(A) NI-308.18F7 VH (variable heavy chain sequence VH)

FR1----------CDR1-FR2----------CDR2--------
CQVQLQESGPGLVKPSETLSVTCTVSGGSINDYYWNWIRQPAGKGLEWIGRIYASGTINYNP
----FR3----------CDR3----------FR4--------
SLQSRVTMSIDTSKNQFSLDLISVSAADTAVYYCARWGQVVGDYYYGMDVWGQGTTVTVSS

NI-308.18F7 VK (variable light chain sequence VK)

FR1----------CDR1----------FR2----------CDR2---
CEIVLTQSPLSLSVTPGEPASISCKSSQSLQHTNGYNYLDWYLLKPGQSPQLLIFLTSNRAS
FR3----------CDR3--FR4---------
GVPDRFSGSGSGTNFTLKISRVEAEDVGVYYCMEGIQLWTFGQGTKLEIK

NI-308.18F7 VK - PIMC (variable light chain sequence VK)

FR1----------CDR1----------FR2----------CDR2---
C**D**IV**M**TQSPLSLSVTPGEPASISCKSSQSLQHTNGYNYLDWYLLKPGQSPQLLIFLTSNRAS
FR3----------CDR3–FR4----------
GVPDRFSGSGSGTNFTLKISRVEAEDVGVYYCMEGIQLWTFGQGTK**V**EIK

(B) NI-308.15O7 VH (variable heavy chain sequence VH)

FR1----------CDR1-FR2----------CDR2--------
CQVQLVESGGGVVQPGRSLRLSCAASGFTFSNHAMHWVRQAPGKGLEWVAVISYDGENTYYA
-----FR3----------CDR3----------FR4--------
DSIEGRFTISRDNFKNTLFLQMYSLTADDTAMYFCARGGRRGHFTSYYLDYWGQGTLVTVSS

NI-308.15O7 VK (variable light chain sequence VK)

FR1----------CDR1-------FR2----------CDR2---FR3--
CDIQMTQSPSSLSASVGDRVTITCRASQNIDKYLNWYQQIPGKAPKLLIYAASSLHSGVPSR
----------CDR3-----FR4-------
FSGSGSGTDFSLTISSLQPEDFAIYYCQQSYSSFRTFGQGTKLEIK

NI-308.15O7 VK - PIMC (variable light chain sequence VK)

FR1----------CDR1-------FR2----------CDR2---FR3--
CDIQMTQSPSSLSASVGDRVTITCRASQNIDKYLNWYQQIPGKAPKLLIYAASSLHSGVPSR
----------CDR3-----FR4-------
FSGSGSGTDFSLTISSLQPEDFAIYYCQQSYSSFRTFGQGTK**V**EIK

Fig. 1

(C) NI-308.28G1 VH (variable heavy chain sequence VH)

```
FR1----------CDR1------FR2----------CDR2--------
CQVQLQESGPRLVKPSETLSLTCTVAGGSVNSYYWTWIQQSPGKGLEWLGRIYIAGRTNYNP
----FR3----------CDR3----------FR4--------
SLTSRIALSVDTSRNQLSLKLTSVTAADTAIYYCARWGAESGDYYYGVDVWGPGTLVTVSS
```

NI-308.28G1 VK (variable light chain sequence VK)

```
FR1----------CDR1----------FR2----------CDR2---
CEIVMTQSPLSLPVTPGEPASISCKSSEGLLHSNGYTYLDWYLQKPGQAPQLLIFLASNRAA
FR3----------CDR3-----FR3-------
GVPDRFSGSGSGTDFTLKISRVEAEDVGVYYCMQAIQSPWTFGPGTKVEIK
```

NI-308.28G1 VH - PIMC (variable heavy chain sequence VH)

```
FR1----------CDR1------FR2----------CDR2--------
CQVQLQESGPRLVKPSETLSLTCTVAGGSVNSYYWTWIQQSPGKGLEWLGRIYIAGRTNYNP
----FR3----------CDR3----------FR4--------
SLTSRIALSVDTSRNQLSLKLTSVTAADTAIYYCARWGAESGDYYYGVDVWGPGTTVTVSS
```

NI-308.28G1 VK - PIMC (variable light chain sequence VK)

```
FR1----------CDR1----------FR2----------CDR2---
CDIVMTQSPLSLPVTPGEPASISCKSSEGLLHSNGYTYLDWYLQKPGQAPQLLIFLASNRAA
FR3----------CDR3-----FR3-------
GVPDRFSGSGSGTDFTLKISRVEAEDVGVYYCMQAIQSPWTFGPGTKVEIK
```

(D) NI-308.45C2 VH (variable heavy chain sequence VH)

```
FR1----------CDR1--------FR2----------CDR2------
CQLQLQESGPGLVKPSQILSLTCAISGDSVFSNSAAWNWIRQSPSRGLEWLGRTYYRSKWDN
--------FR3----------CDR3----------FR4---
DYAPSVKSRISINPDTSKNQFSLQLNSVTPEDTAVYYCAREVAYCGGDCYSVSFDYWGQGTL
-----
VTVSS
```

NI-308.45C2 VK (variable light chain sequence VK)

```
FR1----------CDR1----------FR2----------CDR2---
CEIVMTQSPLSLPVTPGEPASISCRSSQSLLQSNGYTYLDWYLQKPGQSPQLLIYLGSNRAS
FR3----------CDR3-----FR4-------
GVPDRFSGSGSGTDFTLKISRVEAEDVGVYYCMQALQTPLTFGGGTKVEIK
```

NI-308.45C2 VH - PIMC (variable heavy chain sequence VH)

```
FR1----------CDR1--------FR2----------CDR2------
CQLQLQQSGPGLVKPSQILSLTCAISGDSVFSNSAAWNWIRQSPSRGLEWLGRTYYRSKWDN
--------FR3----------CDR3----------FR4---
DYAPSVKSRISINPDTSKNQFSLQLNSVTPEDTAVYYCAREVAYCGGDCYSVSFDYWGQGTL
-----
VTVSS
```

Fig. 1 (continued)

NI-308. 45C2 VK - PIMC (variable light chain sequence VK)

FR1----------CDR1----------FR2----------CDR2---
C**D**IVMTQSPLSLPVTPGEPASISCRSSQSLLQSNGYTYLDWYLQKPGQSPQLLIYLGSNRAS
FR3----------CDR3-----FR4-------
GVPDRFSGSGSGTDFTLKISRVEAEDVGVYYCMQALQTPLTFGGGTKVEIK

(E) NI-308.24E11 VH (variable heavy chain sequence VH)

FR1----------CDR1------FR2----------CDR2--------
CQVQLQESGPGLVKPSETLSLTCTVSTTSLRSYFWSWIRQPPGKGLEWIGYVYYSGSTIYNP
----FR3----------CDR3----------FR3-----
SLKNRVTISIDTSKNQFSLNLRSVTAADTAMYFCARGVPAETDARDFPPYYFDHWGQGTLVT

VSS

NI-308.24E11 VK (variable light chain sequence VK)

FR1----------CDR1-------FR2----------CDR2---FR3--
CDIQLTQSPSSLSASVGNRITFTCQASQDIRYYLNWYQQKPGKAPKLLIYDVSNLDTGVPPR
----------CDR3-----FR4-------
FSGSGSGTNFTFTISSLQPEDIAVYYCQQYEGLPVTFGGGTKVEIK

NI-308.24E11 VK - PIMC (variable light chain sequence VK)

FR1----------CDR1-------FR2----------CDR2---FR3--
CDIQ**M**TQSPSSLSASVGNRITFTCQASQDIRYYLNWYQQKPGKAPKLLIYDVSNLDTGVPPR
----------CDR3-----FR4-------
FSGSGSGTNFTFTISSLQPEDIAVYYCQQYEGLPVTFGGGTKVEIK

(F) NI-308.5G2 VH (variable heavy chain sequence VH)

FR1----------CDR1------FR2----------CDR2--------
CEVQLVQSGADLRNPGASVTVSCTASGYHFADNAINWLRQAPGQRLEWMGWINIYSGNTKYS
-----FR3----------CDR3----------FR4--------
QNFQGRVTFTRNTSASTAFMHLRSLRSEDTAVYFCARDPDSSGYYLPYFDYWGQGTLVTVSS

NI-308.5G2 VK (variable light chain sequence VK)

FR1----------CDR1----------FR2----------CDR2--
CDIQLTQSPDSLTLSLGERATINCKSSQSVFYNSNNKNYLAWYQQKPGQPPKLLMYWASTRE
-FR3----------CDR3-----FR4-------
SGVTDRFSGSGSGTDFTLTITNLQAEDVAVYYCQQFYSSPLTFGGGTKVEIK

NI-308.5G2 VH - PIMC (variable heavy chain sequence VH)

FR1----------CDR1------FR2----------CDR2--------
C**Q**VQLVQSGADLRNPGASVTVSCTASGYHFADNAINWLRQAPGQRLEWMGWINIYSGNTKYS
-----FR3----------CDR3----------FR4--------
QNFQGRVTFTRNTSASTAFMHLRSLRSEDTAVYFCARDPDSSGYYLPYFDYWGQGTLVTVSS

Fig. 1 (continued)

NI-308.5G2 VK-PIMC (variable light chain sequence VK)

FR1----------CDR1----------FR2----------CDR2--
CDI**VM**TQSPDSLTLSLGERATINCKSSQSVFYNSNNKNYLAWYQQKPGQPPKLLMYWASTRE
-FR3----------CDR3-----FR4-------
SGVTDRFSGSGSGTDFTLTITNLQAEDVAVYYCQQFYSSPLTFGGGTKVEIK

(G) NI-308.46E9 VH (variable heavy chain sequence VH)

FR1----------CDR1--------FR2----------CDR2------
CQVQLQESGPGLVKPSETLSLTCTVSGASISGSTYYWGWIRQPPGKGLEYIGRIYYSGSTYY
------FR3----------CDR3---------FR4--------
NPSLKSRATISVDTSKNQLSLTLSSVTAADTAVYYCVRPFYAGSGNSPFDYWGQGTLVTVSS

NI-308.46E9 VK (variable light chain sequence VK)

FR1----------CDR1-------FR2----------CDR2---FR3--
CEIVLTQSPATVSVSPGERATLSCRASQSVSTNLAWYQQKPGQPPRLLIYGASTRATGIPAR
----------CDR3-----FR4-------
FSGSGSGAEFTLTISSLQSEDFVVYYCQQYNNWPPAFGGGTKVEIK

NI-308.46E9 VH - PIMC (variable heavy chain sequence VH)

FR1----------CDR1--------FR2----------CDR2------
CQ**L**QLQESGPGLVKPSETLSLTCTVSGASISGSTYYWGWIRQPPGKGLEYIGRIYYSGSTYY
------FR3----------CDR3---------FR4--------
NPSLKSRATISVDTSKNQLSLTLSSVTAADTAVYYCVRPFYAGSGNSPFDYWGQGTLVTVSS

NI-308.46E9 VK - PIMC (variable light chain sequence VK)

FR1----------CDR1-------FR2----------CDR2---FR3--
CEIV**M**TQSPATVSVSPGERATLSCRASQSVSTNLAWYQQKPGQPPRLLIYGASTRATGIPAR
----------CDR3-----FR4-------
FSGSGSGAEFTLTISSLQSEDFVVYYCQQYNNWPPAFGGGTKVEIK

(H) NI-308.6B11 VH (variable heavy chain sequence VH)

FR1----------CDR1------FR2----------CDR2--------
CEVQLVQSGAEVKRPGASVKVSCKVSGYTLTELSMHWVRQAPGKGLEWMGGFDPEDGETVYA
-----FR3----------CDR3---------FR4---
QKFQGRVTMTEDTSTDTAYMELSSLRSEDTALYHCATYGSSWHWNEGNEGSYYFDYWGQGTL

VTVSS

NI-308.6B11 VK (variable light chain sequence VK)

FR1----------CDR1-------FR2----------CDR2---FR3--
CDIQMTQSPSSLSASVGDRVTITCQASQDISIYLNWYQQKPGKAPKLLIYDASNLETGVPSR
----------CDR3-----FR4-------
FSGSGSGTDFTFTISGLQPEDVARYYCQQYDDLPITFGQGTRLEIK

Fig. 1 (continued)

NI-308.6B11 VH - PIMC (variable heavy chain sequence VH)

```
FR1----------CDR1------FR2----------CDR2--------
CQVQLVQSGAEVKRPGASVKVSCKVSGYTLTELSMHWVRQAPGKGLEWMGGFDPEDGETVYA
-----FR3-----------------------------CDR3---------------FR4---
QKFQGRVTMTEDTSTDTAYMELSSLRSEDTALYHCATYGSSWHWNEGNEGSYYFDYWGQGTL
-----
VTVSS
```

(I) NI-308.46F8 VH (variable heavy chain sequence VH)

```
FR1----------CDR1------FR2----------CDR2--------
CQVQLVESGGGVVRPGGSLRLSCTASGFTFDEYGMSWVRQVPGKGLEWVSGINWNGATTRYA
-----FR3-----------------------------CDR3------------FR4------
DSVKGRFTISRDNAKNSLYLQMNSLRAEDTALYHCARDGCRNTSCYIWDWFDPWGQGTLVTV
--
SS
```

NI-308.46F8 VL (variable light chain sequence VL)

```
FR1--------------------CDR1---------FR2------------CDR2---FR3-
CQSVLTQPPSVSAAPGQKVTISCSGSSSNIGNNYVCWYQNLPGTAPKLLIYDDNKRPSGIPD
----------------------------CDR3-------FR4-------
RFSGSKSGTSATLGITGLQTGDEADYYCGTWDSSLSVVVFGGGTKLTVL
```

NI-308.46F8 VH - PIMC (variable heavy chain sequence VH)

```
FR1----------CDR1------FR2----------CDR2--------
CEVQLVESGGGVVRPGGSLRLSCTASGFTFDEYGMSWVRQVPGKGLEWVSGINWNGATTRYA
-----FR3-----------------------------CDR3------------FR4------
DSVKGRFTISRDNAKNSLYLQMNSLRAEDTALYHCARDGCRNTSCYIWDWFDPWGQGTLVTV
--
SS
```

(J) NI-308.4M1 VH (variable heavy chain sequence VH)

```
FR1----------------------------CDR1-FR2----------CDR2--------
CEVQLVETGGGVVQPGGSLRLSCEASGFTIGTYAMHWVRQFPGKGLDWVAVISFDGTTEYYT
---FR3-------------------------------CDR3-------------FR4-----
DAVKGRFTISRDNAKNTLYLQMNYLRGDDTAIYFCARDFTSSGETGSWTQVPDLWGQGTLVT
---
VSS
```

NI-308.4M1 VK (variable light chain sequence VK)

```
FR1---------------------CDR1-------FR2------------CDR2---FR3--
CEIVMTQSPATLSLSPGERATLSCRASQSVTKYLAWYQQKPGQAPRLLIYDVSYRAAGTPAR
--------------------------CDR3------FR4-------
FSGSGSGTDFTLTISSLEPEDFAVYYCHQRSSWPPVTFGGGTKVEIK
```

Fig. 1 (continued)

(J) NI-308.4M1 VH - PIMC (variable heavy chain sequence VH)

FR1----------CDR1-FR2----------CDR2--------
C**Q**VQLVE**S**GGGVVQPGGSLRLSCEASGFTIGTYAMHWVRQFPGKGLDWVAVISFDGTTEYYT
---FR3----------CDR3----------FR4-----
DAVKGRFTISRDNAKNTLYLQMNYLRGDDTAIYFCARDFTSSGETGSWTQVPDLWGQGTLVT

VSS

NI-308.4M1 VK - PIMC (variable light chain sequence VK)

FR1----------CDR1-------FR2----------CDR2---FR3--
CEIV**L**TQSPATLSLSPGERATLSCRASQSVTKYLAWYQQKPGQAPRLLIYDVSYRAAGTPAR
----------CDR3------FR4-------
FSGSGSGTDFTLTISSLEPEDFAVYYCHQRSSWPPVTFGGGTKVEIK

(K) NI-308.12A3 VH (variable heavy chain sequence VH)

FR1----------CDR1-FR2----------CDR2--------
CEVQLVETGGGLVQPGGSLRLSCVGSGFLFSDFEMDWVRQAPGKGLEWISYISGDGNIIYQT
-----FR3----------CDR3----------FR4------
DSVKGRFTISRDNAKNSLFLQMDSLTVEDTAVYYCARDARENCGGDCYSTSFDFWGQGTLVT

VSS

NI-308.12A3 VK (variable light chain sequence VK)

FR1----------CDR1----------FR2----------CDR2--
CDIQMTQSPDSLAVSLGERATINCKSSQSLLYTANNRNYLAWYQKKAGQPPKLLIHWASTRA
-FR3----------CDR3-----FR4-------
SGVPDRFSGSGSGTDFILTISSLQAEDVAVYFCQHYYNSPRTFGQGTKVEIK

NI-308.12A3 VH - PIMC (variable heavy chain sequence VH)

FR1----------CDR1-FR2----------CDR2--------
CEVQLVE**S**GGGLVQPGGSLRLSCVGSGFLFSDFEMDWVRQAPGKGLEWISYISGDGNIIYQT
-----FR3----------CDR3----------FR4------
DSVKGRFTISRDNAKNSLFLQMDSLTVEDTAVYYCARDARENCGGDCYSTSFDFWGQGTLVT

VSS

NI-308.12A3 VK - PIMC (variable light chain sequence VK)

