# Renal duplex system in pediatric population: management and long-term follow-up

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**Background.** Duplex system is a duplication of renal parenchyma, pelvis and collecting system. It could be complete, if ureters lead to bladder separately or incomplete, if they joint before coming out. This study aims duplex system management evaluation, defining indications of conservative or demolitive therapy, and results.
**Materials and methods.** At the section of pediatric surgery of University of Siena we have observed 27 patients with duplex system from January 1980 to May 2011: 7 male (26%) and 20 female (74%), 18 (67%) with complete duplicity, 9 (33%) with incomplete one. Patients were divided into 2 groups: the first one was composed by 12 children (44%), they had negative diagnostic exams for alterations of renal function and associated diseases and no symptoms;the second group had 15 children (56%) whose diagnostic-therapeutic iter was based on associated malformations and symptoms of each case. We found: 5 RVU (33%) with 1 Hutch diverticulum; 5 ureteroceles (33%); 3 ectopic ureters (20%); 4 megaureters (26%), 6 renal dysplasia and upper pole function <10% (40%).
**Results.** Children belonging to second group were treated in different ways. 5 babies (33%) with RVU were approached with submeatal infiltration; 1 baby (7%) with ureterocele was treated with excision of the malformation and Cohen reimplatation; 2 ureteroceles (13%) were incised by transurethral approach and RVU appeared, 1 of them was then treated with eminephroureterectomy; in 1 case of ureterocele (7%)and 1 of ectopic ureter (7%) no treatment was undertaken and 1 baby with ureterocele (7%) needed eminephoureterectomy; 4 kidneys (26%) with upper pole impaired function required eminephroureterectomy. All second group patients, except 3, had a 2-16 years follow-up and they showed normal growth and no symptoms. First group children had negative exams and excellent clinic conditions.
**Conclusions.** Our results suggest that management should be decided on patient's age, clinic presentation and associated diseases.
**Keywords:** duplex system; associated diseases; individualized management.

## Introduction

The expression "renal duplex system" means a set of congenital anomalies of urinary upper and lower tract with partial or complete duplication of collecting system, pelvis and renal parenchyma. The kidney appears divided into two separated halves and ureters can be duplicated partially or totally. They can joint before the vesical outlet ("bifid system" or "bifid ureter") or they can separately lead to bladder ("complete ureteral duplication"). The incidence of the malformation is about 0,8%, female sex is more affected than the male one and the bifid ureter is present on the 60% of cases (complete ureteral duplication in the remaining 40% of cases). Duplex system arises by an embryologic anomaly due to a duplication or a split of the ureteral bud. Morphological and functional alterations, such as inversion of ureteral meatus and upper pole parenchymal dysplasia, are described respectively by Weigert-Mayert and Mackie-Stephen laws. Anomaly of ureteral bud implies an inadequate interaction between the mesonephros and the metanephic blasthema and an incorrect renal maturation. This is the reason of many associated to duplex system diseases; most frequent ones are: lateral ureteral ectopy, ectopic ureter, ureterocele, renal dysplasia and others. According to associated diseases, main symptoms can be attributed to urinary tract infection (UTI), incontinence, epididymis-orchitis, abdominal mass, urinary retention or growing retardation (1). Duplex system can remain asymptomatic all life long and so no treatment is necessary. If it becomes symptomatic because of associated disease, its treatment could be a medical approach or a surgical one and it depends on the kind of pathology and the age of patient (2). Our study aims to define the different management of duplex system basing on clinic; therapeutic option could be conservative, wait or reconstruction, or demolitive. We also aim to analyze results basing on undertaken choices by time.

