FIGURE 1

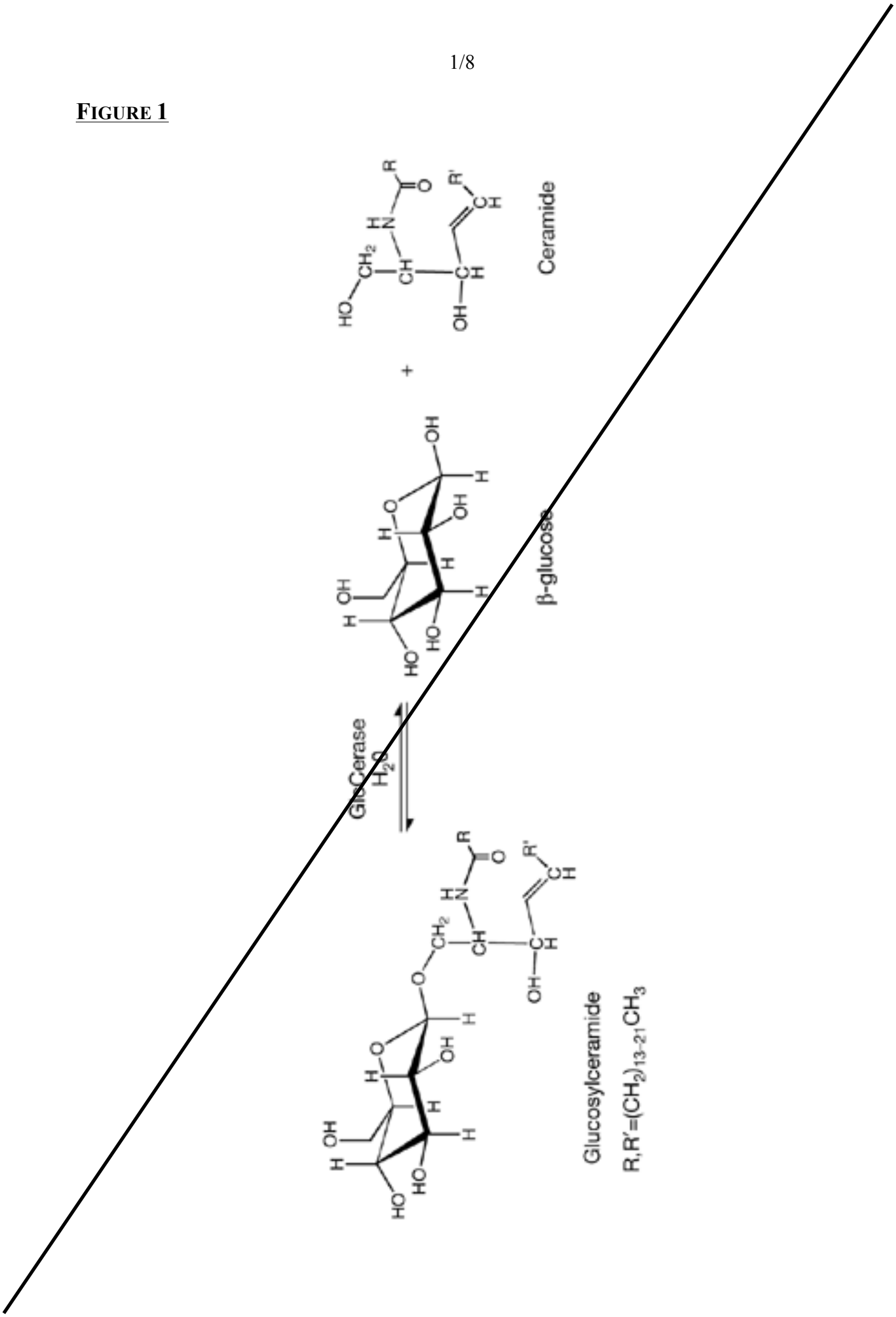

Ceramide
β-glucose
GlcCerase
H_2O
Glucosylceramide
$R,R'=(CH_2)_{13\text{-}21}CH_3$