FR1----------CDR1----------FR2----------CDR2--
CDI**V**MTQSPDSLAVSLGERATINCKSSQSLLYTANNRNYLAWYQKKAGQPPKLLIHWASTRA
-FR3----------CDR3-----FR4-------
SGVPDRFSGSGSGTDFILTISSLQAEDVAVYFCQHYYNSPRTFGQGTKVEIK

Fig. 1 (continued)

(L) NI-308.16C10 VH (variable heavy chain sequence VH)

FR1----------CDR1--FR2----------CDR2--------
CEVQLVETGGGVVQPGMSLSLSCAATGFTFSSYGMHWVRQGPGKGPEWVAGIWYDGTNKYYG
-----FR3----------CDR3----------FR4-------
DSVTGRVTISRDNSKNTLFLQMINVRVEDTAVYYCVKDAERVQKWASYIMDVWGQGTTVTVS
-
S

NI-308.16C10 VK (variable light chain sequence VK)

FR1----------CDR1--------FR2----------CDR2---FR3-
CEIVLTQSPGILSLSRGNRVALSCRASRSVNSSYLNWYQQKPGQAPRLLIYGASKRATGISD
----------CDR3--FR4-------
RFRGTGSGTDFTLTVARLEPEDIAVYYCQHYGAFGQGTKLEIK

NI-308.16C10 VH - PIMC (variable heavy chain sequence VH)

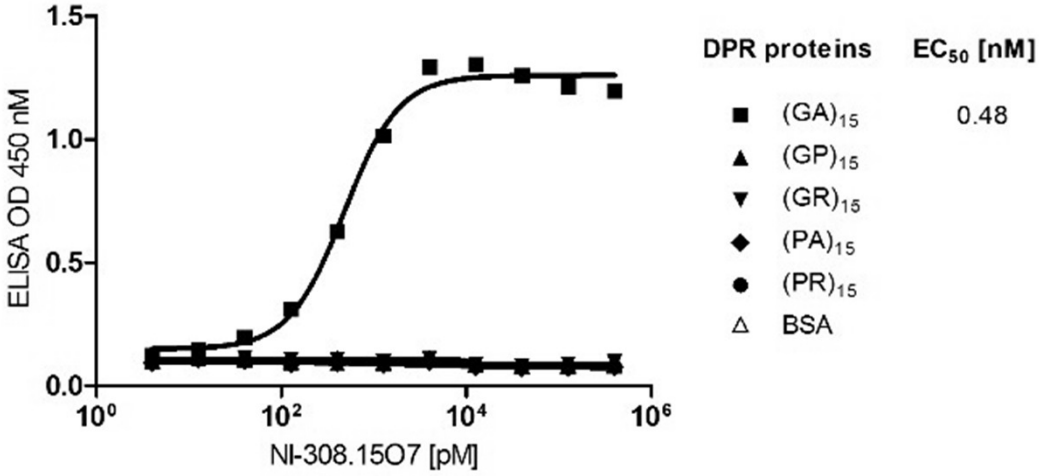
FR1----------CDR1--FR2----------CDR2--------
C**Q**VQLVE**S**GGGVVQPGMSLSLSCAATGFTFSSYGMHWVRQGPGKGPEWVAGIWYDGTNKYYG
-----FR3----------CDR3----------FR4-------
DSVTGRVTISRDNSKNTLFLQMINVRVEDTAVYYCVKDAERVQKWASYIMDVWGQGTTVTVS
-
S

NI-308.16C10 VK - PIMC (variable light chain sequence VK)

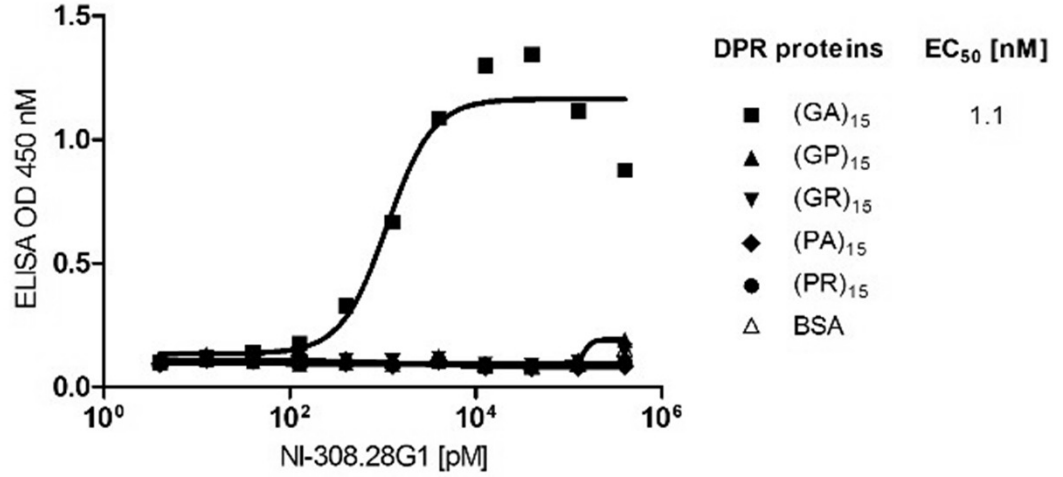
FR1----------CDR1--------FR2----------CDR2---FR3-
CEIVLTQSPGILSLSRGNRVALSCRASRSVNSSYLNWYQQKPGQAPRLLIYGASKRATGISD
----------CDR3--FR4-------
RFRGTGSGTDFTLTVARLEPEDIAVYYCQHYGAFGQGTK**V**EIK

Fig. 1 (continued)

(B) NI-308.15O7



(C) NI-308.28G1



(D) NI-308.45C2

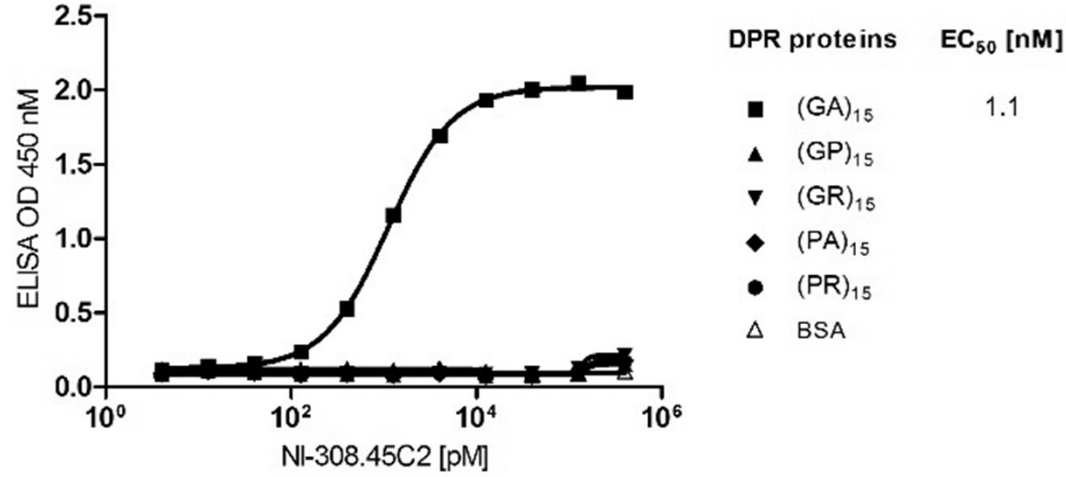
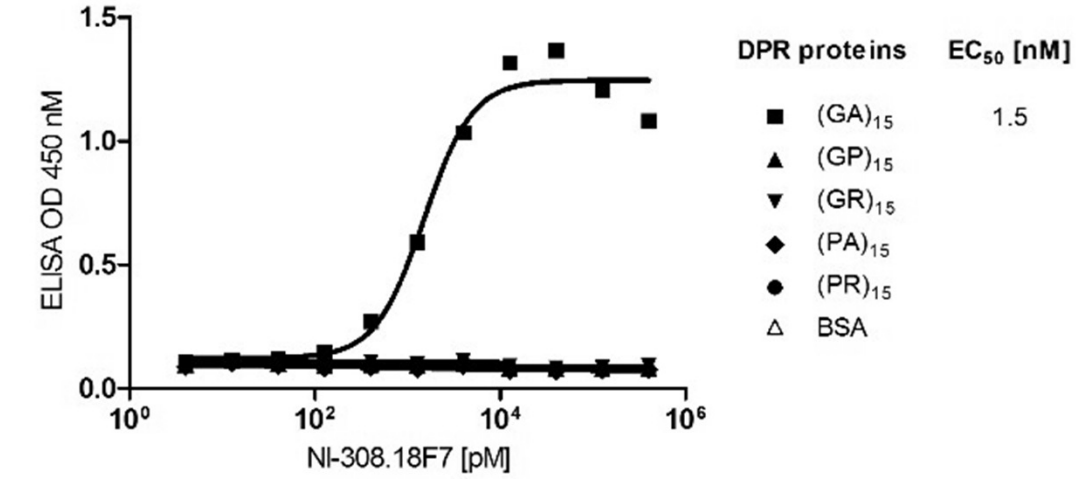
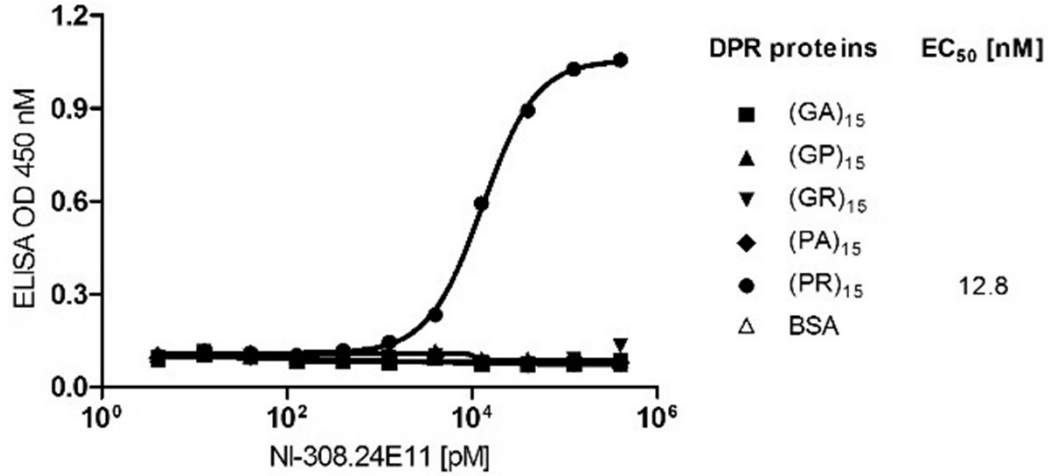


Fig. 2

(E) NI-308.18F7



(F) NI-308.24E11



(G) NI-308.5G2

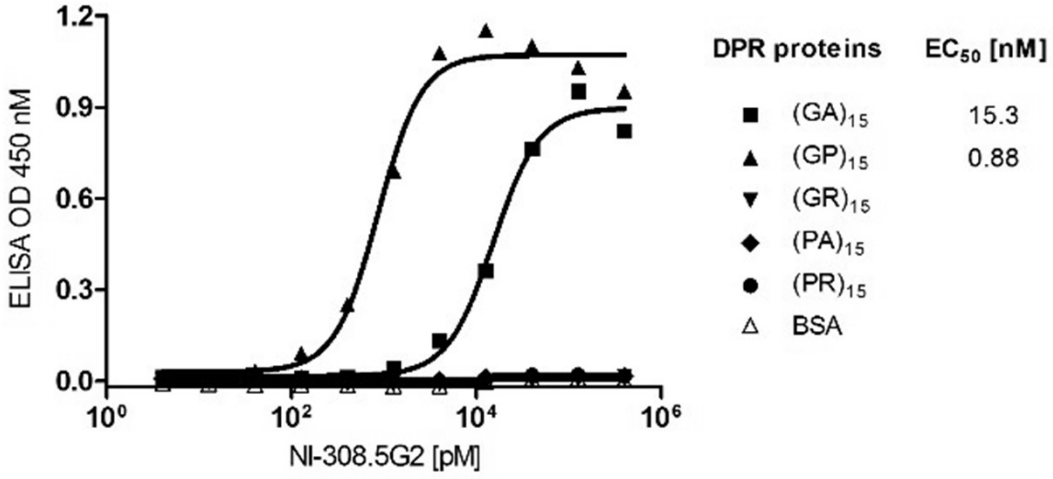
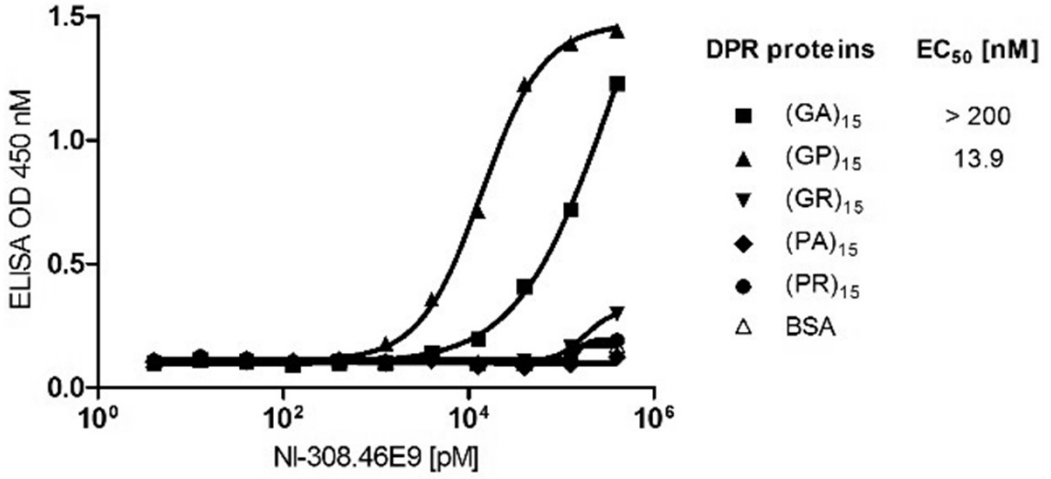
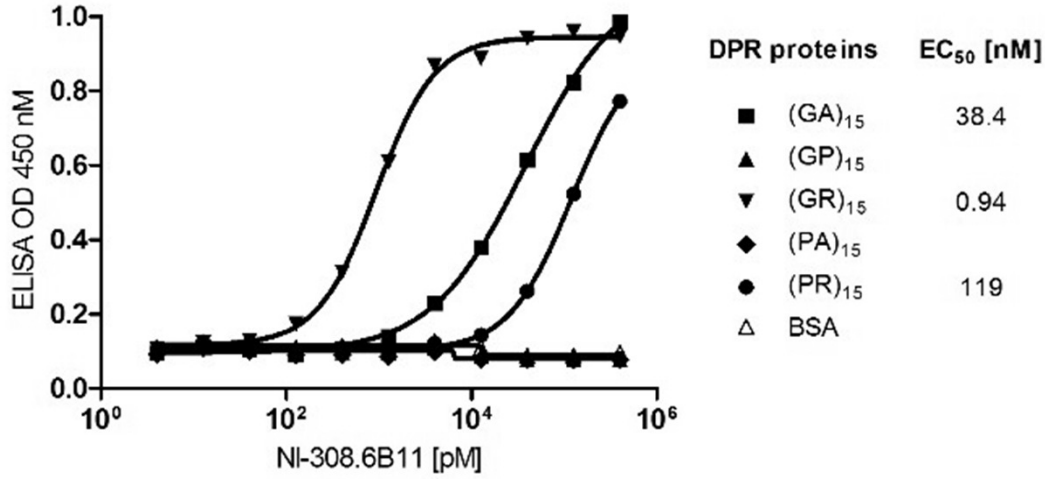


Fig. 2 (continued)

