SUPPLEMENTARY TABLE 1. Complement profiles in members of families PER and BRA.

		Status	C3[1,2] (77-210mg/dL)	C4[2] (14-47mg/dL)	Factor H[3] (12-56mg/dL)	Factor B[3] (7.5-28mg/dL)	Factor I[3] (75-115 %)	MCP[4] (91-109%)
PER	I-2	Healthy	68	24	38	15	n.a.	n.a.
	II-5	Healthy	75	25	37	16	n.a.	n.a.
	II-7 (H55P)	Affected	46	29	36	17	118	100
	II-10	Healthy	122	28	35	22	n.a.	121
	II-11	Healthy	123	28	38	26	n.a.	n.a.
	III-7 (H55)	Affected	91	36	32	16	104	110
	III-12	Healthy	67	16	42	13	n.a.	n.a.
	III-13 (H 85)	Affected	60	19	23	12	98	118
	III-14	Healthy	117	21	33	12	n.a.	98
	IV-1	Healthy	134	17	27	15	n.a.	n.a.
	IV-2 (H 112)	Affected	65	25	24	22	95	66
BRA	I-1	Healthy	102	28	38	11	84	n.a.
	I-2	Healthy	89	28	28	12	85	n.a.
	II-1 (H21)	Affected	23	32	26	21	103	84

1) Normal range of variation in controls for each variable is shown between brackets.
2) C3 and C4 were determined by nephelometry.
3) Factor H, Factor B and Factor I plasma levels were determined by ELISA. Factor I levels are referred to a reference serum; each value corresponds to the mean of three independent determinations.
4) Levels of MCP in PBLs (MFI, mean fluorescence intensity) were determined by flow cytometry and referred to a series of control samples drawn and analysed the same day. The average MFI value for the control samples was set to 100%. Range of variation for all control samples (n=17) was within 10% of the average value (91 to 109%).
5) Pedigree codes as in Figure 2. *BF* mutation carriers are labelled with black triangles. HUS patients are identified by their code in parenthesis.
6) n.a.: not available.