## Materials and methods

At the department of Clinic of Pediatric Surgery of the

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University of Siena, we analyze 27 patients with renal duplex system from January 1980 to May 2011. In 18 patients (67%) the duplex system was complete, in 9 patients (33%) it was incomplete. Patients were 7 males (26%) and 20 females (74%); among males, there were 4 complete duplex systems (57%) and 3 incomplete ones (43%); among females, there were 14 complete duplex systems (70%) and 6 incomplete ones (30%). Duplex system was in 20 cases (74%) on the left side: 13 cases with complete renal system (65%) and 7 with incomplete (35%); 14 cases (70%) on female sex: 10 complete (71%) and 4 incomplete (29%); and 6 cases on male sex (30%): 3 complete (50%) and 3 incomplete (50%). On the right side we found 7 double districts (26%): 5 with complete duplex system (71%) and 2 with incomplete one (29%); 6 cases (86%) involve females: 4 complete (67%) and 2 incomplete (33%); only 1 case (14%) of complete duplex system was noticed on male sex (table I).

**tab.1.** *Distribution of our cases based on sex, side and completeness*

| | Male | Female |
|---|---|---|
| Right complete | 1 (4%) | 4 (15%) |
| Right incomplete | 0 | 2 (7%) |
| Left complete | 3 (11%) | 10 (37%) |
| Left incomplete | 3 (11%) | 4 (15%) |

In 2 cases (7%), the diagnosis of duplex system dated to prenatal age, 15 patients (56%) were analyzed at perinatal time, and the remaining 10 babies (37%) were joined up in preschool time. We performed urinary ultrasound, urinanalysis and urine culture to all patients at their entry. Then lab and instrumental checks were performed, such as blood exams, complete urine culture, ultrasounds, voiding cystourethrogram, renal scan, uro-NMR and cystoscopy. They were decided and performed with different timing and modality, basing on severity and onset of eventual symptoms. Our 27 patients were divided into 2 groups. The first group was composed by 12 babies (44%), 5 males (42%) and 7 females (58%), without any symptoms and with negative diagnostic exams for renal function's alteration and associated disease of urinary tract. Only 1 asymptomatic baby needed cystostomy and colostomy at birth because of the presence of cloaca; the baby was lost on follow-up. The second group was composed by remaining 15 cases (56%), 13 females (87%) and 2 males (13%). Their diagnostic and therapeutic iter was personalized according to found associated diseases and consequent symptoms. At the entry, all patients were submitted to ultrasounds: we found 9 hydronephrosis (60%), 4 megaureters (26%), 5 ureteroceles (33%), 1 ectopic ureter (7%), 1 cystic structure (7%), 2 decreases of upper pole cortical thickness (13%) and 6 increases of renal size compared to contralateral one (40%). At the same time there were 7 positivities of urine culture (47%), but only 5 cases (33%) were associated with hyperpyrexia. We performed voiding cystourethrogram in all babies and we found 5 vesical-ureteral reflux (VUR) (33%), 1 of them (7%) associated with Hutch diverticulum, and 5 ureteroceles (33%). To complete diagnostic iter, we performed cystoscopy in 5 patients (33%) that showed 4 complete duplex systems (26%), in 1 case (7%) there was an associated ectopic ureter and the outlet was on the bladder neck, and 1 incomplete duplex system (7%). All the 5 double districts were on the left side (33%). At the beginning of our experience we executed urography in only 2 cases (13%) and we could see hydronephrosis, megaureteres and delayed contrast removal from upper pole, especially in 1 of them (7%). In only 1 case (7%) we detected ureterocele. In 10 cases (67%) it was necessary to have a nuclear scan to complete diagnosis, in 6 babies (40%) with diethylene triamine pentaacetic acid (DTPA ) and in 5 ones (33%) with dimercapto-succinic acid (DMSA), only 1 patient (7%) needed both exams. All 6 DTPA-scans (40%) showed a decreased transit of the nuclear marker on the upper pole; on the contrary only 4 (26%) of the 5 DMSA-scans showed the same find. The conclusion was that 5 (33%) patients had a residual upper pole functionality under 10%. The clinic was hard in 7 patients, so we performed uro-MR. The results presented the association of a renal duplex system of hydronefrosis in 6 cases (40%), ectopic ureter in 3 cases (20%), ureterocele in 2 cases (13%) and megaureter in 1 case (7%). Concluding our study, we found: 5 VURs (33%), 1 (7%) on the right side and 4 (26%) on the left side (among the last group we also had an Hutch diverticulum); 5 ureteroceles (33%), 4 on the left (26%) and 1 on the right side (7%); 3 megaureteres (26%), all on the left side; finally 6 upper poles (40%) with functionality under 10%, all on the left side (table II).

