

Supplementary Methods

In the present study we observe seven haplotypes HYPA, LYPA, LYQA, LXPA, HYPD, LYPB and LYQC from the six SNPs H/L, X/Y, P/Q, A/D, A/B and A/C. We will refer to these six SNPs by their rare allele: H, X, Q, D, B and C, respectively. The latter three are positioned in *MBL2* exon 1 and never occurs in the same chromosome, i.e. these can be seen as one 4-allelic marker and explains why the haplotypes can be represented by 4 alleles. Formally these would be HYPAAA, LYPAAA, LYQAAA, LXPA, HYPDAA, LYPABA and LYQAAC. We will code genotypes and multilocus genotypes by the number of rare alleles, i.e. 0 (C/C), 1 (C/R) and 2 (R/R) where C and R denotes the common and rare allele, respectively. Obviously, each individual carry exactly two of these haplotypes: 0, 1 or 2 of each. Thus, with the 0/1/2 coding the sum of the seven possible haplotypes will be two for each individual. Clearly, the following set of seven equations is therefore true:

$$H = HYPA + HYPD = 2 - (LYPA + LYQA + LXPA + LYPB + LYQC) = 2 - L$$

$$X = LXPA$$

$$Q = LYQA + LYQC$$

$$D = HYPD$$

$$B = LYPB$$

$$C = LYQC$$

$$2 = HYPA + LYPA + LYQA + LXPA + HYPD + LYPB + LYQC$$

A direct calculation now gives that the model for multiple SNPs with a trend parameter for each of the six single markers, $\text{logit}(p) = \alpha_0 + \alpha_1 H + \alpha_2 X + \alpha_3 Q + \alpha_4 D + \alpha_5 B + \alpha_6 C$, can be re-parameterised to the model containing a trend parameter for each of the seven haplotypes, $\text{logit}(p) = \beta_0 + \beta_1 LYPA + \beta_2 LYQA + \beta_3 LXPA + \beta_4 HYPD + \beta_5 LYPB + \beta_6 LYQC$, with $\beta_0 = \alpha_0 + 2\alpha_1$, $\beta_1 = -\alpha_1$, $\beta_2 = \alpha_3 - \alpha_1$, $\beta_3 = \alpha_2 - \alpha_1$, $\beta_4 = \alpha_4$, $\beta_5 = \alpha_5 - \alpha_1$, and $\beta_6 = \alpha_6 + \alpha_3 - \alpha_1$.

Supplementary Tables

Supplementary table legends

Supplementary Table 1. Estimated median MBL concentration in serum.

Median MBL concentration in serum (ng/ml) estimated from Tobit regressions (on log-transformed data) with patients and controls specific means and an additive effect of haplotypes (Table 3 model 4). The 95% confidence intervals in the parentheses were estimated by use of the normal approximation on results from ordinary bootstrapping with 10000 replicates (Davison, A.C., Hinkley, D.V., Canty, A.J., *Bootstrap methods and their application*, Cambridge University Press, Cambridge, 1997). Results from using only the two markers X/Y and A/O (Table 3 model 3) are given before the corresponding four-marker multilocus genotype groups (e.g. YA/YA corresponds to YA=2, XA=YO=0). The results in the first row (Any) are from the model with only patient/control status as a factor (Table 3 model 1).

Supplementary Table 2. Estimated median MASP-2 concentration in serum.

Median MASP-2 concentration in serum (ng/ml) estimated from regressions analysis (on log-transformed data) with patients, controls and *MASP2* genotype specific means (interaction effect) and a linear effect of the O variant (A/O) in *MBL2* exon 1 (Table 4 model 4). The 95% confidence intervals (in the parentheses) were estimated by use of the normal approximation on results from ordinary bootstrapping with 10000 replicates. Results obtained from Table 4 model 3 (patient/control; *MASP2* genotype and the interaction effect) are given before the corresponding combination with *MBL2* genotype (*MASP2* A/A and *MASP2* A/G rows)). The results in the first row (Any) are from Table 4 model 1 which only includes the patient/control factor (i.e. any genotype combination).

Supplementary Table 1. Estimated median MBL concentration in serum.

MBL2 genotype	Controls (N = 349)	Patients (N = 98)
Any	584 (443 - 717)	578 (296 - 831)
YA/YA	4210 (3649 - 4754)	6357 (4907 - 7729)
HYP A/HYP A	4092 (3438 - 4733)	6698 (5017 - 8277)
HYP A/LYP A	2920 (1819 - 3923)	4780 (2897 - 6474)
HYP A/LYQ A	4412 (3863 - 4961)	7221 (5514 - 8836)
LYP A/LYP A	2084 (413 - 3485)	3411 (723 - 5658)
LYQ A/LYP A	3148 (1965 - 4229)	5153 (3103 - 7000)
LYQ A/LYQ A	4757 (3553 - 5731)	7785 (5437 - 9979)
YA/XA	1126 (990 - 1261)	1701 (1370 - 2019)
HYP A/LXP A	1116 (974 - 1253)	1826 (1449 - 2179)
LYP A/LXP A	796 (493 - 1069)	1303 (805 - 1749)
LYQ A/LXP A	1203 (1 030 - 1370)	1969 (1523 - 2386)
XA/XA	301 (221 - 378)	455 (322 - 580)
LXP A/LXP A	304 (228 - 375)	498 (357 - 627)
YA/YO	188 (156 - 219)	285 (221 - 344)
HYP A/LYP B	138 (109 - 165)	226 (169 - 279)
LYP A/LYP B	99 (56 - 136)	161 (93 - 222)
LYQ A/LYP B	149 (117 - 179)	244 (180 - 303)
HYP A/LYQ C	136 (31 - 226)	223 (43 - 375)
LYQ A/LYQ C	147 (35 - 243)	240 (47 - 405)
HYP A/HYP D	385 (259 - 498)	630 (380 - 851)
LYP A /HYP D	275 (139 - 393)	449 (216 - 650)
LYQ A /HYP D	415 (278 - 538)	679 (405 - 922)
XA/YO	50 (38 - 62)	76 (56 - 95)
LXP A/HYP D	105 (68 - 138)	172 (102 - 233)
LXP A/LYP B	38 (28 - 47)	62 (44 - 78)
LXP A/LYQ C	37 (7 - 63)	61 (10 - 104)
YO/YO	8 (5 - 11)	13 (8 - 17)
HYP D/HYP D	36 (10 - 58)	59 (13 - 97)
HYP D/LYP B	13 (8 - 18)	21 (12 - 29)
HYP D/LYQ C	13 (2 - 22)	21 (2 - 37)
LYP B/LYP B	5 (3 - 6)	8 (4 - 10)
LYP B/LYQ C	5 (1 - 8)	8 (1 - 13)

Supplementary Table 2. Estimated median MASP-2 concentration in serum.

<i>MBL2</i> genotype	Controls (N = 349)	Patients (N = 98)
Any	412 (395 - 428)	442 (394 - 488)
<i>MASP2</i> A/A	431 (415 - 448)	487 (438 - 533)
A/A	409 (390 - 428)	457 (409 - 503)
A/O	459 (437 - 480)	513 (461 - 563)
O/O	515 (466 - 563)	576 (502 - 648)
<i>MASP2</i> A/G	260 (223 - 295)	206 (146 - 261)
A/A	247 (214 - 279)	194 (139 - 245)
A/O	278 (239 - 315)	217 (157 - 274)
O/O	312 (259 - 362)	244 (173 - 309)