Fig. 1

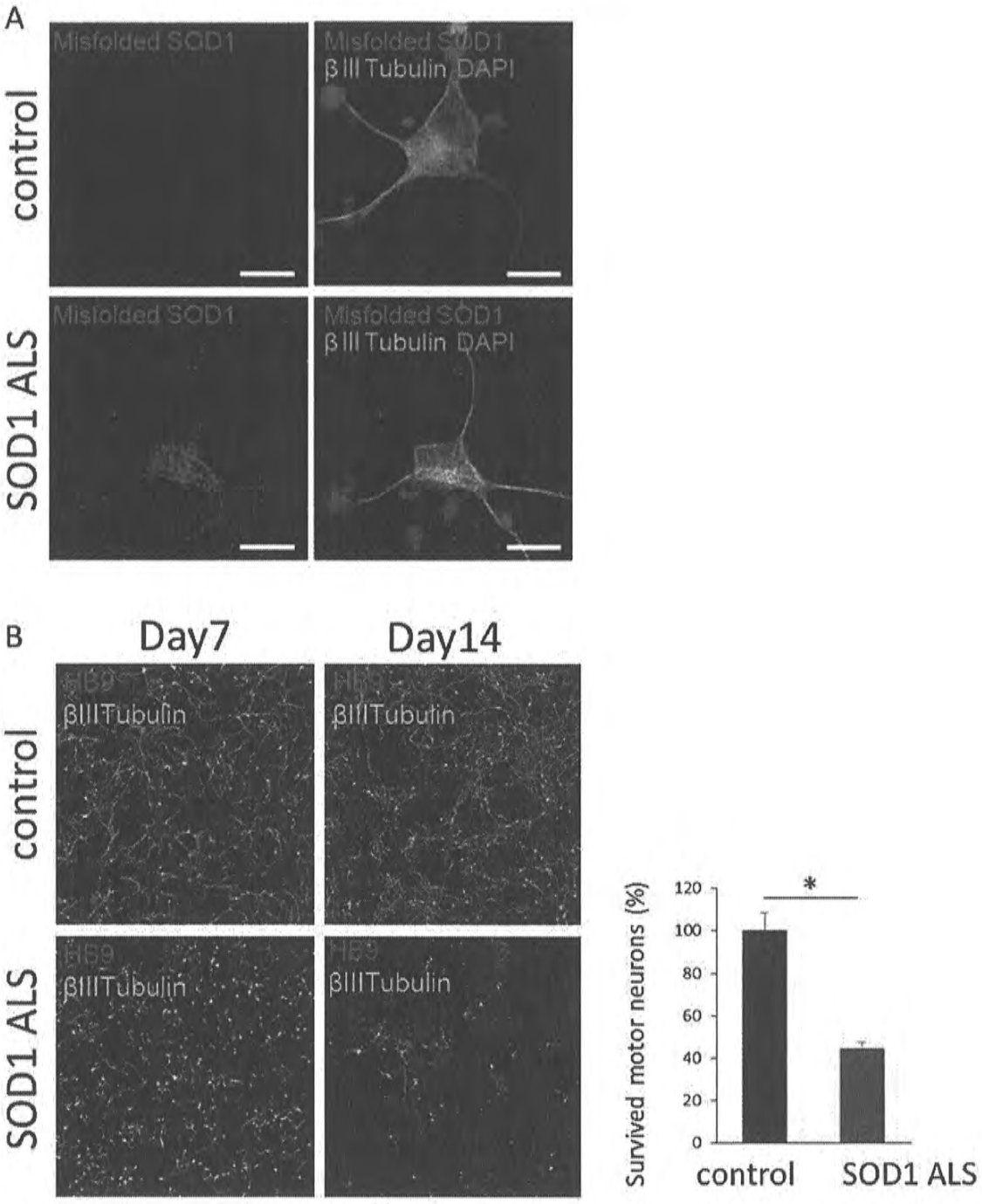

A
control
SOD1 ALS
Misfolded SOD1
Misfolded SOD1
βIII Tubulin DAPI
B
Day7
Day14
βIIITubulin
Survived motor neurons (%)
120
100
80
60
40
20
0
*
control
SOD1 ALS

