Is a complete urological evaluation necessary in all newborns with asymptomatic renal ectopia?

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Aim: To evaluate if a complete urological screening is justified by potential urological anomalies in newborns or infants with asymptomatic renal ectopia (RE).

Methods: The database records of 60 consecutive neonatal cases of RE diagnosed at the authors’ hospital from 1990 to 2004 were retrospectively reviewed.

Results: At diagnosis, mean patient age was 1.6 months. In 58 of 60 patients, the suspected diagnosis of RE was made during prenatal or postnatal screening ultrasonography, including two newborns with anorectal malformation. There were 24 patients with crossed RE (C-RE) and 36 patients with simple RE (S-RE). A solitary RE was present in two patients. The most frequent associated urological abnormality was vesico-ureteral reflux (37.5% of C-RE and 16.6% of S-RE). Hydronephrosis was detected in seven RE and five contralateral kidneys. An obstructive meauraguer was present in one patient with C-RE. In 40% of S-RE and 92.9% of C-RE, the 99mTc DMSA documented reduction of function of RE. The longitudinal diameter of the ectopic kidney was significantly smaller than the contralateral one. Other non-renal diseases were present in 15% of patients, of which the most frequent was cryptorchidism (6.6%).

Conclusions: A complete urological evaluation is necessary in newborns with C-RE for the high incidence of associated urological anomalies, of which VUR is the most frequent. A complete urological evaluation is also reasonable in patients with S-RE who have a pelvic dilatation. No diagnosis or treatment of the associated urological diseases, in consideration of congenital decreased function of the ectopic kidney, might predispose these children to improvement of renal function impairment.

Key words: hydronephrosis, renal ectopia, vesico-ureteral reflux.

Introduction

Renal ectopia (RE) describes a kidney that lies outside the renal fossa. Simple RE (S-RE) refers to kidney that remains in the ipsilateral retroperitoneal space. The most common position is in the pelvis opposite the sacrum and below the aortic bifurcation. By definition, crossed RE (C-RE) exists when a kidney is located on the opposite side of the midline from where the ureter enters the bladder. Ninety percent of C-RE kidneys are fused from below to the normally located kidney.

Renal ectopia has been reported in approximately 1 in 1000 autopsies.1 Crossed ectopic kidney is a rare anomaly, with an estimated incidence of 1 in 7500 autopsies or 1 in 14 000 pediatric admissions.2–4 Some screening studies have reported the incidence of RE to be 1 in 5000 patients.1,5 Most cases of RE remain asymptomatic and the condition is incidentally discovered during screening for associated congenital anomalies or during imaging for other purposes.6

In previous studies, a high incidence of associated urological abnormalities was reported in children with RE.7 Hydronephrosis is the most common significant finding in RE,1 and the presence of associated vesico-ureteral reflux (VUR) is estimated in 25–70% of cases.5,6 For these reasons, prenatal or incidental diagnosis of RE is considered an absolute indicator for an early and complete investigation of the urinary tract, even in asymptomatic children.

The aim of this study was to evaluate if a complete urological screening is justified by potential urological associated anomalies in newborns or infants with asymptomatic ectopic kidney.

Methods

From 1990 in our hospital a facultative screening program has offered hip and kidney ultrasound (US) to all newborns in their first 4 months of life. From 1990 to 2004, 32 221 newborns were screened at our institution and a RE was diagnosed in 60 patients (0.2%), consisting of 39 boys (65%) and 21 girls (35%). Horseshoe kidneys were not considered in this study. At diagnosis, patient age ranged from newborn to 4 months (mean 1.6 months).

The final diagnosis in each patient was confirmed by static 99mtechnetium dimercapto-succinic acid (DMSA) scan. In two patients a magnetic resonance urography (MRU) was necessary, and in another two patients an intravenous urography (IVU) was performed. Voiding cystourethrogram (VCUG) was performed on all male patients and a color-Doppler cystosonography with echocontrast (CystoUS) was performed on all female patients. VUR was graded according to the International Reflux Committee guidelines.10 We used the Society for Fetal Urology classification system to grade dilatation of the upper urinary tract.11 Differential renal function was calculated by DMSA scan. In the right and left kidneys, DMSA uptake was measured 2 h after intravenous injection of radiisotope, as previously reported.12

For the ectopic kidney, attenuation correction was achieved using the geometric mean of the anterior and posterior view.
Statistical analysis

Comparisons of demographic and clinical parameters among groups were performed using the chi-squared test and unpaired t-test. Statistical significance was considered achieved if \( P < 0.05 \). Data are expressed as mean \( \pm \) SEM.

