Fig. 1

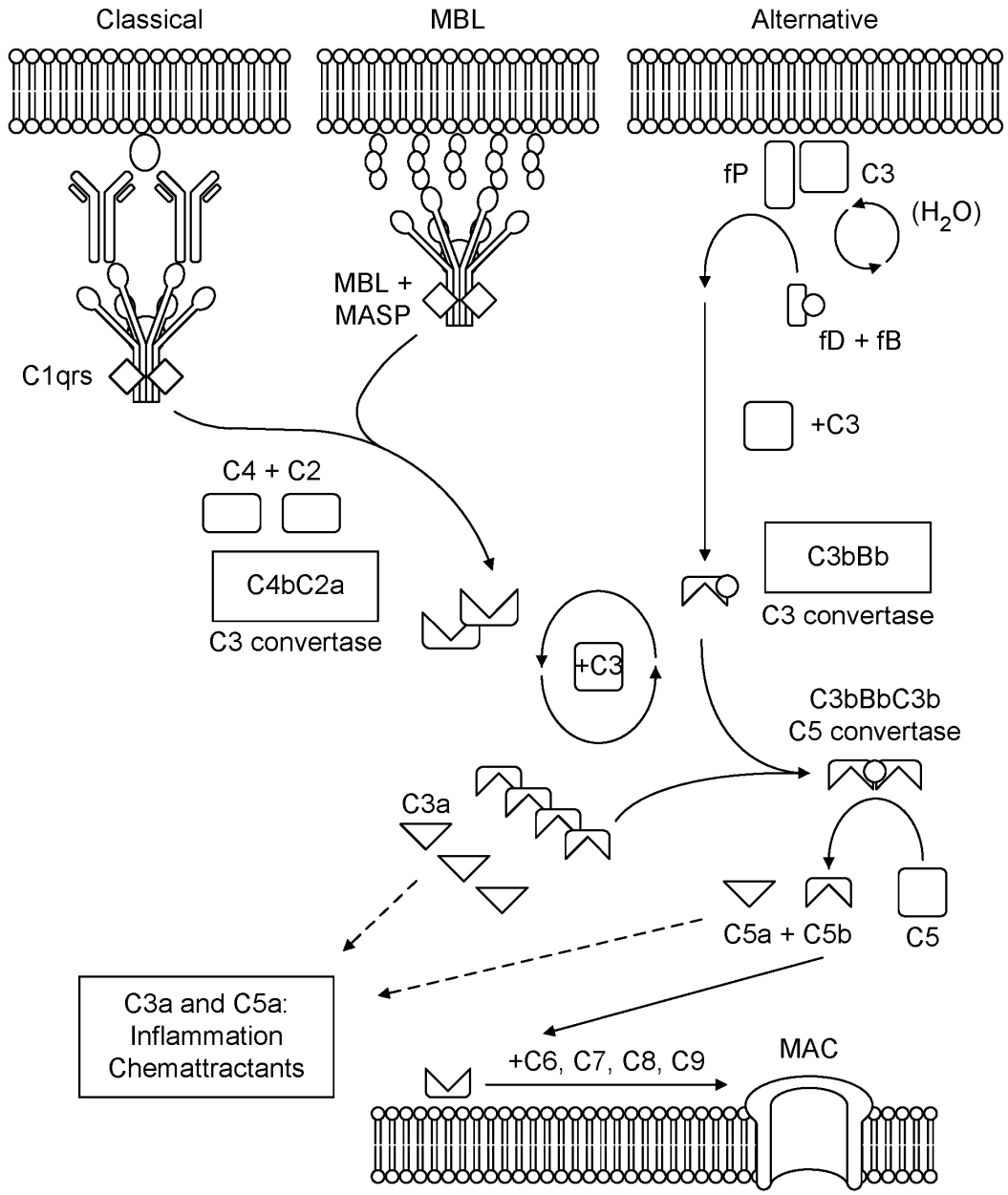

Classical
MBL
Alternative
fP
C3
(H2O)
MBL +
MASP
fD + fB
C1qrs
+C3
C4 + C2
C4bC2a
C3 convertase
C3bBb
C3 convertase
+C3
C3bBbC3b
C5 convertase
C3a
C5a + C5b
C5
C3a and C5a:
Inflammation
Chemattractants
+C6, C7, C8, C9
MAC

Fig. 2

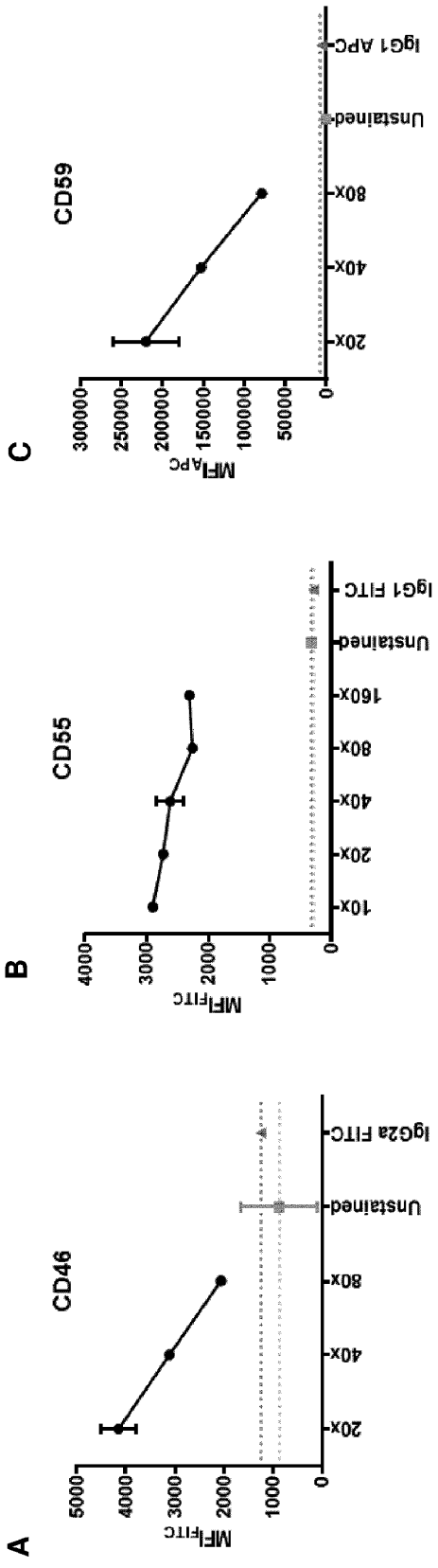

A
CD46
MFIFITC
5000
4000
3000
2000
1000
0
20x
40x
80x
Unstained
IgG2a FITC
B
CD55
MFIFITC
4000
3000
2000
1000
0
10x
20x
40x
80x
160x
Unstained
IgG1 FITC
C
CD59
MFIAPC
300000
250000
200000
150000
100000
50000
0
20x
40x
80x
Unstained
IgG1 APC

Fig. 2 (continued)

