



INTRODUCTION AND OBJECTIVE

Congenital solitary functioning kidney (CSFK) is associated to other congenital anomalies. Most of them affect urogenital tract, cardiac, skeletal and central nervous system. There are also some syndromes associated with renal malformations. The objective of this study was to determine prevalence of associated malformations in children with CSFK.

METHODS

We reviewed electronic processes of 134 children with CFSK followed in a tertiary department of paediatric nephrology during five years (2012 – 2016). The congenital malformations found were grouped into system of organs.

RESULTS

	Total	Antenatal diagnosis		Cause	
		Yes	No	Agenesis	Other causes
CSFK	134	106	28	47	87
With at least one other malformation	41 (30,6%)	34 (33,0%)	7 (25,0%)	37 (78,7%)	26 (29,9%)

Urologic	Genital	Gastro-intestinal	Cardio-vascular	Neurological	Otorhinolaryngological	Muscle and skeleton	Ophthalmic
14	9	9	8	6	4	3	1
Vesicoureteral reflux (n=5)	Imperforated hymen (n=1)	Esophageal atresia (n= 3)	Tetralogy of Fallot (n=2)	Caudal regression syndrome (n=2)	Deafness (n=2)	Vertebral malformation (n=2)	Coloboma
Pelvic ureteric junction obstruction (n=3)	Uterine didelphys (n=3)	Anorectal malformation (n= 4)	Aortic stenosis (n=1)	Spina bifida (n=1)	Inner ear malformation (n=1)	Scoliosis (n=1)	
Ectopic ureter (n=1)	Vaginal septum (n=2)	Anorectal atresia (n=1)	Ventricular septal defect (n=1)	Neurogenic bladder (n=1)	Cleft palate (n=1)	Abdominal wall defect (n=1)	
Ureterocele (n=1)	Cryptorchidism (n=1)	Ectopic anus (n=1)	Single umbilical artery (n=5)	Dysmorphic cerebellum (n=1)		Limb malformation (n=1)	
Obstructive megaureter (n=2)	Hypospadias (n= 2)			Hydrocephalus (n=1)			
Ureteral stenosis (n=2)							

Identified syndromes (n=14)

VATER/VACTERL (n=2)	Manick Fraser
Herlyn-Werner-Wunderlich (n=2)	Barakat
Williams (n=2)	CHARGE
Mayer Rokitansky Kuster Hauser	Acrorenal mandibular
Prune Belly	Polymalformative syndrome without specific diagnosis
Turner	

All identified syndromes were associated with renal anomalies

Majority of cases with renal agenesis had another congenital malformation

Three cases with other congenital malformation had chronic renal disease:

- Polymalformative syndrome (1)
- Ureteral stenosis (1)
- Anorectal malformation (1)

DISCUSSION

This study intends to recall that CSFK may not be the only congenital malformation in a child. It also highlights that there are other anomalies, besides urogenital tract, that may be present and must be investigated, especially if there is a diagnosis of true renal agenesis. A good prenatal care and careful follow-up of children with CSFK are essential.