

Prognostic Insights into Congenital Solitary Functioning Kidneys among the Turkish Pediatric Population: A Comparative Study of Renal Agenesis and Multicystic Dysplastic Kidney Disease

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ABSTRACT

Objective: Our study aims to assess the outcomes of solitary functioning kidneys (SFKs) resulting from unilateral renal agenesis (URA) and multicystic dysplastic kidney (MCDK) in order to identify the factors influencing kidney damage in these patients and to elucidate the potential contrasts between these two conditions.

Methods: The study retrospectively analyzes 154 pediatric patients (ages 0-18) with SFK treated at a tertiary center in Türkiye.

Results: Among the 154 SFK patients, 91 are male, 74 were diagnosed with MCDK, and 80 with URA. The median age is one month at diagnosis and 69.5 months at the last follow-up. Most MCDK cases were identified antenatally, while URA was most commonly detected incidentally. Congenital anomalies of the kidney and urinary tract (CAKUT) in the functioning kidney were identified in 21.6% of MCDK patients and 7.5% of URA patients, with a significantly higher occurrence in MCDK patients ($p = 0.012$). Vesicoureteral Reflux (VUR) was the most prevalent CAKUT, occurring in 45% of patients. A low glomerular filtration rate (GFR) was observed in 12.4% of patients, and 15% exhibited signs of renal damage. No significant disparity was found in low GFR or kidney damage between MCDK and URA patients. Those with a low GFR showed increased rates of hydronephrosis, CAKUT in the functioning kidney, recurrent urinary tract infections (UTIs), renal scarring, hypoplastic kidneys, proteinuria, and hypertension (HT).

Conclusions: The findings underline that CAKUT in the functioning kidney, recurrent UTIs, and renal scarring significantly influence GFR and kidney injury. VCUG is useful in select cases for identifying CAKUT, especially VUR. No significant distinction was observed between MCDK and URA concerning the eventual renal injury.

Keywords: Solitary functioning kidney, unilateral renal agenesis, multicystic dysplastic kidney disease, chronic kidney disease

INTRODUCTION

Congenital anomalies of the kidney and urinary tract (CAKUT) represent a leading cause of childhood chronic kidney disease (CKD), contributing to as many as 50-60% of cases (1). Solitary functioning kidney (SFK) may arise as a manifestation of CAKUT, particularly conditions such as unilateral renal agenesis (URA) and unilateral multicystic dysplastic kidney (MCDK), or may result from unilateral nephrectomy due to various underlying causes. Patients with SFK typically exhibit a 50% reduction in renal mass and nephron count, thereby inducing a state of hyperfiltration (2, 3). Schreuder et al. established a connection between a 20% decrease in nephrons and increased blood pressure, proteinuria, and glomerulosclerosis in animal models (4, 5). Furthermore, they found that half of the patients

with congenital reductions in renal mass (as seen in URA and MCDK) were hypertensive and displayed microalbuminuria (6), a finding that was later corroborated by multiple studies (7-9). Despite these observations and the seemingly favorable prognoses in long-term kidney donor follow-up studies (10), some adult studies reported 25-30% of patients with a single kidney to have a glomerular filtration rate (GFR) of less than 60 ml/min (11-13).

Multicystic dysplastic kidney (MCDK) is the most severe form of cystic renal dysplasia often identified via antenatal ultrasonography (USG) and is characterized by noncommunicating cysts separated by dysplastic tissue. The incidence rate of MCDK ranges from 1:3600 to 1:4300 live births, depending on the study and country (14). Unilateral

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Renal Agenesis (URA), with a prevalence of around 1:2000, results from a failed ureteric bud leading to the absence of one kidney (14).

In most recent reports, SFK, MCDK, and URA outcomes are typically studied collectively. However, some studies have begun to contrast these conditions, as they seemingly have different pathogeneses (15). This study aims to examine the clinical features and prognosis of patients diagnosed with URA and MCDK, two conditions that comprise a large portion of the clinic's SFK patient cohort. It aims to explore the factors influencing renal damage in these patients and to elucidate any potential differences between the two conditions.