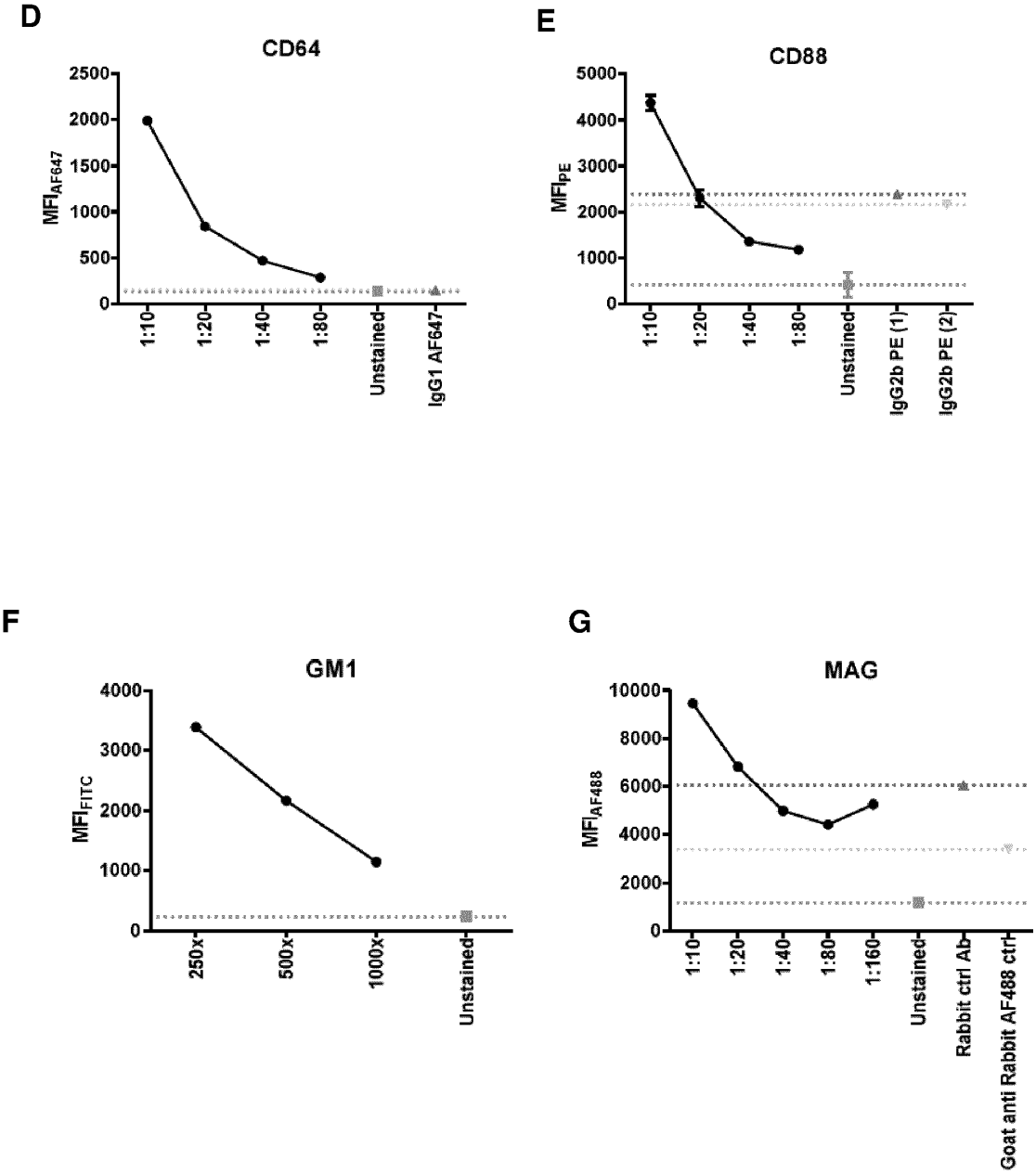


Fig. 3

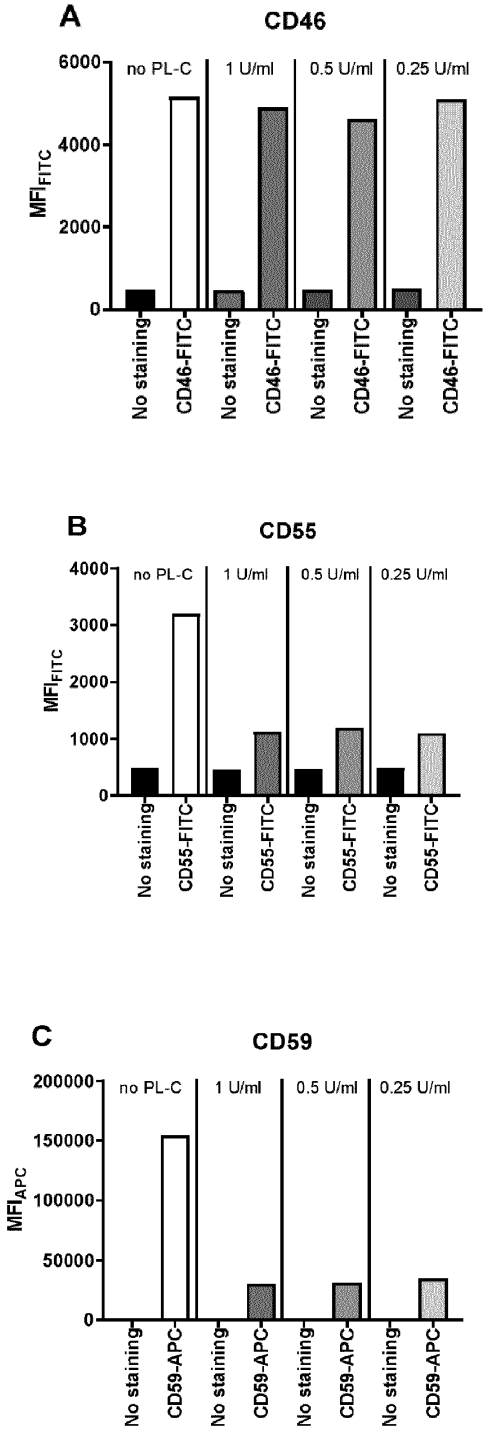

A
CD46
no PL-C
1 U/ml
0.5 U/ml
0.25 U/ml
6000
4000
2000
0
MFI FITC
No staining
CD46-FITC
B
CD55
4000
3000
2000
1000
CD55-FITC
C
CD59
200000
150000
100000
50000
MFI APC
CD59-APC

Fig. 4

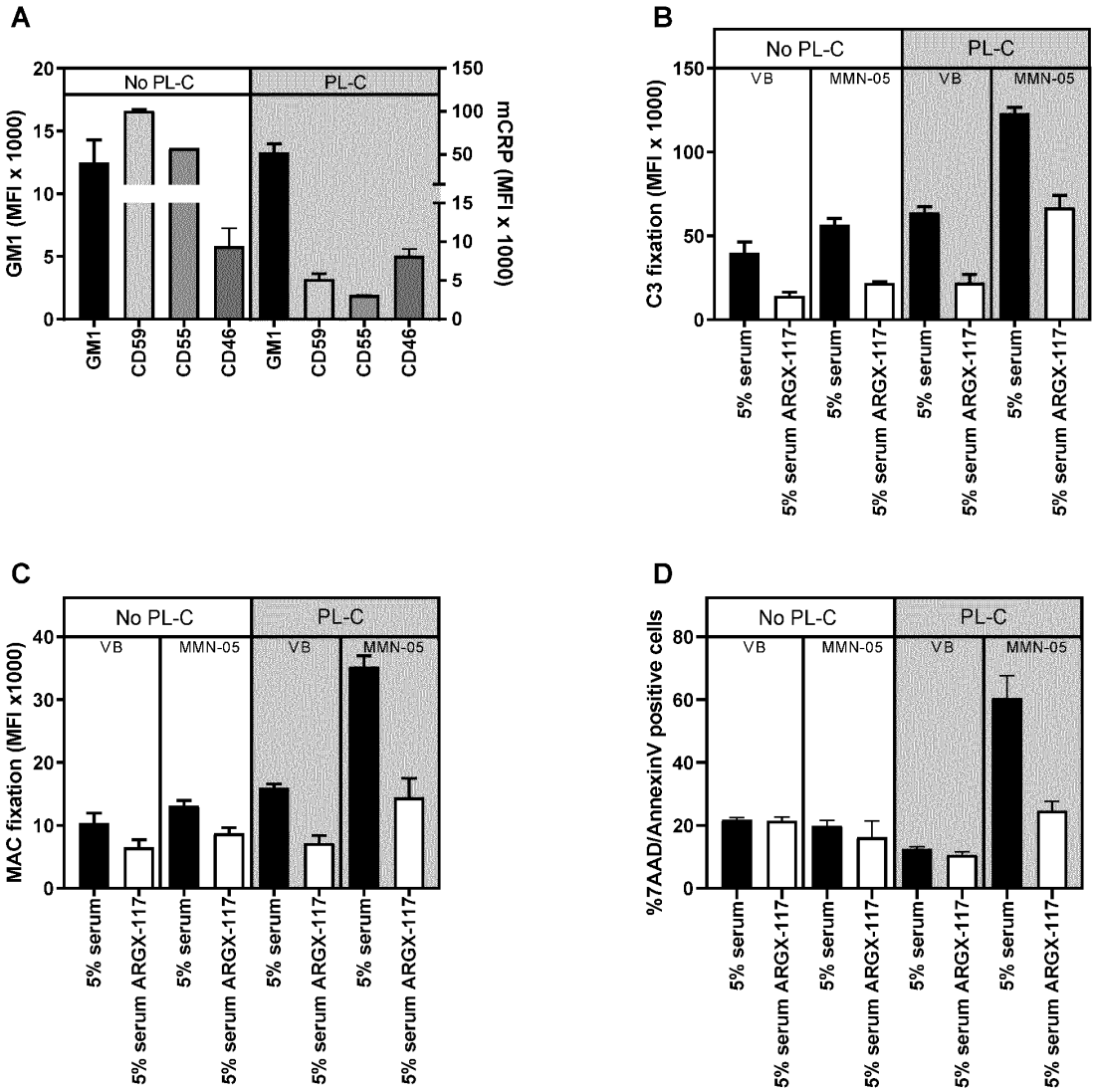

A
No PL-C
PL-C
GM1 (MFI x 1000)
mCRP (MFI x 1000)
GM1
CD59
CD55
CD46
B
No PL-C
PL-C
VB
MMN-05
C3 fixation (MFI x 1000)
5% serum
5% serum ARGX-117
C
MAC fixation (MFI x1000)
D
%7AAD/AnnexinV positive cells

