Clinical features of solitary functioning kidney

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Solitary functioning kidney (SFK) indicates having either 1 anatomical kidney or 2 kidneys of which 1 does not function. SFK may be an isolated congenital malformation or it may be associated with a chromosomal syndrome [1] or with various nonchromosomal anomalies such as genital [2], gastrointestinal [3], and urological [4] abnormalities. However, there are few reports about the incidence of any anomalies associated with SFK. Herein, we enrolled 14 children hospitalized at our institution between 2000 and 2017 (Table 1). There were 6 boys and 8 girls whose ages at diagnosis ranged from at birth to 11 years. The follow-up duration ranged from 1 to 17 years. Multicystic dysplastic kidney (MCDK) as indicated by prenatal ultrasonography was observed in 4 patients (cases 1–4, right: 2, left: 2 cases). In case with MCDK, opposite kidneys often have genital or urological abnormalities, however, there were no abnormalities in 4 MCDK cases. In case 1, the MCDK spontaneously involuted at the age of 2 years. In other cases, the right and left kidneys were absent in 4 and 6 patients, respectively. The SFK diagnosis was based on high fever, failure to thrive, hearing loss, urinary tract infection, abdominal

pain, school urinary screening, and urinary frequency. Chromosomal anomalies were observed in 2 cases (case 7: William syndrome and case 8: Branchio-oto-renal syndrome). Hearing loss, gastrointestinal and genital disorders were each detected in 2 (cases 8 and 11), 3 (cases 9, 10, and 11), and 2 (cases 13 and 14) cases, respectively. The disorders of the gastrointestinal system were inguinal hernia (case 9) and intractable constipation (cases 10 and 11). Constipation plays a major role in the function and dysfunction of the urinary tract. However, the relationship between constipation and SFK remains unclear. The genital disorders observed were Herlyn–Werner–Wunderlich syndrome (case 13) and uterus bipartitus as revealed by MRI after 13 years (case 14). In this report, renal function was normal in any case for the duration of follow-up. SFK is apparently associated with little or no renal damage in infancy or childhood [5]. However, disorders of other organ systems are found frequently; therefore, SFK patients should be further examined for other organ disorders. In female patients, a work-up for uterine and vaginal abnormalities should be performed.

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Table 1 Associated anomalies in patients with SFK

Case. Age/sex	Background of diagnosis	Absent kidney	Other organs with disorders	Type of anomaly	Follow-up (years)	
1. After birth/F	Prenatal US	MCDK (L)	Genital/Urological	None	4	L: MCDK spontaneous involution at 2 years of age
2. After birth/F	Prenatal US	MCDK (R)	Genital/Urological	None	1	
3. After birth/F	Prenatal US	MCDK (L)	Genital/Urological	None	1	
4. After birth/F	Prenatal US	MCDK (R)	Genital/Urological	None	3	
5. After birth/M	Prenatal US	R	Urological	L kidney: Hydronephron	6	
6. 5 M/M	High fever, UTI	L	Urological	R kidney: Hydronephron	2	
7. 1 Y/M	Failure to thrive	L	Cardiac/Ophthalmological	SAS/esotropia	15	WS
8. 4 Y/F	Hearing loss	L	Ear	Malformations of the pinnae, Branchial arch remnants	8	BOR
9. 6 Y/M	UTI	L	Urological/Gastrointestinal/eye/mouth	R: VUR (IV) L: inguinal hernia/L: dyskinesia/L: corner palsy	6	
10. 9 Y/F	Abdominal pain, UTI	R	Gastrointestinal		11	Intractable constipation
11. 10 Y/M	Abdominal pain, hearing loss	R	Gastrointestinal/ear		17	Intractable constipation
12. 8 Y/M	School urinary screening	L			1	Chance hematuria
13. 11 Y/F	Urinary frequency	R	Genital	Bicornate uterus, R: obstructed hemivagina	4	HWWS
14. 5 M/F	High fever	L	Hematological	Neutrocytopenia	17	Uterus bipartitus as revealed by MRI at 13 years of age

M male, *F* female, *R* right, *L* left, *MCDK* multicystic dysplastic kidney, *US* ultrasound study, *UTI* urinary tract infection, *SAS* supravalvular aortic stenosis, *WS* William syndrome, *BOR* Branchio-oto-renal syndrome, *HWWS* Herlyn Werner-Wunderlich syndrome

Compliance with ethical standards

Conflict of interest All the authors have declared no competing interest.

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee at which the studies were conducted and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from all individual participants included in the study.

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