**Table 2.** *Associated diseases to our population found by diagnostic exams.*

| | Total | Left complete | Left incomplete | Right complete | Right incomplete |
|---|---|---|---|---|---|
| VUR | 5 (33%) | 2 (13%) | 2 (13%) | | 1 (7%) |
| Hutch diverticulum | 1 (7%) | | 1 (7%) | | |
| Ectopic ureter | 3 (20%) | 3 (20%) | | | |
| Ureterocele | 5 (33%) | 4 (26%) | | 1 (7%) | |
| Megaureter | 4 (26%) | 4 (26%) | | | |
| Non functioning upper pole | 6 (40%) | 6 (40%) | | | |

## RESULTS

Our 15 patients of the second group were controlled basing on symptoms and found associated disease; if there was the need of a surgical approach (13 on 15 cases), this varied from endoscopy to laparotomy or a minimally invasive approach. The 5 (33%) VURs were treated with submeatal infiltration of Deflux® by cystoscopy. In 1 case (7%) of ureterocele the treatment was the excision of the malformation and the Cohen reimplantation of both of duplex system's ureters; the decision was taken basing on the obstruction of the urine flow at the age of two years. The same problem was observed in other 2 cases (13%) at the age of two and five years respectively; both of them were treated with the transuretral incision of ureterocele. After the incision, VUR developed and it was confirmed by control voiding cystourethrogram, so we dispensed antibiotic therapy. 1 case (7%) of ureterocele was without symptoms so any therapeutic step was necessary. In 6 cases (40%) upper pole function was under 10% so the approach was an upper pole eminephroureterectomy: in 5 cases (33%) this was in laparotomy and only in 1 case (7%) in retroperitoneoscopy. The retroperitoneoscopic eminephroureterctomy was performed in 1 case with ureterocele, which was treated previously by endoscopic incision and which developed VUR and UTI after this procedure. In 1 laparotomic eminephrectomy, an ureterocele was present and it was decompressed by the upper pole ureter's removal. We found only 1 isolated ectopic megaureter (7%) with the outlet under the bladder neck; we doesn't have still decided its most suitable management, waiting for the conclusion of the diagnostic iter (renal nuclear scan) and because of absence of symptoms. (table III)

Patients follow-up had a minimal time of 2 years (range: 2-16 years, average: 5 years). 3 patients were excluded from the follow-up: 1 patient with ectopic megaureter with an unconcluded diagnostic iter; 2 babies, lost during the observation time, 1 with an ureterocele without any treatment and 1 with developed after ureterocele incision. We performed urine exams, urine culture and urinary system ultrasonography to all patients during time of controls. Basing on every undertaken management or clinic of the baby at the moment of control, we added further exams such as blood exams, voiding cystourethrogram, nuclear scan, uro-MR and cystoscopy. We executed Deflux® infiltration in 5 babies and they don't present any symptoms related to VUR; we could perform cystourethrogram only after 6-12 months since infiltration, so we had the exam only in 3 cases of them and the results were negative. The baby, who underwent ureterocele excision and ureters reimplantation, had a negative urinary clinic. On opposite, we performed ultrasonographies every 3 or 6 months and they showed the increase of pelvis dilatation and the decrease of cortical thickness until 4 mm (the last one 4 months ago). Also renal nuclear scan showed that function of left kidney decreased and it was about 35-38%; the exam confirmed hydronephrosis with positivity to furosemide test. In 6 patients we performed upper pole eminephroureterectomies and the results are good, confirmed by negative clinic, urine colture, blood exams, voiding cystourethrogram and valid function of remaining lower pole, showed by nuclear scan. The other 12 patients don't present any associated diseases or symptoms and they always had negative control exams and excellent clinic conditions during follow-up.

