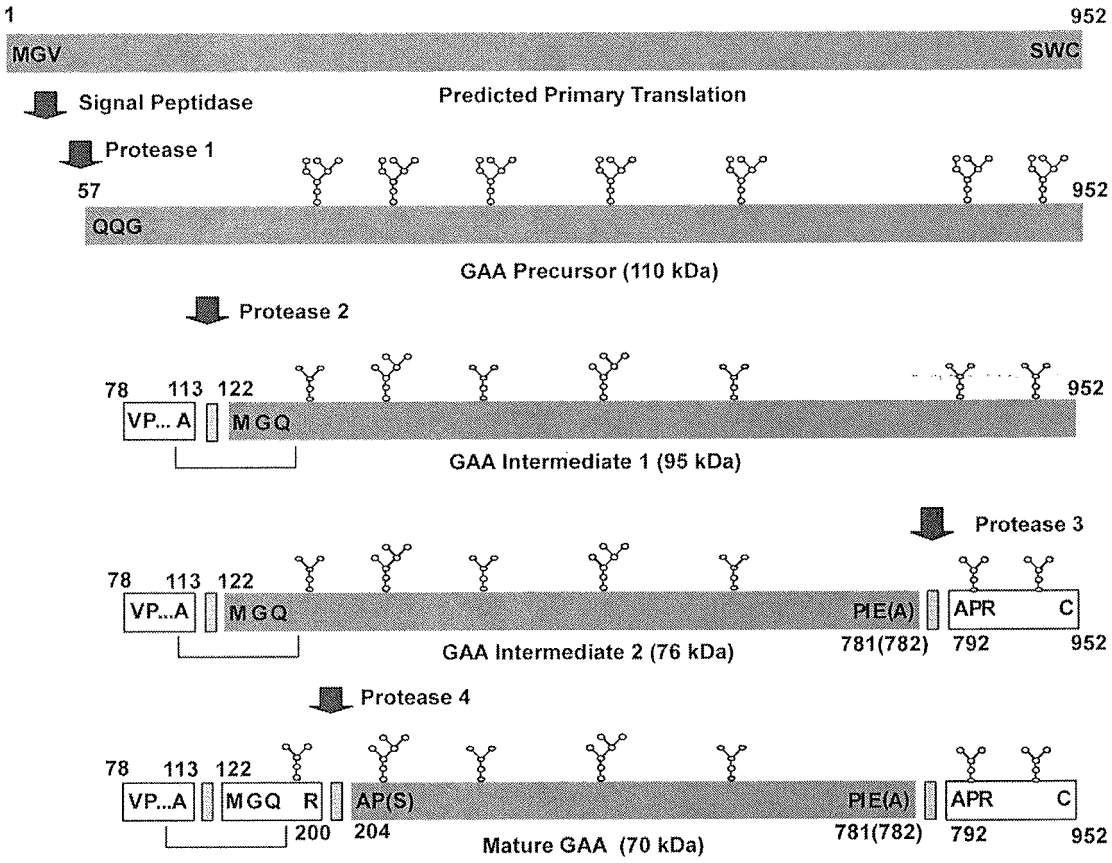

1
952
MGV
SWC
Signal Peptidase
Predicted Primary Translation
Protease 1
57
952
QQG
GAA Precursor (110 kDa)
Protease 2
78 113 122
952
VP... A
MGQ
GAA Intermediate 1 (95 kDa)
Protease 3
78 113 122
VP...A
MGQ
PIE(A)
APR
C
781(782)
792
952
GAA Intermediate 2 (76 kDa)
Protease 4
78 113 122
VP...A
MGQ R
AP(S)
PIE(A)
APR
C
200
204
781(782)
792
952
Mature GAA (70 kDa)