Fig. 5

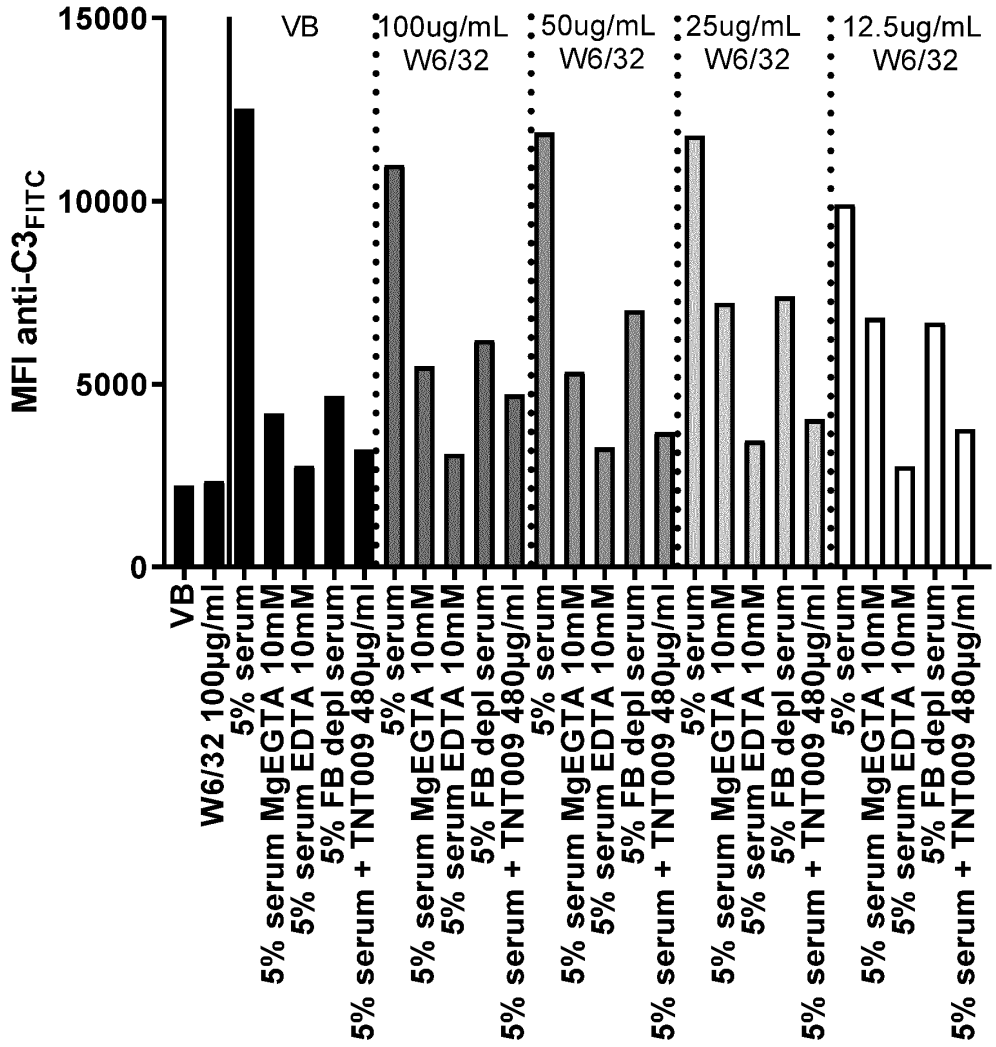

15000
10000
5000
0
MFI anti-C3FITC
VB
100ug/mL W6/32
50ug/mL W6/32
25ug/mL W6/32
12.5ug/mL W6/32
VB
W6/32 100µg/ml
5% serum
5% serum MgEGTA 10mM
5% serum EDTA 10mM
5% FB depl serum
5% serum + TNT009 480µg/ml
5% serum
5% serum MgEGTA 10mM
5% serum EDTA 10mM
5% FB depl serum
5% serum + TNT009 480µg/ml
5% serum
5% serum MgEGTA 10mM
5% serum EDTA 10mM
5% FB depl serum
5% serum + TNT009 480µg/ml
5% serum
5% serum MgEGTA 10mM
5% serum EDTA 10mM
5% FB depl serum
5% serum + TNT009 480µg/ml
5% serum
5% serum MgEGTA 10mM
5% serum EDTA 10mM
5% FB depl serum
5% serum + TNT009 480µg/ml

Fig. 6

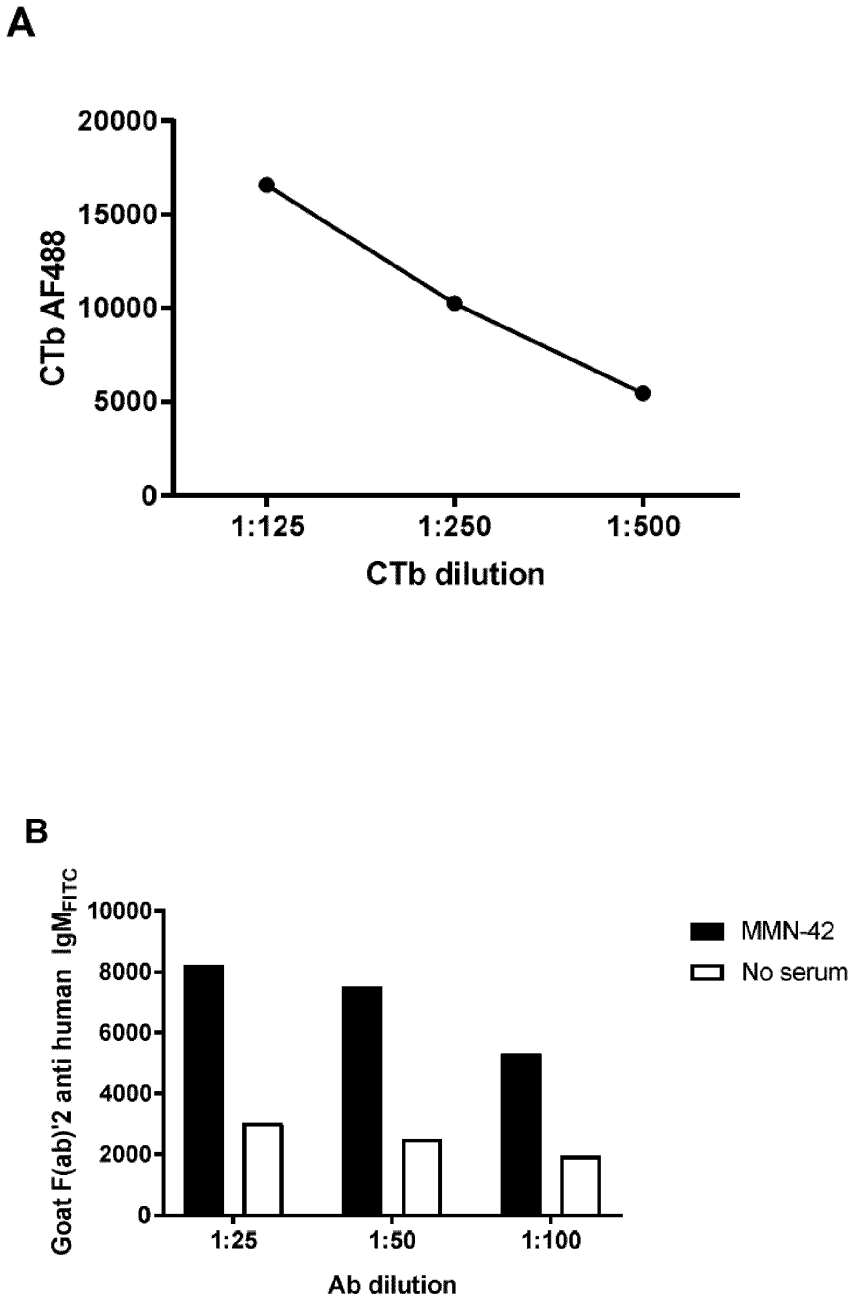
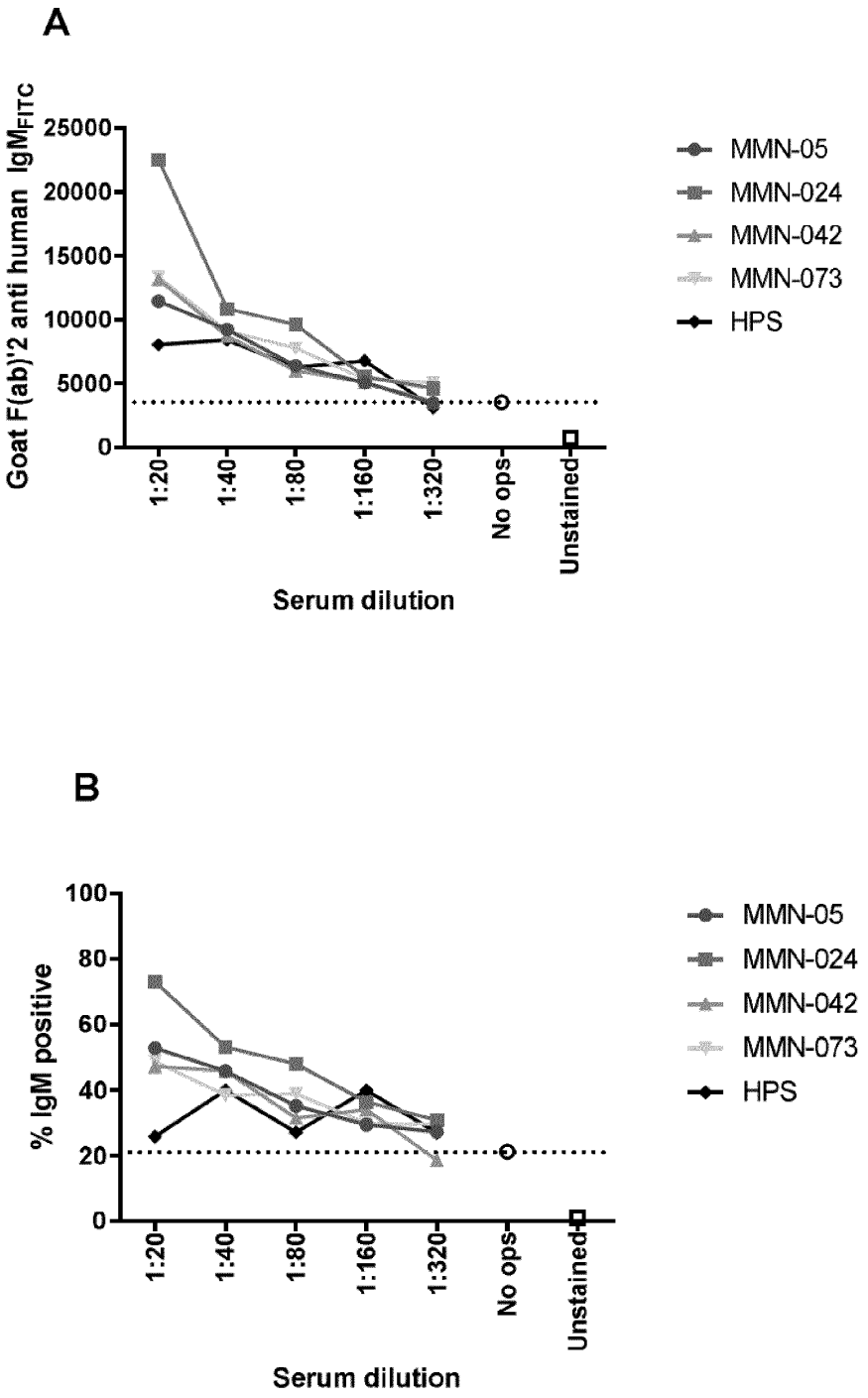

