

Suppl. Table 1. Genetic and clinical data of 2 families with SRNS and mutations in *KIRREL1*

Family-Individual	Nucleotide change	Amino acid change	Zygoty	Exon	PPH2	SIFT	MT	Amino acid conserved to species	gnomAD hom/het/all (allele frequency in %)	EVS hom/het/all (allele frequency in %)	Sex	Ethnic origin	PC	Age of onset at (yrs)	Extra renal manifestations	Biopsy (age)	Renal phenotype	Renal function	Therapy and response (Y/N)	Follow up (yrs)
<u>KIRREL1</u>																				
A666-21 [^]	c.1318C>T	p.Arg440Cys	Hom	11	0.999	Del	DC	<i>D. rerio</i>	0/1/251320 (0.0003979)	not reported	M	Arabic	Y	5	None	MCD (5)	SRNS	normal	CS (N) CP (N) CsA (N) MMF (N) ACE-I and ARB	12,5
B742-21	c.1718C>T	p.Ser573Leu	Hom	13	0.825	Del	DC	<i>D. rerio</i>	1/220/282054 (0.078)	0/12/6491 (0.0923)	M	Italian	Y	14	None	FSGS (14)	SRNS	CKD Stage 2	CS (N) Tac (partially Y) ACE-I and CCB	4

The following NCBI GenBank annotations were used: *KIRREL1* (NM_018240.6). [^]Younger sister had similar clinical phenotype, but no DNA sample could be obtained.

ACE-I, angiotensin-converting enzyme inhibitor; ARB, angiotensin II receptor blocker; CCB, calcium channel blocker; CKD, chronic kidney disease (eGFR calculated using CKD-EPI equation); CP, cyclophosphamide; CS, corticosteroid; CsA, cyclosporine A; DC, disease causing; Del, deleterious; *D. rerio*, *Danio rerio*; EVS, Exome Variant Server; FSGS, focal segmental glomerulosclerosis; gnomAD, Genome Aggregation Database (<http://gnomad.broadinstitute.org>); Hom, homozygous; M, male; MCD, minimal change disease; MMF, mycophenolate mofetil; MT, Mutation Taster (prediction software); N, no; PC, parental consanguinity; PPH2, PolyPhen-2 (prediction software); SIFT, Sorting Intolerant From Tolerant (prediction software); SRNS, steroid-resistant nephrotic syndrome; Tac, tacrolimus; Y, yes; yrs, years.