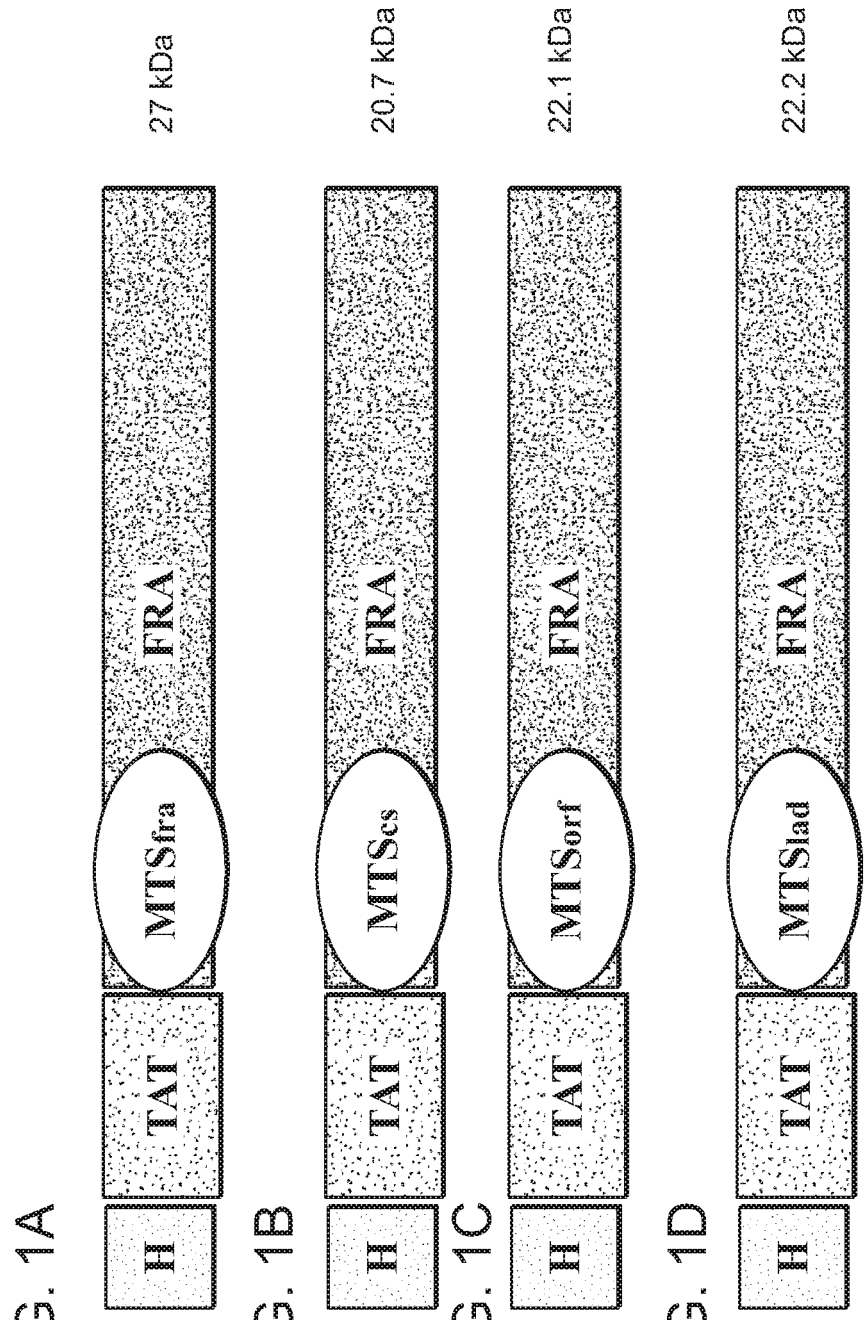


FIG. 1A

FIG. 1B

FIG. 1C

FIG. 1D

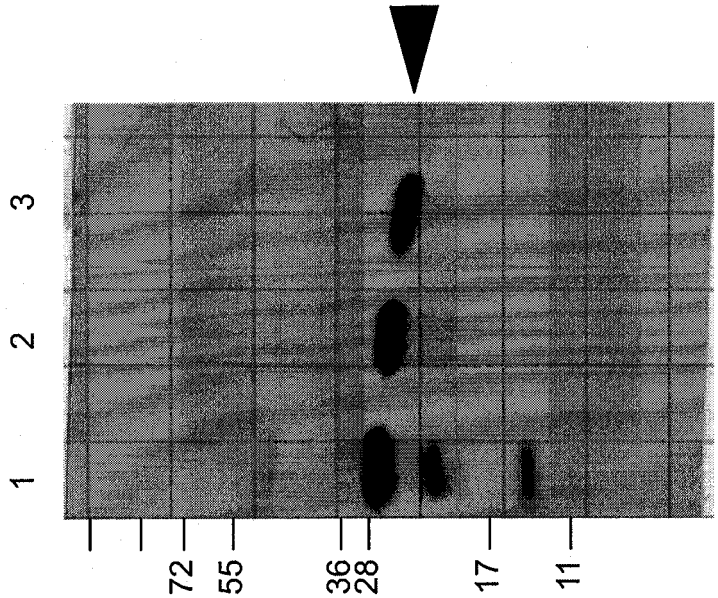


FIG. 2B

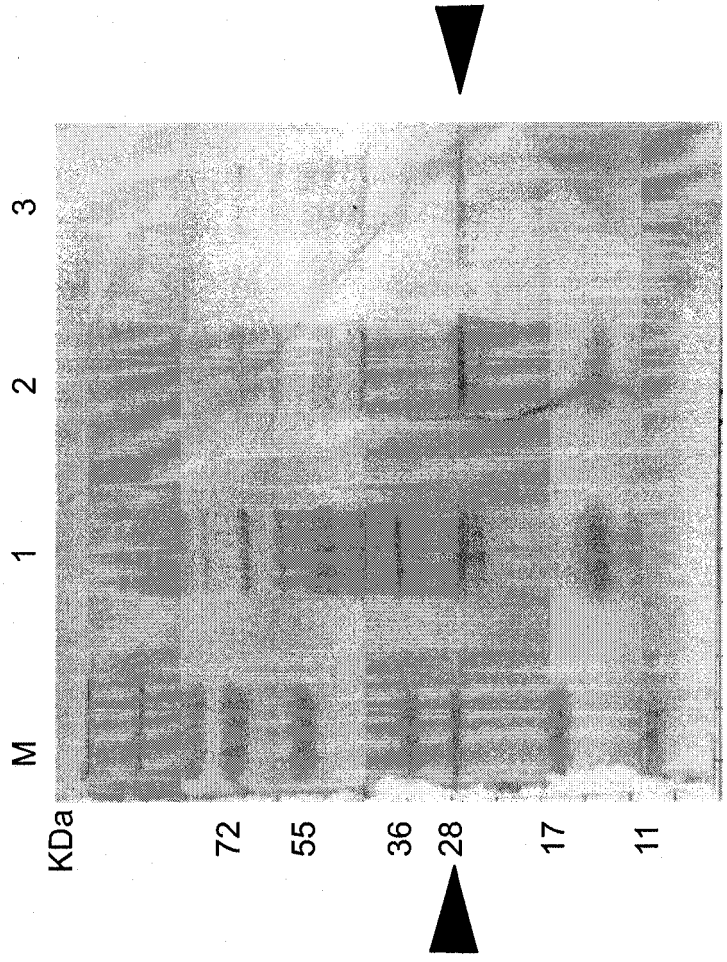


FIG. 2A

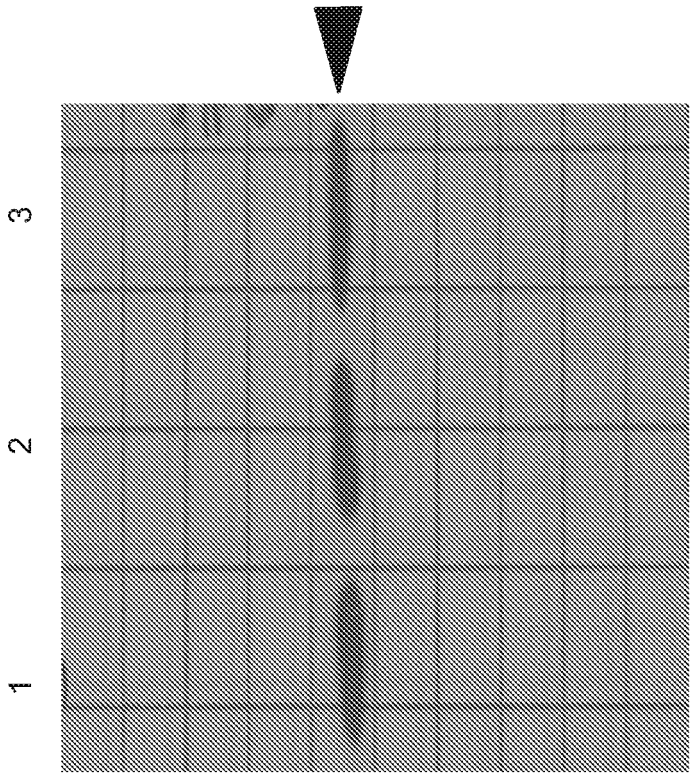


FIG. 2D

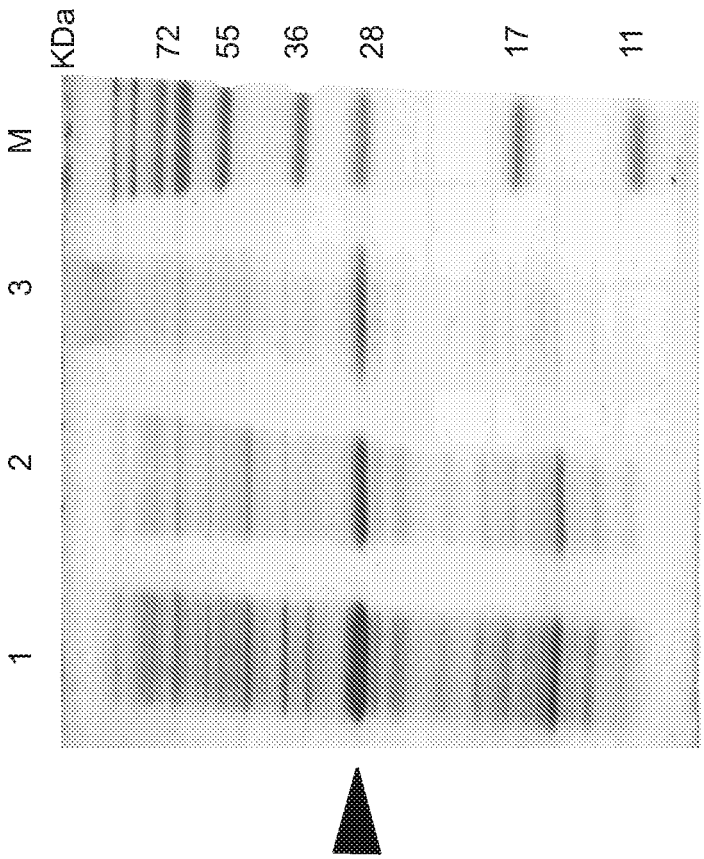


FIG. 2C

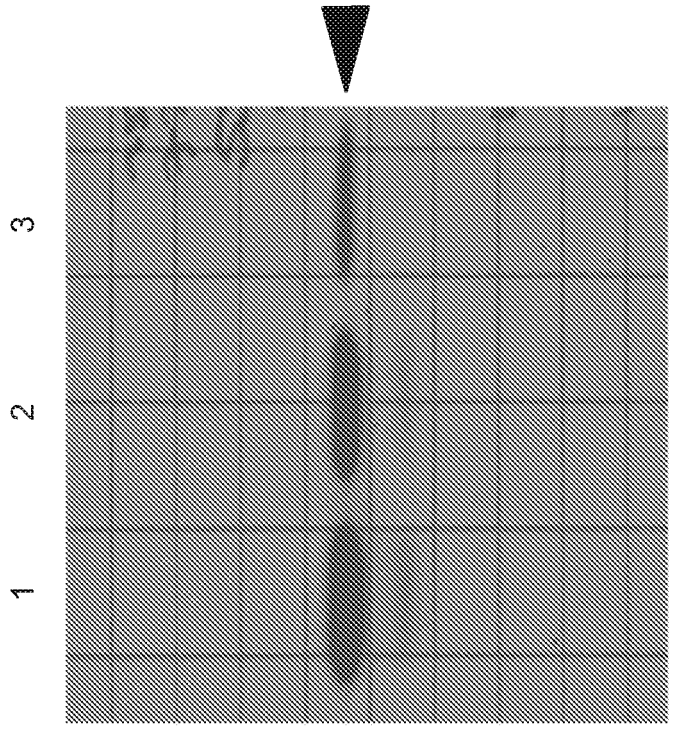