MATERIALS and METHODS

The study incorporates 154 patients between 0-18 years of age diagnosed with MCDK and URA. These patients visited the Pediatric Nephrology Department of a tertiary referral hospital in Samsun, Türkiye, from January-December 2022.

Patient data were obtained retrospectively from hospital records, documenting primary diagnosis (URA or MCDK), gender, the side of the affected kidney, age at diagnosis and at last follow-up, clinical presentation, hypertension (HT), recurrent urinary tract infection (UTI), proteinuria, and GFR at diagnosis and at last follow-up, as well as any accompanying extrarenal anomalies. Factors potentially impacting renal damage in the functional kidney were also recorded; these included the size of the functioning kidney, hydronephrosis (HN), hypodysplasia, the presence of ureterocele in USG, vesicourethral reflux (VUR) in voiding cystourethrography (VCUG), ureteropelvic junction (UPJ) obstruction detected with Mercaptoacetyltriglycine (MAG)-3 scan, and renal scars as seen with a dimercaptosuccinic acid (DMSA) scan.

A UTI was identified based on the presence of pyuria and/or leukocyte esterase positivity, nitrite positivity, and significant growth in urine culture, alongside substantial clinical symptoms. Febrile infections were noted when recording the patients' previous UTIs. Those with two or more UTIs were classified as having recurrent UTIs. Blood pressure values were recorded, with HT defined as >130/80 mmHg for patients over 13 and above the 95th percentile for those under 13 (16). In cases of proteinuria, spot urine protein, spot urine creatinine, and 24-hour urine protein and volume values were noted for patients with positive protein results in the complete urinalysis. Proteinuria was identified when the spot urinary protein to creatinine ratio exceeded 0.2 mg/mg creatinine and/or 24-hour urine protein exceeded 4 mg/m²/hr.

Kidney function was evaluated based on the GFR and calculated using the Schwartz method and serum creatinine levels (17). Normal GFR levels vary depending on age, sex, and body size (18). GFR increases from infancy to adolescence, reaching adult average values at two years (19). This study focused on children over two years of age due to unreliable GFR values for those under two. Hence, GFR values were analyzed for 37 patients over two years old at admission and for 121 patients over the

age of two at last checkup. Patients were categorized into groups of GFR < 90 ml/min/1.73 m² and GFR > 90 ml/min/1.73 m². GFR < 90 ml/min/1.73 m² was taken as stage 2 chronic kidney disease (indicative of kidney damage with moderate GFR reduction) according to the KDOQI guideline (20). As per reviews and estimated GFR studies, the pediatric threshold for hyperfiltration was set at 135 mL/min/1.73 m² for children aged > 2 years (21).

The criteria for hydronephrosis (HN) and hydroureteronephrosis (HUN) included calyceal dilatation or a renal pelvic diameter > 10 mm or the presence of ureteral dilation as indicated by ureteral visualization in USG (22). During the final follow-up, the size of the functional kidney was logged in the USG, and the kidney percentile values were determined according to the 2023 publication of Obrycki et al. (23). Hypoplasia was defined as below the 2.5th percentile, while hyperplasia was above the 97.5th percentile. VCUG and DMSA were not routinely applied to all patients. VUR was graded per the International Reflux Study in Children (24). The DMSA scan was carried out at least 12 weeks after the recent febrile UTI, and any patients with scars were noted.

Statistical methods

Data entry and analysis are performed using IBM SPSS Statistics 22.0 (IBM Corp., Armonk, New York, USA). Categorical variables are described as numbers and percentages. Continuous variables have been evaluated for normal distribution using histogram and analytic (Shapiro-Wilk) tests. Parameters not fitting a normal distribution were defined through the median value with the distribution (lower-upper limit). Non-normally distributed continuous variables were compared using the Mann-Whitney U test. Categorical variables were compared using the χ^2 test. The nonparametric variant analysis Kruskal-Wallis test has been used for parameters that do not show a normal distribution in order to compare more than two groups, with a *p*-value < 0.05 being considered significant.

This study was approved by Samsun Ondokuz Mayıs University Clinical Research Ethics Committee with Decision No. 2023/108 dated 27/04/2023.

