Online Supplement

Table S-1: Genetic diagnoses in 212 children with hereditary SRNS.

Causative gene	**N (%)**
NPHS2	103 (48.6)
WT1	43 (20.3)
SMARCAL1	14 (6.6)
PLCE1	11 (5.2)
INF2	7 (3.3)
ADCK4	6 (2.8)
LMX1B	5 (2.4)
NPHS1	4 (1.9)
MYO1E	4 (1.9)
LAMB2	3 (1.4)
COQ6	2 (0.9)
PTPRO	2 (0.9)
COL4A5	2 (0.9)
COQ2	1 (0.5)
TRPC6	1 (0.5)
COL4A3	1 (0.5)
CLCN5	1 (0.5)
$tRNA^{LEU}$	1 (0.5)
duplication at chromosome 6p	1 (0.5)
All	212

Table S-2: Characteristics of patients with hereditary SRNS and reported responsiveness to intensified immunosuppression (IIS).

Gene	Mutation	Age at disease onset (yrs)	Serum albumin at disease onset (g/l)	Histo-pathology	First year response to IIS	Immuno-suppression (± oral Pred.)	RAS Antagonist	Follow-up (yrs)	Remission status at last observation	Status at last observation
WT1	c.1351T>C p.(Phe451Leu) previously referred as p.F383L	3.2	32.0	FSGS	Complete rem.	CSA, Pred.pulses	-	12.0	No rem.	CKD 2
NPHS2	c.[365G>C];[851C>T] p.[(Trp122Ser)];[(Ala284Val)]	5.4	28.0	MCGN	Complete rem.	CSA	Enalapril, Losartan	4.8	No rem.	Peritoneal dialysis
NPHS2	c.[868G>A];[868G>A] p.[(Val290Met)];[(Val290Met)]	7.9	30.0	MCGN	Partial rem.	CSA	Enalapril	1.5	Partial rem.	CKD 1
NPHS2	c.[249del];[538G>A] p.[(Leu84Trpfs*15)];[(Val180Met)]	3.9	37.0	MesPGN	Partial rem.	CSA	-	2.5	Partial rem.	CKD 1
NPHS2	c.[419delG];[506T>C] p.[(Gly140Aspfs*41)];[(Leu169Pro)]	1.0		FSGS	Partial rem.	CSA, Pred.pulses	-	0.2	Partial rem.	-
NPHS2	c.[353C>T];[(353C>T)] p.[Pro118Leu(;)Pro118Leu]	1.3	29.0	FSGS	Partial rem.	Pred.pulses	Ramipril	4.3	No rem.	CKD 1
WT1	c.1432+4C>T previously referred as IVS9+4C>T	8.0	29.0	FSGS	Partial rem.	CSA, Pred.pulses	Enalapril	5.6	No rem.	CKD 3
WT1	c.1372C>T p.(Arg458*) previously referred as p.390X	15.3	25.0	FSGS	Partial rem.	CSA	-	2.7	No rem.	CKD 4
COQ6	c.[1058C>T];[(1058C>T)] p. [Ala353Asp(;)Ala353Asp]	7.0	28.0	unknown	Partial rem.	CSA	-	3.1	No rem.	CKD 5
WT1	c.1432+5G>A previously referred as IVS9+5G>A	8.1	37.0	FSGS	Partial rem.	CSA	Enalapril	8.6	No rem.	CKD 5

The reference transcripts for mutation description are: *COQ6* - NM_182480.2; *NPHS2* - NM_014625.2; *WT1* - NM_024426.4;

Figure S-1: **Renal survival by response to intensified immunosuppression (IIS), irrespective of genetic findings and familial disease occurrence.** Patient group with unknown treatment response is included.

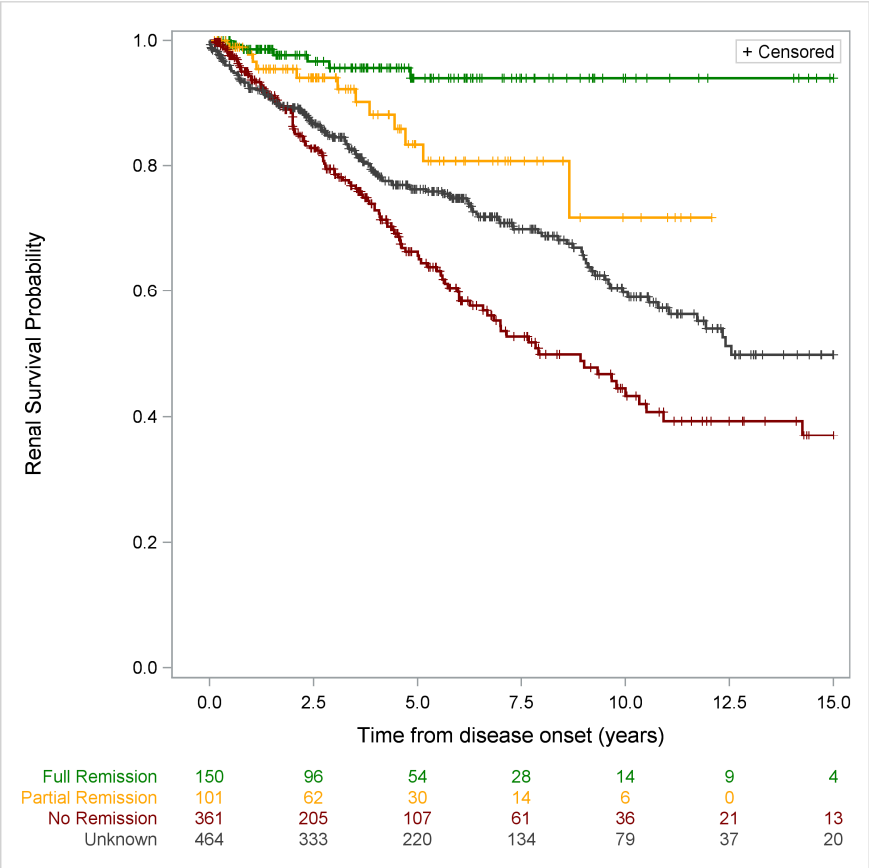

Figure S-2: Renal survival by initial treatment response, genetic findings and familial disease occurrence. Patient groups with sporadic disease occurrence and unknown treatment response are included.