## DISCUSSION

Renal duplex system, both complete and incomplete, can't be approached by a standardized diagnostic-therapeutic management. Indeed we have to plan the management basing on "when" and "how" diagnosis is defined. Analyzing our population, diagnostic exams and surgical treatment were different among children considering their clinic and their primary disease to solve.

Literature refers that first signs of a duplex system are related to UTI due to VUR. The main treatment of this condition is submeatal infiltration of Deflux® in VUR of low degree, because their resolution is similar than the single system reflux. Only a quarter of so treated patients needs an open approach in the future. So, according with Literature, we performed this procedure also in babies of our study (3). When the VUR is IV-V degree, it is better to perform an open approach, because the healing rate after infiltration is too low (4). In general, Literature suggests the open surgery when a "golf hole" ureteral orifice, a perpendicular to bladder intramural ureter and a VUR associated to ureterocele are present (5).

Management of ureterocele is difficult because it depends on the kind of ureterocele, the eventual damage

**Table 3.** *Different treatments performed on babies of II group.*

| | Total | Left complete | Left incomplete | Right complete | Right incomplete |
|---|---|---|---|---|---|
| Infiltration | 5 (33%) | 2 (13%) | 2 (13%) | | 1 (7%) |
| Ureterocele excision +Cohen reimplantation | 1 (7%) | 1 (7%) | | | |
| Eminephroureterectomy | 6 (40%) | 6 (40%) | | | |
| Ureterocele incision | 2 (13%) | 2 (13%) | | | |

on the renal parenchyma and the age of the baby. The treatment could involve only the bladder, only the kidney or both of them, to reduce the reoperation rate.
Controversy is still present about the evolution of upper pole parenchymal dysplasia and, by consequence, its treatment. Authors think dysplasia is an irreversible phenomenon linked to an alteration during embryological process; Others suggest the correction of associated disease to prevent the worsening of the situation (6,7).
According to Hendren and Mitchell, the gold standard of ureterocele treatment is the combined approach because of its lower rate of reoperation; it contemplates eminephrectomy, ureterocele excision and ureters reimplantation (8). When the upper pole functionality is not preserved, it is possible to execute eminephrectomy and extravescical ureterocele resection, followed by the repair of bladder wound without any reimplantation of lower pole ureter (9). Decter and Gonzales confirm the need to treat each case individually. According to their opinion, upper pole should be approached when the ipsilateral ureter of lower pole doesn't present any reflux or if this is of low degree; on the contrary, when a further procedure is necessary (such as with a high degree reflux or a prolapsing ureter) or when a non-responsive to therapy sepsis is present, Authors suggest the previous decompression of ureterocele by endoscopic incision, before the final surgery. The success of treatment vary depending on the intravescical or ectopic ureterocele's position and on the presence or absence of reflux. Extravescical ureteroceles need a further operation in a percentage of 40% of limited to upper tract approaches. If the degree of reflux is high, the percentage can increase until 53%. On the contrary, the need of a further operation is present in the 33% of cases, if the ureterocele is intravescical, performing the same procedure (10). Most important complications linked to upper urinary tract approach are: permanence or development of reflux, UTI due to ureteral stump, visible flank scar, possible lower tract damages and the need of reoperation of 50-84%. On the contrary, benefits include the decrease of risk of proteinuria, hypertension and cancerogenesis. The reconstruction of low urinary tract presents more benefits than upper tract approach, because it is possible to correct the disease, without a visible flank scar, and to improve renal function in a percentage of 15-50%. Complications include infections, postoperative incontinence, residual renal non-working units, possible following surgery and technical difficulties (11).
The complete urinary tract reconstruction can be considered the gold standard, having the best surgical outcome, but it is affected by an high morbidity, such as urinary incontinence and bladder dysfunctions, linked to low urinary tract involvement. So eminephrectomy or ureteroureterostomy are the best surgical options when babies are young, because their lower urinary tract is easier damaging. This procedure permits to postpone bladder involvement to an older age and a better anatomical maturation (2,12). If reflux is absent, eminephroureterectomy is the only performed procedure in 85% of cases; on the contrary if reflux is present, it is resolutive only in 16% of cases (2,13,14). According to Hussman, intravescical ureteroceles can be approach by endoscopic incision, because the reoperation rate is only about 23%. Instead, if the ureterocele is extravescical and it is incised, the percentage of reoperation is about 100%, so this approach is accepted only in emergency situation such as obstruction of urine flow, sepsis and neonates, where the anatomy is too weak to guarantee the success of a lower tract reconstruction. The possibility of failure of this procedure increases if VUR was present in preoperative time. Not incision neither eminephrectomy can be sufficient to solve disease in a patient with ectopic ureterocele and vesicoureteral reflux; the final act consists in total lower urinary tract reconstruction. The principal benefit of endoscopic approach of ureterocele is to stabilize temporarily urinary tract functionality (2,12,13,14,15,16). Guide lines about approach of ureterocele in duplex system are so defined:

