all valid options but we chose to perform a laparoscopic transperitoneal right upper pole nephrectomy for many reasons – the large working space provided by this technique, the ease of access to the upper renal pole and to the retrovesical inferior ureteral pole and the fact that in our clinic we are more comfortable with the transperitoneal laparoscopic approach than the retroperitoneoscopic one. We also considered cystoscopy necessary at the beginning of surgery in order to locate the ectopic ureter, with double J stent placement in the lower pole ureter. In cases where the upper pole ureter is obviously dilated, double J stent is not mandatory because this ureter can be easily identified during laparoscopic dissection.

The purpose of treatment is to reduce external pressure on the right lower normal functioning renal pole also to reduce the risk of urinary tract infection by excising as much as possible of the tortuous dilated ureter, without damaging retro vesical structures or the normal lower pole ureter.

The surgical treatment is the only choice in this case, without it the urinary tract infection will be often and will lead to renal disease. We expect a good outcome after surgery with good renal function which will provide a good quality of life.

As we begun with cystoscopy, ectopic intravesical implantation of the right upper pole ureter was identified, with the other ureters normally located at the level of the trigone (figure 4).

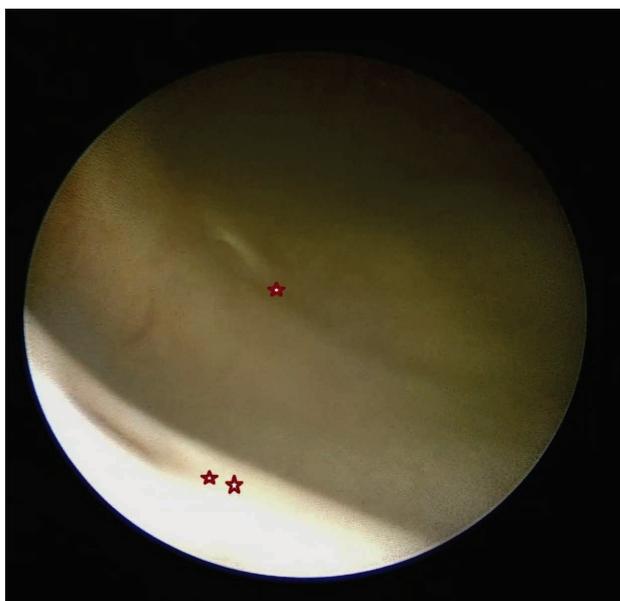


FIGURE 4. Cystoscopic aspect * lower pole ureter, normally implanted ** upper pole ureter near the bladder neck

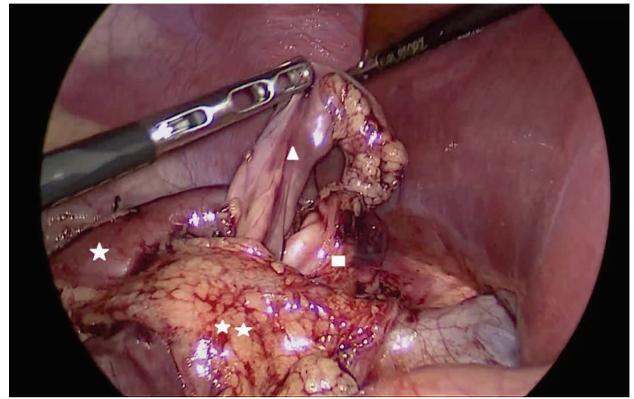


FIGURE 5. Laparoscopic aspect * right lower renal pole **renal main vascular pedicle ▲ dilated upper pole ureter ■ atrophic right upper renal pole

During laparoscopy we preceded in the standard manner: right colon mobilization, ureteral dissection as low as possible in the retrovesical space where we transected the distal ureter, dissection of the para renal fat and Gerota's fascia. To retract the liver we have used a trocar less 3mm instrument. After complete dissection of the upper pole pelvis and ureter, without injuring the renal vessels, we have resected the upper pole using bipolar electro-surgical instrument (figures 5,6).

Total surgery time was 150 minutes.

Follow-up and monitoring

A drain was left in place for 24 hours and the immediate postoperative period was uneventful. The child was discharged 48 hours later, after abdominal ultrasound reevaluation (figure 7).