(H) NI-308.46E9



(I) NI-308.6B11



(J) NI-308.46F8

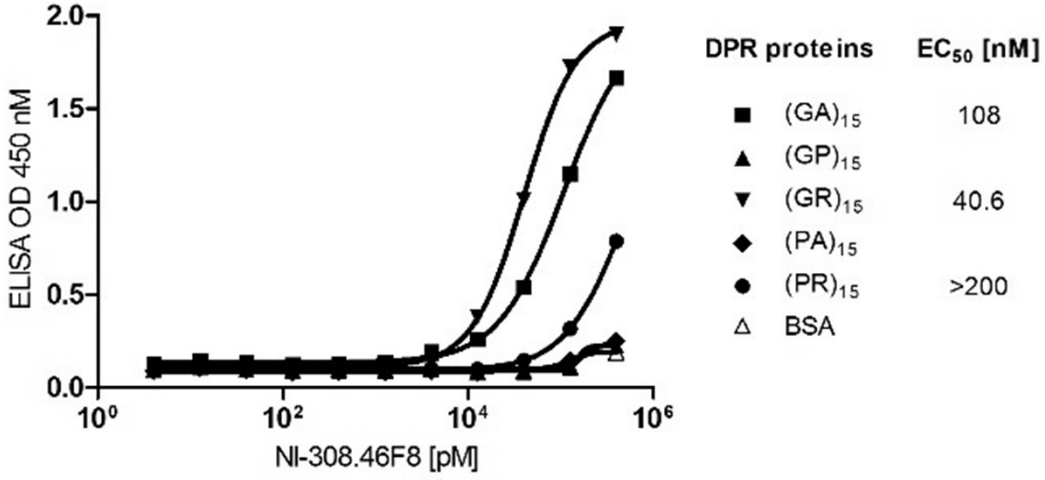
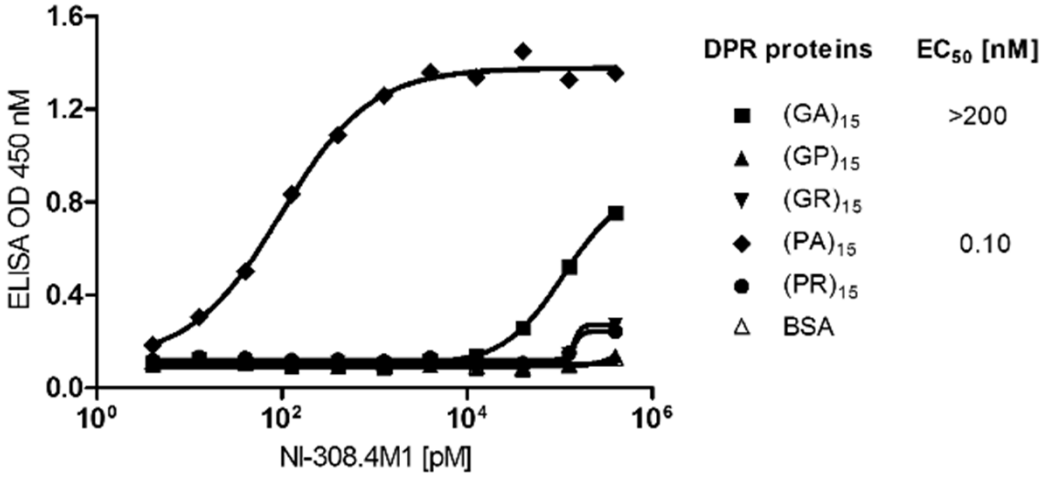
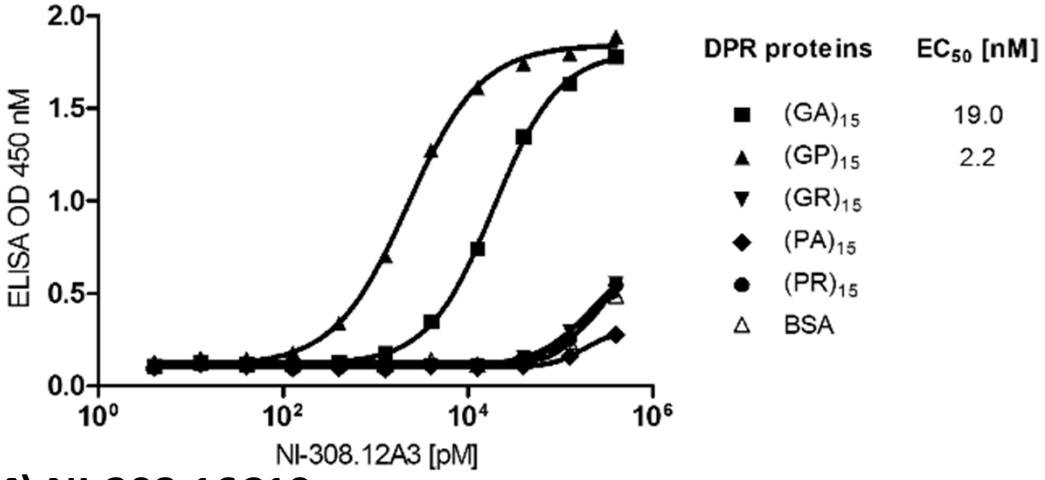


Fig. 2 (continued)

(K) NI-308.4M1



(L) NI-308.12A3



(M) NI-308.16C10

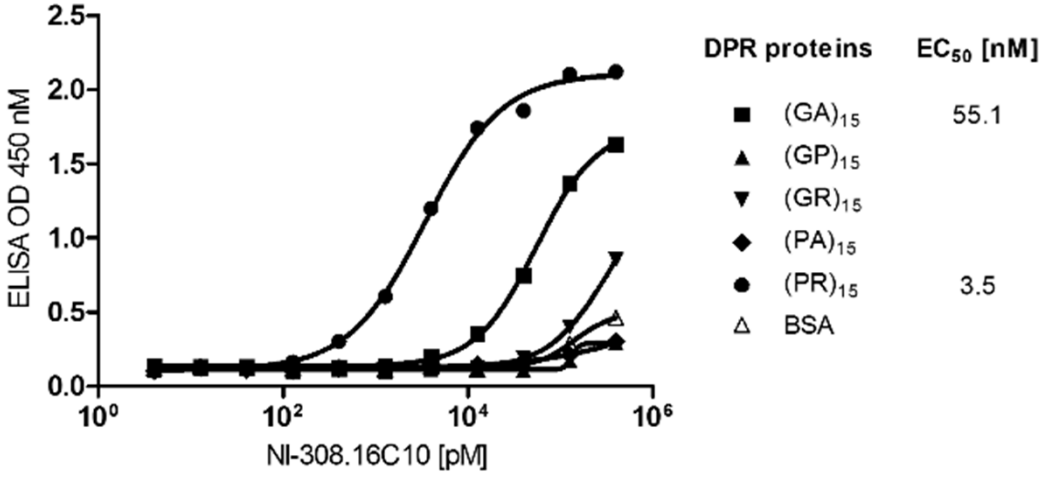
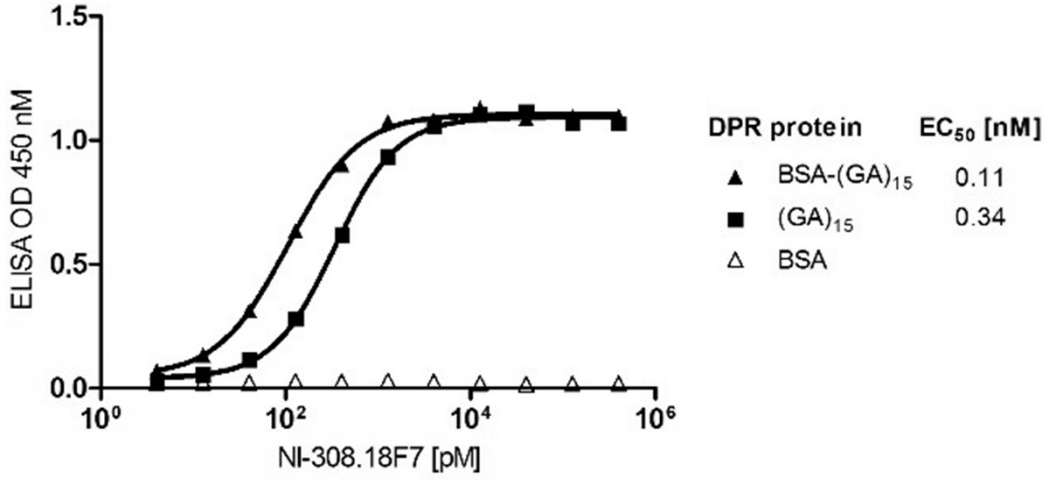


Fig. 2 (continued)

(A)

		EC_{50} [nM]	
Antibody	**Peptide**	**BSA-coupled peptide**	**Uncoupled peptide**
NI-308.18F7	$(GA)_{15}$	0.11	0.34
NI-308.15O7	$(GA)_{15}$	0.41	0.18
NI-308.28G1	$(GA)_{15}$	0.66	2.0
NI-308.45C2	$(GA)_{15}$	15.2	1.1
NI-308.24E11	$(PR)_{15}$	> 200	11.3
NI-308.16C10	$(PR)_{15}$	91.1	3.5
NI-308.5G2	$(GP)_{15}$	0.21	0.40
NI-308.12A3	$(GP)_{15}$	6.1	2.2
NI-308.46E9	$(GP)_{15}$	12.1	22.0
NI-308.6B11	$(GR)_{15}$	NA	1.3
NI-308.46F8	$(GR)_{15}$	NA	90.3
NI-308.4M1	$(PA)_{15}$	0.14	0.10

(B) NI-308.18F7



(C) NI-308.15O7

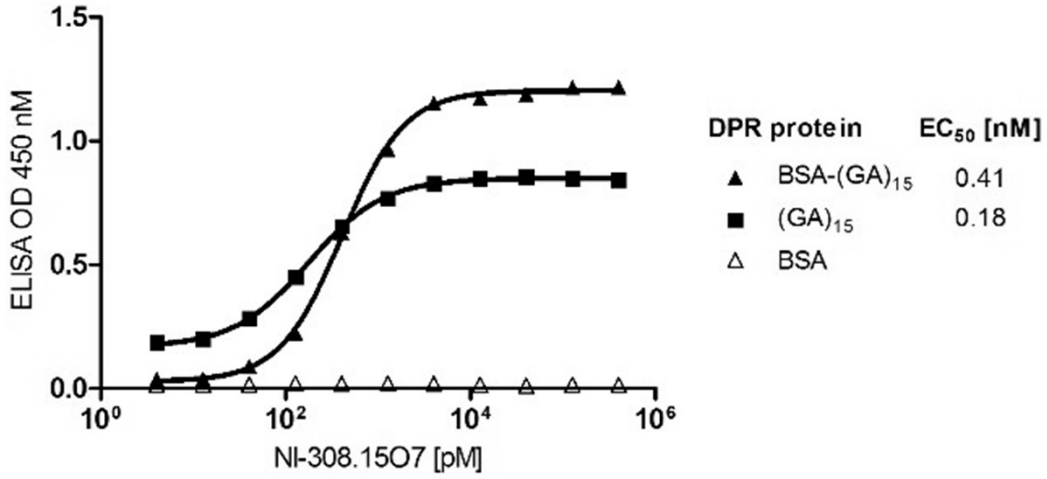
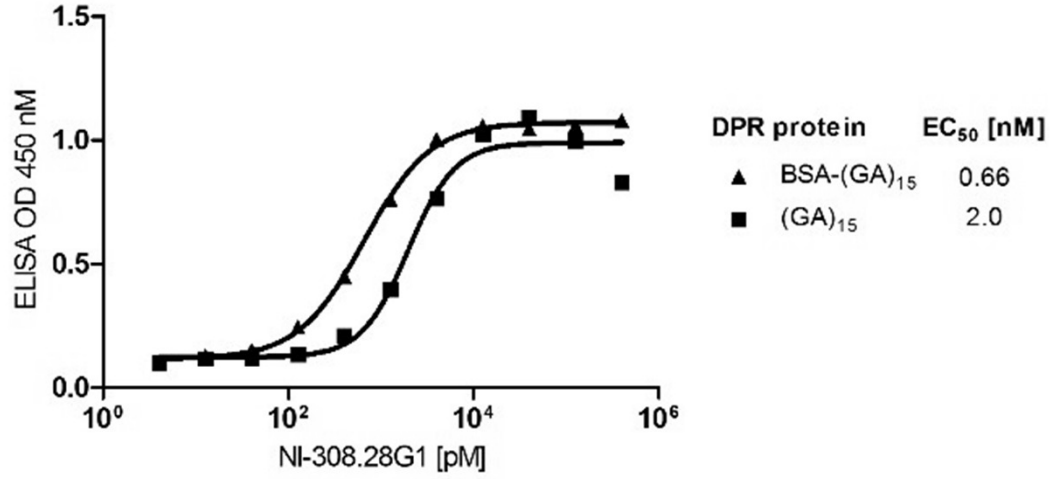
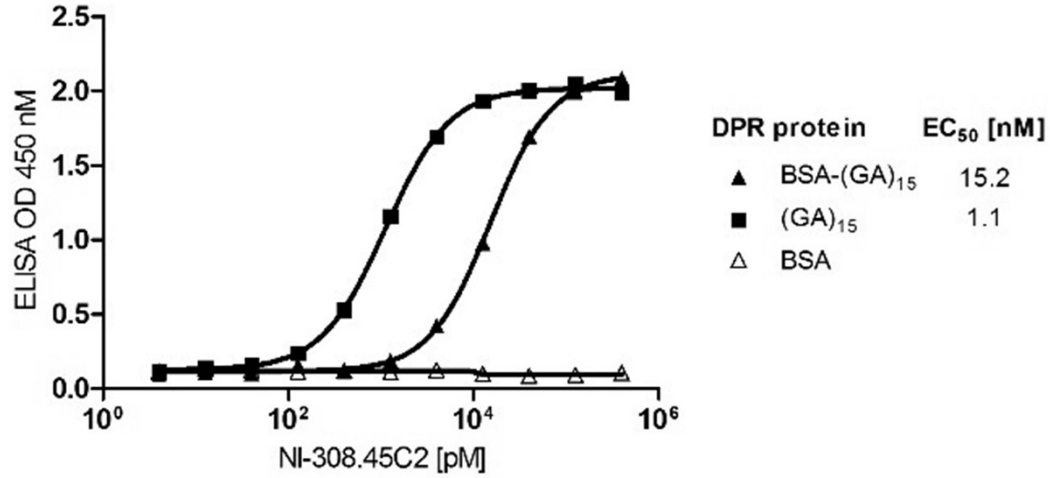


Fig. 3

(D) NI-308.28G1



(E) NI-308.45C2



(F) NI-308.24E11

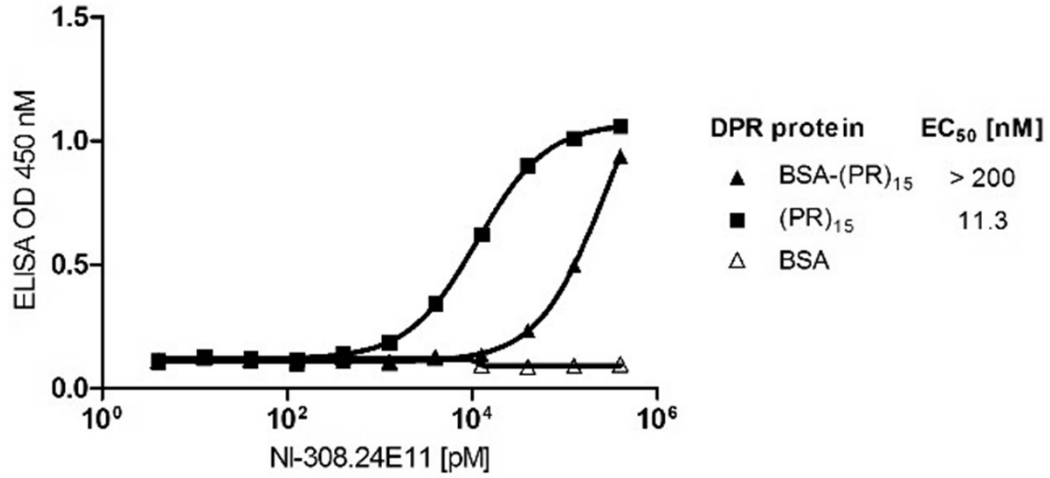
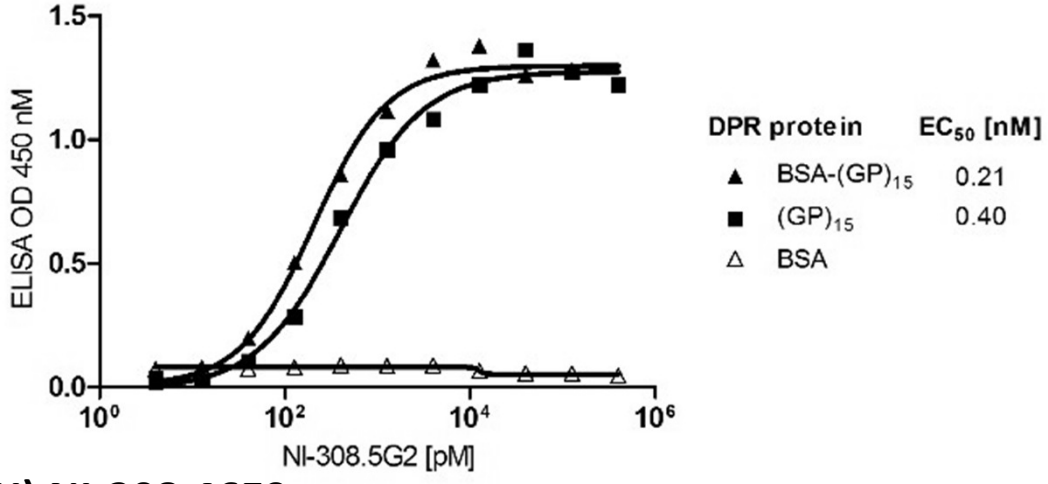
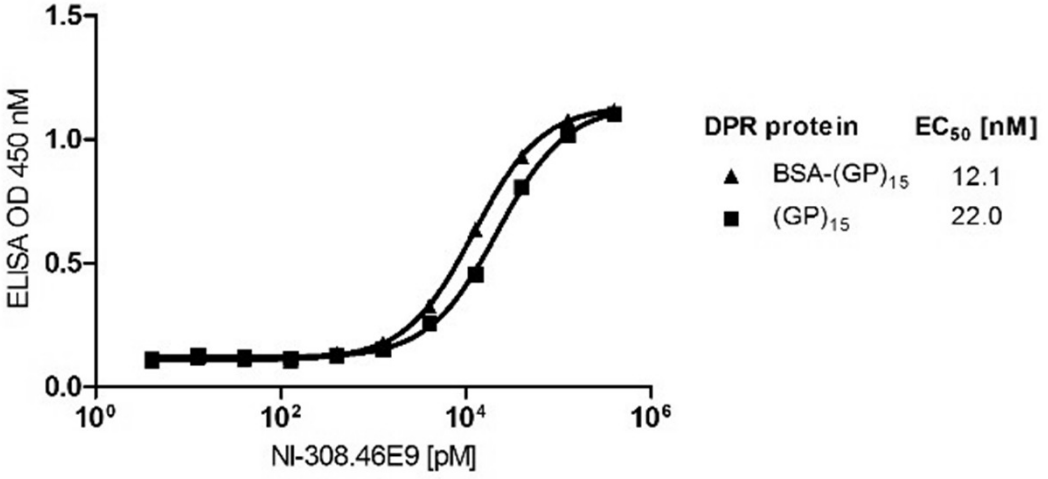