FIG. 1

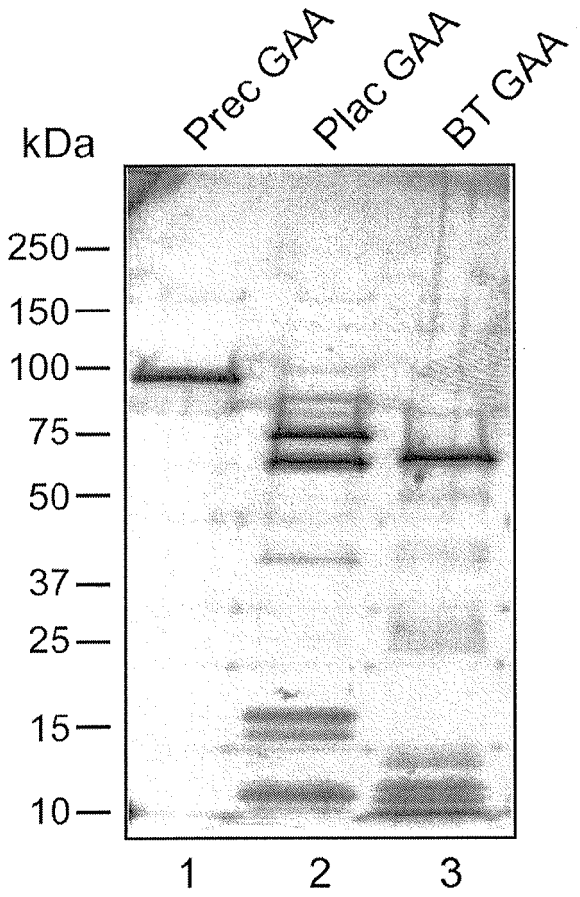


FIG. 2

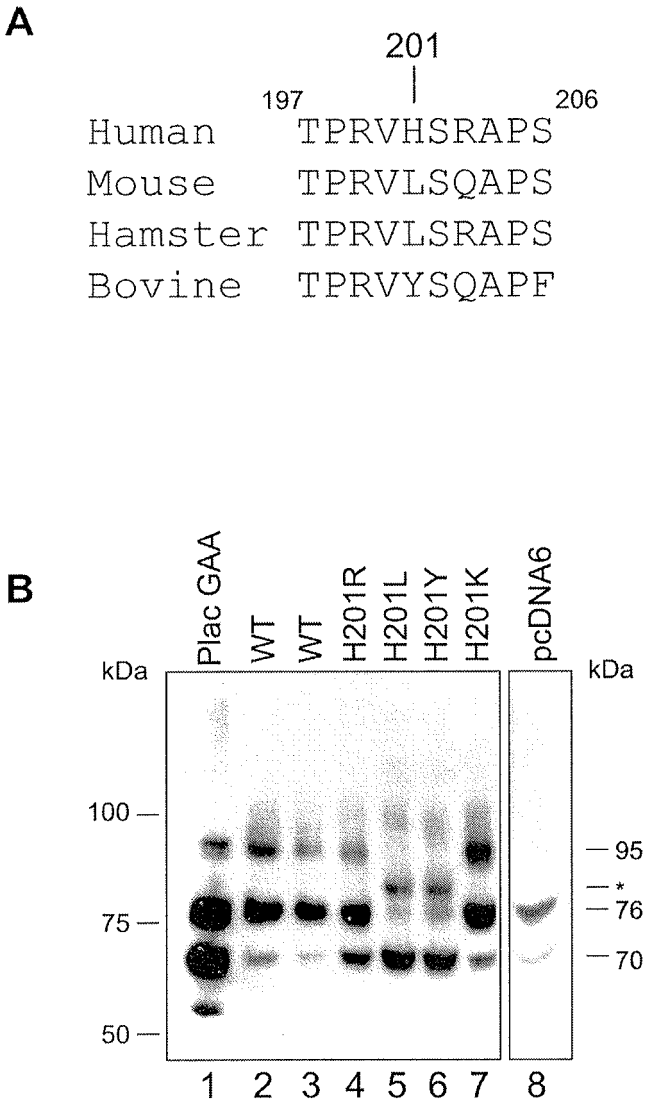

A
201
197
206
Human TPRVHSRAPS
Mouse TPRVLSQAPS
Hamster TPRVLSRAPS
Bovine TPRVYSQAPF
B
Plac GAA
WT
WT
H201R
H201L
H201Y
H201K
pcDNA6
kDa
kDa
100
75
50
95
*
76
70
1 2 3 4 5 6 7 8