RESULTS

General characteristics of the SFK population

The study's cohort of 154 SFK patients is comprised of 91 males (59.1%) with a median age of one month at diagnosis. The median age at last follow-up is 69.5 months, spanning a median follow-up period of 50 months. Of these patients, 74 (48.1%) were diagnosed with MCDK and 80 (51.9%) with URA, with the left kidney being the most commonly affected side in 94 patients (61%). Most cases were identified antenatally (57.8%, *n* = 89), while 47 patients (30.5%) were incidentally found to have SFK. Twenty-two patients (14.3%) demonstrated an additional CAKUT in the functional kidney, with VUR being the most frequent and occurring in 45% of these cases. Additionally, 21 patients (13.6%) exhibited an extrarenal congenital anomaly. A summary of these findings can be found in Table 1.

Table 1: Characteristics of the patient group

	Total	MCDK	URA	p
Number of patients (%)	154	74 (48.1)	80 (51.9)	
Male (%)	91 (59.1)	44 (59.4)	47 (58.7)	0.929
Affected kidney: left (%)	94 (61.0)	46 (62.2)	48 (60)	0.783
Age at presentation (months) Median (min-max)	1 (1-178)	1 (1-178)	8 (1-165)	<0.001
Age at last follow-up Median (min-max)	69.5 (1-216)	59 (2-216)	81.3 (1-216)	
Clinical presentation (%)				
Antenatal diagnosis	89 (57.8)	64 (86.5)	25 (31.3)	<0.001
Incidental	47 (30.5)	7 (9.5)	40 (50.0)	
UTI	10 (6.5)	2 (2.7)	8 (10)	
Voiding disorder	3 (1.9)	-	3 (3.8)	
Screening for congenital anomalies	3 (1.9)	-	3 (3.8)	
Symptom of HT	1 (0.6)	-	-	
Family history	1 (0.6)	1 (1.4)	-	
CAKUT in the functional kidney (%)	22 (14.3)	16 (21.6)	6 (7.5)	0.012
Extrarenal congenital anomaly (%)	21 (13.6)	11 (14.9)	10 (12.5)	0.669

MCDK: Multicystic dysplastic kidney, URA: Unilateral renal agenesis, UTI: Urinary tract infection, HT: Hypertension, CAKUT: Congenital anomalies of the kidney and urinary tract

Comparison between URA and MCDK

Of the 74 MCDK patients, 44 are males, and of the 80 URA patients, 47 are male, thus revealing no significant gender difference between the two conditions. The left kidney was the one most commonly affected in both groups. The median age at diagnosis is one month for MCDK (Range = 1-178 months) and eight months for URA (Range = 1-165 months), indicating a significantly earlier diagnosis for MCDK ($p < 0.001$). Most MCDK cases (64 out of 74) were diagnosed antenatally, with most occurring at one month of age. In contrast, incidental detection of a cystic kidney was the second most frequent clinical presentation for MCDK ($n = 7$ patients; 9.5%), with other uncommon presentations being UTI, voiding disorders, congenital anomaly screenings, HT symptoms, and family history of cystic kidney disease. For URA, the most common reason for diagnosis was an incidentally detected single kidney on USG ($n = 40$, 50.0%), followed by an antenatal presentation ($n = 25$), UTI ($n = 2$), and family history of kidney anomaly ($n = 1$). These findings are summarized in Table 1.

CAKUT in the functioning kidney was detected in 16 MCDK patients (21.6%) and 6 URA patients (7.5%), occurring significantly higher in MCDK patients ($p = 0.012$). Seven patients had VUR, four had hypodysplasia, two had UPJ obstruction, and three had ureterocele without VUR. Other minor pathologies were simple cysts in one patient and kidney stones in two patients. In the URA group, three patients had VUR, one had hypodysplasia, one had ureterocele without VUR, one had a posterior urethral valve (PUV), three had simple cysts, and two had kidney stones. VUR was the most prevalent CAKUT, occurring in 45% of patients (10 out of 22), with seven cases belonging to

the MCDK group. One patient had grade 1 VUR, one had grade 2 VUR, and the remaining eight had grades 3-5 (high grade) VUR. In summary, high-grade VUR in functioning kidneys was more common in MCDK patients in the study's SFK-diagnosed cohort.

