Urological and Nephrological Findings of Renal Ectopia

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Abbreviations and Acronyms

DMSA = dimercapto-succinic acid

GFR = glomerular filtration rate

VCUG = voiding cystourethrogram

VUR = vesicoureteral reflux

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Purpose: Urological characteristics of renal ectopia have been addressed previously but little is known about the functional consequences. We sought to study renal function, blood pressure, proteinuria and urological abnormalities in children with renal ectopia. As a secondary objective, we compared these parameters between simple and crossed ectopia.

Materials and Methods: For this retrospective, single center, observational study we reviewed case documents and radiological records. We also analyzed longitudinal data on blood pressure, proteinuria and kidney function.

Results: Renal ectopia was diagnosed in 41 cases, of which 26 (63%) were simple renal ectopia, ie unilateral pelvic kidney. In 32% of patients the diagnosis was made during prenatal screening. Median patient age was 0.24 years at diagnosis and 7.7 years at the most recent control visit. Associated urological abnormalities were found in 66% of patients. Voiding cystourethrography was performed in all patients, with vesicoureteral reflux shown in 13. In 8 of 10 cases with unilateral reflux the condition manifested in the orthotopic kidney. The relative function of the ectopic kidney on dimercapto-succinic acid scan was 38%, and in 22% of patients glomerular filtration rate was less than 90 ml per minute per 1.73 m^2. Albuminuria and proteinuria were absent in most cases. Longitudinal analysis of blood pressure, glomerular filtration rate and albuminuria revealed a stable course for all parameters. No substantial difference was observed between simple and crossed renal ectopia.

Conclusions: Our data suggest no adverse effects on blood pressure or kidney function in children with renal ectopia. However, periodic followup seems warranted, at least until young adulthood.

Key Words: kidney, urogenital abnormalities, urologic diseases, vesico-ureteral reflux

Renal ectopia is a rare congenital defect where the kidney is not located in the renal fossa. In rare cases the kidney is even located in the thorax.[1] Simple renal ectopia implies that the kidney lies ipsilateral in the pelvis, the result of defective ascension of the affected kidney during embryogenesis. In crossed renal ectopia the kidney is located contralateral to the side where the ureter enters the bladder, usually below the orthotopic organ. In the majority of cases the orthotopic and ectopic kidneys are fused.[2] During embryogenesis interaction between the ureteral buds and nephrogenic cords is essential for kidney development. Crossed renal ectopia has been speculated to result from fusion of the ureteral buds with only 1

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nephrogenic cord after lateral flexion of the "tail" of the embryo. In this position the wolffian duct (and with it the ureteral bud) crosses over the midline and fuses with the contralateral nephrogenic cord.[3]

Renal ectopia is asymptomatic in most patients, which explains why the incidence in autopsy series (1:1,000) is much higher than with clinical presentation (1:10,000).[4] Several studies have addressed the urological characteristics of renal ectopia.[4–8] However, little is known about the functional consequences. Therefore, we studied renal function, blood pressure and proteinuria in children diagnosed with renal ectopia. As a secondary objective, we compared these parameters between simple and crossed ectopia.

MATERIALS AND METHODS

The study was designed as a retrospective, single center, observational trial. Institutional review board approval and patient consent were waived. Patients were diagnosed with an ectopic kidney and followed at the pediatric renal center of VU University Medical Center between 1994 and 2008.

We reviewed the case documents and radiological records, and calculated GFR according to the method described by Schwartz et al (ml per minute per 1.73 m^2),[9] using a k-value of 49 for children older than 2 years, which has been validated by inulin clearance at our institution (unpublished data). We used serial data on spot urine samples to calculate protein-to-creatinine and albumin-to-creatinine ratios (mg/mmol), and analyzed these values using a general linear model with age as independent variable. ^{99m}Technetium DMSA scan was used to confirm the diagnosis. Differential renal function was calculated as the geometric mean of the anterior and posterior views on DMSA scan. In patients with crossed renal ectopia with fusion the regions of interest for the calculation of split function were drawn by comparing ultrasound images. In 5 cases no clear separation could be made, and these cases were excluded from analysis. VCUG was performed in all patients and VUR was graded according to the International Reflux Committee guidelines.[10] Pelvic dilatation was diagnosed by abdominal ultrasound and graded according to the Society for Fetal Urology classification.[11]

Blood pressure was measured on the right arm using appropriately sized cuffs after at least 10 minutes of rest.[12] A minimum of 3 consecutive blood pressure measurements were taken and the mean was used for further analysis. The blood pressure reading was converted into a z-score with an algorithm based on data from the Fourth Report on the Diagnosis, Evaluation, and Treatment of High Blood Pressure in Children and Adolescents,[12] using height z-scores for Dutch children.[13]

Statistical analysis was performed using JMP® and SPSS®, version 16.0. Quantitative data are presented as median (IQR). Findings in the simple and crossed renal ectopia groups were compared by Mann-Whitney U test or chi-square test, as applicable. Longitudinal data on blood pressure, proteinuria and kidney function were analyzed by multilevel analysis in a general linear model. The relationship between GFR and proteinuria/albuminuria was studied using linear regression analysis. For this analysis only data beyond age 2 years were included because of age related differences in reference values below this age. A p value of less than 0.05 was considered statistically significant.

RESULTS

Demographic data are summarized in table 1. A total of 41 patients were diagnosed with renal ectopia, of whom 26 (63%) had simple renal ectopia. Gender distribution was equal. In about two-thirds of the cases the left kidney was ectopic. Renal ectopia was diagnosed on prenatal screening in 13 patients (32%), and during evaluation for dysmorphism (single umbilical artery, preauricular pits, skin tags) in 10 (24%). In only 3 of these patients was a defined syndrome diagnosed, namely Mayer-Rokitansky-Kuster syndrome, Goldenhar syndrome and Treacher Collins syndrome. In 11 patients (27%) renal ectopia was symptomatic with urinary tract infection, hematuria, a palpable abdominal mass or renal insufficiency with hypertension. In only 7 patients (17%) the diagnosis was made coincidentally during abdominal ultrasound for a non-nephrological reason.

Median patient age was 0.24 years (IQR 0 to 1.84) at diagnosis, 1.55 years (0.2 to 7.6) at referral to our institution and 7.7 years (4.1 to 14.1) at the most

Table 1. *Patient characteristics*

	Simple Renal Ectopia		Crossed Renal Ectopia		p Value
No. gender:					1.00
M	12		7		
F	14		8		
No. side:					1.00
Lt	16		10		
Rt	10		5		
No. prenatal diagnosis/total No. (%)	11/26	(42)	2/15	(13)	0.08
Mean yrs age at diagnosis (range)	0.17	(0–2.74)	0.25	(0–1)	0.82
Mean yrs age at latest visit (range)	6.6	(3.4–11.8)	14.4	(4.5–17.5)	0.08
No. UTI/total No. (%):					
At presentation	4/26	(15)	3/15	(20)	0.69
During followup	9/26	(35)	10/15	(67)	0.059
No. VUR:					
Ectopic	2		0		
Orthotopic	6		2		
Bilat	2		1		
Grade IV–V	3		3		0.65
Overall/total No. (%)	10/26	(38)	3/15	(20)	0.067
% Differential renal function on DMSA (range)	39	(31–44)	38	(35–41)	0.92