FIG. 3

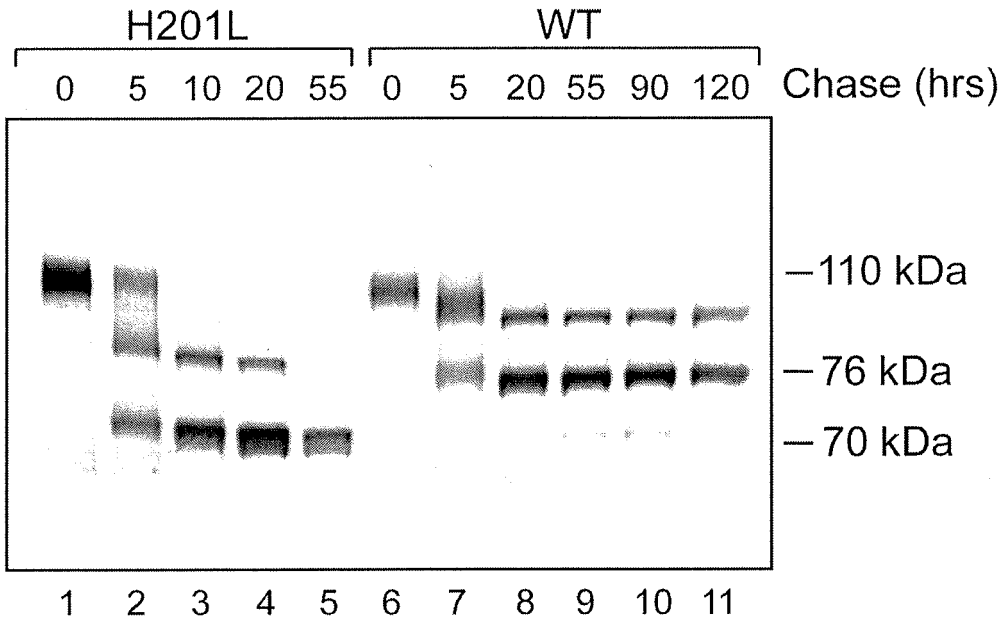

H201L
WT
0 5 10 20 55 0 5 20 55 90 120
Chase (hrs)
110 kDa
76 kDa
70 kDa
1 2 3 4 5 6 7 8 9 10 11

