

CLAIMS

The embodiments of the invention in which an exclusive property or privilege is claimed are defined as follows:

1. Use of a MASP-2 inhibitory monoclonal antibody, or antigen-binding fragment thereof in the manufacture of a medicament for treating a human subject suffering from plasma therapy-resistant aHUS.
2. The use of claim 1, wherein the antibody or fragment thereof is selected from the group consisting of a recombinant antibody, an antibody having reduced effector function, a chimeric antibody, a humanized antibody, and a human antibody.
3. The use of claim 1 wherein the subject has previously undergone, or is currently undergoing, treatment with a terminal complement inhibitor that inhibits cleavage of complement protein C5.
4. The use of claim 3, wherein the terminal complement inhibitor is a humanized anti-C5 antibody or antigen-binding fragment thereof.
5. The use of claim 1, wherein the MASP-2 inhibitory antibody does not substantially inhibit the classical pathway.
6. The use of claim 1, wherein the MASP-2 inhibitory antibody inhibits C3b deposition in 90% human serum with an IC_{50} of 30 nM or less.
7. The use of claim 1, wherein the MASP-2 inhibitory antibody is to be delivered to the subject systemically.
8. The use of claim 1, wherein administration of the MASP-2 inhibitory antibody improves at least one or more of the following clinical parameters associated with aHUS: (i) an increase in platelet count, (ii) a decrease in LDH, (iii) an increase in haptoglobin, (iv) an increase in hemoglobin, and/or (v) a decrease in creatinine.
9. The use of claim 1, wherein the MASP-2 inhibitory antibody or antigen-binding fragment thereof, comprises a heavy-chain variable region comprising CDR-H1, CDR-H2 and CDR-H3 of the amino acid sequence set forth as SEQ ID NO:67 and a light-chain variable

region comprising CDR-L1, CDR-L2 and CDR-L3 of the amino acid sequence set forth as SEQ ID NO:70.

10. The use of claim 1, wherein the MASP-2 inhibitory antibody or antigen binding fragment thereof, comprises a heavy chain variable region comprising SEQ ID NO:67 and a light chain variable region comprising SEQ ID NO:70.