FIG. 3B

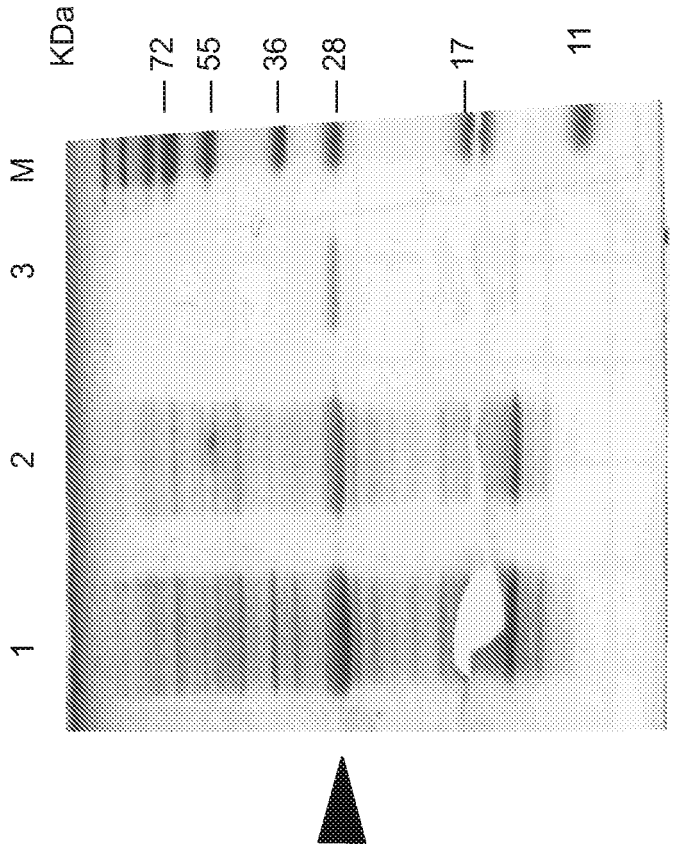


FIG. 3A

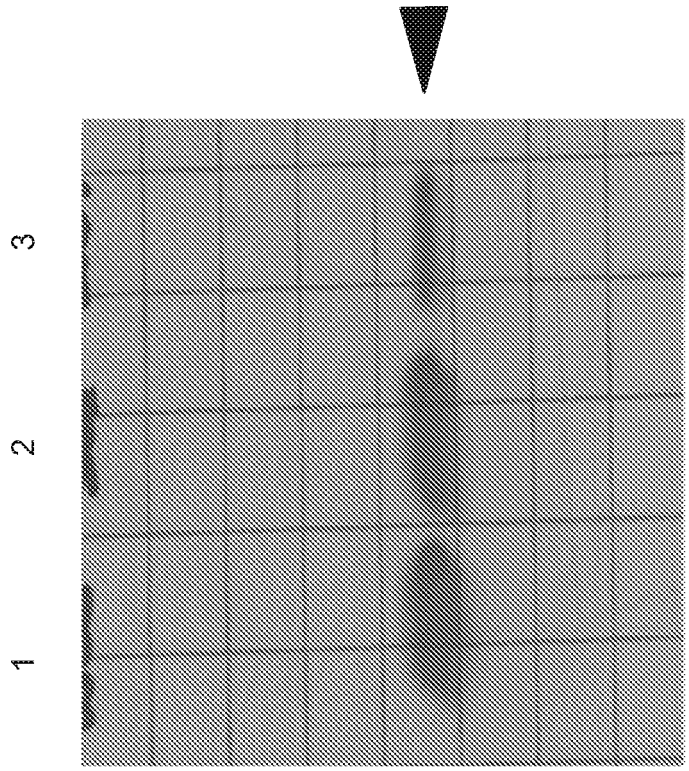


FIG. 3D

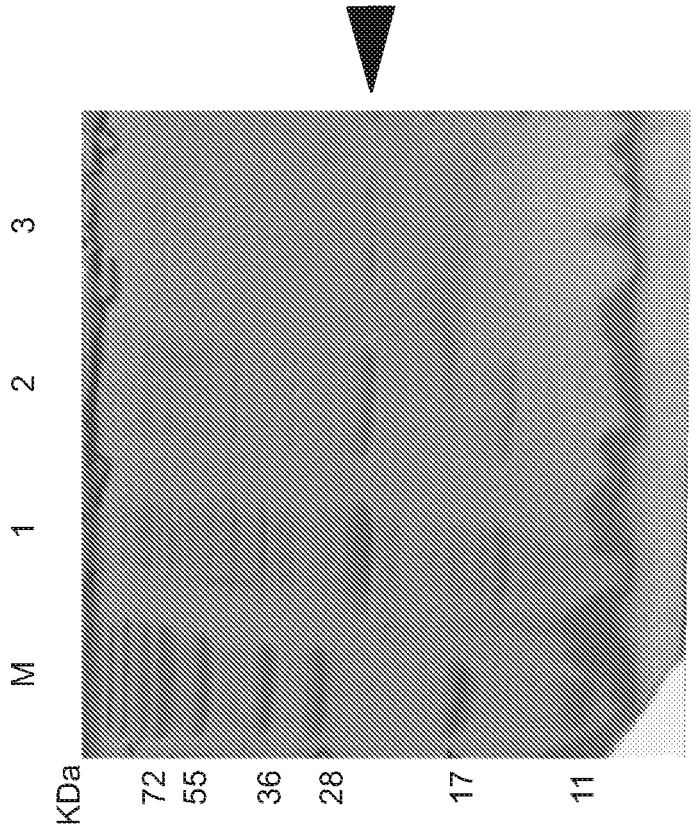


FIG. 3C

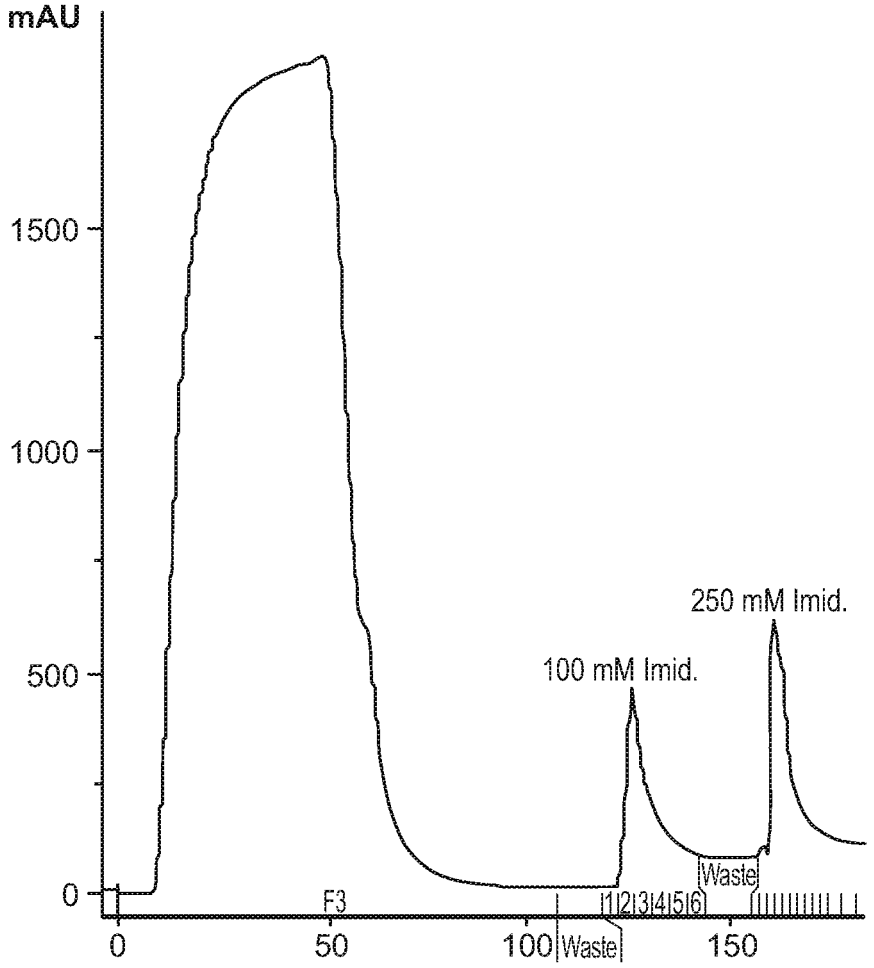

mAU
1500
1000
500
0
250 mM Imid.
100 mM Imid.
Waste
F3
1|2|3|4|5|6
0
50
100
Waste
150