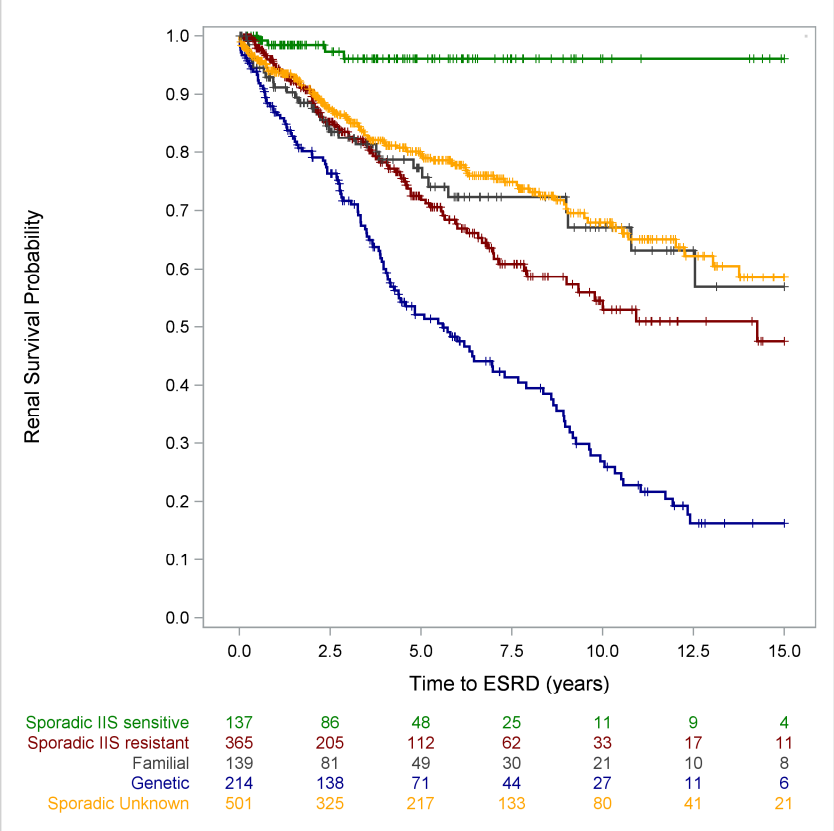

Figure S-3: Renal survival by major genetic entities in 212 children with hereditary SRNS.

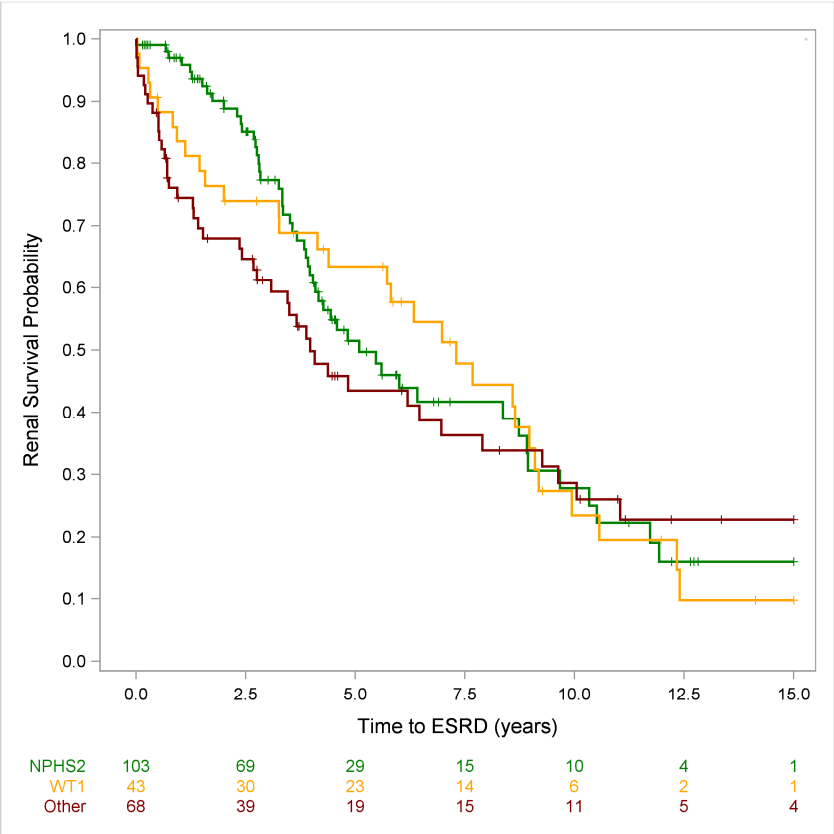

Figure S-4: Renal survival by underlying histopathological diagnoses in 1155 children with performed kidney biopsy.