

Supplementary Table 1. Restriction fragment length polymorphism procedures for C3 and CFH mutation analysis.

Gene name	Target mutation	Primer sequence	Restriction enzyme	PCR product size (bp)
C3	p.R425C	F: 5'- CAA TTC CCA GGT CTC AGG GA - 3' R: 5'- GAG AGA AAA GGA GAA AGG G - 3'	<i>Hha I</i>	Wild type : 493+250 Mutated : 493+170+80
C3	p.S562L	F: 5'- CAA TTC CCA GGT CTC AGG GA - 3' R: 5'- GAG AGA AAA GGA GAA AGG G - 3'	<i>Ban II</i>	Wild type : 229+175+102 Mutated : 229+155+102
CFH	p.Y355S	F: 5'- GCA AGT GAA ACC TTG TGA TTA TCC -3' R: 5'- GGT GTA ATC ACT TAT GTG CTC TCC -3'	<i>BseR I</i>	Wild type : 281 Mutated : 168+113
CFH	p.C926F	F: 5'- TGT TCA CAA CCA CCT CAG A -3 R: 5'- TGC CAG AAT ACA AAG TGA CTC -3'	<i>Ale I</i>	Wild type : 382 Mutated : 217+165
CFH	p.C1077W	F: 5'- AAT GAA CAC TAG GTG GAA CCA -3' R: 5'- TCT ACC TTT GCA TTG AGG TGG -3'	<i>BseR I</i>	Wild type : 223 Mutated : 152+71
CFH	p.Q1139X	F: 5'- CTG TGT TTG CGT TTG CCT TA -3' R: 5'- CAC CCA GCC CTA AAG AGA AA -3'	<i>Bfa I</i>	Wild type : 437 Mutated : 251+186
CFH	p.R1215G	F: 5'- ATC CGT GTG TAA TAT CCC GAG A - 3' R: 5'- GCA CAA GTT GGA ATC TCC AGT - 3'	<i>BsrD I</i>	Wild type : 144+49 Mutated : 193

Abbreviation: C3, complement 3 CFH complement factor H.

Supplementary Table 2. Characteristics of aHUS patients with genetic abnormalities.

Gene abnormality	Age (yr)	Sex	ADAMTS13 activity (%)	Hgb (g/dL)	PLT ($\times 10^9/L$)	Cr. (mg/dL)	LDH (mg/dL)	Outcome
CFH c.3572C>T, p.Ser1191Leu	53	F	91	8.4	14	1.58	2,146	Alive
C3 c.1273C>T, p.Arg425Cys	23	F	91	10.6	98	0.68	602	Alive
C3 c.1273C>T, p.Arg425Cys	88	F	42	12.2	91	2	369	Dead at HD 11
C3 c.1273C>T, p.Arg425Cys	66	F	83	11.2	19	1.25	1,192	Alive
C3 c.1685C>T, p.Ser562Leu	64	F	61	8.2	7	1.4	3,769	Alive
Plasminogen c.1858G>A, p.Ala620Thr	56	F	67	9	52	4.1	1,332	Dead at HD 117
Plasminogen c.1858G>A, p.Ala620Thr	27	M	27	7.8	25	11.93	162	Recurrence
THBD c.1456G>T p.Asp486Tyr	31	M	31	7.8	92	19.5	2,445	Alive
CFB c.1598A>G p.Lys533Arg	33	F	94	5.1	10	2.64	2,302	Alive

Abbreviations: ADAMTS13, a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13; aHUS, atypical hemolytic uremic syndrome; C3, complement 3; CFB, complement factor B; CFH, complement factor H; Cr., serum creatinine; HD, hospital day; Hgb, hemoglobin; LDH, lactate dehydrogenase; PLT, platelet; THBD, thrombomodulin.

Supplementary Table 3. Correlation between the PLASMIC score and ADAMTS13 activity.

PLASMIC score	N (%)	ADAMTS13 activity		OR	95% CI	P
		$\leq 10\%$ (N=48)	$> 10\%$ (N=50)			
6 or 7	46 (47)	35	11	9.545	3.791-24.038	< 0.001
≤ 5	52 (53)	13	39			
		Value (%)		95% CI		
Sensitivity		73		0.579-0.843		
Specificity		78		0.637-0.880		
Positive predictive value		76		0.609-0.869		
Negative predictive value		75		0.608-0.855		

Abbreviations: ADAMTS13, a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13; CI, confidence interval; OR, odds ratio.