FIG. 4A

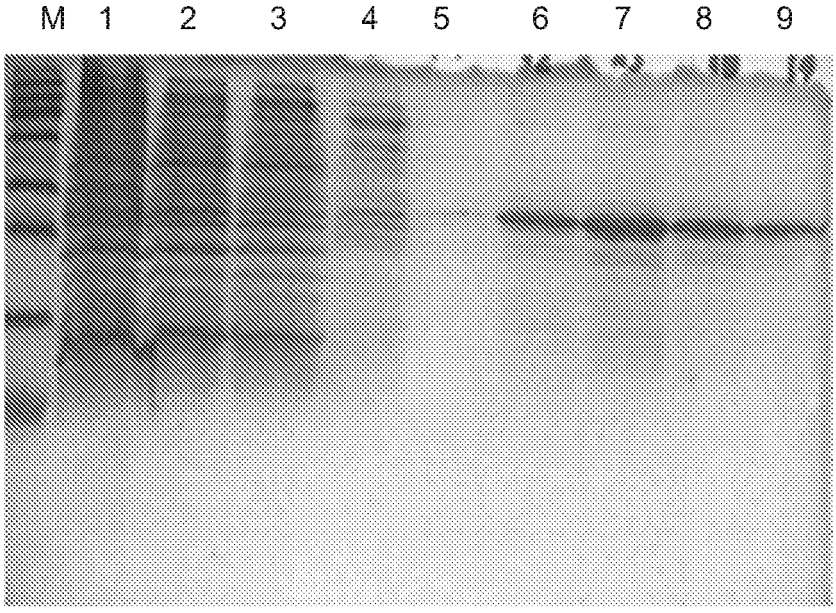

M 1 2 3 4 5 6 7 8 9

FIG. 4B

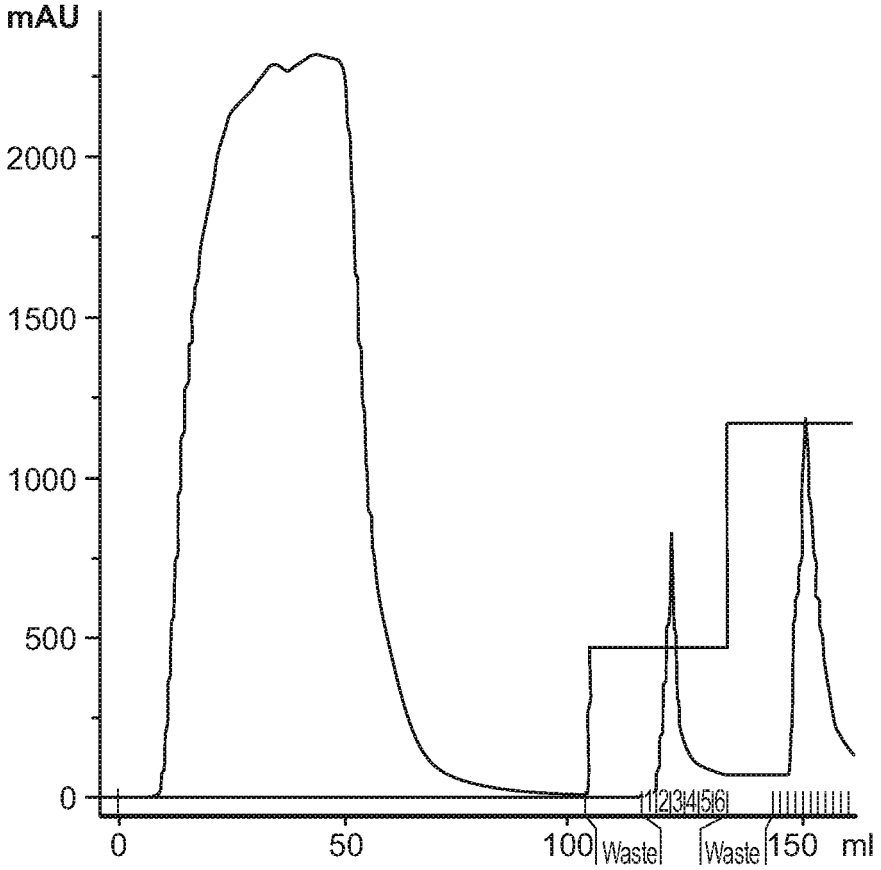

mAU
2000
1500
1000
500
0
0
50
100
Waste
Waste
150
ml
1|2|3|4|5|6|

FIG. 4C

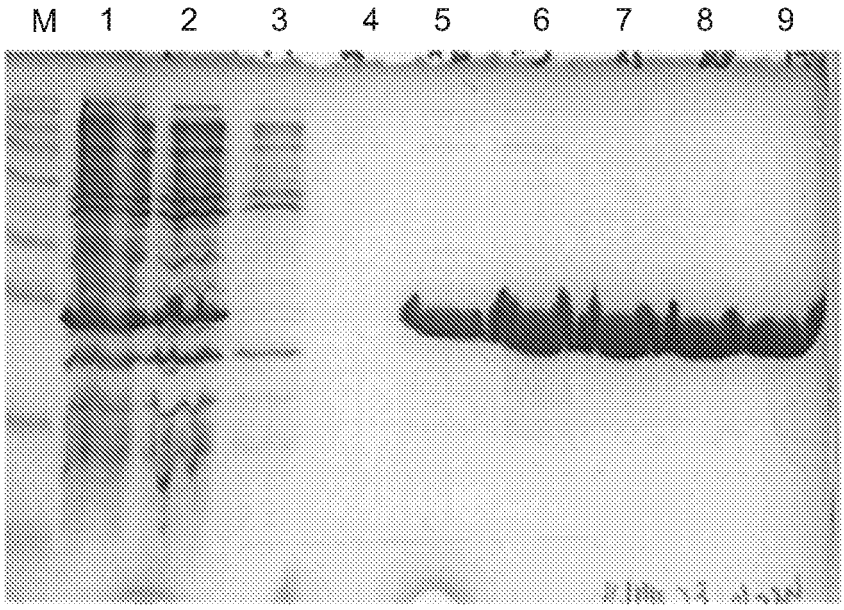

M
1
2
3
4
5
6
7
8
9

FIG. 4D

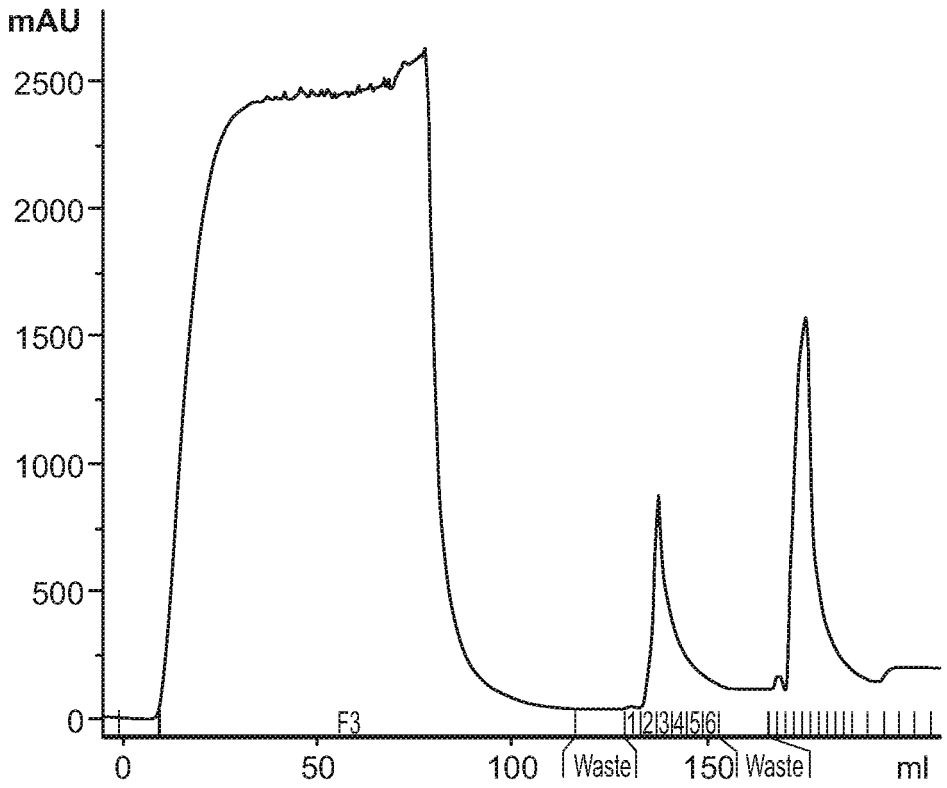

mAU
2500
2000
1500
1000
500
0
F3
1|2|3|4|5|6|
0
50
100
Waste
150
Waste
ml

FIG. 5A

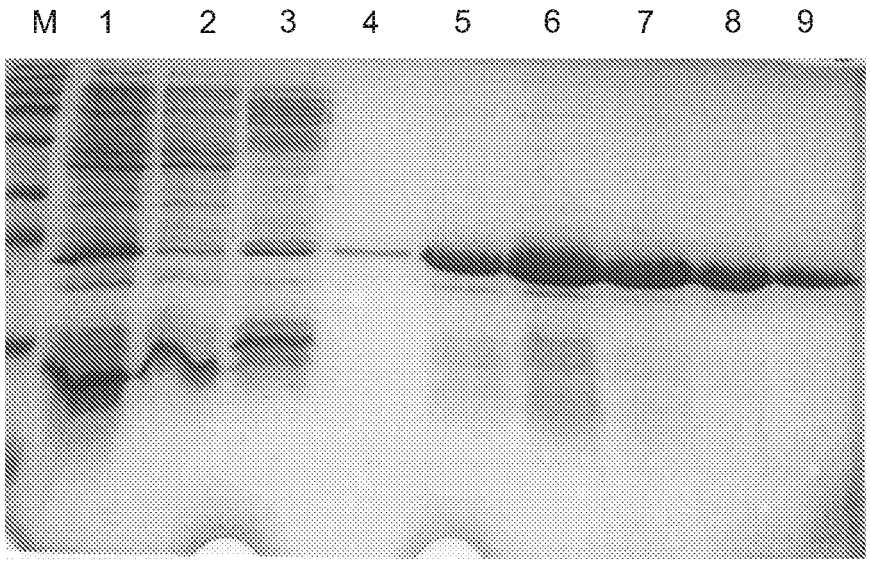

M 1 2 3 4 5 6 7 8 9

FIG. 5B

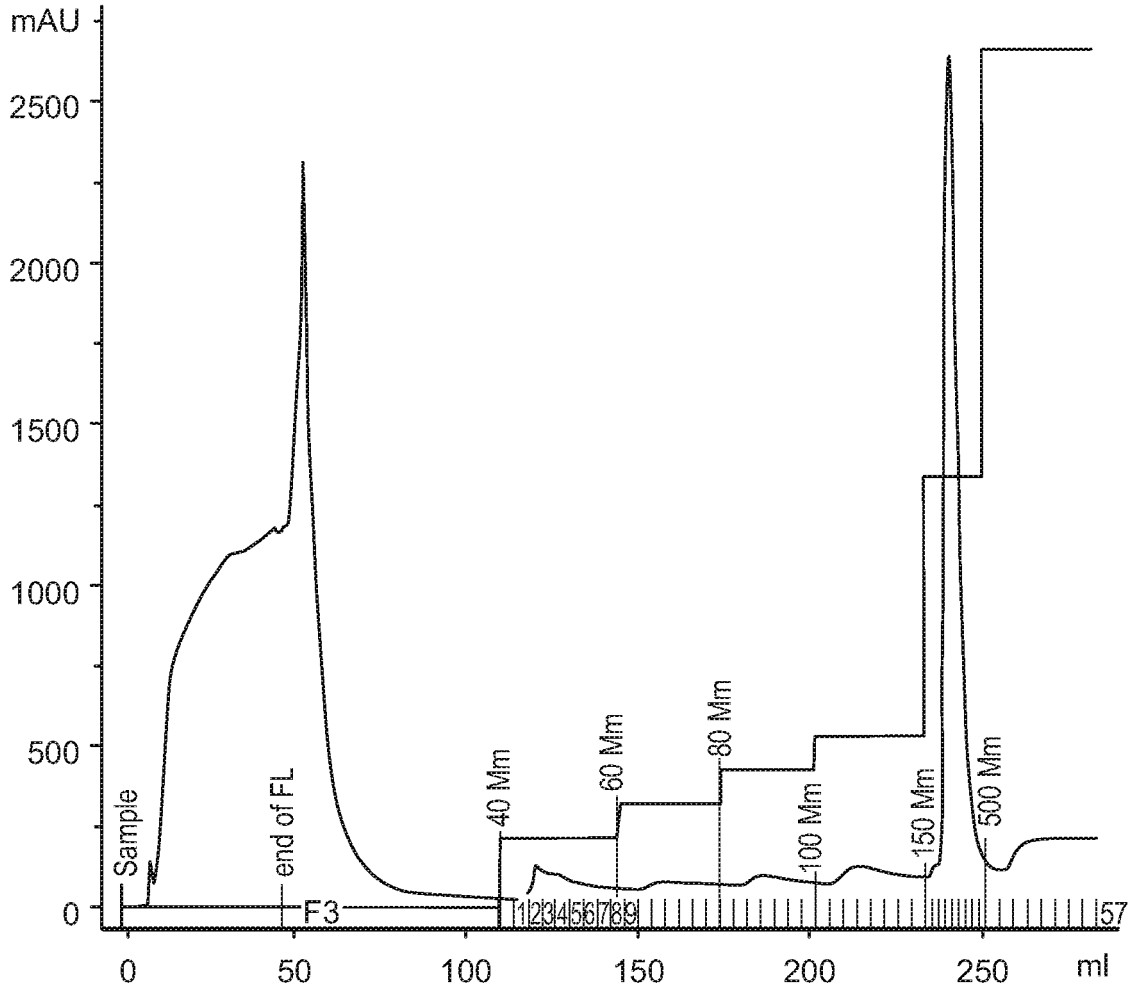

mAU
2500
2000
1500
1000
500
0
Sample
end of FL
F3
40 Mm
60 Mm
80 Mm
100 Mm
150 Mm
500 Mm
1 2 3 4 5 6 7 8 9
57
0
50
100
150
200
250
ml