Fig. 2

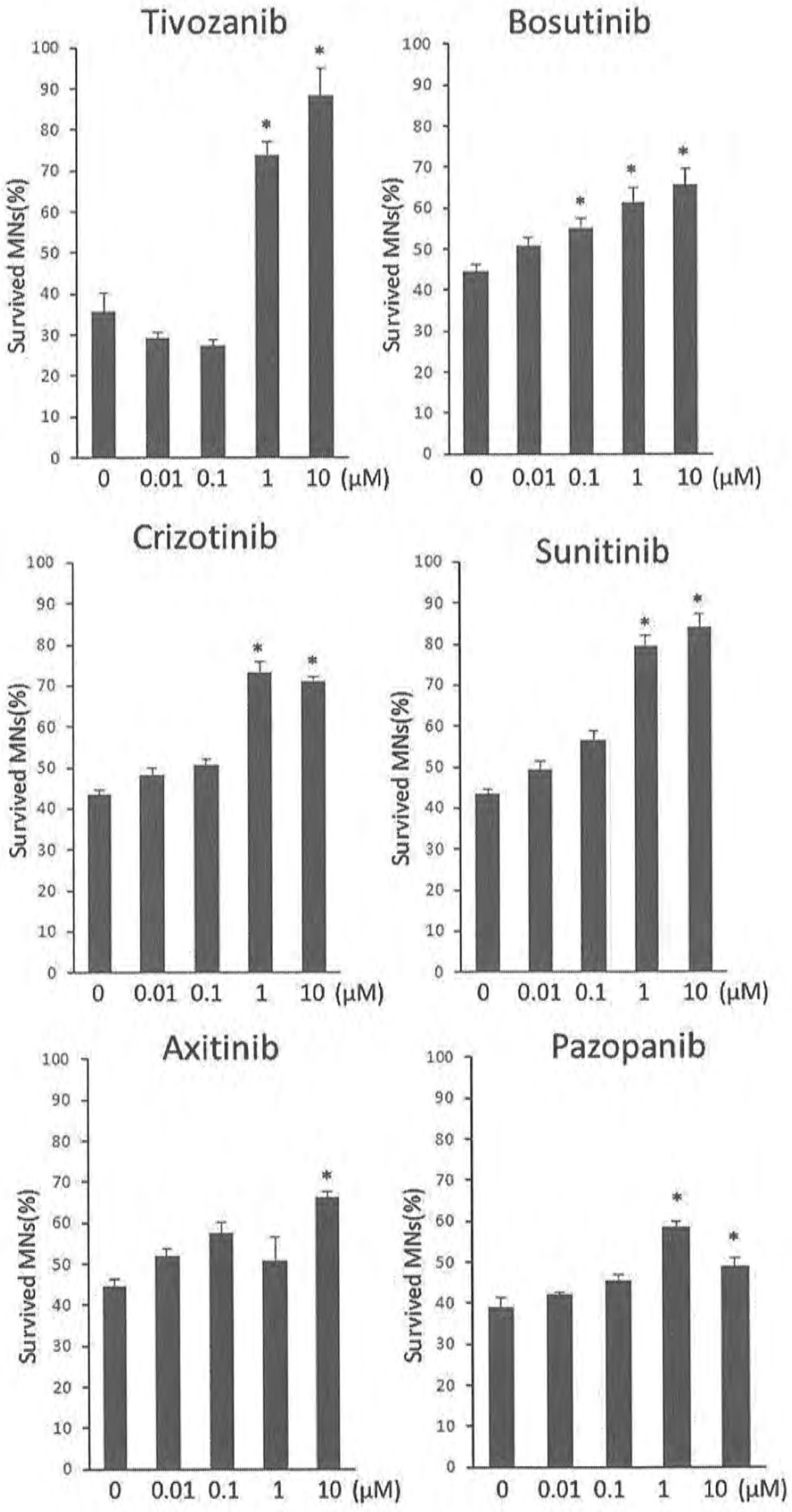

Tivozanib
Bosutinib
Crizotinib
Sunitinib
Axitinib
Pazopanib
Survived MNs(%)
0 0.01 0.1 1 10 (μM)