Fig. 3 (continued)

(G) NI-308.5G2



(H) NI-308.46E9



(I) NI-308.6B11

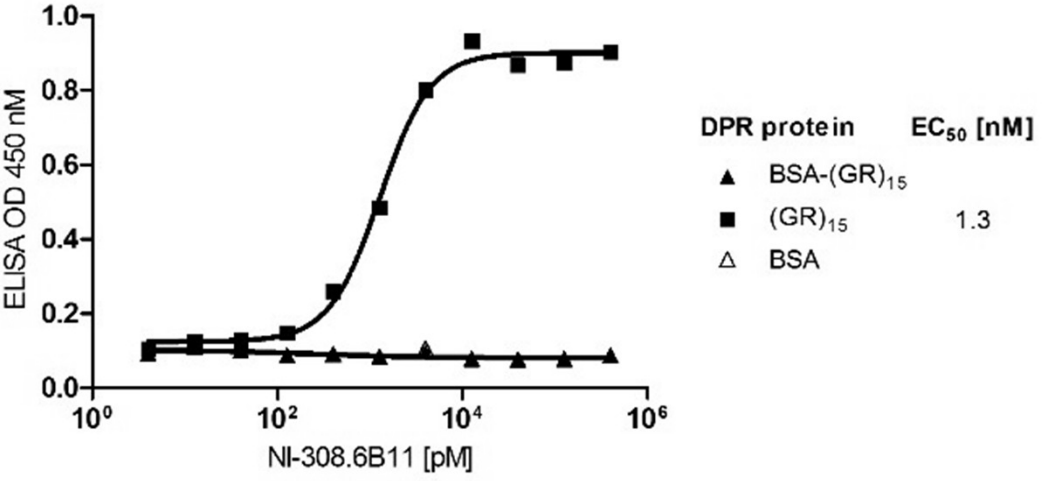
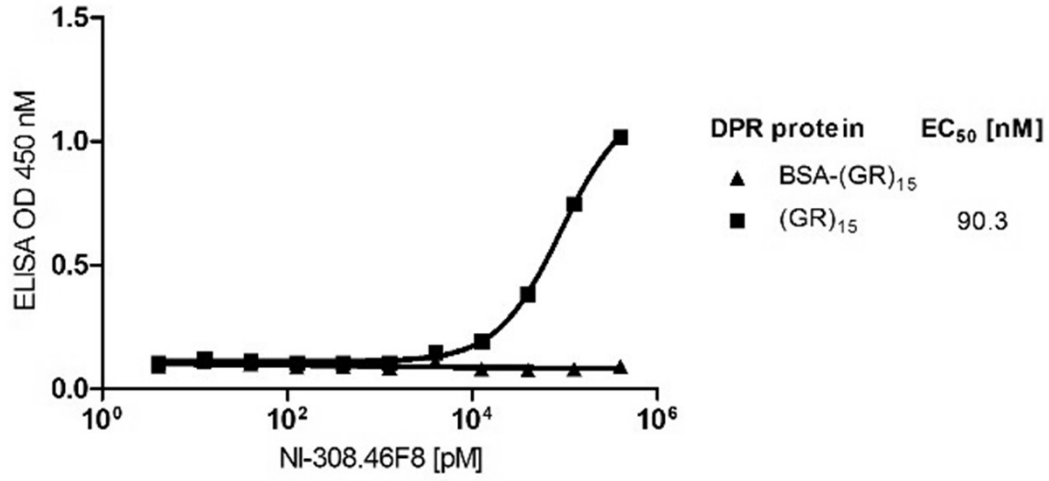
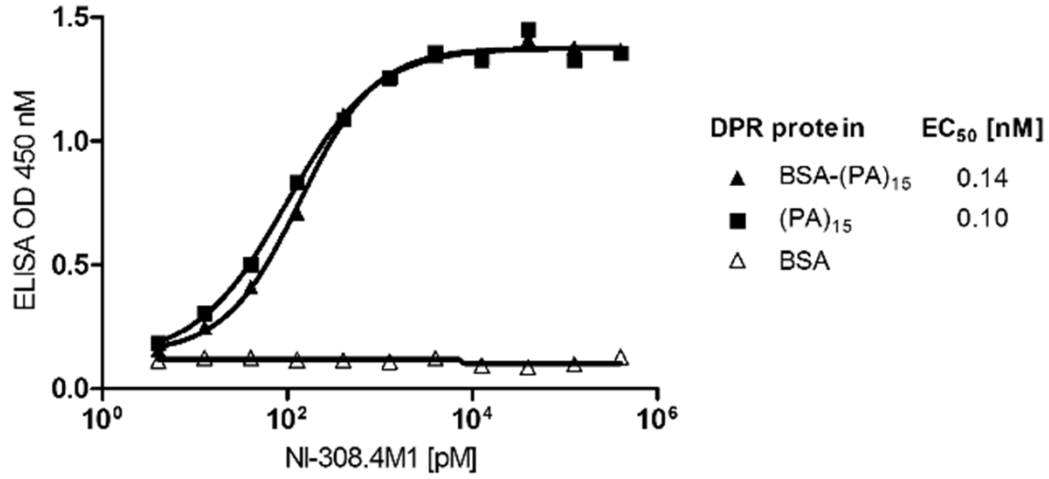


Fig. 3 (continued)

(J) NI-308.46F8



(K) NI-308.4M1



(L) NI-308.12A3

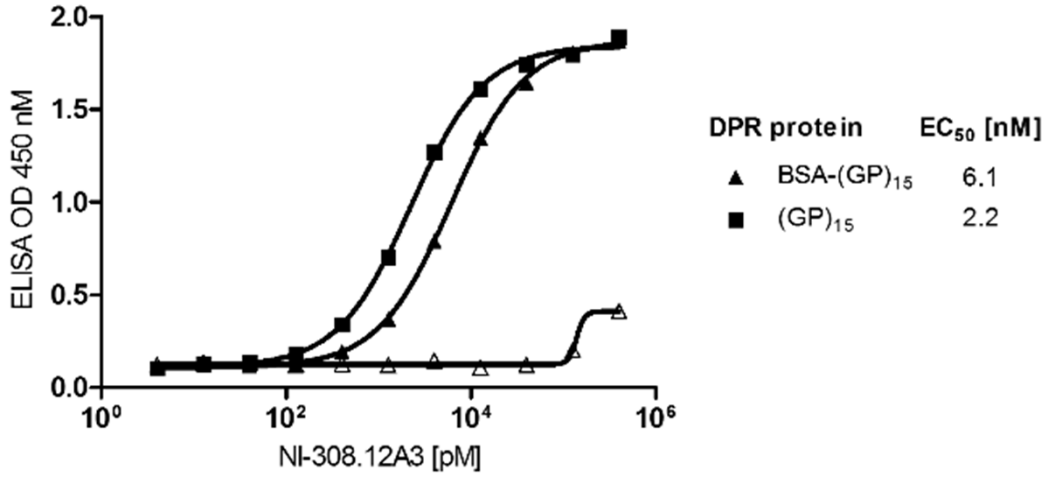


Fig. 3 (continued)

(M) NI-308.16C10

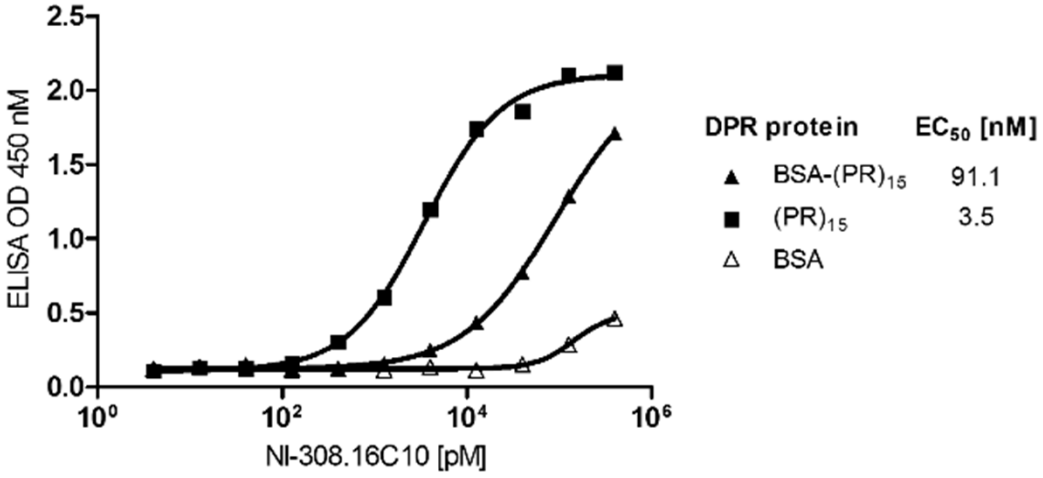


Fig. 3 (continued)

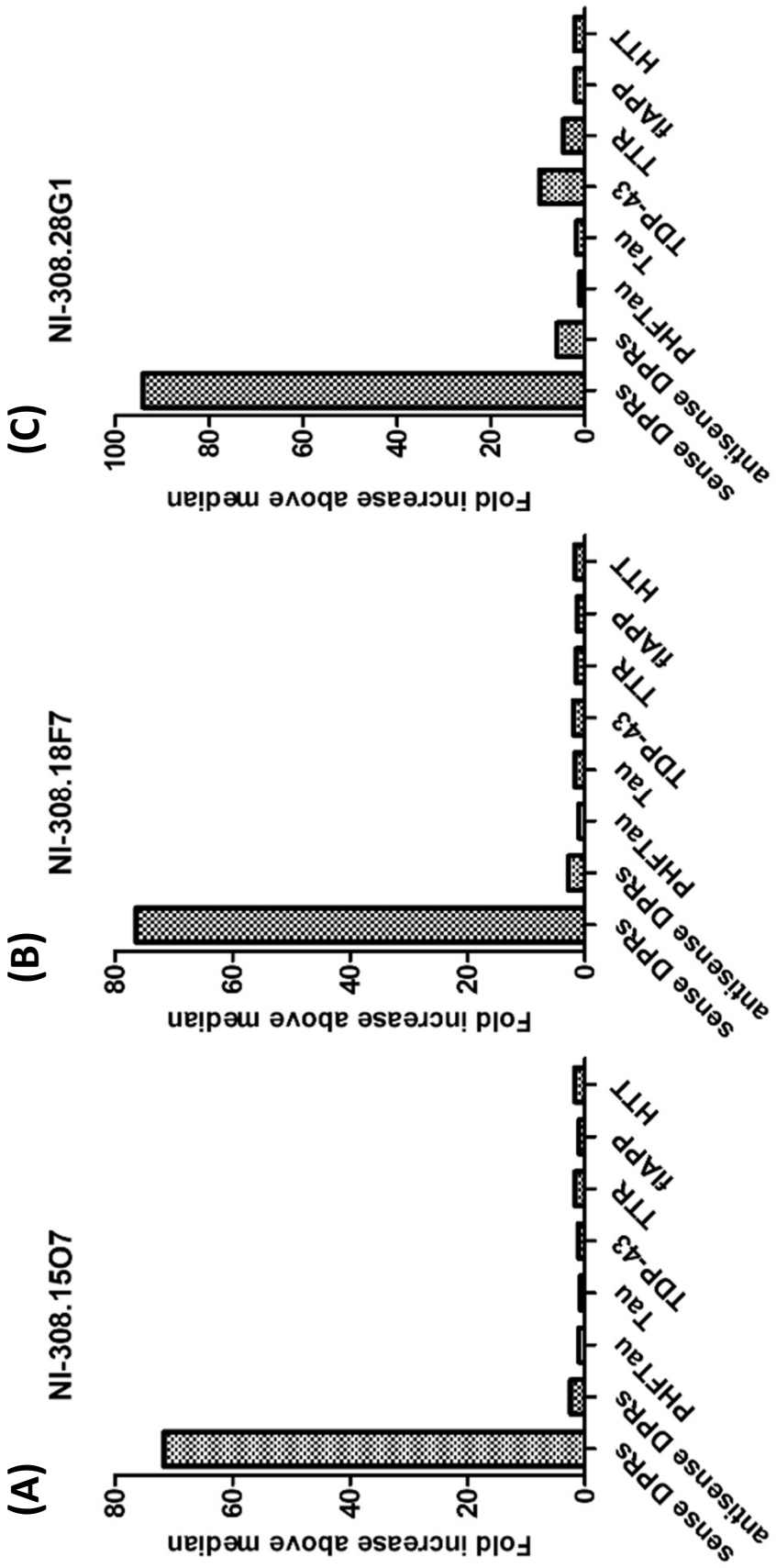

(A)
NI-308.15O7
(B)
NI-308.18F7
(C)
NI-308.28G1
Fold increase above median
0
20
40
60
80
100
sense DPRS
antisense DPRS
PHFTau
Tau
TDP-43
TTR
fIAPP
HTT

Fig. 4

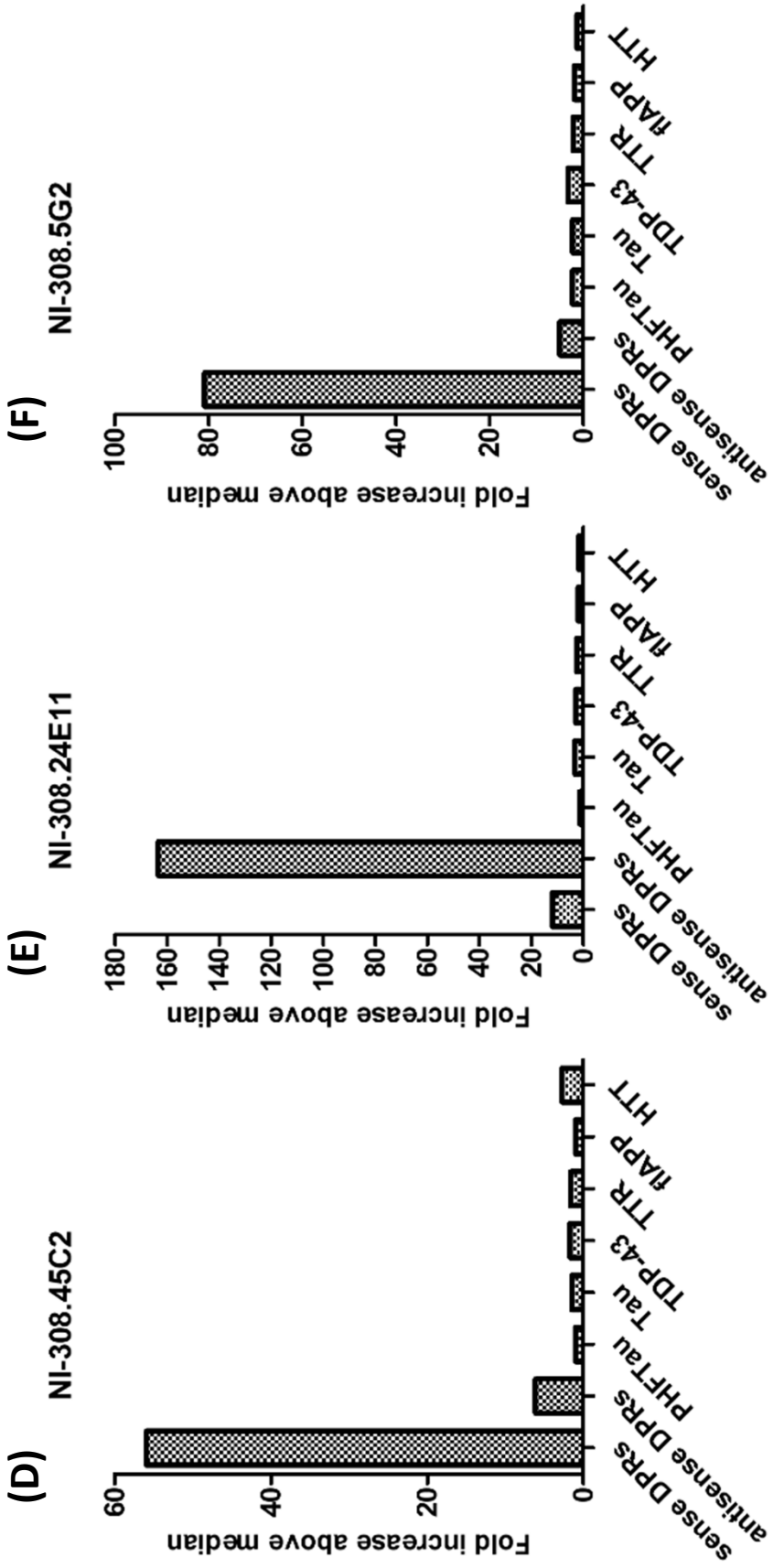


Fig. 4 (continued)

