

FIG.1A



FIG.1B

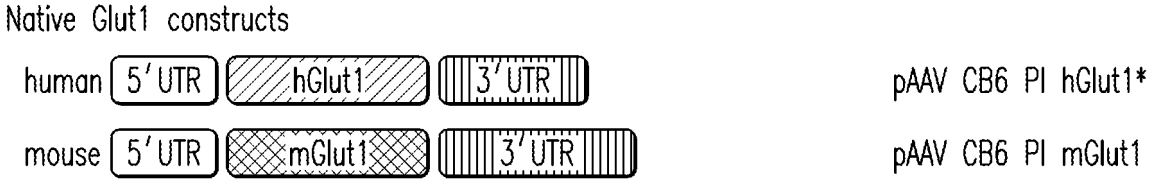


FIG.1C

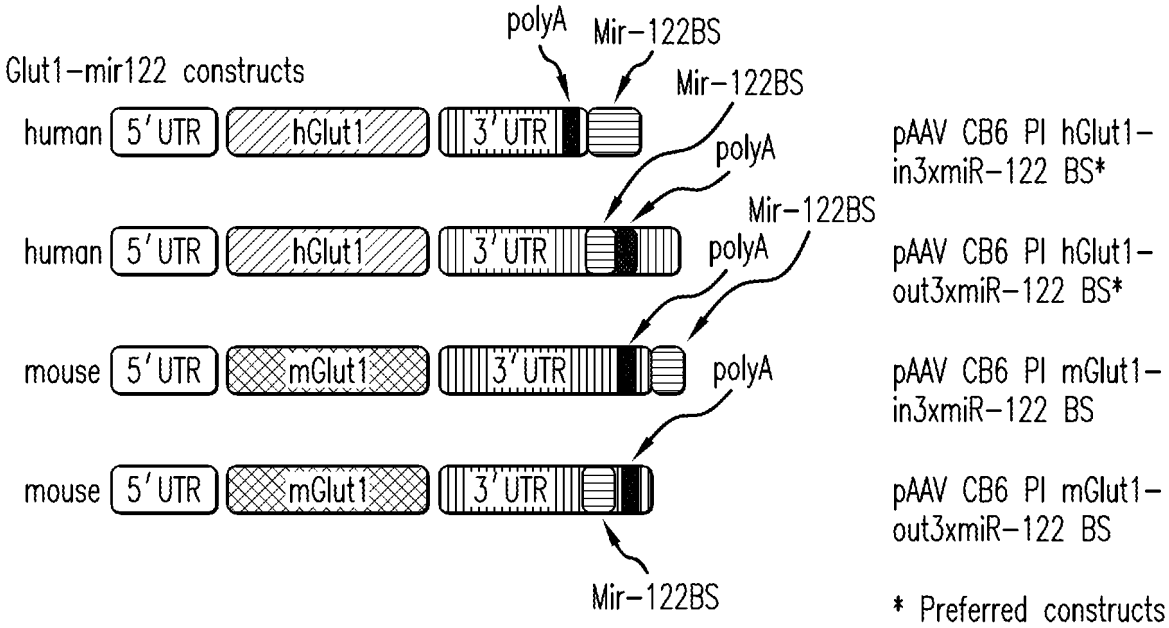


FIG.1D

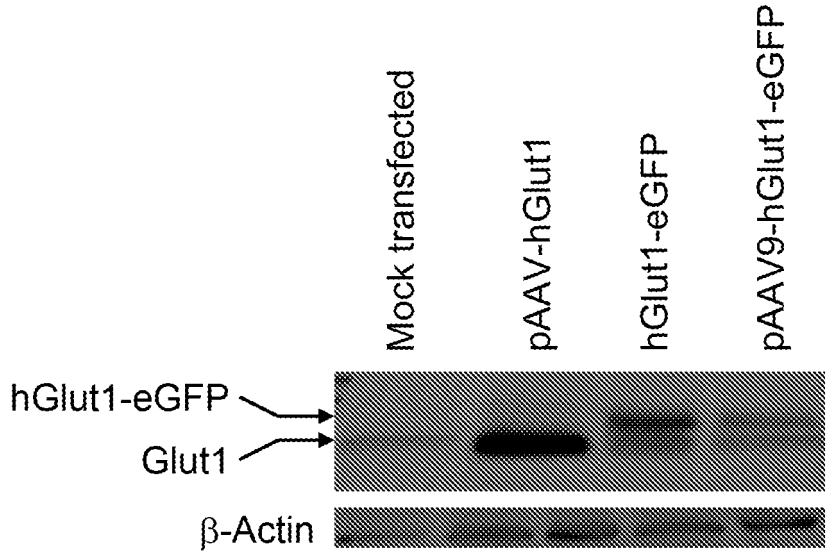


FIG.2A

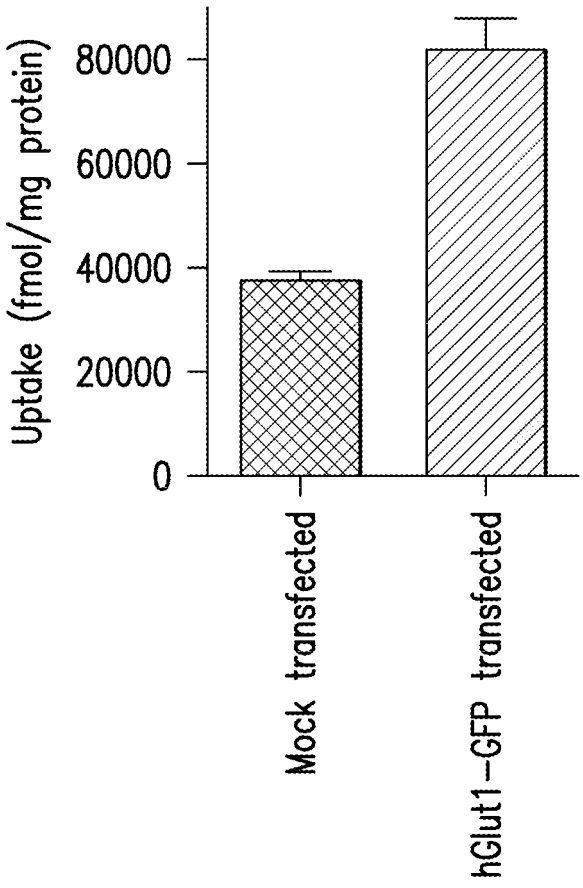


FIG.2B

