ChiKD CHILDHOOD KIDNEY DISEASES

Review Article

Child Kidney Dis 2024;28(3):99-105
pISSN 2384-0242 · eISSN 2384-0250
https://doi.org/10.3339/ckd.24.013

Congenital solitary functioning kidney: evaluations to do which, when, and how

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Congenital solitary functioning kidney (CSFK) is characterized by an anatomical or functional absence of one kidney from birth. When suspected on perinatal ultrasonography (US), repeat US after birth should be performed for confirmation. Although postnatal ^{99m}Tc-dimercaptosuccinic acid scintigraphy (DMSA scan) is the gold standard for confirming CSFK, it carries the risk of radiation exposure; US alone is sufficient when performed by an experienced radiologist. One-third of patients with CSFK have additional congenital anomalies of the kidney and urinary tract at the solitary functioning kidney, the most common of which is vesicoureteral reflux. As evidence regarding vesicoureteral reflux with normal kidney US is correlated with significant urinary tract infection is lacking, voiding cystourethrogram may be considered in patients with CSFK with abnormal US findings. Furthermore, approximately 30% of patients with CSFK have extrarenal malformations. Moreover, up to 10% of them have syndromic features. In particular, examining for female genitalia malformations, which can have potential for complications from untreated obstructive malformations, is important. In conclusion, DMSA scan and voiding cystourethrogram are not necessary for all patients with CSFK, and the risk of each patient should be assessed to determine which test is needed during follow-up. The presence of extrarenal manifestations should also always be considered.

Keywords: Evaluation study; Multicystic dysplastic kidney; Solitary kidney

Introduction

Congenital solitary functioning kidney (CSFK) is characterized by an anatomical or functional absence of one kidney from birth. It includes unilateral renal agenesis (URA), aplasia, and multicystic dysplastic kidney (MCDK). Renal agenesis is characterized by the absence of one kidney, detected via ultrasonography (US) performed at 18–22 gestational weeks, and confirmed postnatally. Aplasia refers to the presence of a rudimentary kidney, observed during the same gestational period, and has a relative function below 5%. MCDK manifests as multiple non-communicating cysts of various sizes within a lobulated kidney contour, pelvis, and parenchyma.

The prevalence of CSFK is approximately 1 in 1,500 newborns [1-3]. Two systematic reviews analyzing large numbers of patients revealed that the prevalence of URA and MCDK is 1 in 2,000 and 1 in 4,300 newborns, respectively [2,3]. Between 6% and 60% of the patients with CSFK have decreased kidney function by the age of 15 [4-7]. However, the possibility of impaired kidney function in CSFK remains debated because of the limited number of studies. Furthermore, CSFK frequently has additional malformations of the kidney, urinary tract, and ex-

Received: July 3, 2024; Revised: August 10, 2024; Accepted: August 19, 2024

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trarenal organs. However, as to which, when, and how to test for CSFK remain unclear.

This review aimed to describe the publications and recent guidelines, with a focus on optimal evaluation methods for CSFK.

When CSFK is suspected prenatally, should it be reevaluated after birth?

CSFK should be confirmed using neonatal US after birth when suspected prenatally because prenatal US may not detect ectopic kidney, which may be mistaken for a solitary kidney, or severe hydronephrosis (HN), which may be misdiagnosed as MCDK [8]. Additionally, one-third of patients with CSFK may have extrarenal malformations [2-3,7,9-11]. Therefore, even if a child is diagnosed with CSFK on prenatal US, performing repeat US in the neonatal period to examine for kidney, urinary tract, and other organ malformations is important.

Is US alone sufficient for diagnosing CSFK after birth? If not, should kidney DMSA scan be selected in all cases?

