

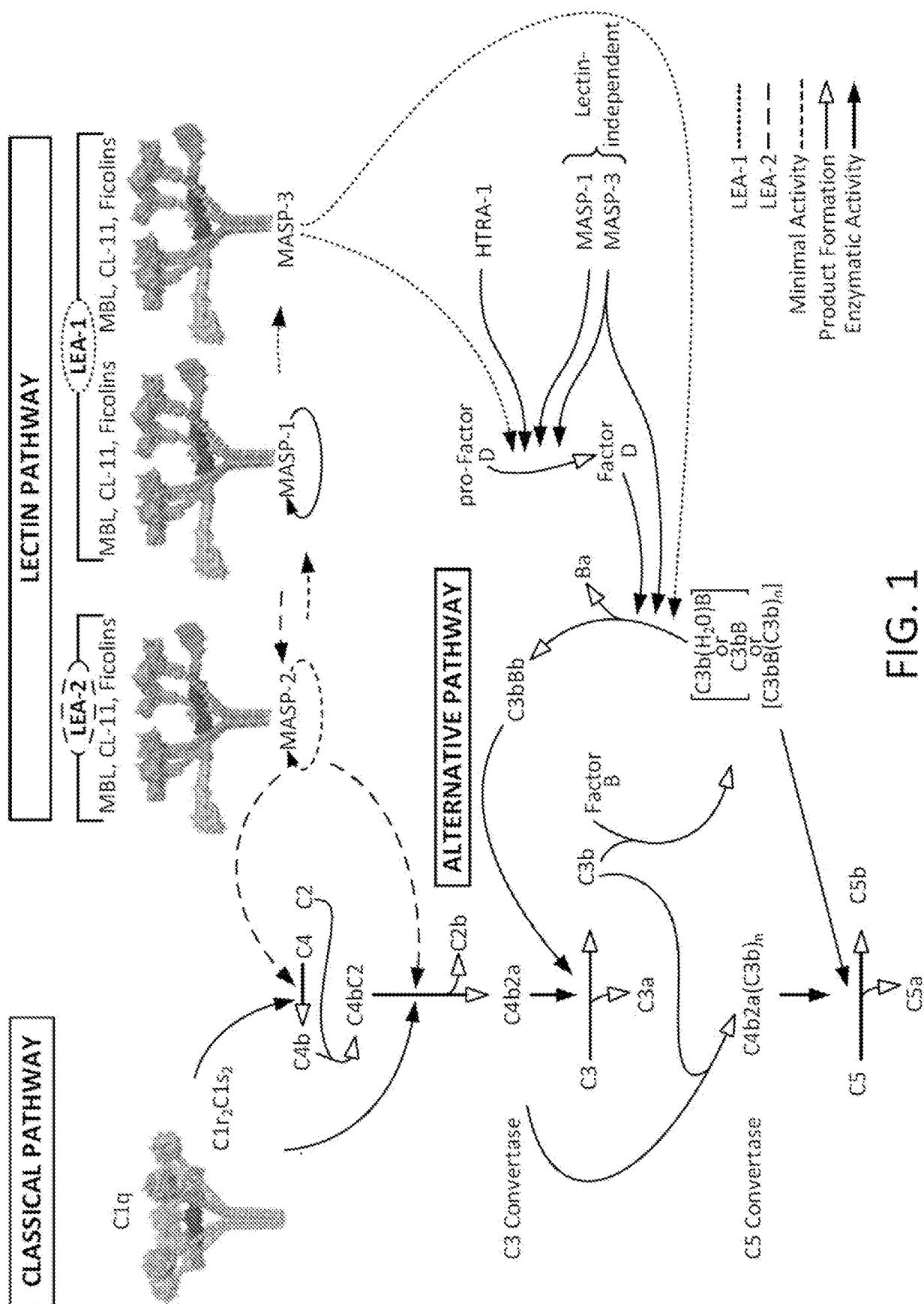


FIG. 1

FIG. 2

MAASP 1 and 3

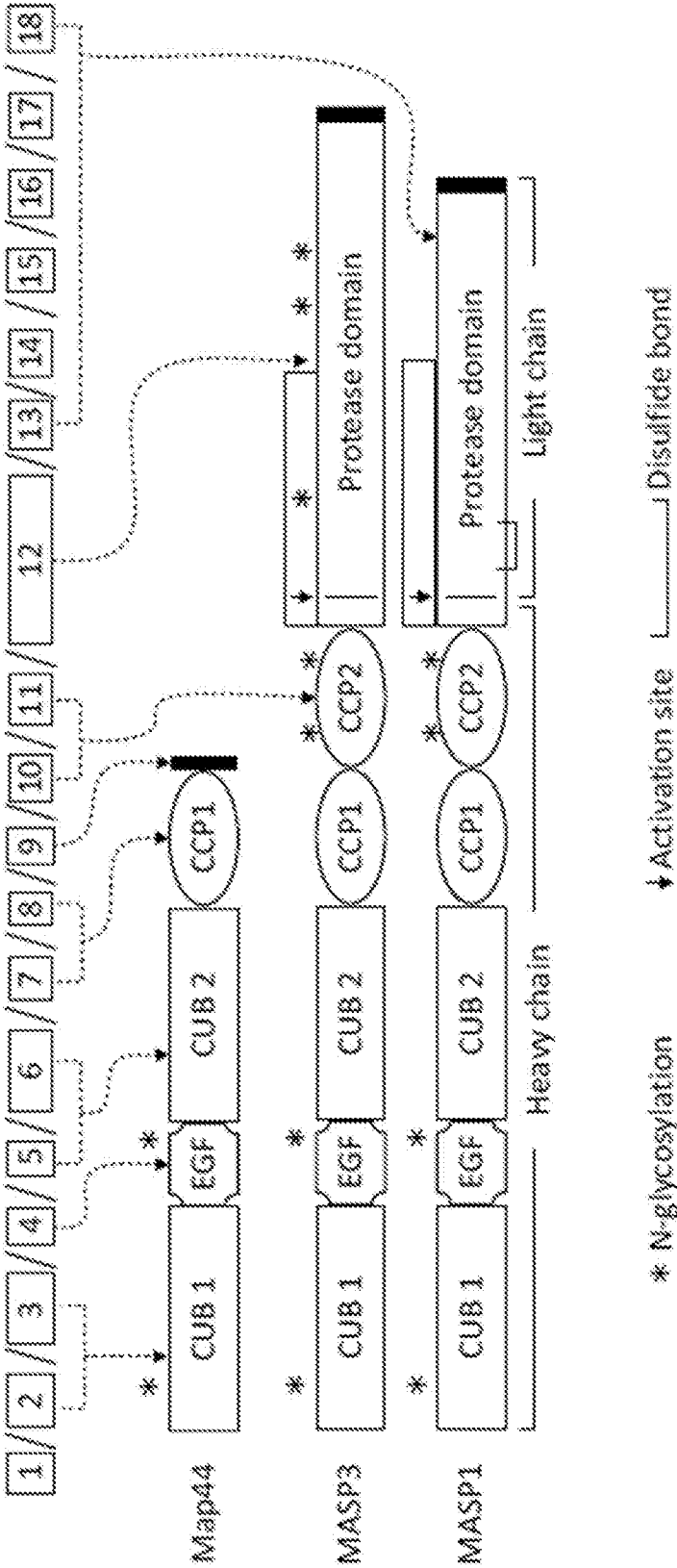


FIG. 3

Human MASP-3 (SEQ ID NO:2)

MRWLLYYALCFSLSKASAHTVELNNMFGQIQSPGYPDSPSPDSEVTWNITVPDGFR IKLYFMHFNLESSYLCEYD
 YVKVETEDQVLATFCGRETTDTEQTGQEVVLSPGSFMSITFRSDFSNEERFTGFD AHYMAVDVDECKEREDEELS
 CDHYCHNYIGGYCSCRFGYILHTDNRTCRVECDNLFTQRTGVI TSPDFPNPYPKSSECLYTIELEEGFMVNLQF
 EDIFDIEDHPEVPCPYDIKIKVGPKVLGPFCEGEKAPEPISTQSHSVLILFHSDNSGENRGWRLSYRAAG
 NECPELQPPVHGKIEPSQAKYFFKDQVLVSCDTGYKVLKDNVEMDTFQIECLKDGTWSNKIPTCKIVDCRAPGELE
 HGLITFSTRNNLT TYKSEIKYSCQEPYYKMLNNNTGIYTCSAQGVWMNKVLGRSLPTCLPECGQPSRSLPSLVKRI
 IGGRNAEPGLFPWQALIVVEDTSRVPNDKWFSGGALLSASWILTAAHVLRQRD TTVIPVSKEHVTYVLGLHDVR
 DKSGAVNSSAARVVLHPDENIQYNHDI ALVQLQEPVPLGPHVMPVCLPRLEPEGPAPHMLGLVAGWGISNPNVTY
 DEIISSGTRTLDVLQYVKLPVVPHAECKTSYESRSGNYSVTENMFCAGYYEGGKDTCLGDSGGAFVIFDDLSQRW
 VVQGLVSWGPEECGSKQVYGVT KVSNIYVDWVWEQMGLPQSVVEPQVER

} CUB1-EGF-CUB2
 } CCP1-CCP2-SP

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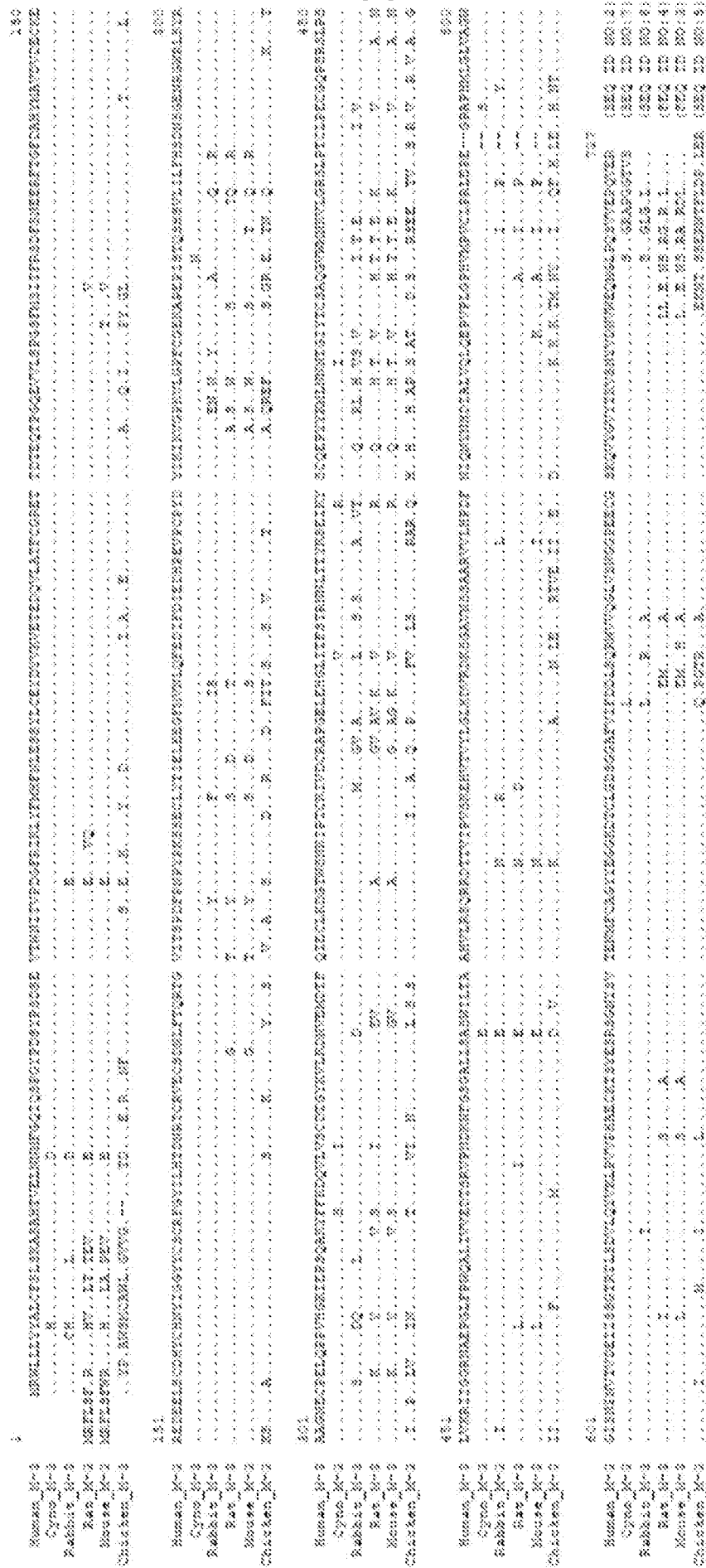


FIG. 5
MASP-3 SP domain: multispecies alignment

Human_M0_SP	1	IGGNAEPGLEFNGALIVVEDTAEVNDKMFSGALLSAWILTAHLLASQSDITVIVAEKHHVVLGLGRDVRKNS	80
Cyno_M0_SP		E.....	
Rabbit_M0_SP		E.....N.....A.....	
Rat_M0_SP		I.....E.....N.....D.....	
Mouse_M0_SP		L.....E.....N.....	
Chicken_M0_SP		F.....M.....D.....V.....K.....A.....N.....L.....	
Human_M0_SP	81	GAVNSAARVVLKEPTNQNNHDLALVQGEVLEGEHVEVLELEED--GFAFNDLGLTAAGLSENVVDEELIS	160
Cyno_M0_SP		I.....~.....~.....~.....	
Rabbit_M0_SP		I.....A.....I.....P.....~.....	
Rat_M0_SP		I.....K.....A.....I.....P.....~.....	
Mouse_M0_SP		E.....RVEII.....E.....D.....KKKKTWNY.....I.....QFHLDE.....N.....NT.....I.....	
Chicken_M0_SP		I.....	
Human_M0_SP	161	SGRTSLDVLQVNLPEVVEKAEKTSVIESRSGHVSIVTENKFACTVGGGKDTCLGGSGARVIFDLSQNNVQGLVNG	240
Cyno_M0_SP		I.....	
Rabbit_M0_SP		S.....A.....~.....~.....~.....L.....A.....A.....	
Rat_M0_SP		S.....A.....~.....~.....~.....EM.....A.....	
Mouse_M0_SP		M.....I.....	
Chicken_M0_SP		M.....I.....	
Human_M0_SP	241	GPEZCGSNQVGEVTVNVEVWVEWQSLDQ--SVVEFQVER	280
Cyno_M0_SP		~.....~.....~.....S.....--GPAIGGVTS	
Rabbit_M0_SP		~.....~.....~.....S.....--GLG.....L.....	
Rat_M0_SP		~.....~.....~.....LL.....E.....NS.....R.....G.....R.....L.....	
Mouse_M0_SP		~.....~.....~.....L.....E.....NS.....R.....~.....A.....FDL.....	
Chicken_M0_SP		~.....~.....~.....EENT.....SSEKNTFLD.....EL.....	