FIG. 5C

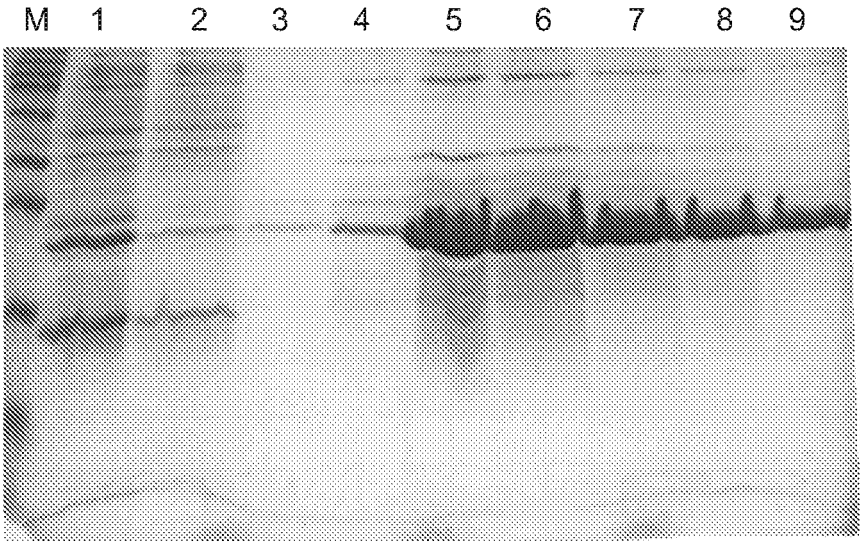

M 1 2 3 4 5 6 7 8 9

FIG. 5D

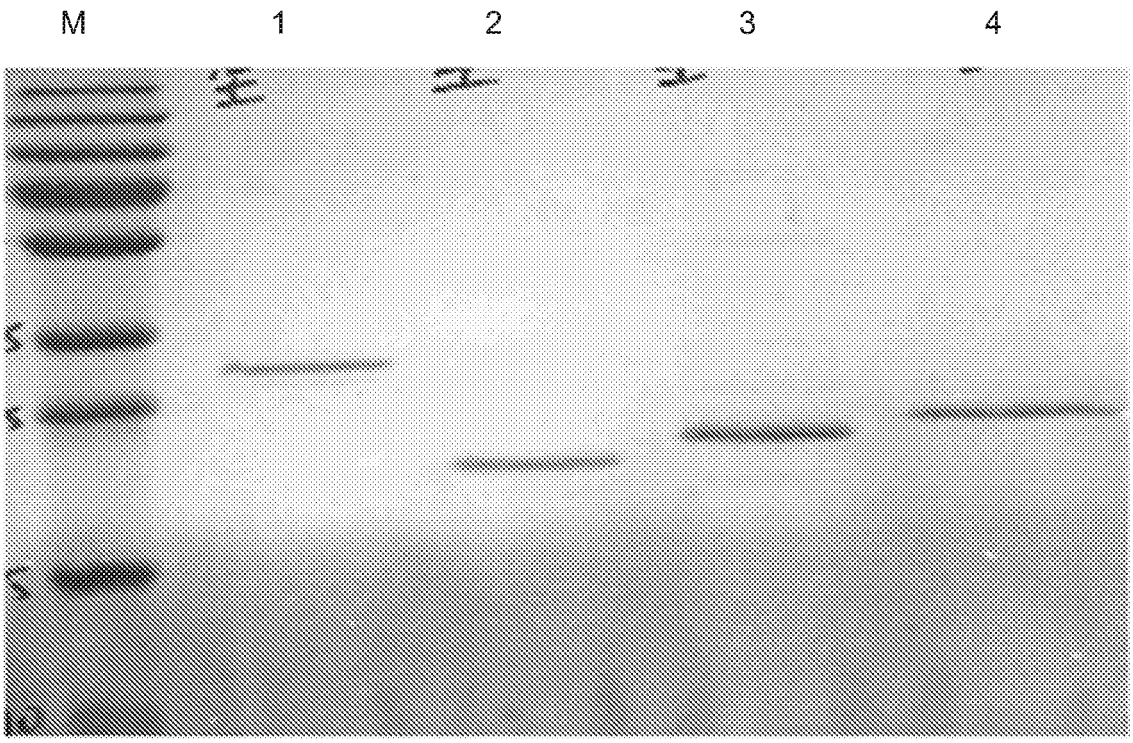


FIG. 6A

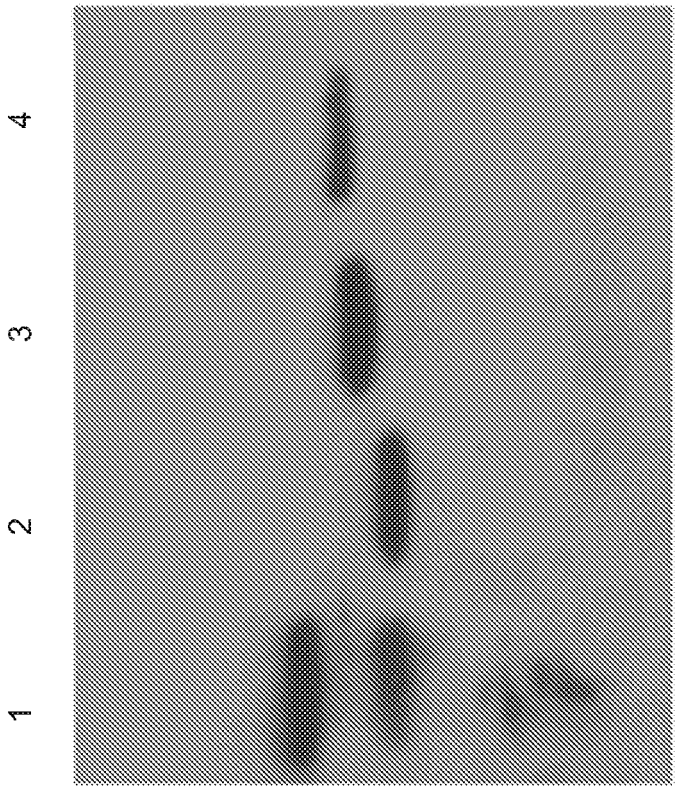


FIG. 6C

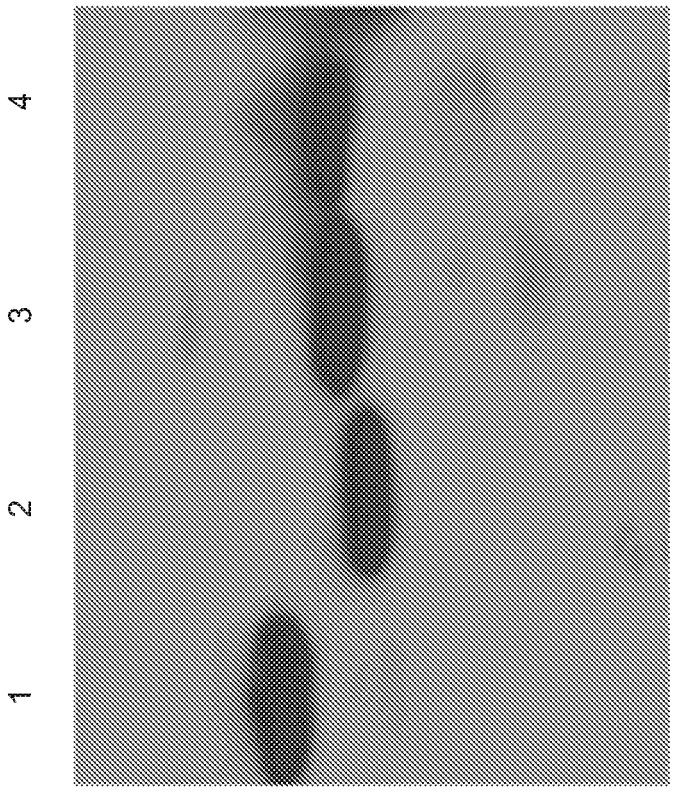


FIG. 6B

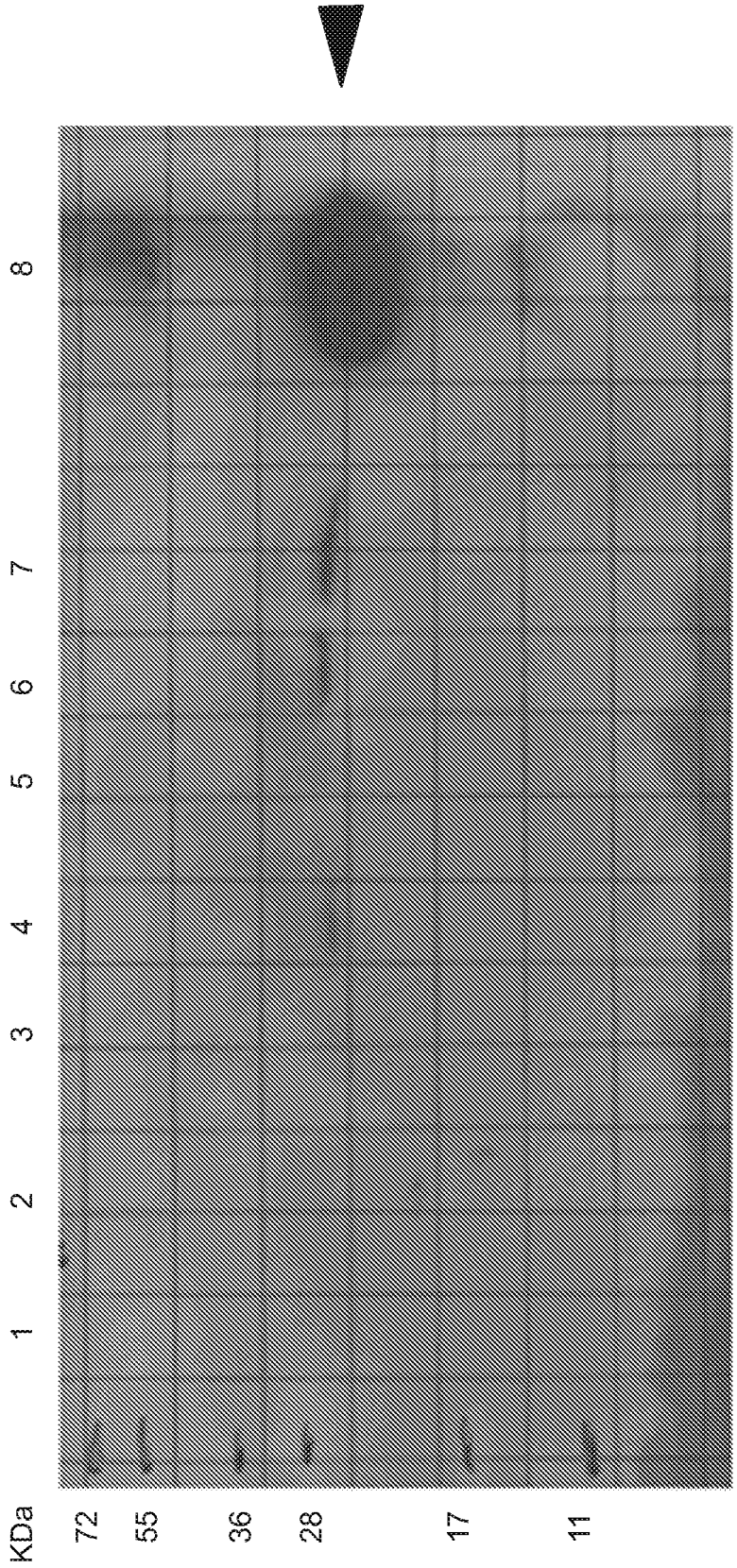


FIG. 7A

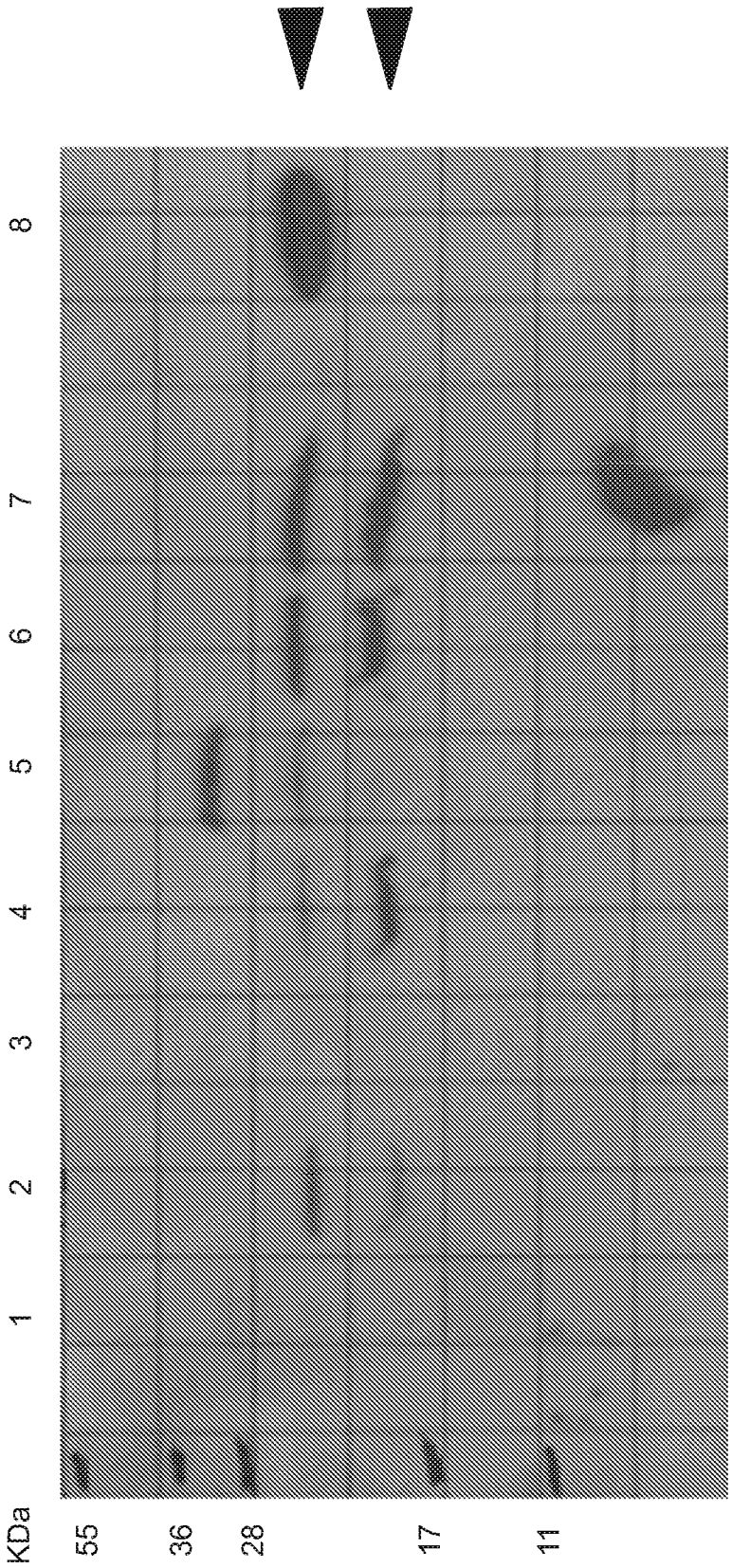


FIG. 7B

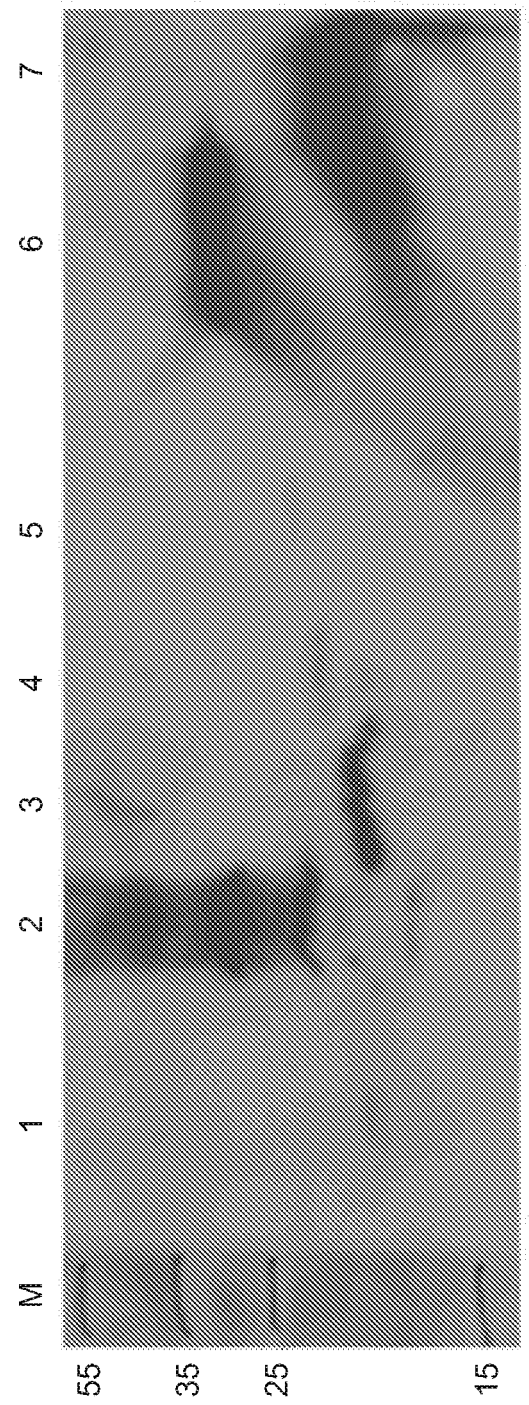