Additional extrarenal anomalies were present in 21 patients (13.6%): 11 with MCDK and 10 with URA. Genital abnormalities were found in eight MCDK and one URA patient, while cardiac anomalies were found in two MCDK and three URA patients. One URA patient had a combined cardiac and genital abnormality. Midline defects such as esophageal atresia, anal atresia, tracheoesophageal fistula, and cleft lip and palate were present in one MCDK patient and six URA patients. One URA patient had a triple x chromosomal anomaly. Genital abnormalities in this study's cohort were more common in MCDK, while midline defects were more common in URA.

In the study's cohort of 154 patients, all had undergone USG, with HN identified in 22.1%. This occurrence was significantly higher among MCDK patients ($p = 0.010$). VCUG was performed on only 56 patients, revealing VUR in 10 patients (17.9%). No significant difference occurred in the detection rates of VUR between MCDK and URA patients (see Table 2). MAG-3 scintigraphy was only performed on 21 patients, with obstructive findings detected in only two patients diagnosed with MCDK. Consequently, the results indicate MCDK patients to exhibit more VUR and UPJ obstruction, which explains the higher incidence of hydronephrosis in this group.

Proteinuria, a sign of kidney damage, was identified in 7.8% (12 patients) of the cohort, while HT was found in 3% (4 patients). The length of the functioning kidney at initial visit and the final

Table 2: Comparison of clinical, laboratory, and radiological findings of MCDK and URA

	Total	MCDK	URA	p
GFR at presentation (%) <90 ml/min/1.73m ²	3/37 (8.1)	0/8 (0.0)	3/29 (10.3)	0.343
Hyperfiltration at presentation (%) GFR>135 ml/min/1.73m ²	1/37 (2.7)	0/8 (0.0)	1/29 (3.4)	0.594
GFR at last follow-up (%) <90 ml/min/1.73m ²	15/121 (12.4)	7/61 (11.5)	8/60 (13.3)	0.757
GFR at last follow-up (%) <60 ml/min/1.73m ²	6/121 (5)	2/61 (3.3)	4/60 (6.7)	0.391
Hyperfiltration at last follow-up (%) GFR>135 ml/min/1.73m ²	21/121 (17.4)	9/61 (14.8)	12/60 (20)	0.446
Recurrent UTI (%)	16/154 (10.4)	9/74 (12.2)	7/80 (8.8)	0.488
Hydronephrosis in USG (%)	34/154 (22.1)	23/74 (31.1)	11/80 (13.8)	0.010
VUR in VCUG (%)	10/56 (17.9)	7/38 (18.4)	3/18 (16.7)	0.873
Renal scar in DMSA scan (%)	11/136 (8.1)	4/70 (5.7)	7/66 (10.6)	0.296
Obstruction in MAG-3 scan (%)	2/21 (9.5)	2/14 (14.3)	0/7 (0.0)	0.417
Proteinuria (%)	12/154 (7.8)	5/74 (6.8)	7/80 (8.8)	0.645
Hypertension (%)	4/133 (3.0)	3/62 (4.8)	1/71 (1.4)	0.248
Length of SFK at last follow-up (%)				
Measured kidney side: Left	60/152	26/72	34/80	
<2.5 p (Hypoplasia)	9/152 (5.9)	5/72 (6.9)	4/80 (5.0)	0.241
2.5-97.5 p	72/152 (47.4)	29/72 (40.3)	43/80 (53.8)	
>97.5 p (hypertrophy)	71/152 (46.7)	38/72 (52.8)	33/80 (41.3)	
Renal damage*	20/133 (15)	8/62 (12.9)	12/71 (16.9)	0.520

Available data is specified separately in each row due to variability.