FIG. 6

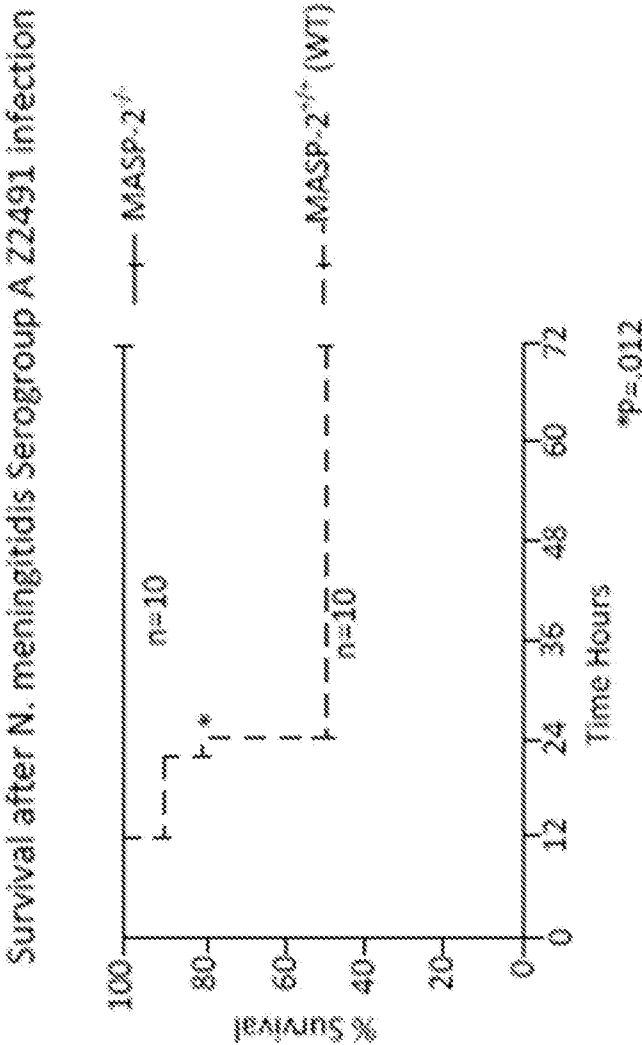


FIG. 7

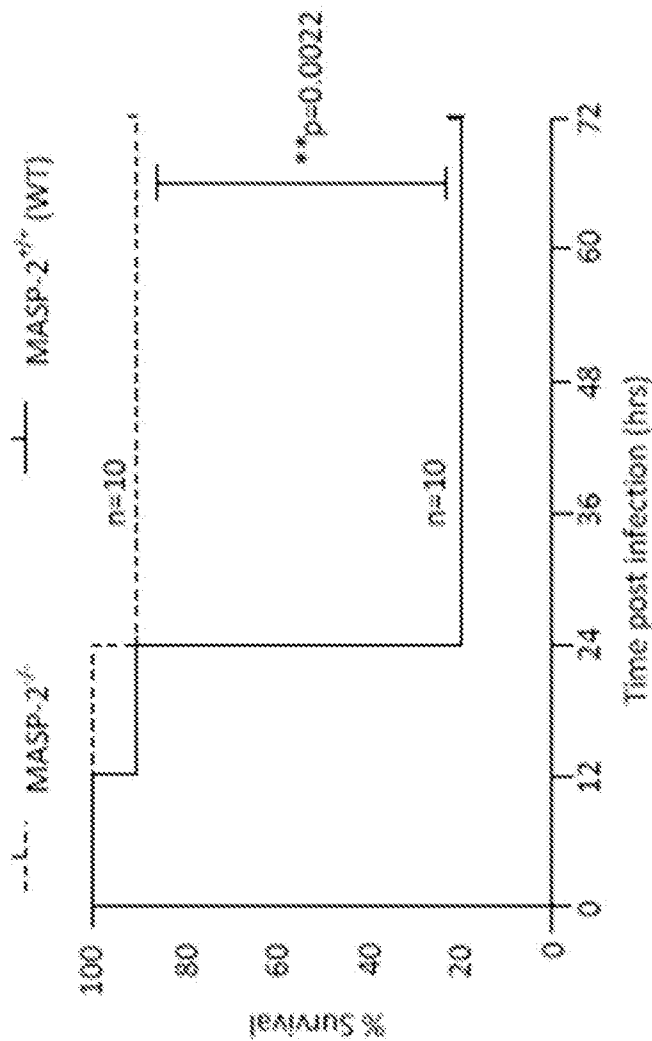


FIG. 8

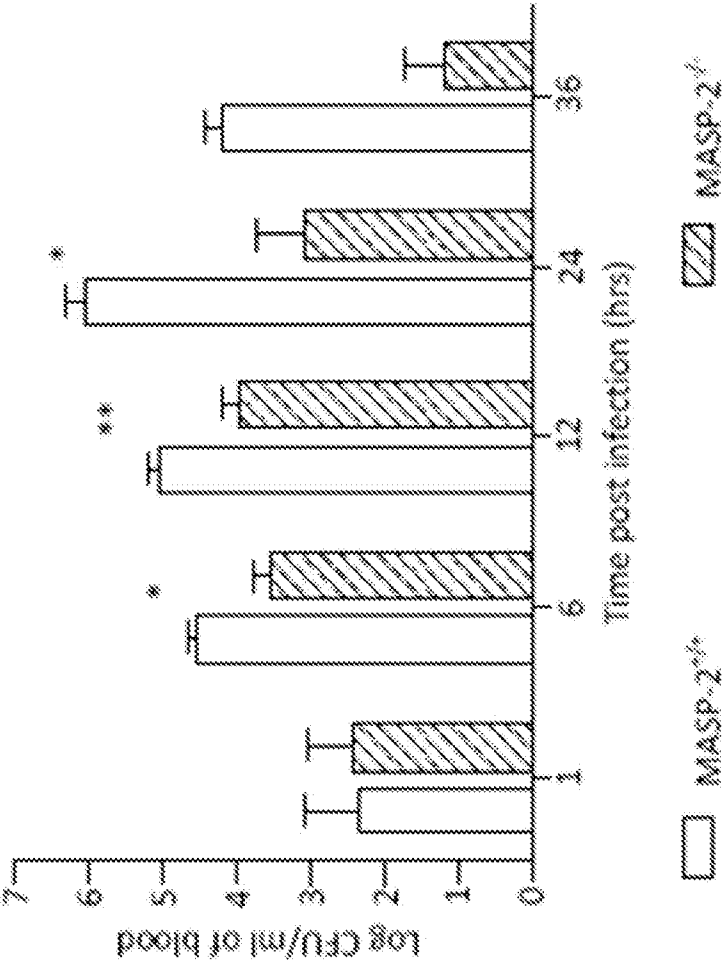


FIG. 9

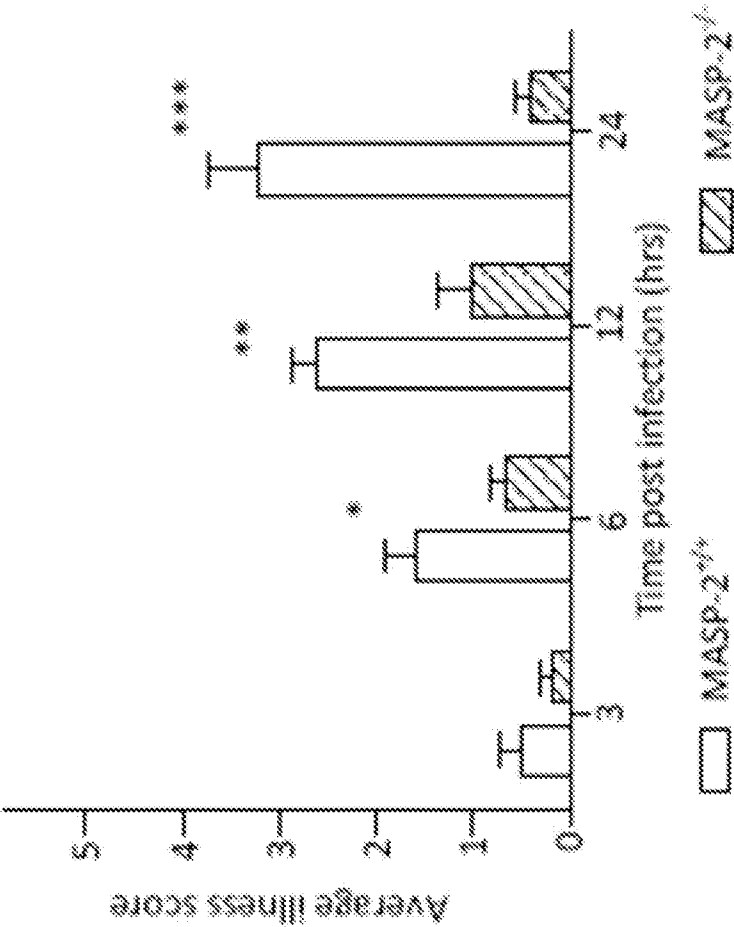


FIG. 10

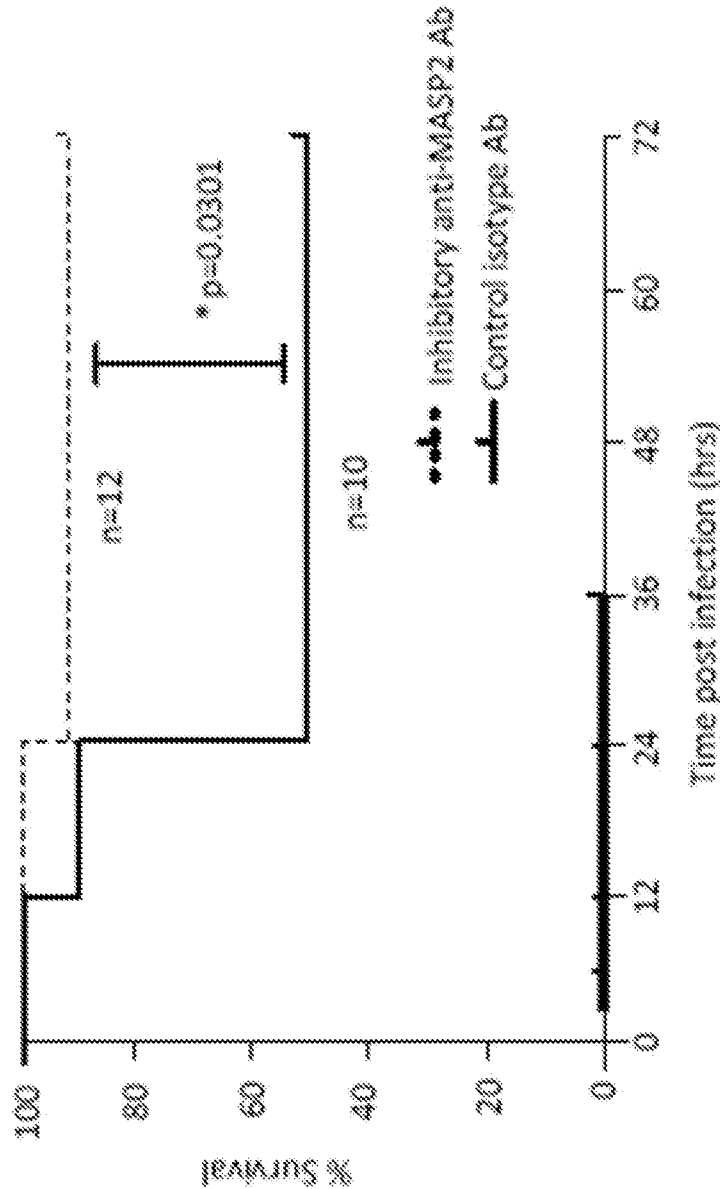


FIG. 11

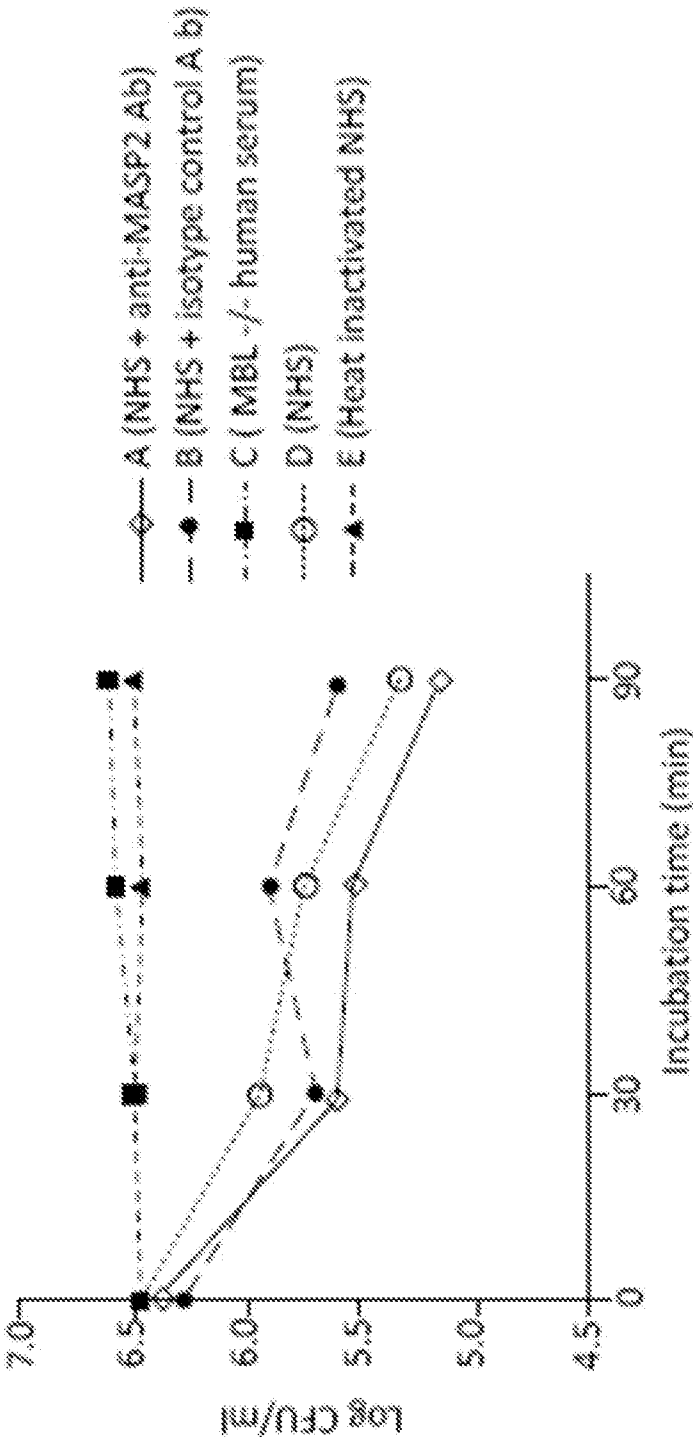


FIG. 12

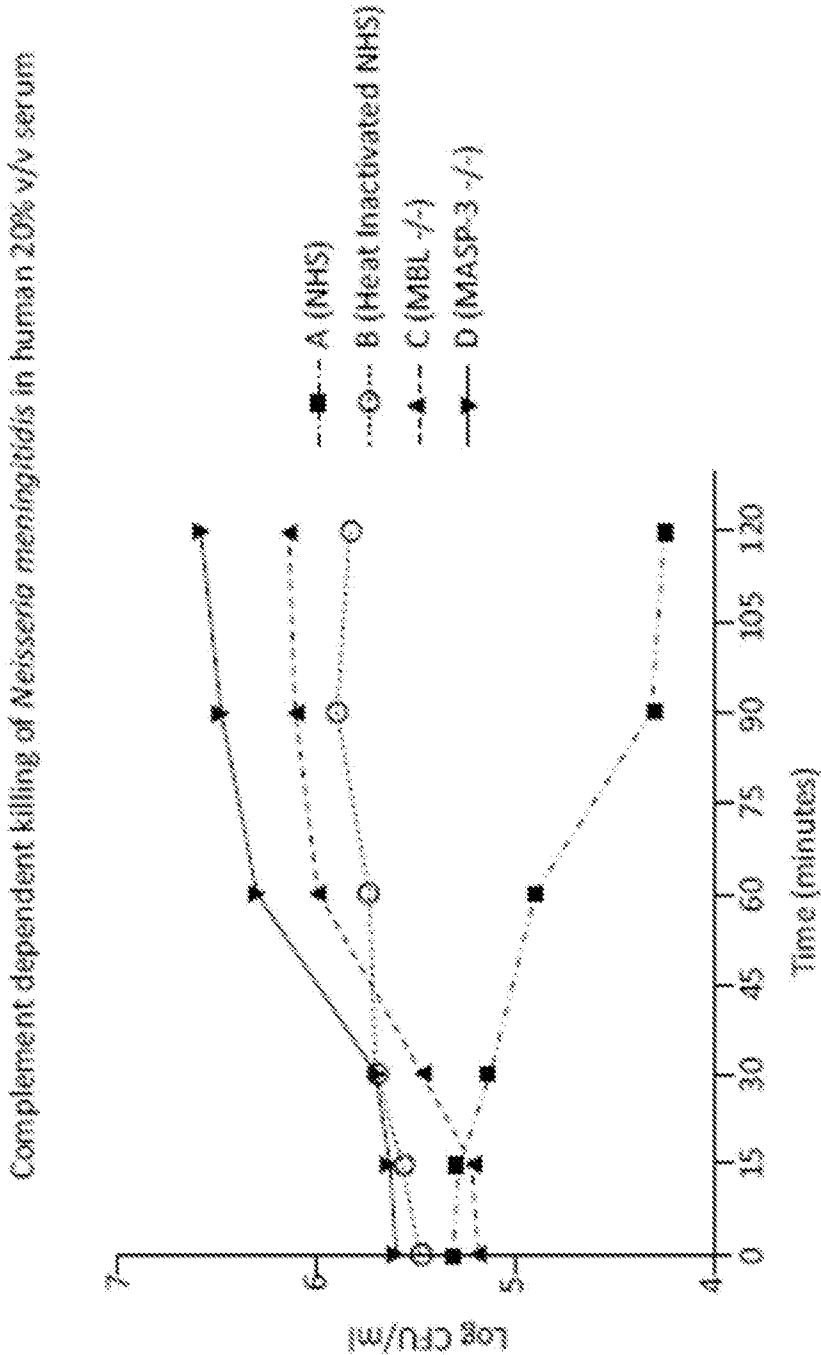


FIG. 13

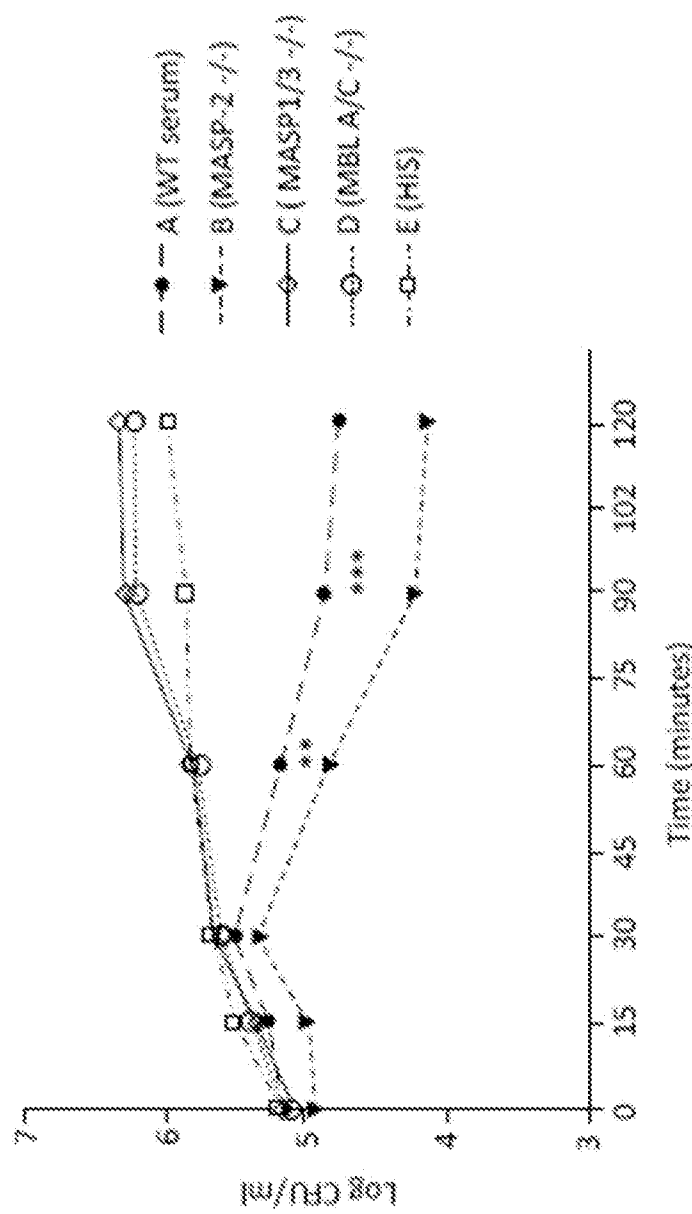


FIG. 14

MASP-1/3 deficient mice are not deficient of LP functional activity

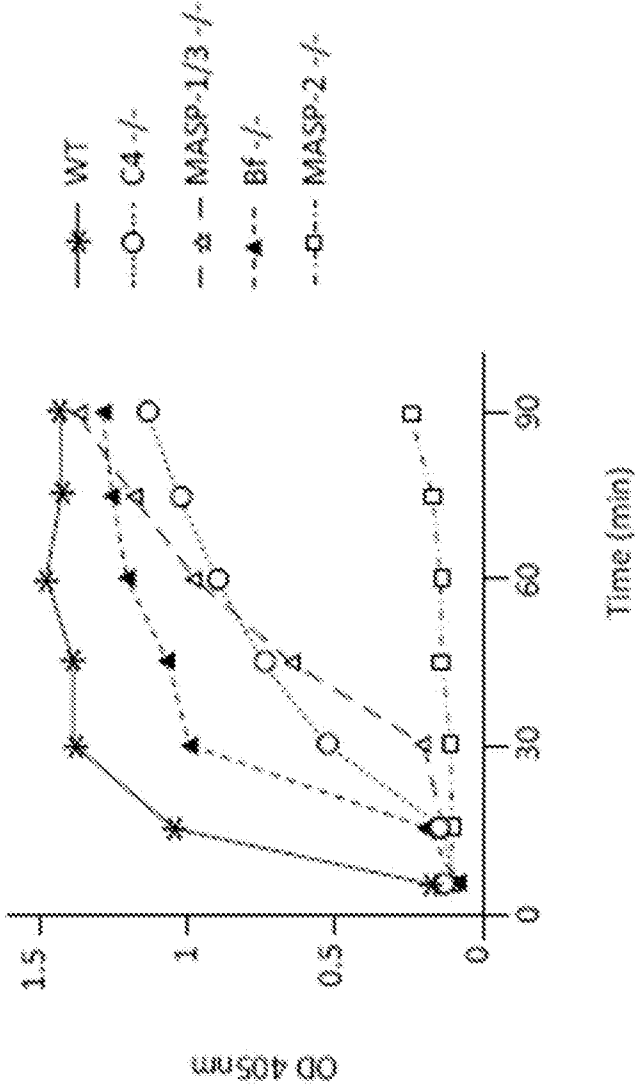


FIG. 15

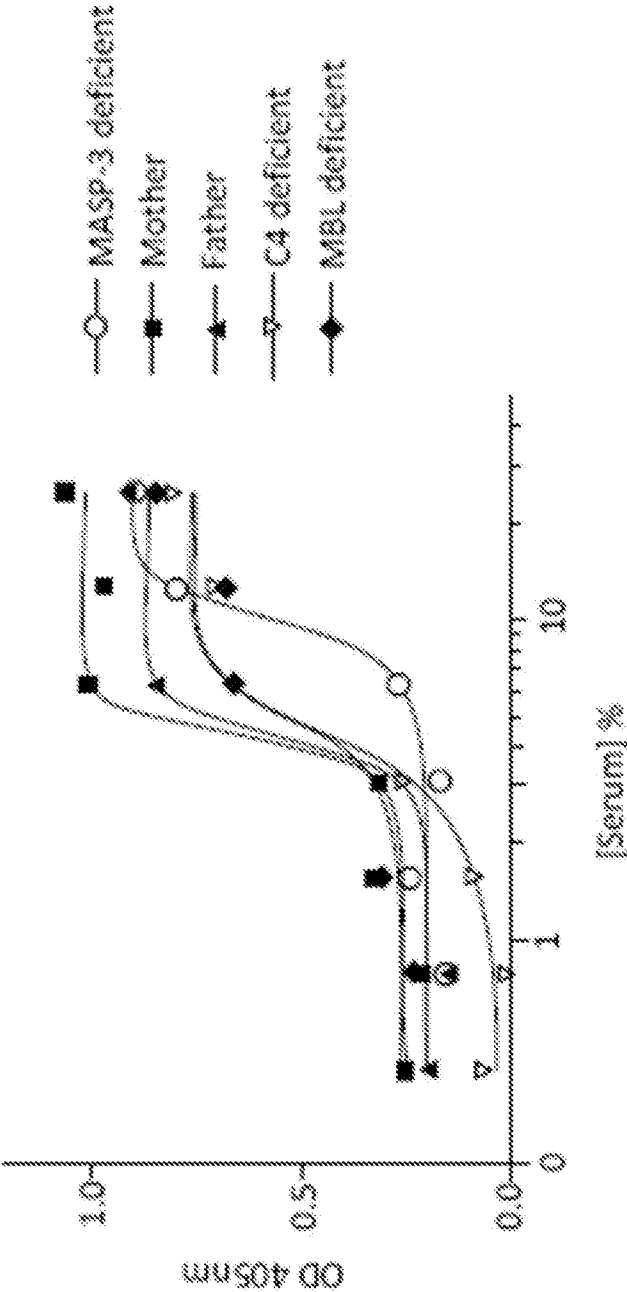


FIG. 16

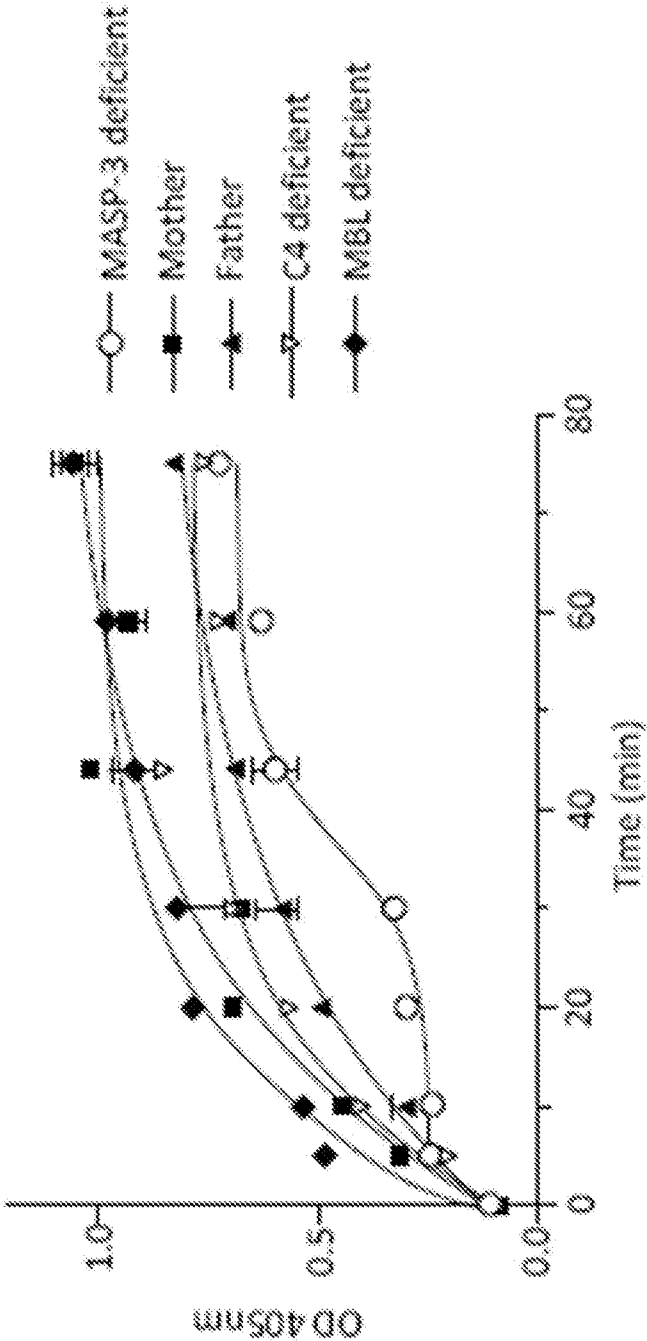


FIG. 17A

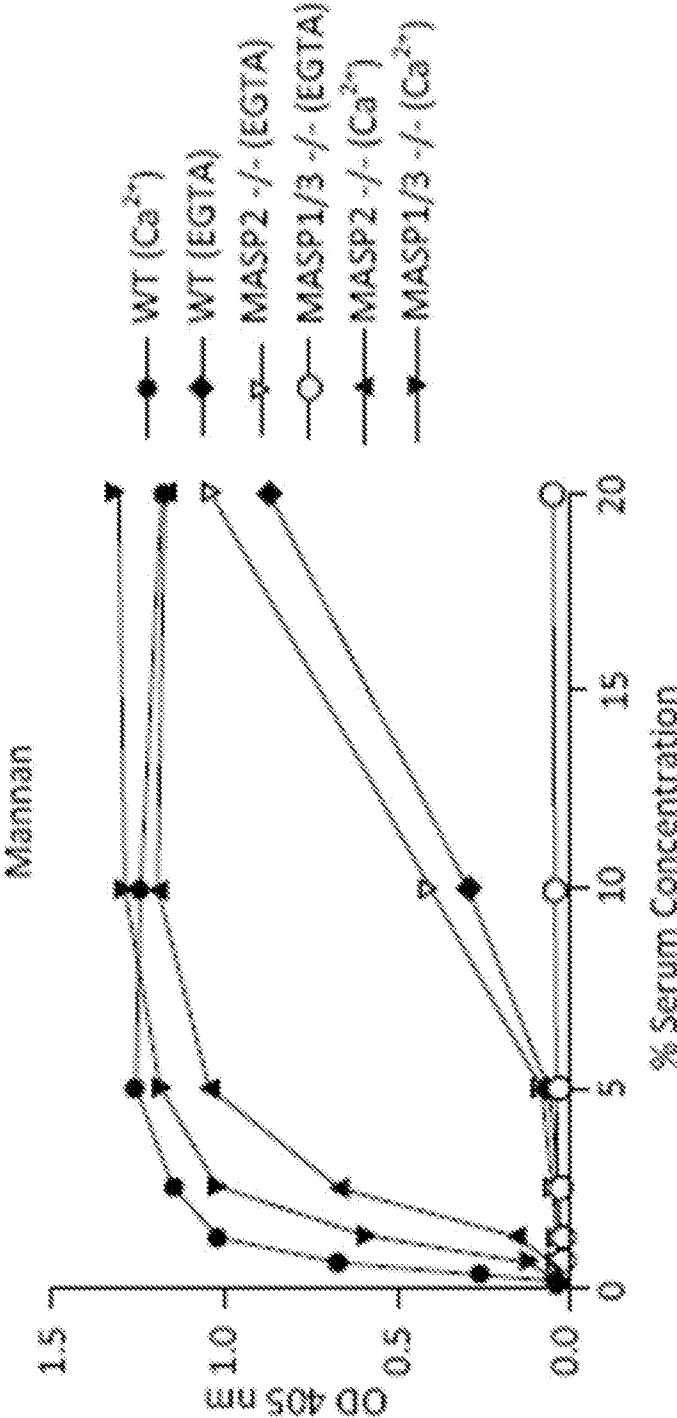


FIG. 17B

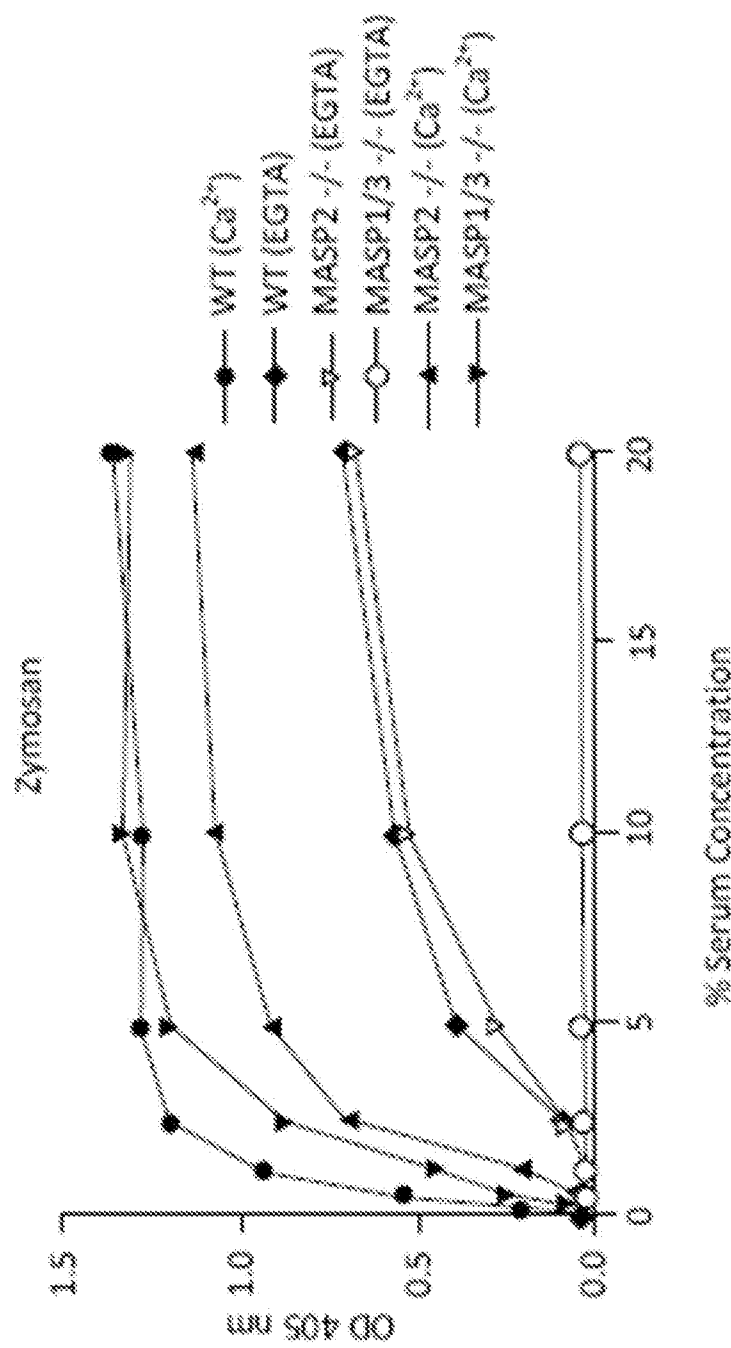


FIG. 17C

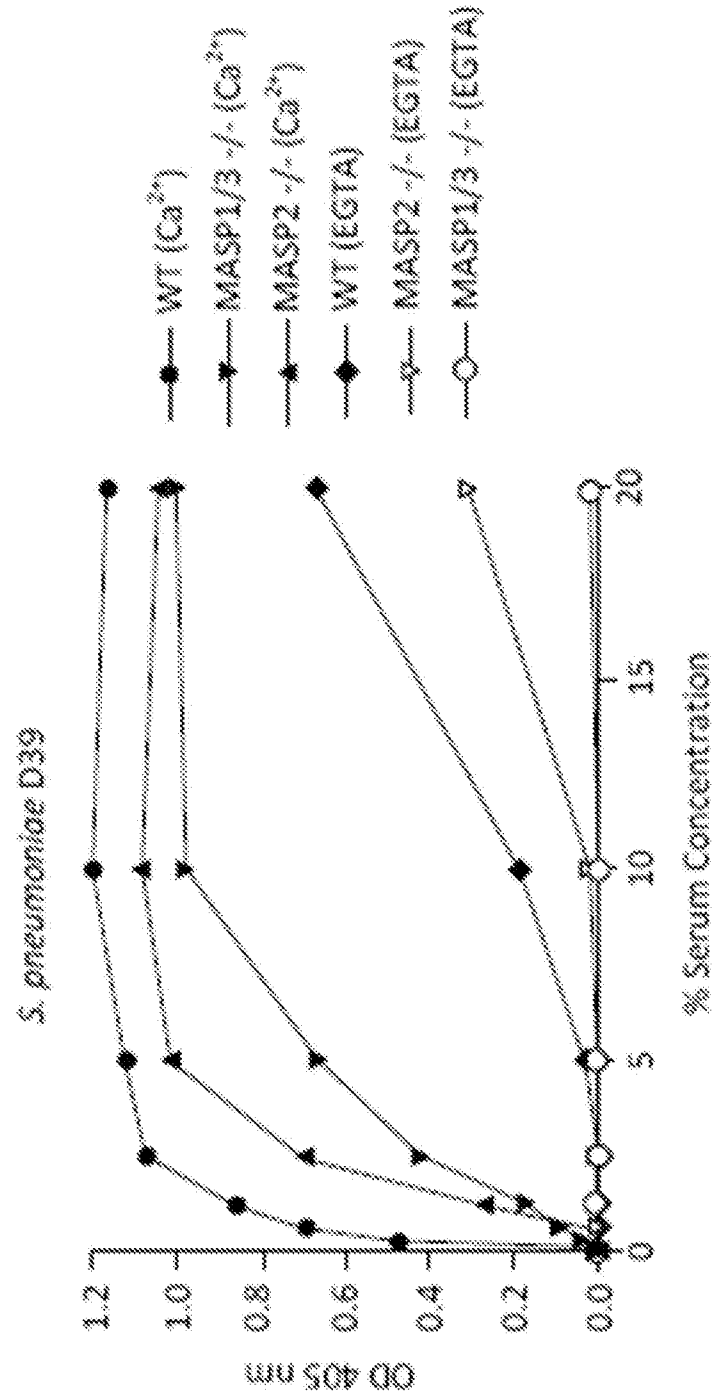


FIG. 18A

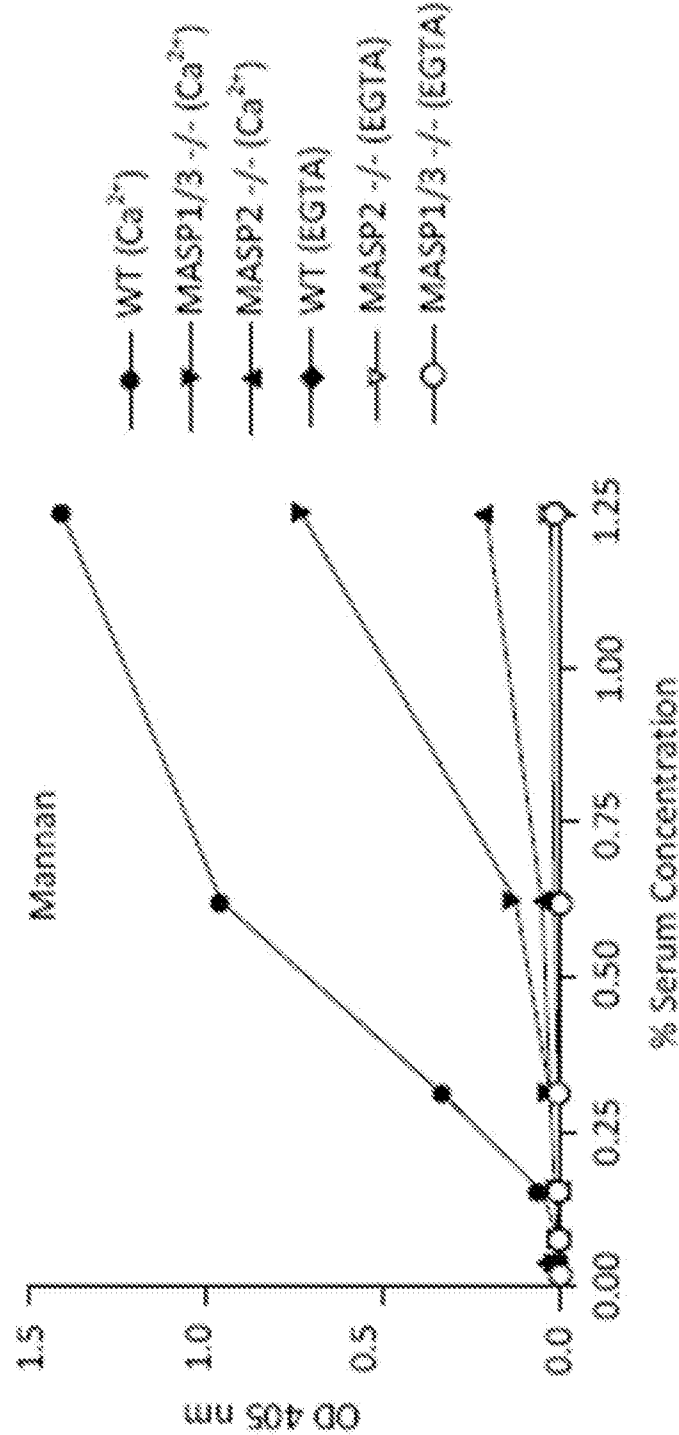


FIG. 18B

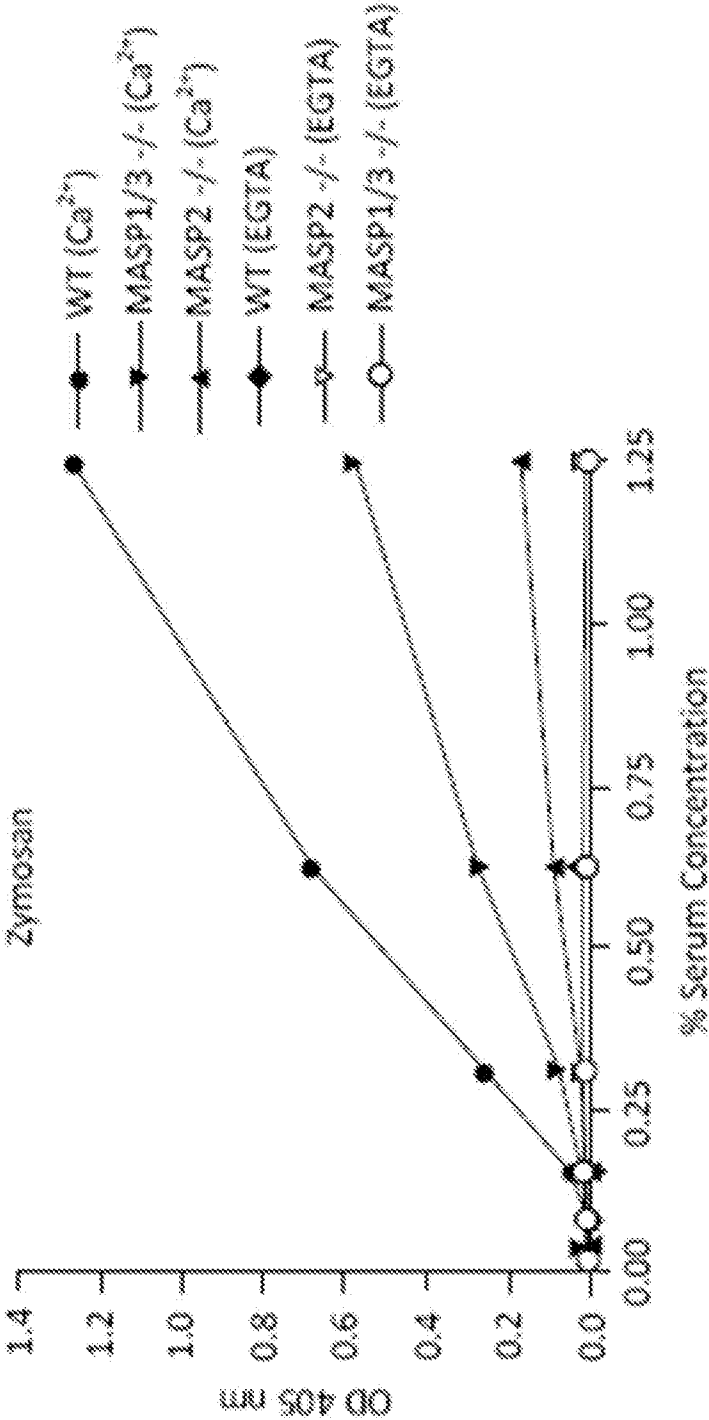


FIG. 18C

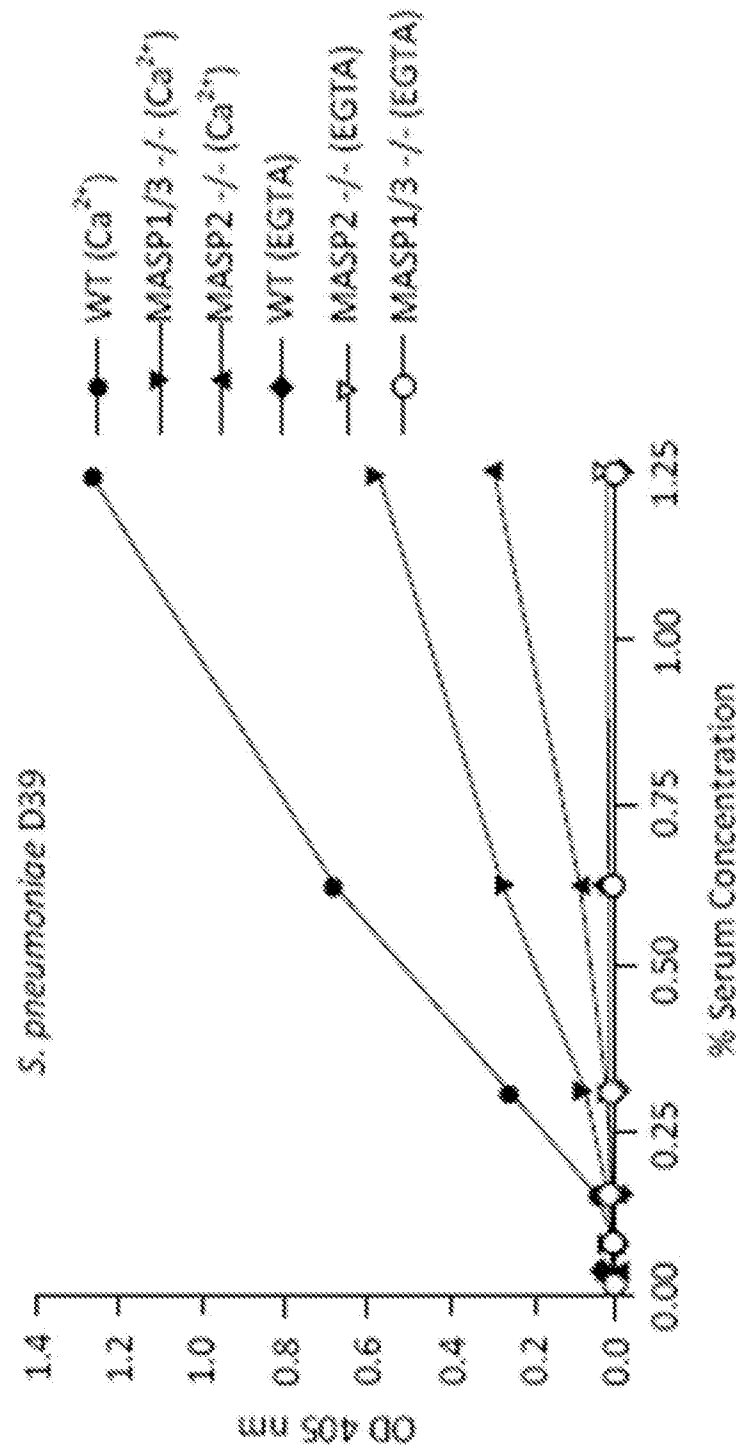


FIG. 19

