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Early kidney damage in patients born with unilateral renal agenesis[☆]



Daño renal precoz en pacientes nacidos con agenesia renal unilateral

To the Editor:

Unilateral renal agenesis, or solitary kidney, is a common disease (1/720 births) with a predominance of male patients. It affects the left kidney most frequently. Its aetiology and pathogenesis are unknown, with the literature describing possible genetic and environmental mechanisms.¹ Given its frequent association with other malformations, some authors hypothesise that it may be part of a syndrome.^{1,2} The diagnosis is made by ultrasonography, usually before birth. Performance of a nuclear medicine assessment is recommended to rule out nephrourologic comorbidities. These patients are at higher risk of developing proteinuria, chronic kidney disease (CKD) and/or high blood pressure, and unilateral renal agenesis is a frequent cause of CKD in children aged less than 5 years.^{3,4} In this article, we describe our experience with this disease.

We conducted a retrospective, observational and descriptive study by collecting data from the health records of children born with unilateral renal agenesis in our hospital between 2008 and 2015. We found records for 21 patients (57% male) with a mean age of 3.8 years for the period under study. Forty-five percent of the patients had a prenatal diagnosis of unilateral renal agenesis, which was confirmed postnatally by renal ultrasound in all. Of the remaining 55%

that received the diagnosis after birth, 66% had some type of malformation at birth (most frequently gastrointestinal), and underwent a renal ultrasound examination for the purpose of ruling out associated nephrourologic malformations (which were found in 45%, with a predominance of pyelocaliectasis [29%] and vesicoureteral reflux [VUR] [21%]). Other, less frequent reasons that led to diagnosis were oligohydramnios, spina bifida or acute pyelonephritis in the early days of life (11%).

There were more cases of left-sided renal agenesis (65%), and a solitary hypertrophic kidney was detected in 55% of cases. All patients underwent a workup that included measurement of plasma urea and creatinine levels, urinary sediment analysis and calculation of the albumin/creatinine ratio, and the results were normal in all at the time of testing. The evaluation was completed with imaging of the kidney by ^{99m}Tc-mercaptoacetyltriglycine (MAG3) renography in 80% and serial voiding cystourethrography (VCUG) in 30% of patients. Eighty-one percent of patients were followed up in outpatient services (median duration, 3 years; range, 0–7 years), with periodic checkups including measurement of blood pressure, renal ultrasound examination, renal function panel, albumin/creatinine ratio, urinary sediment analysis and calculation of the glomerular filtration rate (GFR) using the Schwartz formula updated in 2009. We found no cases of high blood pressure. Thirty-five percent of patients had at least one episode of acute pyelonephritis that responded well to antibiotic treatment. Fifty percent of these patients had VUR and received prophylactic antibiotherapy. Three patients (17%) developed early CKD, associated with microalbuminuria in 1 (Table 1). None of the patients needed renal replacement therapy.

One of the salient findings of our study was the high morbidity associated with unilateral renal agenesis, which was consistent with the literature. For instance, Westland et al.⁵ analysed 2684 cases of unilateral renal agenesis in children and adults and found high blood pressure, microalbuminuria and chronic kidney disease with a GFR of less than 60 mL/min/1.73 m² in 16%, 21% and 10% of

[☆] Please cite this article as: Castellano-Martinez A, Rodriguez-Gonzalez M, Roldan-Cano V. Daño renal precoz en pacientes nacidos con agenesia renal unilateral. *An Pediatr (Barc)*. 2017;87:171–173.

Table 1 Data of patients with unilateral renal agenesis and development of kidney damage.

	Patient 1	Patient 2	Patient 3
Renal agenesis	Right	Right	Left
Sex	Male	Female	Male
Age renal damage was detected	3 years	3 years	3 years
Comorbidities	Microtia Fistulae in 2nd and 3rd pharyngeal arches	Single umbilical artery Dysplasia of 5th finger Hemivertebra Hypotrophic rib Megaureter with grade III VUR Post-APN scar	Ventricular septal defect Mild pyelocaliectasis
Symptoms	Asymptomatic (one episode of APN)	Recurrent APN (3 episodes)	Asymptomatic
Height	96.5 cm	110 cm	112 cm
GFR	CKD III GFR 42 mL/min/1.73 m ²	CKD II GFR 65 mL/min/1.73 m ²	CKD II GFR 70 mL/min/1.73 m ²
Albumin/creatinine ratio	Normal	Normal	Microalbuminuria Albumin/creatinine 163 mg/g
Urea and creatinine	Urea 95 mg/dL Creatinine 0.94 mg/dL	Urea 61 mg/dL Creatinine 0.7 mg/dL	Urea 54 mg/dL Creatinine 0.66 mg/dL
Single kidney ultrasound	Normal	Normal	Renal hypertrophy
Treatment	Oral bicarbonate Oral ferrous sulfate Oral alfacalcidol	Prophylactic antibiotherapy	Enalapril

APN, acute pyelonephritis; CKD, chronic kidney disease; GFR, glomerular filtration rate; VUR, vesicoureteral reflux.

participants, respectively (the estimated mean age of the onset of complications in the paediatric subset was 9.1 years). The development of these complications can be explained by the hyperfiltration hypothesis proposed by Brenner et al.⁶ on the basis of animal studies, according to which hypertrophy would be a compensatory adaptation to the reduced number of nephrons in individuals with unilateral renal agenesis.^{3,4} In our series, 17% of patients developed complications early (mean age, 3 years). Thirty-three percent of them also presented with a solitary hypertrophic kidney, while 66% had some type of associated nephrourologic abnormality (grade III VUR, pyelocaliectasis and/or recurrent acute pyelonephritis), which led us to believe that the early development of renal damage in our series may have been related not only to renal hypertrophy but also to the presence of these comorbidities. Furthermore, due to ethical concerns few studies on the hyperfiltration theory have been conducted in children, and all are case series with small sample sizes and highly variable and inconclusive results.⁴ Therefore, we believe that it is important to continue reporting cases and to perform longitudinal studies with larger samples in children with congenital unilateral renal agenesis with the purpose of elucidating the pathophysiology of this disease. Last of all, we ought to underscore that these patients may be asymptomatic, and they must be under followup with regular checkups for the early detection and treatment of complications, even in the absence of associated kidney or urologic malformations or acute pyelonephritis, as renal

damage, as we found in our series, may be silent and occur in seemingly healthy patients.