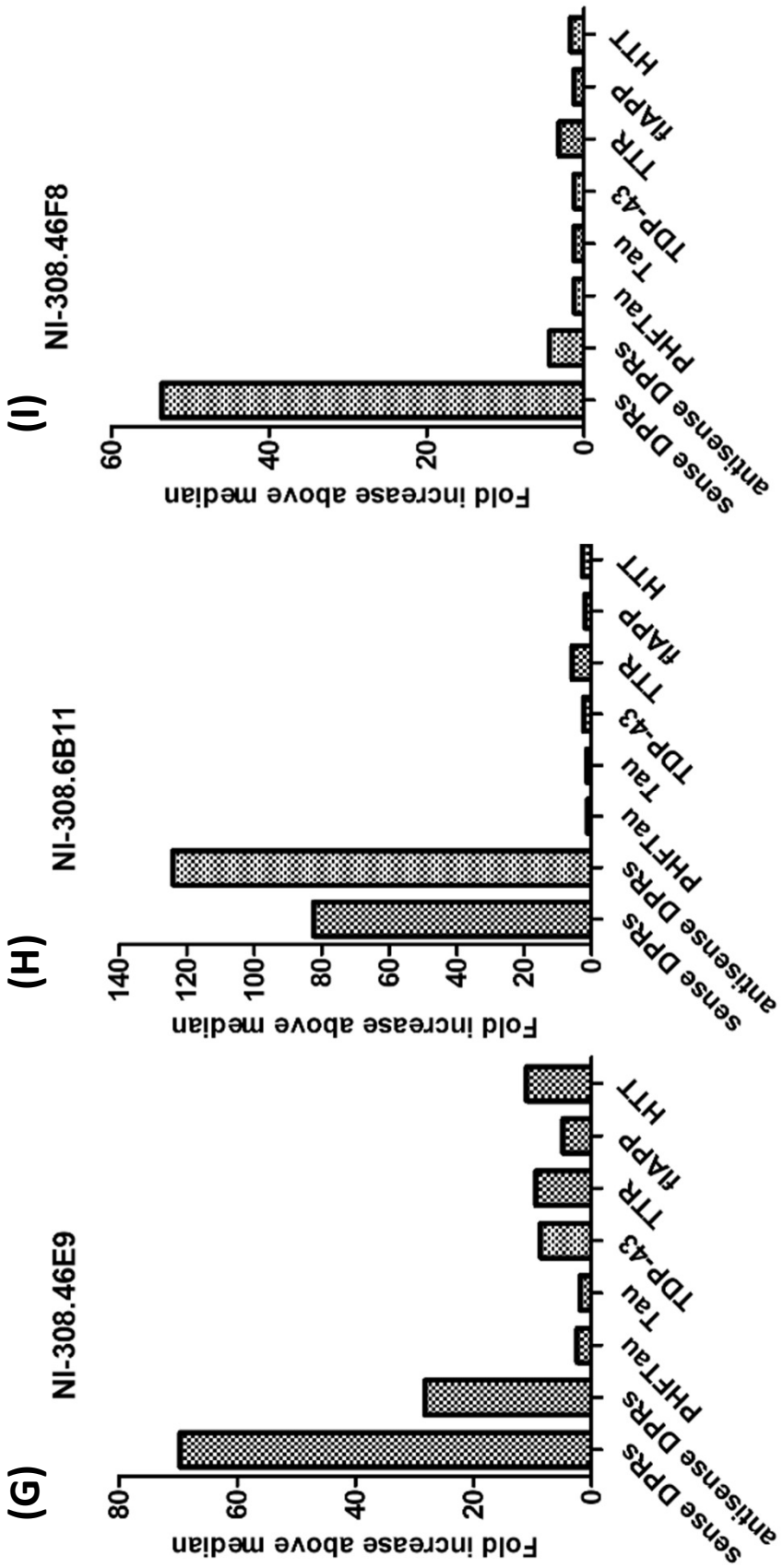


Fig. 4 (continued)

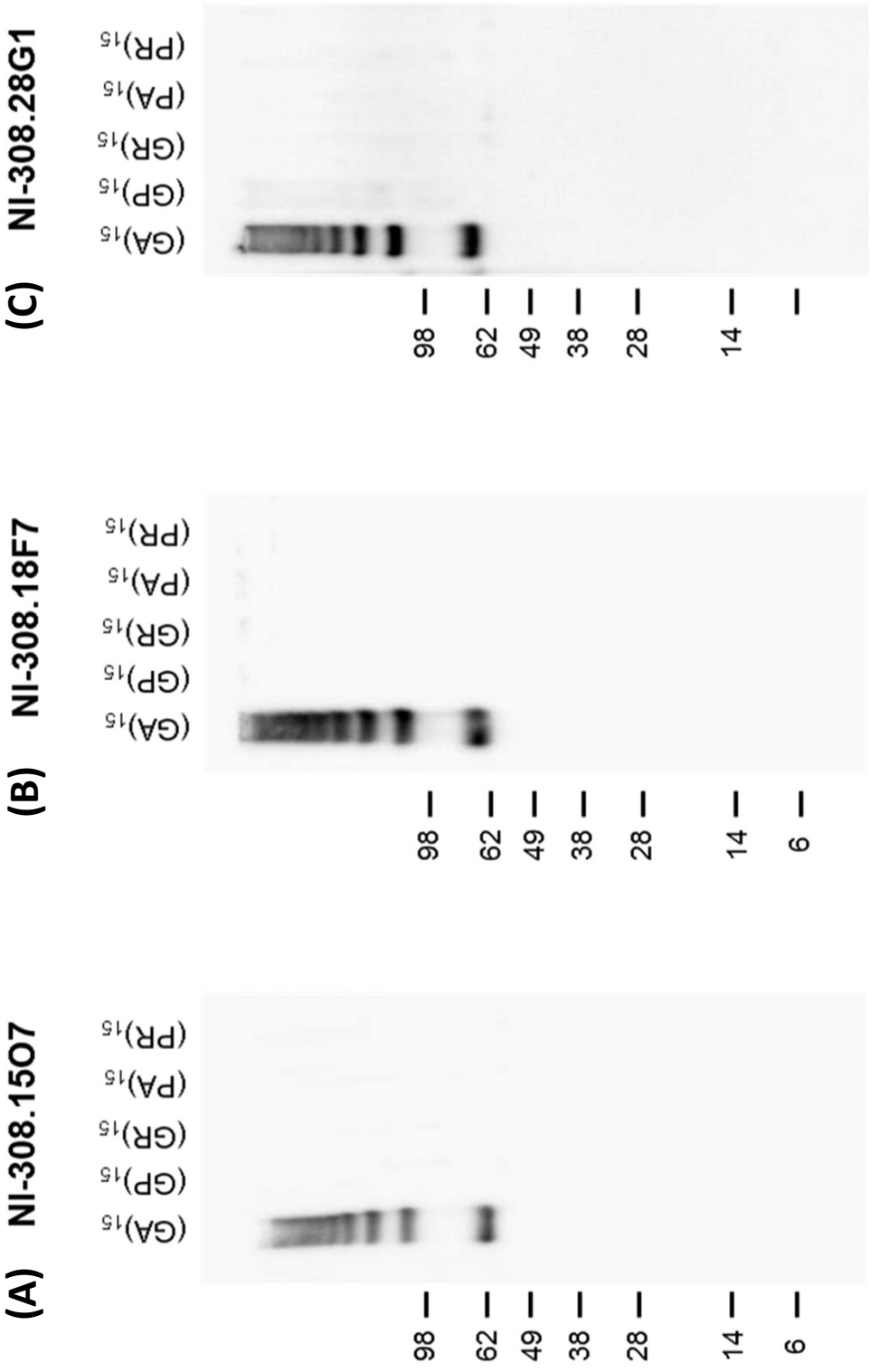

(A) NI-308.15O7
(B) NI-308.18F7
(C) NI-308.28G1
(GA)15
(GP)15
(GR)15
(PA)15
(PR)15
98
62
49
38
28
14
6

Fig. 5

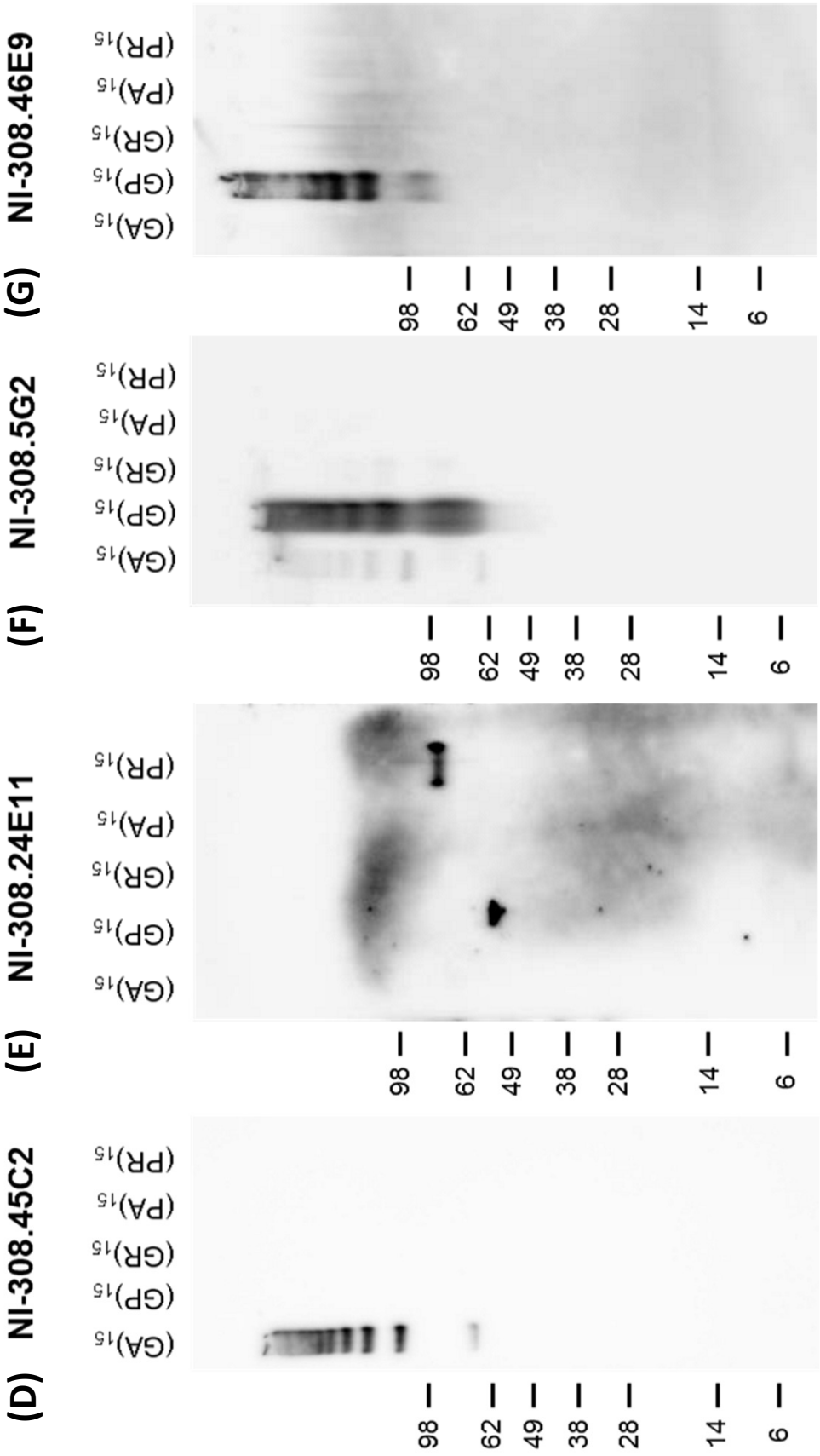


Fig. 5 (continued)

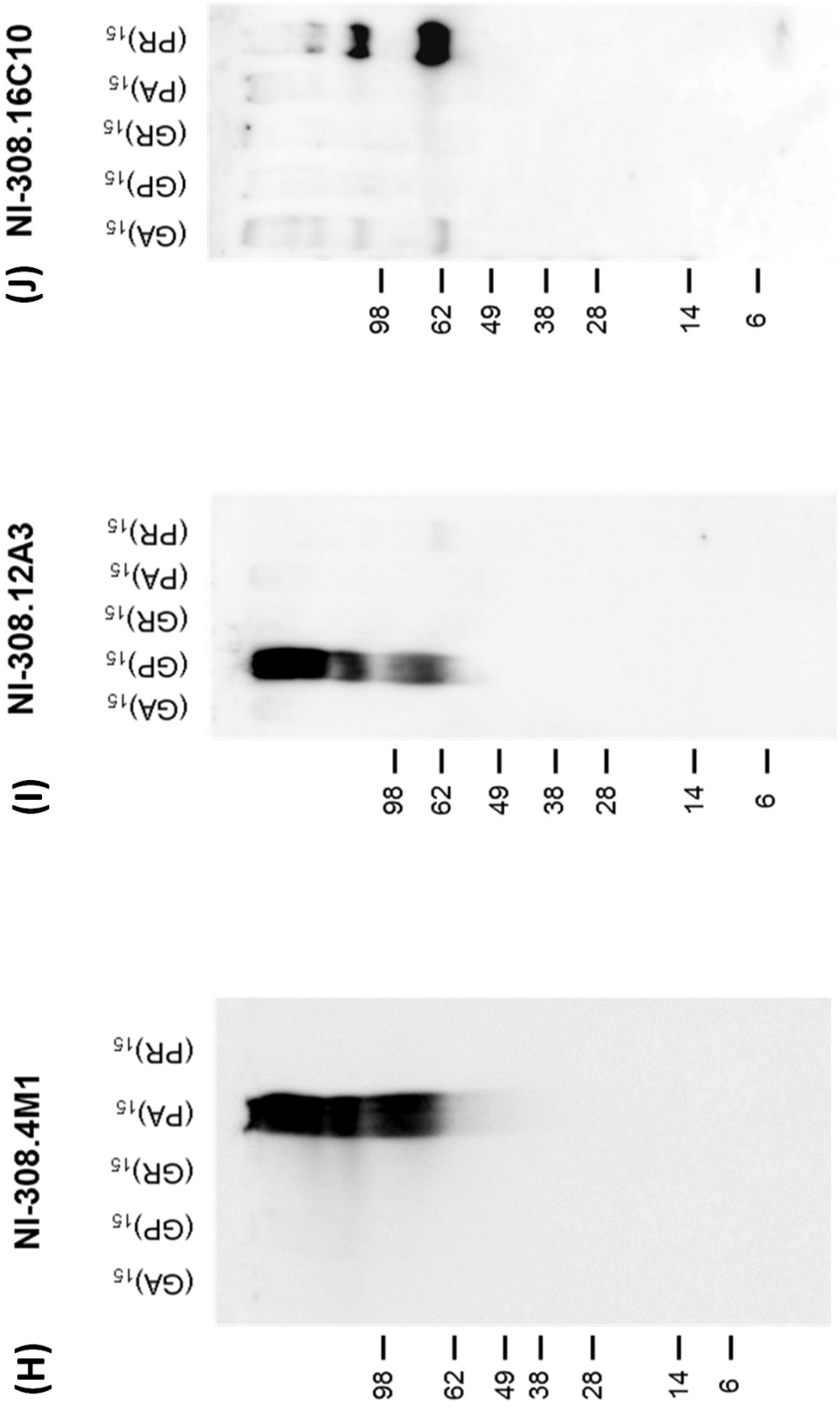


Fig. 5 (continued)

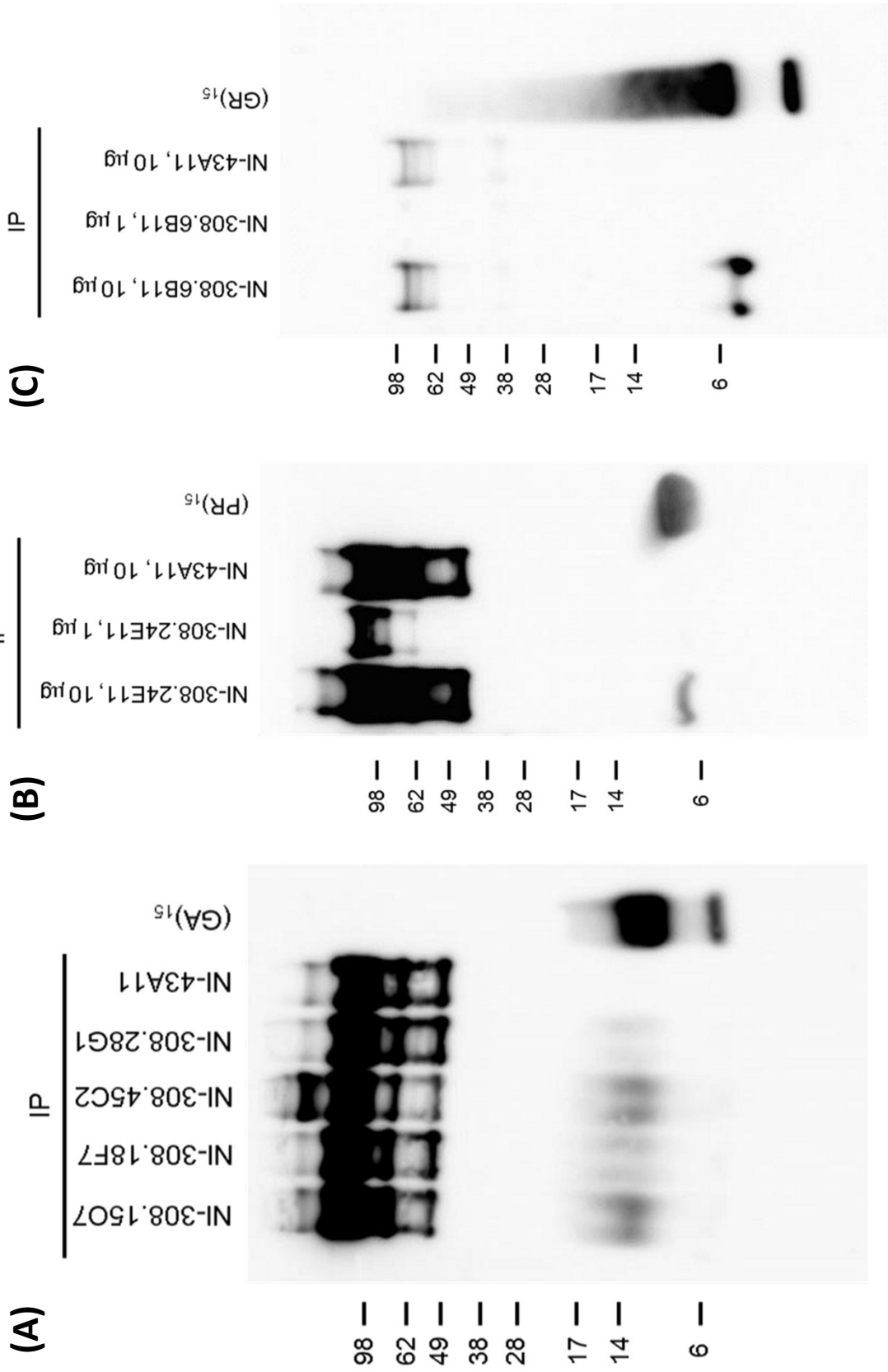

(A)
IP
NI-308.15O7
NI-308.18F7
NI-308.45C2
NI-308.28G1
NI-43A11
(GA)15
98
62
49
38
28
17
14
6
(B)
IP
NI-308.24E11, 10 µg
NI-308.24E11, 1 µg
NI-43A11, 10 µg
(PR)15
98
62
49
38
28
17
14
6
(C)
IP
NI-308.6B11, 10 µg
NI-308.6B11, 1 µg
NI-43A11, 10 µg
(GR)15
98
62
49
38
28
17
14
6