1. Emergency treatment (obstruction to urine flow, sepsis and renal insufficiency): endoscopic incision.

2. Elective treatment:
• Intravescical ureterocele: endoscopic incision
• Ectopic ureterocele, functional renal upper pole and absence of VUR in lower pole ureter: ureteroureterostomy or uretero-pyelostomy from upper to lower pole
• Ectopic ureterocele, poor functional renal upper pole and absence of VUR in lower pole ureter: upper pole eminephroureterectomy (final treatment in 85% of cases)
• Ectopic ureterocele, poor functional renal upper pole and VUR in lower pole ureter or in contralateral one: upper pole eminephroureterectomy and bladder reconstruction (excision of ureterocele and bilateral ureteral reimplantation). With these features, the performance of ureterocele incision needs a reoperation in the 50-100% of cases; the upper pole nephrectomy needs a reoperation in the 84-90% of cases (17,18).

Looking at our therapeutic behavior, it is not so different than the one described in Literature. The approach of 5 children with ureterocele was different basing on clinical features. In 1 case, we perform ureterocele excision and ureters remplantation even if the baby was only few months old, because of an important obstruction of urine flow; in 2 cases we incised ureteroceles because of relapsing UTI and their lack of sensitive to antibiotic therapy; in 1 case we had a "wait and see attitude", because of the absence of any symptoms; finally in 1 case we executed eminephroureterectomy. Both ureteroceles incisions caused VUR in lower pole and one of them was treated with eminephroureterctomy, considering the poor functionality of upper pole. In six cases with renal functionality under 10% we performed eminephroureterectomy. According to Lit-

erature, we executed a careful ureterectomy, to avoid any possible VUR in the ureteral stump. In the last case we analyzed, the eminephrectomy was performed by a retroperitoneoscopic technique. This approach guarantee a lower postoperative pain, a faster intestinal recanalization, a shorter hospitalization and a better esthetic result than the laparotomic one (2,19).
According to our opinion, when the renal duplex system is found accidentally and any symptomatology is present, it is sufficient routine controls of urinary system.

## CONCLUSIONS

Our results could be considered satisfactory regarding both esthetics and functionality.
Nowadays, all our patients present a regular height-weight growth and they are symptoms-free. Diagnostic and therapeutic management of renal duplex system is complex and vary, indeed any gold standard doesn't exist.
It should be based on the way of the clinic presentation and on the age of each patient. It is important to define associated diseases to correct them quickly by miniinvasive techiques; this could reduce the postoperative hospital and allow the earliest recovery for the patient.

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