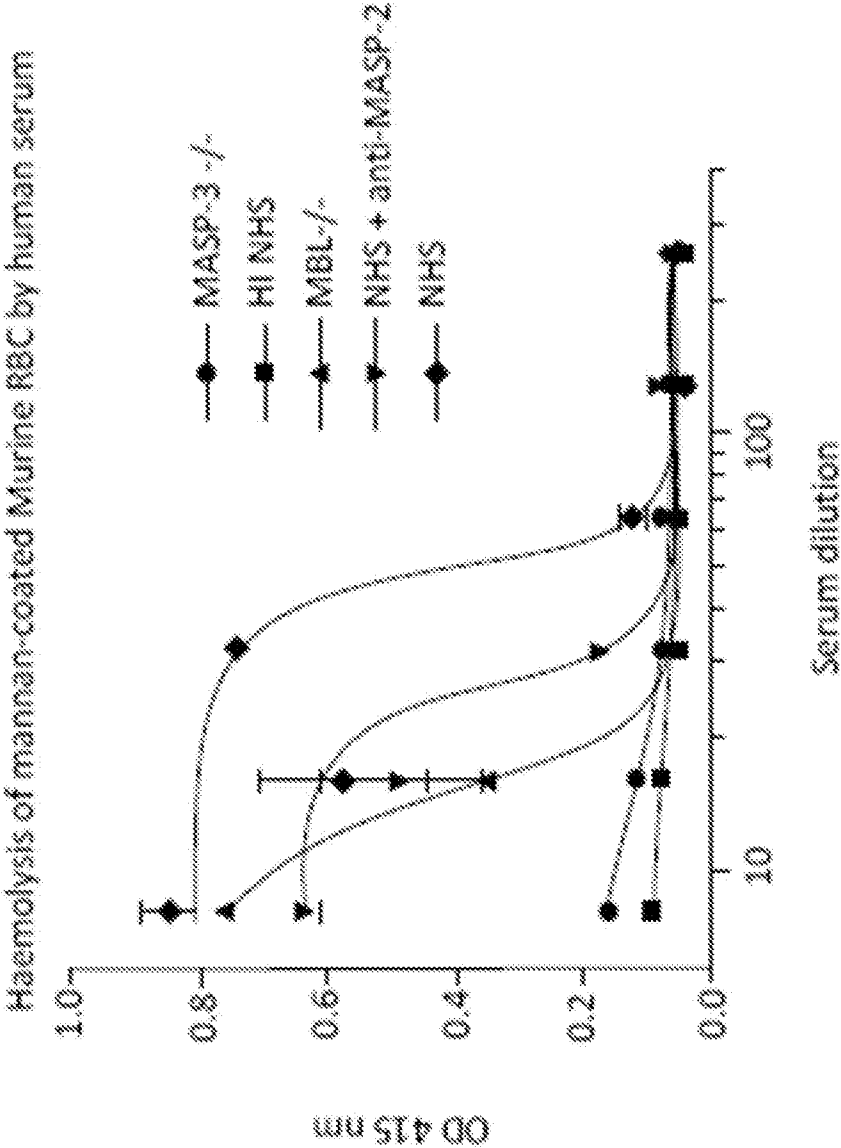


FIG. 20

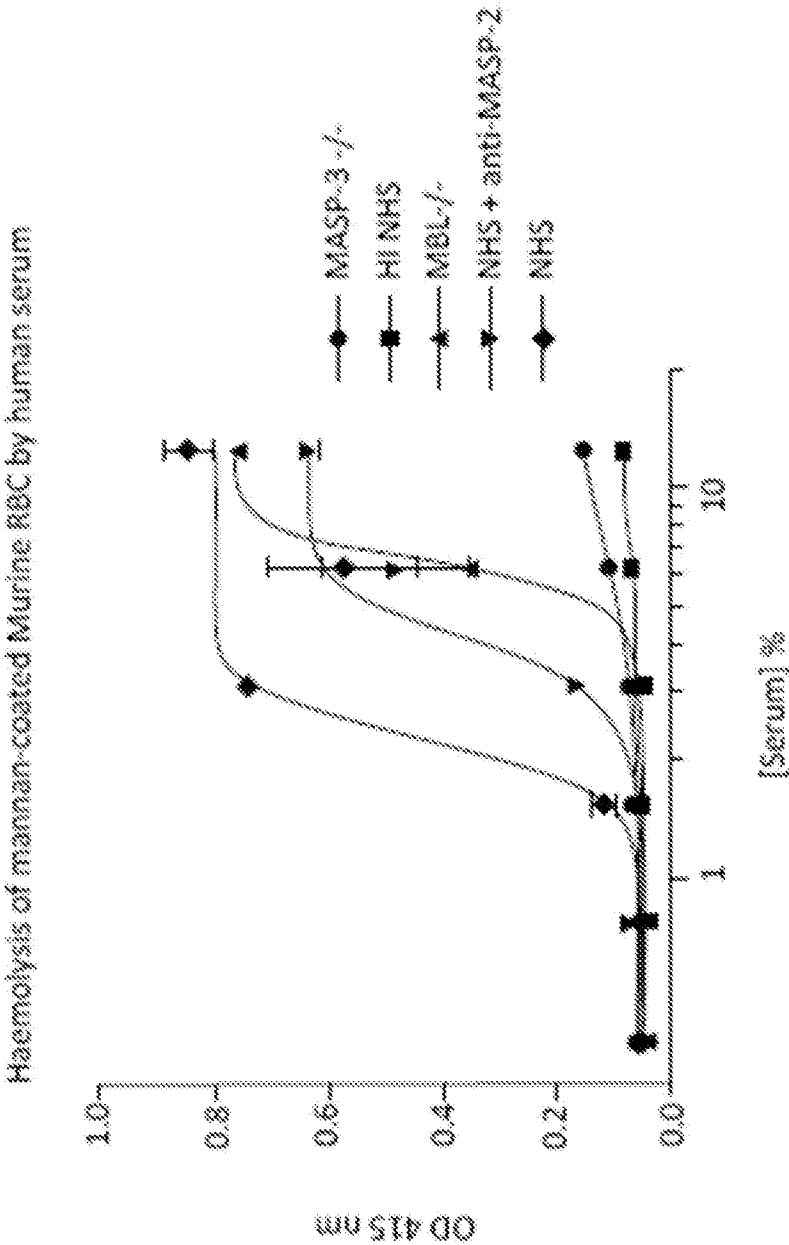


FIG. 21

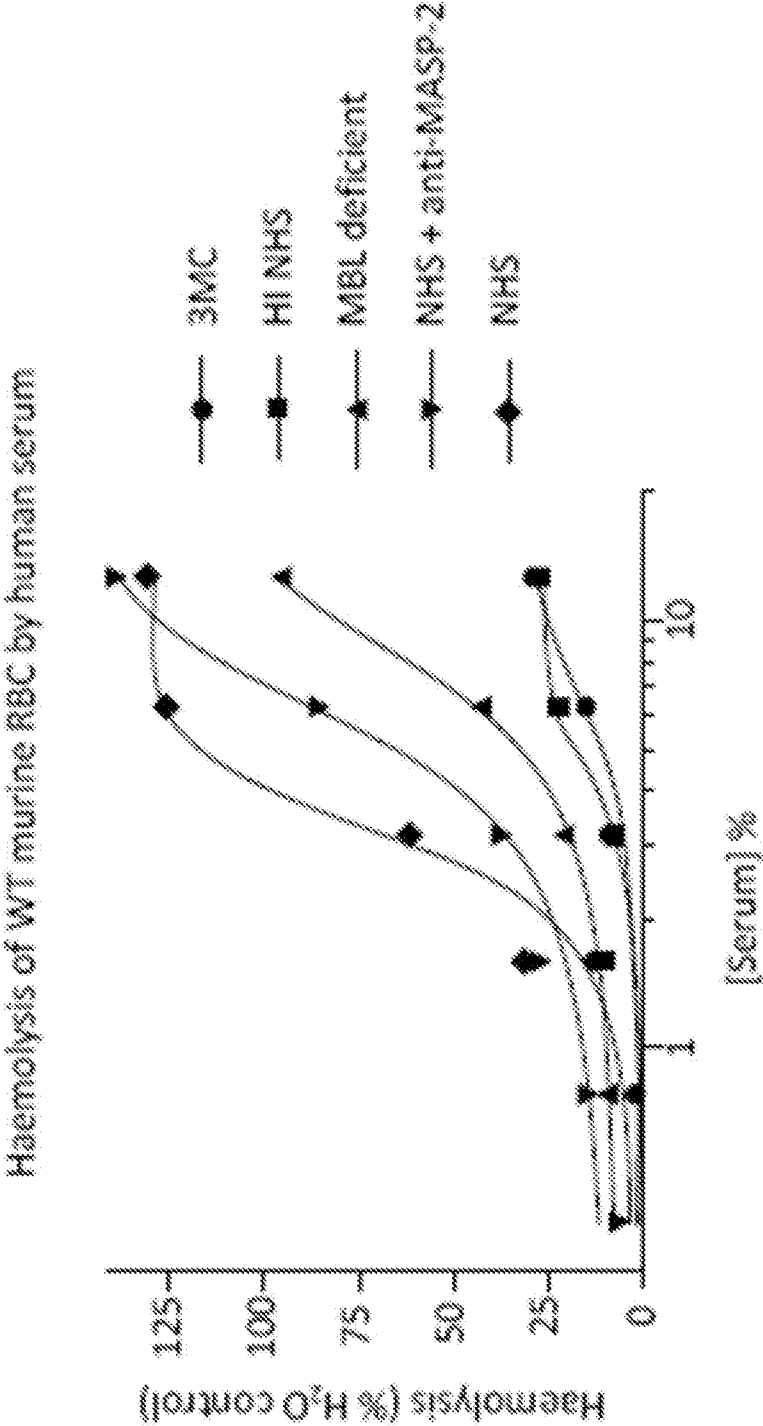


FIG. 22

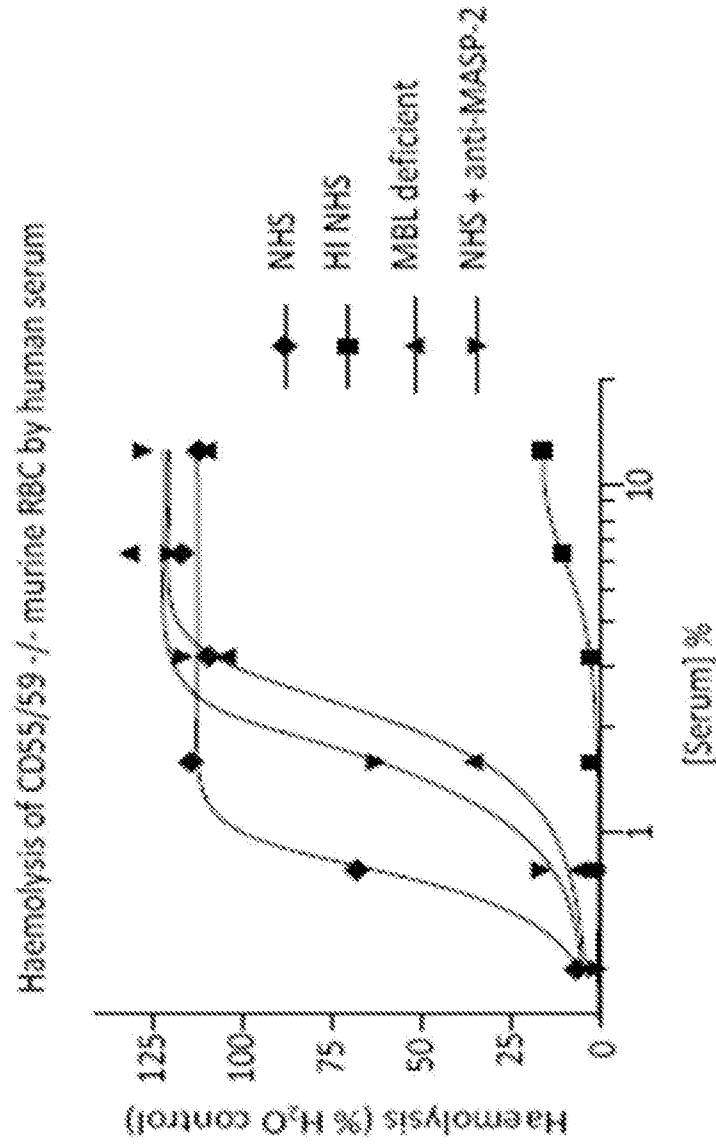


FIG. 23

Haemolytic assay using Rabbit RBCs with WT and MASP-1/3^{-/-} mouse sera.

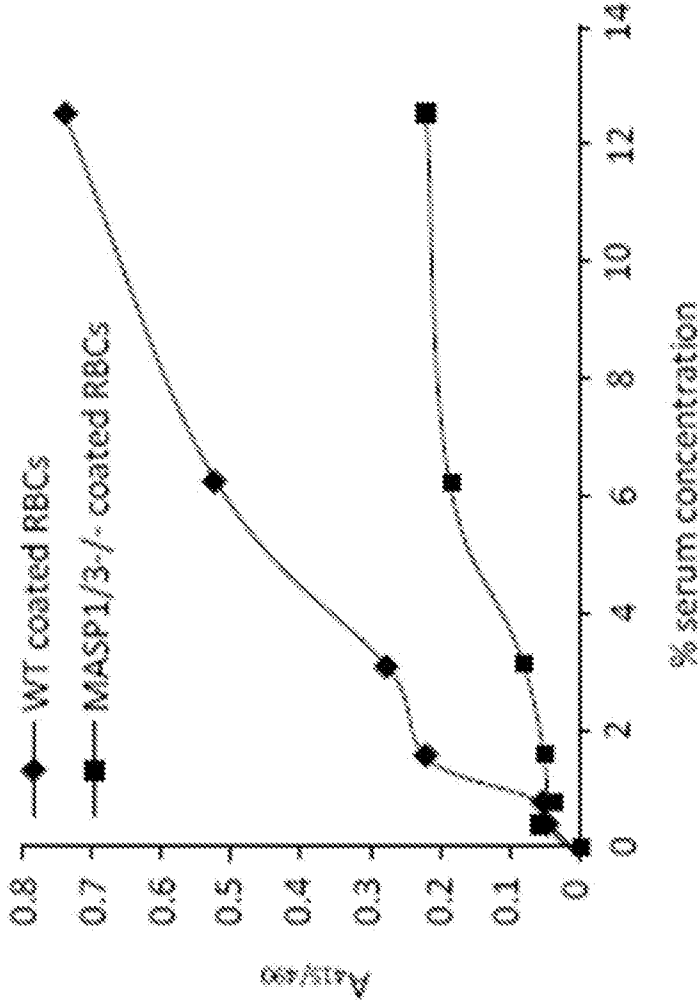


FIG. 24B

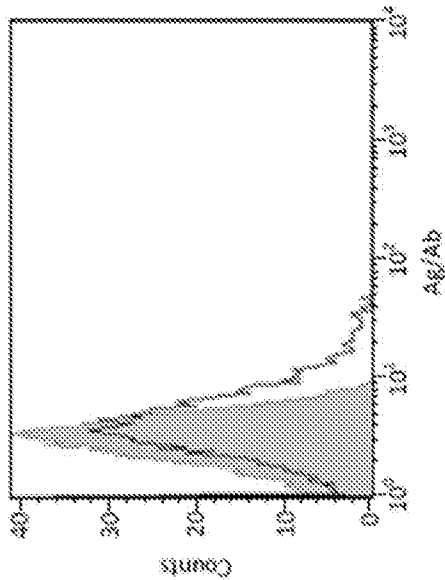


FIG. 24A

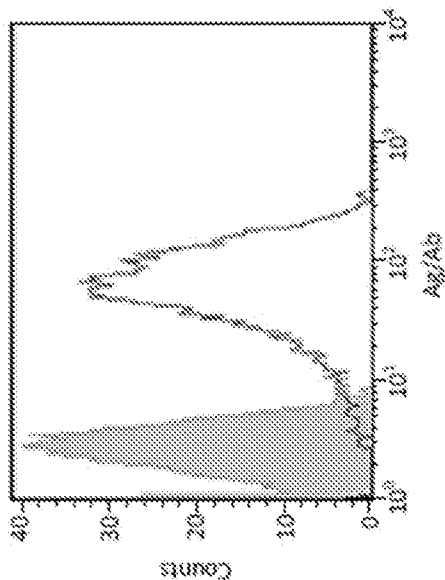


FIG. 25

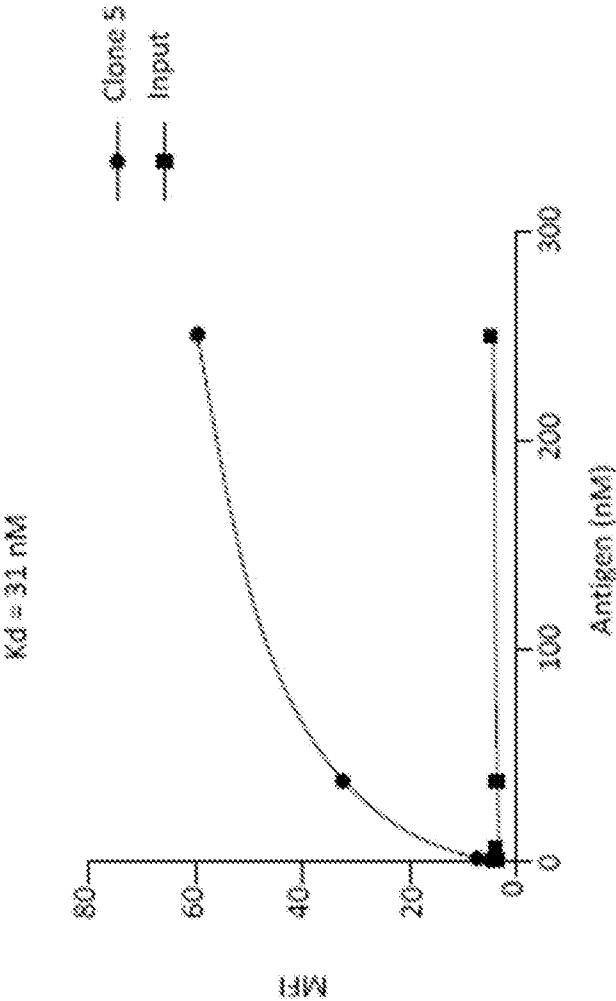


FIG. 26A

1	65
VH	AVTLDESGGGLQTPGGALS LVCKASGFTFSNANMCHNRQAPGKGLEWVAGID-DDGS GTRYAPAV
M3J5	G.....Y.....M.....Y.....R-S...F.L...T...
M3M1	G.....D...YQ.N...I.....F...A.N-RF.NS.GHGA...
D14	Y.....Y.....H.....Y-KS.A...N.....
1E10	YD.V.....F.....SRN...RY.E.GS...

66	129
VH	KGRATISRONGQSTLRQLNNLRAEDTGTYCTKAYSS-GCDYEGG--YIDANGHGTEVIVSS
M3J5	V.....A..F..RSG.....NV...D.....
M3M1	D...V...S.....A...A.GV.GYC.SYSCC.VDT.....
D14	V.....A...TTG.GCSSG.RAE.....
1E10	V.....A...ARD.GG.-AYWFDA...-Q.....

[illegible]