As an ectopic kidney may be missed on US, postnatal ^{99m}Tc-dimercaptosuccinic acid scintigraphy (DMSA) scan is the gold standard for confirming that one kidney is nonfunctional. However, DMSA scan poses a risk of radiation exposure compared with US. Postnatal US may be sufficient for diagnosing CSFK when it is performed by an experienced pediatric radiologist [8,12,13]. Postnatal US should describe the kidney length, echogenicity, parenchymal thickness, features of the calyces, and anteroposterior diameter of the kidney pelvis [14]. If possible, it should also include the maximal ureter diameter, bladder wall thickeness, and both pre- and post-void bladder volumes [14]. However, if US cannot accurately confirm CSFK, for example, cannot differentiate between MCDK and severe HN or cannot rule out an ectopic kidney, then further evaluation including DMSA scan or magnetic resonance imaging (MRI) is suggested.

After confirming CSFK after birth, what further examinations are required?

One-third of patients with CSFK have additional congenital anomalies of the kidney and urinary tract (CAKUT) at the solitary functioning kidney [1,2]. Therefore, additional anomalies in the contralateral kidney of MCDK or solitary kidney of renal agenesis should be excluded.

How can we examine if the solitary functioning kidney is normal?

CSFK without additional CAKUT has compensatory growth. It can be detected as early as 20 weeks of gestational age [15] and is frequently established in the first year of life or beyond in some cases [16,17]. Whether the compensatory growth is caused by the hypertrophy of pre-existing nephrons or hyperplasia that increases the number of nephrons remains debated [9]. Once CSFK has undergone compensatory enlargement, it does not subsequently regress in length and size [16].

Compensatory growth is assessed by measuring the kidney length using US compared with normative data. The compensatory growth of CSFK in children is defined as a kidney length for age of ≥2 or 2.5 standard deviation or a kidney length for height of ≥95th percentile [9,12,16,18], whereas in adults, the expected value of compensatory growth is >120 mm of the kidney length [19,20]. As physical growth varies even at the same age, using the kidney length for height rather than for age is frequently considered more appropriate. We can use the nomograms of kidney normal length or web-based tool [21,22]. If the kidney shows abnormalities in morphology or position including HN, duplex collecting system, or ectopic kidney, length measurement may not reflect parenchymal enlargement. In those cases, other methods for parenchymal area measurement should be considered.

The rate of kidney growth is most rapid during the first 2 years of life. It subsequently slows down between 2 and 5 years and only 2–3 mm per year throughout adolescence [23]. If insufficient compensatory enlargement is observed at 1 year old, we can wait until 2 years old for compensatory growth. The 2022 CSFK guidelines in Italy [14] have stated that kidney length with sufficient compensation is above the 50th percentile for age until 2 years old and above the 95th percentile for age after 2 years old.

The prevalence and types of additional CAKUT in CSFK

Among the CSFK-associated urologic abnormalities, vesicoureteral reflux (VUR) is the most common [2,3,9,16,24]. Previous studies on the prevalence of CSFK-associated CAKUT are sum-

https://doi.org/10.3339/ckd.24.013

marized in Table 1 [14]. Among these studies, two systematic reviews analyzing large populations of patients with MCDK or URA were included [2,3]. In 2009, Schreuder et al. [2] conducted a meta-analysis of 67 cohort studies with more than 3,500 patients with MCDK. One-third of the patients with MCDK had a contralateral kidney anomaly, with VUR being the most common anomaly and accounting for 20% of the cases. Of the patients with VUR, 40% had high grades of over 3. Conversely, a meta-analysis of 43 studies with 2,684 patients with URA conducted by Westland et al. in 2013 [3] showed anomalies at the solitary kidney in approximately 30% of the patients, which is consistent with the results of previous meta-analyses on MCDK. VUR, accounting for 24%, was the most common urologic anomaly. However, the VUR grade was not analyzed in this study.

Is routinely performing VCUG for detecting VUR necessary?

US has a low predictive value in low-grade VUR detection. This leads us to consider performing voiding cystourethrogram (VCUG) in patients with CSFK; however, whether it should be performed in all patients with CSFK is debatable.

We examine for VUR not because we are concerned about the presence of VUR itself but because we are concerned about the damage to a solitary functioning kidney caused by urinary tract infection (UTI). If the evidence is clear that low-grade VUR is associated with a lower frequency of UTI and a lower likelihood of kidney scarring, and US is a fairly good indicator of high-grade VUR, we may only perform VCUG in patients with abnormal US findings.

