

Supplemental Table 1. Biomarker Assays

Assay	Method	Normal Range	Company/Reference
C3	Radial immunodiffusion	0.9~1.8 g/L	The Binding Site
C3a	Direct sandwich ELISA	<122 µg/L	Quidel Corp
C3c	Direct sandwich ELISA	2.12~4.92 mg/L	Palarasah, <i>et al</i> 2010
C3d	Direct sandwich ELISA	0.3~0.68 mg/L	ProGen Biologics
Factor B	Indirect sandwich ELISA	155~250 mg/L	Abcam
Ba	Direct sandwich ELISA	<0.9 mg/L	Quidel Corp
Bb	Direct sandwich ELISA	<1.5mg/L	Quidel Corp
Properdin	Indirect sandwich ELISA	10~33 mg/L	Hycult Biotech
C5	Indirect sandwich ELISA	55~125 mg/L	Abcam
C5a	Direct sandwich ELISA	0~16 µg/L	Quidel Corp
C6	Indirect sandwich ELISA	63~145 mg/L	Abcam
C7	Indirect sandwich ELISA	48~109 mg/L	Abcam
C8	Indirect sandwich ELISA	24~121 mg/L	Abcam
C9	Indirect sandwich ELISA	30~93 mg/L	Abcam
sC5b-9 ^a	Direct sandwich ELISA	<0.3 mg/L	Quidel Corp
C3CSA ^b ; C3CSAP ^c	Hemolytic assay	<20%	Zhang, <i>et al</i> 2012
IFE ^d	Electrophoresis	<7.5%	Zhang, <i>et al</i> 2012
FHAA ^e	Direct ELISA	<1:50+	Goodship, <i>et al</i> 2012

^a sC5b-9, soluble C5b-9; ^b C3CSA, C3 convertase stabilizing assay; ^c C3CSAP, C3 convertase stabilizing assay with properdin; ^d IFE, immunofixation electrophoresis; ^e FHAA, Factor H autoantibodies

Supplemental Table 2. Mutations and Autoantibodies in DDD Patients

DDD	Mutation ^a	FH Risk Allele		C3 Risk Allele		C3CSA (%) ^b	C3CSAP (%) ^c	IFE (%) ^d	FHAA ^e
1	-	p.Val62	0	p.Gly102	0	20.6	20.5	7.2	Negative
		p.His402	1	p.Leu314	0				
2	-	Not Done		Not Done		54.2	40.6	12.2	Negative
3	-	p.Val62	2	p.Gly102	1		53.3	37.2	Negative
		p.His402	2	p.Leu314	1				
4	-	p.Val62	2	p.Gly102	1	66.6	34.5	30.4	Negative
		p.His402	1	p.Leu314	1				
5	-	p.Val62	2	p.Gly102	0	66.4	38.4	29.1	Negative
		p.His402	2	p.Leu314	0				
6	-	p.Val62	2	p.Gly102	1	77.1	82.3	30.1	Negative
		p.His402	1	p.Leu314	1				
7	-	p.Val62	2	p.Gly102	0	40.0	38.3	17.5	Negative
		p.His402	1	p.Leu314	0				
8	-	Not Done		Not Done		16.0	19.4	3.0	Negative
9	<i>FI</i> p.R406H	p.Val62	1	p.Gly102	1	8.5	34.0	0.9	Negative
		p.His402	0	p.Leu314	0				
10	-	p.Val62	0	p.Gly102	1	22.6	32.8	13.0	Negative
		p.His402	0	p.Leu314	1				
11	-	p.Val62	2	p.Gly102	0	111.6	71.1	77.2	Negative
		p.His402	1	p.Leu314	0				
12	-	p.Val62	2	p.Gly102	1	5.3	24.7	18.4	Negative
		p.His402	2	p.Leu314	1				
13	-	p.Val62	2	p.Gly102	1	24.6	41.4	21.9	Negative
		p.His402	1	p.Leu314	1				
14	-	p.Val62	1	p.Gly102	0	26.0	27.6	10.3	Negative
		p.His402	1	p.Leu314	0				
15	<i>FH</i> p.V941F	Not Done		Not Done		98.9	117.4	79.7	Negative
16	<i>FH</i> p.V837I; p.E1145D	p.Val62	1	p.Gly102	0	23.6	63.0	5.9	Negative
		p.His402	0	p.Leu314	0				
17	-	p.Val62	2	p.Gly102	0	17.9	44.2	15.4	Negative
		p.His402	1	p.Leu314	0				

^a Genes screened: *FH*, *FI*, *FB*, *C3*, *MCP*; ^b C3CSA, C3 convertase stabilizing assay, normal reference value <20%; ^c C3CSAP, C3 convertase stabilizing assay with properdin, normal reference value <20%; ^d IFE, immunofixation electrophoresis, normal reference value <7.5%; ^e FHAA, Factor H autoantibodies

Supplemental Table 3. Mutations and Autoantibodies in C3GN Patients

C3GN	Mutation ^a	FH Risk Allele		C3 Risk Allele		C3CSA (%) ^b	C3CSAP (%) ^c	IFE (%) ^d	FHAA ^e
1	-	p.Val62	0	p.Gly102	1	16.3	18.9	8.4	Negative
		p.His402	1	p.Leu314	1				
2	-	p.Val62	2	p.Gly102	1	11.3	16.5	11.8	Negative
		p.His402	2	p.Leu314	1				
3	-	Not Done		Not Done		43.9	52.3	9.2	Negative
4	-	p.Val62	2	p.Gly102	1	12.4	19.8	13.2	Negative
		p.His402	2	p.Leu314	1				
5	Not Done	Not Done		Not Done		14.2	18.9	10.8	Negative
6	-	p.Val62	2	p.Gly102	0	20.4	20.1	4.3	Negative
		p.His402	0	p.Leu314	0				
7	<i>FH</i> p.R567G	p.Val62	2	p.Gly102	2	12.9	53.9	19.7	Negative
		p.His402	2	p.Leu314	2				
8	-	p.Val62	2	p.Gly102	0	13.3	20.6	4.5	Negative
		p.His402	1	p.Leu314	0				
9	-	p.Val62	2	Not Done		17.0	19.6	10.0	Negative
		p.His402	0						
10	-	p.Val62	2	p.Gly102	1	16.0	15.8	12.9	Negative
		p.His402	1	p.Leu314	0				
11	-	p.Val62	2	p.Gly102	0	19.4	16.1	3.2	Negative
		p.His402	2	p.Leu314	0				
12	-	p.Val62	2	p.Gly102	1	16.3	20.3	4.6	Negative
		p.His402	1	p.Leu314	1				
13	-	p.Val62	2	p.Gly102	1	16.3	18.5	7.1	Negative
		p.His402	2	p.Leu314	1				
14	-	p.Val62	1	p.Gly102	0	14.9	43.9	12.1	Negative
		p.His402	0	p.Leu314	0				
15	-	p.Val62	2	p.Gly102	0	20.8	44.6	4.8	Negative
		p.His402	2	p.Leu314	0				
16	-	p.Val62	2	p.Gly102	1	16.2	20.5	0	Negative
		p.His402	1	p.Leu314	1				
17	-	p.Val62	1	Not Done		24.9	29.9	3.5	Negative
		p.His402	1						

^a Genes screened: *FH*, *FI*, *FB*, *C3*, *MCP*; ^b C3CSA, C3 convertase stabilizing assay, normal reference value <20%; ^c C3CSAP, C3 convertase stabilizing assay with properdin, normal reference value <20%; ^d IFE, immunofixation electrophoresis, normal reference value <7.5%; ^e FHAA, Factor H autoantibodies