GFR: Glomerular filtration rate by Schwartz (ml/min/1.73m²), patients aged > 2 years were analyzed

MCDK: Multicystic dysplastic kidney, URA: Unilateral renal agenesis, UTI: Urinary tract infection, USG: Ultrasonography, SFK: Solitary functional kidney, VCUG: Voiding cystourethrography, VUR: Vesicourethral reflux, DMSA: Dimercaptosuccinic acid scan, MAG-3: Mercaptoacetyltriglycine scan

*Renal damage was defined as having at least one of these; Hypertension, proteinuria, low GFR

follow-up was determined using age-adjusted percentiles and then categorized into three groups. At the final follow-up, nine patients (5.9%) were below the 2.5th percentile, and 71 (46.7%) were above the 97.5th percentile, as outlined in Table 2. These findings indicate that compensatory hypertrophy had developed in nearly half of the patients by an average age of 69.5 months (5.8 years). No significant differences were found in the rates of hypoplasia and hypertrophy between the MCDK and URA patient groups at the end of the follow-up period.

When examining the kidney sizes of 87 infants (60 MCDK and 27 URA) who'd been admitted within their first month of life due to an antenatal diagnosis or UTI, 8 (9.2%) were found to have hypoplastic kidneys (<2.5th percentile) and 41 (47.1%) to have hyperplastic kidneys (>97.5th percentile). This suggests that hyperplasia can be detected within the first month of life in nearly half the patients. Hyperplasia was observed in 31 MCDK patients (51.7%) and 10 URA patients (37%) at one month of age, a difference that is not statistically significant.

Factors affecting GFR in SFK patients

Glomerular filtration rate (GFR) was assessed in patients aged two years and above. Table 2 reveals that three patients had a

low GFR at the initial consultation, while 15 patients displayed a low GFR at the last follow-up. A total of 22 patients showed signs of renal damage, which is defined in this study as having at least one instance of HT, proteinuria, or a low GFR. No significant difference was seen in low GFR or kidney damage between the MCDK and URA patients ($p = 0.757$, $p = 0.520$, respectively).

Given this, Table 3 presents the details of the study's evaluation of the factors impacting low GFR without categorizing SFK patients. The patients with a low GFR displayed higher rates of hydronephrosis, CAKUT in the functioning kidney, recurrent UTIs, renal scarring, hypoplastic kidneys, proteinuria, and HT. Of these 15 patients, four had hypodysplasia, three had VUR in the functioning kidney, one had a PUV, and one had a ureterocele. Upon stratifying the 15 patients with GFR < 90 ml/min/1.73m² into stages of CKD, two patients were found to be in stage four, four in stage three, and nine in stage two. One of the patients with left MCDK and a severely hypo/dysplastic right kidney had undergone a preemptive kidney transplant and, as such, was not included in the low GFR group.

The study also examined the effect of extrarenal anomalies on GFR and found no difference in the frequency of these

Table 3: Factors affecting GFR in SFK patients

	GFR<90 ml/min/1.73m ² at last follow-up	GFR>90 ml/min/1.73m ² at last follow-up	p
Gender (male)	8/15 (53.3)	65/106 (61.3)	0.554
MCDK	7/15 (46.7)	54/106 (50.9)	0.757
URA	8/15 (53.3)	52/106 (49.1)	
Recurrent UTI	6/15 (40.0)	9/106 (8.5)	0.001
Proteinuria	6/15 (40.0)	4/106 (3.8)	<0.001
Hypertension	2/15 (13.3)	2/106 (1.9)	0.020
Hydronephrosis in USG	9/15 (60.0)	23/106 (21.7)	0.002
CAKUT in functional kidney	10/15 (66.7)	10/106 (9.4)	<0.001
Renal scar in DMSA scan	6/14 (42.9)	5/93 (5.4)	<0.001
Length of SFK at last follow-up			
<2.5 p (Hypoplasia)	3/15 (20.0)	4/104 (3.8)	
2.5-97.5 p	7/15 (46.7)	44/104 (42.3)	0.056
>97.5 p (hypertrophy)	5/15 (33.3)	56/104 (53.8)	
Extrarenal anomaly	4/15 (26.7)	14/106 (13.2)	0.170

Available data is specified separately in each row due to variability.

GFR Glomerular filtration rate by Schwartz (ml/min/1.73m²), patients aged > 2 years were analyzed

MCDK: Multicystic dysplastic kidney, URA: Unilateral renal agenesis, UTI: Urinary tract infection, USG: Ultrasonography, VCUG: Voiding cystourethrography, VUR: Vesicourethral reflux, DMSA: Dimercaptosuccinic acid, SFK: Solitary functional kidney

abnormalities between MCDK and URA patients. Furthermore, the study found no significant effect of extrarenal anomalies on low GFR, as illustrated in Table 3.