FIG. 8A

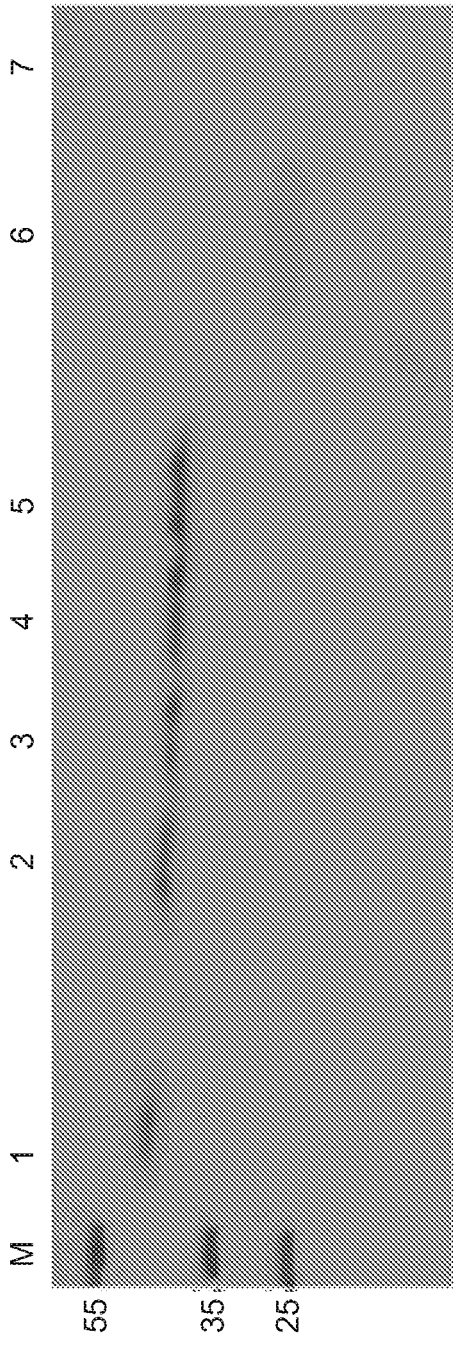


FIG. 8B

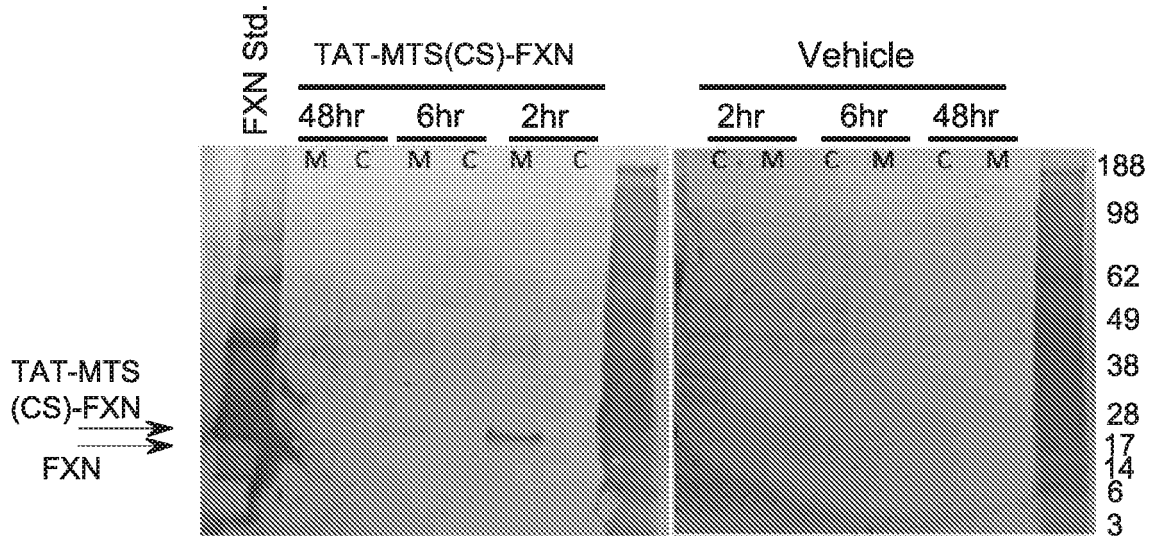

FXN Std.
TAT-MTS(CS)-FXN
48hr
6hr
2hr
Vehicle
2hr
6hr
48hr
M C M C M C C M C M C M
188
98
62
49
38
28
17
14
6
3
TAT-MTS
(CS)-FXN
FXN

FIG. 9

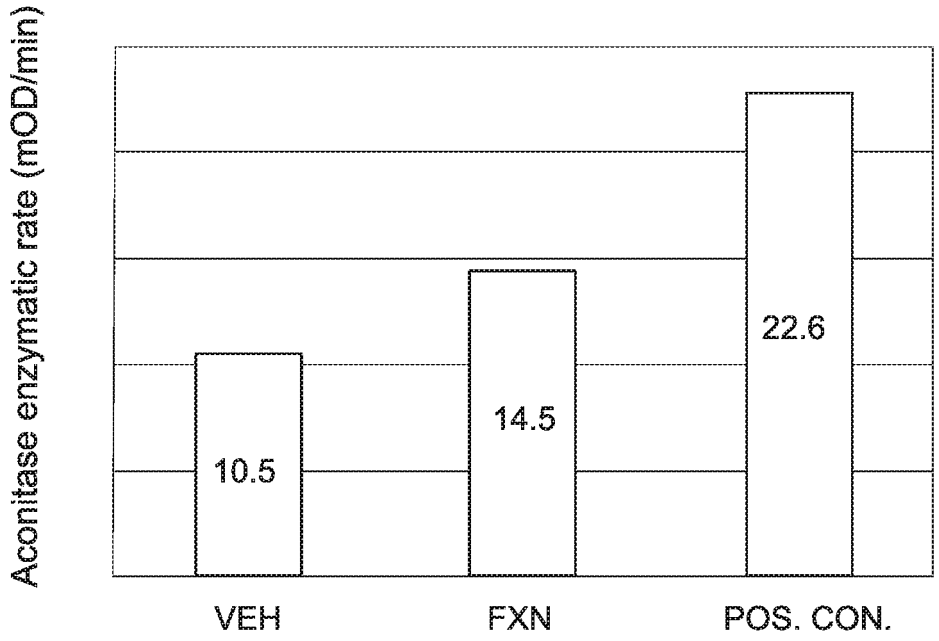

Aconitase enzymatic rate (mOD/min)
10.5
14.5
22.6
VEH
FXN
POS. CON.