Results

An S-RE was found in 36 patients (60%), and a C-RE was found in 24 patients (40%). S-RE was pelvic in 33 patients (91.6%) and lumbar in three patients (8.4%). The S-RE was left side in 14 patients (39%), right side in 18 (50%) and bilateral in two (5.5%). A solitary ectopic kidney was present in two patients (5.5%). In C-RE, ectopic kidneys were crossed and fused in 18 of 24 patients (75%), while they were crossed but not fused in six (25%). In all crossed but non-fused kidneys, the ectopic kidney was in the pelvic position. The C-RE was left side in 18 patients (75%) and right side in six (25%).

In 58 of 60 patients, the suspected diagnosis of RE was performed during prenatal (23 patients, 38.3%) or postnatal screening US (35 patients, 58.3%). Two newborns were affected by anorectal malformation, so diagnosis of RE was carried out during preoperative genitourinary US. In 30% of patients, a renal agenesis was initially suspected.

Associated non-renal diseases existed in nine patients (15%). Of the patients with S-RE, three had cryptorchidism (8.3%) and one had absent vagina (2.8%). Of patients with C-RE, two had imperforate anus with sacral agenesis (8.3%), one had bowel malrotation (4.2%) and one had hypospadias with cryptorchidism (4.2%).
Associated urological abnormalities were recorded in 24 patients with RE (40%). In particular, these were present in 15 of 24 patients (62.5%) with C-RE (eight fused and seven non-fused kidneys) and in nine of 26 patients (25%) with S-RE (all in pelvic position).

A VUR was documented in 15 patients with RE (25%). VUR was bilateral in four patients (three girls and one boy) and unilateral in 11 (eight girls and three boys). A VUR grade II–III was found in nine patients (37.5%) with C-RE (three bilateral and six unilateral, five fused and four non-fused) (Figs 1, 2) and in six (16.6%) with S-RE (one bilateral and five unilateral, all in pelvic position). Of patients with unilateral RE, VUR involved the orthotopic kidney in 80%. A significantly greater incidence of VUR was recorded in girls than in boys (11

**Fig. 3** Case of left crossed renal ectopia (C-RE) in a patient with bowel malrotation. (a) Magnetic resonance urography showing a left C-RE with hydrouretheronephrosis and weak renal parenchyma. The right kidney appears rotated along the vertical axis and fused in middle line with the contralateral kidney. The small bowel is left localized. (b) Renal ecocolor Doppler demonstrating two renal adherent parenchymas in the right renal fossa. (c) Renal scan (DMSA) showing a unique renal formation (fused kidneys) in right paramedian site.
girls and four boys, respectively, \( P < 0.005 \). The grade of VUR was significantly greater in girls than in boys (\( P < 0.05 \)). No significant difference in the incidence of VUR was recorded between right or left side of RE. At 2 years, a spontaneous recovery of VUR was observed in three of four boys (75%) and in six of 11 girls (54.5%), four with S-RE and two with C-RE. One boy (25%) and five girls (45.4%) required operative treatment, performed at a mean age of 38 ± 8 months. Transstrigonal reimplantation was performed in a boy with grade III bilateral VUR, and four girls underwent endoscopic treatment.

A renal pelvic dilatation was detected on prenatal or neonatal ultrasound in seven RE non-refluxing units (grade III in two patients and grade II in five patients) and in five orthotopic kidneys (grade III in two patients and grade II in three patients). These patients underwent further investigations (DTPA with furosemide test in all patients, IVU in two patients and MRU in two patients) that documented ureteropelvic junction obstruction (UPJO) affecting the ectopic kidney in one patient with C-RE (non-fused kidneys) and the contralateral orthotopic kidney in two patients (one patient with S-RE and one patient with C-RE, non-fused). According to Anderson-Hynes, patients affected by UPJO underwent a pyeloplasty when they were 23 ± 4.6 months (range 18–27 months). A uretero-vesical junction obstruction (ectopic ureter) was recorded in one patient with C-RE. In this patient, the ectopic kidney was poorly functioning, while the contralateral orthotopic kidney was malrotated, with fetal appearance (Fig. 3). This patient required transstrigonal reimplantation with tailoring of the ectopic megaureter when he was 6 months old.

In 40% of the patients with unilateral S-RE, DMSA scan documented decreased function of the ectopic kidney (mean 37.5 ± 5.6%; range 29–47%). In C-RE, measurement of differential renal function using DMSA scan was possible in 14 patients; however, the superimposition of the fused kidney prevented the differential evaluation of renal function in remaining C-RE. In 13 patients (92.9%) with unilateral C-RE, DMSA scan documented decreased function of the ectopic kidney (mean 33.2 ± 3.8%; range 28–38%).