Hyperfiltration was detected in 21 patients at final follow-up (see Table 2), with no significant difference observed between MCDK and URA patients. When comparing the GFR values of the patient group with renal hypertrophy to those with normal kidney size, no significant difference was found regarding hyperfiltration rates. Among those aged two years and above, 5 out of the 25 patients (20%) with compensatory hypertrophy and 16 out of the 93 patients (17.2%) with normal kidney sizes had a GFR > 135 ml/min/1.73m² (p = 0.746).

DISCUSSION

CAKUT is frequently recognized as the leading cause of end-stage renal disease in various studies. SFK can result from numerous congenital renal anomalies, notably MCDK and URA. According to many reports, a 25-30% reduction in renal function and GFR is anticipated by adulthood. Thus, identifying these patients’ risk factors for renal damage is vital for their ongoing management and determining appropriate treatment options during childhood.

This study has examined 154 patients diagnosed with SFK. While some studies identified no male predominance in MCDK (15, 25), the current study’s patient group aligned more with most of the literature, showing males to be more likely to be affected regarding both MCDK and URA (14, 26). Also in line with previous research, the left kidney was more frequently affected (27), and the most common presentation occurred in the antenatal diagnosis.

The incidence of CAKUT in patients with SFK varies from 25-50%, according to different studies (9, 15, 25). The current study’s findings indicate a lower incidence of 21% for MCDK and 7.5% for URA, primarily notable for URA compared to the existing literature. The SOFIA study group, which examined the largest cohort of SFK patients to date (including 308 MCDK and 150 URA among 715 congenital SFK cases), reported a similar severe CAKUT comorbidity rate of 21% (including grade 3 or 4 hydronephroses, grade 3-5 VUR, parenchymal abnormalities or defects, and/or dysplasia detected via USG, VCUG, or nuclear scan), which aligns more closely with the current findings (28). The discrepancy in results might be attributed to the differing definitions of CAKUT across studies, with some considering simple cysts and mild hydronephrosis as CAKUT and others not. Similar to the SOFIA study group, this study also discounted simple cysts and mild hydronephrosis and found a similar CAKUT rate for MCDK. Consistent with many other studies, VUR was also identified as the current study’s most common renal anomaly (9, 29, 30).

The prevalence of extrarenal congenital anomalies in patients with MCDK and URA has been reported to vary widely. Notable associations with the central nervous system (e.g., meningomyelocele), ears, nose, throat, cardiac, genitourinary, gastrointestinal, musculoskeletal, and pulmonary anomalies have been documented (15, 25, 28). These studies observed a higher association with URA. Furthermore, genetic syndromes were reported in 15% of URA cases and 6% of MCDK cases (15). However, the current study found a lower frequency of extrarenal congenital anomalies, observing a higher occurrence of extrarenal congenital anomalies in MCDK than in URA, which contradicts the findings from other studies. Genital anomalies

were the most common type of extrarenal congenital anomaly this study encountered, with only one patient being found to have a genetic syndrome.

The necessity for routine VCUG and DMSA scans for patients with MCDK and URA is currently debated. VCUG is more commonly accepted for selected patients, whereas many centers have made DMSA scans a routine part of their protocol. In the current study, VCUG was performed on approximately one-third of the patients, with VUR being detected in 17.9% of them, a finding that aligns with another study in Türkiye (31). In children who've undergone VCUG due to recurrent UTIs, the likelihood of detecting VUR varies between 15-50% depending on the age group (32, 33). Given the current results, this study argues that routine VCUG may not be necessary for patients with SFK.