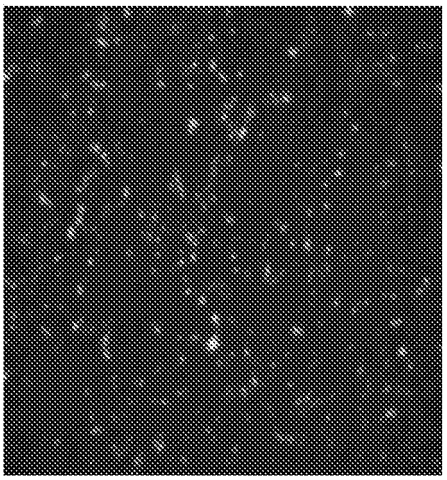


FIG.2C

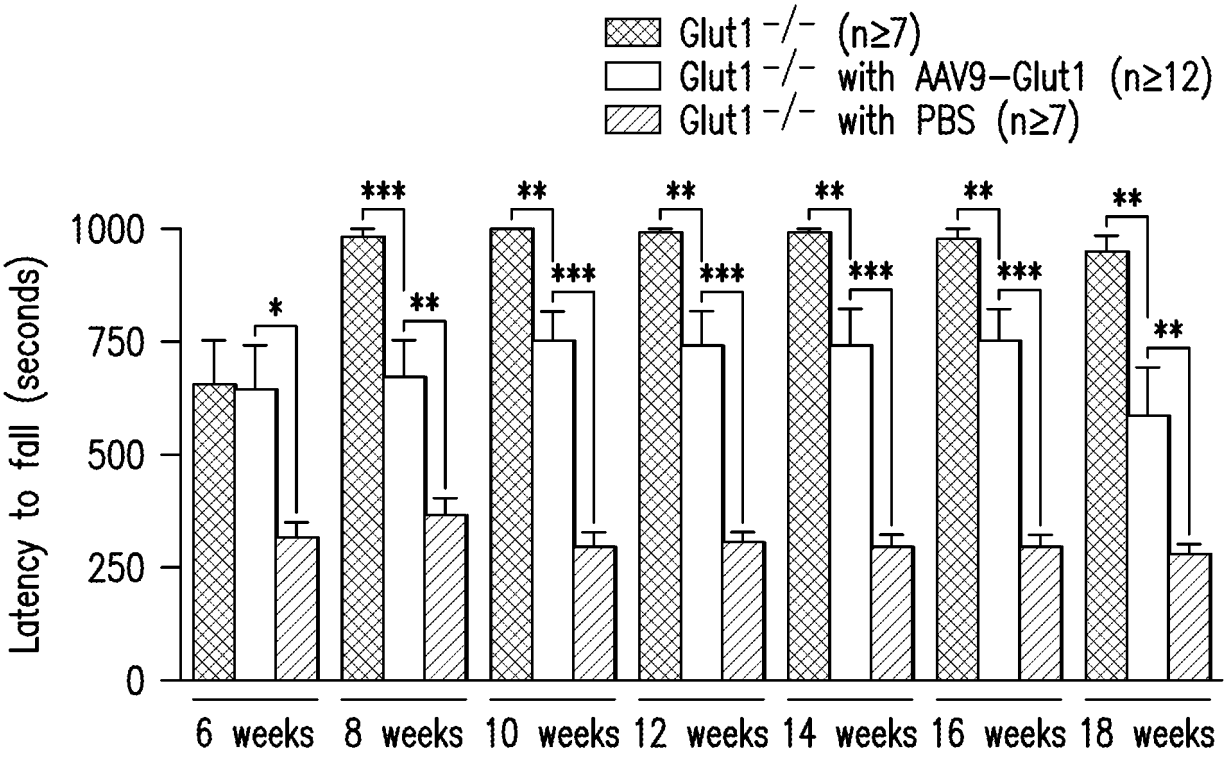

Glut1 −/− (n≥7)
Glut1 −/− with AAV9−Glut1 (n≥12)
Glut1 −/− with PBS (n≥7)
Latency to fall (seconds)
1000
750
500
250
0
*
***
**
6 weeks