FIGURE 2

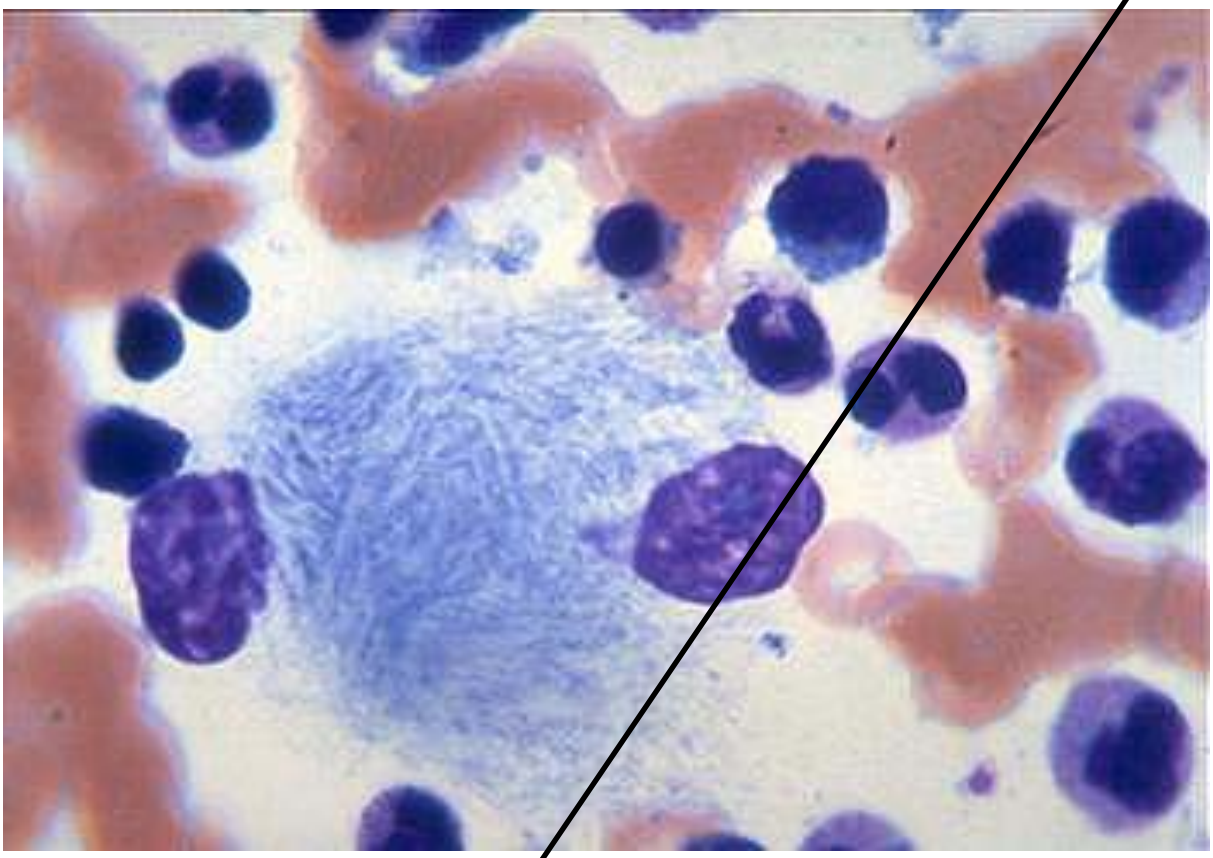


FIGURE 3



FIGURE 4

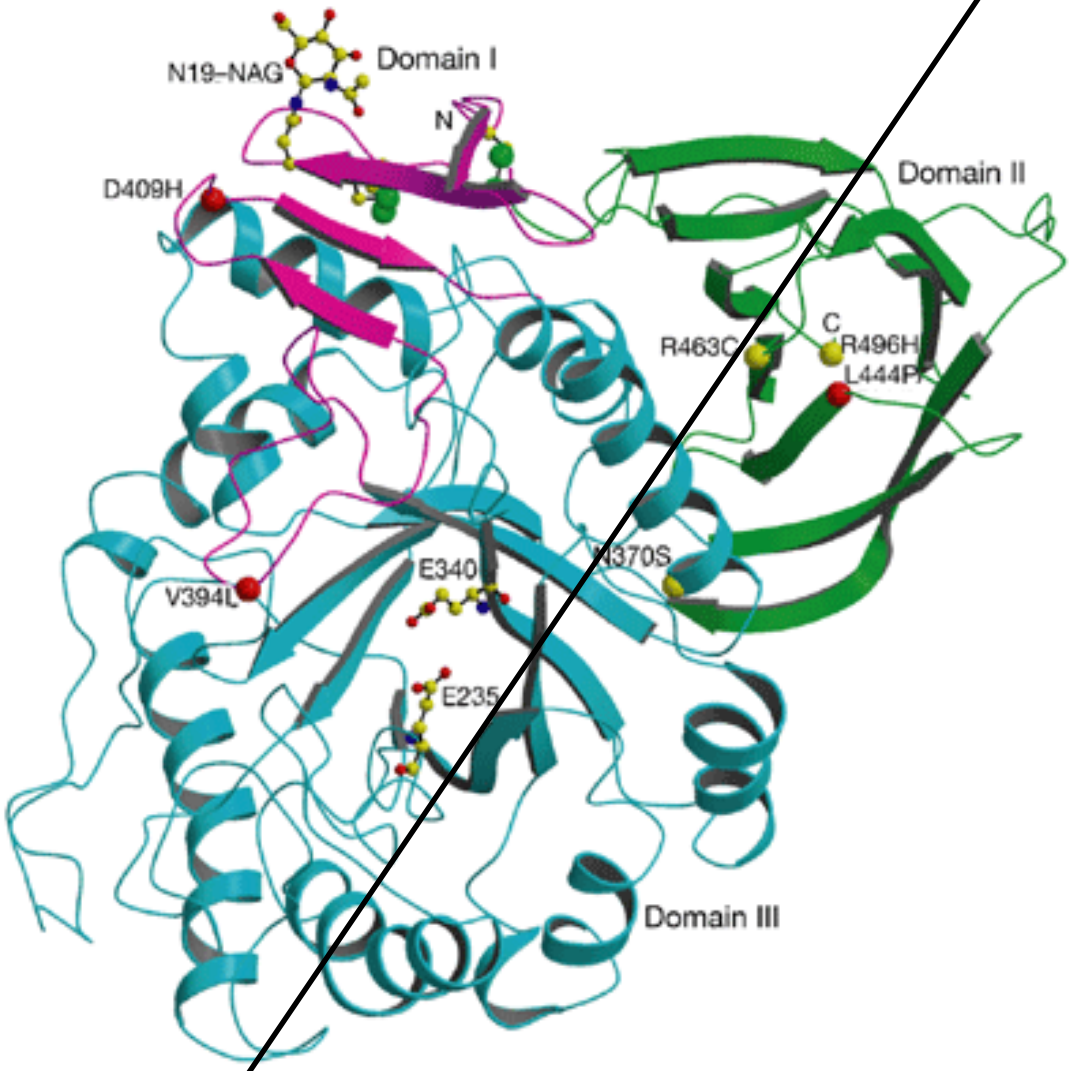
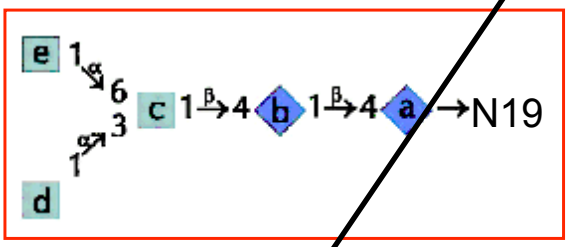

N19–NAG
Domain I
N
D409H
Domain II
R463C
C
R496H
L444P
V394L
E340
I370S
E235
Domain III