Fig. 6

(A)

	EC_{50} [nM]						
Antibody	$(GA)_2$	$(GA)_3$	$(GA)_4$	$(GA)_5$	$(GA)_6$	$(GA)_{10}$	$(GA)_{20}$
NI-308.15O7	-	-	-	>150	7.9	0.37	0.54
NI-308.18F7	-	-	-	-	>200	0.78	0.83
NI-308.28G1	-	-	-	>200	6.3	0.9	1.1
NI-308.45C2	-	-	-	-	>200	7.6	9.6
NI-308.6B11	-	-	-	-	>200	27.7	33.6
NI-308.5G2	-	-	-	-	>200	5.7	18.2

(B)

	EC_{50} [nM]						
Antibody	$(GP)_2$	$(GP)_3$	$(GP)_4$	$(GP)_5$	$(GP)_6$	$(GP)_{10}$	$(GP)_{20}$
NI-308.5G2	-	-	-	>200	26.8	0.94	0.47
NI-308.46E9	-	-	-	-	-	63.3	15.5

(C)

	EC_{50} [nM]						
Antibody	$(GR)_2$	$(GR)_3$	$(GR)_4$	$(GR)_5$	$(GR)_6$	$(GR)_{10}$	$(GR)_{20}$
NI-308.6B11	-	14.3	13.2	0.47	0.33	1.3	0.25

(D)

	EC_{50} [nM]						
Antibody	$(PR)_2$	$(PR)_3$	$(PR)_4$	$(PR)_5$	$(PR)_6$	$(PR)_{10}$	$(PR)_{20}$
NI-308.24E11	-	-	-	-	>200	31.7	11.3

(E)

	EC_{50} [nM]						
Antibody	$(PA)_2$	$(PA)_3$	$(PA)_4$	$(PA)_5$	$(PA)_6$	$(PA)_{10}$	$(PA)_{20}$
NI-308.4M1	-	-	-	26.9	4.3	0.06	0.06

(F) NI-308.15O7

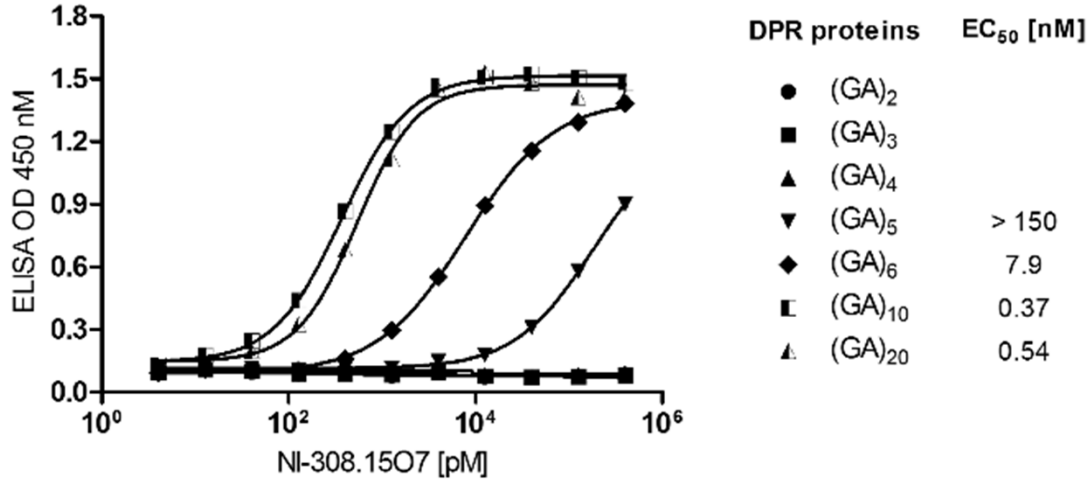
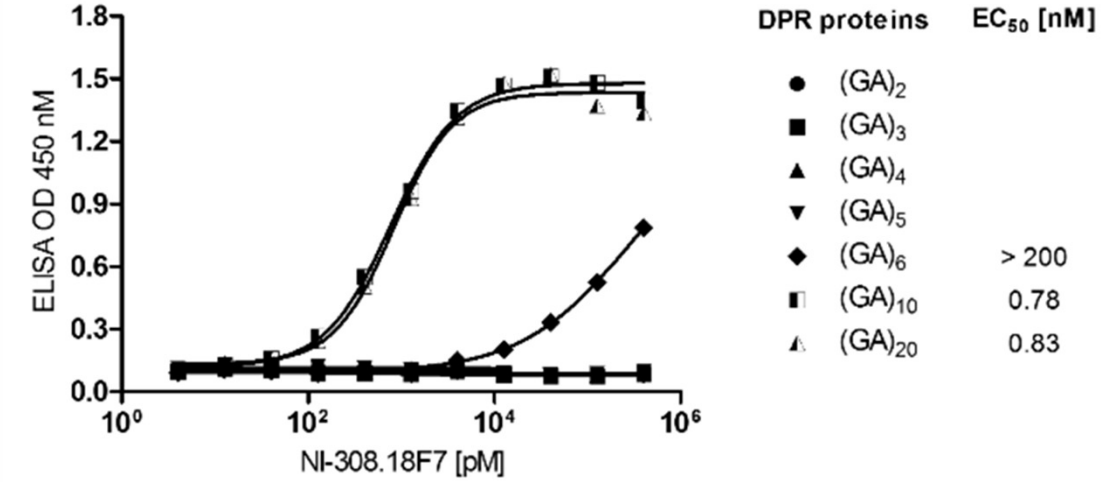
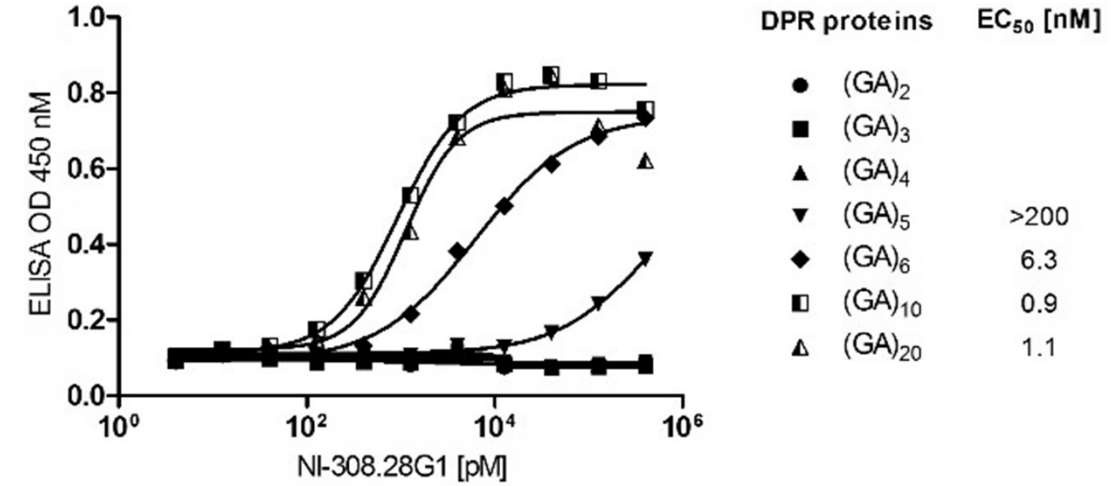


Fig. 7

(G) NI-308.18F7



(H) NI-308.28G1



(I) NI-308.45C2

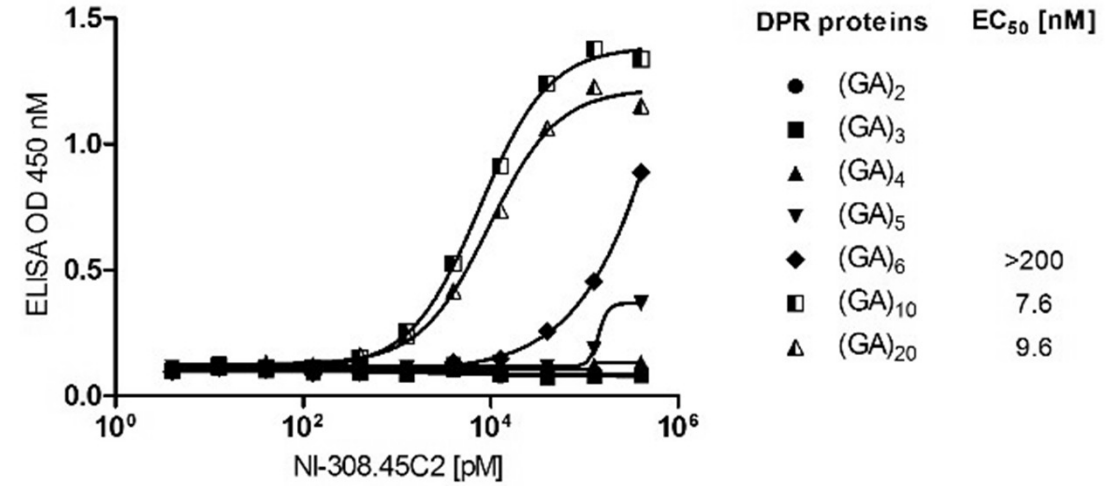
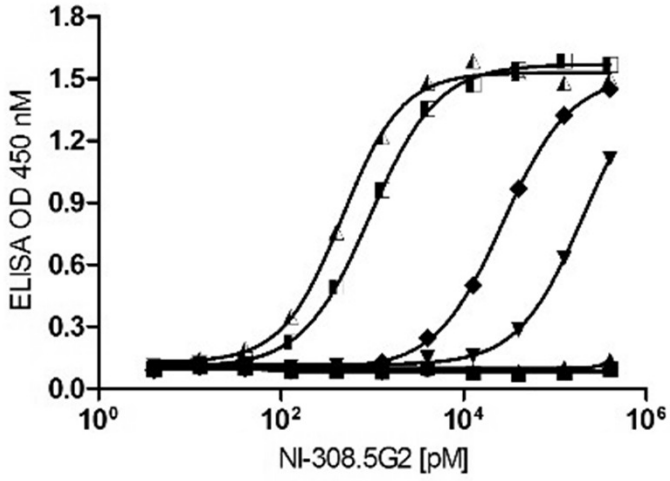


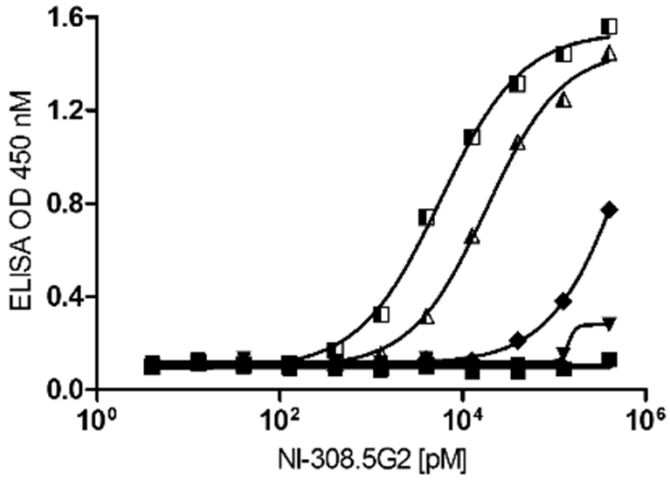
Fig. 7 (continued)

(J) NI-308.5G2



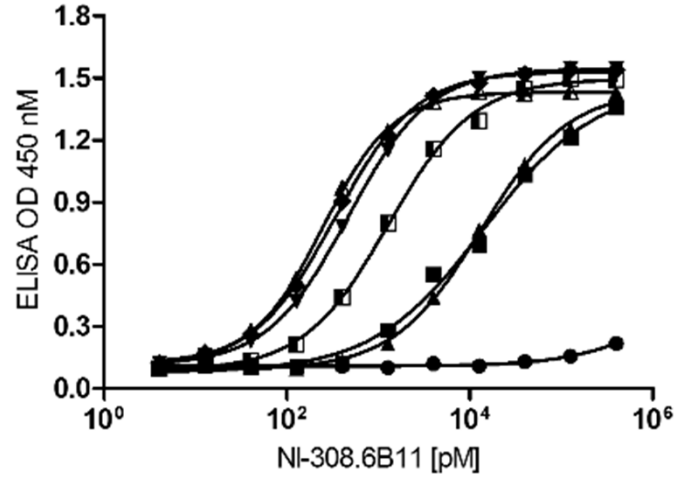
DPR proteins	EC_{50} [nM]
$(GP)_2$	
$(GP)_3$	
$(GP)_4$	
$(GP)_5$	> 200
$(GP)_6$	26.8
$(GP)_{10}$	0.94
$(GP)_{20}$	0.47

(K) NI-308.5G2



DPR proteins	EC_{50} [nM]
$(GA)_2$	
$(GA)_3$	
$(GA)_4$	
$(GA)_5$	
$(GA)_6$	>200
$(GA)_{10}$	5.7
$(GA)_{20}$	18.2

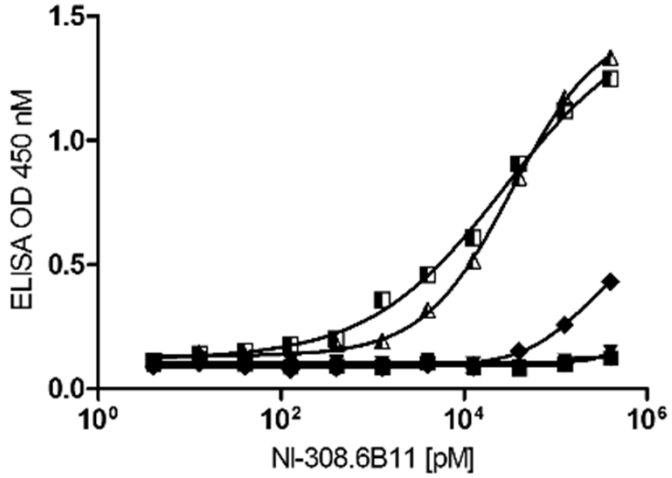
(L) NI-308.6B11



DPR proteins	EC_{50} [nM]
$(GR)_2$	
$(GR)_3$	14.3
$(GR)_4$	13.2
$(GR)_5$	0.47
$(GR)_6$	0.33
$(GR)_{10}$	1.28
$(GR)_{20}$	0.25

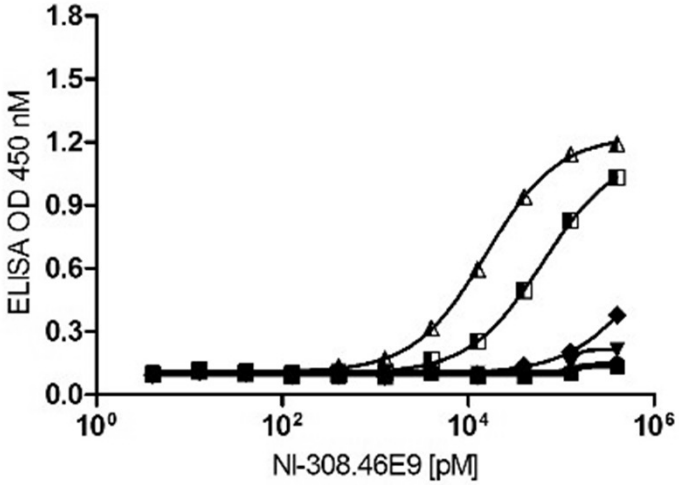
Fig. 7 (continued)

