

<b>Patient number</b>	1 (IV:1)	2 (IV:8)	3 (V:9)	4	5	6	7	8	9	10	11	12	13	14
<b>Sex</b>	M	F	M	F	M	F	M	M	M	F	F	M	M	M
<b>Age at presentation (years)</b>	40	21	16 months	22	7	30	23	28 months	6	2	23	10 months	8 months	3
<b>C3 (mg/L)</b>	570	550	380	479	380	613	483	510	416	555	540	350	570	611
<b>FH (g/l)</b>	0.70	0.55	0.50	0.50	0.56	0.56	0.64	0.60	0.58	0.49	0.67	0.64	0.61	0.42
<b>FI (mg/l)</b>	83	61	65	89	45	70	65	56.4	88	64	61	49	53	64
<b>Nucleotide change</b>	1774 C>T	1774 C>T	1774 C>T	1775 G>A	1775 G>A	1775 G>A	2203 C>T	2203 C>T	2562 C>G	3281 C>T	3343 G>A	3474 C>G	3481 C>A	4390 C>G
<b>Exon</b>	14	14	14	14	14	14	17	17	20	26	26	27	27	37
<b>Amino acid change</b>	R570W	R570W	R570W	R570Q	R570Q	R570Q	R713W	R713W	Y832X	A1072V	D1093N	C1136W	Q1139K	H1442D
<b>Sporadic/Familial</b>	F	F	F	S	F	F	F	S	S	S	S	S	S	F
<b>Follow-up (y)</b>	13	29	12	3	32	20	30	16	4	13	19	2	4	10
<b>Renal outcome</b>	ESRD	ESRD	ESRD	ESRD	ESRD	ESRD	Recovered	ESRD	Recovered	Recovered	ESRD	CKD	Recovered	Recovered
<b>Number of transplants</b>	2	1	2	0	3	2	0	0	0	0	2	0	0	0
<b>Number of transplants affected by recurrent HUS</b>	2	0	2	0	0	1	0	0	0	0	0	0	0	0
<b>Healthy carriers</b>	n/a	n/a	Father (IV:9)	Mother	n/a	n/a	n/a	n/a	Mother	Father	n/a	Mother	n/a	Mother

Abbreviations. ESRD, end stage renal disease; CKD, chronic kidney disease not requiring dialysis.

Normal ranges: C3 680-1380 mg/l, FH 0.34-0.68 g/L, FI 42 –78 mg/L.

The figures in parentheses after patient numbers 1, 2, and 3 refer to the pedigree shown in Fig. S1.

n/a, not available