FIG. 27

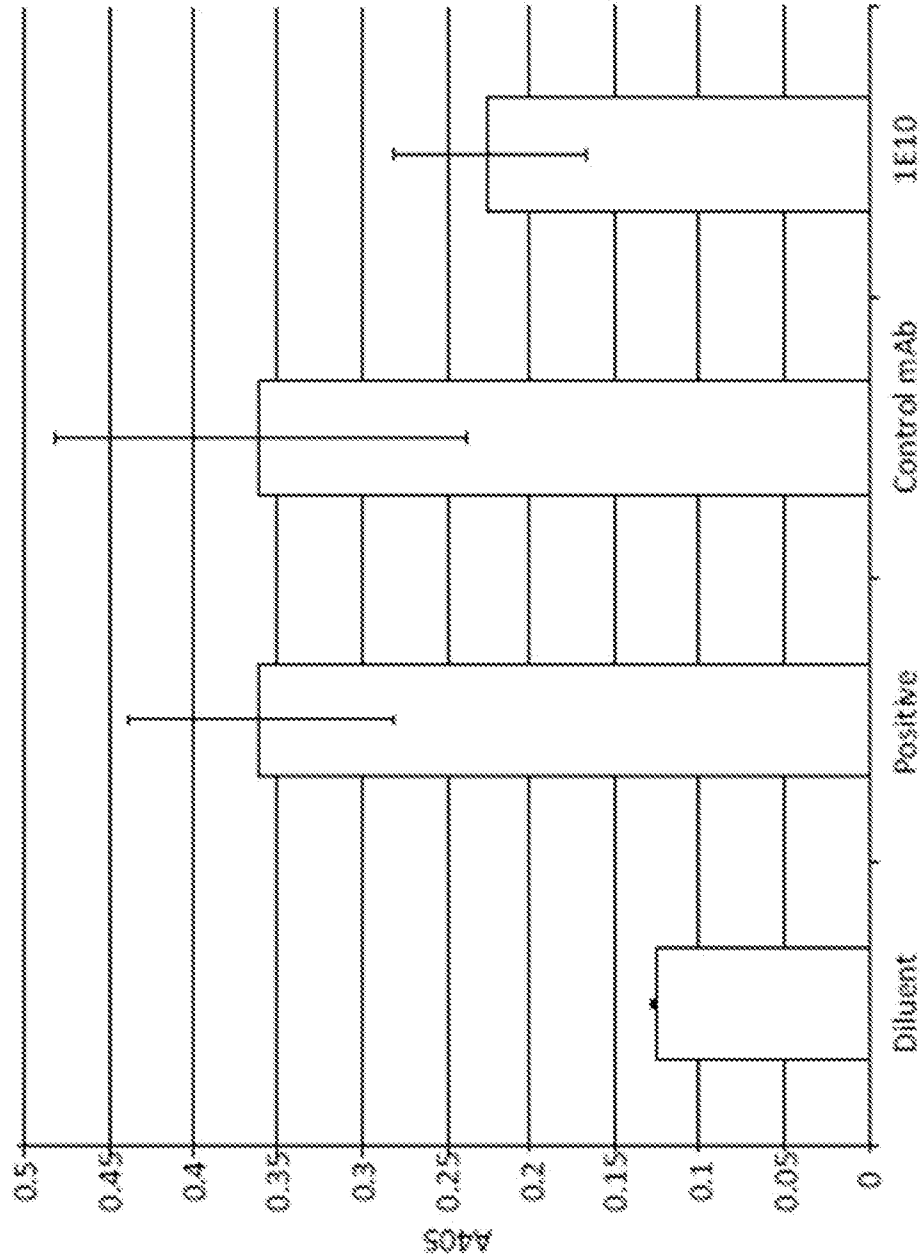


FIG. 28A

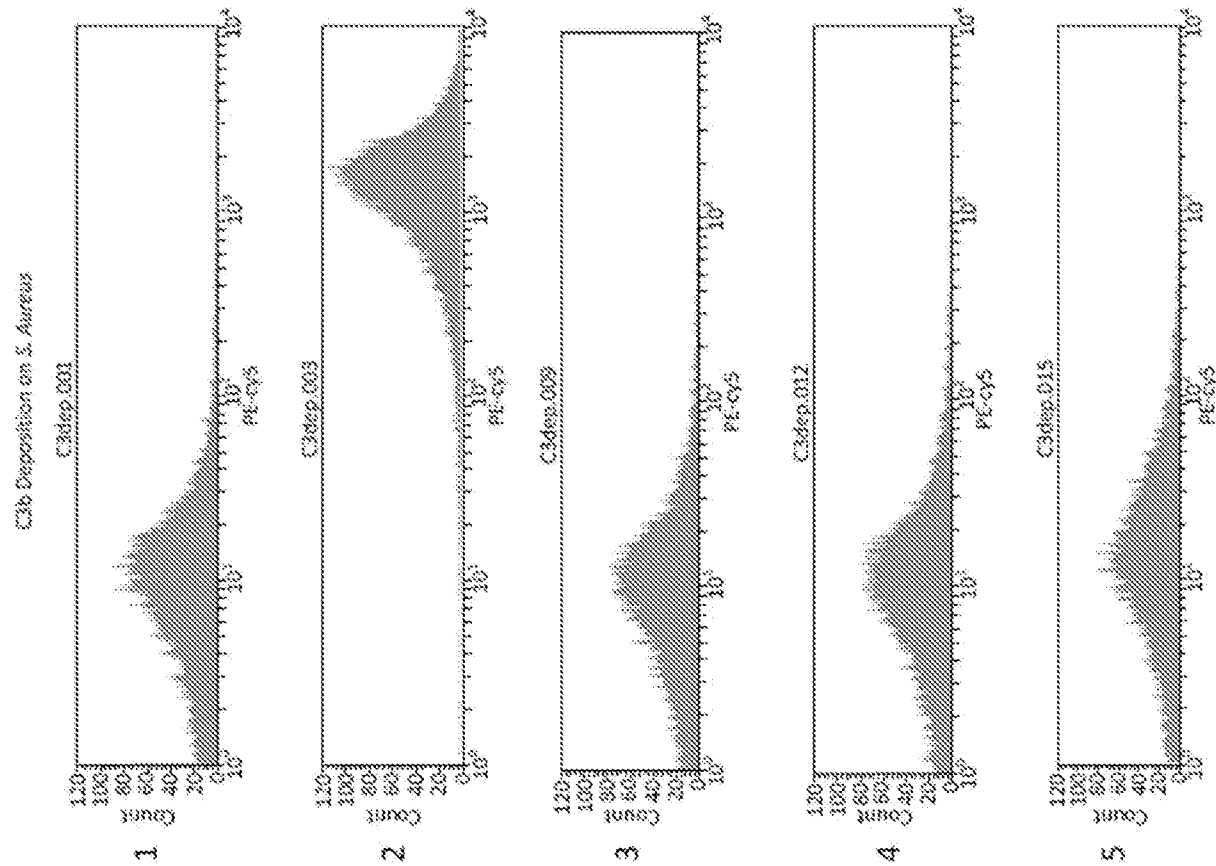


FIG. 28B

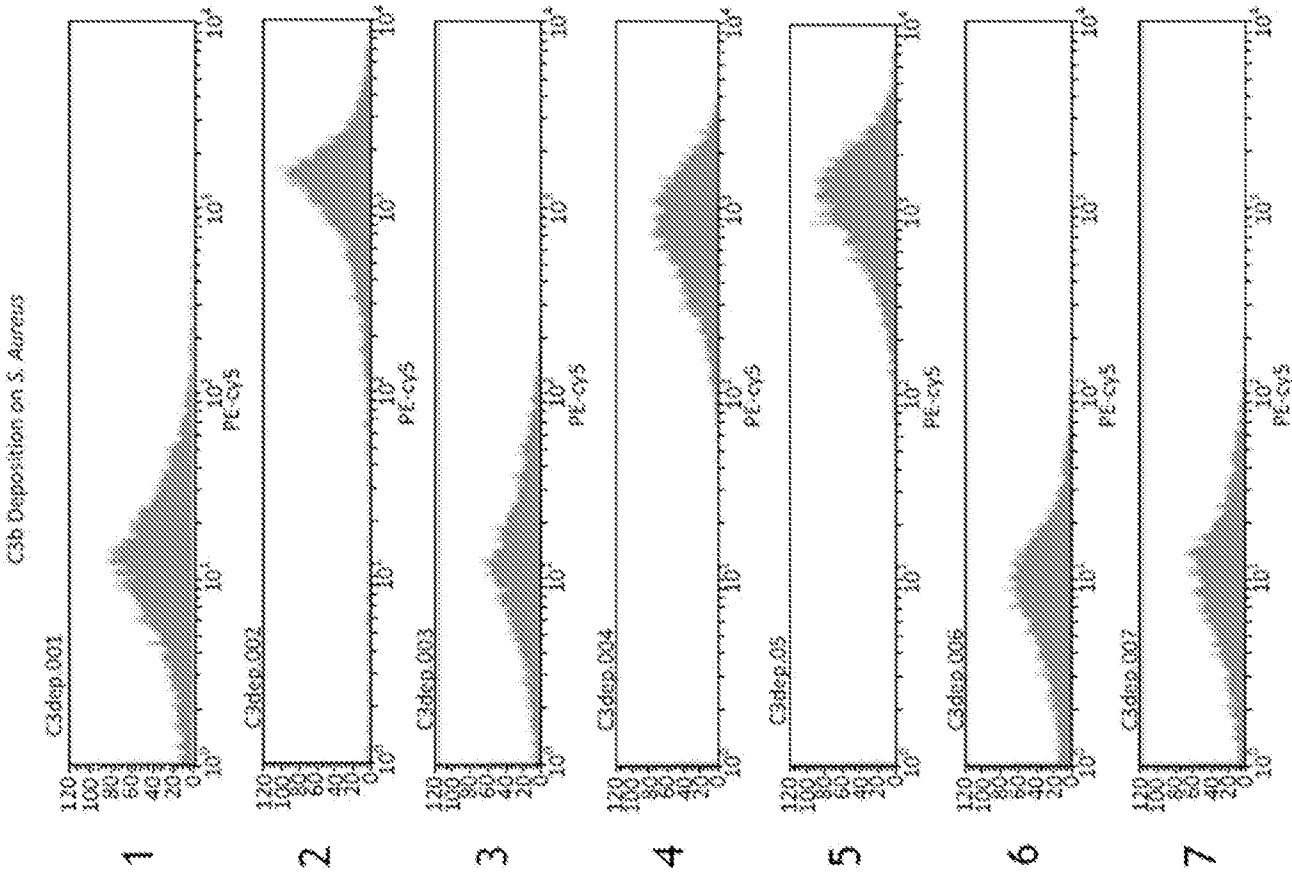


FIG. 29

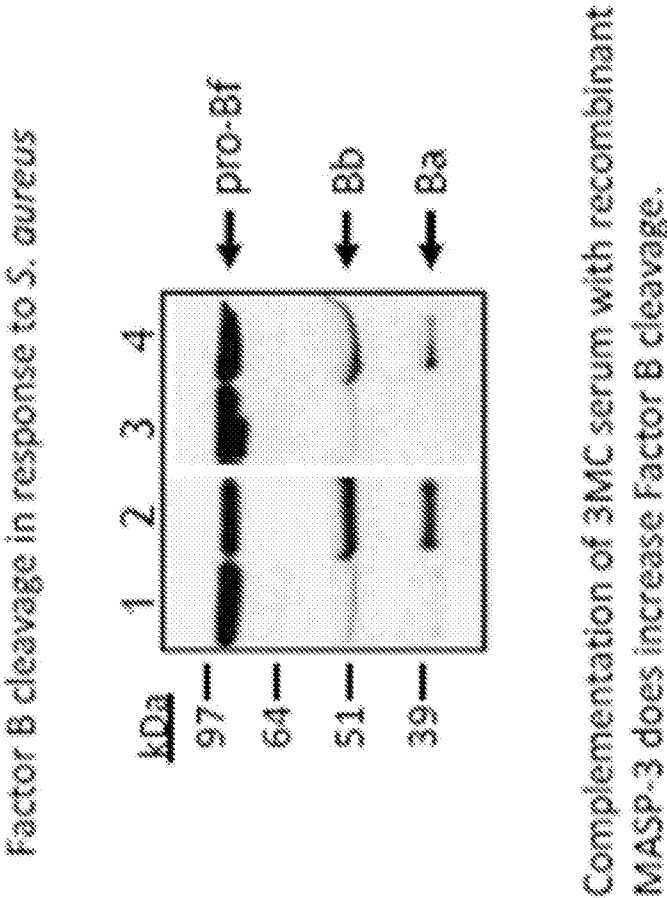


FIG. 30

MASP-3 directly cleaves pro-Bf

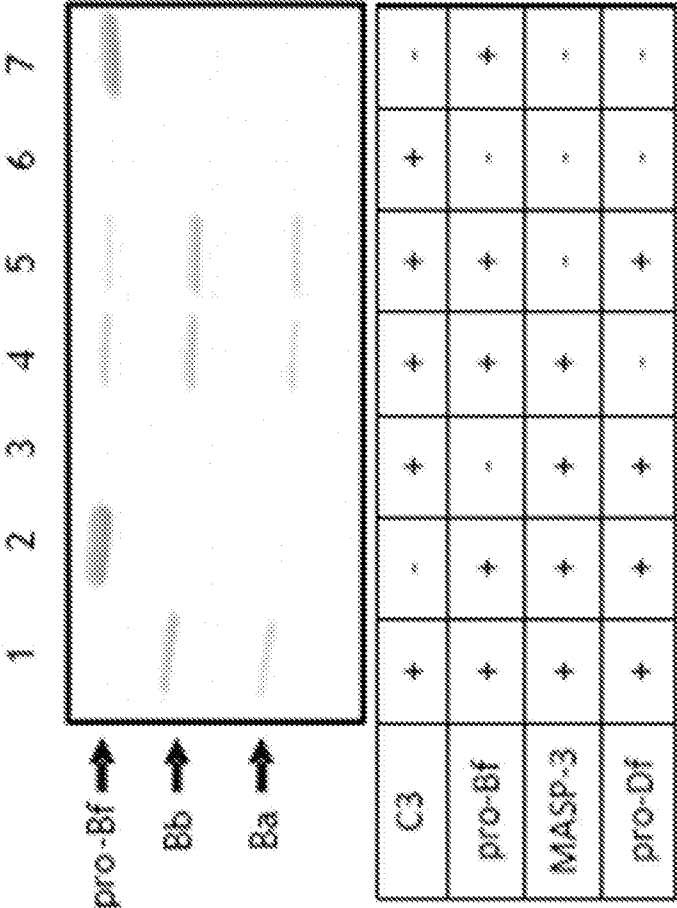


FIG. 31

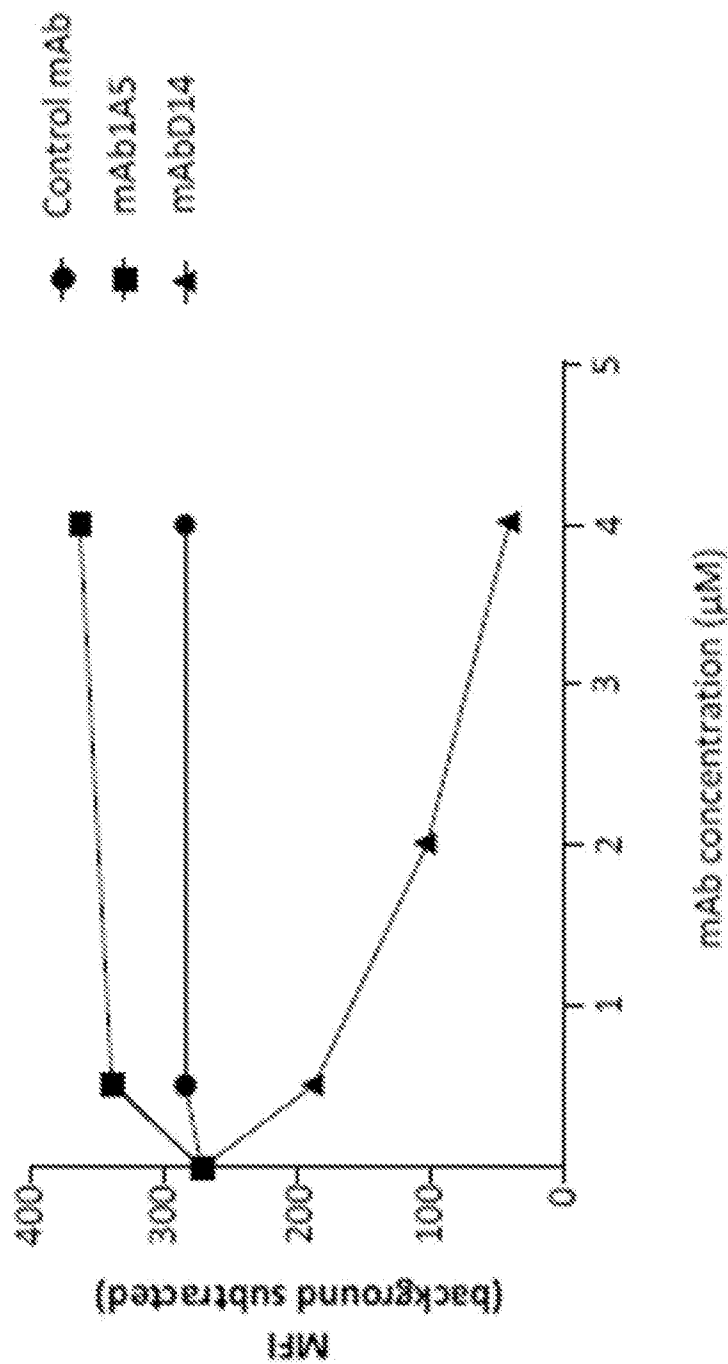


FIG. 32

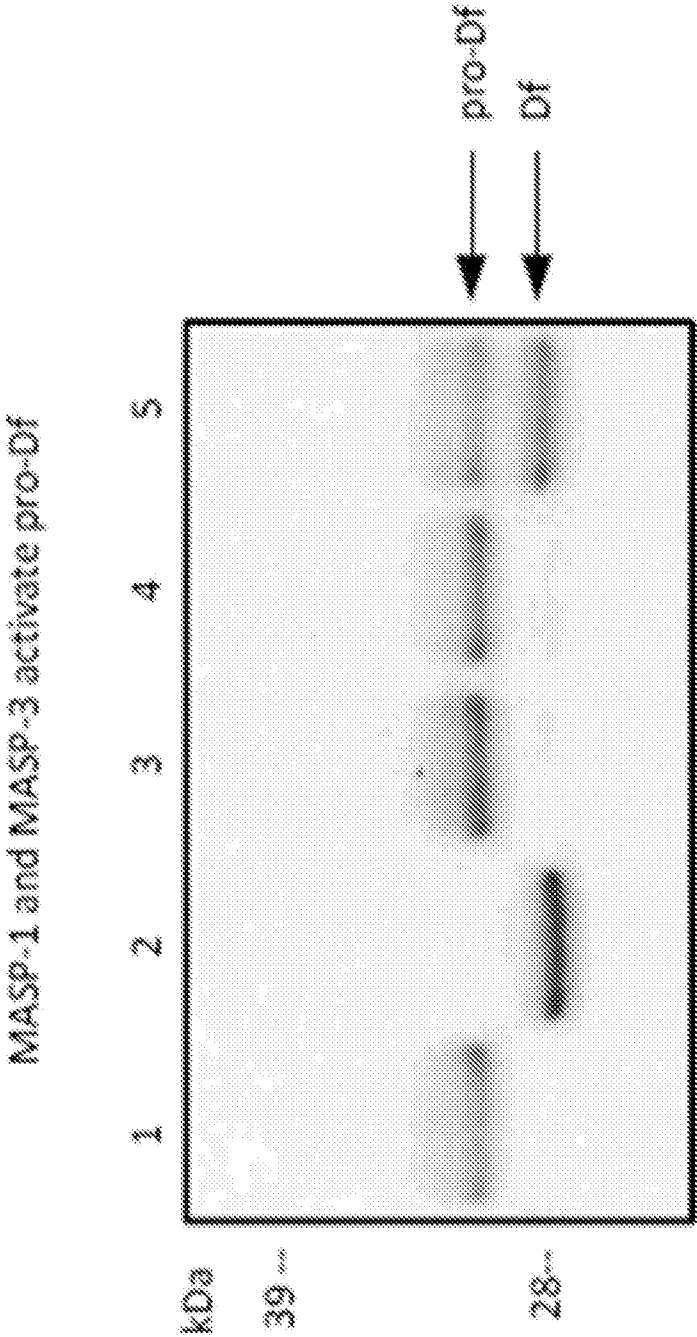


FIG. 33

Partial inhibition of MASP-3 dependent
Cleavage of Pro-Df with MASP-3 antibodies



FIG. 34

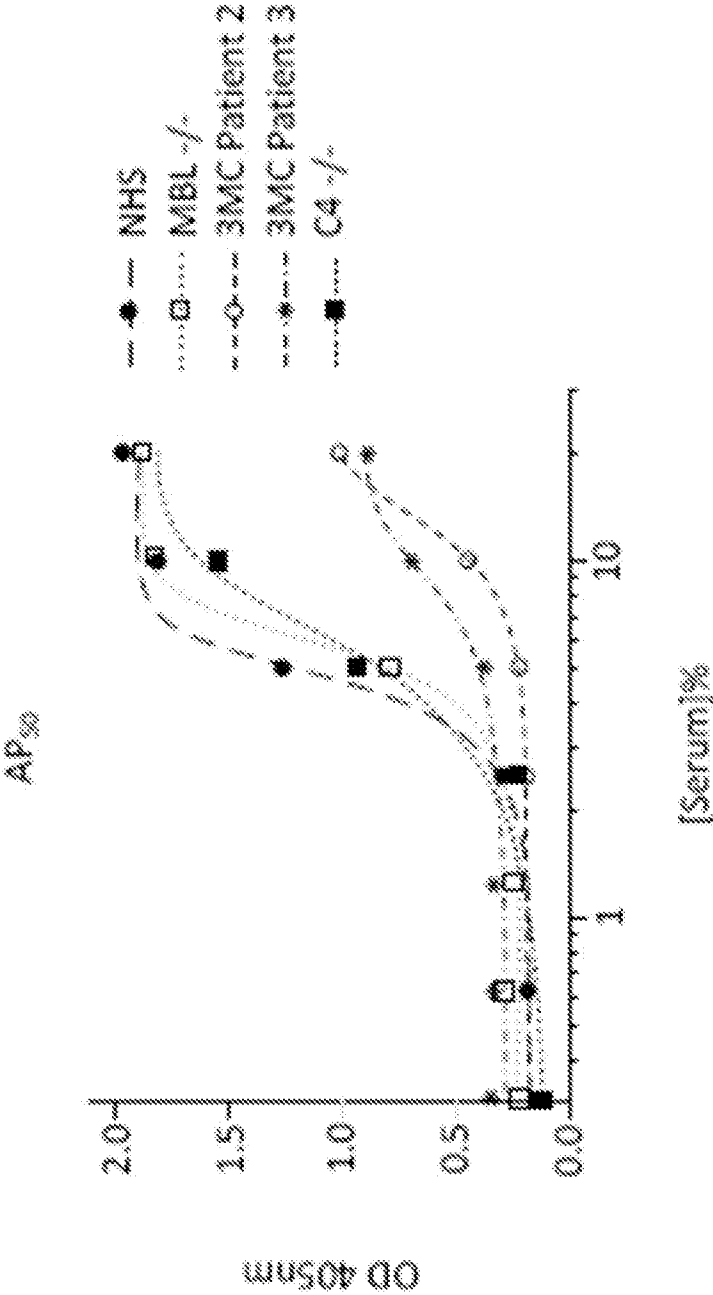


FIG. 35A

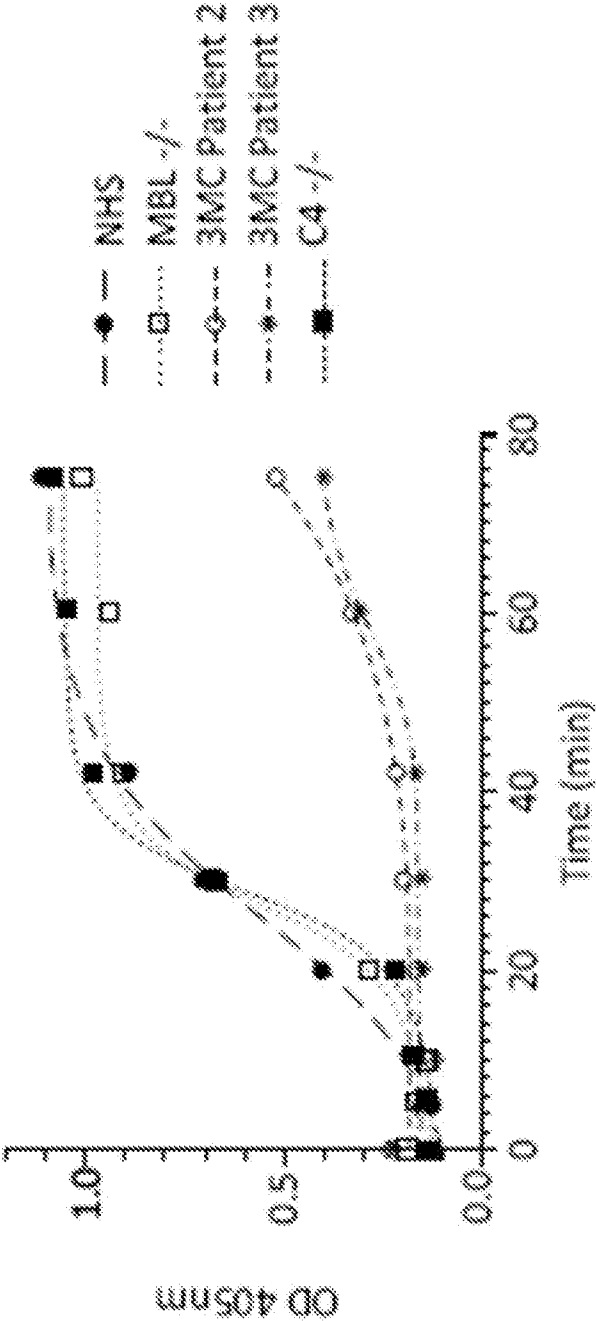


FIG. 35B

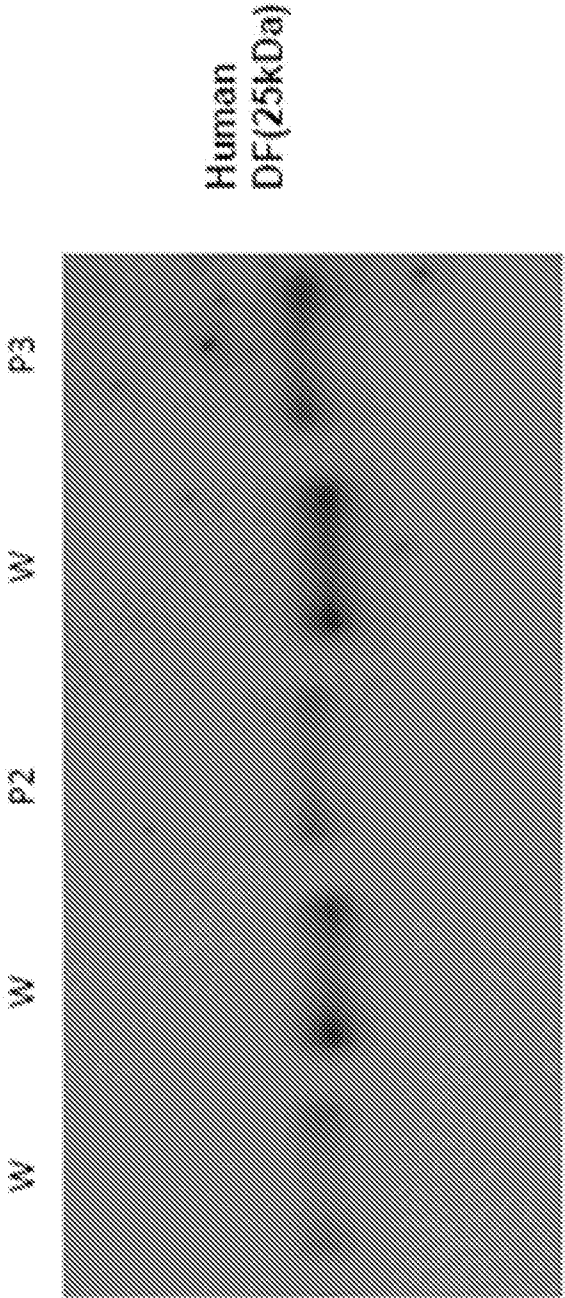


FIG. 35C

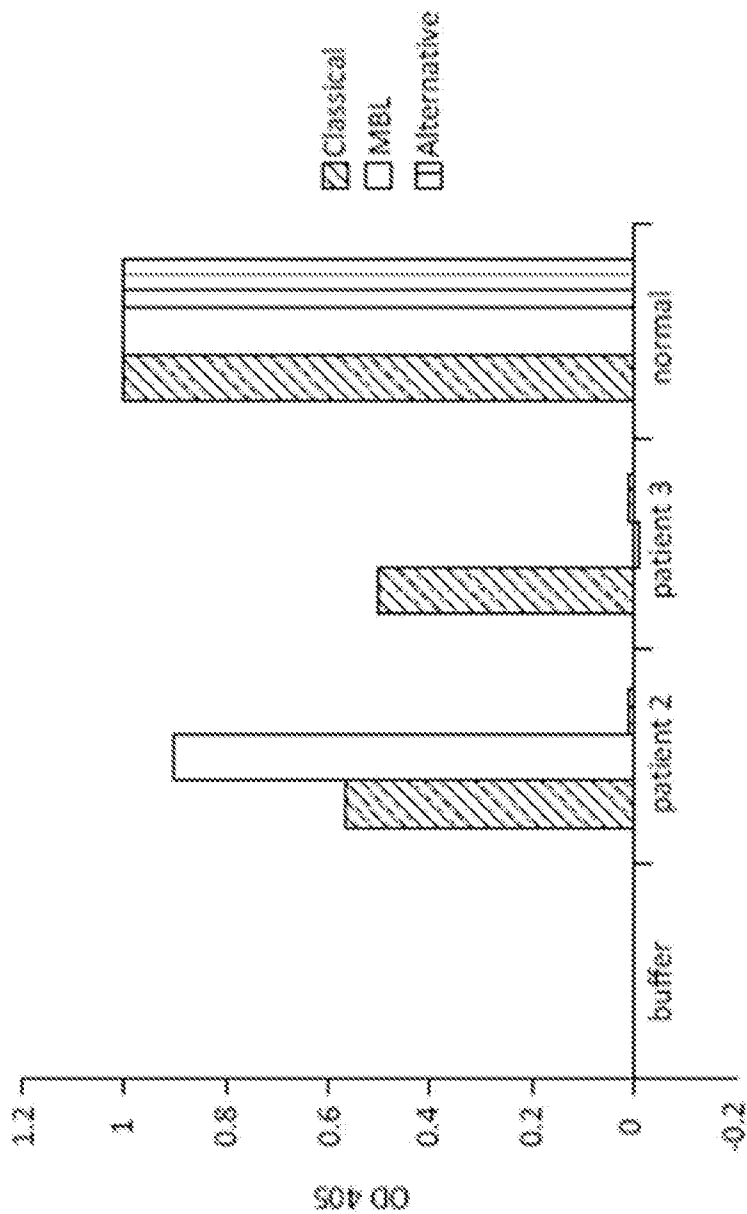


FIG. 36

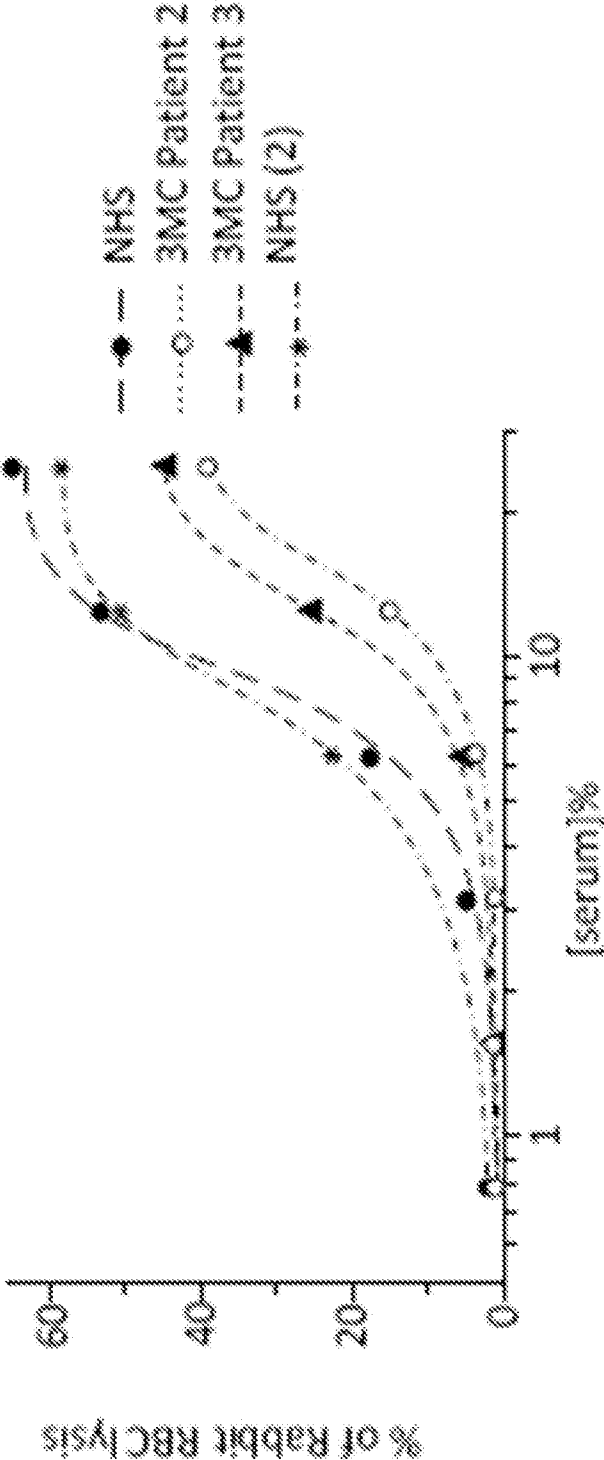


FIG. 37

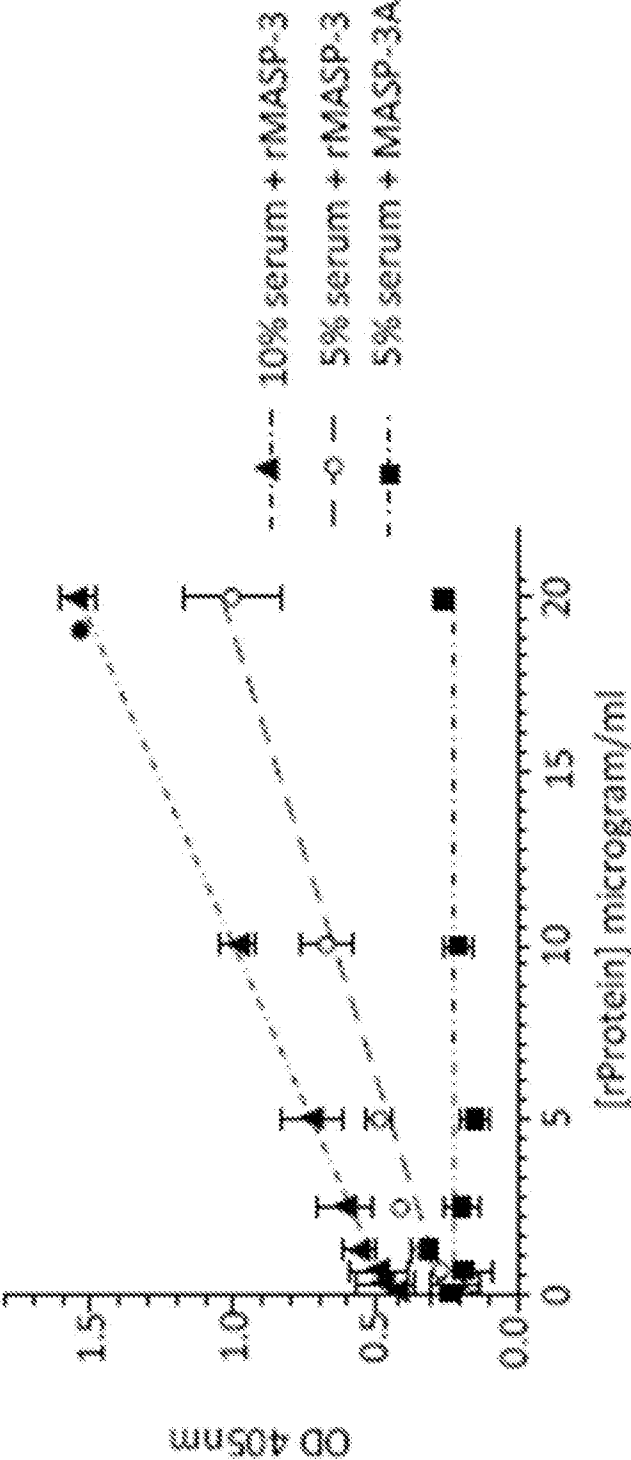


FIG. 38

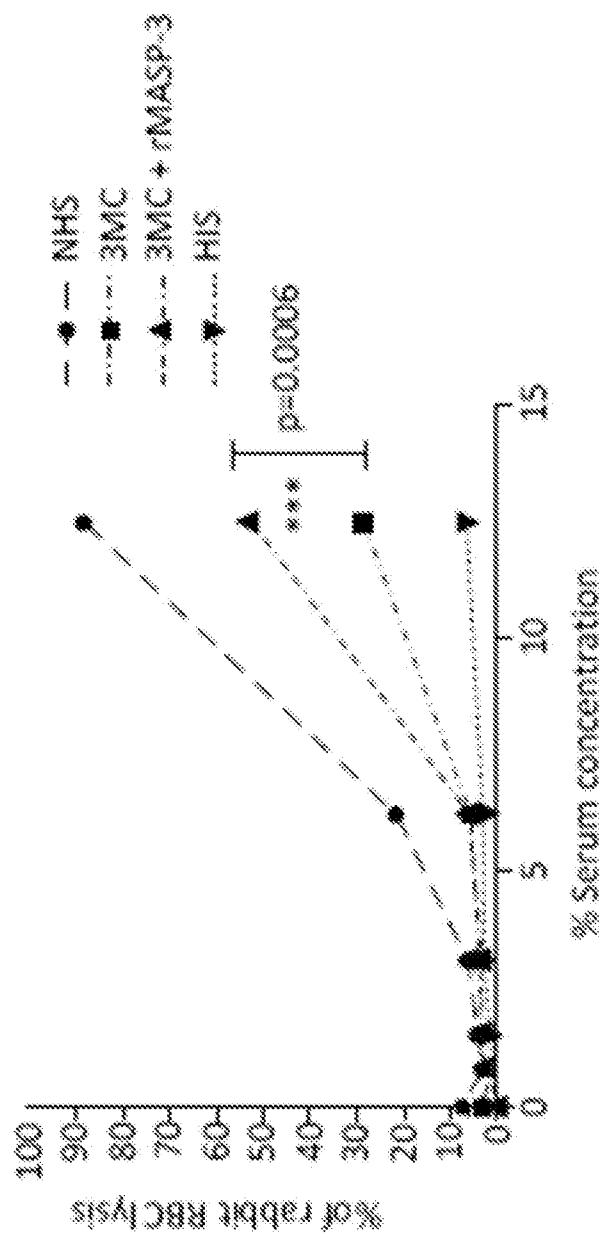


FIG. 39

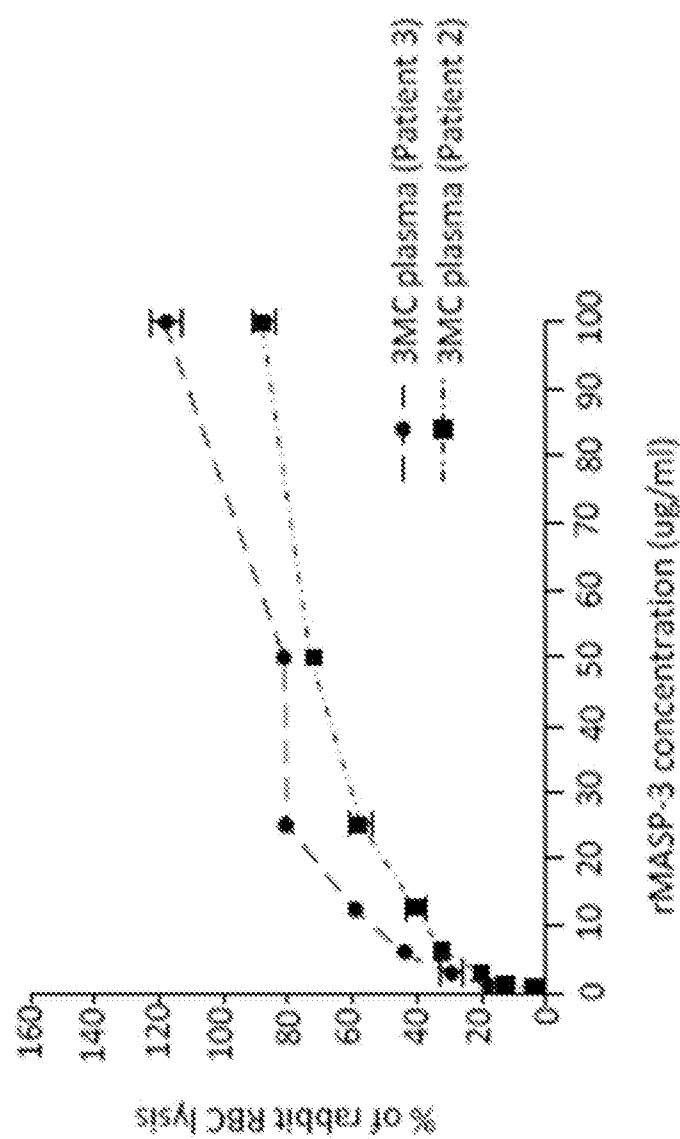


FIG. 40

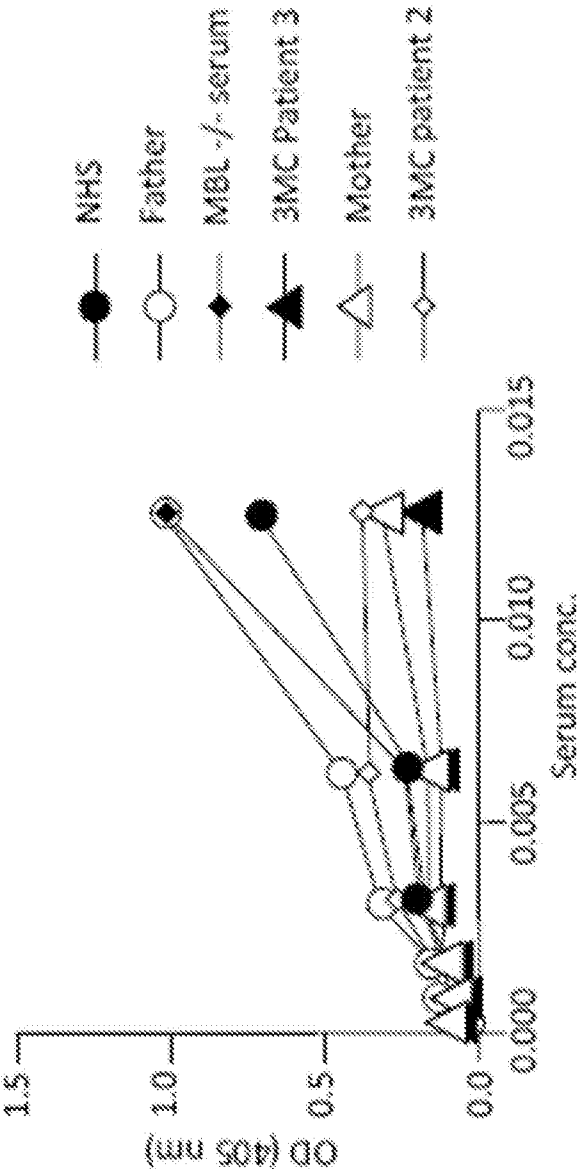


FIG. 41

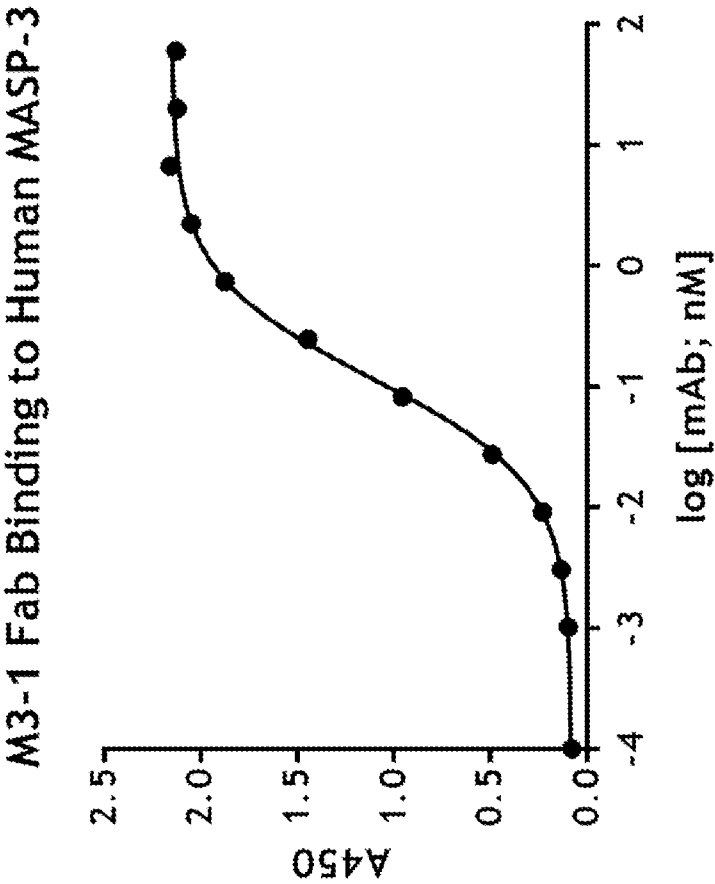


FIG. 42

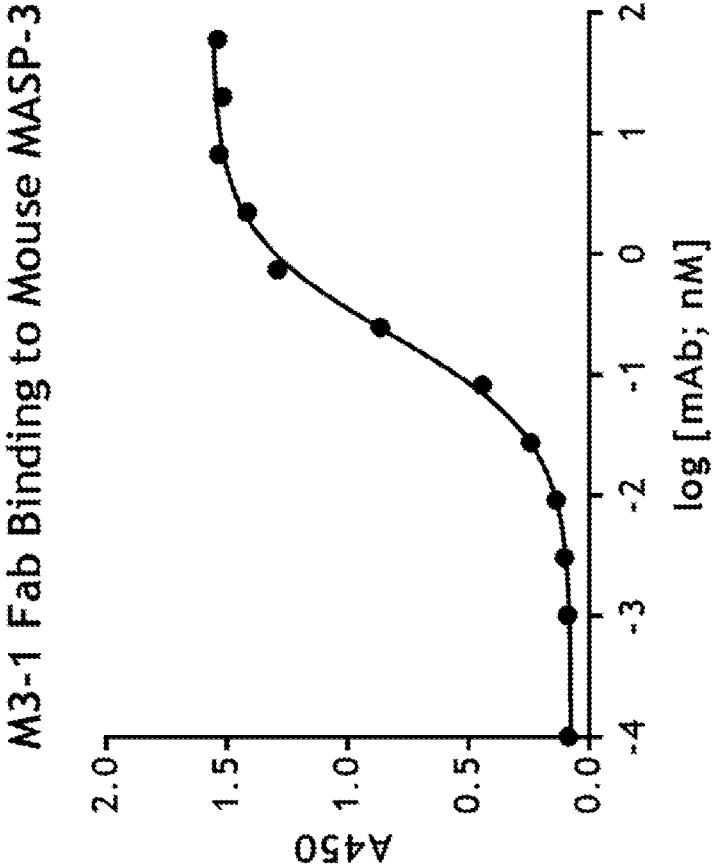


FIG. 43

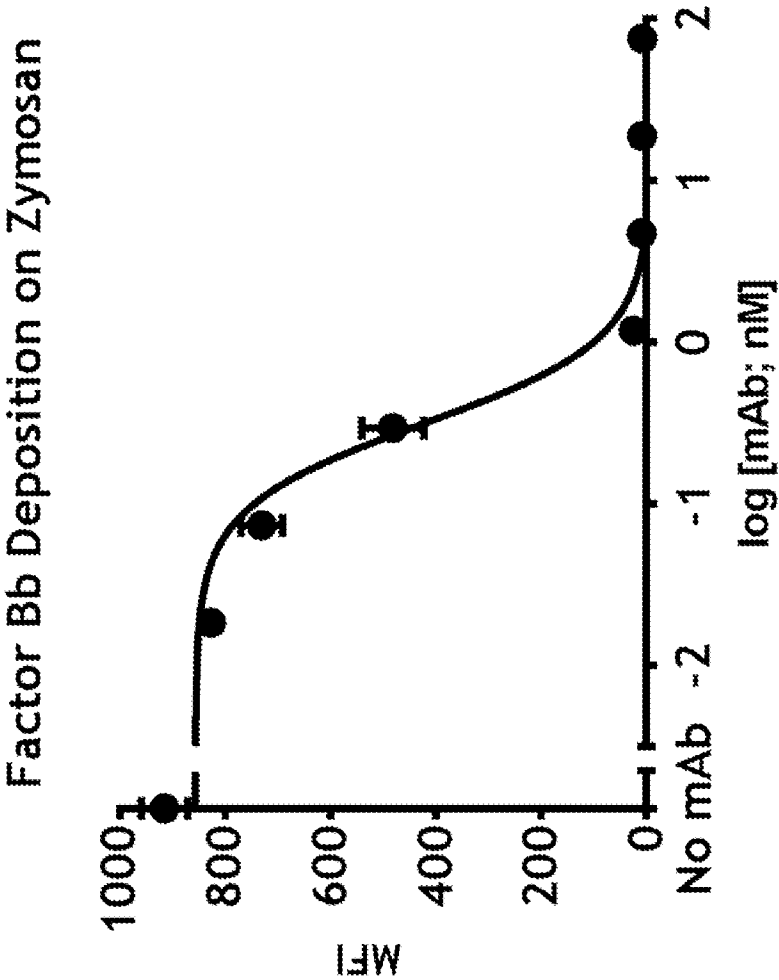


FIG. 44

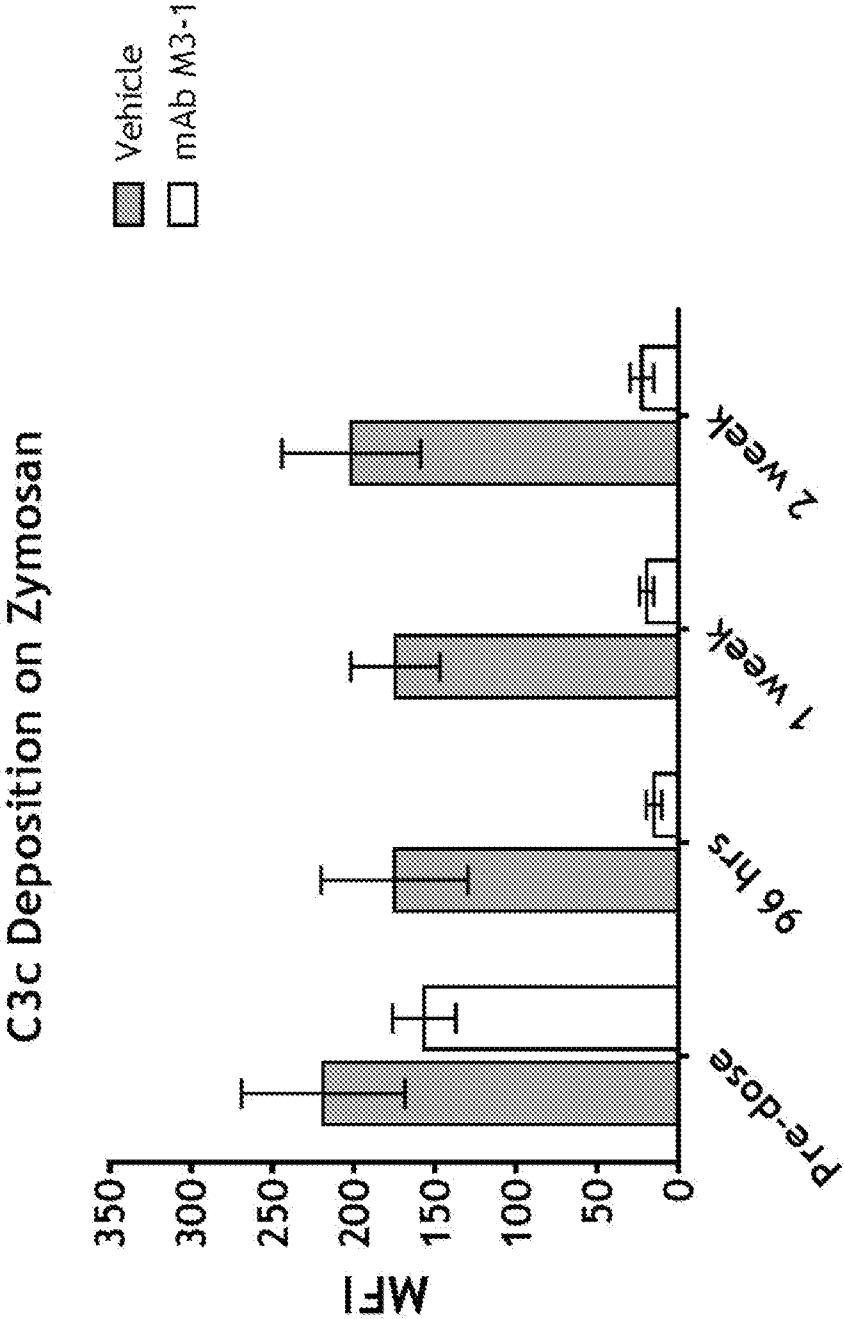


FIG. 45

Ongoing PNH Study

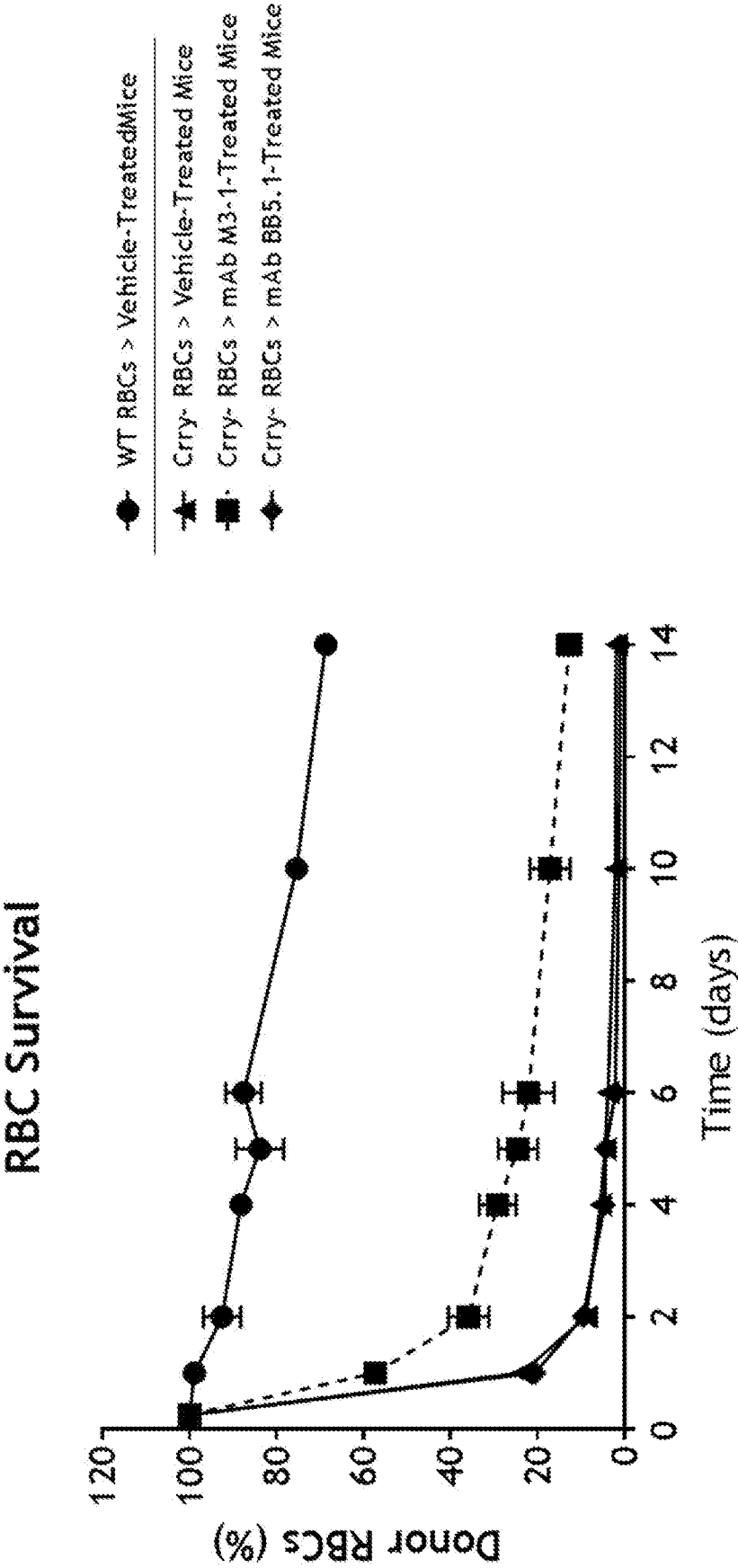


FIG. 46
RBC Survival in Treated Mice

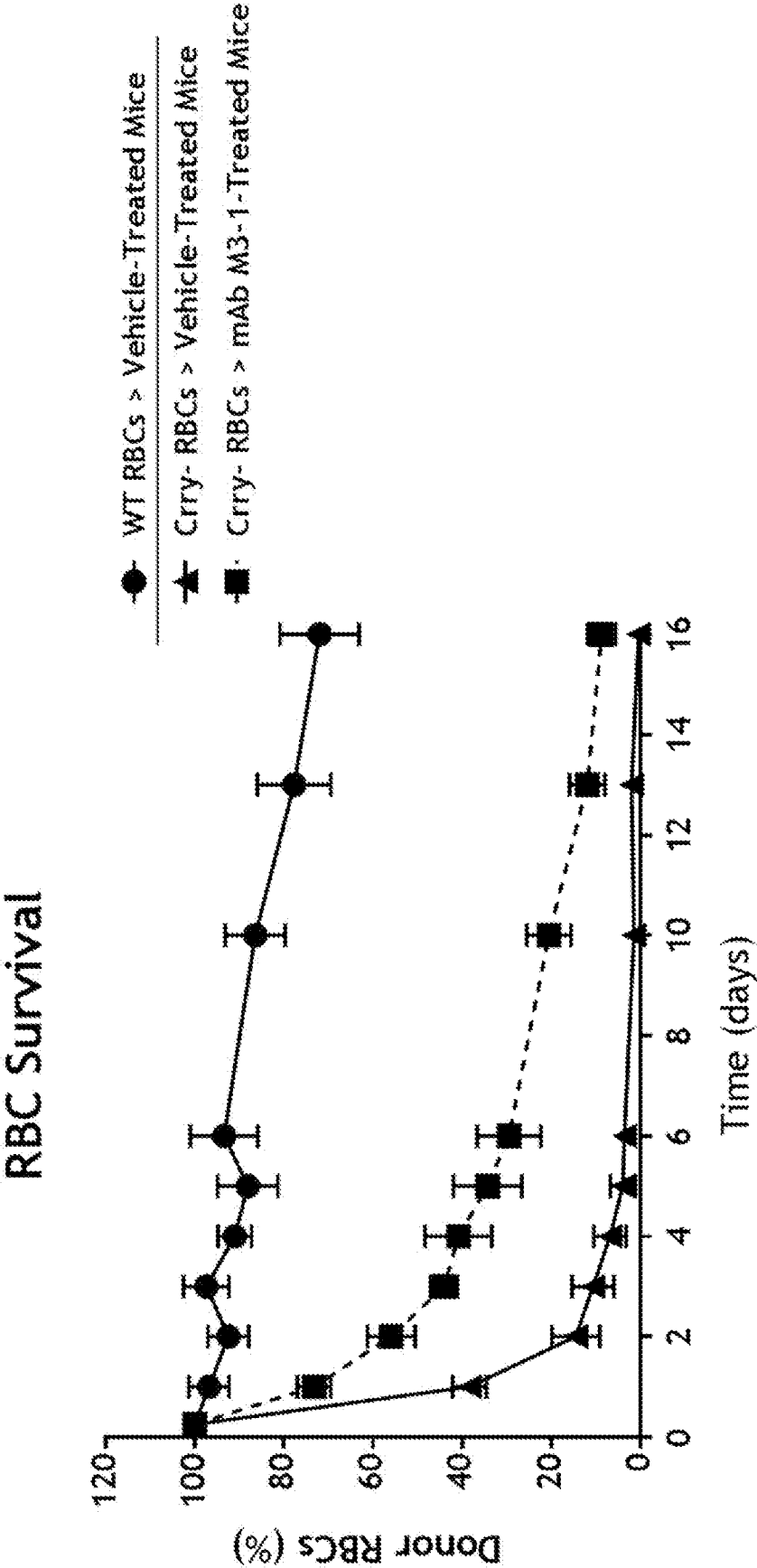


FIG. 47

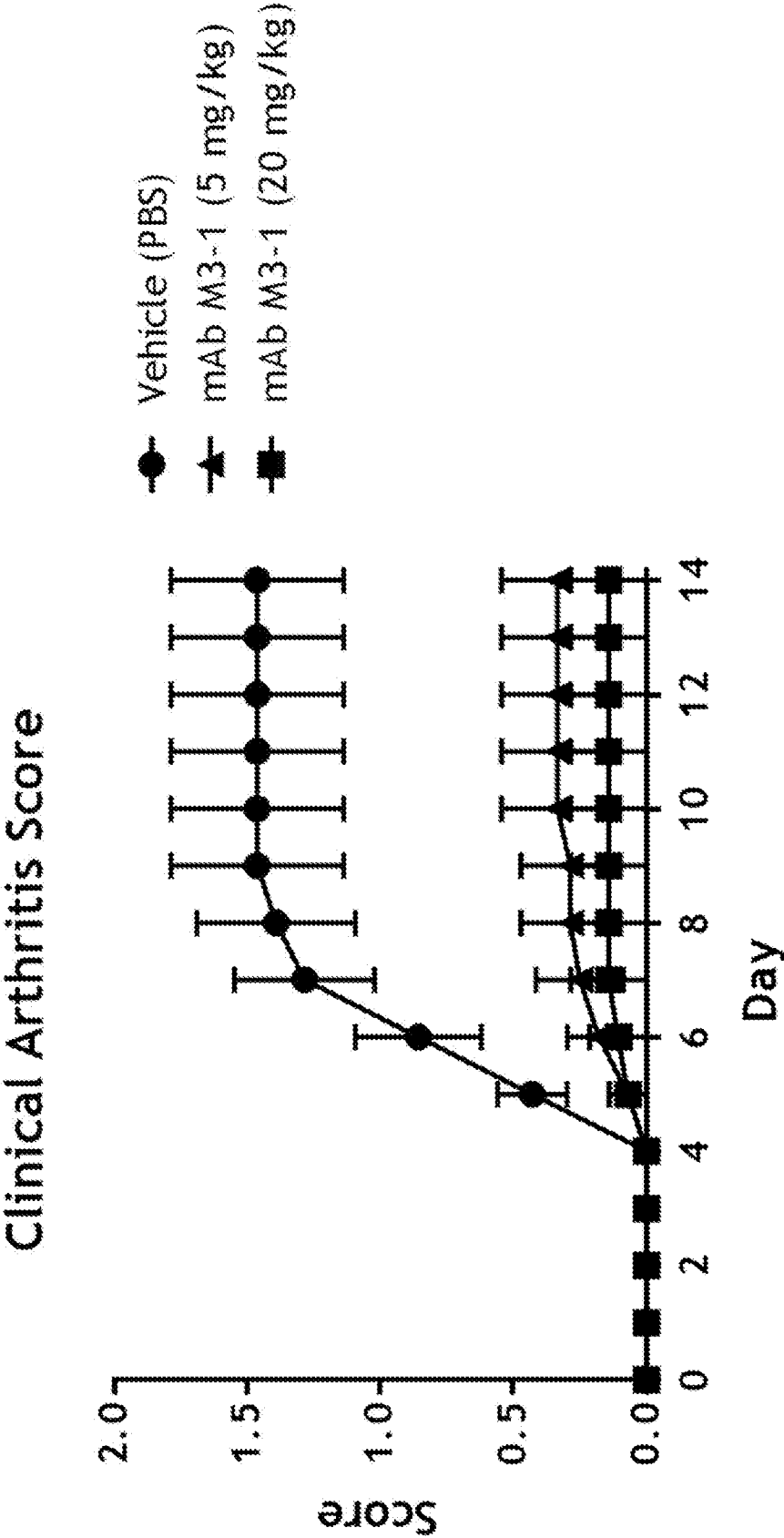
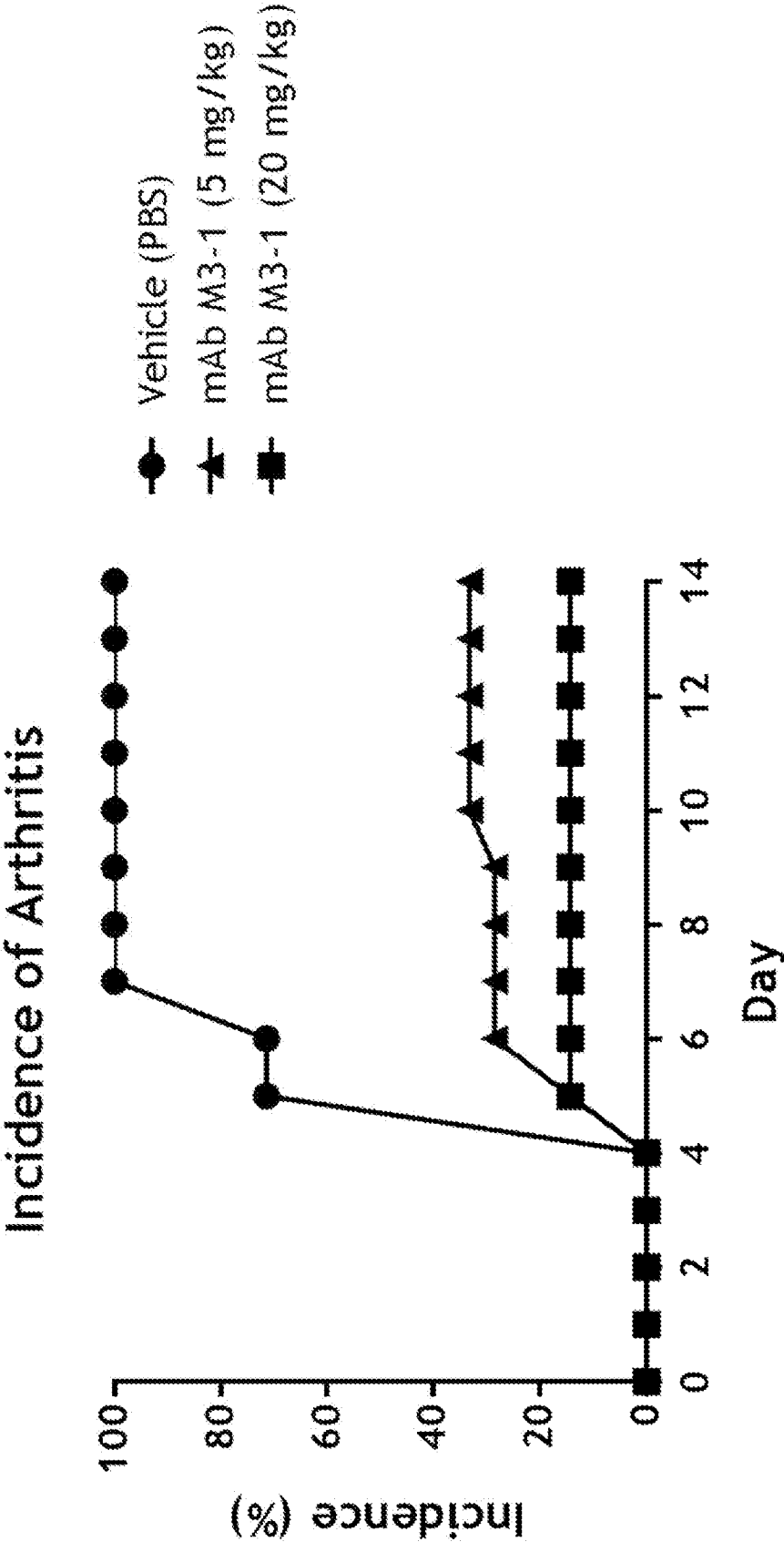


FIG. 48



High Affinity MASP-3 mAbs: Heavy Chain Variable Regions

[illegible]

FIG. 49B
High Affinity MASP-3 mAbs: Light Chain Variable Regions

Seq	24	34	40	58	69	87	
184_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:41)
484_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:42)
486_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:43)
186_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:44)
184_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:45)
484_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:46)
486_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:47)
186_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:48)
184_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:49)
484_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:50)
486_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:51)
186_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:52)
184_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:53)
484_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:54)
486_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:55)
186_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:56)
184_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:57)
484_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:58)
486_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:59)
186_VL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	LVNAGGNT	WYVGGGGL	(829:60)