FIGURE 5

N-linked glycan on Cerezyme by Genzyme



N-linked glycan, if the glucocerebrosidase is produced in GnT I mutant CHO cells

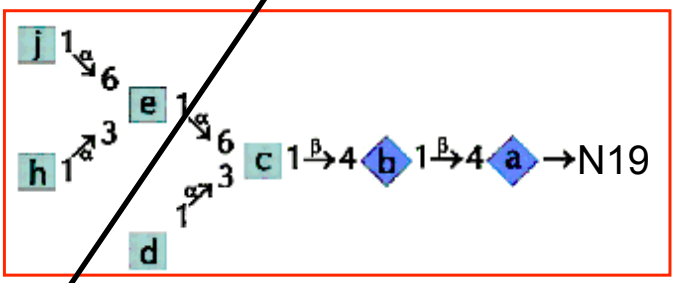


FIGURE 6

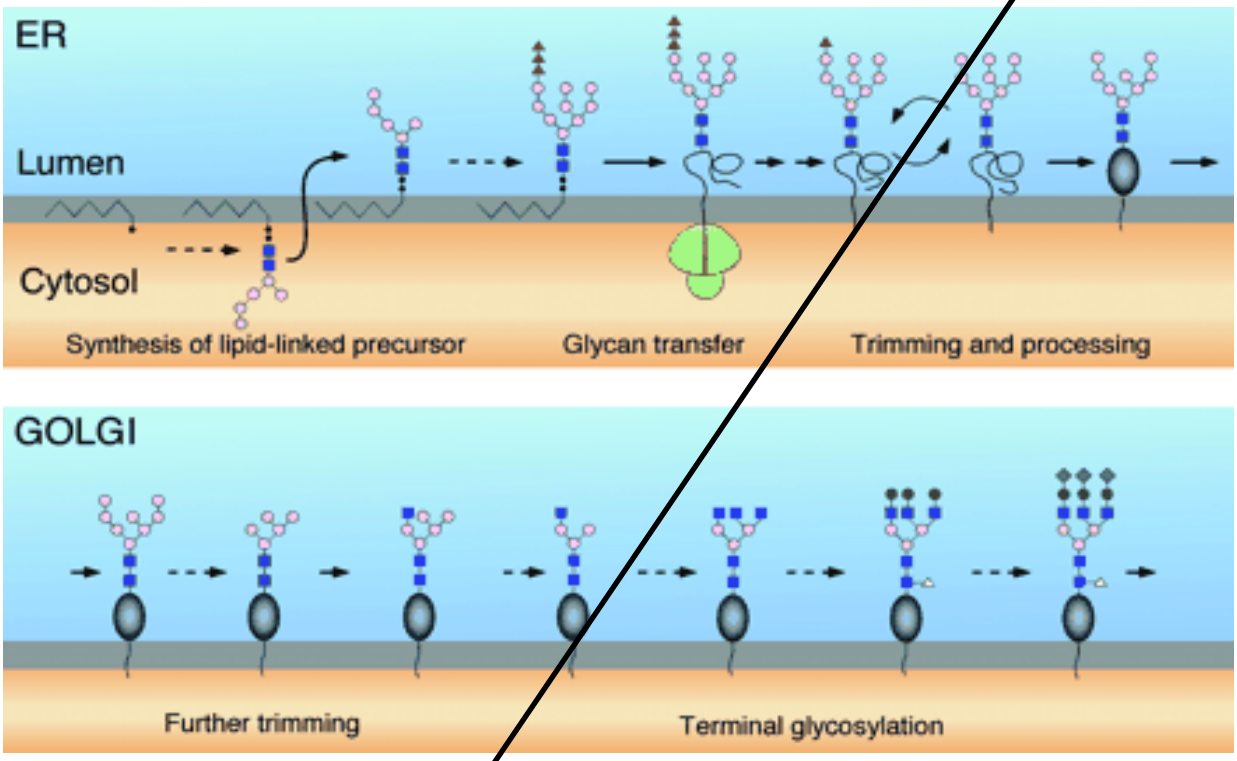

ER
Lumen
Cytosol
Synthesis of lipid-linked precursor
Glycan transfer
Trimming and processing
GOLGI
Further trimming
Terminal glycosylation

