

Supplementary Appendix

This appendix has been provided by the authors to give readers additional information about their work.

Supplement to: Stengaard-Pedersen K, Thiel S, Gadjeva M, et al. Inherited Deficiency of Mannan-Binding Lectin-Associated Serine Protease 2. *N Engl J Med* 2003;349:554-60.

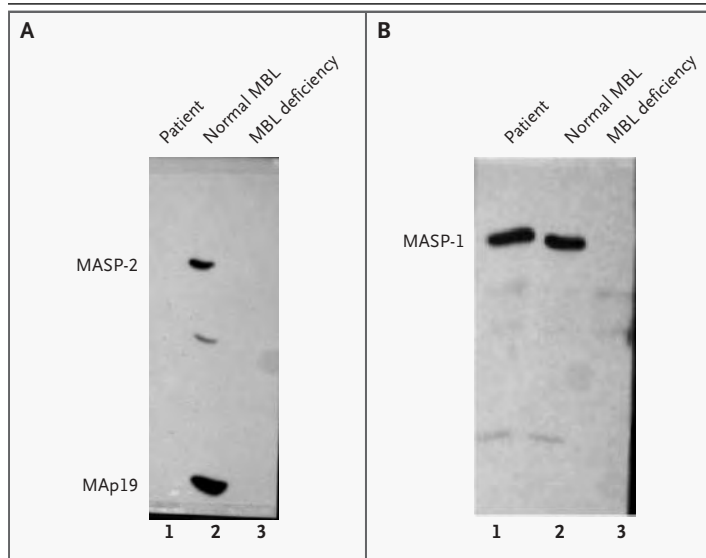
Supplementary Appendix 1. Complement Profile in the Patient.

Component	Patient's Value	Reference Value
C1q (% of normal)	11	76–136
C4 (g/liter)	0.06	0.12–0.33
C3 (g/liter)	0.36	0.77–1.38
Properdin (% of normal)	54	57–153
C1 inhibitor (% of normal)	123	72–146
C1 inhibitor function (% of normal)	116	70–130
C3dg fragments (mg/liter)	13	<5
Terminal component complexes (SC5b–C9) (U/liter)	43	<20
Classic-pathway function (% of normal)*	6	90–112
Alternative-pathway function (% of normal)†	22	65–161
Anti-C1q autoantibodies (U/liter)‡	55	<16

* Values reflect the extent of hemolysis of antibody-coated sheep erythrocytes.

† Values reflect the extent of hemolysis of rabbit erythrocytes.

‡ Anti-C1q autoantibodies were determined by enzyme-linked immunosorbent assay. The antibodies were not detectable by Western blot analysis.



Supplementary Appendix 2. Western Blot Analysis of Materials Eluted from Anti-Mannan-Binding Lectin-Coated Wells after Incubation with Plasma from the Patient.

The blots were developed with antibody against MAp19 and mannan-binding lectin-associated serine protease 2 (MASP-2) (Panel A) and antibody against MASP-1 (Panel B). In each panel, lane 1 shows plasma from the patient, lane 2 plasma from a subject with normal levels of mannan-binding lectin (MBL), and lane 3 plasma from a subject with MBL deficiency.