FIG. 50A

Dendrogram of the VH regions of High Affinity MASP-3 mAbs

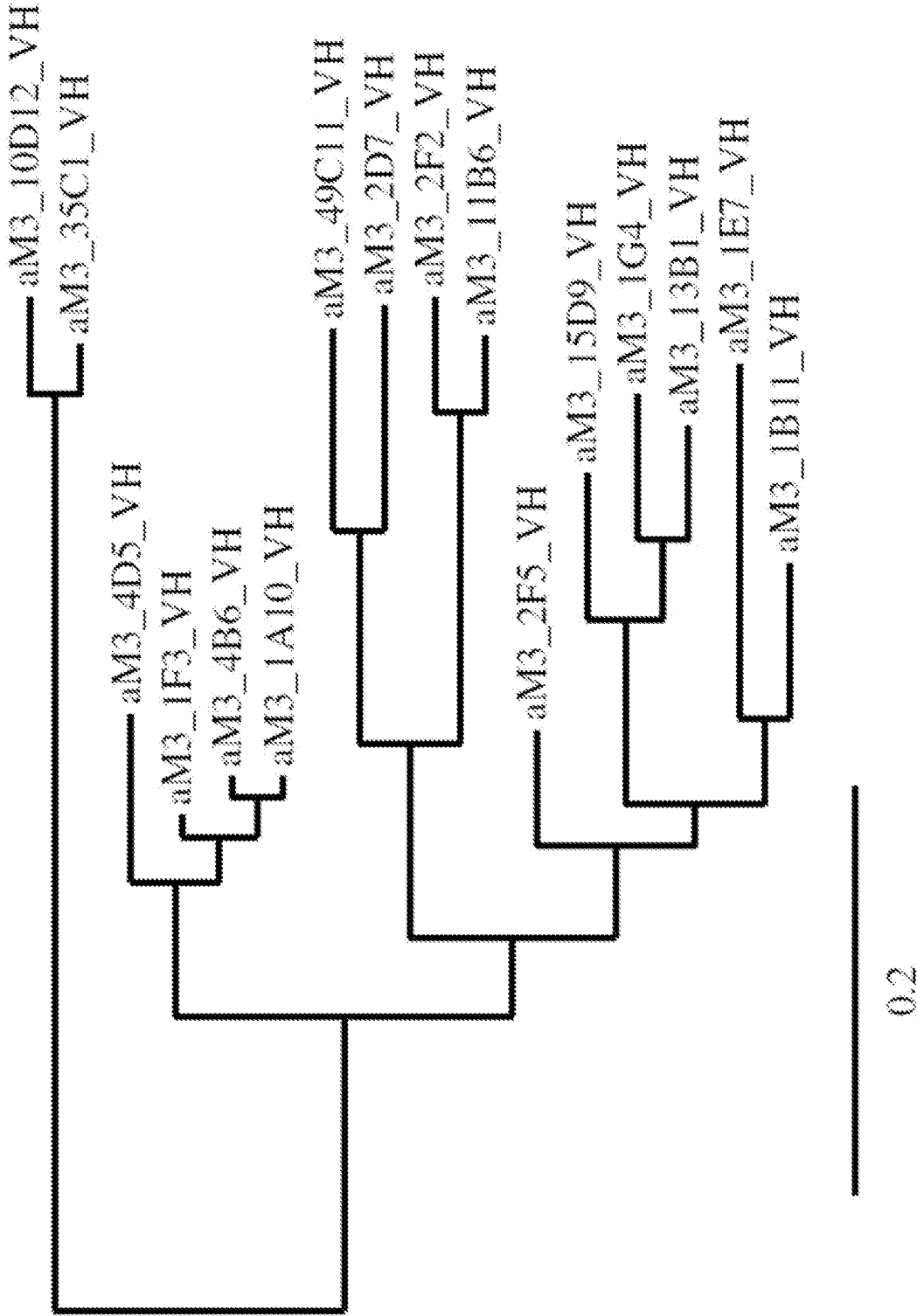


FIG. 50B

Dendrogram of the VL regions of High Affinity MASP-3 mAbs

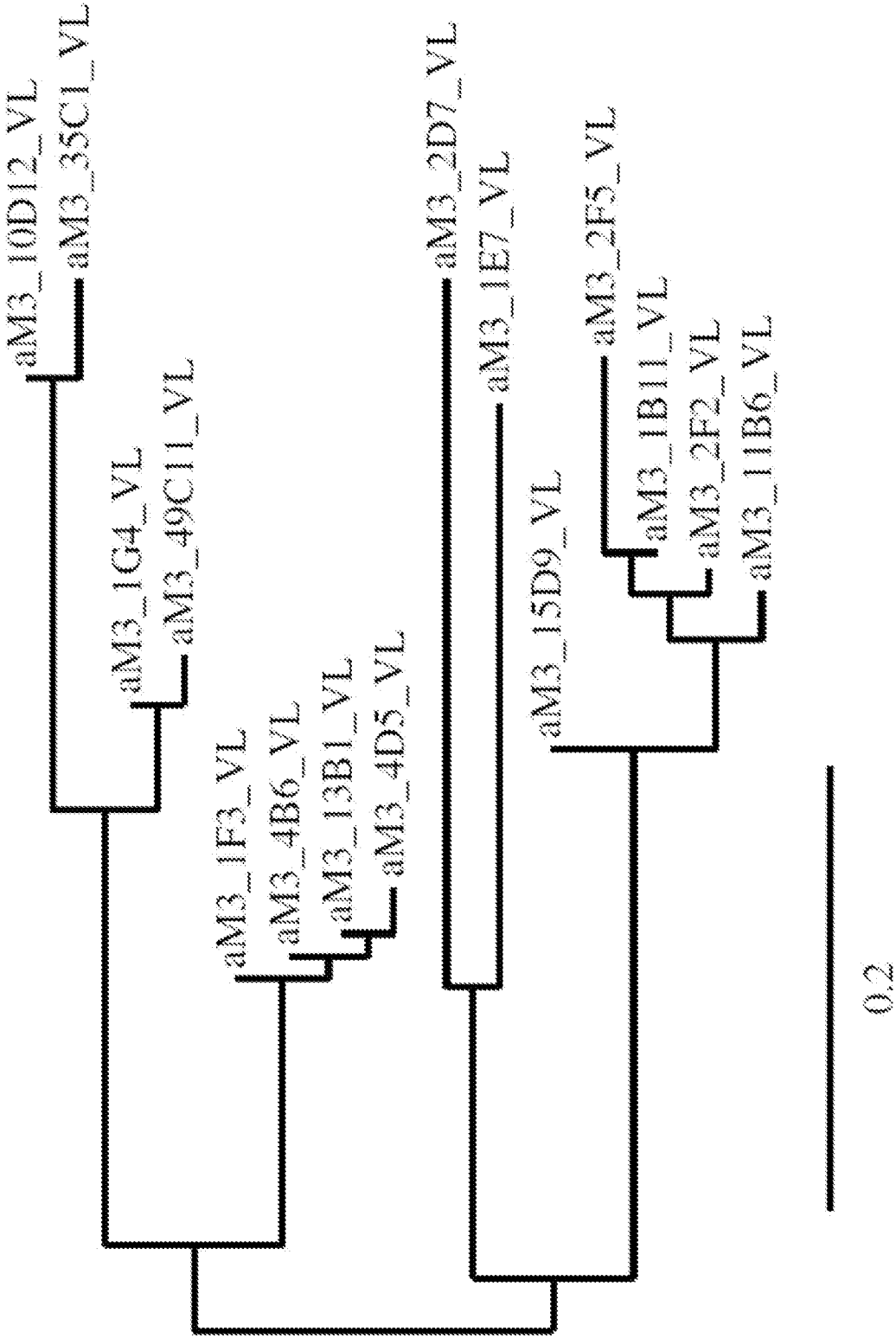


FIG. 51A

Binding of purified recombinant MASP-3 mAbs to human MASP-3

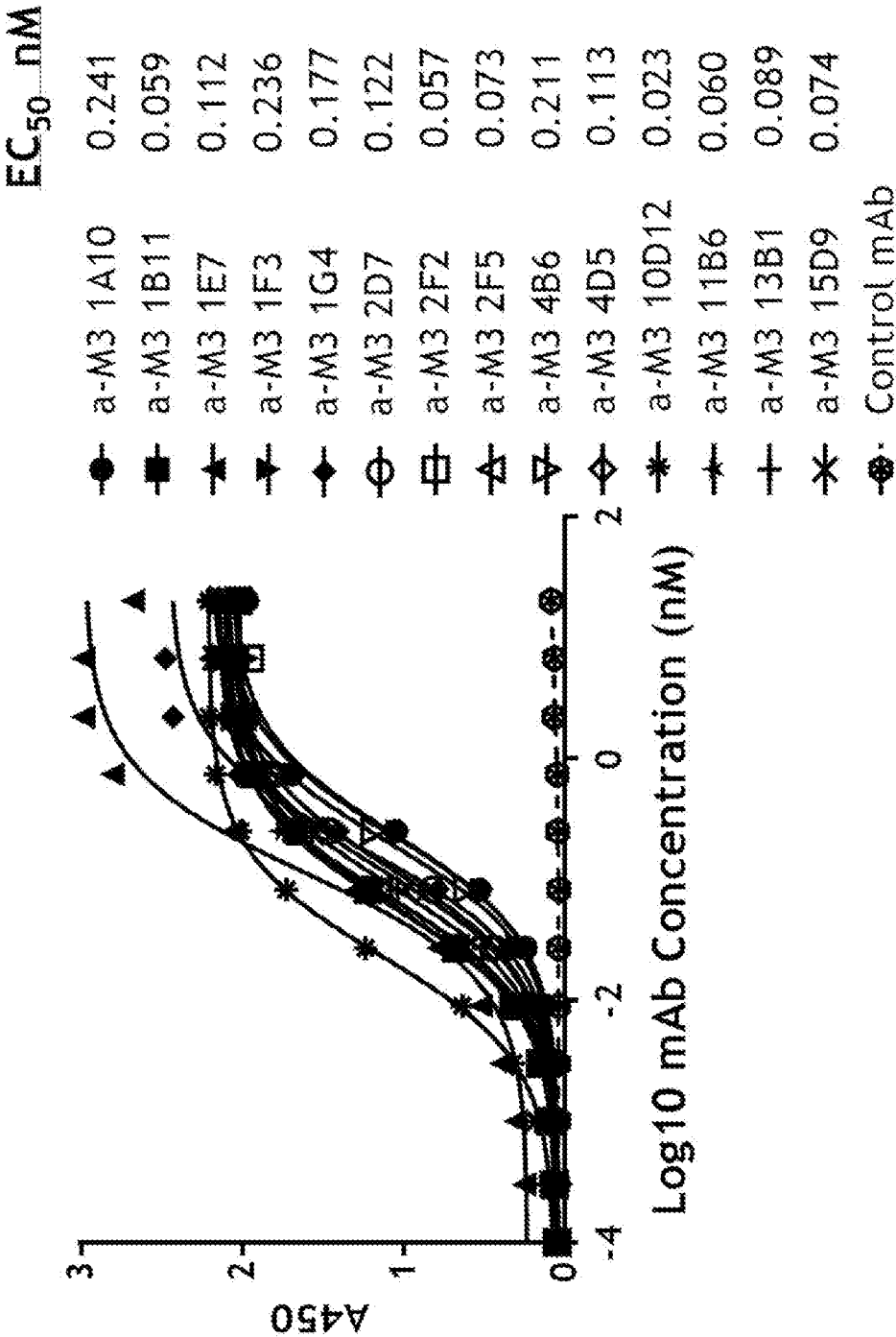


FIG. 51B

Binding of purified recombinant MASP-3 mAbs to human MASP-3

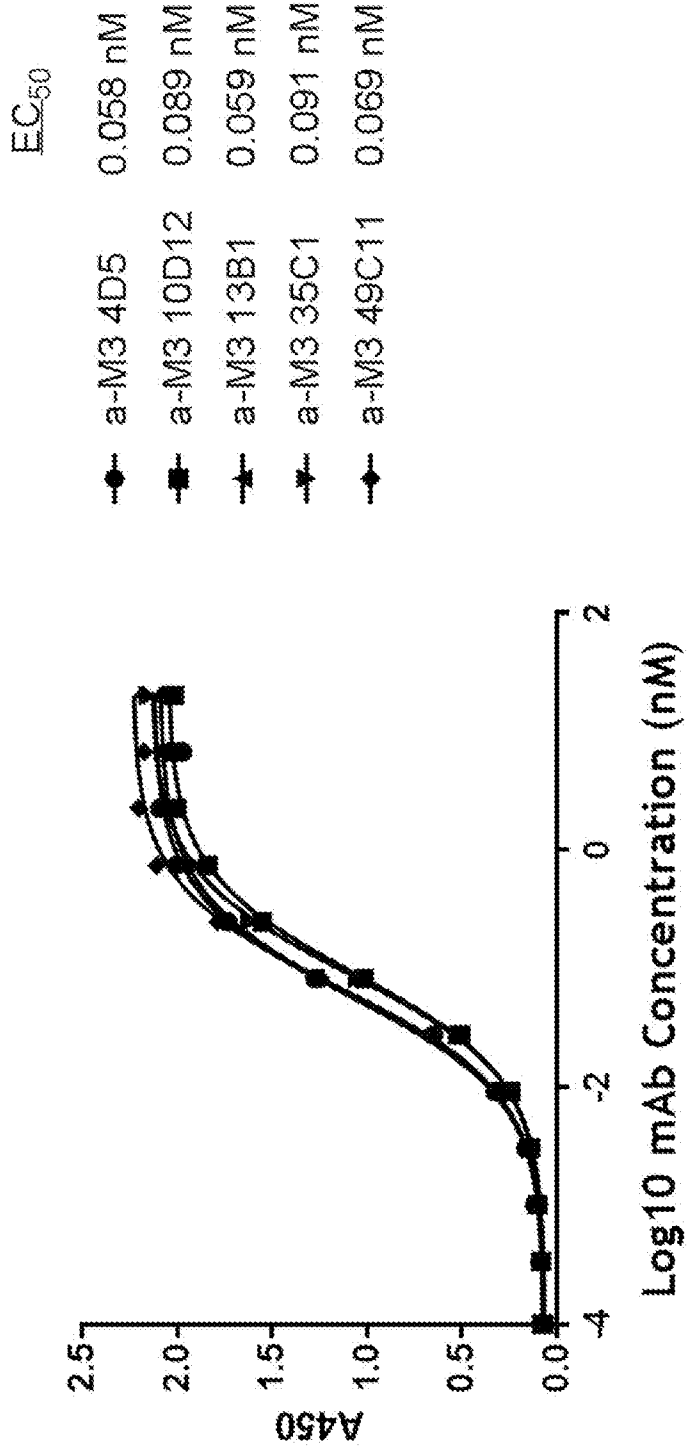


FIG. 51C

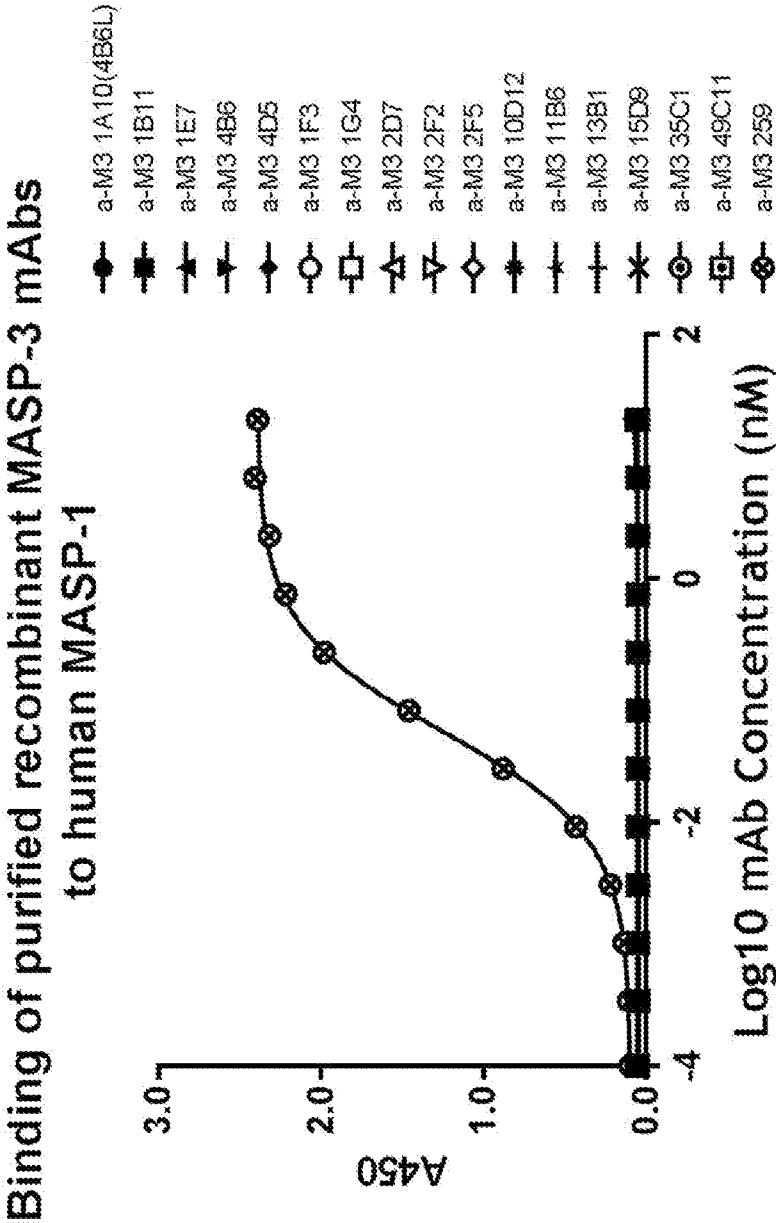


FIG. 51D

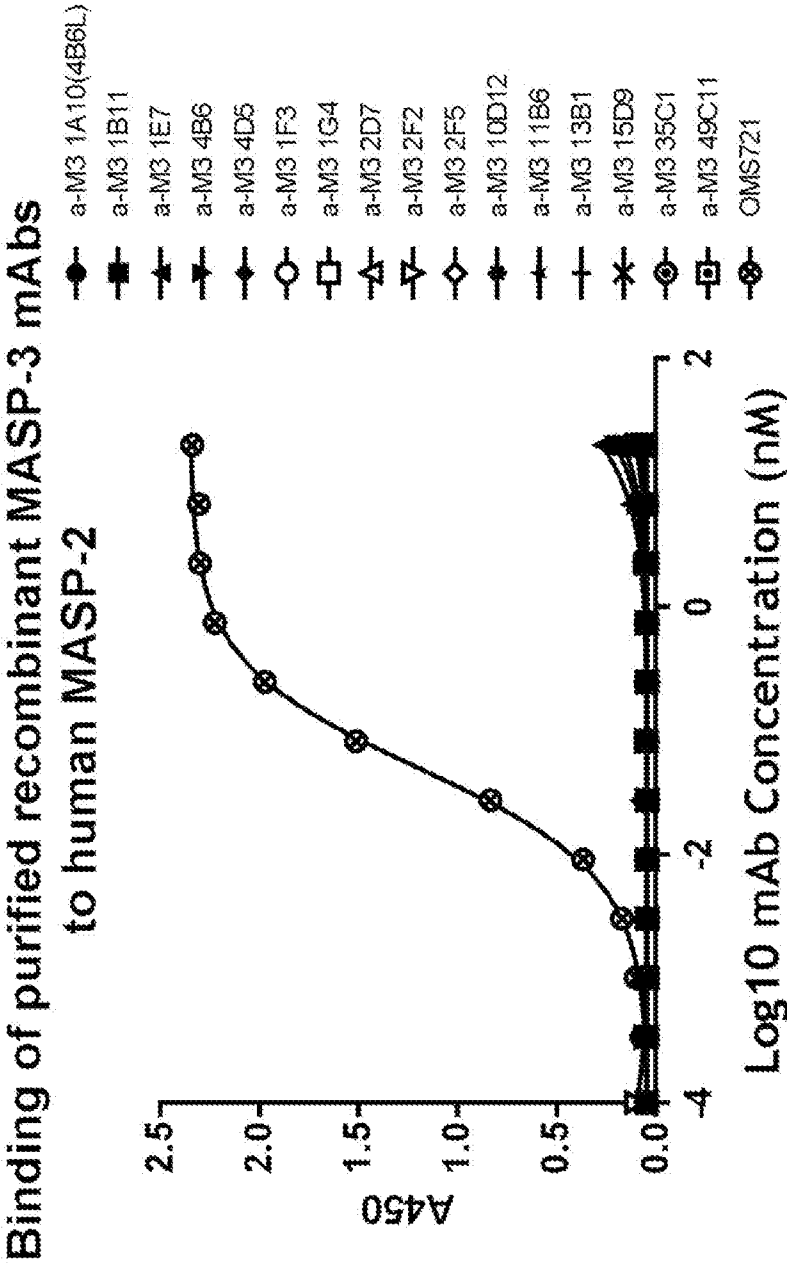


FIG. 52
Binding of MASP-3 mAbs to full-length mouse MASP-3

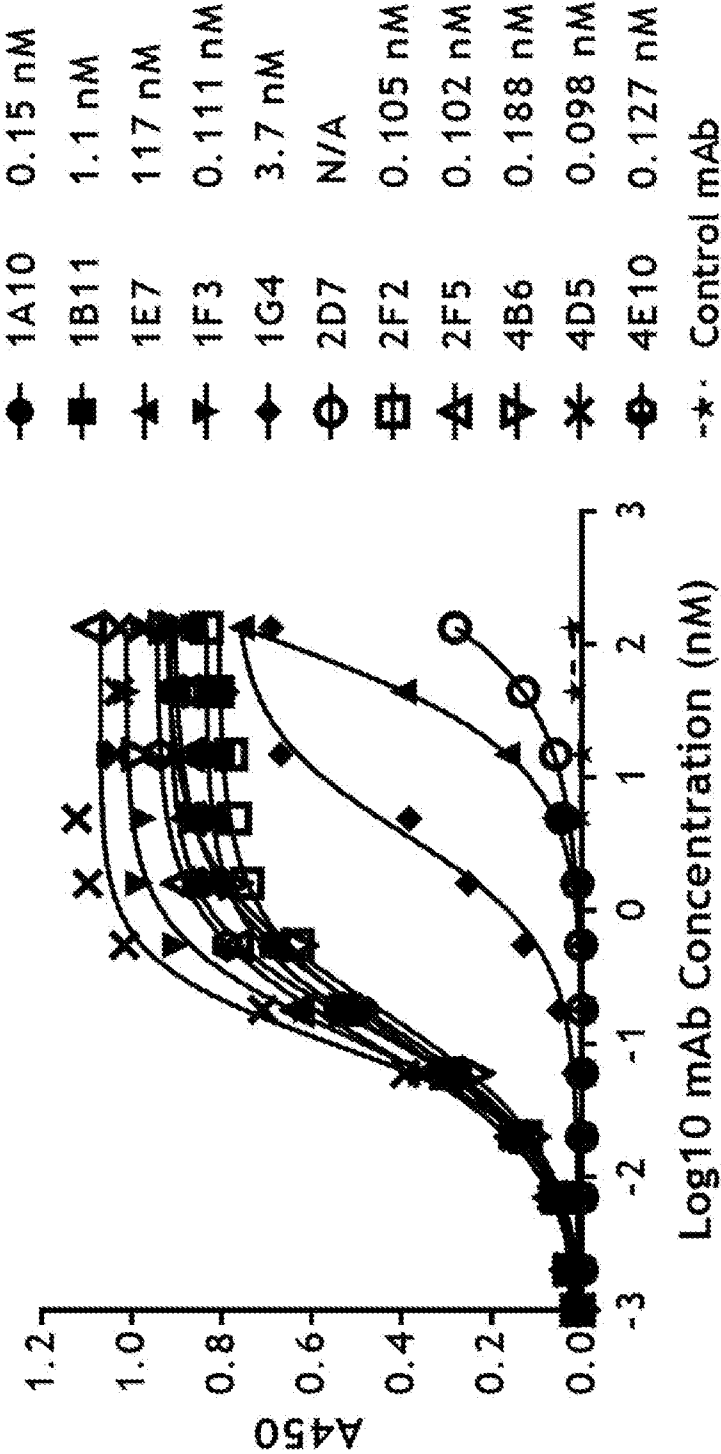


FIG. 54
High Affinity MASP-3 mAbs inhibit CFD Cleavage by rMASP-3

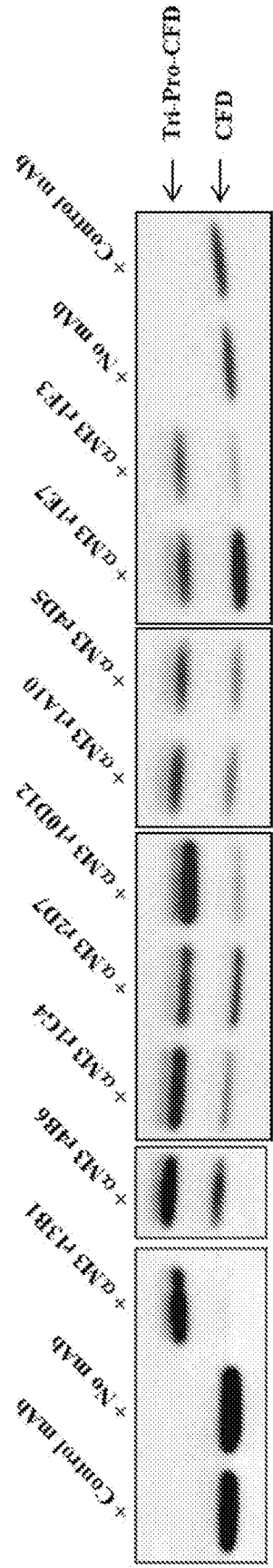
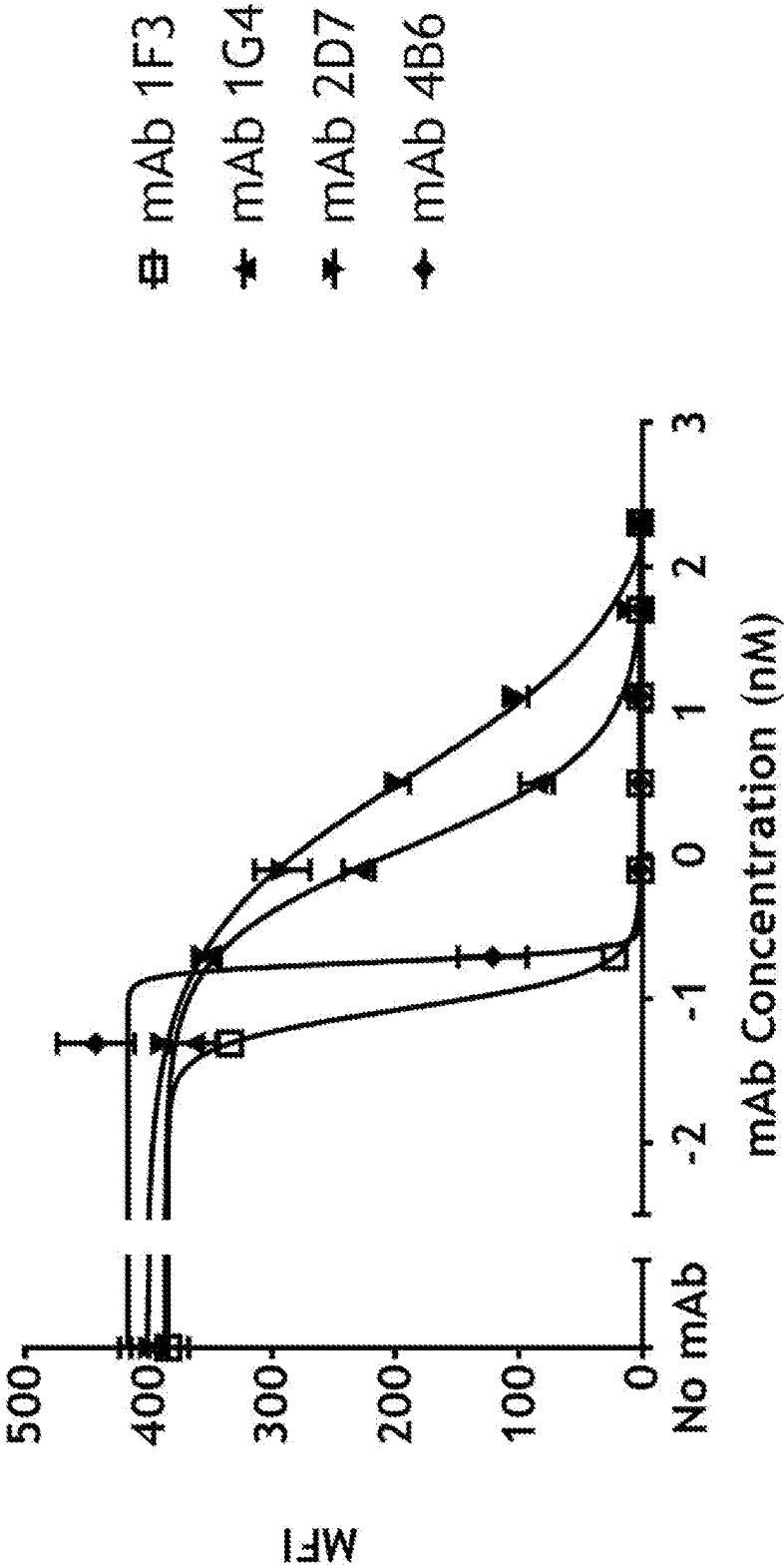


FIG. 55A

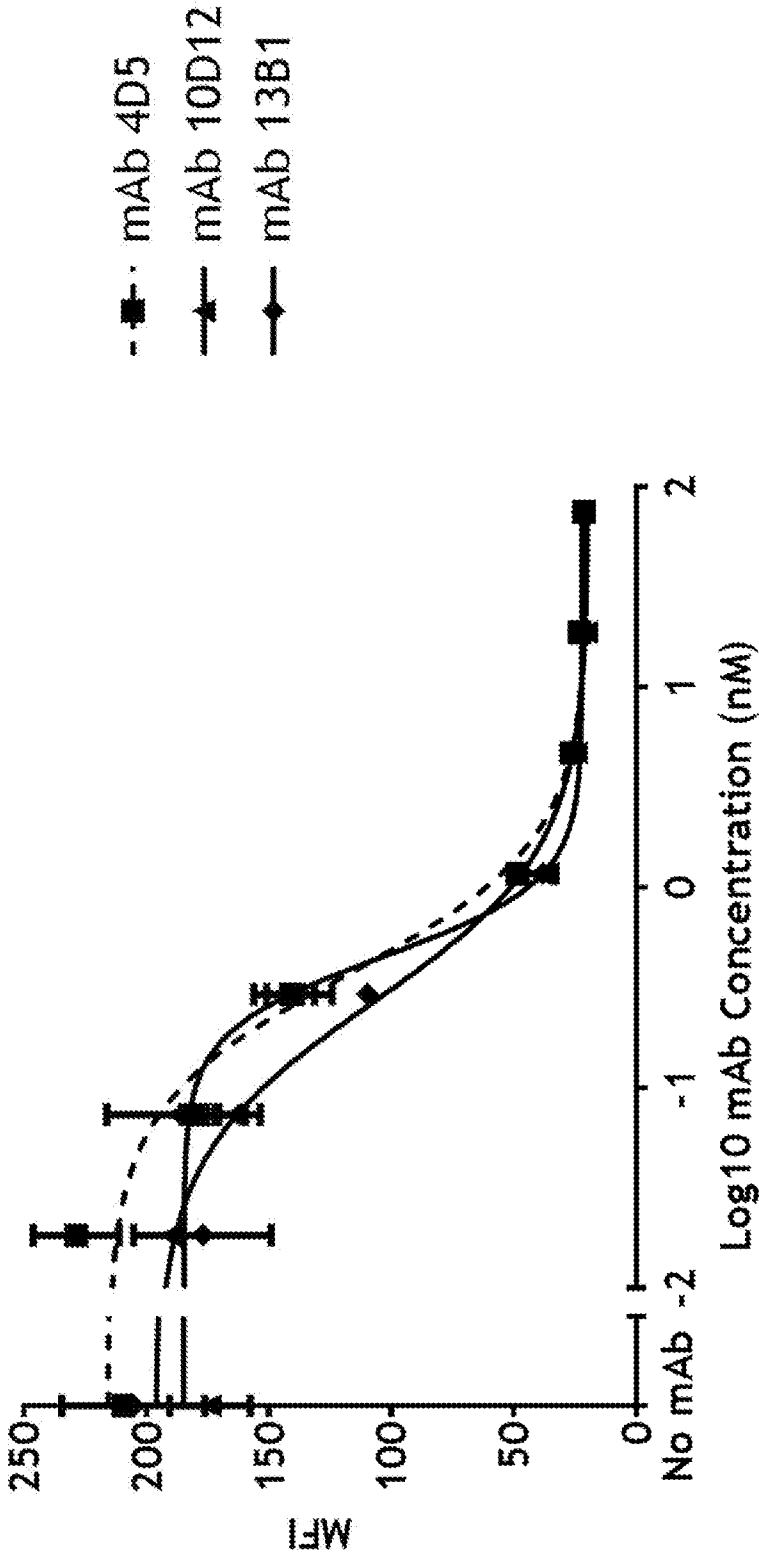
Inhibition of Factor Bb Deposition on Zymosan by MASP-3 mAbs



	mAb 1F3	mAb 1G4	mAb 2D7	mAb 4B6
IC50 (nM)	0.1	1.1	3.5	0.2

FIG. 55B

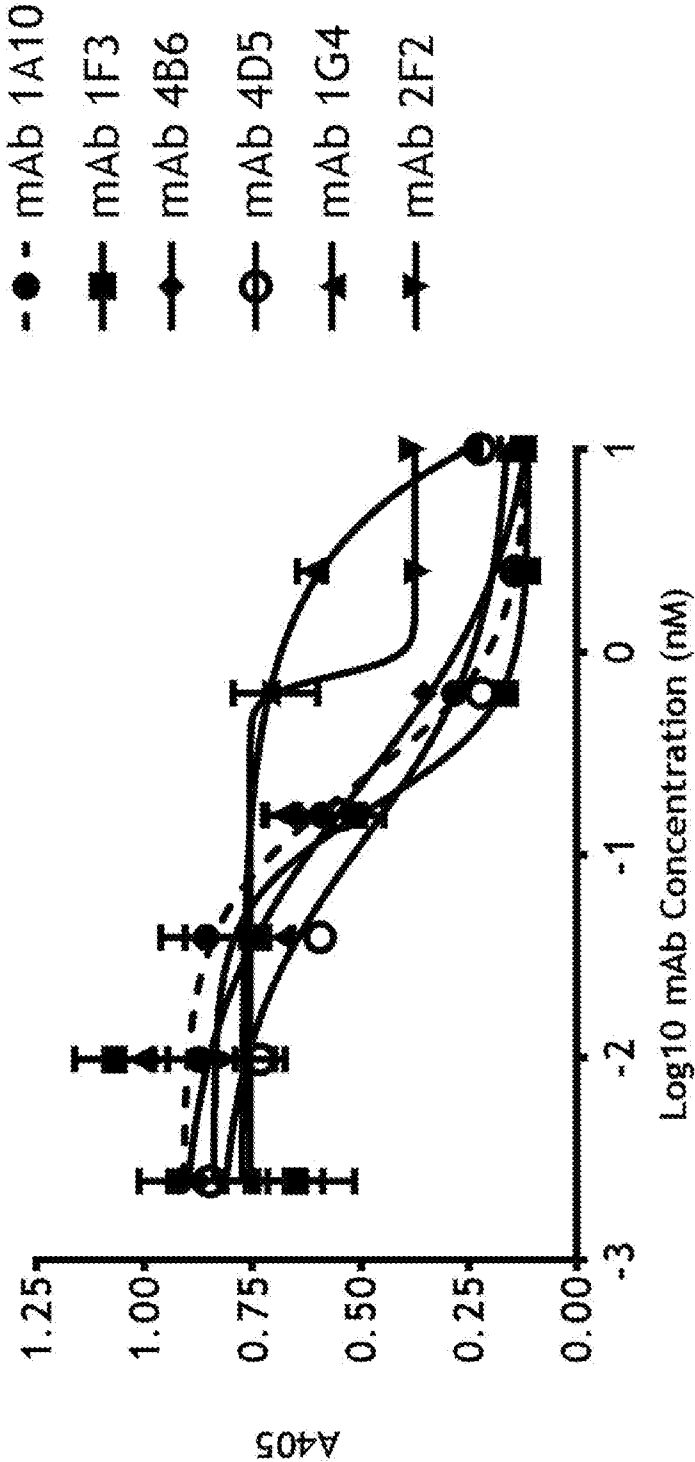
Inhibition of Factor Bb Deposition on Zymosan by MASP-3 mAbs



	mAb 4D5	mAb 10D12	mAb 13B1
IC50 (nM)	0.4	0.5	0.3

FIG. 56A

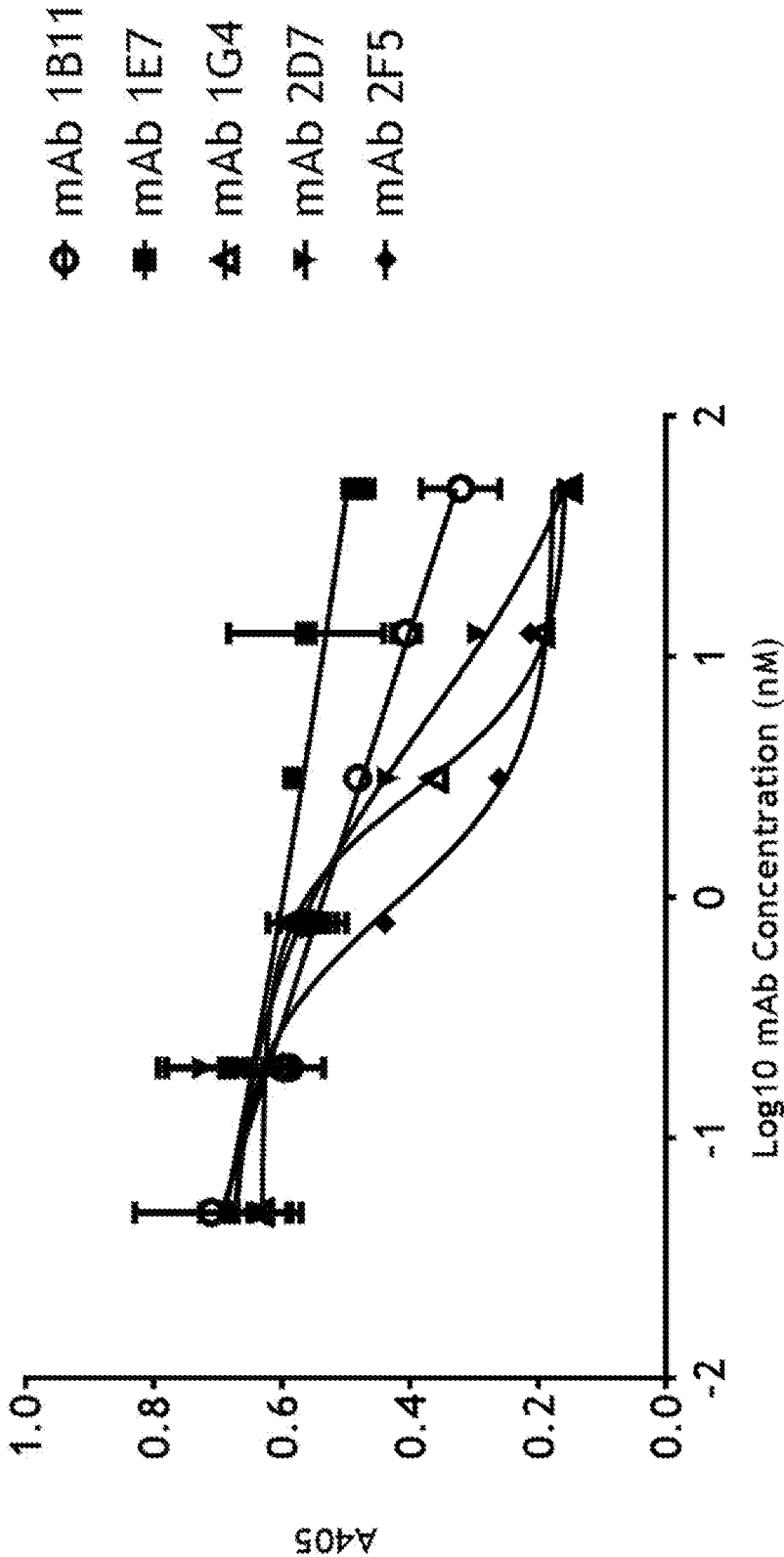
Inhibition of Rabbit Erythrocyte Lysis by MASP-3 mAbs