8 weeks
10 weeks
12 weeks
14 weeks
16 weeks
18 weeks

FIG.3A

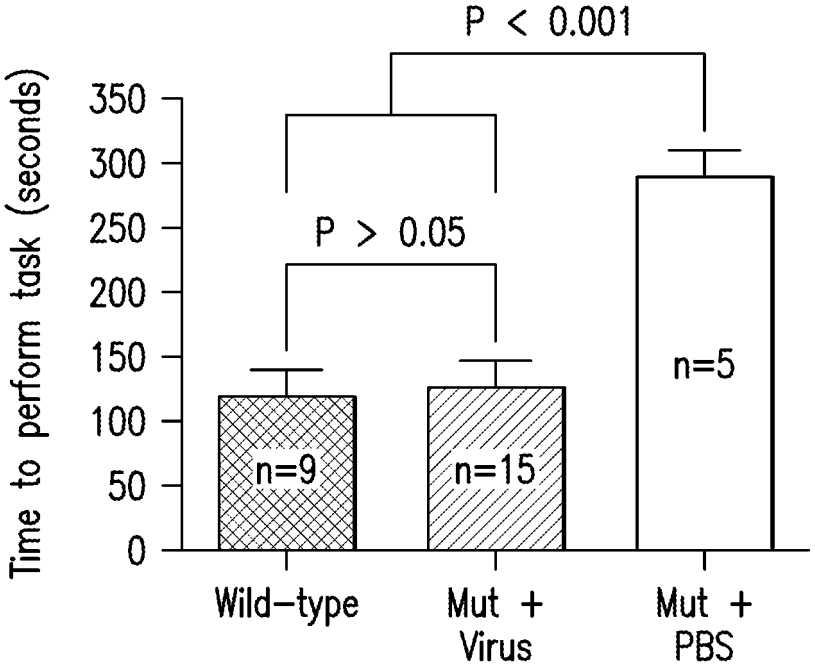

P < 0.001
P > 0.05
Time to perform task (seconds)
350
300
250
200
150
100
50
0
n=9
n=15
n=5
Wild−type
Mut + Virus
Mut + PBS