FIGURE 7

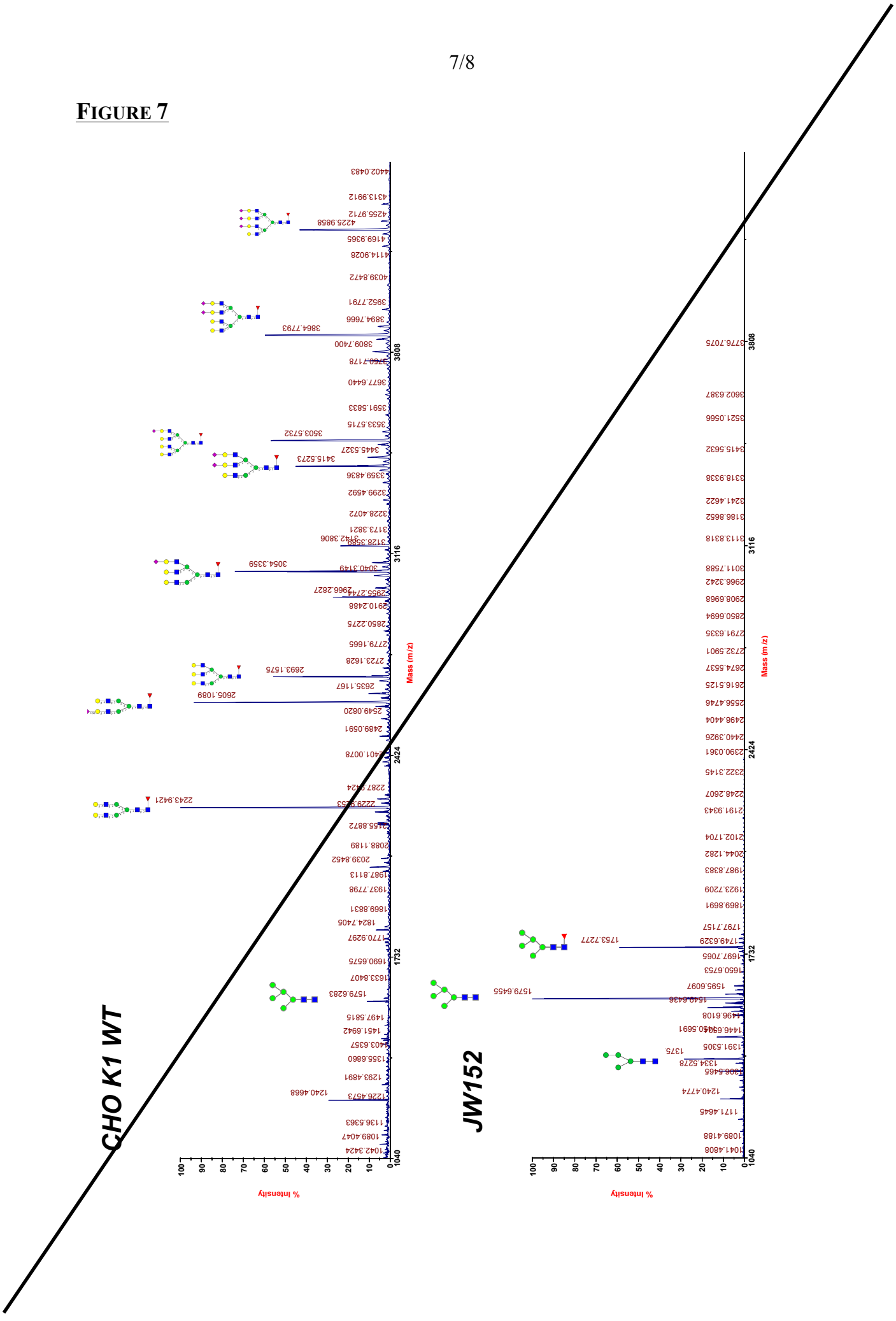


FIGURE 8

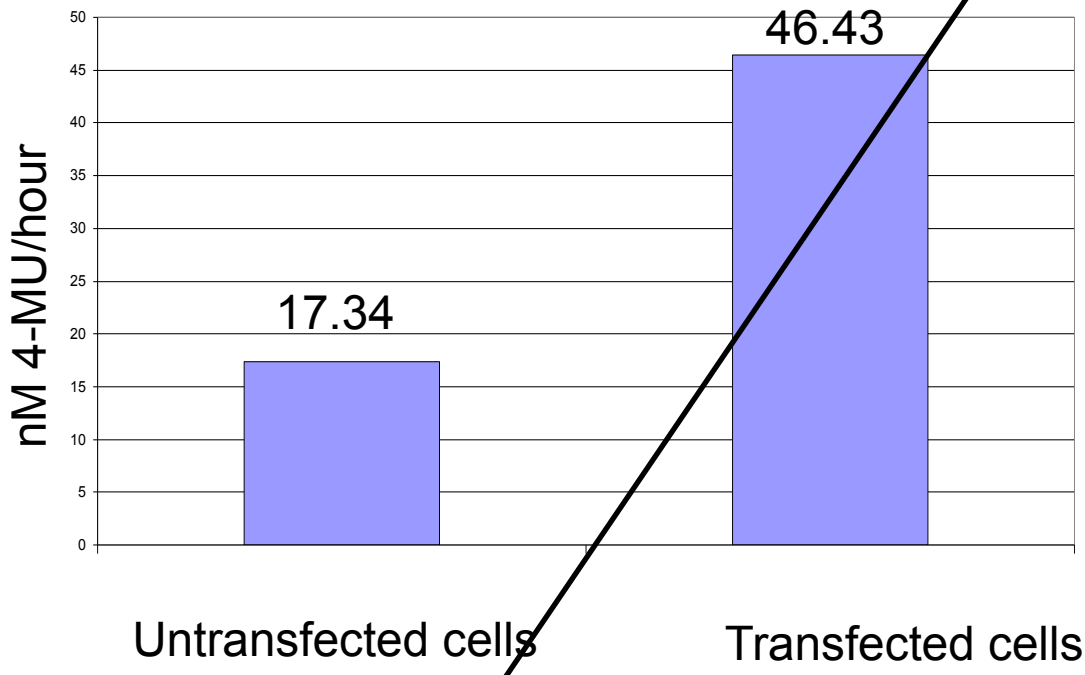

50
45
40
35
30
25
20
15
10
5
0
nM 4-MU/hour
17.34
46.43
Untransfected cells