	mAb 1A10	mAb 1F3	mAb 1G4	mAb 2F2	mAb 4B6	mAb 4D5
IC50 (nM)	0.2	0.2	-	0.8	0.2	0.1

FIG. 56B

Inhibition of Rabbit Erythrocyte Lysis by MASP-3 mAbs



	mAb 1B11	mAb 1E7	mAb 1G4	mAb 2D7	mAb 2F5
IC50 (nM)	-	-	2.7	5.4	0.9

FIG. 57

Factor D in 3MC Serum

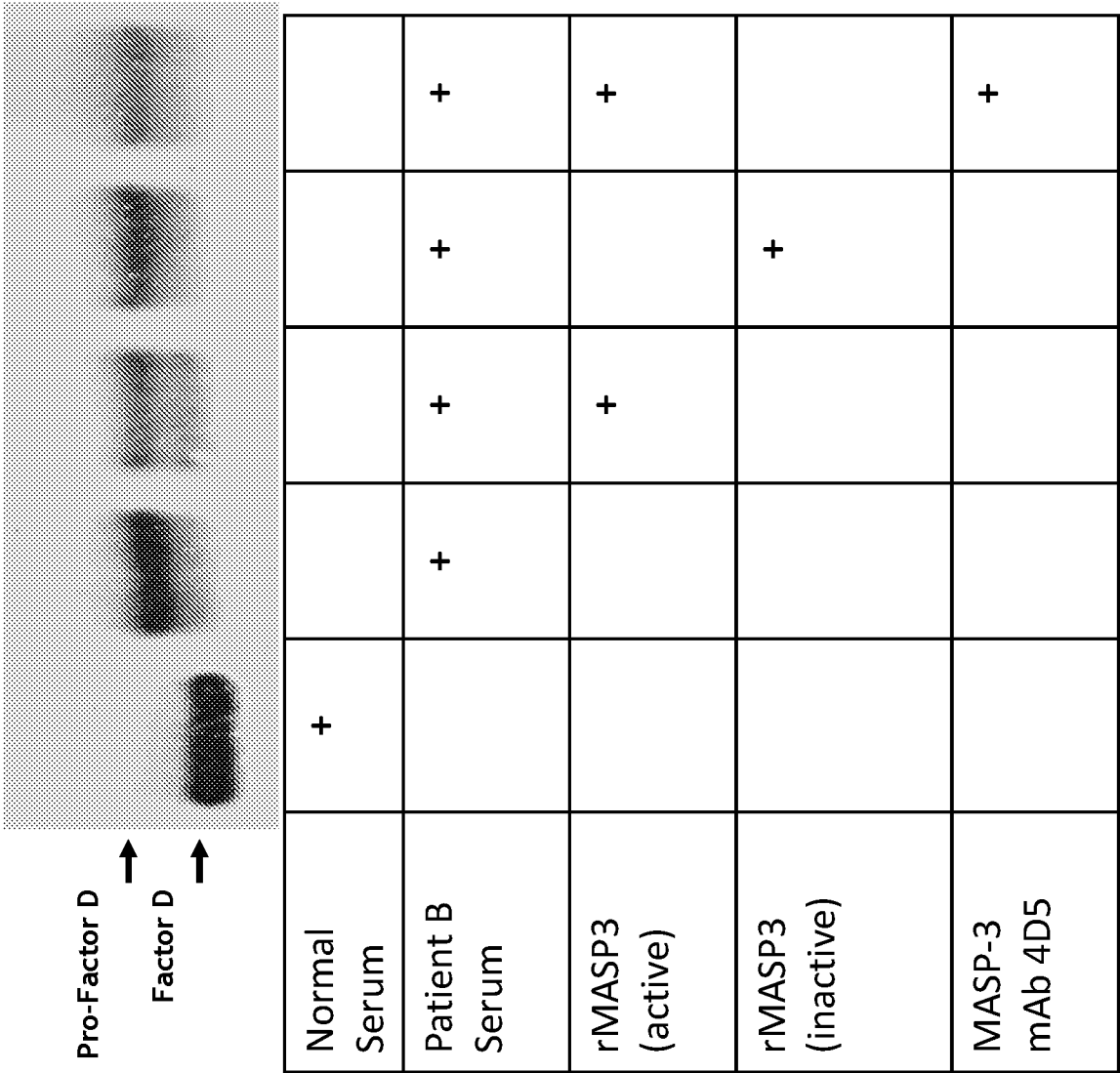


FIG. 58

MASP-3 mAbs inhibit the APC *in vivo*

C3/C3b/iC3b Deposition on Zymosan

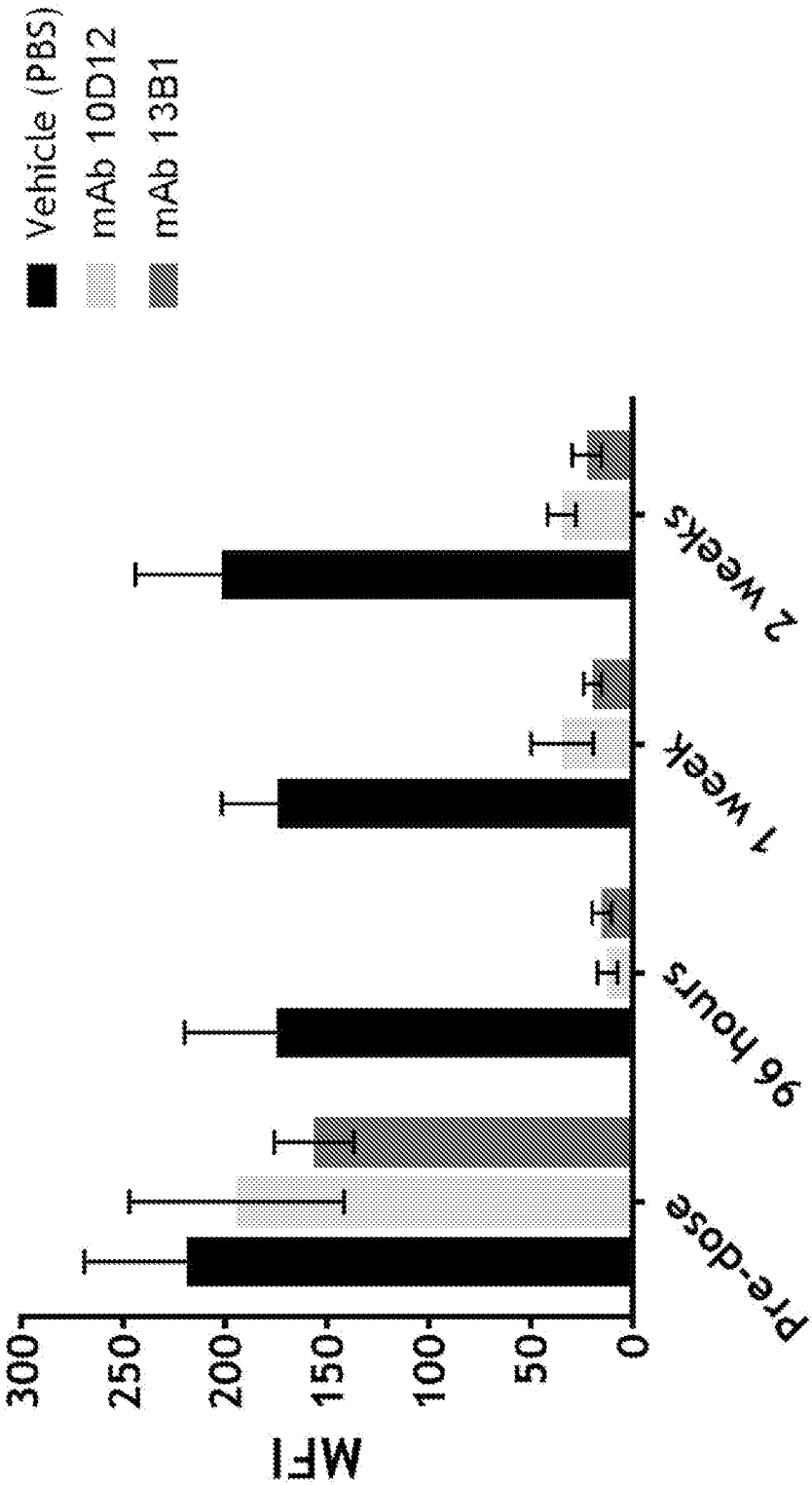


FIG. 59

Status of the Ba fragment of Factor B in Mice
treated with mAb 10D12

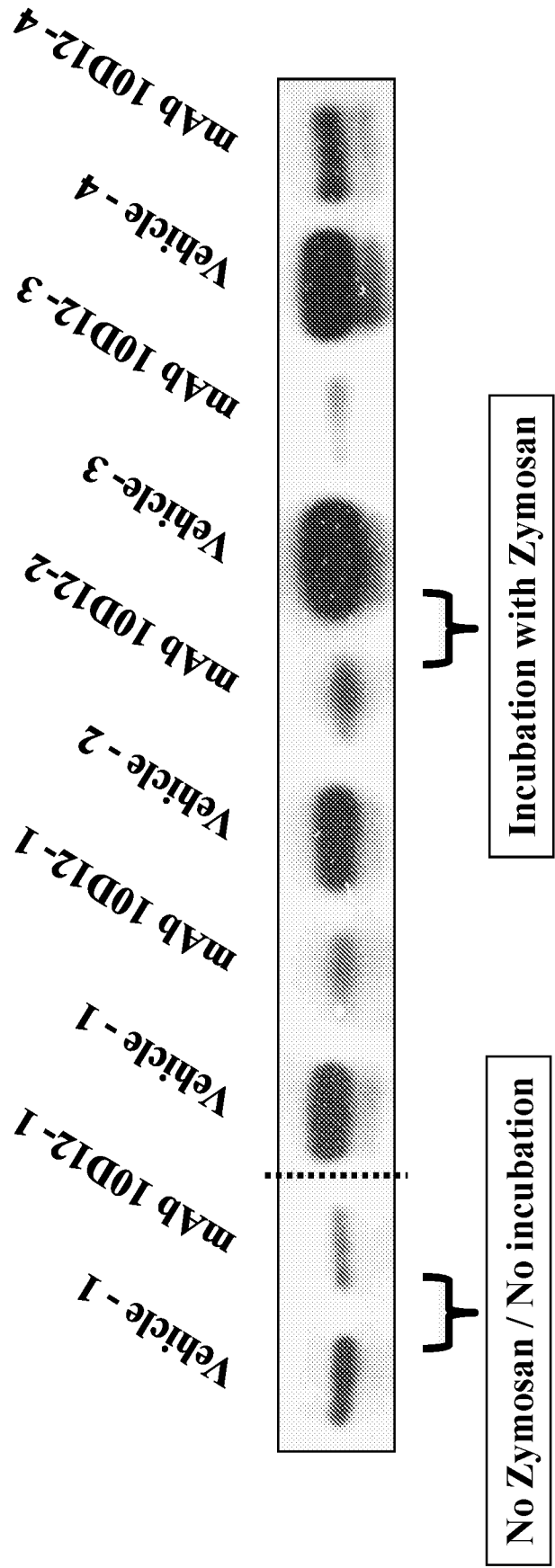


FIG. 60

Serum from mice treated with mAb 10D12 shows inhibition of hemolysis

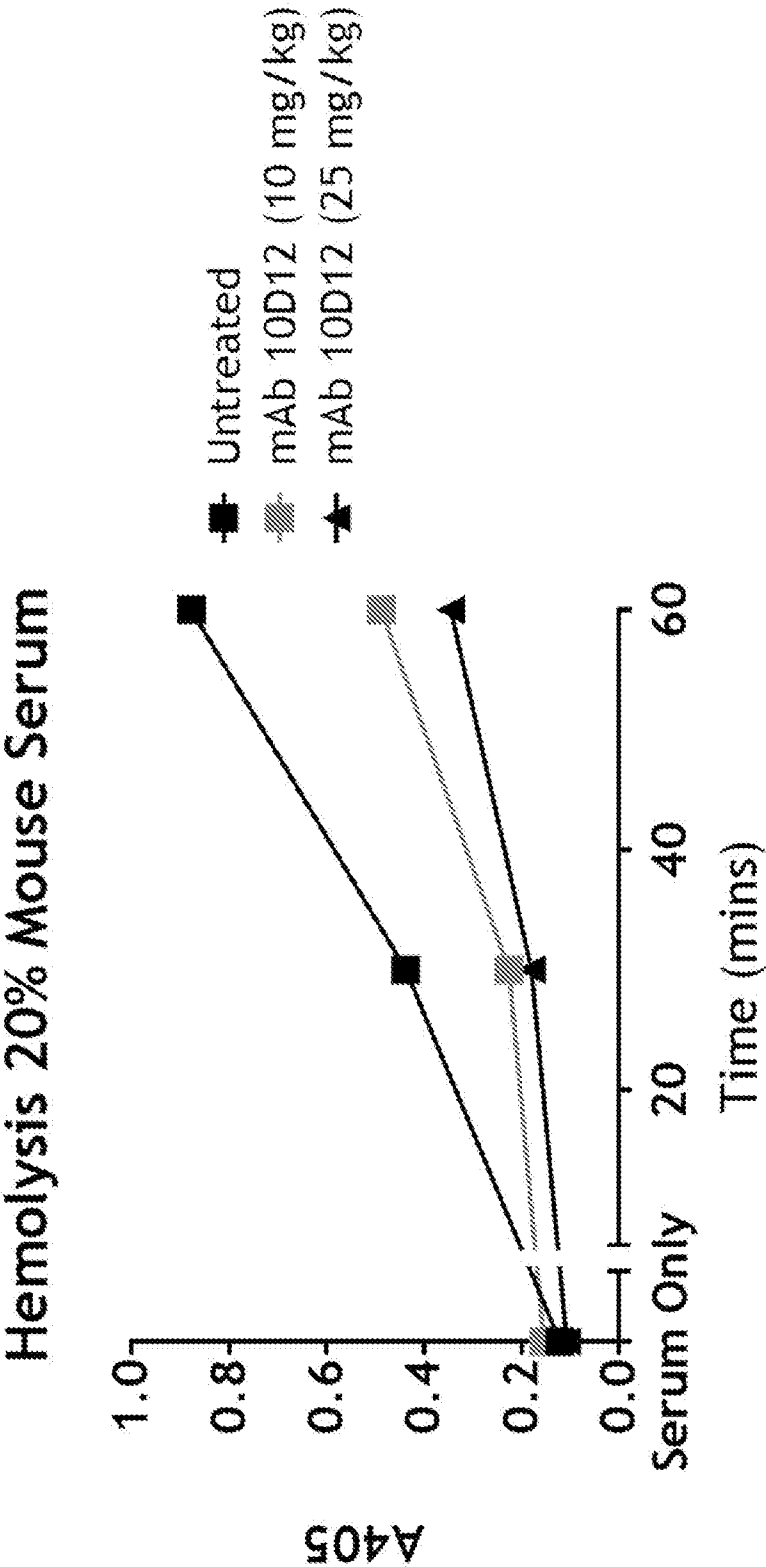


FIG. 61A

Competition binding analysis identifies MASP-3 mAbs (IgG4) that block the interaction between mAb 4D5 (IgG1) and MASP-3

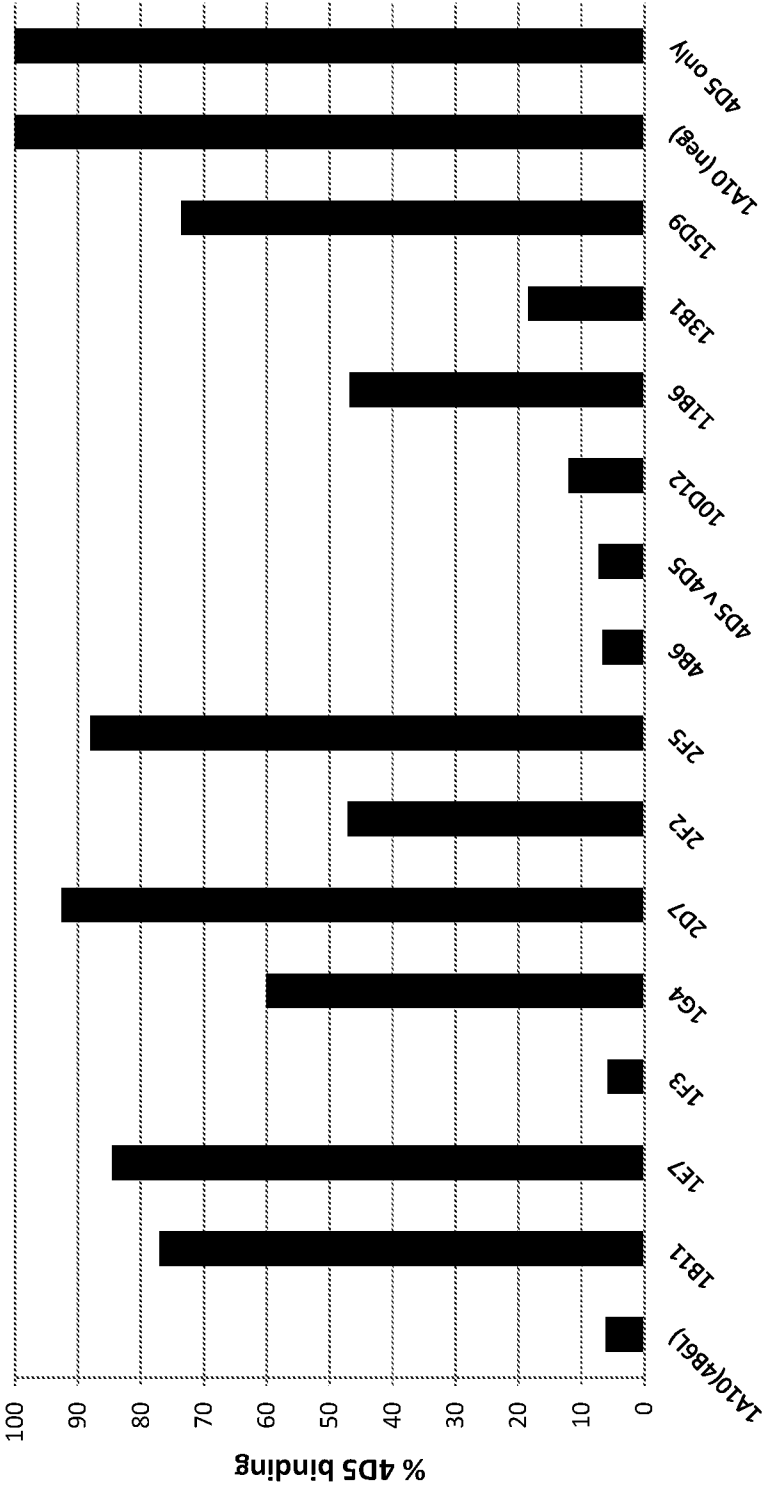


FIG. 61B

Competition binding analysis identifies MASP-3 mAbs (IgG4) that block the interaction between mAb 10D12 (IgG1) and MASP-3

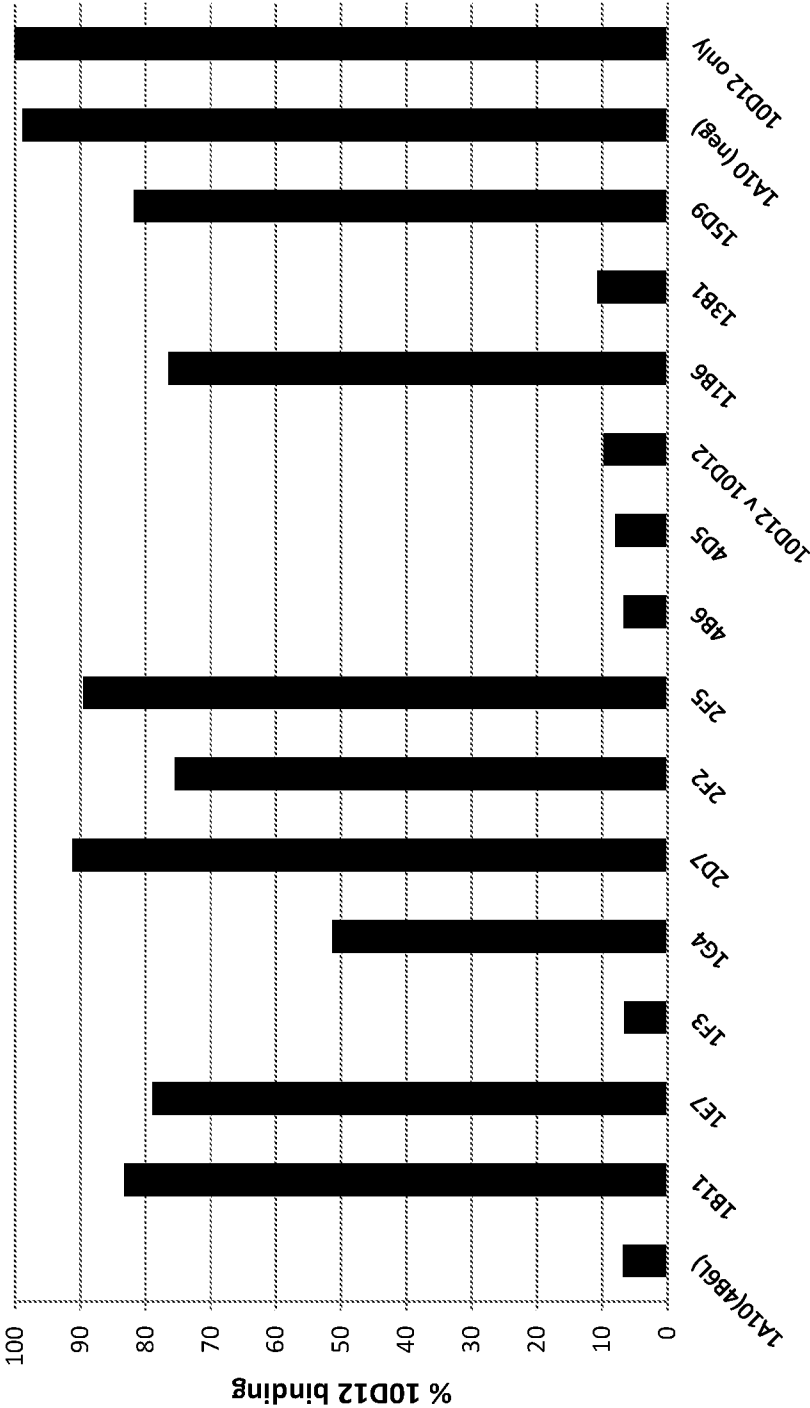


FIG. 61C

Competition binding analysis identifies MASP-3 mAbs (IgG4) that block the interaction between mAb 13B1 (IgG1) and MASP-3

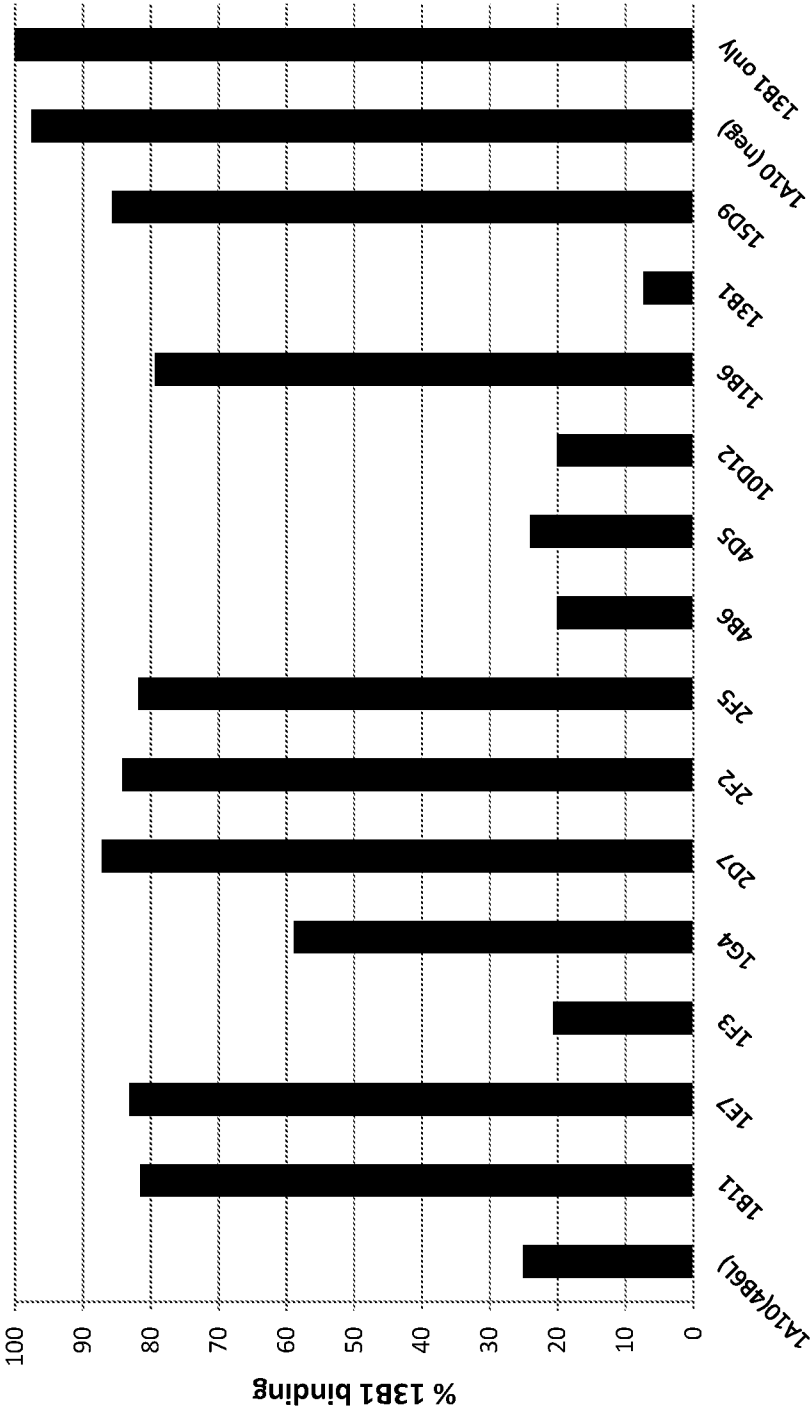


FIG. 61D

Competition binding analysis identifies MASP-3 mAbs (IgG4) that block the interaction between mAb 1F3 (IgG4) and MASP-3

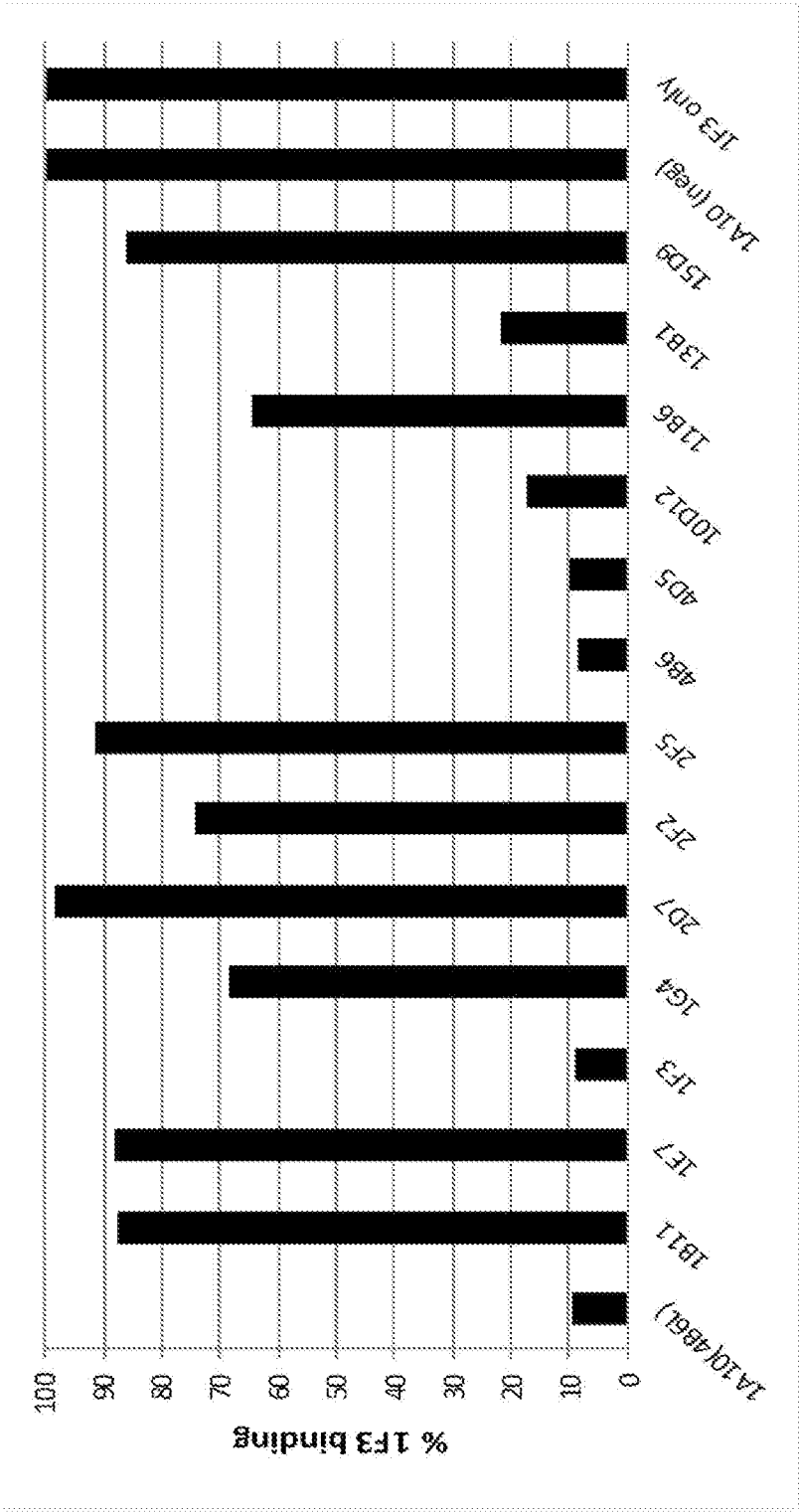


FIG. 61E

Competition binding analysis identifies MASP-3 mAbs (IgG4) that block the interaction between mAb 1G4 (IgG1) and MASP-3

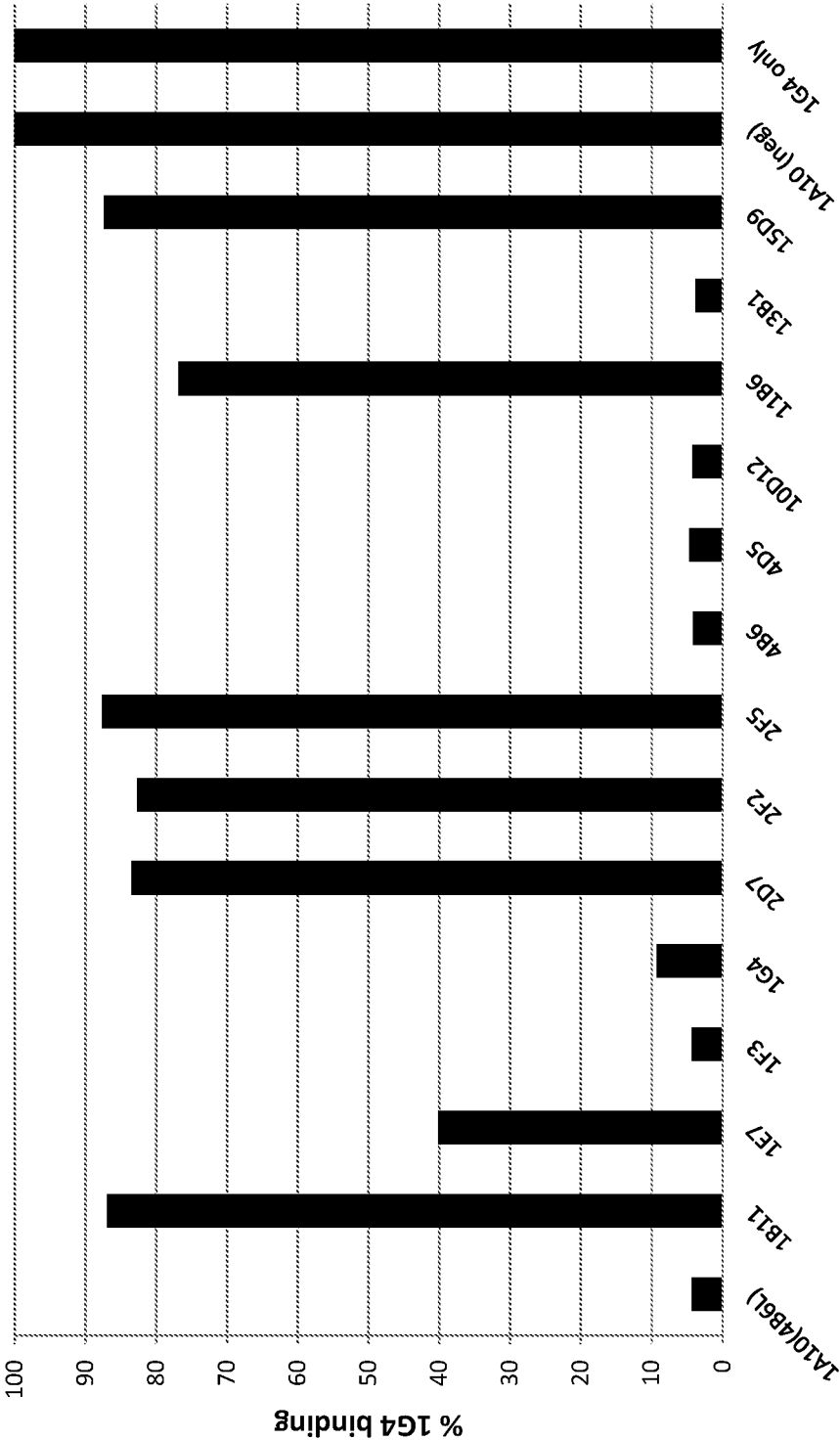
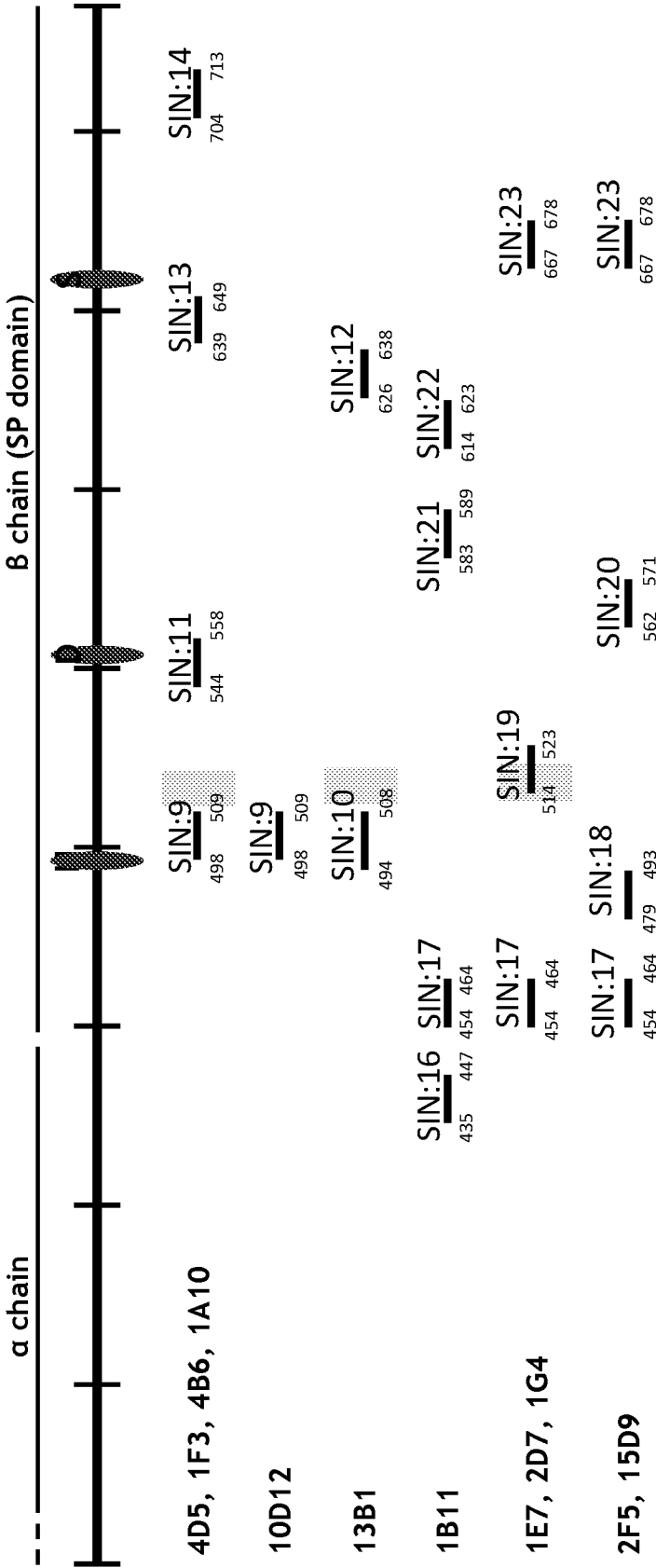


FIG. 62
Regions of contact on MASP-3 by the MASP-3 mAbs

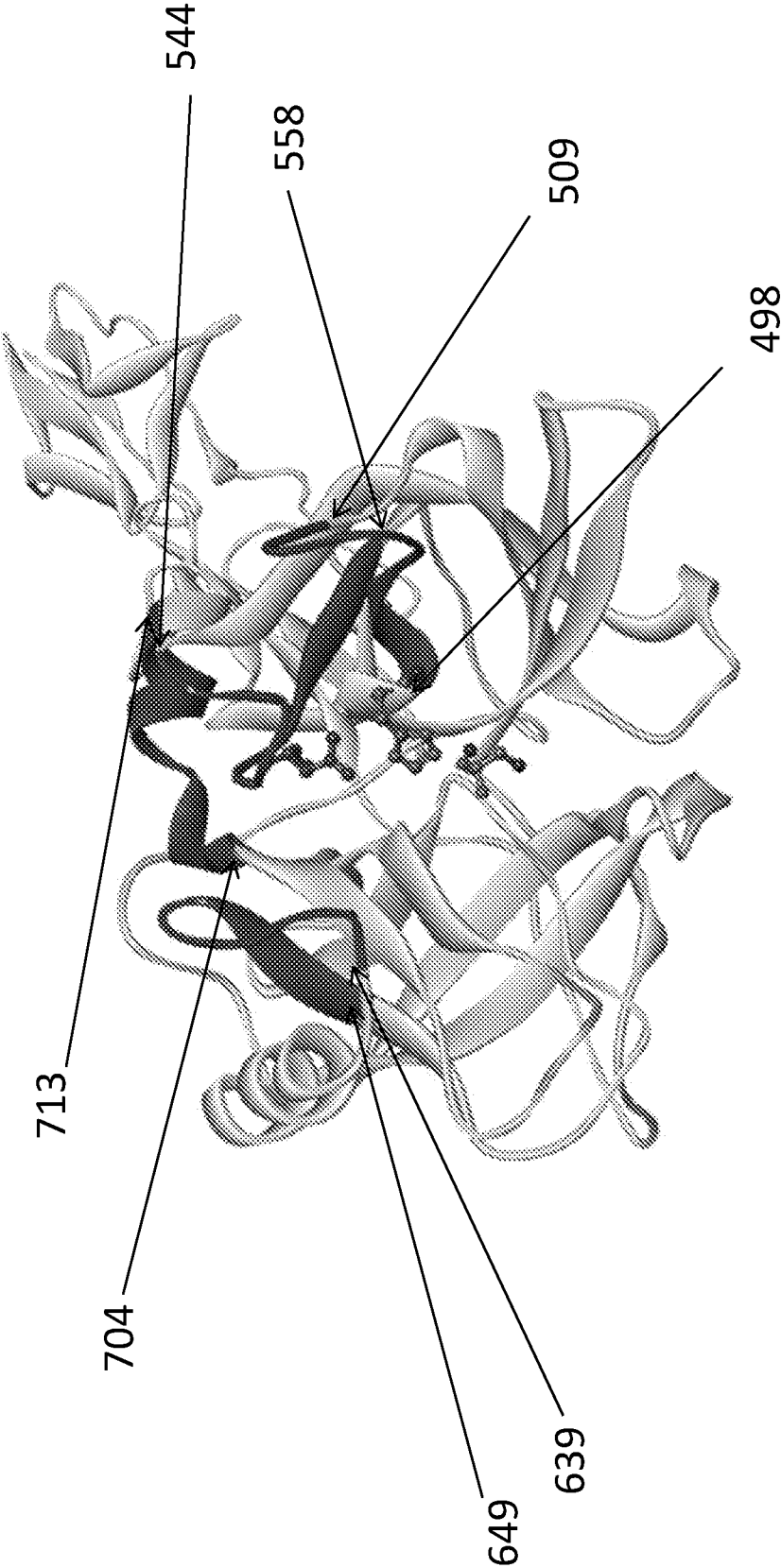


H=497
D=553
S=664

Note: #9 to #23 "SIN" refer to SEQ ID NO:s

FIG. 63A

Regions of contact on human MASP-3 for mAbs 1F3, 4B6, 4D5 and 1A10



SEQ ID NO:9 = aa residues 498-509 of MASP-3

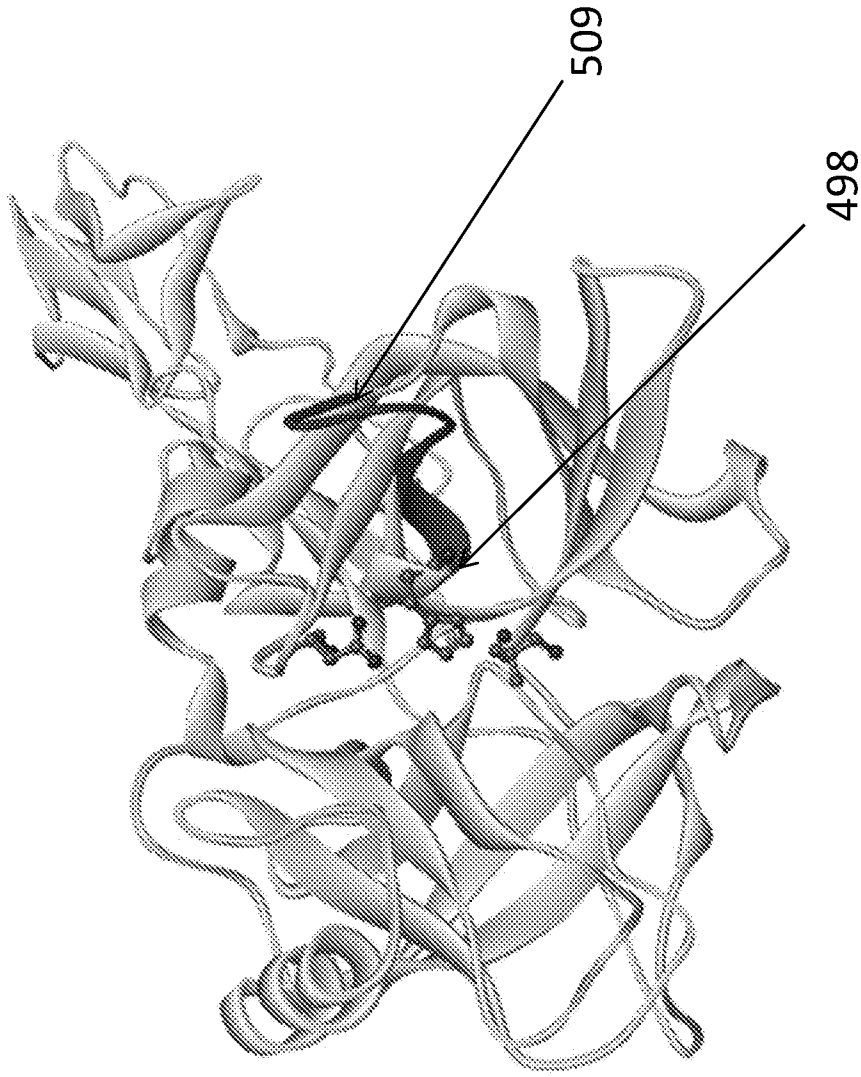
SEQ ID NO:11= aa residues 544-558 of MASP-3