FIG.3B

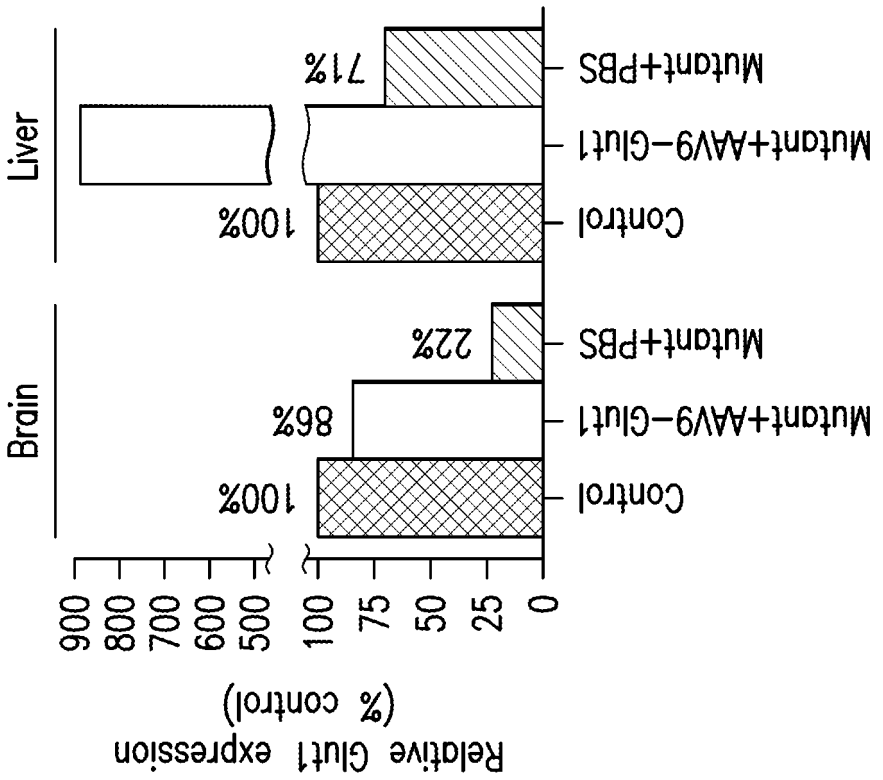

Relative Glut1 expression
(% control)
900
800
700
600
500
100
75
50
25
0
Brain
Liver
100%
86%
22%
100%
71%
Control
Mutant+AAV9–Glut1
Mutant+PBS
Control
Mutant+AAV9–Glut1
Mutant+PBS