A
20000
15000
10000
5000
0
CTb AF488
1:125
1:250
1:500
CTb dilution
B
Goat F(ab)'2 anti human IgM FITC
10000
8000
6000
4000
2000
0
1:25
1:50
1:100
Ab dilution
MMN-42
No serum

Fig. 7


A
Goat F(ab)'2 anti human IgM FITC
25000
20000
15000
10000
5000
0
1:20
1:40
1:80
1:160
1:320
No ops
Unstained
Serum dilution
MMN-05
MMN-024
MMN-042
MMN-073
HPS
B
% IgM positive
100
80
60
40
20
0
1:20
1:40
1:80
1:160
1:320
No ops
Unstained
Serum dilution
MMN-05
MMN-024
MMN-042
MMN-073
HPS

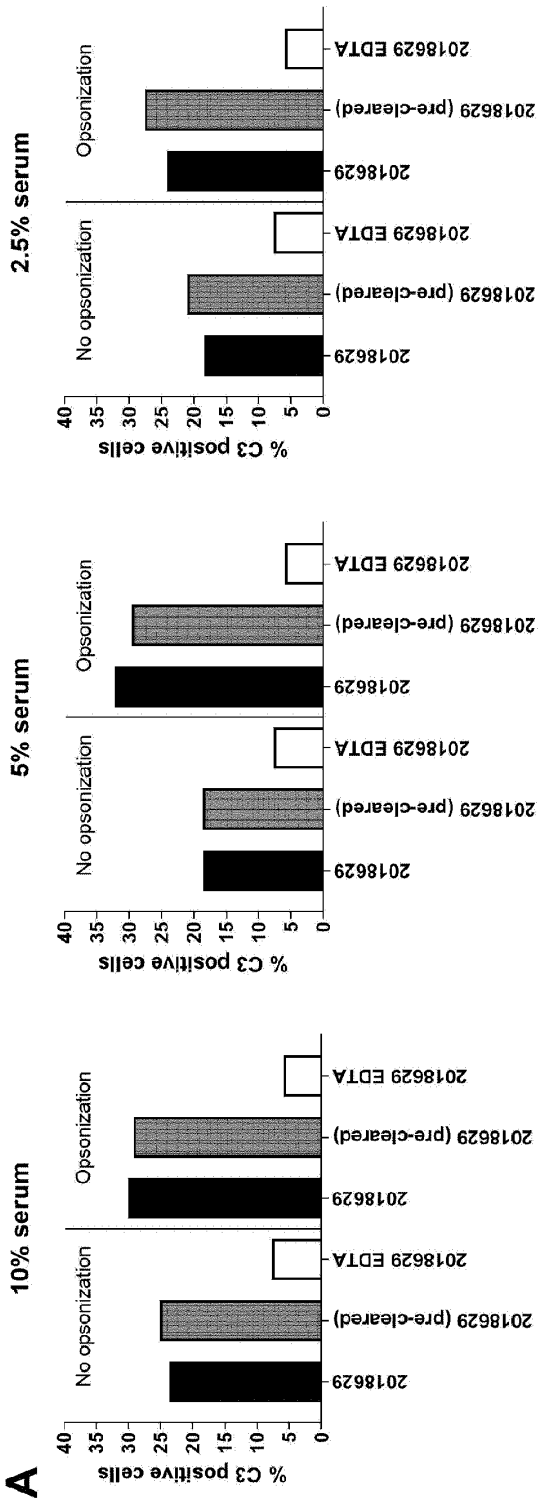

A
10% serum
5% serum
2.5% serum
% C3 positive cells
0
5
10
15
20
25
30
35
40
No opsonization
Opsonization
2018629
2018629 (pre-cleared)
2018629 EDTA

Fig. 8

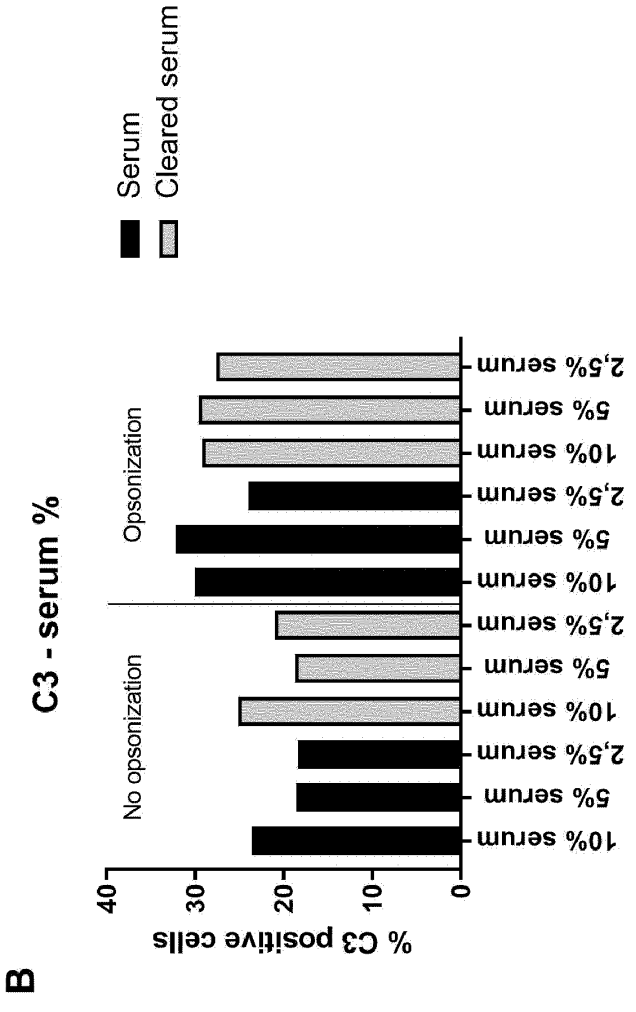


Fig. 8 (continued)

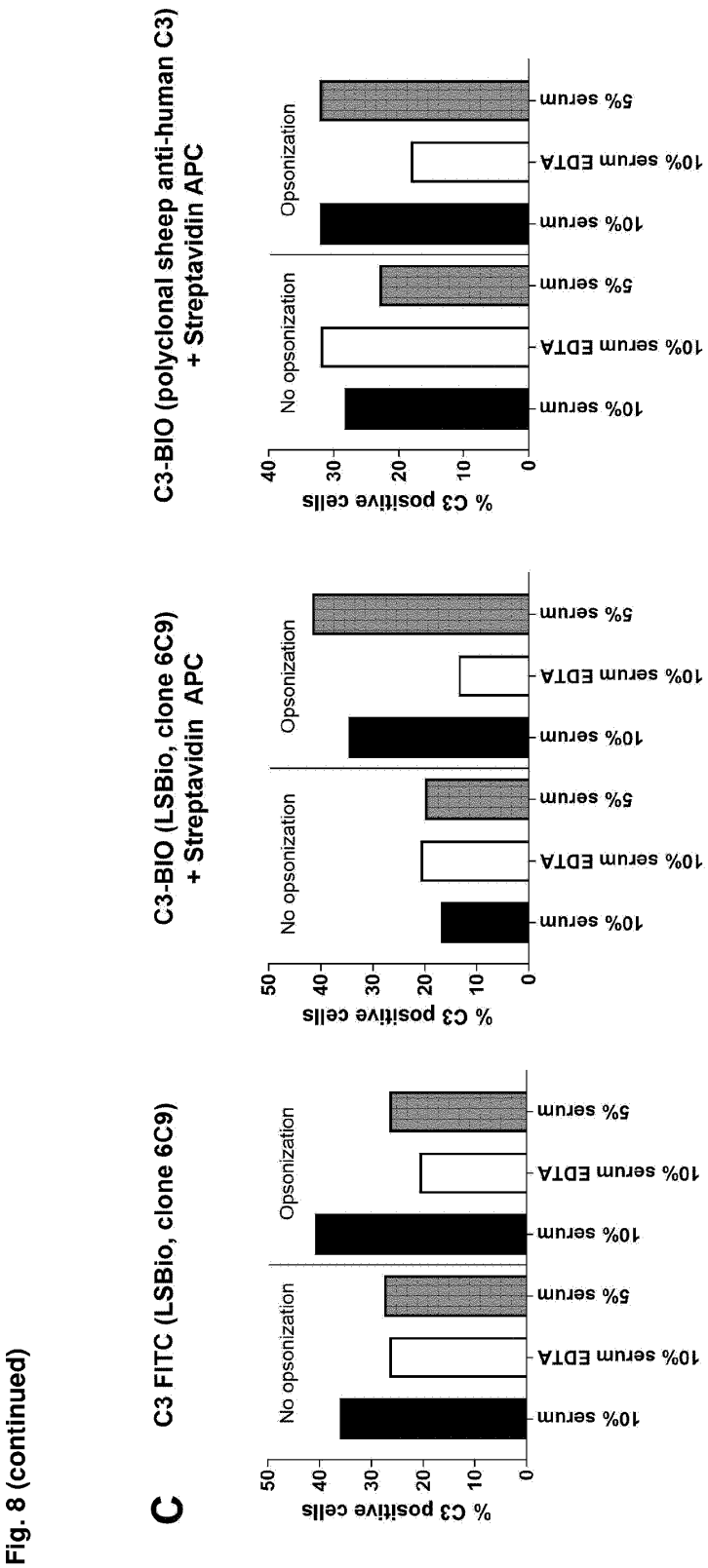


Fig. 8 (continued)