Some researchers have reported evidence of this. One study analyzed 156 patients with unilateral MCDK [24] and observed that only 1.7% of the patients with normal contralateral kidneys on US had high-grade VUR (grades 4–5), compared with 13.2% of those with abnormal US findings. This suggested that children with MCDK who have normal contralateral kidneys on US have a low incidence of clinically significant high-grade VUR. Moreover, they described the significant association between abnormal CSFK on US, including HN, duplex configuration, ureterocele, hydroureter, or uroepithelial thickening, and severe VUR. However, the only factor that was statistically significant in the comparison of patients with and without UTI was female gender, not VUR or abnormal US findings. Another study [25] described VUR in 77 children with MCDK examined using VCUG. The study showed no significant association between severe VUR and UTI and concluded that VCUG did not lead to changes in the management of the patients (e.g., antibiotic prophylaxis) before and after acquiring knowledge about the presence of VUR. However, due to the limited number of available studies and enrolled patients, the decision to perform VCUG is left to the opinion of the clinician.

How do we decide the type and timing of laboratory tests?

To date, several recommendations on the type and timing of laboratory tests for patients with CSFK are available [9,14,27-30]. These papers do not recommend routine blood tests when CSFK has good compensatory growth; however, they have rec-

Table 1. Reported prevalence of additional CAKUT in children with CSFK

Author (year)	No. of pts	CSFK type[a]	Associated CAKUT (%)	Total VUR (%)	VUR grades 3-4 (%)	UPJO (%)	UVJO (%)
Schreuder et al. (2009) [2][b]	3,557	MCDK (100)	31.3 (of 2,415 pts)	15 (of 2,104 pts)	8	4.8 (of 2,159 pts)	NR
Westland et al. (2013) [3][b]	1,093	URA (100)	32	24 (of 770 pts)	NR	6 (of 615 pts)	7 (of 605 pts)
Ross et al. (2015) [26]	138	MCDK (63), URA (37)	14.6	36	17	NR	NR
La Scola et al. (2016) [9]	146	MCDK (38), URA (29), URAP (16), undefined (18)	NR	11.5	10	2	3
Marzuillo et al. (2017) [7]	322	MCDK (48), URA (52)	33	9.3	5.6	0.3	4
Brown et al. (2019) [25]	165	MCDK (100)	NR	17 (of 77 pts)	NR	NR	NR
Blachman-Braun et al. (2020) [24]	156	MCDK (100)	NR	16	6	NR	NR

CAKUT, congenital anomalies of the kidney and urinary tract; CSFK, congenital solitary functioning kidney; pts, patients; VUR, vesicoureteral reflux; UPJO, ureteropelvic junction obstruction; UVJO, ureterovesical junction obstruction; MCDK, multicystic dysplastic kidney; URA, unilateral renal agenesis; URAP, unilateral renal aplasia; NR, not reported.
[a]The numbers in parentheses are percent values. [b]These are meta-analyses.
Adapted from La Scola et al. Pediatr Nephrol 2022;37:2185-207 [14].

ommended urinalysis to examine for proteinuria at diagnosis. However, if the patient with CSFK has abnormal US findings or additional indications including obstructive uropathy, UTI, preterm birth, or low birth weight, they have recommended evaluating serum creatinine and quantifying proteinuria. This is because those with no additional CAKUT and sufficient compensatory growth were less likely to have high blood pressure, proteinuria, or decreased kidney function [9,27,28]. The published recommendations are summarized in Fig. 1.

What are the methods for detecting extrarenal malformations?

Literatures have suggested that approximately 30% of patients with CSFK have extrarenal malformations [2,3,7,9,10]. Extrarenal malformations can develop in the heart, gastrointestinal tract, musculoskeletal system, and genital tract. Regarding additional CAKUT in the contralateral kidney, the studies by Schreuder et al. [2] and Westland et al. [3] included the largest number of patients with CSFK and reported that approximately 15% and 31% of the patients with MCDK and URA have extrarenal manifestations, respectively, and up to 10% have syndromic features. The study by Schreuder also observed that up to 11% of female patients with URA have genital malformations, including obstructive hemivagina, uterine didelphys, and Müllerian duct aplasia-hypoplasia.