FIG.4B

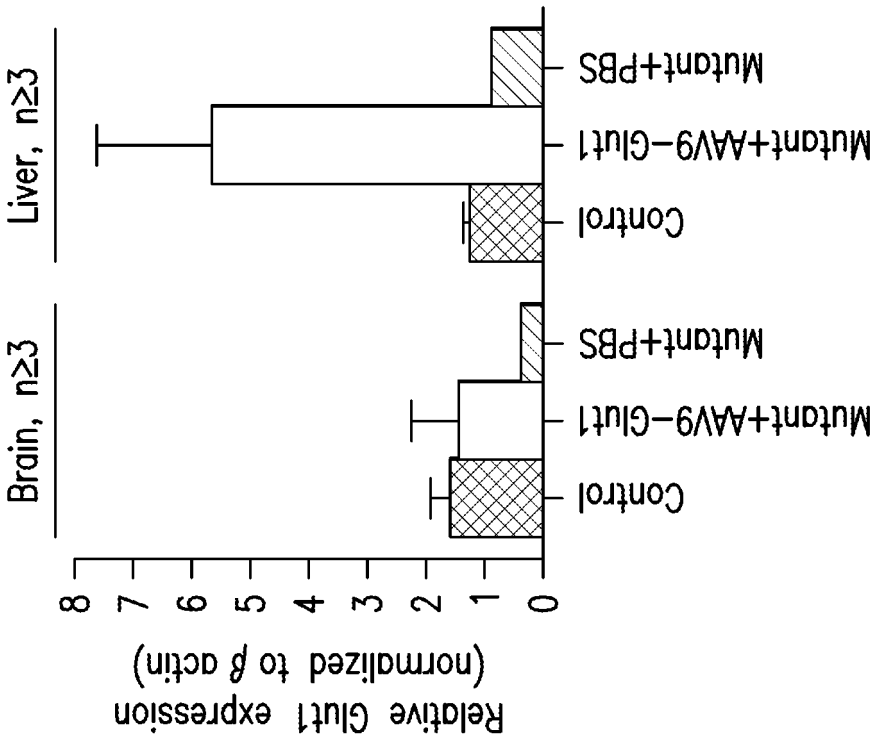

Relative Glut1 expression
(normalized to β actin)
8
7
6
5
4
3
2
1
0
Brain, n≥3
Liver, n≥3
Control
Mutant+AAV9–Glut1
Mutant+PBS
Control
Mutant+AAV9–Glut1
Mutant+PBS

FIG.4A

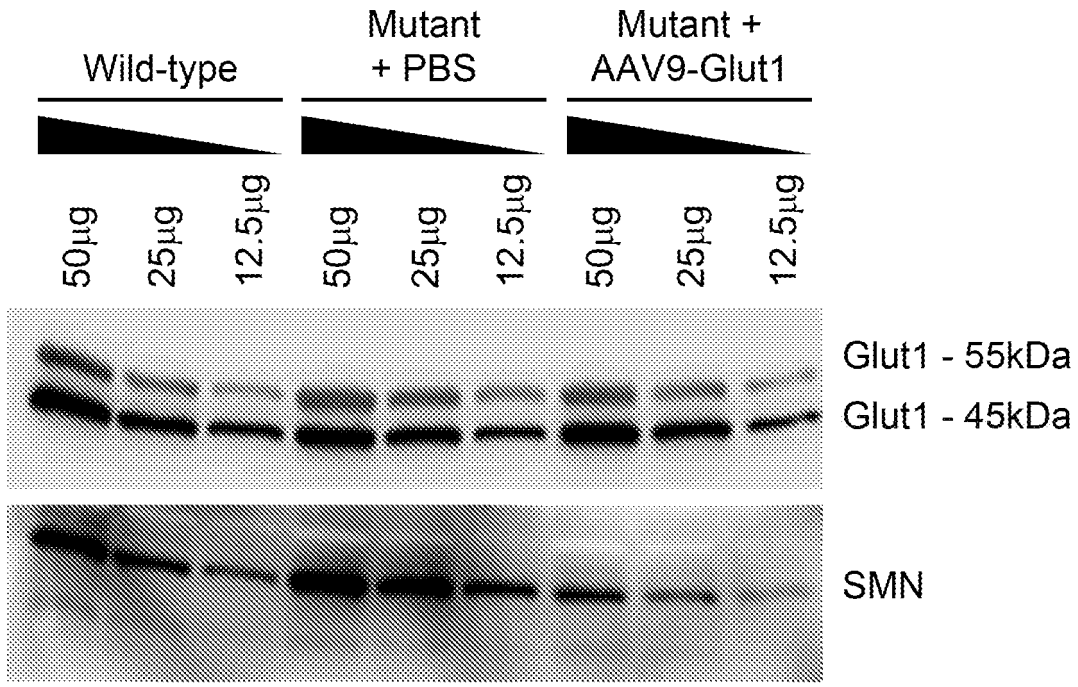

Wild-type
Mutant + PBS
Mutant + AAV9-Glut1
50µg
25µg
12.5µg
50µg
25µg
12.5µg
50µg
25µg
12.5µg
Glut1 - 55kDa
Glut1 - 45kDa
SMN