SEQ ID NO:13= aa residues 639-649 of MASP-3

SEQ ID NO:14= aa residues 704-713 of MASP-3

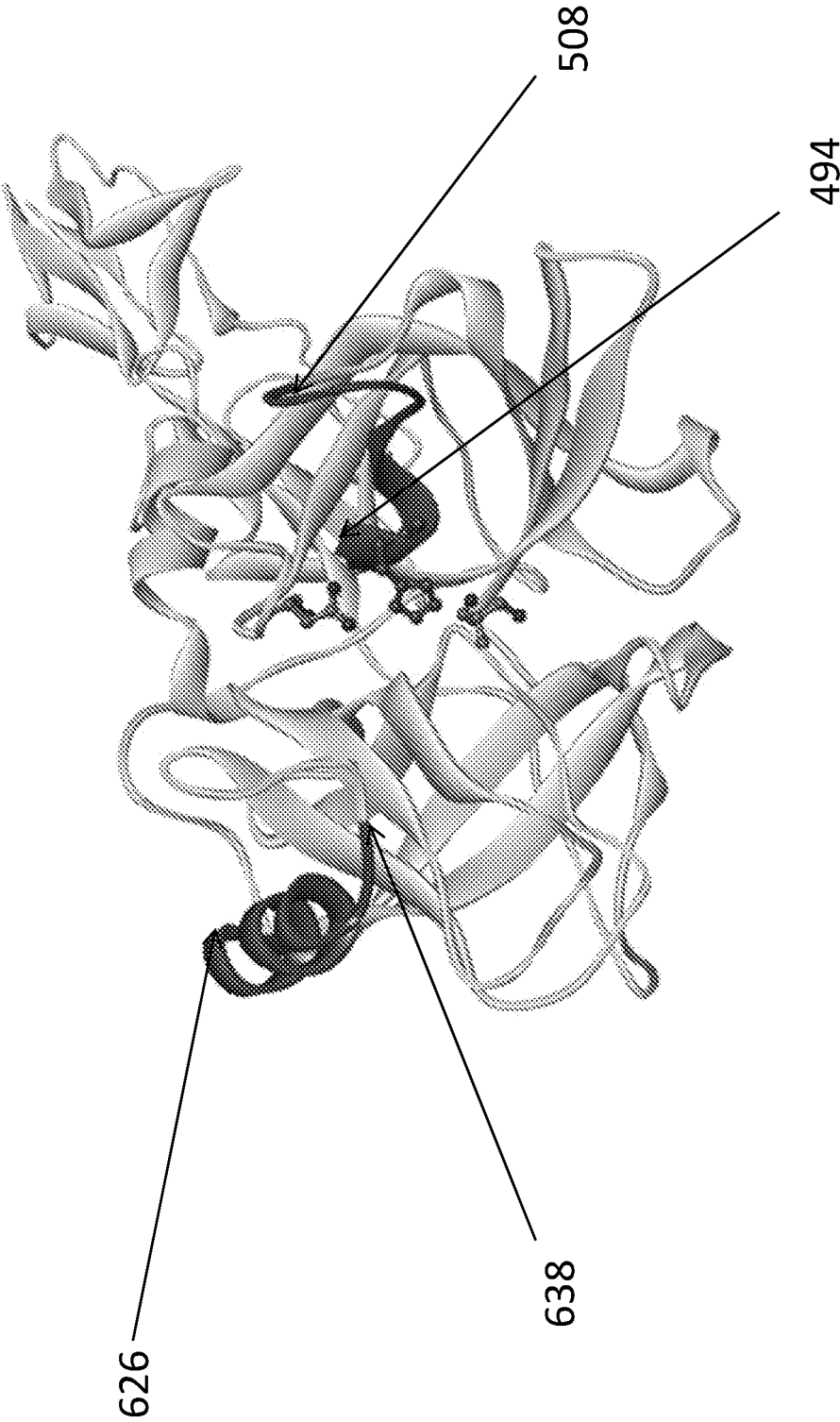
FIG. 63B

Regions of contact on human MASP-3 for mAb 10D12



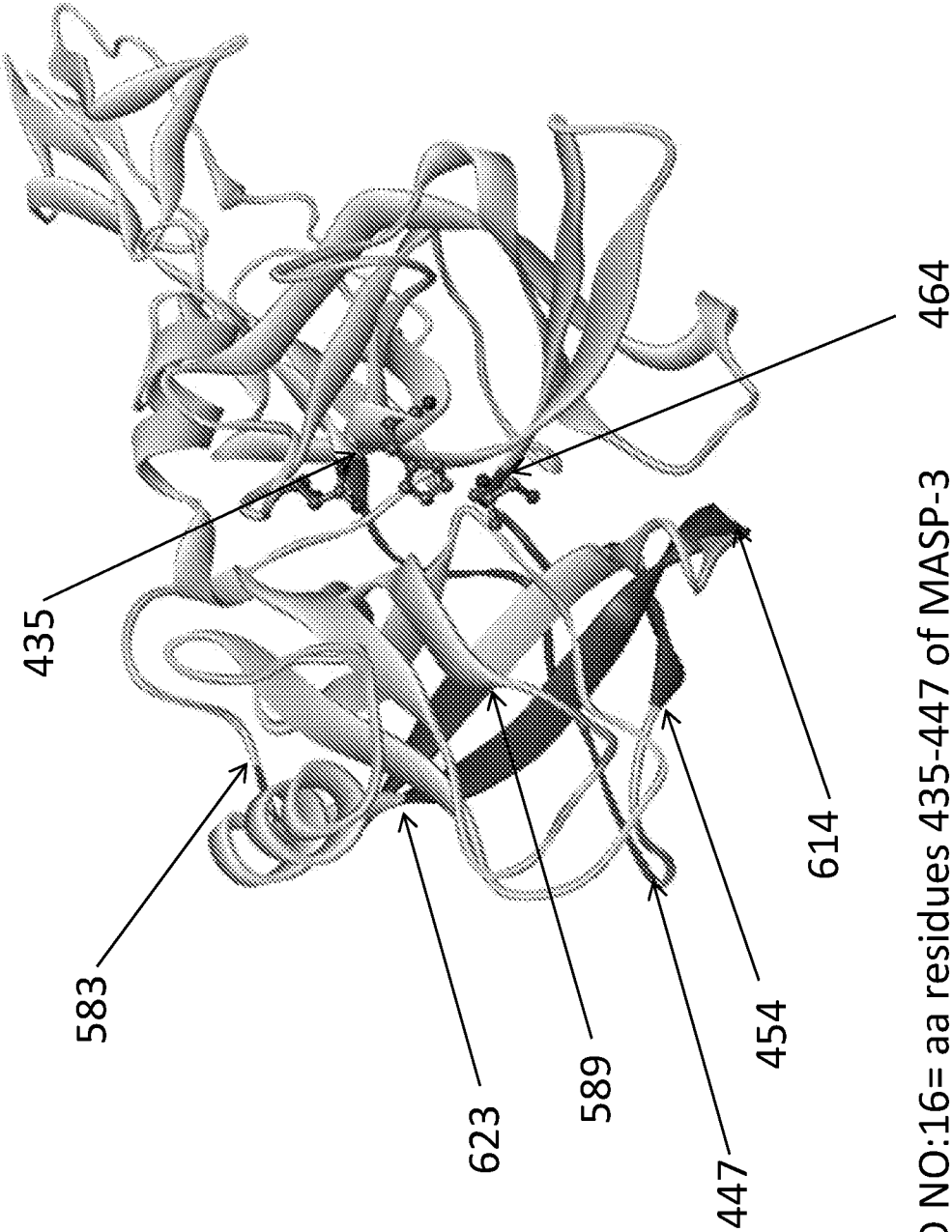
SEQ ID NO:9 = aa residues 498-509 of MASP-3

FIG. 64
Regions of contact on human MASP-3 for mAb 13B1



SEQ ID NO:10= aa residues 494 to 508 of MASP-3
SEQ ID NO:12= aa residues 626 to 638 of MASP-3

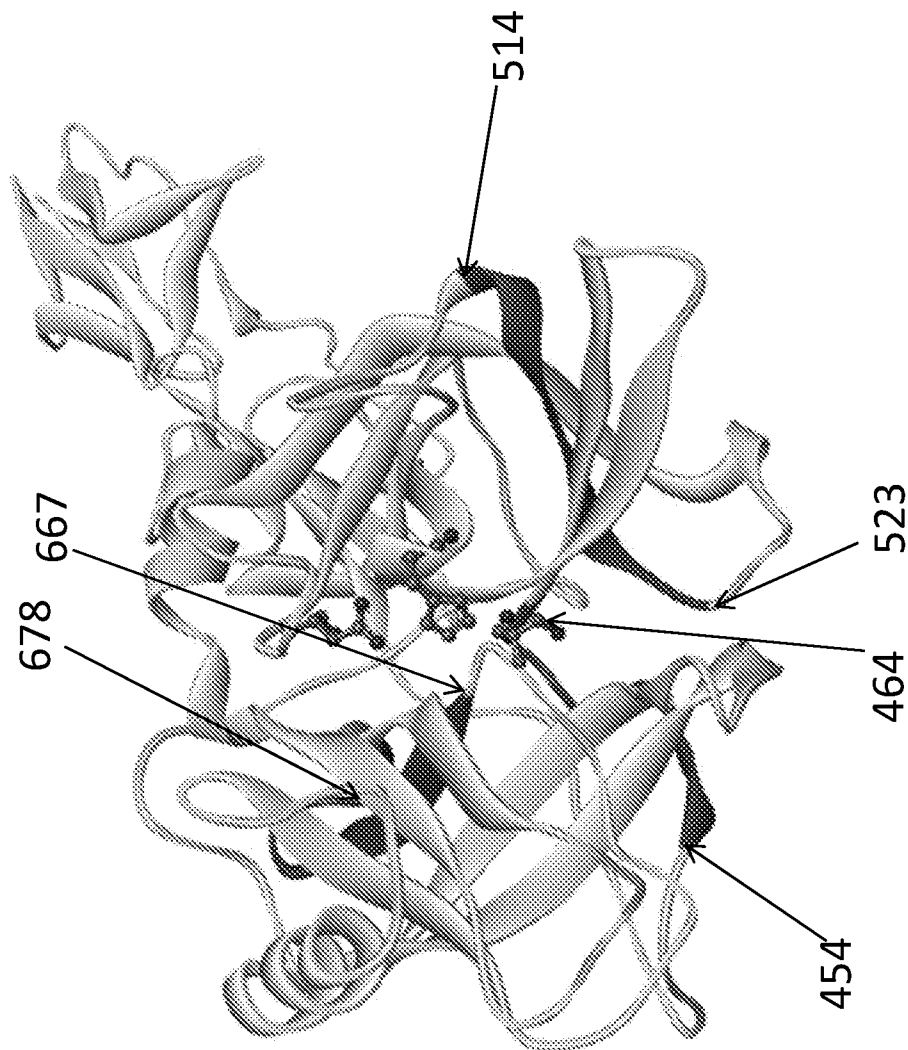
FIG. 65
Regions of contact on human MASP-3 for mAb 1B11



SEQ ID NO:16= aa residues 435-447 of MASP-3
SEQ ID NO:17= aa residues 454-464 of MASP-3
SEQ ID NO:21= aa residues 583-589 of MASP-3
SEQ ID NO:22= aa residues 614-623 of MASP-3

FIG. 66

Regions of contact on human MASP-3 for mAb 1E7, 1G4 and 2D7



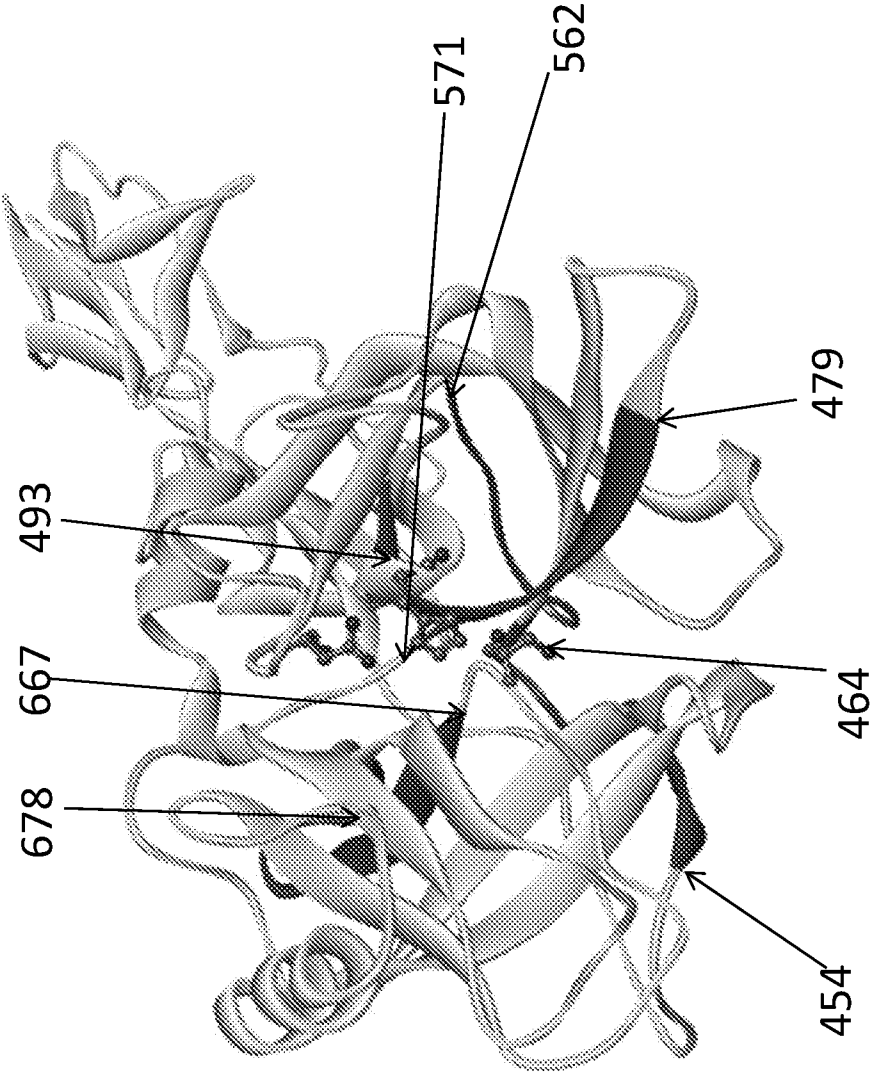
SEQ ID NO:17= aa residues 454 to 464 of MASP-3

SEQ ID NO:19= aa residues 514 to 523 of MASP-3

SEQ ID NO:23= aa residues 667 to 678 of MASP-3

FIG. 67

Regions of contact on human MASP-3 for mAb 15D9 and 2F5



SEQ ID NO:17= aa residues 454 to 464 of MASP-3
SEQ ID NO:18= aa residues 479 to 493 of MASP-3
SEQ ID NO:20= aa residues 562 to 571 of MASP-3
SEQ ID NO:23= aa residues 667 to 678 of MASP-3

FIG. 68

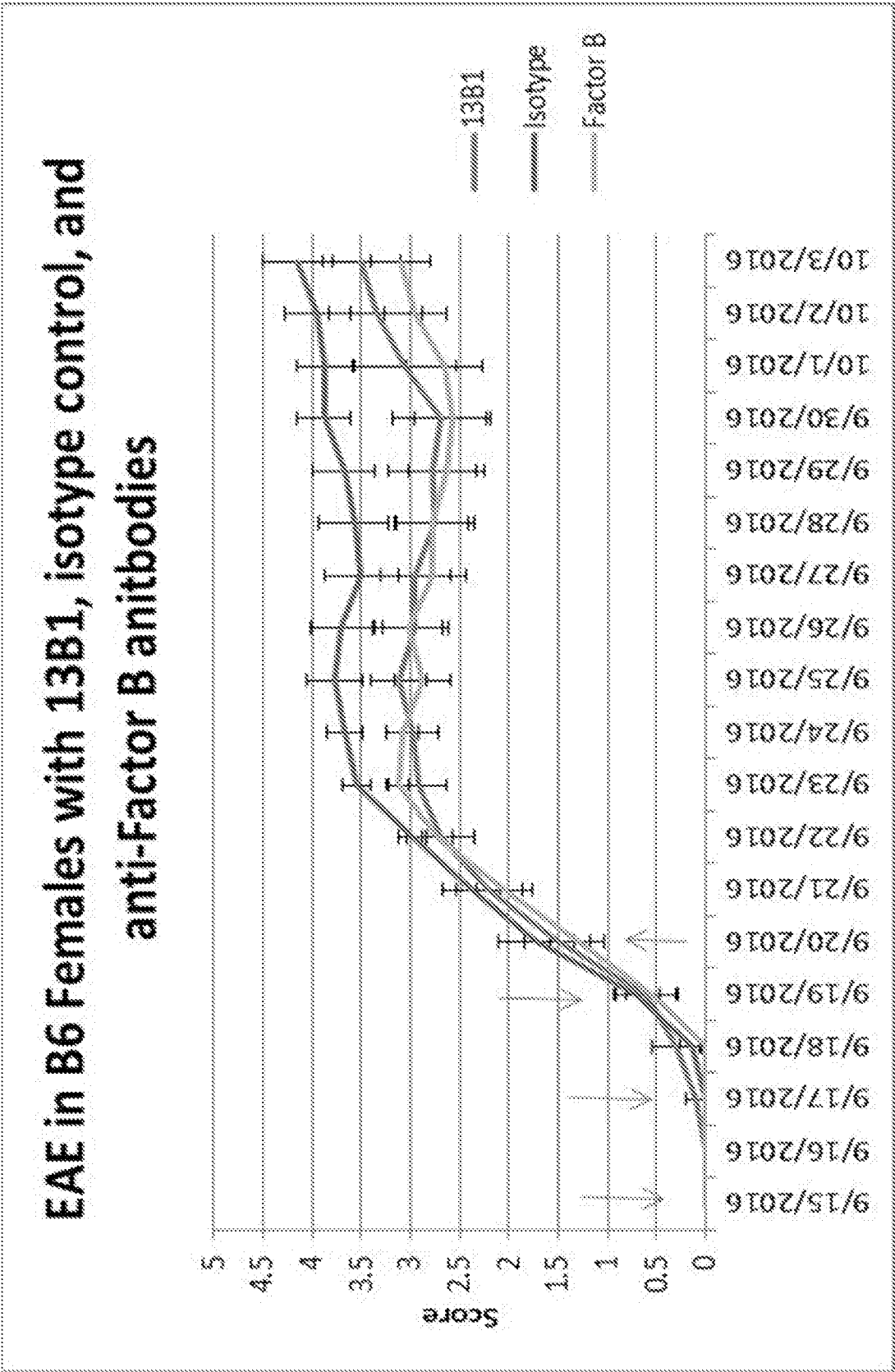


FIG. 69

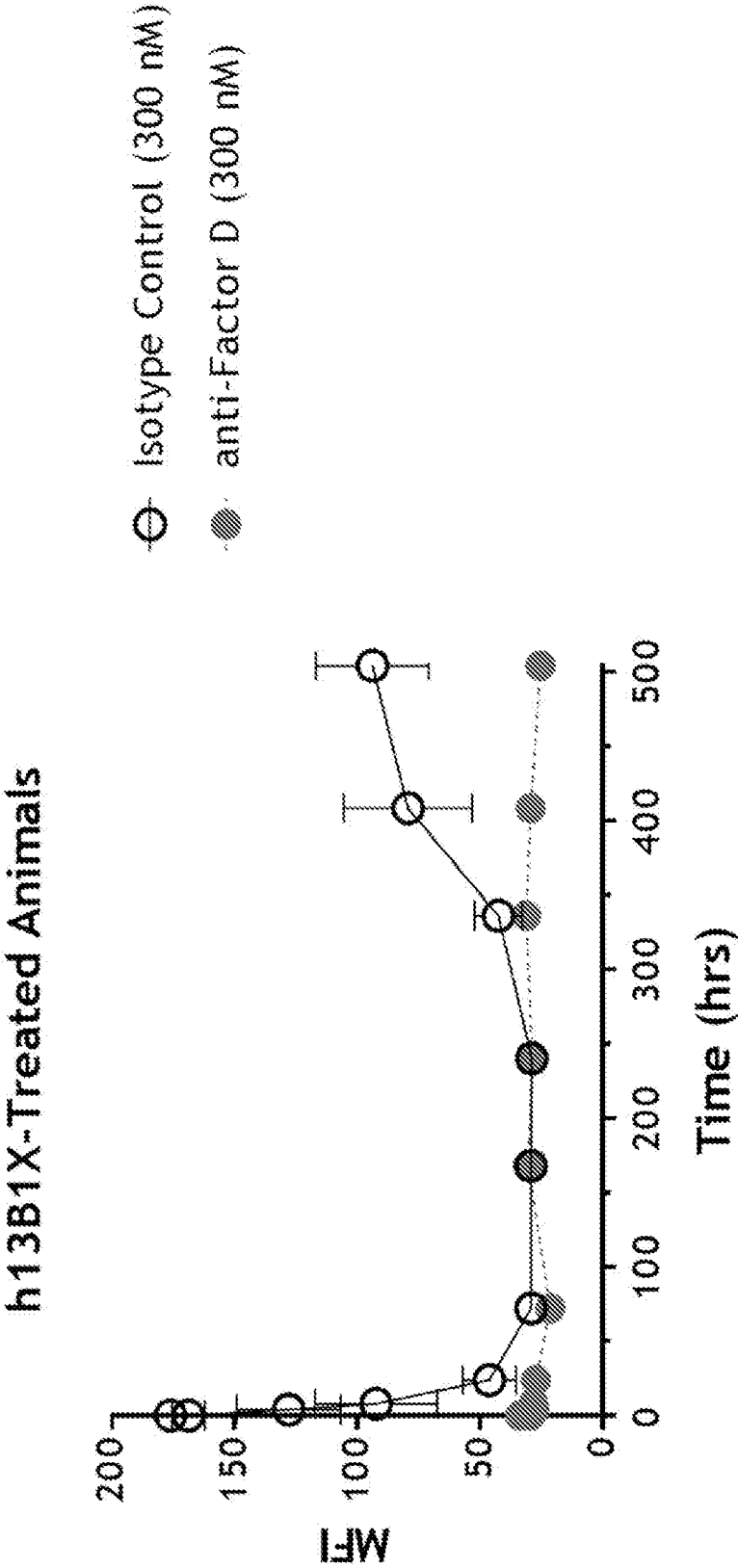


FIG. 70

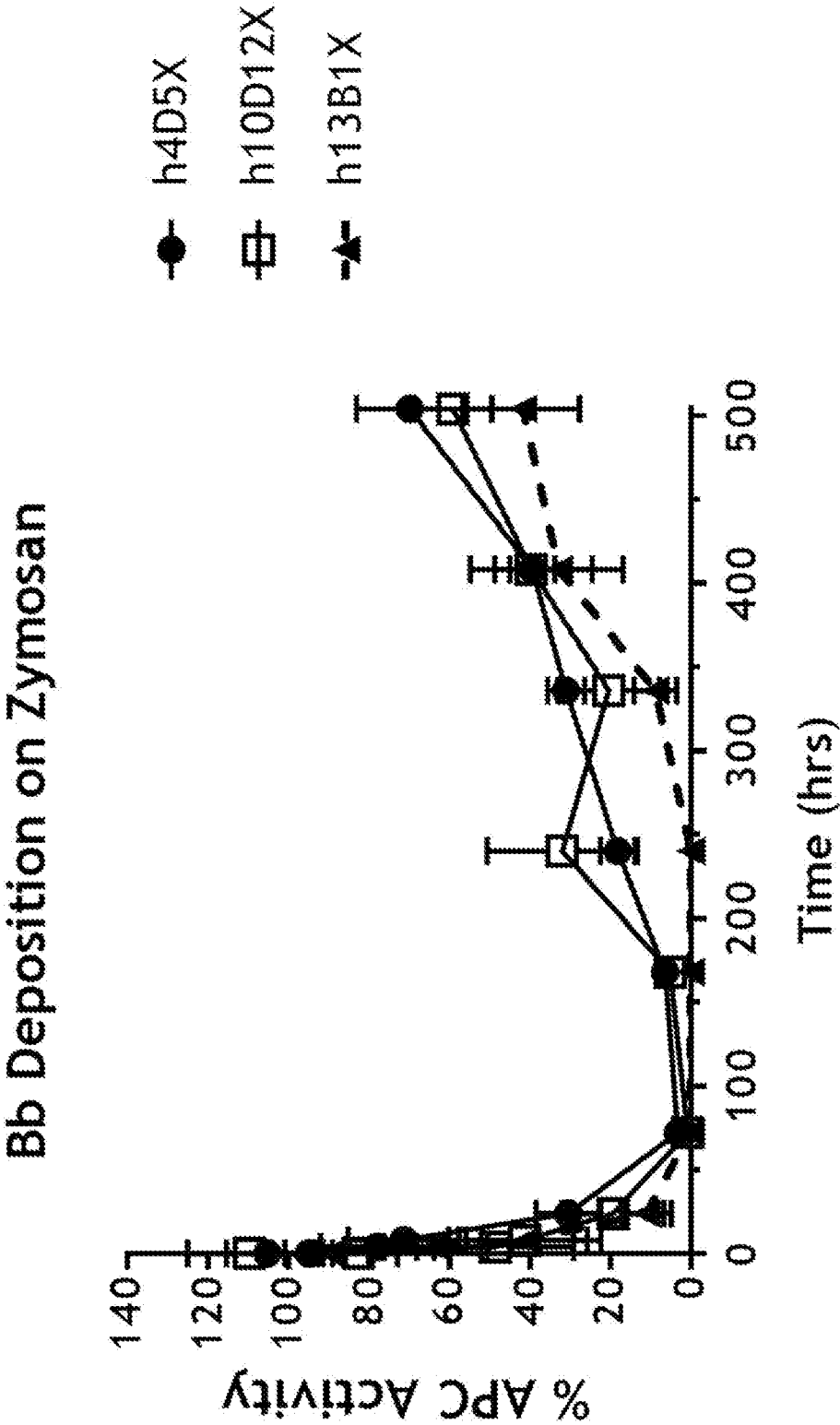


FIG. 71A

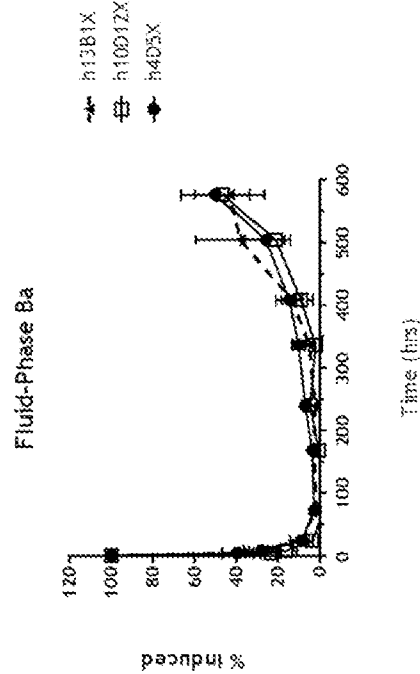


FIG. 71B

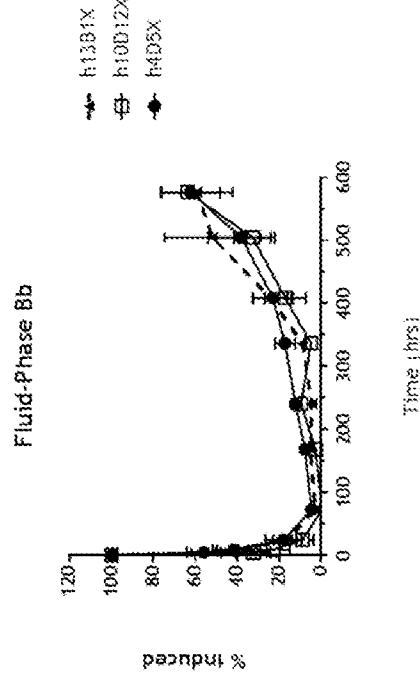


FIG. 71C

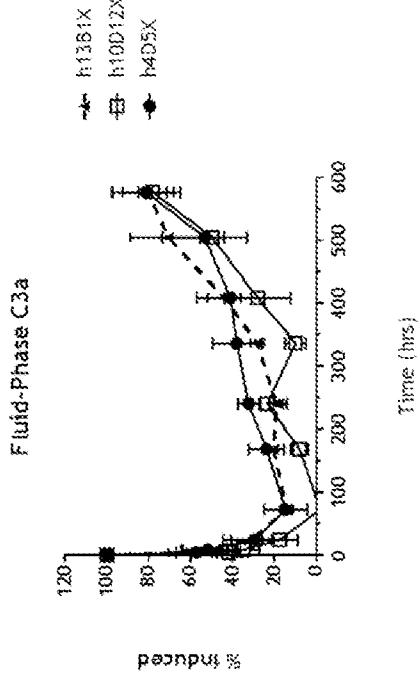


FIG. 72A

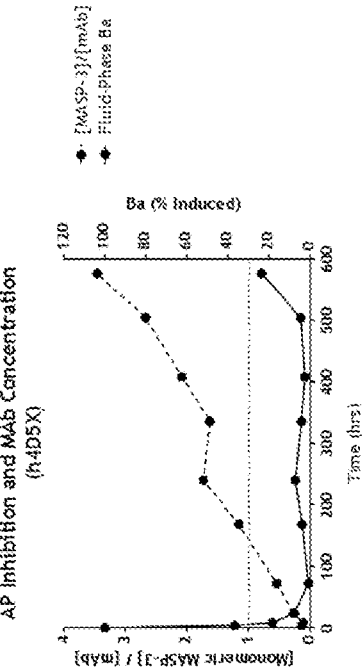


FIG. 72B

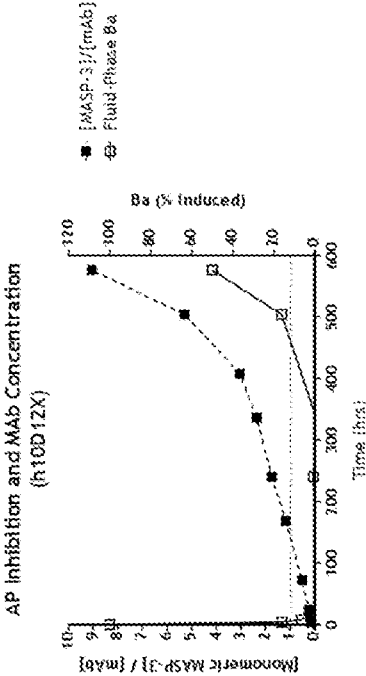


FIG. 72C

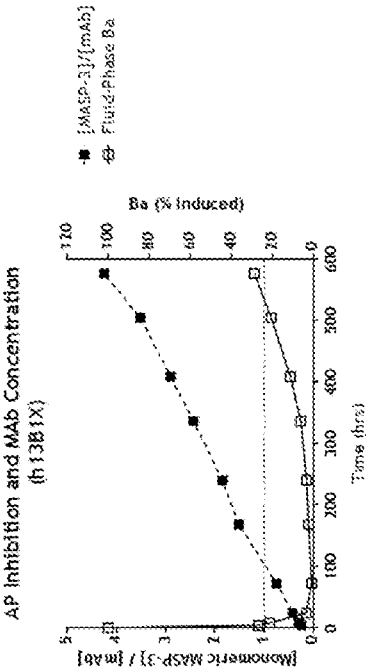


FIG. 73
Analysis of Pro-Df/Mature Df in Serum from Monkey Treated with 13B1X