FIG. 10

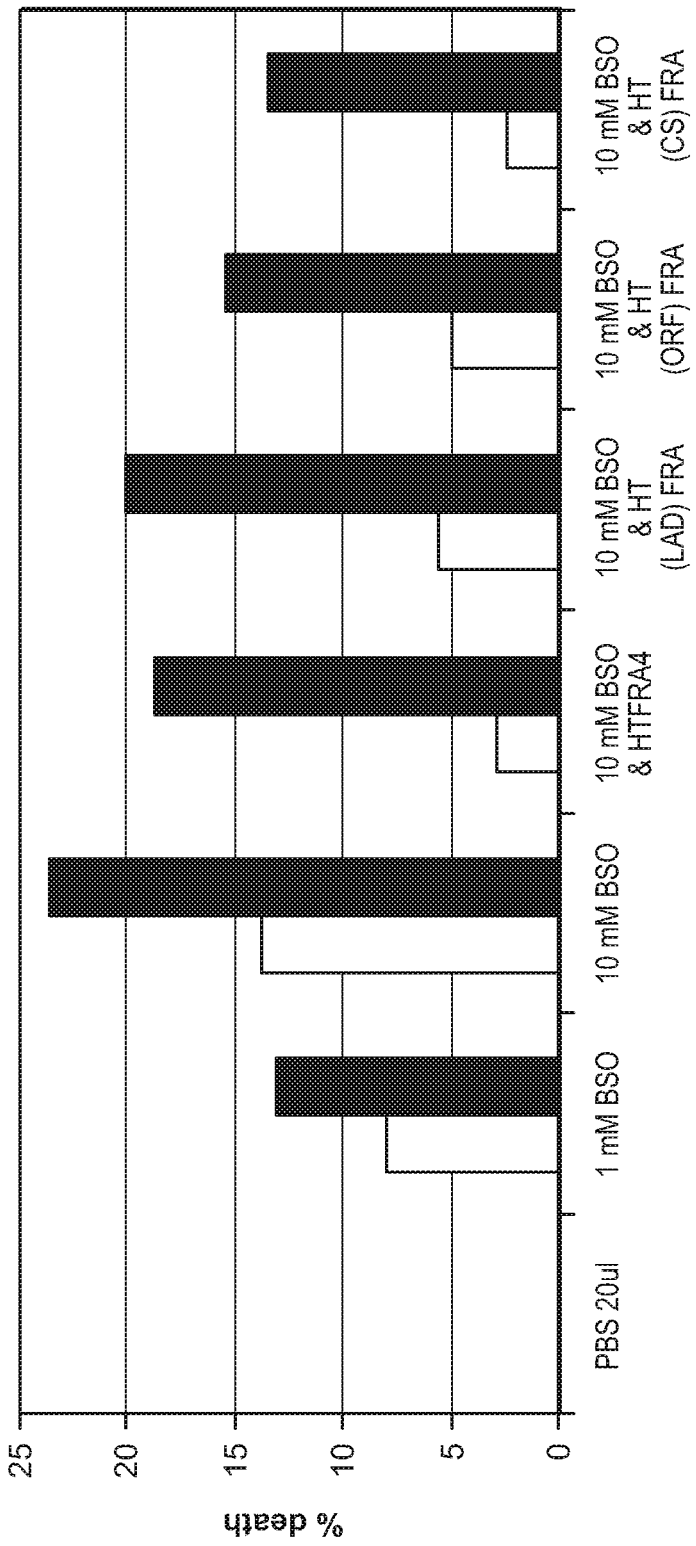

% death
25
20
15
10
5
0
PBS 20ul
1 mM BSO
10 mM BSO
10 mM BSO & HTFRA4
10 mM BSO & HT (LAD) FRA
10 mM BSO & HT (ORF) FRA
10 mM BSO & HT (CS) FRA
YC normal lym
FRA lym

FIG. 11A

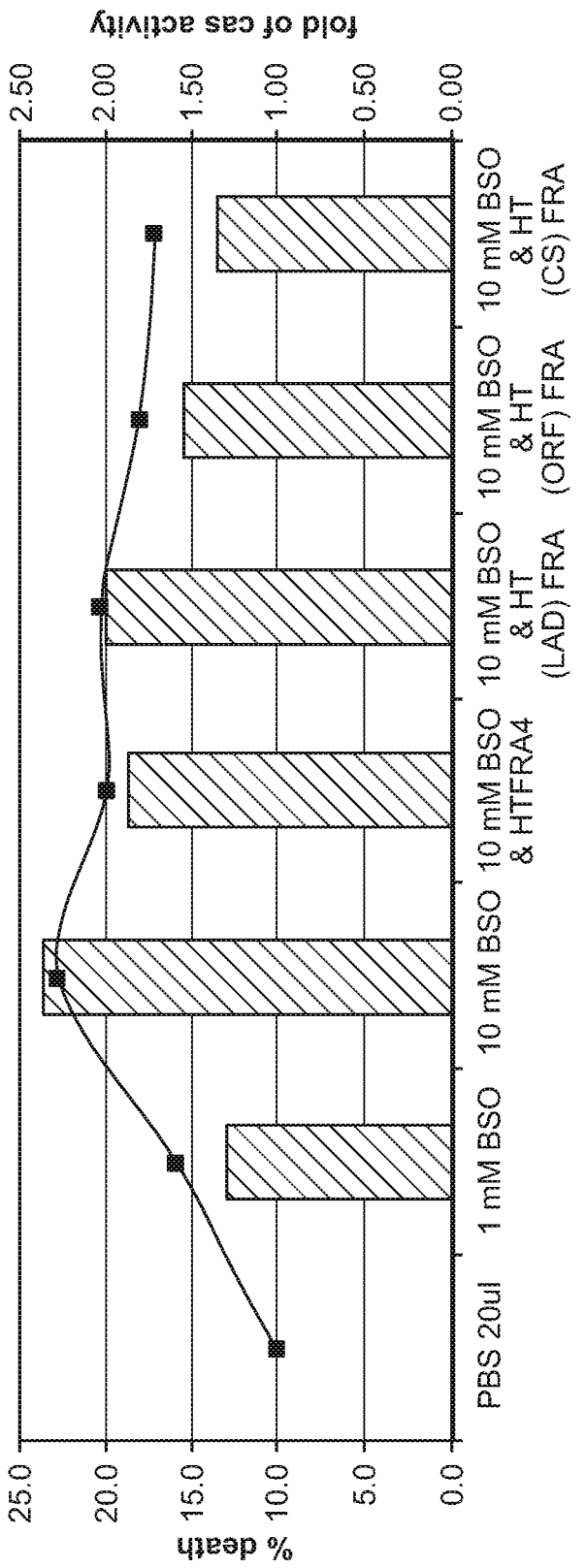

fold of cas activity
2.50
2.00
1.50
1.00
0.50
0.00
% death
25.0
20.0
15.0
10.0
5.0
0.0
PBS 20ul
1 mM BSO
10 mM BSO
10 mM BSO & HTFRA4
10 mM BSO & HT (LAD) FRA
10 mM BSO & HT (ORF) FRA
10 mM BSO & HT (CS) FRA
% DEATH
CAS

