

### **Supplementary Table 1. IgA1 myeloma proteins used in this study**

<b>Myeloma protein</b>	<b>IgA isotype</b>	<b>Molecular form</b>
Mce	IgA1	Polymer
Mce1	IgA1	Polymer
Ale mono	IgA1	Monomer
Ale poly	IgA1	Polymer
Fab-IgA1	IgA1	Fab fragment of IgA1 (Ste) containing part of the hinge region

## Supplementary Table 2. Serum levels of antigen-specific IgG

Cohort from southeastern USA

Antigen	IgAN (n = 16)	Controls (n = 16)
dd-IgA1	2.256 ± 0.112*	1.995 ± 0.146
Fab-IgA1	2.136 ± 0.163*	1.724 ± 0.184

Cohort from Japan

Antigen	IgAN (n = 20)	Controls (n = 20)
Fab-IgA1	2.021 ± 0.202**	1.532 ± 0.229

Data are expressed as optical density at 490 nm and shown as mean ± SD

\*,  $P < 0.001$ , \*\*,  $P < 0.0001$

dd-IgA1, enzymatically desialylated and degalactosylated IgA1

Fab-IgA1, Fab fragment of Gd-IgA1 containing part of the hinge region with *O*-glycans

IgAN, patients with IgA nephropathy; Controls, healthy controls

**Supplementary Table 3. Primer sequences for mutagenesis**

#1123	Sense		Antisense	
PCR 1	Age1-VH3	ACTGCAACCGGTGTACATTCCGAGGTGCAGCTGGTGGAGTC	R-mutated	AGGGGCGACACACTTTCGCACAGTAATATATGGCCG
PCR 2	F-mutated	ATATATTACTGTGCGAAAGTGTGTCGCCCCCTGG	Sal1-JH3	CTGCGAAGTCGACGCTGAAGAGACGGTGACCATTG
Overlap PCR	Age1-VH3	ACTGCAACCGGTGTACATTCCGAGGTGCAGCTGGTGGAGTC	Sal1-JH3	CTGCGAAGTCGACGCTGAAGAGACGGTGACCATTG
#9017	Sense		Antisense	
PCR 1	Age1-VH3	ACTGCAACCGGTGTACATTCCGAGGTGCAGCTGGTGGAGTC	R-mutated	ATCATAGCGCTGGACTCTGGAACAGTAATACACAGCCGTG
PCR 2	F-mutated	TGTGTATTACTGTTCCAGAGTCCAGCGCTATGATAGCACTG	Sal1-JH145	CTGCGAAGTCGACGCTGAGGAGACGGTGACCAGGG
Overlap PCR	Age1-VH3	ACTGCAACCGGTGTACATTCCGAGGTGCAGCTGGTGGAGTC	Sal1-JH145	CTGCGAAGTCGACGCTGAGGAGACGGTGACCAGGG

**Supplementary Table 4. Clinical characteristics of study population**

	Cohort		Age	Male	Female	Race	serum IgG mg/ml	serum IgA mg/ml	SCr <sup>a</sup> mg/dl	UP/Cr <sup>b</sup>
IgAN	US	28	40.0 ± 15.2	W16, B1	W9, B2	W25, B3	12.3 ± 2.7	4.4 ± 2.4	1.4 ± 0.9	1.2 ± 1.5
	Japan	32	30.3 ± 7.0	12	20	-	12.1 ± 2.9	3.5 ± 1.1	1.2 ± 0.4	1.4 ± 1.7
	total	60	34.8 ± 12.5	29	31	W25, B3, J32	12.2 ± 2.8	3.9 ± 1.9	1.3 ± 0.6	1.3 ± 1.6
Disease controls	US	7	33.7 ± 16.4	W5	W1, B1	W6, B1	17.2 ± 6.3	3.4 ± 1.2	1.0 ± 0.3	1.5 ± 2.6
	Japan	13	35.7 ± 8.4	7	6	-	15.5 ± 3.5	3.8 ± 1.8	1.2 ± 0.4	1.6 ± 1.7
	total	20	35.0 ± 11.4	12	8	W6, B1, J20	16.1 ± 4.6	3.7 ± 1.6	1.1 ± 0.4	1.6 ± 1.9
Healthy controls	US	30	38.6 ± 17.9	W12, B2	W12, B4	W24, B6	10.4 ± 2.4	3.1 ± 1.7	1.0 ± 0.2	0.1 ± 0.1
	Japan	10	36.1 ± 10.0	4	6	-	10.9 ± 2.9	2.8 ± 1.5	0.8 ± 0.2	0.1 ± 0.0
	total	40	38.0 ± 16.2	18	22	W24, B6, J10	10.5 ± 2.5	3.0 ± 1.7	0.9 ± 0.2	0.1 ± 0.1

Data expressed as mean ± SD. <sup>a</sup>Serum creatinine concentration. <sup>b</sup>Urinary protein/creatinine ratio

W, white, B, African-American, J, Japanese