Fig. 3

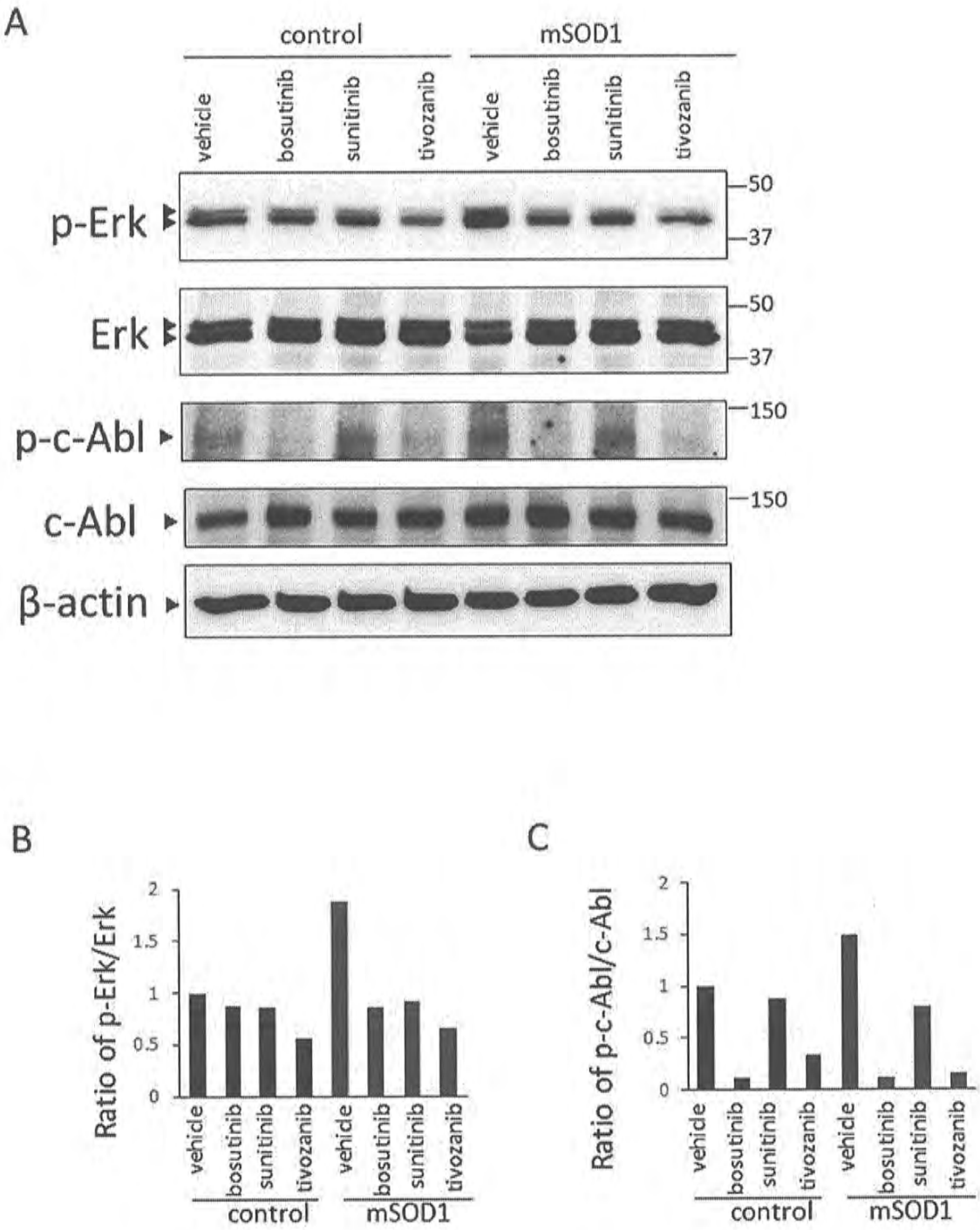

A
control
mSOD1
vehicle
bosutinib
sunitinib
tivozanib
vehicle
bosutinib
sunitinib
tivozanib
p-Erk
Erk
p-c-Abl
c-Abl
β-actin
50
37
50
37
150
150
B
Ratio of p-Erk/Erk
2
1.5
1
0.5
0
vehicle
bosutinib
sunitinib
tivozanib
vehicle
bosutinib
sunitinib
tivozanib
control
mSOD1
C
Ratio of p-c-Abl/c-Abl
2
1.5
1
0.5
0
vehicle
bosutinib
sunitinib
tivozanib
vehicle
bosutinib
sunitinib
tivozanib
control
mSOD1