FIG. 11B

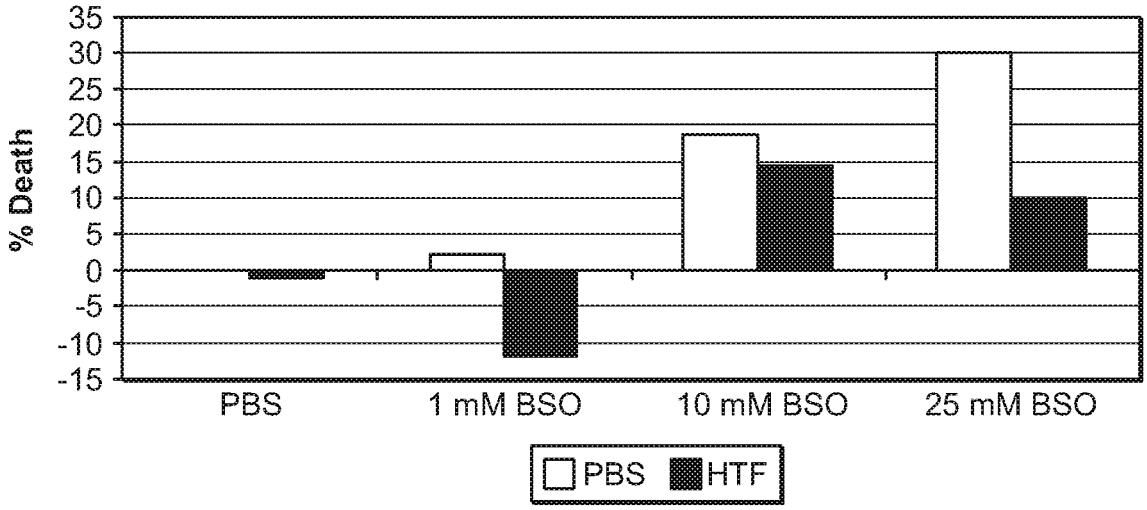


FIG. 12A

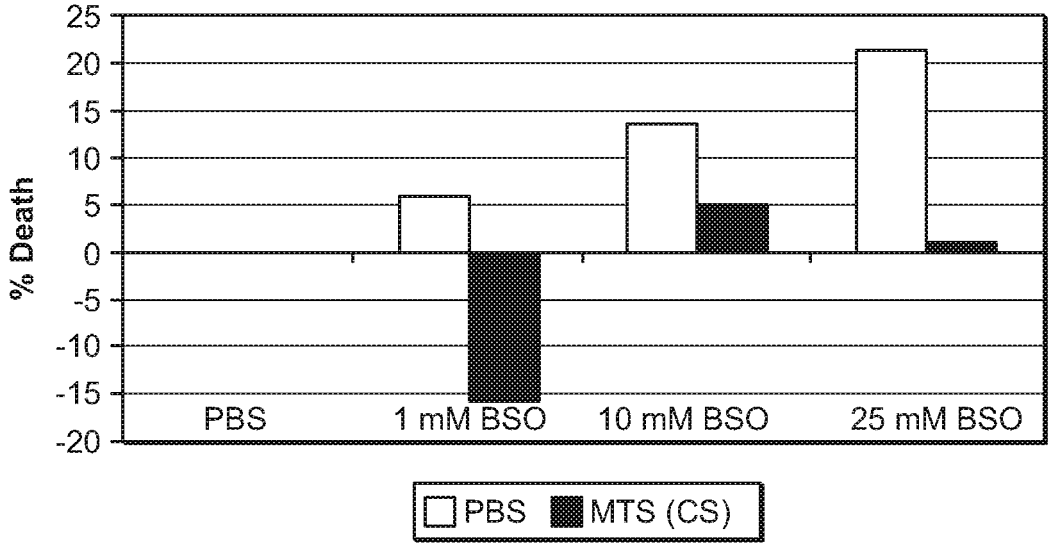


FIG. 12B

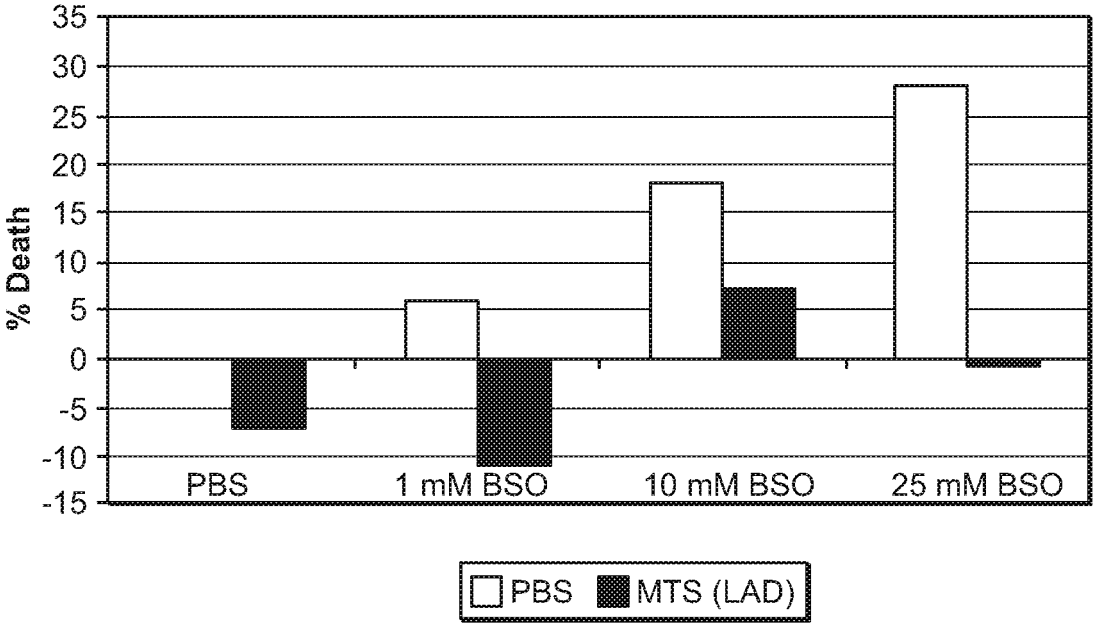


FIG. 12C

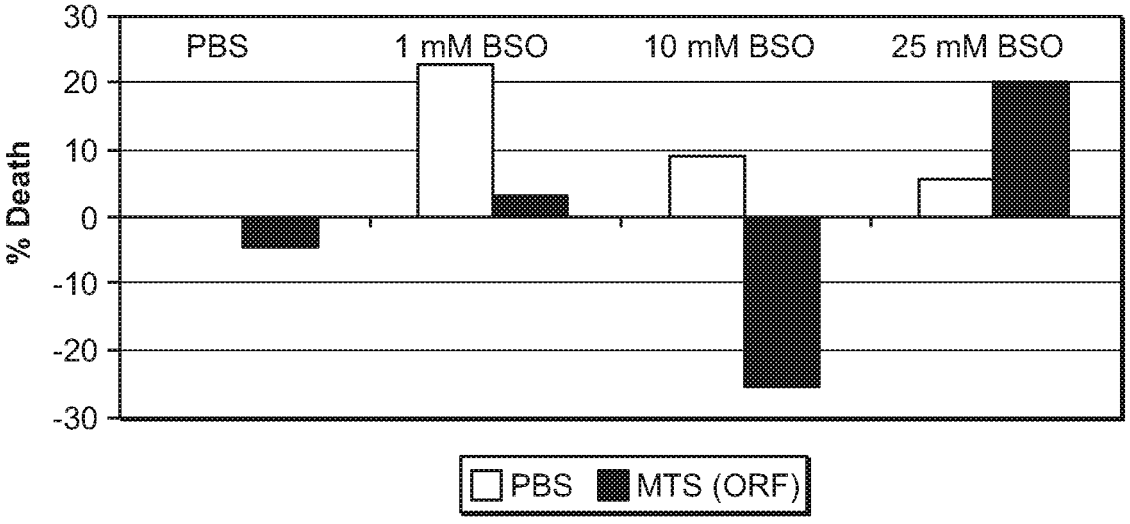


FIG. 12D

FIG. 13A

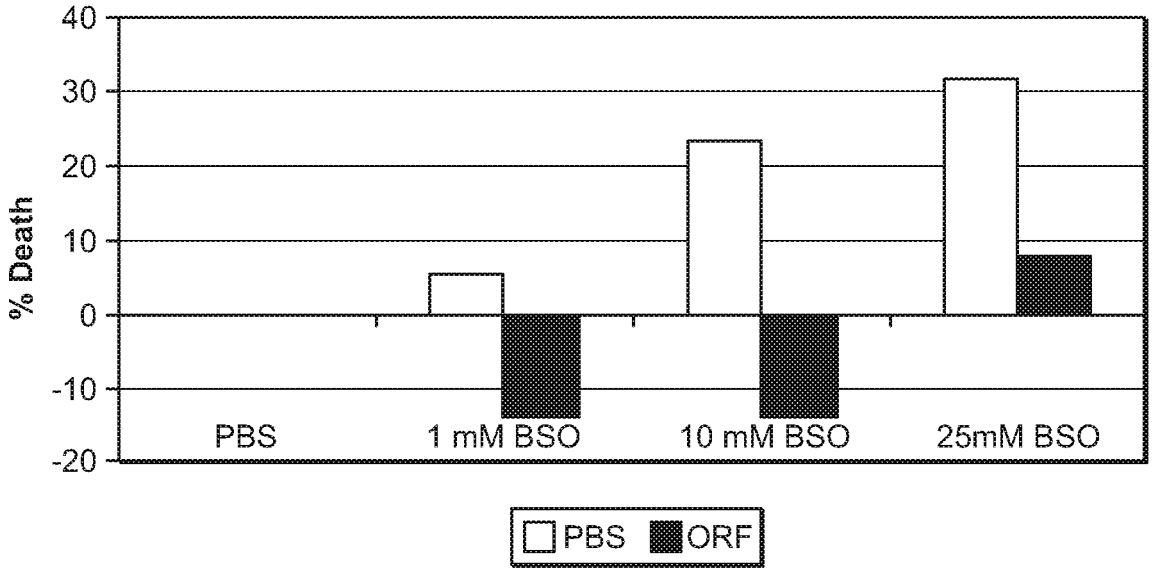
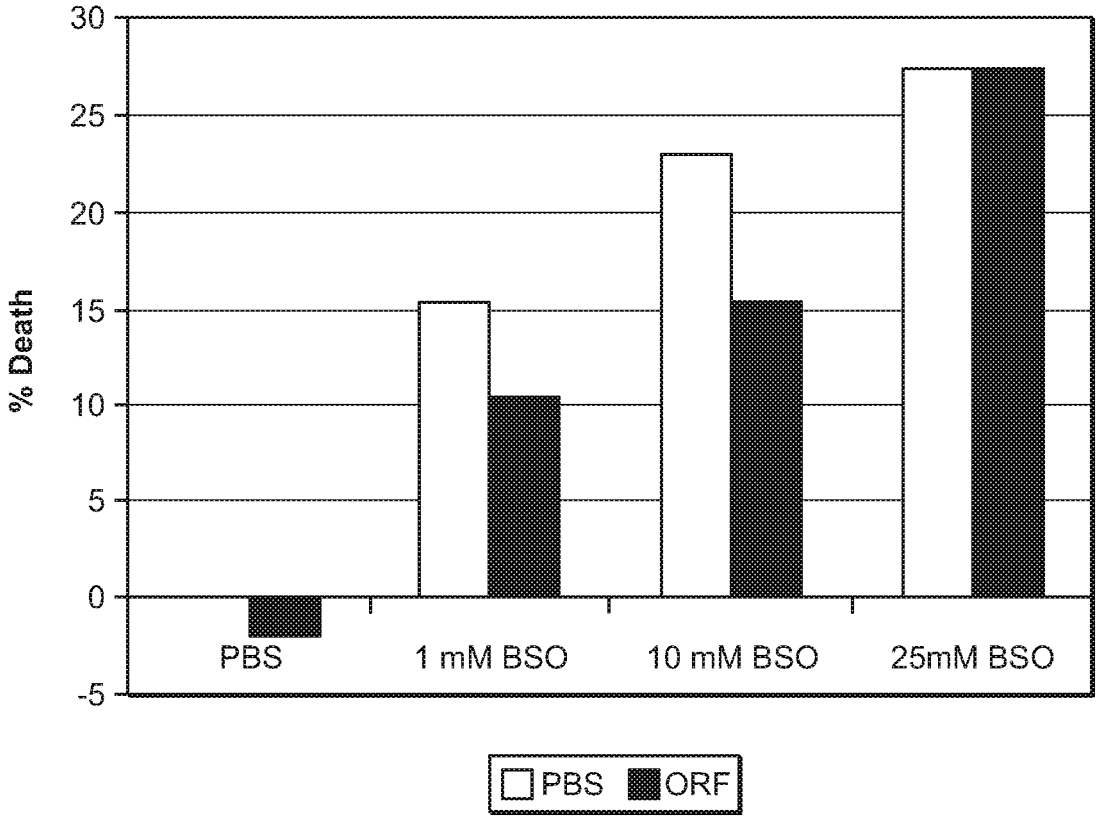


FIG. 13B



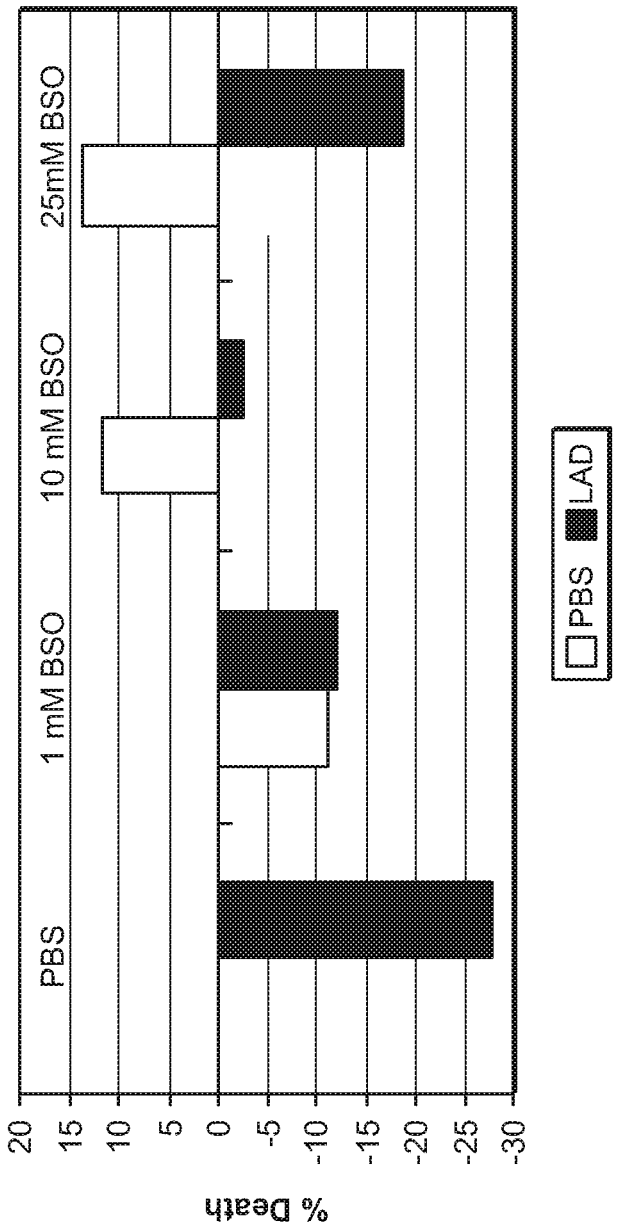

% Death
20
15
10
5
0
-5
-10
-15
-20
-25
-30
PBS
1 mM BSO
10 mM BSO
25mM BSO
PBS
LAD

FIG. 14A

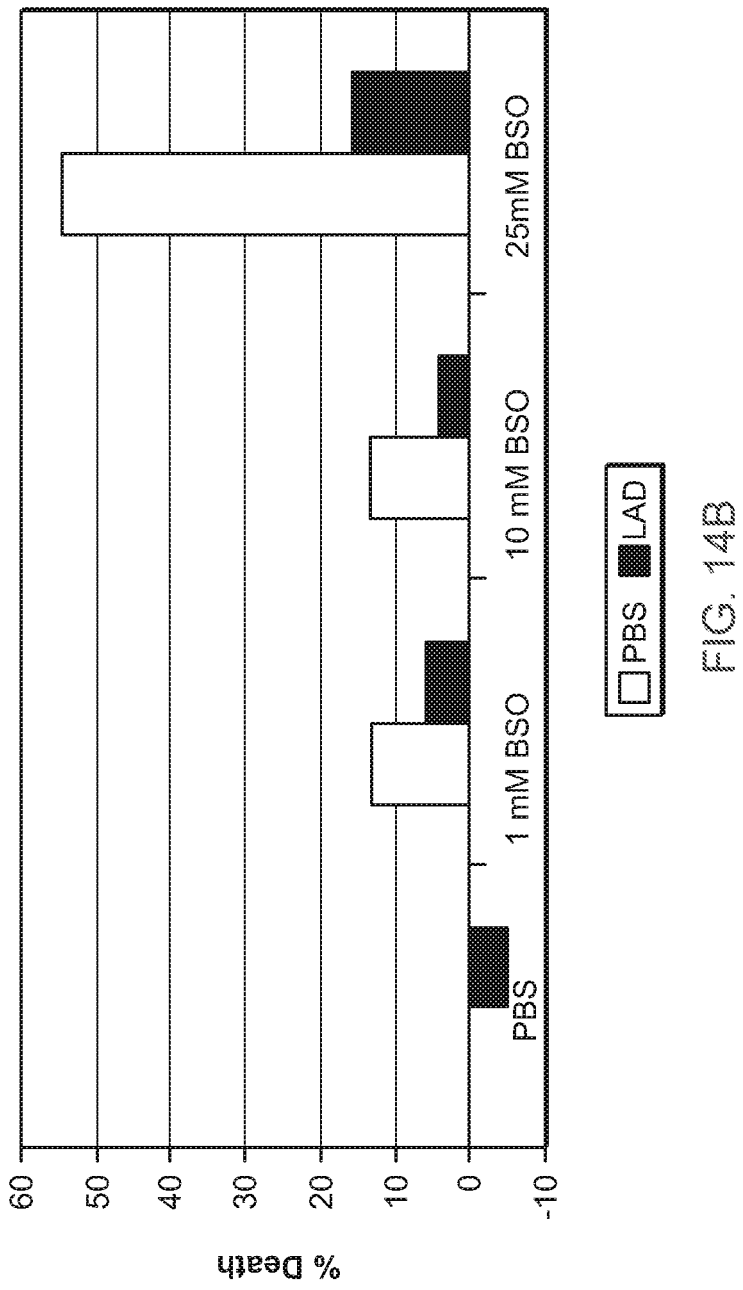


FIG. 14B

FIG. 15A

MRKKRRQRRRGSDPWTLGRRAVAGLLASPSPAQAQTLTRVPRPAELAPLCGRR
GLRTDIDATCTPRRASSNQRGLNQIWNVKKQSVYLMNLRKSGTLGHPGSLDETT
YERLAEETLDSLAEFFEDLADKPYTFEDYDVSFGSGVLTVKLGGDLGTYVINKQT
PNKQIWLSSPSSGPKRYDWTGKNWVYSHDGVSLHELLAAELTKALKTKLDLSSL
AYSGKDA