(M) NI-308.6B11



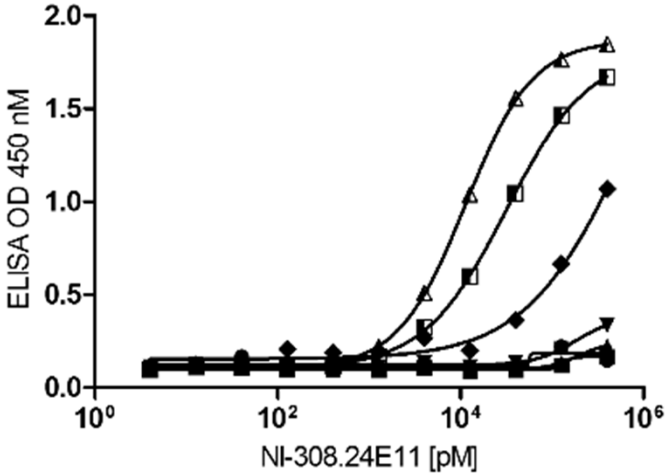
DPR proteins	EC_{50} [nM]
$(GA)_2$	
$(GA)_3$	
$(GA)_4$	
$(GA)_5$	
$(GA)_6$	>200
$(GA)_{10}$	27.7
$(GA)_{20}$	33.6

(N) NI-308.46E9



DPR proteins	EC_{50} [nM]
$(GP)_2$	
$(GP)_3$	
$(GP)_4$	
$(GP)_5$	
$(GP)_6$	>200
$(GP)_{10}$	63.3
$(GP)_{20}$	15.5

(O) NI-308.24E11



DPR proteins	EC_{50} [nM]
$(PR)_2$	
$(PR)_3$	
$(PR)_4$	
$(PR)_5$	
$(PR)_6$	>200
$(PR)_{10}$	31.7
$(PR)_{20}$	11.3

Fig. 7 (continued)

(P) NI-308.4M1

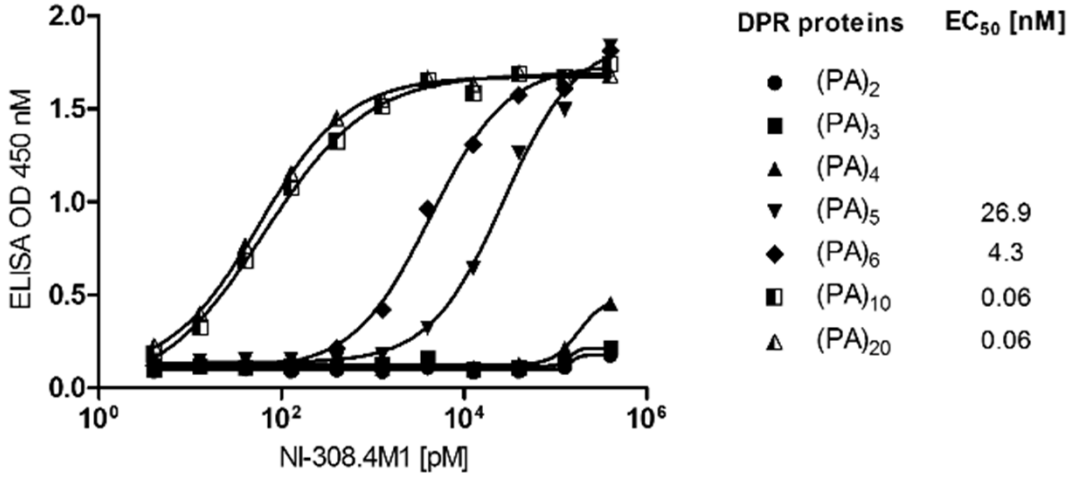


Fig. 7 (continued)

(A)

	EC50 [nM]						
Antibody	**$(GA)_2$**	**$(GA)_3$**	**$(GA)_4$**	**$(GA)_5$**	**$(GA)_6$**	**$(GA)_{10}$**	**$(GA)_{20}$**
NI-308.15O7	-	-	-	-	-	>400	9.0
NI-308.18F7	-	-	-	-	-	0.34	0.45
NI-308.28G1	-	-	-	-	-	0.46	0.40

(B)

	EC50 [nM]						
Antibody	**$(GA)_2$**	**$(GA)_3$**	**$(GA)_4$**	**$(GA)_5$**	**$(GA)_6$**	**$(GA)_{10}$**	**$(GA)_{20}$**
NI-308.5G2	-	-	-	-	-	-	38.2

(C) NI-308.15O7

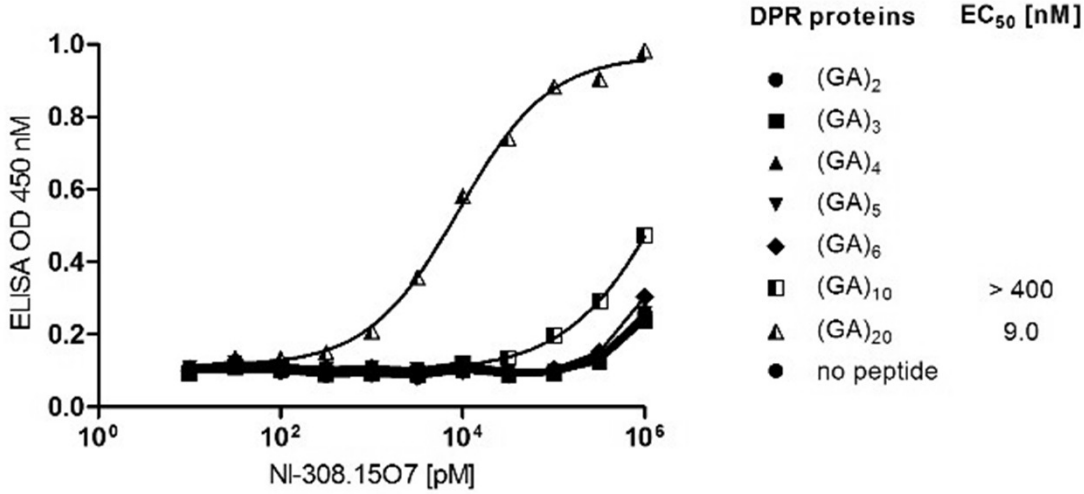
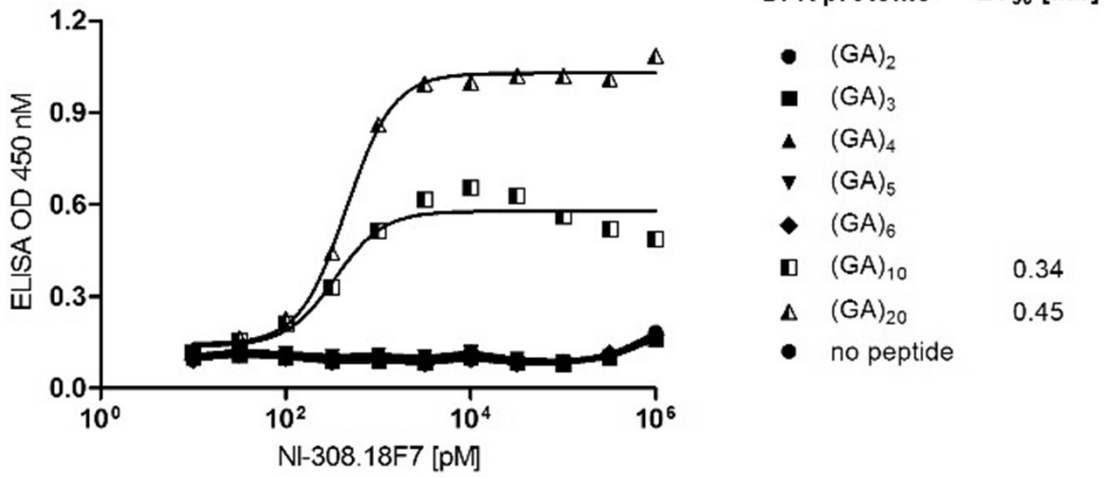
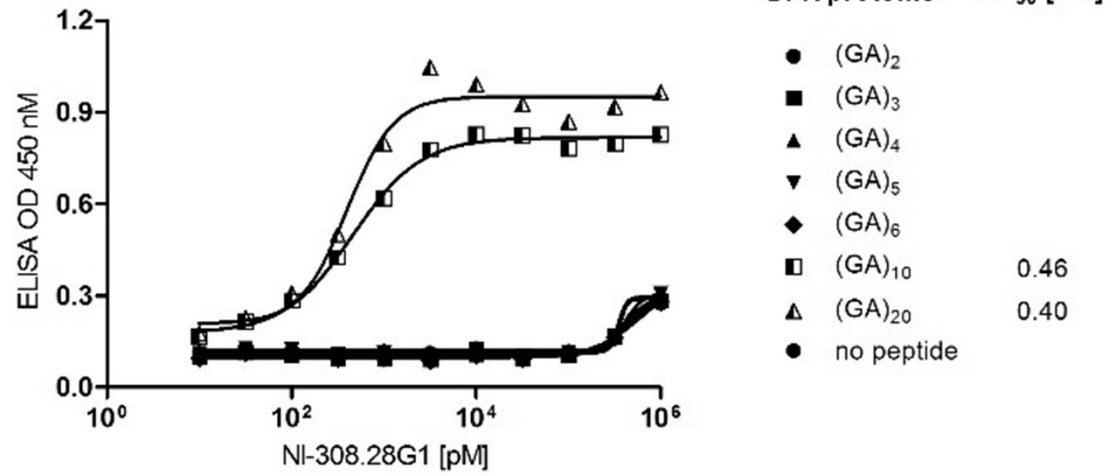


Fig. 8

(D) NI-308.18F7



(E) NI-308.28G1



(F) NI-308.5G2

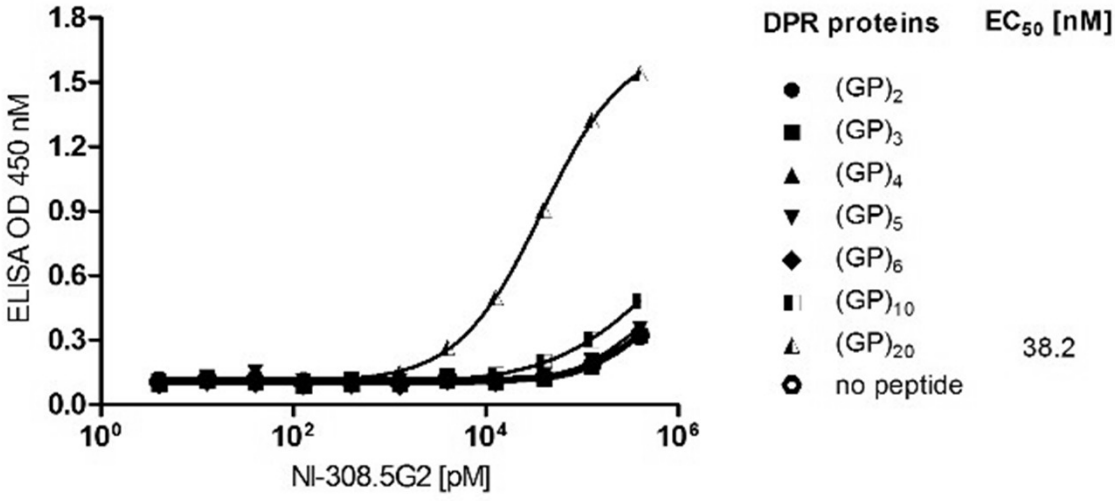


Fig. 8 (continued)

	EC_{50} [nM]			
	indirect ELISA		Sandwich ELISA	
Antibody	monomeric $(GA)_{15}$	aggregated $(GA)_{15}$	monomeric $(GA)_{15}$	aggregated $(GA)_{15}$
NI-308.18F7	0.87	0.89	0.64	2.9
NI-308.15O7	0.53	0.78	6.3	8.1

Fig. 9

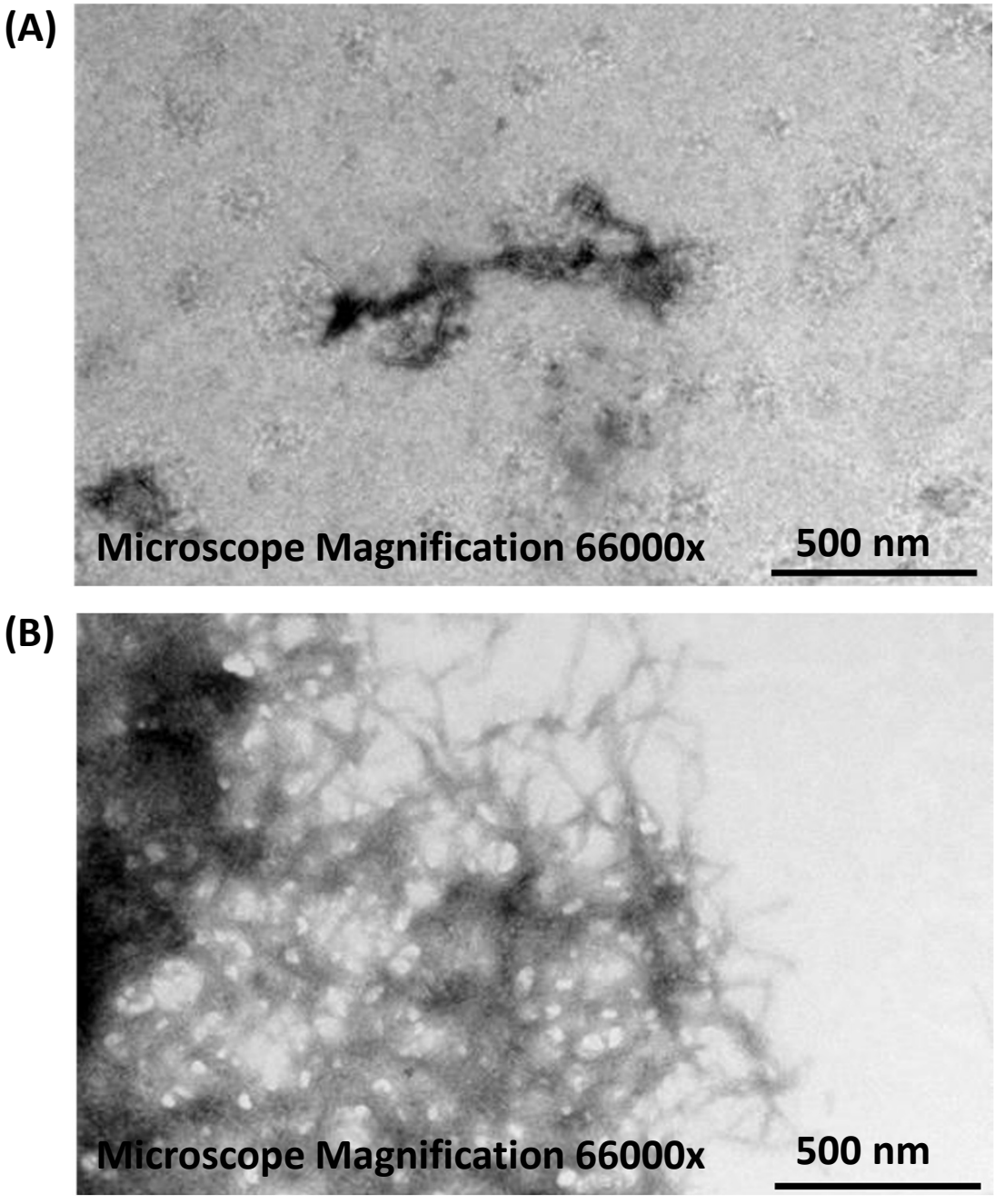
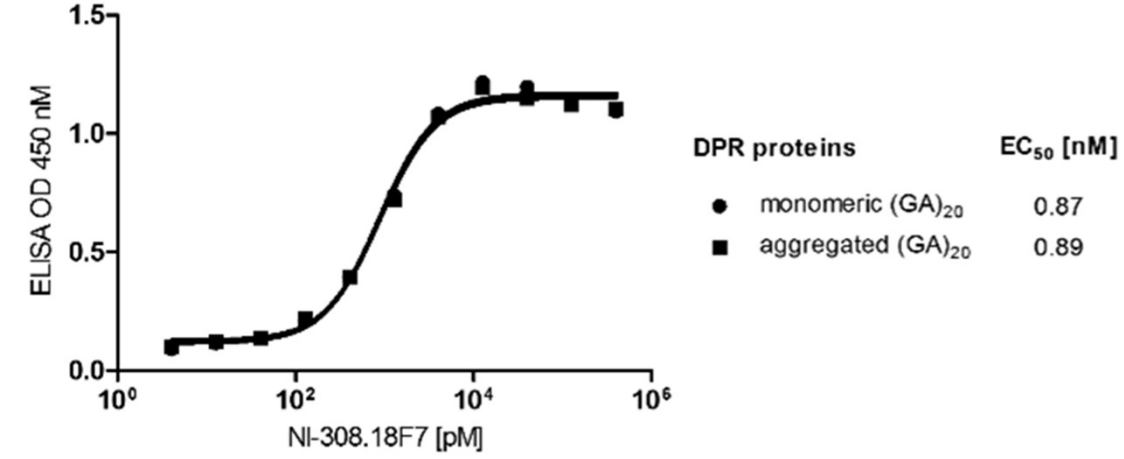