Fig. 4

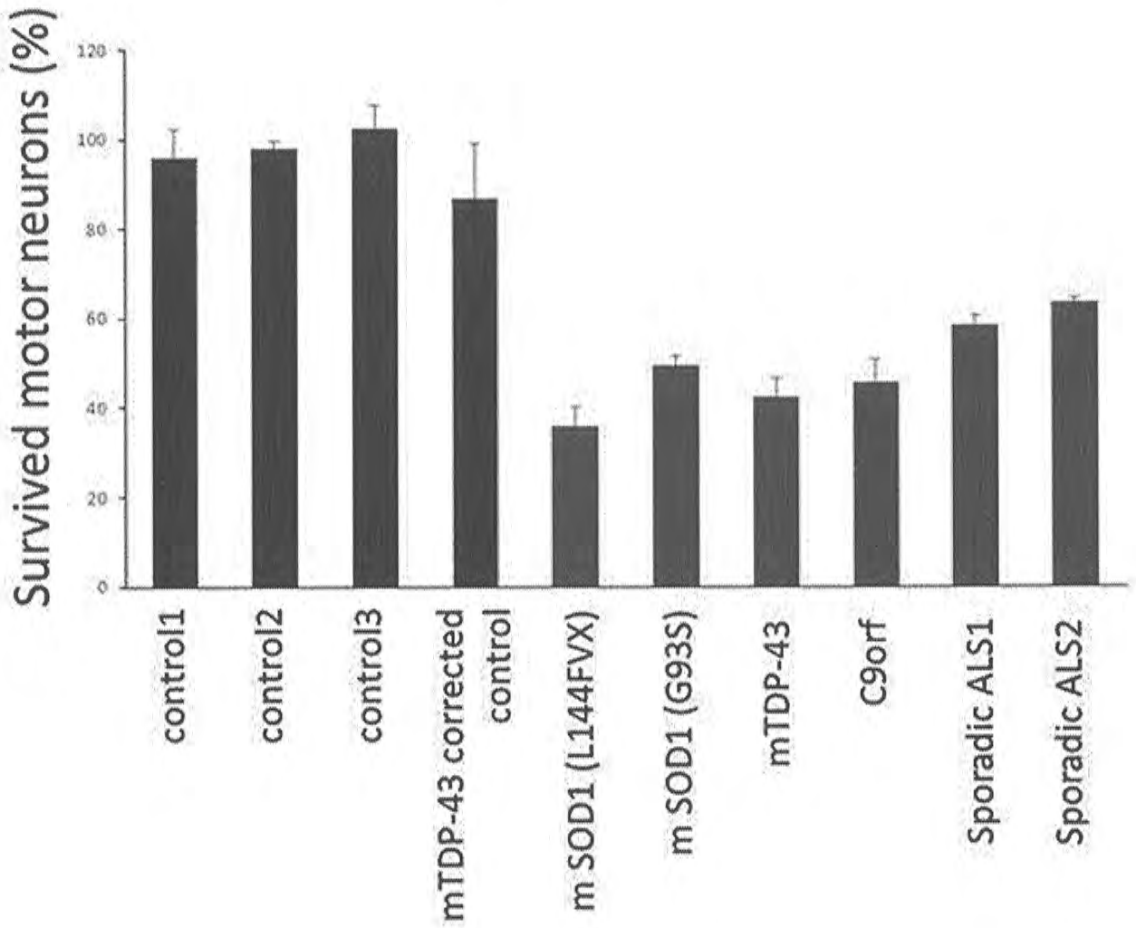

Survived motor neurons (%)
120
100
80
60
40
20
0
control1
control2
control3
mTDP-43 corrected control
m SOD1 (L144FVX)
m SOD1 (G93S)
mTDP-43
C9orf
Sporadic ALS1
Sporadic ALS2