FIG. 15B

MRKKRRQRRRGSDPALLTAAARLLGTKNASCLVLAARHASSGTLGHPGSLDETT
YERLAEETLDSLAEFFEDLADKPYTFEDYDVSFGSGVLTVKLGGDLGTYVINKQT
PNKQIWLSSPSSGPKRYDWTGKNWVYSHDGVSLHELLAAELTKALKTKLDLSSL
AYSGKDA

FIG. 15C

MRKKRRQRRRGSDPQSWSRVYCSLAKRGHFNRISHGLQGLSAVPLRTYASGTLG
HPGSLDETTYERLAEETLDSLAEFFEDLADKPYTFEDYDVSFGSGVLTVKLGGDL
GTYVINKQTPNKQIWLSSPSSGPKRYDWTGKNWVYSHDGVSLHELLAAELTKAL
KTKLDLSSLAYSGKDA

FIG. 15D

MRKKRRQRRRGSDPGALVIRGIRNFNLENRAEREISKMKPSVAPRHPSSGTLGHP
GSLDETTYERLAEETLDSLAEFFEDLADKPYTFEDYDVSFGSGVLTVKLGGDLGT
YVINKQTPNKQIWLSSPSSGPKRYDWTGKNWVYSHDGVSLHELLAAELTKALKT
KLDLSSLAYSGKDA

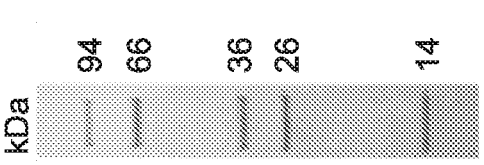


FIG. 16E

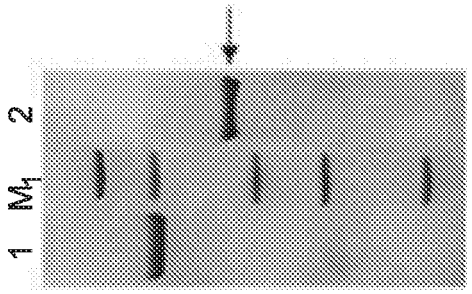


FIG. 16D

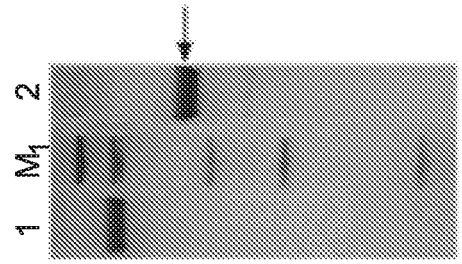


FIG. 16C

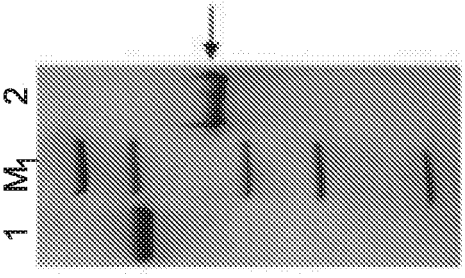


FIG. 16B

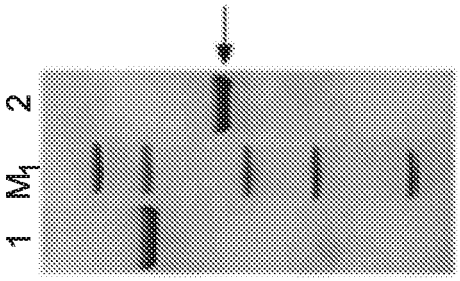


FIG. 16A

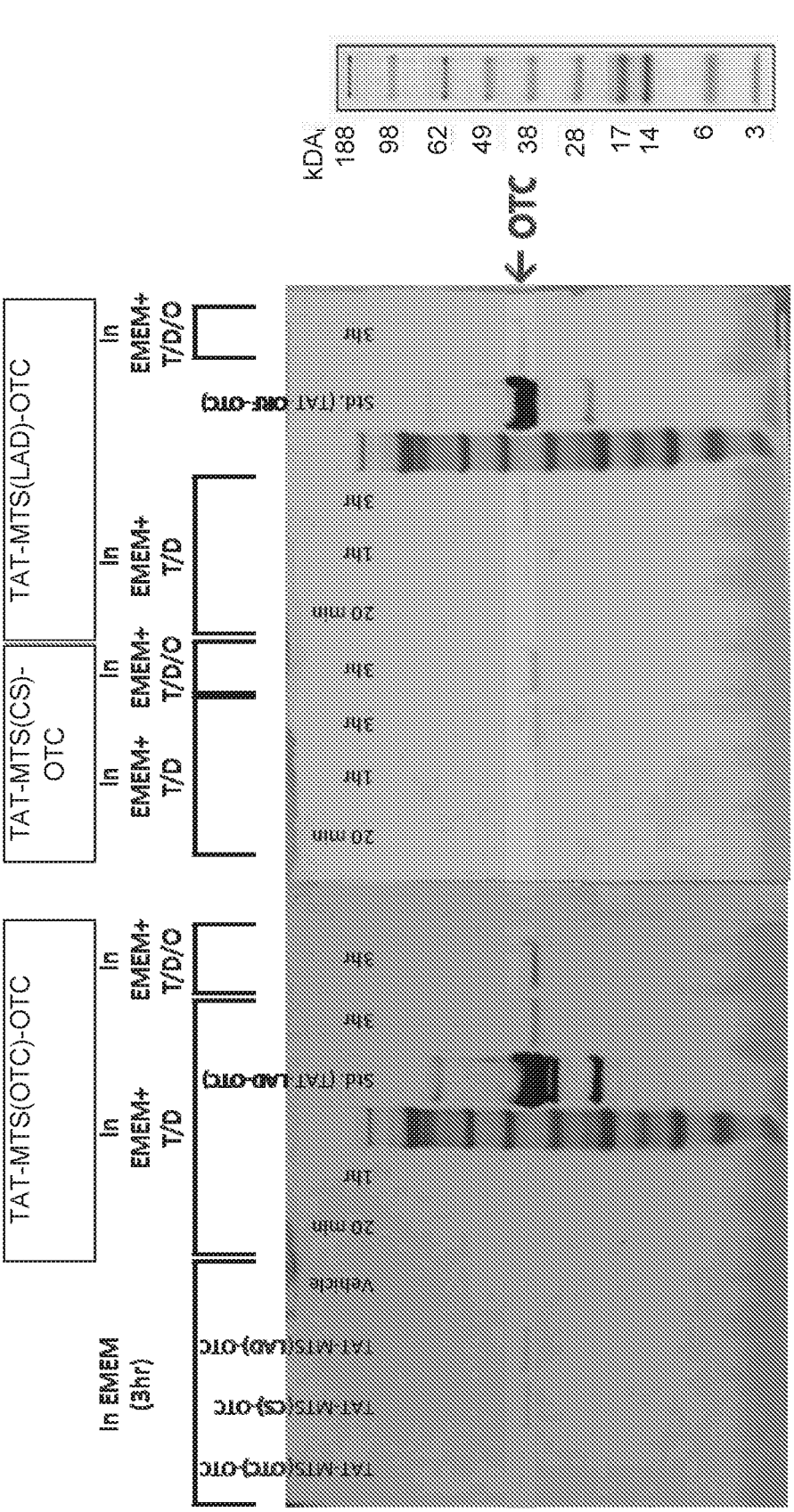


FIG. 17A

FIG. 17B

Protein [TAT-MTS(x)-OTC], X=	Incubation Medium	Incubation Time
OTC	EMEM	3 hr
Vehicle	EMEM	3 hr
CS	EMEM	3 hr
OTC	EMEM+T/D	20 min
LAD	EMEM	3 hr
OTC	EMEM+T/D	3 hr
OTC	EMEM+T/D	3 hr
OTC	EMEM+T/D/O	3 hr
CS	–	–

FIG. 18A

Protein [TAT-MTS(x)-OTC], X=	Incubation Medium	Incubation Time
CS	EMEM+T/D	20 min
CS	EMEM+T/D	1 hr
CS	EMEM+T/D	3 hr
CS	EMEM+T/D/O	3 hr
LAD	EMEM+T/D	20 min
LAD	EMEM+T/D	1 hr
LAD	EMEM+T/D	3 hr
ORF	–	–
		M
LAD	EMEM+T/D/O	3 hr

FIG. 18B

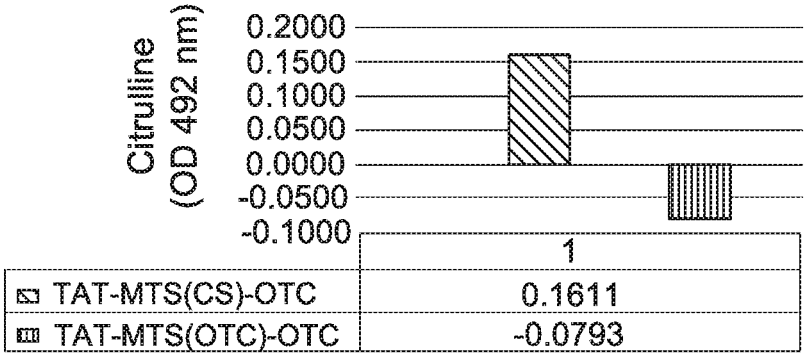

Citrulline (OD 492 nm)
0.2000
0.1500
0.1000
0.0500
0.0000
-0.0500
-0.1000
1
TAT-MTS(CS)-OTC
0.1611
TAT-MTS(OTC)-OTC
-0.0793

FIG. 19

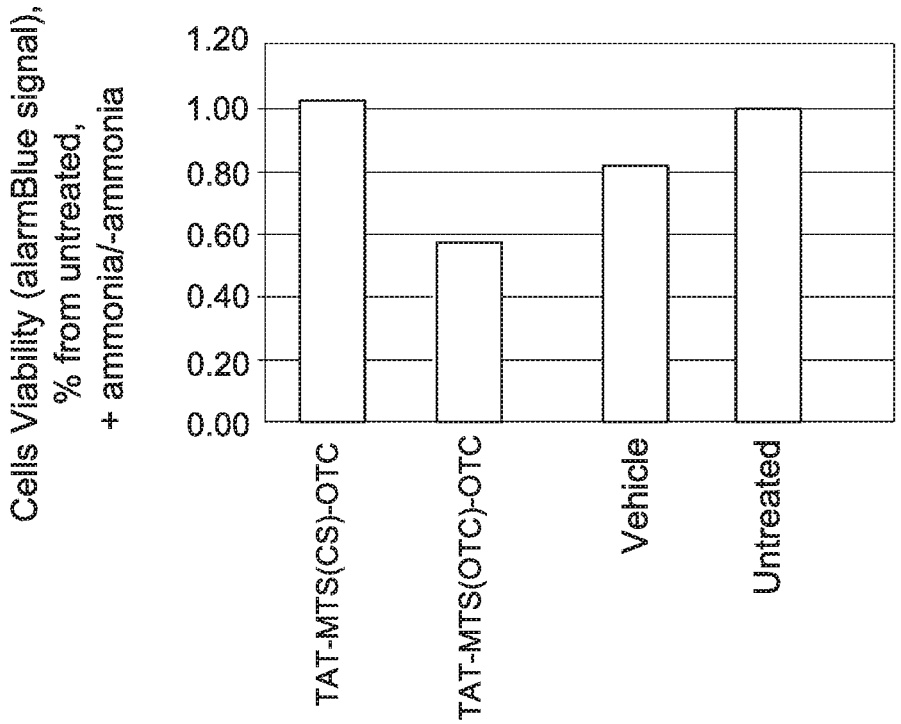

Cells Viability (alarmBlue signal), % from untreated, + ammonia/-ammonia
1.20
1.00
0.80
0.60
0.40
0.20
0.00
TAT-MTS(CS)-OTC
TAT-MTS(OTC)-OTC
Vehicle
Untreated

FIG. 20