FIG.5A

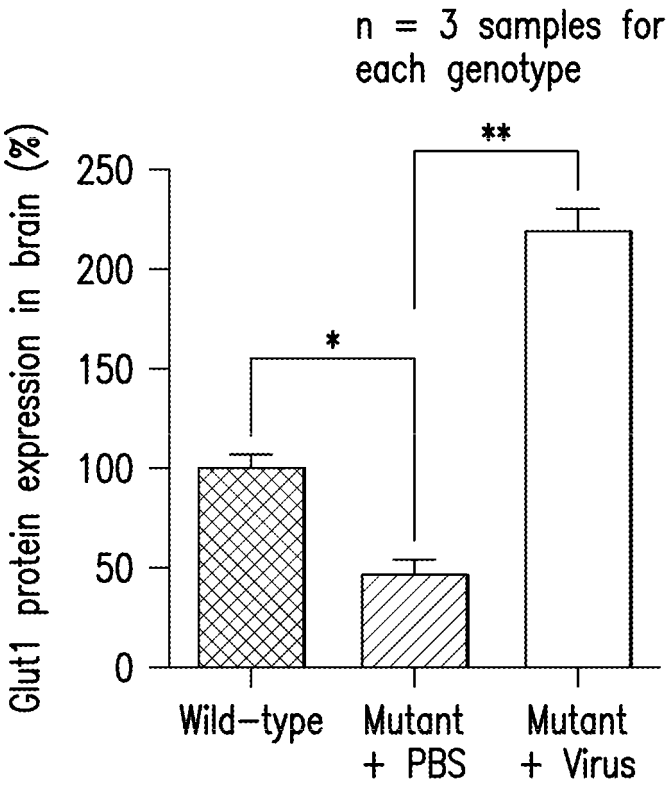

n = 3 samples for each genotype
Glut1 protein expression in brain (%)
250
200
150
100
50
0
*
**
Wild-type
Mutant + PBS
Mutant + Virus

FIG.5B

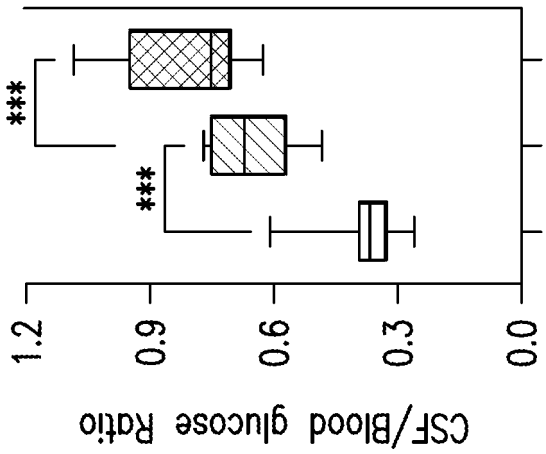

CSF/Blood glucose Ratio
1.2
0.9
0.6
0.3
0.0
***
***
Control
Mutant+AAV9-Glut1
Mutant+PBS

FIG.5D

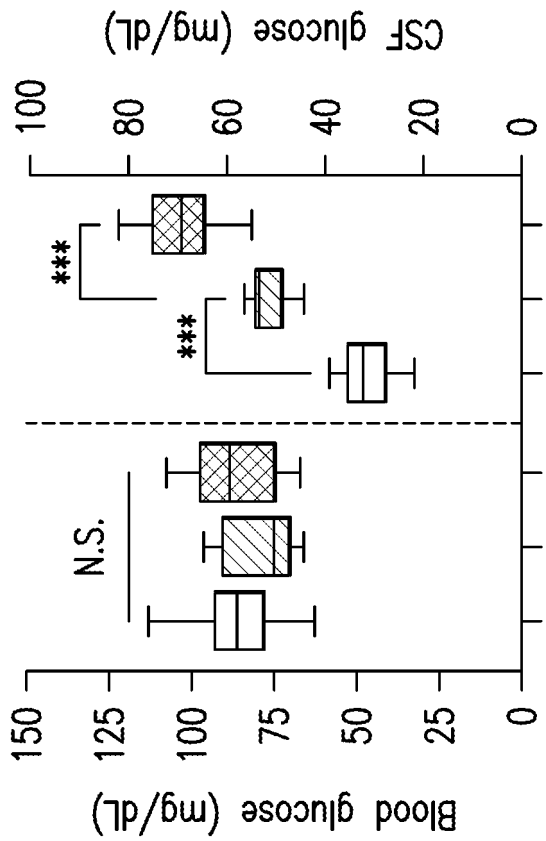

Blood glucose (mg/dL)
150
125
100
75
50
25
0
N.S.
CSF glucose (mg/dL)
100
80
60
40
20
0
***
***
Control
Mutant+AAV9-Glut1
Mutant+PBS
Control
Mutant+AAV9-Glut1
Mutant+PBS

FIG.5C