FIG. 4

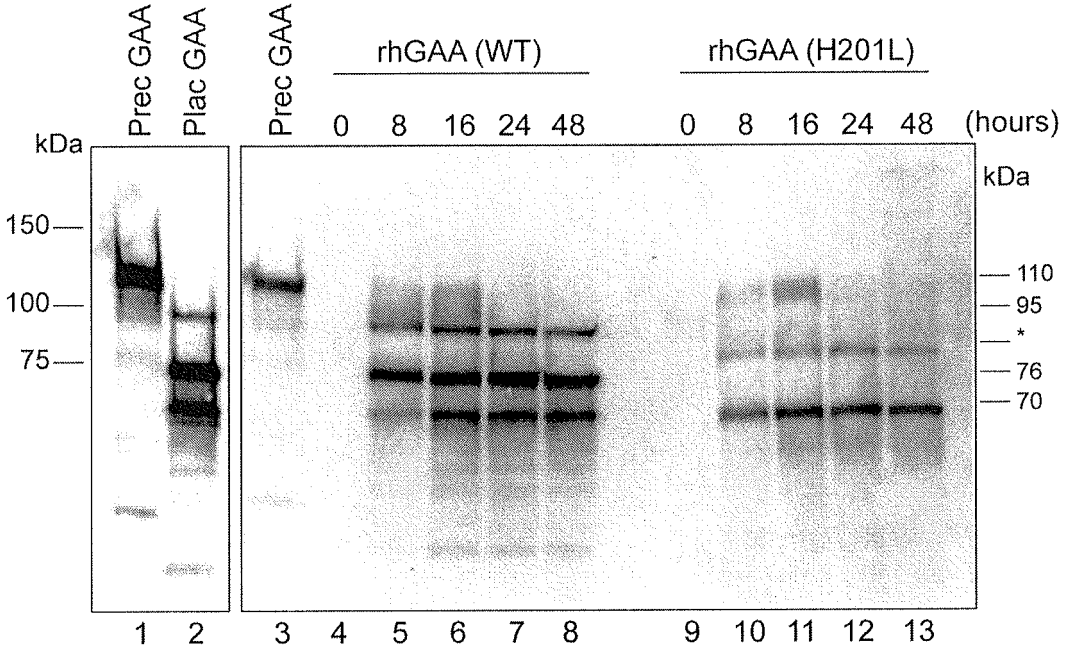

Prec GAA
Plac GAA
Prec GAA
rhGAA (WT)
rhGAA (H201L)
0 8 16 24 48
0 8 16 24 48 (hours)
kDa
150
100
75
kDa
110
95
*
76
70
1 2 3 4 5 6 7 8 9 10 11 12 13

FIG. 5

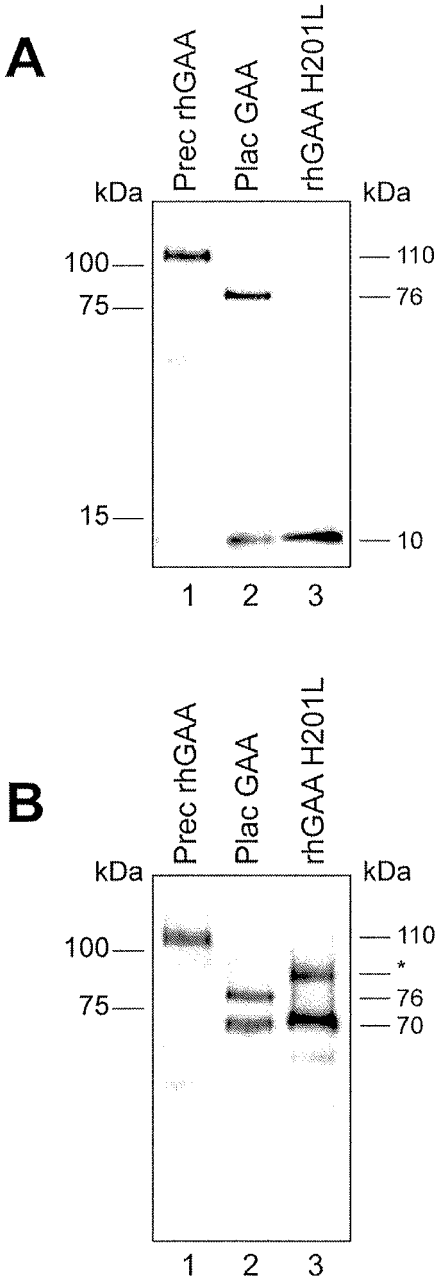

A
Prec rhGAA
Plac GAA
rhGAA H201L
kDa
kDa
100
75
15
110
76
10
1
2
3
B
Prec rhGAA
Plac GAA
rhGAA H201L
kDa
kDa
100
75
110
*
76
70
1
2
3

