

CONGENITAL SOLITARY FUNCTIONING KIDNEY AND OTHER ASSOCIATED CONGENITAL MALFORMATIONS

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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.

RESULIS						
		Antenatal	diagnosis	Cause		
	Total	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	Otorhinola- ryngological	Muscle and skeleton	Ophthalmic
14	9	9	8	6	4	3	1
Vesicoureteral reflux (n=5)	Imperfurated 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 septus (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 syndro	omes (n=14)	All identified syndromes were associated with renal anomalies
VATER/VACTERL (n=2)	Manick Fraser	
Herlyn-Werner-Wunderlich (n=2)	Barakat	Majority of cases with renal agenesis had another congenital malformation
Williams (n=2)	CHARGE	Three cases with other congenital
Mayer Rokitansky Kuster Hauser	Acrorenal mandibular	malformation had chronic renal disease:
Prune Belly	Polymalformative	Polymalformative syndrome (1)
urner syndrome without specific diagnosis		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 urological 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.

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