At the median age of 70 months, the study observed proteinuria in 7.8% of the patients and HT in 3%. Comparatively, studies have shown hypertension in SFK patient groups, which include both MCDK and URA patients, to range from 8%-34%. For patients around four or five years old, hypertension typically ranges between 8-15% (9, 15, 25). However, this rate climbs to 23% in studies with an 11-year follow-up and reaches 34% in those with a 12-year follow-up (28, 34). The current study indicates a lower proteinuria and hypertension prevalence than these reports, which may be attributed to the patients' relatively young median age during their last follow-up. The study's findings highlight the increasing risk of hypertension with age in patients diagnosed with SFK, thereby emphasizing the need for regular blood pressure monitoring during patient follow-ups. This claim is further supported by a study of a Chinese cohort (average age = 32), which reported a high hypertension rate of 38% (13).

Compensatory hypertrophy is a common occurrence in patients with SFK, observable in 24-48% of MCDK cases as early as the 20th gestational week (14, 35). However, it doesn't occur in all patients (36). The current study detected hypertrophy in 50% of patients whose average age is 5.8 years. Additionally, hypertrophy was observed in half of the 87 infants who'd undergone postnatal ultrasonography within the first month. These hypertrophy rates are consistent with those found in many other studies.

Hyperfiltration is a significant consequence of hypertrophy. While some studies associate kidney length with GFR and higher GFR in kidneys with compensatory hypertrophy, claiming that those without hypertrophy are at a higher risk of kidney failure is challenging (36). Furthermore, though the exact mechanism remains unclear, glomerular hyperfiltration is linked to progressive renal damage and loss of kidney function (37). In a study comparing Wilms tumor and MCDK patients regarding compensatory hypertrophy and GFR, both groups exhibited respective hypertrophy rates of 100% and 82% over nine years, and this hypertrophy was associated with an increase in GFR (38). In the current study (see Table 3), patients with kidney length < 2.5th percentile had lower GFR, while those in the >97.5th percentile had higher GFR. However, this difference was

not statistically significant ($p = 0.056$). Hyperfiltration was only detected in 17% of this study's patients and was not associated with proteinuria, hypertension, or GFR. This might be attributed to the shorter follow-up period, necessitating studies with longer follow-ups or even examining adult patients' statuses.

At admission, GFR could only be evaluated in 37 patients due to the small number of patients over two years of age; however, it was able to be assessed in 121 patients at the end of the follow-up. CKD stage two or higher was observed in 12.4% of the study's cohort. CKD rates in SFK vary widely in the literature, with this variation thought to be due to differences in patients' last follow-up age and the CAKUT rates regarding SFK. Also, some studies have reported patients with GFR < 90 ml/min/1.73m² and some with GFR < 60 ml/min/1.73m². In the current study, 5% of patients had a GFR < 60, similar to other studies with a last follow-up age of 4.5-9.5 years (9, 15, 25). A long-term study of 944 SFK patients reported a GFR < 90 ml/min/1.73m² at a rate of 31% (28). This could be due to the patients being older than the current study's cohort. Severe renal damage was reported in 39% of patients in the same study, but only 3% had GFR < 60 ml/min/1.73m².

As presented in Table 3, this study identified the factors affecting GFR in SFK patients as hydronephrosis, CAKUT in the functioning kidney, recurrent UTI, scarring in DMSA, proteinuria, and HT. These findings align with most literature studies. Unilateral renal agenesis (URA), identified as a poor prognostic factor in some studies (15, 28), did not significantly impact the final GFR compared to MCDK in this study.

CONCLUSION

To conclude, this study highlights the key factors that affect GFR and renal damage in patients with SFK, specifically MCDK and URA. The findings indicate CAKUT in the functional kidney, kidney scarring, and recurrent UTIs to significantly impact GFR and renal injury. VCUG has value in selected cases for identifying CAKUT, particularly VUR, the most prevalent CAKUT. Proteinuria and HT are more frequent in patients with lower GFR. No significant difference was observed between MCDK and URA regarding renal injury upon final follow-up. While compensatory hypertrophy commonly develops, further investigations examining long-term prognosis into adulthood are necessary to understand the implications of hyperfiltration in these patients.

Ethics Committee Approval: This study was approved by the ethics committee of Samsun Ondokuz Mayıs University Clinical Research Ethics Committee with Decision No. 2023/108 dated 27/04/2023.

Informed Consent: Since patient data were obtained retrospectively, patient consent was not obtained.

Peer Review: Externally peer-reviewed.

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