The longitudinal diameter of the ectopic kidney was significantly smaller than the contralateral one in S-RE (mean 37.4 ± 4.7 mm vs 49.5 ± 7.0 mm; \( P < 0.0001 \)). In C-RE, ultrasonographic measurement of longitudinal diameter was possible in 12 of 24 patients. The longitudinal diameter of the ectopic kidney was also significantly smaller than the contralateral one in C-RE (mean 25.0 ± 10.0 mm vs 43.0 ± 4.7 mm; \( P < 0.005 \)).

**Discussion**

Diagnosis of RE is usually achieved using DMSA scan, because DMSA remains bound to the cortical renal proximal tubules and is minimally excreted. In terms of using antero-posterior, latero-lateral and oblique projections, DMSA has been shown to be useful for diagnosing RE below L-2 and renal cortical lesions, and uptake measurements provide a marker for the functioning cortical mass.

The primary goal of our study was to evaluate the incidence of associated urological abnormalities in the randomized population of newborns with RE, selected after findings of RE during prenatal or postnatal screening US. A high incidence of associated urological abnormalities has been reported in patients with RE. \(^1\) \(^4\) \(^7\) The increased incidence of VUR in patients with RE was initially reported by Kramer and Kelalis, \(^7\) who studied a small group of symptomatic patients. In their study VCUG indicated an incidence of 52% and 70% in C-RE and S-RE, respectively. Hendren \(^1\) \(^3\) et al. observed reflux in six of nine children and Kheradpir \(^1\) \(^4\) et al. in three of five children with C-RE. Our data, according to Guarino \(^1\) \(^3\) et al., \(^7\) \(^8\) show that the risk of VUR, even if significant, was lower than what has been previously reported and it differs according to the type of RE. In our study, patients with C-RE had a risk of VUR of 37.5%, without any significant difference between fused and non-fused kidneys, while those with S-RE had an incidence of just 16.6%. It was suggestive that all S-RE affected by VUR had a pelvic RE. Of our patients with unilateral RE, the VUR affected the orthotopic kidney in 80%. We found that girls with RE have a higher incidence and grade of reflux. Our data suggest that ectopic kidneys are more disease-prone than are normal kidneys: they are often hypoplastic or dysplastic. \(^3\) On this basis, we encourage a VCUG in boys and a CystoUS in girls affected by C-RE, for the high risk of associated VUR.
and the evidence that postnatal US is a poor predictor of VUR. In our opinion, a cystographic evaluation is necessary in S-RE with a neonatal pelvic dilatation ≥8 mm.

In our patients the ectopic kidneys were significantly smaller than the contralateral, orthotopic ones, in particular in C-RE. Also, C-RE kidneys were significantly smaller than S-RE ones. In a previous study, Guarino et al. documented that in a high percentage of children with RE, the ectopic kidney had a primitive reduction of function. Also in infection. This condition occurs late and it explains the higher outflow with subsequent predisposition to hydronephrosis, calculi and (5%) children required surgery because affected by UPJO.

Previous reports documented a high incidence of hydronephrosis in children with RE. Gleason et al. not an incidence of hydronephrosis of 56% in ectopic kidneys and 26% in non-ectopic contralateral kidneys. Dilatation of the ectopic renal pelvis was the result of UPJO in 37% of patients and ureterovesical junction obstruction in 15%. Kramer and Kelalis reported a 33% incidence of hydronephrosis for UPJO in children with RE. In our study, in accordance to Guarino et al., the incidence of hydronephrosis is lower because we just enrolled asymptomatic newborns. In fact, in our experience, only three (5%) children required surgery because affected by UPJO.

It is clear that an abnormal position of the kidney can lead to poor outflow with subsequent predisposition to hydronephrosis, calculi and infection. This condition occurs late and it explains the higher incidence of hydronephrosis in older patients. On the evidence of high incidence of urological anomalies, in particular in patients affected by C-RE, we propose different evaluation on the basis of renal pelvic dilatation (Fig. 4).

Associated abnormalities of spinal growth have been found in 50% and genital anomalies in 40% of patients with RE. In our series just two patients with C-RE and imperforate anus with recto-urethral fistula showed an agenesis of the sacrum and skeletal anomaly. The incidence of genital anomalies in RE was 10.3% in boys (three patients with cryptorchidism and one with cryptorchidism and hypospadias) and 2.9% in girls (one patient with absent vagina). However, our data are influenced by the age of the studied patients. Many malformations of female genitalia are detected late with the occurrence of gynecologic symptoms (pelvic pain, infertility, repeated abortion).

In conclusion, in prenatal or neonatal diagnosis of C-RE, a complete urological evaluation is always necessary because a high proportion of these patients have associated urological anomalies, of which VUR is the most common. The presence of VUR in the contralateral, normally positioned kidney, together with decreased function of the ectopic kidney, might predispose these children to renal function impairment if the condition is not recognized and treated. In patients with S-RE, a complete urological evaluation is warranted if a renal pelvic dilatation coexists.

References