Fig. 5

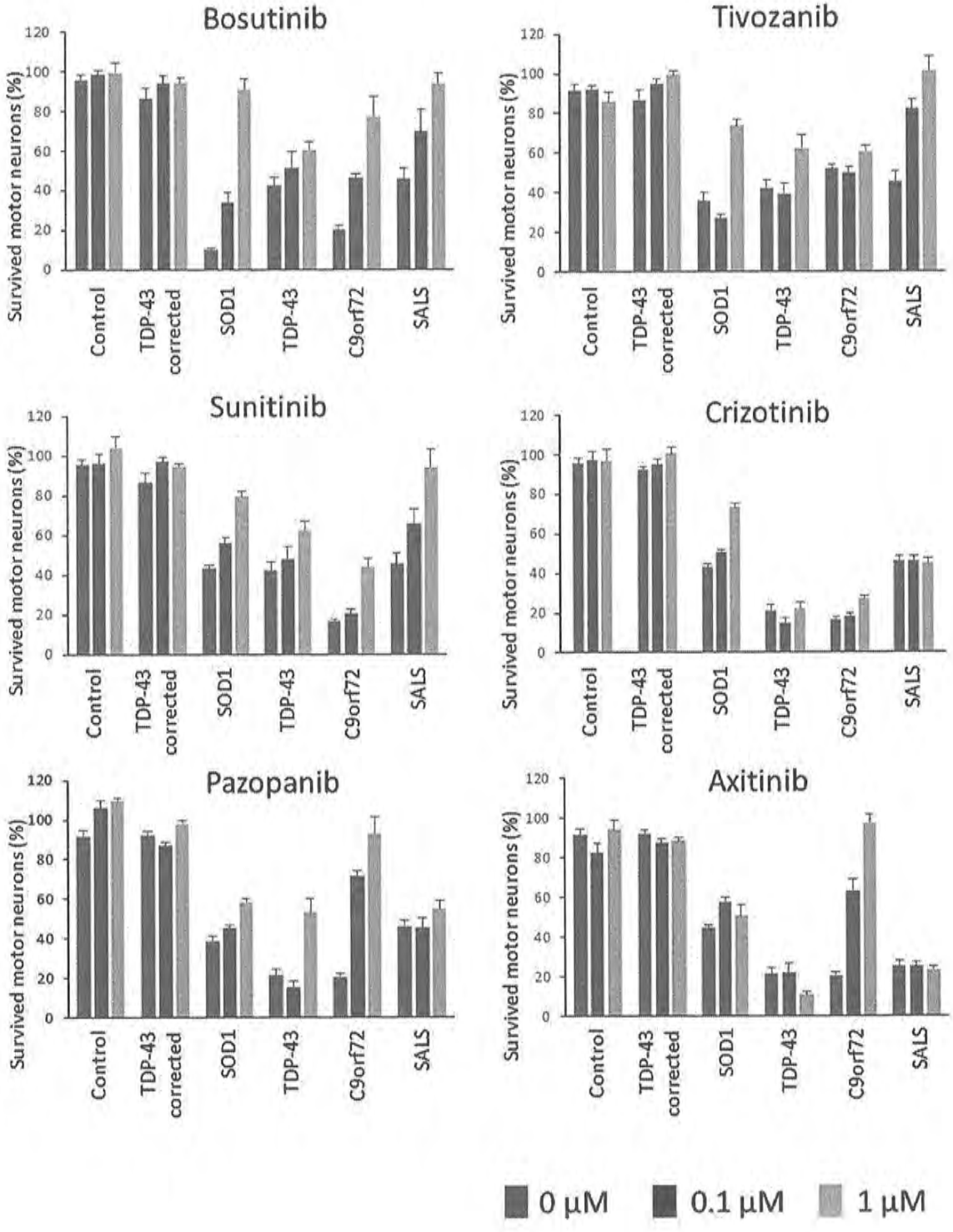

Bosutinib
Tivozanib
Sunitinib
Crizotinib
Pazopanib
Axitinib
Survived motor neurons (%)
0
20
40
60
80
100
120
Control
TDP-43 corrected
SOD1
TDP-43
C9orf72
SALS
0 μM
0.1 μM
1 μM