Fig. 10

(A) NI-308.18F7



(B) NI-308.15O7

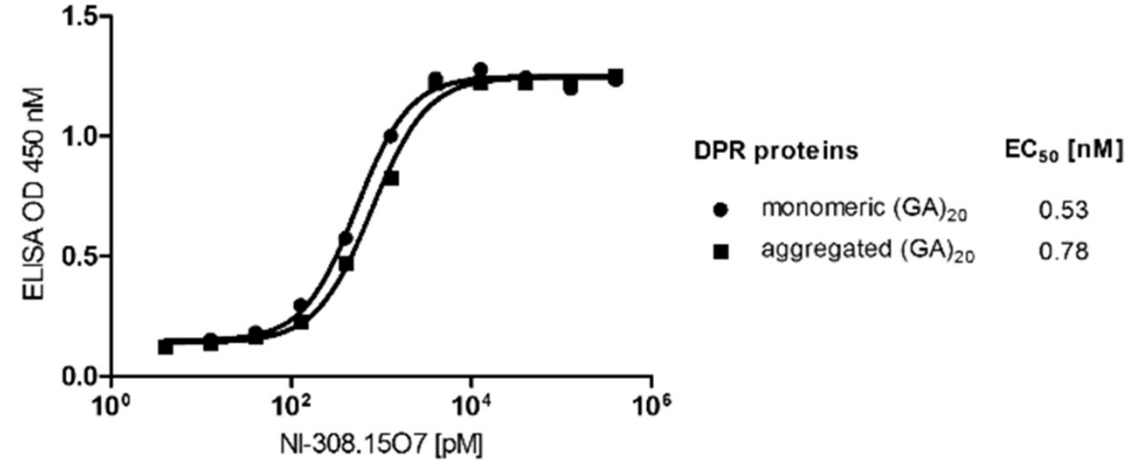


Fig. 11

(A) NI-308.18F7

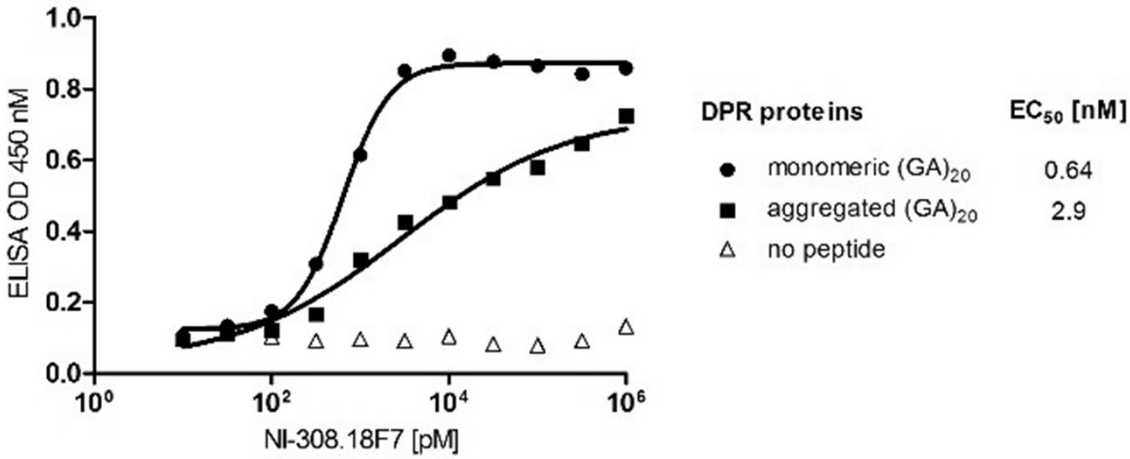


Fig. 12

(B) NI-308.15O7

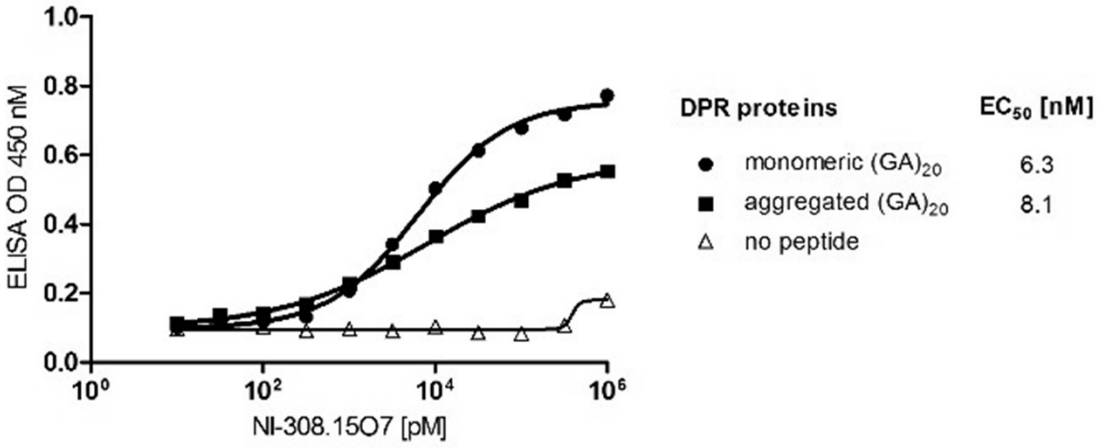


Fig. 12 (continued)

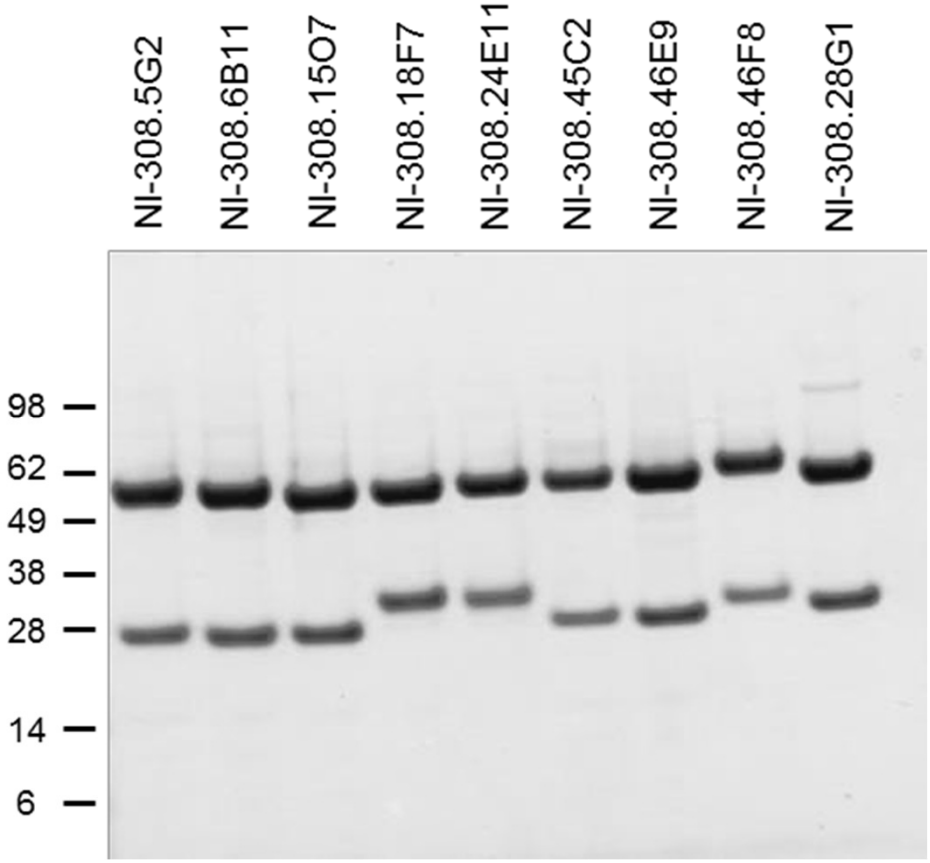


Fig.13

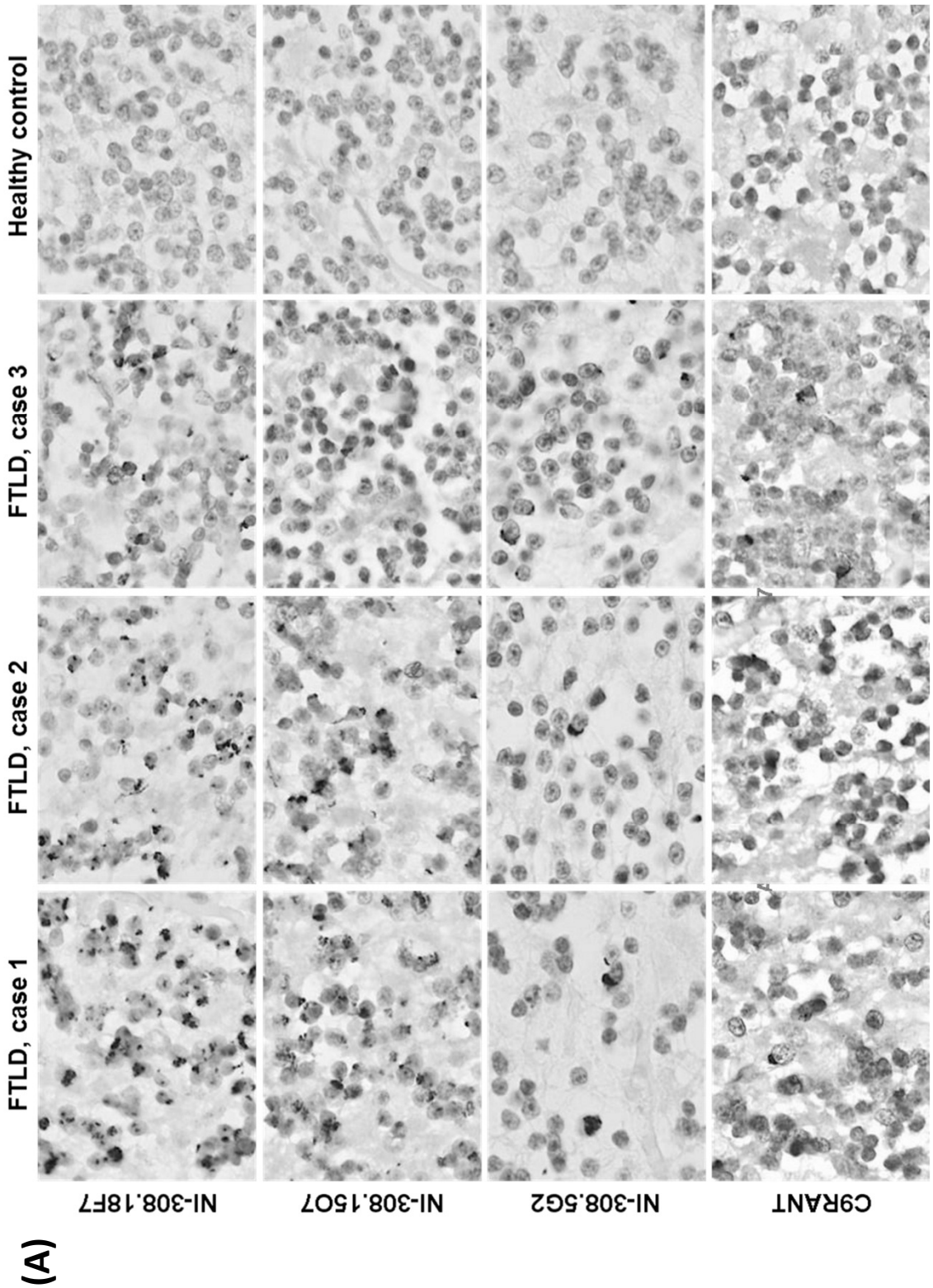

Healthy control
FTLD, case 3
FTLD, case 2
FTLD, case 1
NI-308.18F7
NI-308.15O7
NI-308.5G2
C9RANT
(A)

Fig. 14

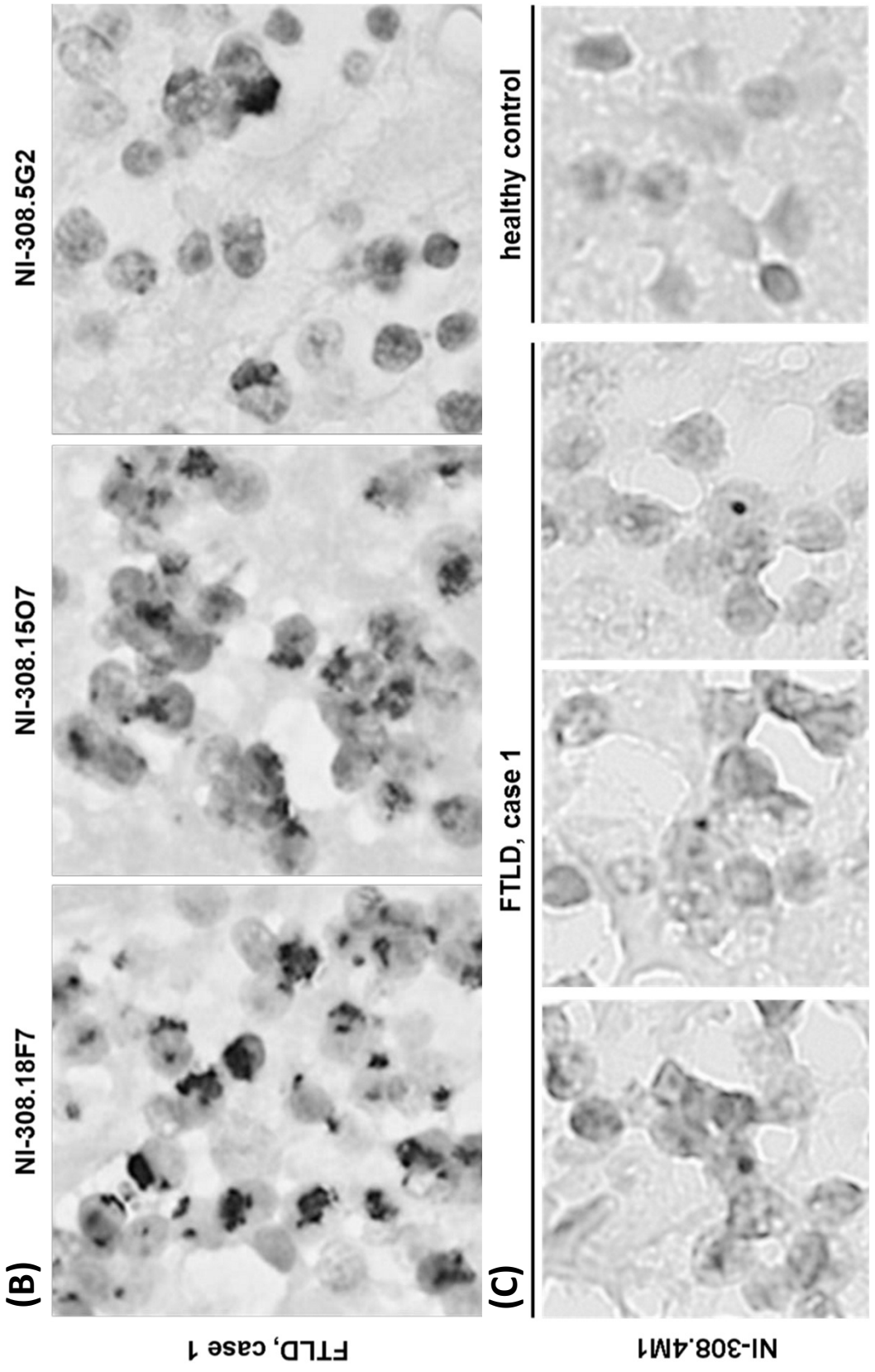


Fig. 14 (continued)

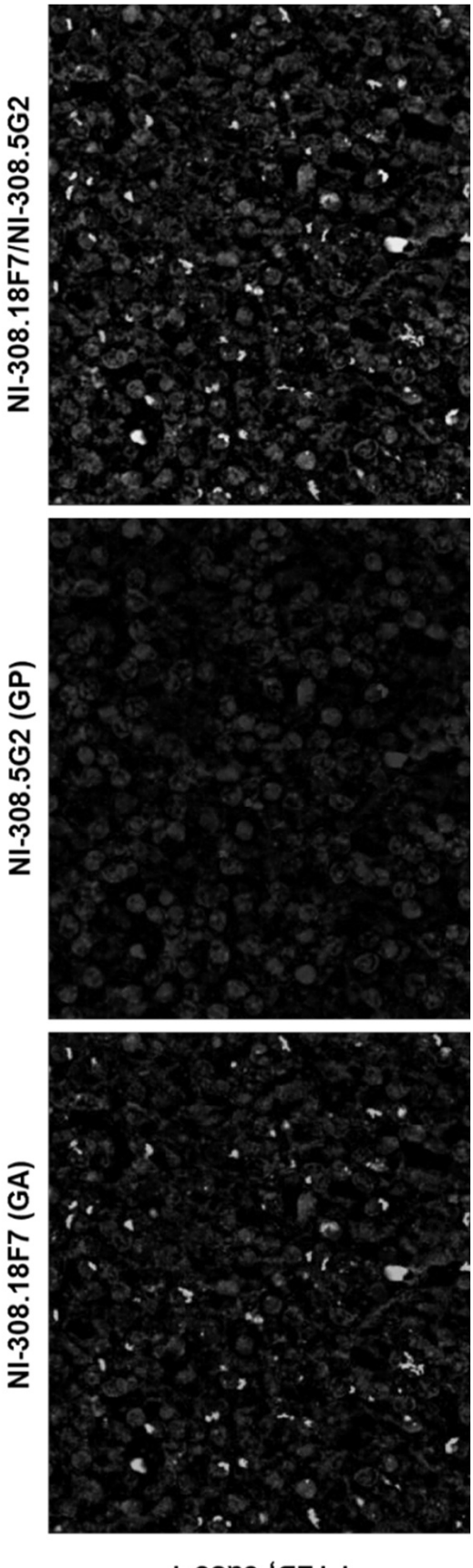


Fig. 15