Fig. 9

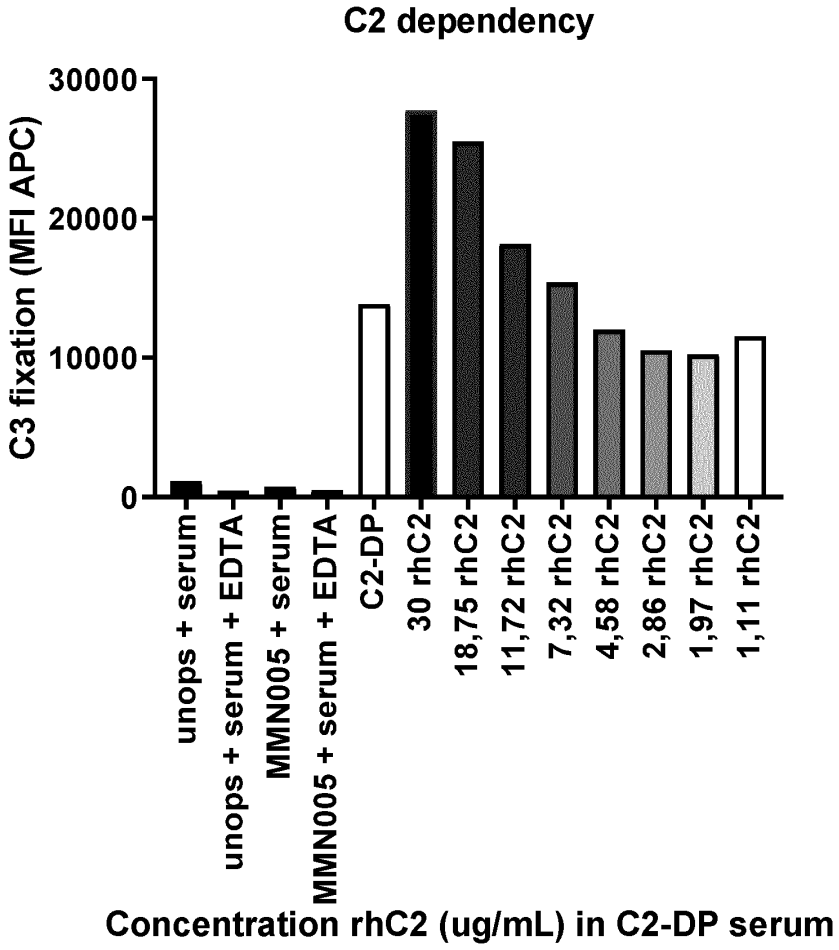
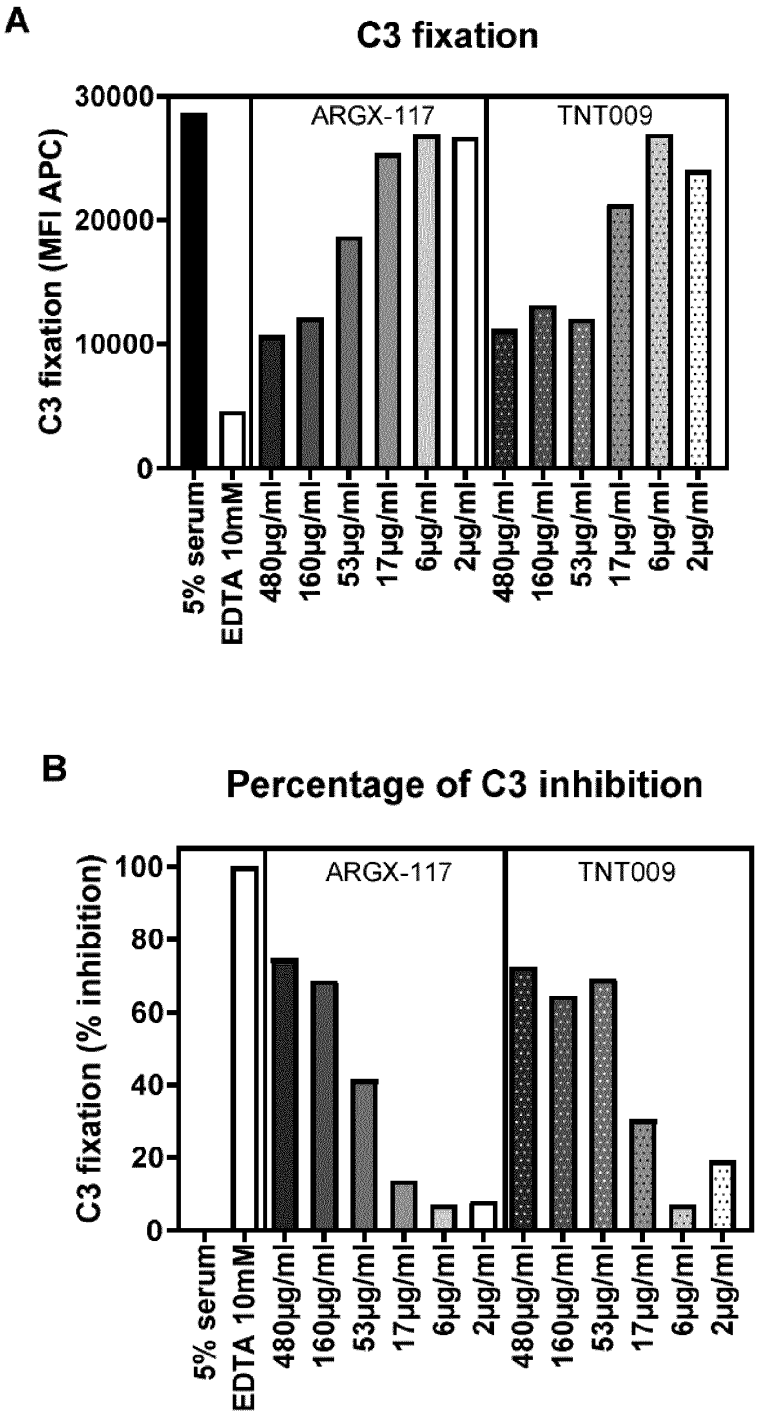
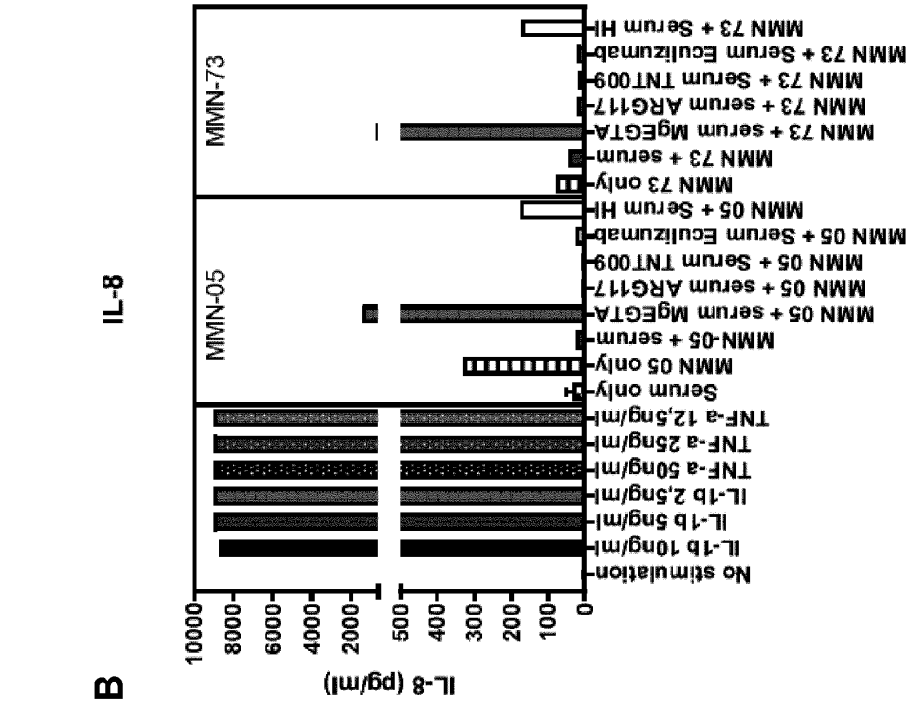