Fig. 6

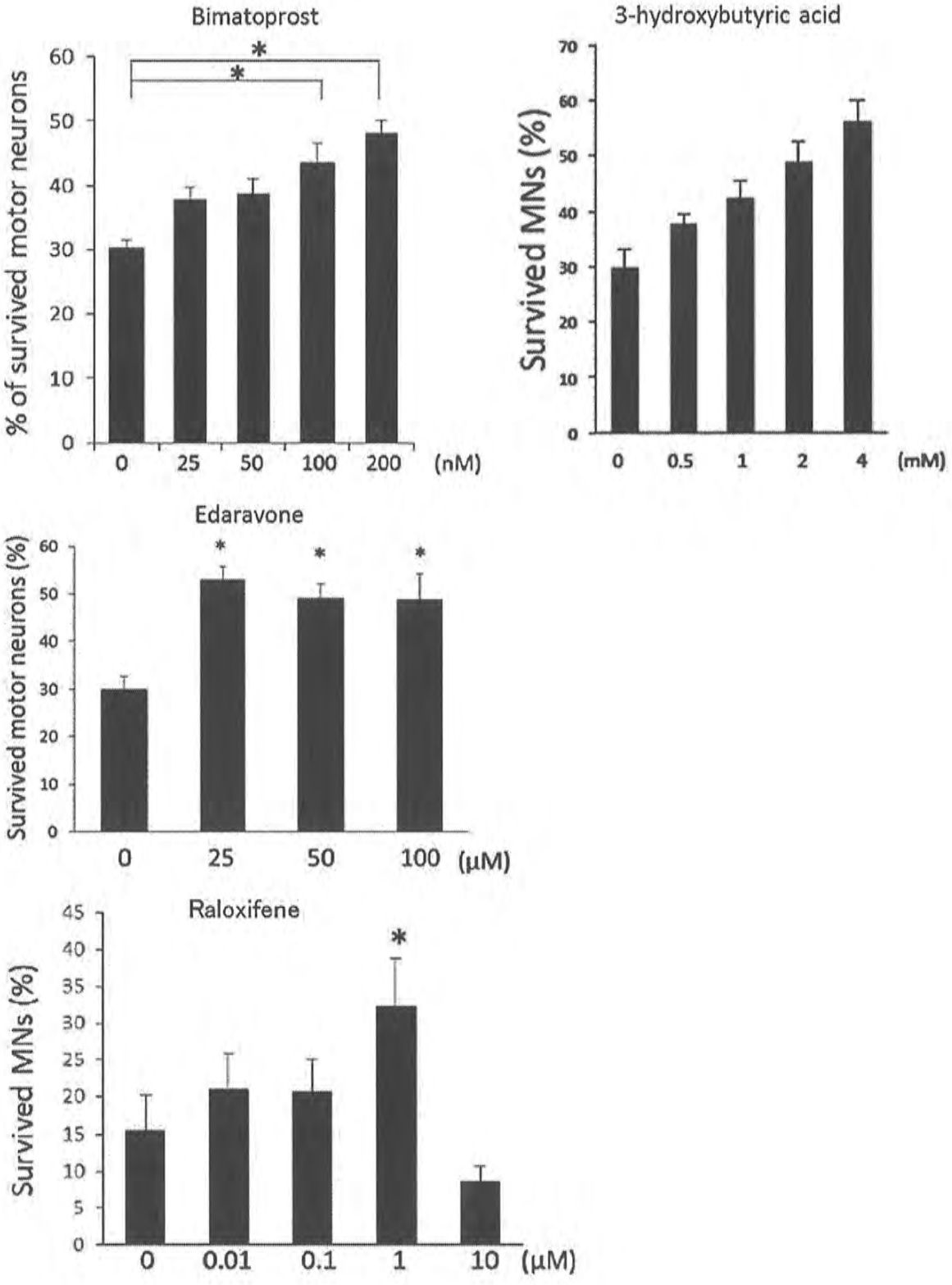

Bimatoprost
% of survived motor neurons
0
10
20
30
40
50
60
*
*
0
25
50
100
200
(nM)
3-hydroxybutyric acid
Survived MNs (%)
0
10
20
30
40
50
60
70
0
0.5
1
2
4
(mM)
Edaravone
Survived motor neurons (%)
0
10
20
30
40
50
60
*
*
*
0
25
50
100
(μM)
Raloxifene
Survived MNs (%)
0
5
10
15
20
25
30
35
40
45
*
0
0.01
0.1
1
10
(μM)