The most reported syndromes are described in Table 2. Among these syndromes, Herlyn–Werner–Wunderlich (HWW) or obstructive hemivagina and ipsilateral renal agenesis (OHVIRA) and Müllerian duct aplasia–renal agenesis-cervicothoracic somite dysplasia (MURCS) have CSFK with female genital malformations and autosomal dominant inheritance.

Why is evaluation of female genital malformations significant?

Genital malformations are more common in female patients with CSFK; however, they may be presented in ipsilateral sem-

Author (year)	No additional CAKUT		Additional CAKUT	eGFR < 60 mL/min/1.73 m^2 or PU/HTN medications
	Compensatory growth	No compensatory growth		
Corbani et al. (2011) [30]	BP: every 2 yr until the age of 14, then every 3–5 yr thereafter Urinalysis: every 2 yr until the age of 14, then every 3–5 yr thereafter SCr and eGFR: every 2 yr until the age of 14, then every 3–5 yr thereafter		BP: yearly Urinalysis: yearly SCr and eGFR: yearly	BP: yearly Urinalysis: yearly SCr and eGFR: yearly
Westland et al. (2014) [29]	BP: yearly Microalbuminuria: yearly SCr and eGFR: every 5 yr US: every 5 yr		BP: 2 times/yr Microalbuminuria: 2 times/yr SCr and eGFR: every 5 yr US: depends on the type and severity of CAKUT	BP: 2–4 times/yr Microalbuminuria: 2–4 times/yr SCr and eGFR: 2–4 times/yr US: depends on the type and severity of CAKUT
	· Last US to be performed at 15–16 yr			
La Scola et al. (2016) [9]	Urinalysis: yearly SCr and eGFR: at diagnosis, then at ages 1, 5, 10, 15 thereafter US: yearly until age 3, then at increasing intervals	Urinalysis: yearly SCr and eGFR: yearly US: yearly	Follow-up schedule depends on the type and severity of CAKUT	-
	· Routine examination for proteinuria is by urinalysis. If the patient shows proteinuria by urinalysis, urine protein per creatinine ratio should be calculated.			
Groen In 't Woud et al. (2021) [28]	BP: yearly Urinalysis: yearly SCr and eGFR: at ages 5 and 10 yr, then every 2 yr thereafter US: at the age of 1, then every 5 yr thereafter		SCr and eGFR: within 1–2 wk of age Then, follow-up by a pediatric nephrologist	Follow-up by a pediatric nephrologist
La Scola et al. (2022) [14]	At diagnosis, urinalysis		At diagnosis, quantitative PU, SCr	-

Fig. 1. Recommended follow-up schedule for patients with congenital solitary functioning kidney in recent publications [9,14,28-30]. CAKUT, congenital anomalies of the kidney and urinary tract; eGFR, estimated glomerular filtration rate; PU, proteinuria; HTN, hypertension; BP, blood pressure; SCr, serum creatinine; US, ultrasonography.