C2 dependency
C3 fixation (MFI APC)
30000
20000
10000
0
unops + serum
unops + serum + EDTA
MMN005 + serum
MMN005 + serum + EDTA
C2-DP
30 rhC2
18,75 rhC2
11,72 rhC2
7,32 rhC2
4,58 rhC2
2,86 rhC2
1,97 rhC2
1,11 rhC2
Concentration rhC2 (ug/mL) in C2-DP serum

Fig. 10


A
C3 fixation
C3 fixation (MFI APC)
30000
20000
10000
0
ARGX-117
TNT009
5% serum
EDTA 10mM
480µg/ml
160µg/ml
53µg/ml
17µg/ml
6µg/ml
2µg/ml
480µg/ml
160µg/ml
53µg/ml
17µg/ml
6µg/ml
2µg/ml
B
Percentage of C3 inhibition
C3 fixation (% inhibition)
100
80
60
40
20
0
ARGX-117
TNT009
5% serum
EDTA 10mM
480µg/ml
160µg/ml
53µg/ml
17µg/ml
6µg/ml
2µg/ml
480µg/ml
160µg/ml
53µg/ml
17µg/ml
6µg/ml
2µg/ml


B
IL-8
IL-8 (pg/ml)
10000
8000
6000
4000
2000
500
400
300
200
100
0
MMN-05
MMN-73
No stimulation
IL-1b 10ng/ml
IL-1b 5ng/ml
IL-1b 2,5ng/ml
TNF-a 50ng/ml
TNF-a 25ng/ml
TNF-a 12,5ng/ml
Serum only
MMN 05 only
MMN-05 + serum
MMN 05 + serum MgEGTA
MMN 05 + serum ARG117
MMN 05 + Serum TNT009
MMN 05 + Serum Eculizumab
MMN 05 + Serum HI
MMN 73 only
MMN 73 + serum
MMN 73 + serum MgEGTA
MMN 73 + serum ARG117
MMN 73 + Serum TNT009
MMN 73 + Serum Eculizumab
MMN 73 + Serum HI

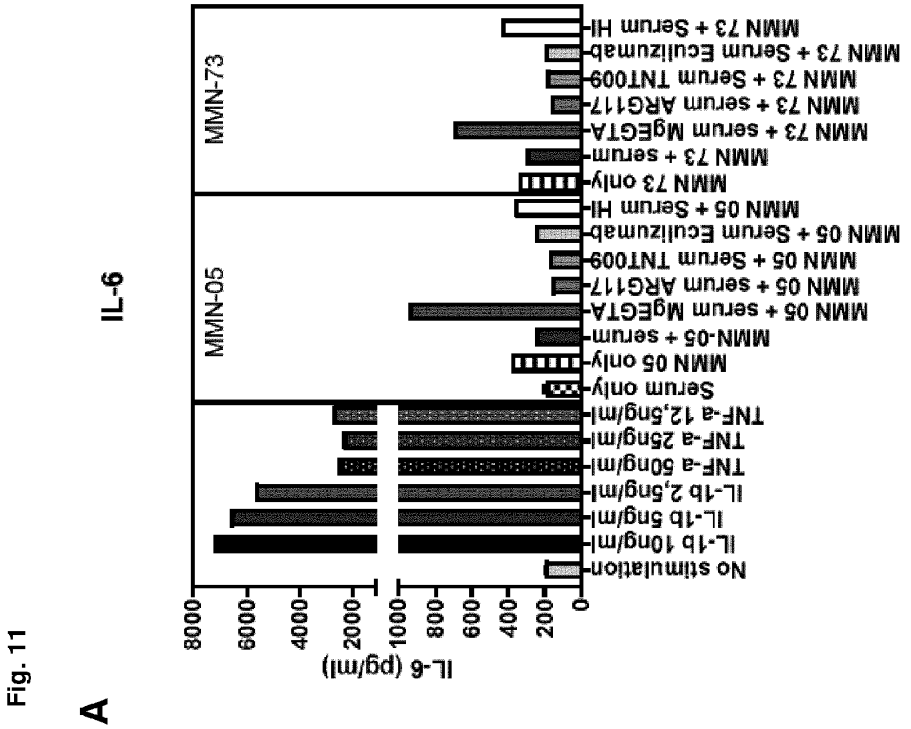
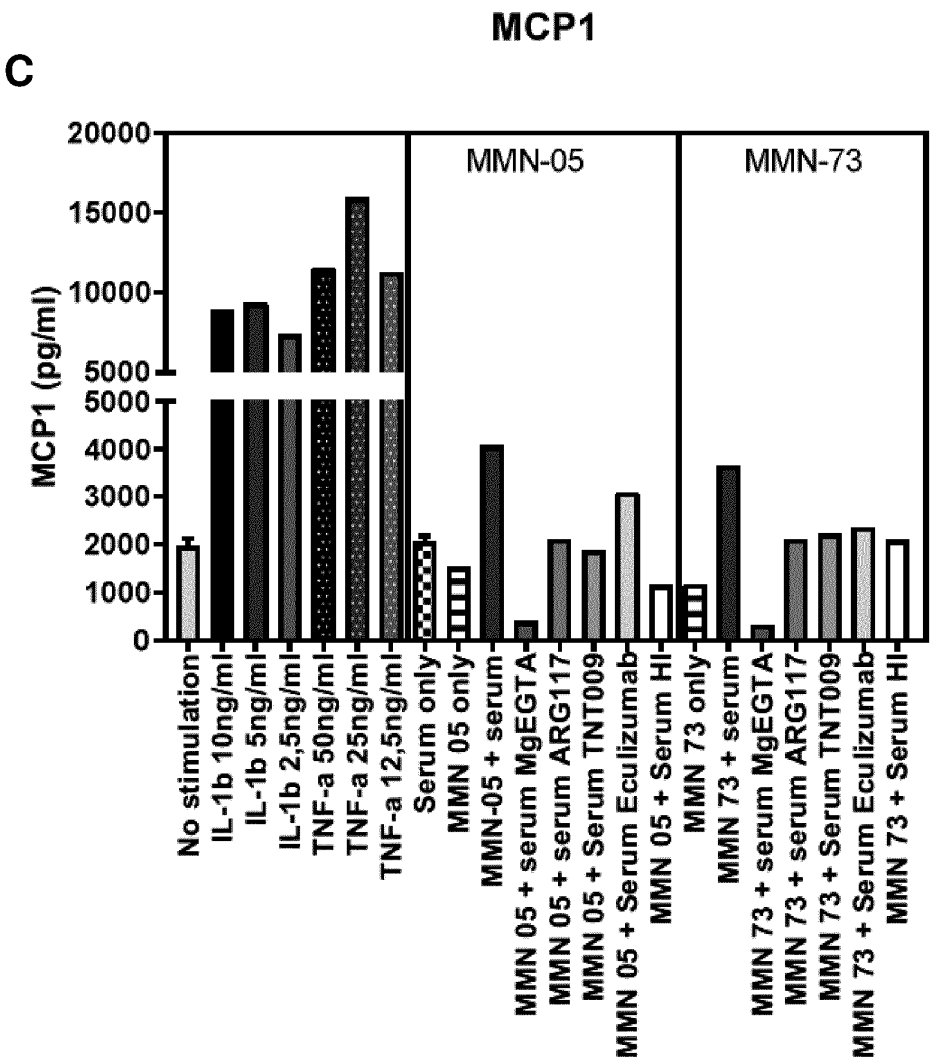

A
IL-6
IL-6 (pg/ml)
8000
6000
4000
2000
1000
800
600
400
200
0
MMN-05
MMN-73
No stimulation
IL-1b 10ng/ml
IL-1b 5ng/ml
IL-1b 2,5ng/ml
TNF-a 50ng/ml
TNF-a 25ng/ml
TNF-a 12,5ng/ml
Serum only
MMN 05 only
MMN-05 + serum
MMN 05 + serum MgEGTA
MMN 05 + serum ARG117
MMN 05 + Serum TNT009
MMN 05 + Serum Eculizumab
MMN 05 + Serum HI
MMN 73 only
MMN 73 + serum
MMN 73 + serum MgEGTA
MMN 73 + serum ARG117
MMN 73 + Serum TNT009
MMN 73 + Serum Eculizumab
MMN 73 + Serum HI

Fig. 11

Fig. 11 (continued)