Fig. 7

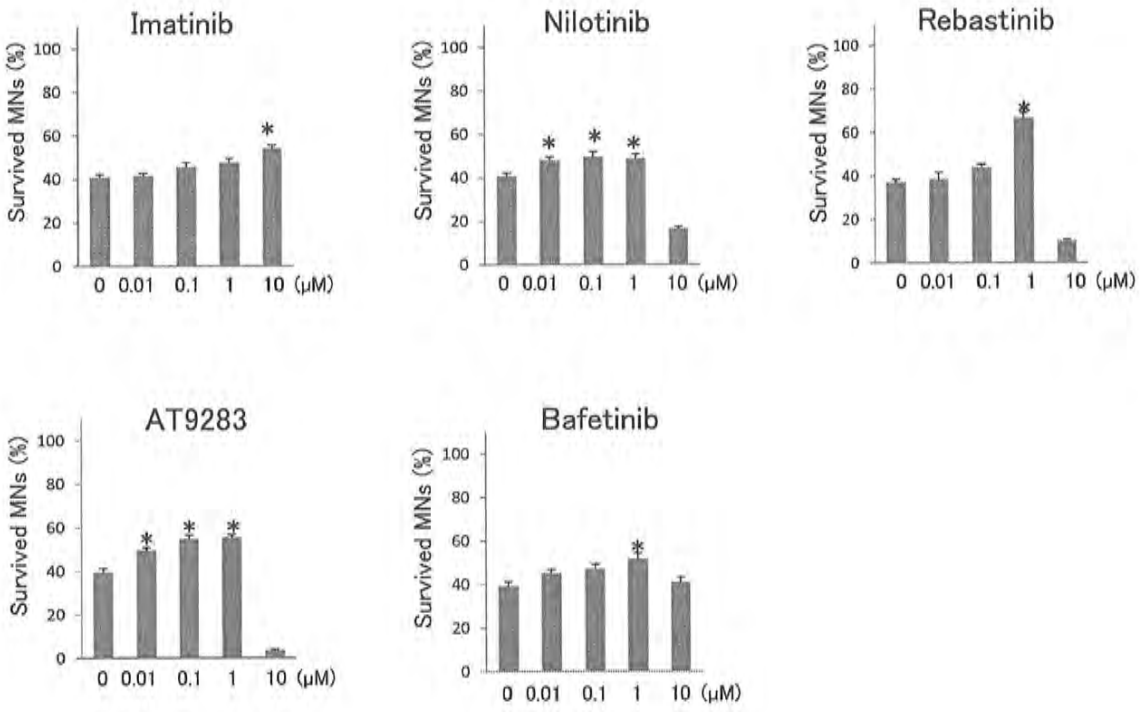
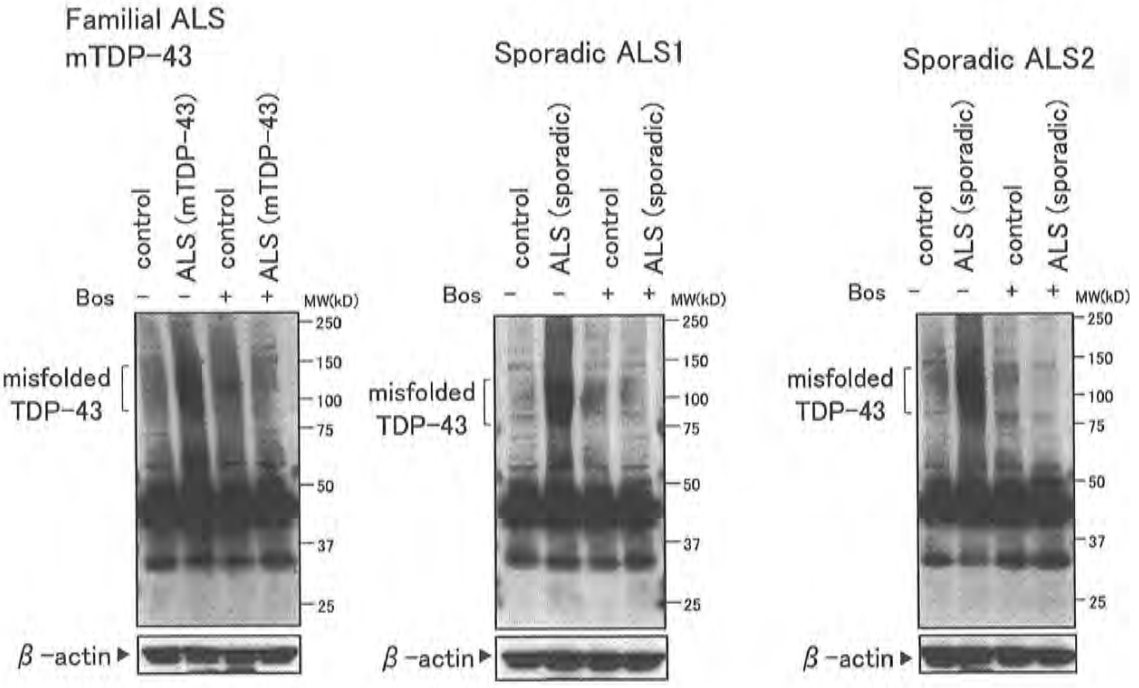

Imatinib
Nilotinib
Rebastinib
AT9283
Bafetinib
Survived MNs (%)
0 20 40 60 80 100
0 0.01 0.1 1 10 (μM)
*

Fig. 8


Familial ALS
mTDP-43
Sporadic ALS1
Sporadic ALS2
control
ALS (mTDP-43)
ALS (sporadic)
Bos
MW(kD)
250
150
100
75
50
37
25
misfolded TDP-43
β-actin