https://doi.org/10.3339/ckd.24.013

Table 2. Reported congenital solitary functioning kidney-associated syndromes

Syndrome	Extrarenal manifestations	Genes	Possible inheritance
Branchio-oto-renal	Sensorineural hearing loss, preauricular pits, branchial cysts, and microtia	*EYA1, SIX1, SIX5*	Autosomal dominant
DiGeorge	Congenital heart disease, hypocalcemia, immunodeficiency, and neurocognitive disorders	22q11 deletion	Autosomal dominant
Fraser	Cryptophthalmos, cutaneous syndactyly, occasional malformations of the larynx, ambiguous genitalia, and mental retardation	*FRAS1, FREM2*	Autosomal recessive
Herlyn–Werner–Wunderlich or OHVIRA	Obstructed hemivagina and uterus didelphys	Unknown	Autosomal dominant
Kallmann 1	Micropenis, bilateral cryptorchidism, and anosmia	*KAL1*	X-linked
Klinefelter	Small, firm testis, gynecomastia, azoospermia, and hypergonadotropic hypogonadism	47, XXY	Sporadic
MURCS	Müllerian anomaly, renal agenesis, cervicothoracic somite dysplasia	Unknown	Autosomal dominant
Renal coloboma	Retinal and optic nerve coloboma	*PAX2*	Autosomal dominant
Renal cysts and diabetes	Maturity-onset diabetes of the young type 5, hyperuricemia, hypomagnesemia, and uterine malformations	*HNF1B*	Autosomal dominant
Townes–Brocks	Thumb anomalies, imperforate anus, and sensorineural hearing loss	*SALL1*	Autosomal dominant
VACTERL association	Vertebral anomalies, anorectal malformations, cardiovascular disease, tracheoesophageal fistula, esophageal atresia, and limb defects	*TRAP1*	Autosomal recessive
Williams–Beuren	Developmental delay, cardiovascular anomalies, mental retardation, and facial dysmorphology	7q11.23 deletion	Autosomal dominant

OHVIRA, obstructive hemivagina and ipsilateral renal agenesis; MURCS, Müllerian duct aplasia–renal agenesis-cervicothoracic somite dysplasia; VACTERL, vertebra-anus-cardiac-tracheoesophageal-renal-limb.
Adapted from La Scola et al. Pediatr Nephrol 2022;37:2185-207 [14].

inal vesicle hypoplasia, absence of the vas deferens in males occasionally. Uterine vaginal agenesis, uterine duplicity, obstructive or blind hemivagina, monoliteral ovarian agenesis, and Gartner duct pseudocyst are the types of anomalies. Moreover, genital anomalies are frequently associated with various syndromes, including HWW/OHVIRA and MURCS syndrome.

As previously described, up to 30% of patients with URA are associated with Müllerian anomalies. Conversely, more than 50% of patients with Müllerian anomalies and 92% to 100% of patients with obstructive hemivagina have kidney anomalies [31-33]. Uterine anomalies occur in one in over 500 females, 43% of whom have URA [34]. Although the kidney anomalies that are most commonly associated with Müllerian anomalies are URA and MCDK, unilateral or bilateral pelvic kidney, horseshoe kidney or kidney crossed ectopia, pyelocaliceal duplicity, ectopic ureter, and bilateral pyelectasis have also been reported [35].

Screening young women with URA and MCDK for Müllerian anomalies is significant owing to the potential for complications from untreated obstructive malformations [31,36-38]. Obstructive anomalies of the female genital tract can lead to retrograde menstruation, which develops significant problems including endometriosis, pelvic inflammatory disease, and infertility. However, prenatal US is unreliable for genital malformation screening, and several patients with obstructive genital anomalies can be asymptomatic during childhood. Therefore, when URA is identified in antenatal periods, physicians must explain to parents the significance of proper screening before menarche. To exclude genital abnormalities between thelarche and menarche, abdominopelvic US must be performed in all females with CSFK. Pelvic MRI is the gold standard of assessment; however, pelvic three-dimensional US may be an alternative for MRI [39]. Genital examination should include checking for a vaginal bulge or lower abdominal mass, indicating mucocolpos. Drainage of hematocolpos and vaginal septum excision comprise the treatments of obstructive genital malformations [37].

Conclusion

An optimized clinical approach is important for the management of patients with CSFK. Unilateral CSFK carries the potential for hypertension, proteinuria, and progression to chronic

kidney disease. However, in those without additional CAKUT and with adequate compensatory growth, routine blood testing is not recommended. Routine DMSA and VCUG are not necessary in all patients with CSFK. In addition, many patients with CSFK are accompanied by extrarenal malformations, among which the Müllerian anomalies can cause infertility, which should always be kept in mind.

Conflicts of interest

Hyun Kyung Lee is an editorial board member of the journal but was not involved in the peer reviewer selection, evaluation, or decision process of this article. No other potential conflicts of interest relevant to this article were reported.

Funding

None.

Author contributions

All the work was done by HKL.

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