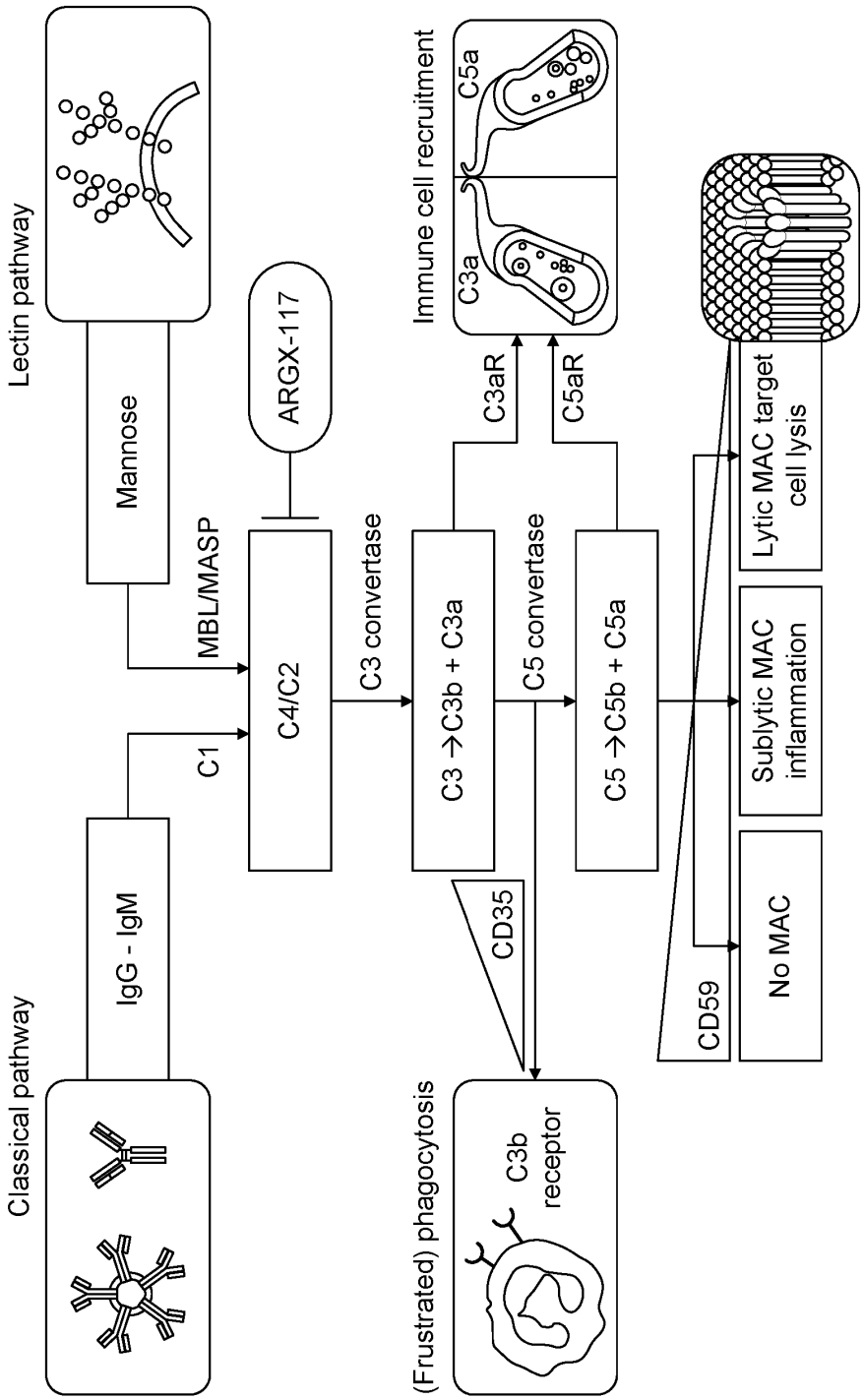

Lectin pathway
Mannose
MBL/MASP
ARGX-117
Immune cell recruitment
C5a
C3a
C3aR
C5aR
Lytic MAC target cell lysis
C4/C2
C3 convertase
C3 →C3b + C3a
C5 convertase
C5 →C5b + C5a
Sublytic MAC inflammation
C1
IgG - IgM
CD35
CD59
No MAC
Classical pathway
(Frustrated) phagocytosis
C3b receptor

Fig. 12

Fig. 13

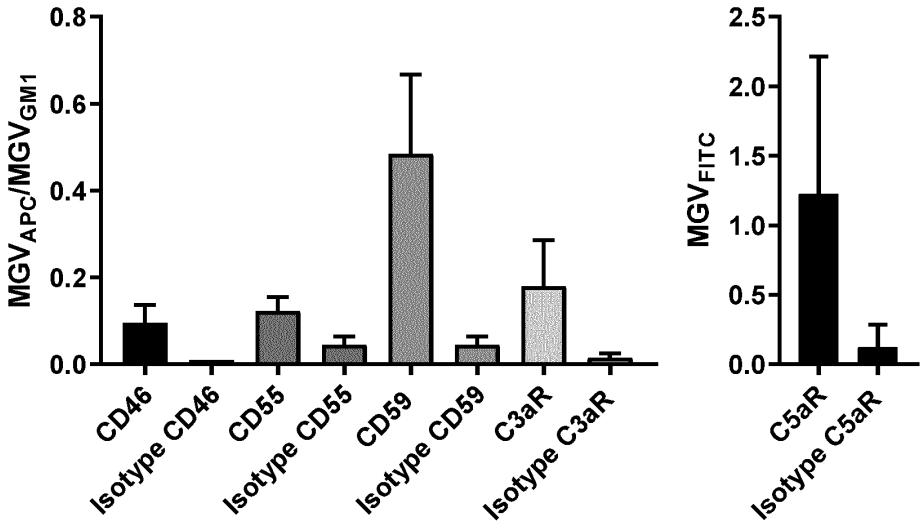

MGV$_{APC}$/MGV$_{GM1}$
0.8
0.6
0.4
0.2
0.0
CD46
Isotype CD46
CD55
Isotype CD55
CD59
Isotype CD59
C3aR
Isotype C3aR
MGV$_{FITC}$
2.5
2.0
1.5
1.0
0.5
0.0
C5aR
Isotype C5aR

Fig. 14

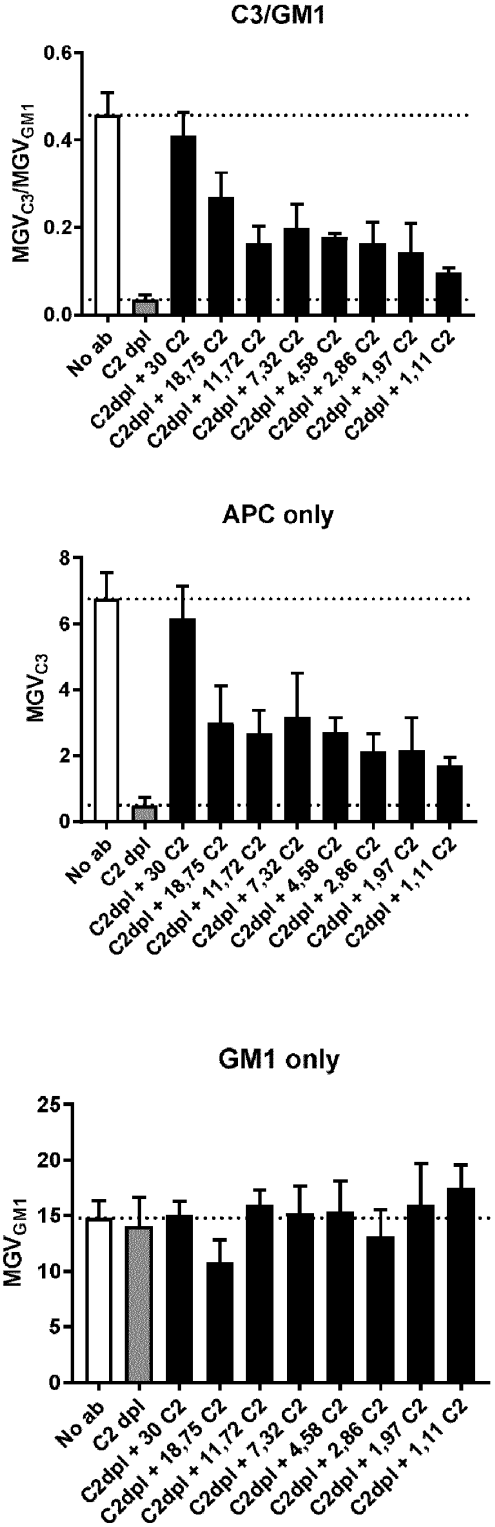
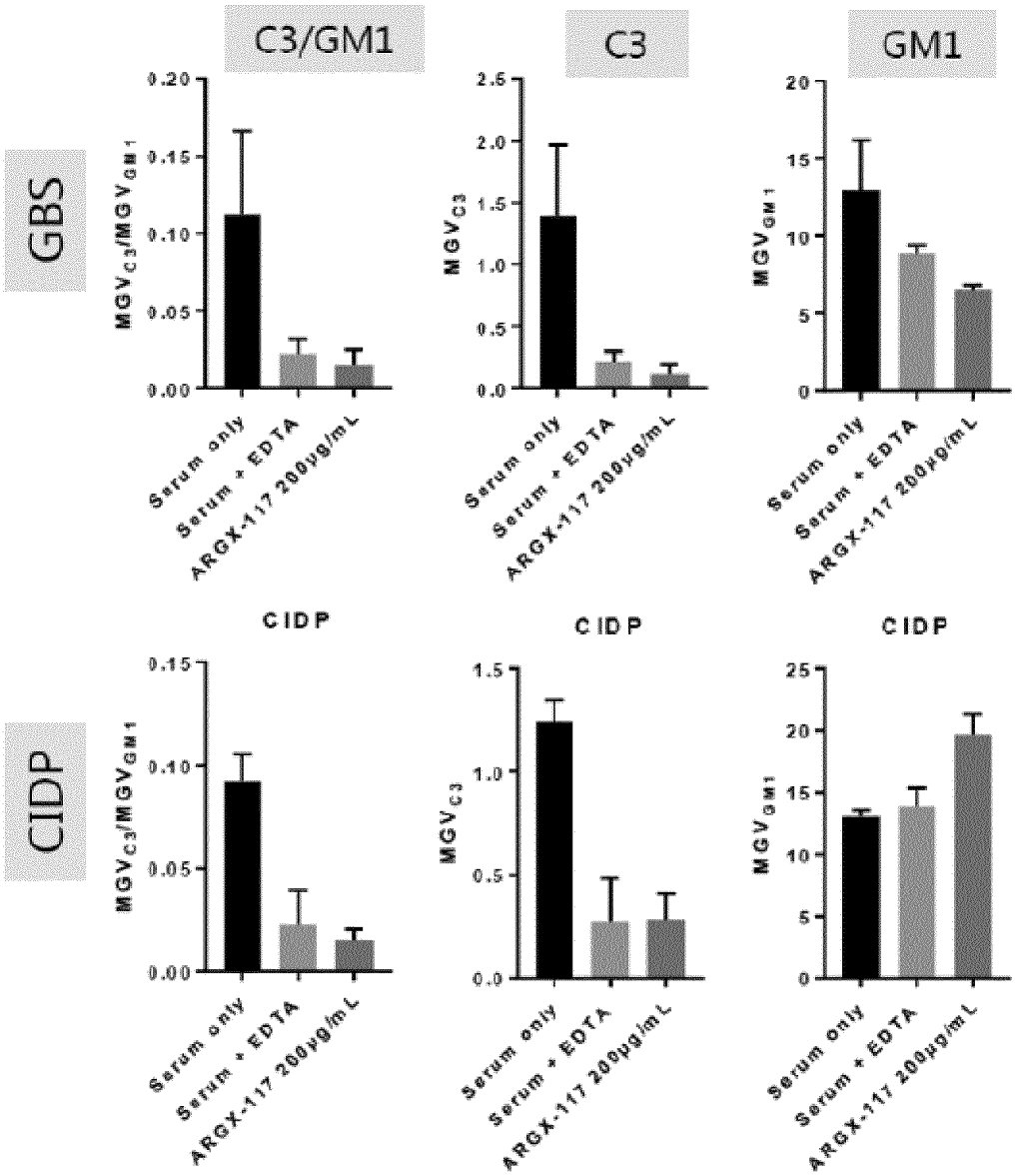