Limitations of the study

Due to lack of documentation in the electronic health records, we could not obtain data on other factors that may be associated with less favourable outcomes in these patients, such as low birth weight, maternal use of pharmaceuticals during pregnancy or use of nephrotoxic drugs in the neonatal period, among others. Similarly, we do not know the reason for the performance of the abdominal ultrasound that led to postnatal diagnosis in 33% of patients.

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2341-2879/

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Experience with junctional atrioventricular reciprocating tachycardia[☆]



Nuestra experiencia con la taquicardia recíprocante permanente de la unión aurículo-ventricular

Dear Editor:

Permanent junctional reciprocating tachycardia, also known as Coumel's tachycardia, is a rare form of supraventricular tachycardia that is usually incessant, characterised by retrograde conduction via an accessory pathway usually located in the posteroseptal region with slow and decremental conduction.¹ Its characteristic electrocardiographic features (Fig. 1) are regular tachycardia with a narrow QRS complex, a long RP interval (RP > PR) and an inverted

P-wave in the inferior leads (II, III and aVF).² It is usually diagnosed during childhood, or even prenatally, and may cause tachycardia-mediated cardiomyopathy, although it may be asymptomatic and diagnosed in the course of a routine examination.³ It is usually refractory to medical treatment, and ablation of the accessory pathway by means of radiofrequency is considered a safe and effective definitive treatment.¹

We conducted a retrospective descriptive study of paediatric patients with a diagnosis of Coumel's tachycardia followed up at a tertiary level hospital in 2014 and 2015 to assess their outcomes.

Seven patients were followed up during these two years, of who 57.1% were boys and 42.9% girls. One patient received an early prenatal diagnosis and 2 received the diagnosis after tachycardia was detected in the foetus during monitoring before birth, requiring urgent caesarean delivery due to uncontrolled foetal tachycardia in all 3 cases.

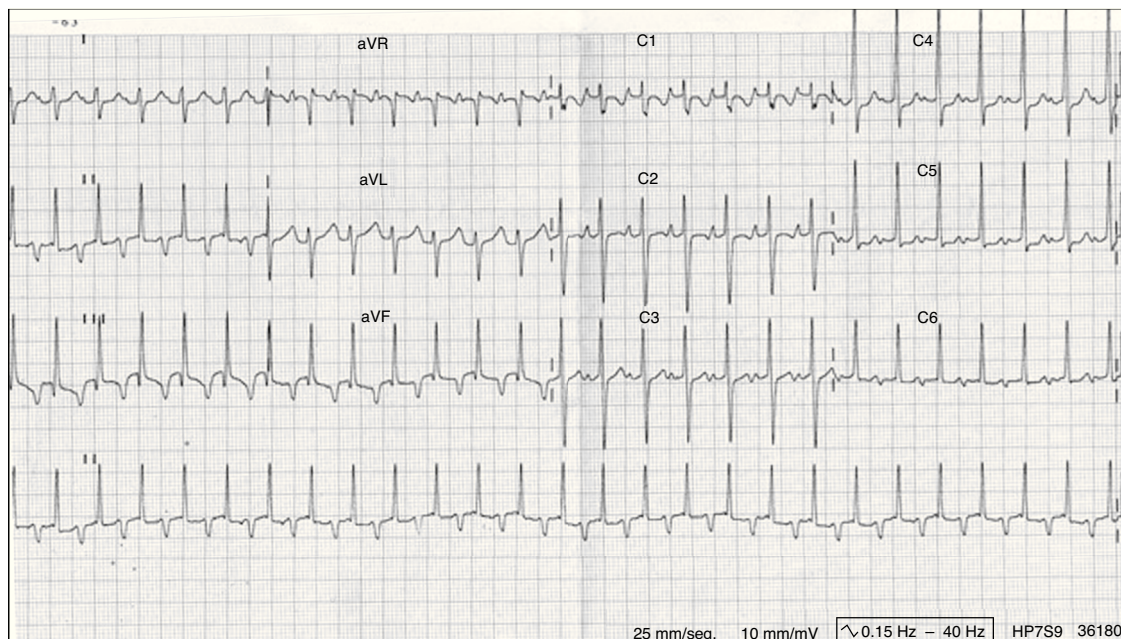


Figure 1 Electrocardiograph showing features of junctional atrioventricular reciprocating tachycardia. Regular tachycardia with narrow QRS complex, long RP interval (RP > PR) and a negative P-wave in the inferior leads (II, III and aVF).

[☆] Please cite this article as: Cardiel Valiente L, Ayerza Casas A, López Ramón M, Jiménez Montañes L, Sarquella-Brugada G. Nuestra experiencia con la taquicardia recíprocante permanente de la unión aurículo-ventricular. *An Pediatr (Barc)*. 2017;87:173–174.