FIG. 6

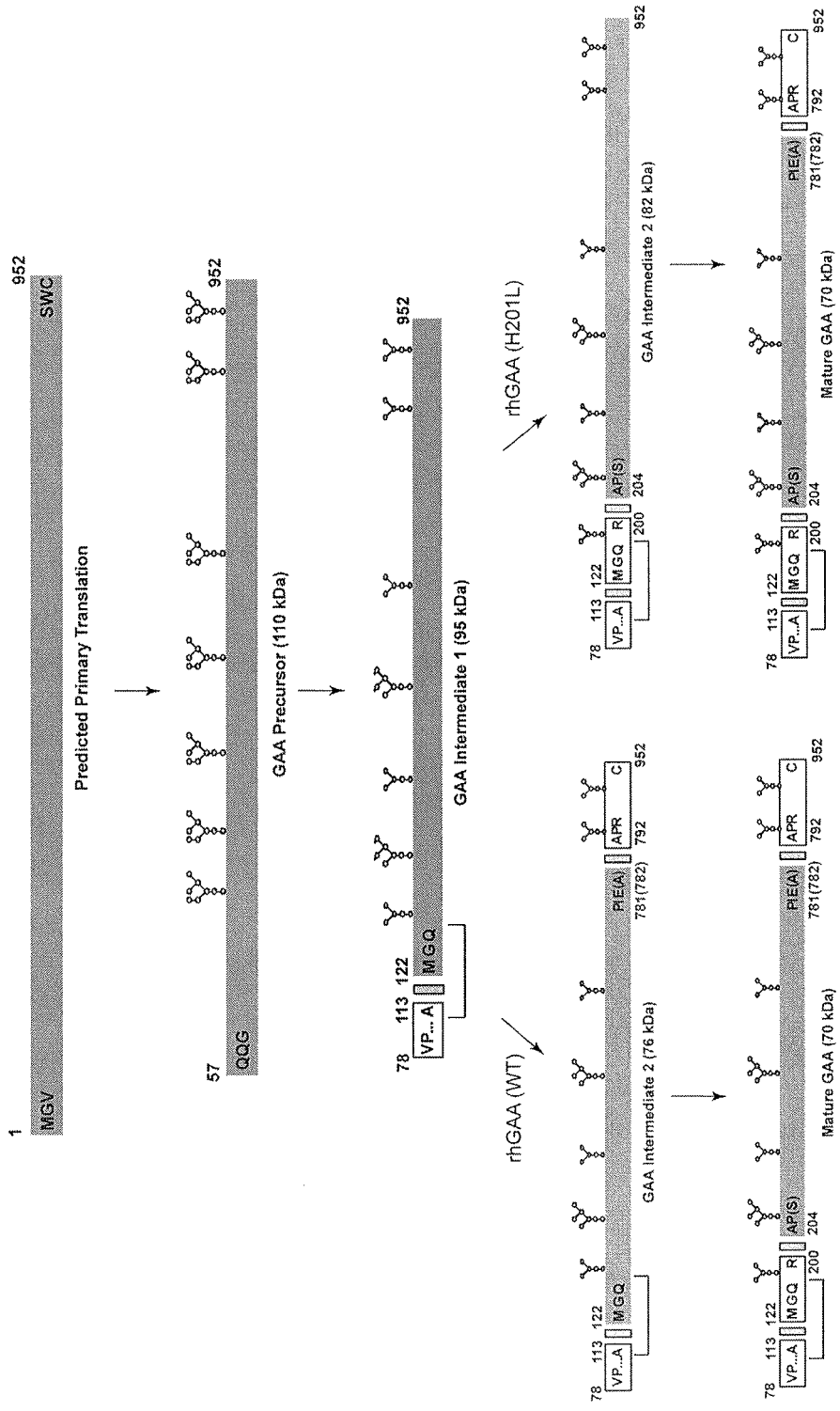

1
MGV
952
SWC
Predicted Primary Translation
57
QQG
952
GAA Precursor (110 kDa)
78
VP...A
113
122
MGQ
952
GAA Intermediate 1 (95 kDa)
rhGAA (H201L)
rhGAA (WT)
AP(S)
204
200
R
GAA Intermediate 2 (82 kDa)
PIE(A)
781(782)
APR
792
C
GAA Intermediate 2 (76 kDa)
Mature GAA (70 kDa)

FIG. 7