C3/GM1
MGVC3/MGVGM1
0.6
0.4
0.2
0.0
No ab
C2 dpl
C2dpl + 30 C2
C2dpl + 18,75 C2
C2dpl + 11,72 C2
C2dpl + 7,32 C2
C2dpl + 4,58 C2
C2dpl + 2,86 C2
C2dpl + 1,97 C2
C2dpl + 1,11 C2
APC only
MGVC3
8
6
4
2
0
GM1 only
MGVGM1
25
20
15
10
5
0

Fig. 15


C3/GM1
C3
GM1
GBS
CIDP
MGV C3/MGV GM1
MGV C3
MGV GM1
Serum only
Serum + EDTA
ARGX-117 200µg/mL

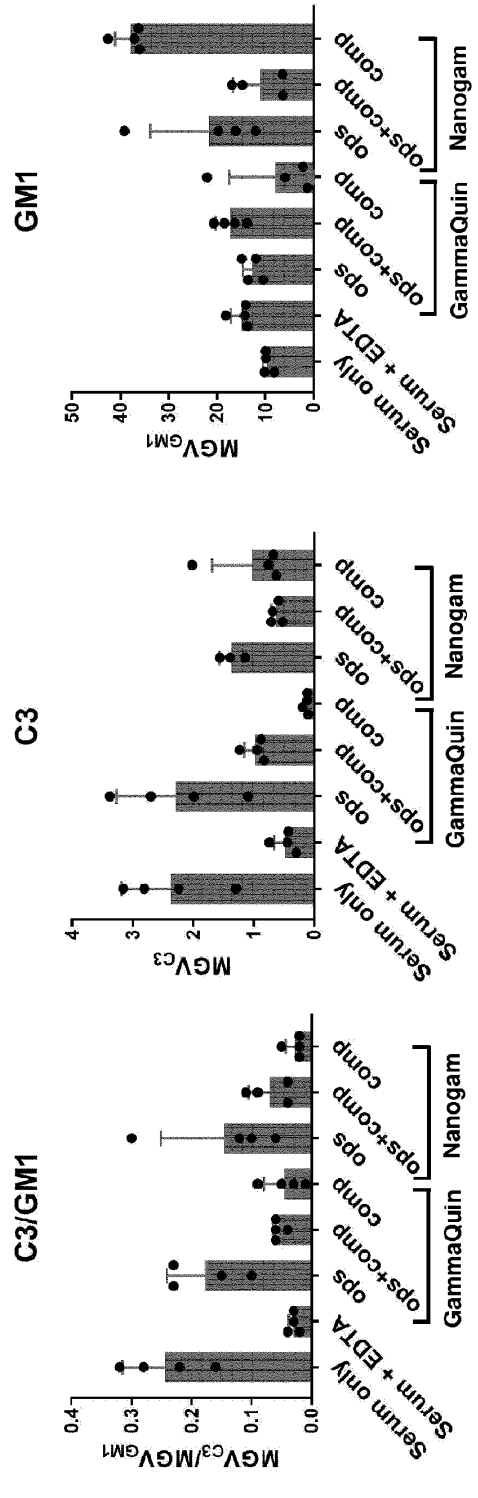
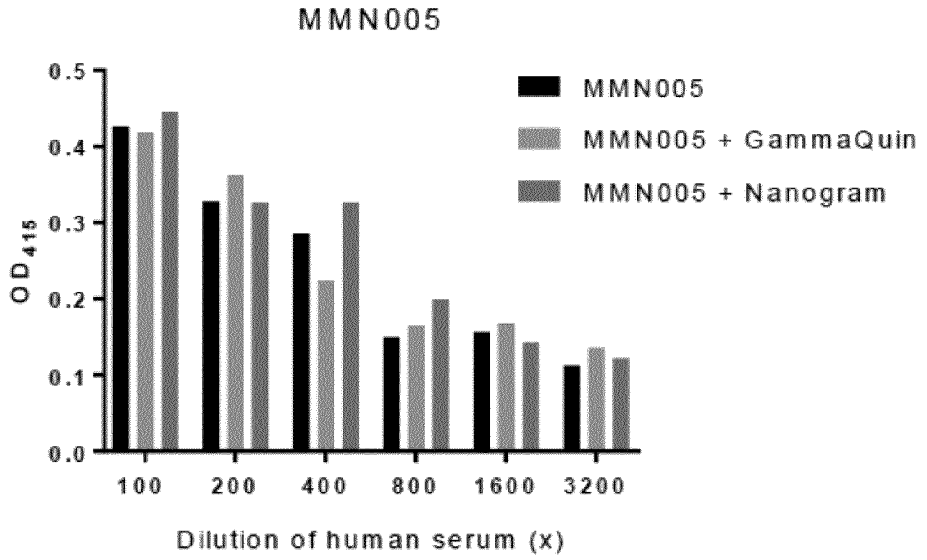
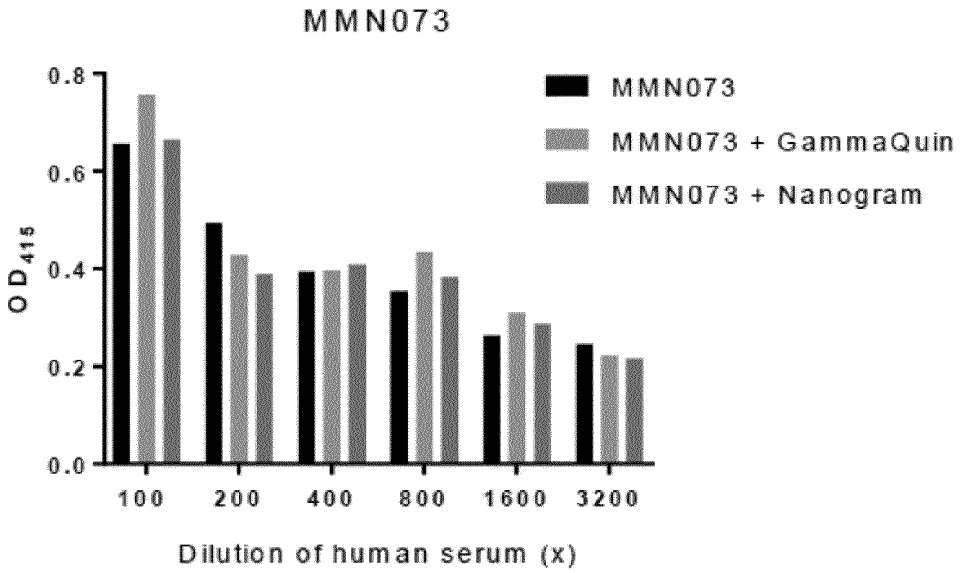

C3/GM1
MGVC3/MGVGM1
0.0
0.1
0.2
0.3
0.4
C3
MGVC3
0
1
2
3
4
GM1
MGVGM1
0
10
20
30
40
50
Serum only
Serum + EDTA
ops
ops+comp
comp
GammaQuin
Nanogam

Fig. 16

Fig. 17


MMN005
0.5
0.4
0.3
0.2
0.1
0.0
OD415
100
200
400
800
1600
3200
Dilution of human serum (x)
MMN005
MMN005 + GammaQuin
MMN005 + Nanogram


MMN073
0.8
0.6
0.4
0.2
0.0
OD415
100
200
400
800
1600
3200
Dilution of human serum (x)
MMN073
MMN073 + GammaQuin
MMN073 + Nanogram