FIGURE 6. Resected upper renal pole with the dilated ureter



FIGURE 7. Postoperative sonographic aspect - hyperechoic structures occupying the fossa of the upper excluded pole, the remaining lower pole, ureteral stump

DISCUSSIONS

Urinary tract infection is a common infection in children. Prompt diagnosis and appropriate treatment are very important to reduce the morbidity associated with this condition as Leung et al. also found (8). Arshad et al. say that urinary tract infections may be the sentinel event for underlying renal abnormality, although normal anatomy is most common. Prompt diagnosis and initiation of treatment is important in preventing long-term renal scarring (9). As Zee et al. say, risk factors for urinary tract infection in children with prenatal hydronephrosis are not clearly defined (10).

Taking into account that the classic voiding cystourethrograme (VCUG) exposes children to considerable amount of ionizing radiation, and that there are studies like the one of Kis et al., that obviate the superiority of contrast enhanced sonography for the diagnosis of vesicoureteral reflux especially in girls (11), in our hospital we have a very dedicated radiologist who is experienced in ultrasonography and so we used this kind of investigation.

Hiorns et al. compares the use of computed tomography (CT) versus magnetic resonance imaging (MRI), which are valuable tools for assessing the urinary tract (12). We think that in children the indication should be very precise for the use of computed tomography because of the radiation and for the use of MRI because of the need for general anesthesia.

Complete renal duplex system when associated with obstructive megaurterer, incontinence form ectopic ureter or vesicoureteral reflux, needs various surgical procedures to restore normal urine evacuation in order to avoid urinary tract infections and chronic renal disease. As in our case, when upper pole obstructive megaturer is associated with nonfunctional renal parenchyma, nefroureterectomy is the treatment of choice. Open and minimal invasive approaches are described with more advantages for the minimal invasive ones, with better

cosmesis, shorter recovery time and shorter hospital stay.

Although it is not clear but it seems that the laparoscopic approach has a lower complication and lower conversion rate than retroperitoneoscopic one, while providing better access to both upper renal pole and inferior pelvic segment of the ureter as Esposito et al. also says (13). As Lopes et al. emphasize that other techniques like clipping the ureter at the level of the bladder while leaving the dilated ureter in place are now being implemented for similar cases, but we think that more studies need to be conducted before validating such techniques (14).

There are discussions among specialist regarding the ureteral stump and its fate. It is known that when there is ureteral reflux, the ureter should be excised up to the level of the bladder in order to avoid stump reflux from the bladder and urinary tract infection but with obstruction and genital tract ectopia, the ureteral stump is left in place, as Ade-Ajayi et al. also consider (15). The risk of damaging the bladder's neck or the normally implanted ureter, (with which the dilated ureter shares vascularization) is considerable. As De Caluwe et al. says that postoperative events like necrosis of the distal part of the remaining ureter or incontinence should be kept in mind when dissecting in the retrovesical area, as they are frequent, while the remaining ureteric stump in obstructive megaurterer is rarely source of complications (16). Using modern vessel sealing devices there is a low risk of bleeding or urine leakage from the remaining kidney.

We hope that our experience in this case – the use of ultrasound of the abdomen for the anatomy, the contrast sonography for the investigation of the vesico-ureteral reflux and also the minimally invasive laparoscopic approach for the treatment – is a starting point for further research and management protocol of these patients.

CONCLUSIONS

In most cases duplex systems are associated with ureterocele which inquires cystoscopic incision during the neonatal period but in this case there was no ureterocele and no lower pole ureter reflux so no other surgical treatment was needed. Under antibioprohylaxis there were no additional urinary tract infections, growth and neurological development were normal and surgery could be scheduled at 1 year of age with lower risks involving general anesthesia and better surgical working space.

Because we have in our team an experienced radiologist, there was not necessary to complete the evaluation with more invasive investigations (voiding cystourethrogram, magnetic resonance imaging, computed tomography) so we avoided preoperative general anesthesia and irradiation.

We consider that in this case the laparoscopic approach is the best option with better access to both upper renal pole and lower ureter than the open or retroperitoneal techniques, good cosmetic results and short hospital stay.

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