Transfected cells

FIGURE 1

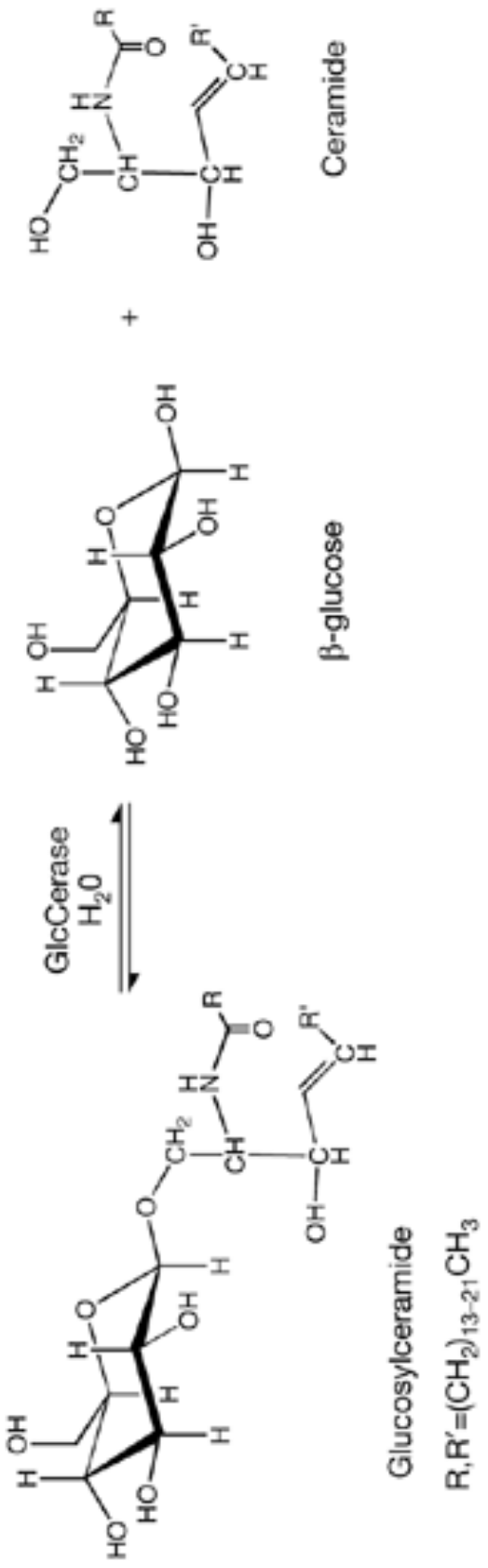

Ceramide
β-glucose
GlcCerase
H_2O
Glucosylceramide
$R,R'=(CH_2)_{13\text{-}21}CH_3$

FIGURE 2

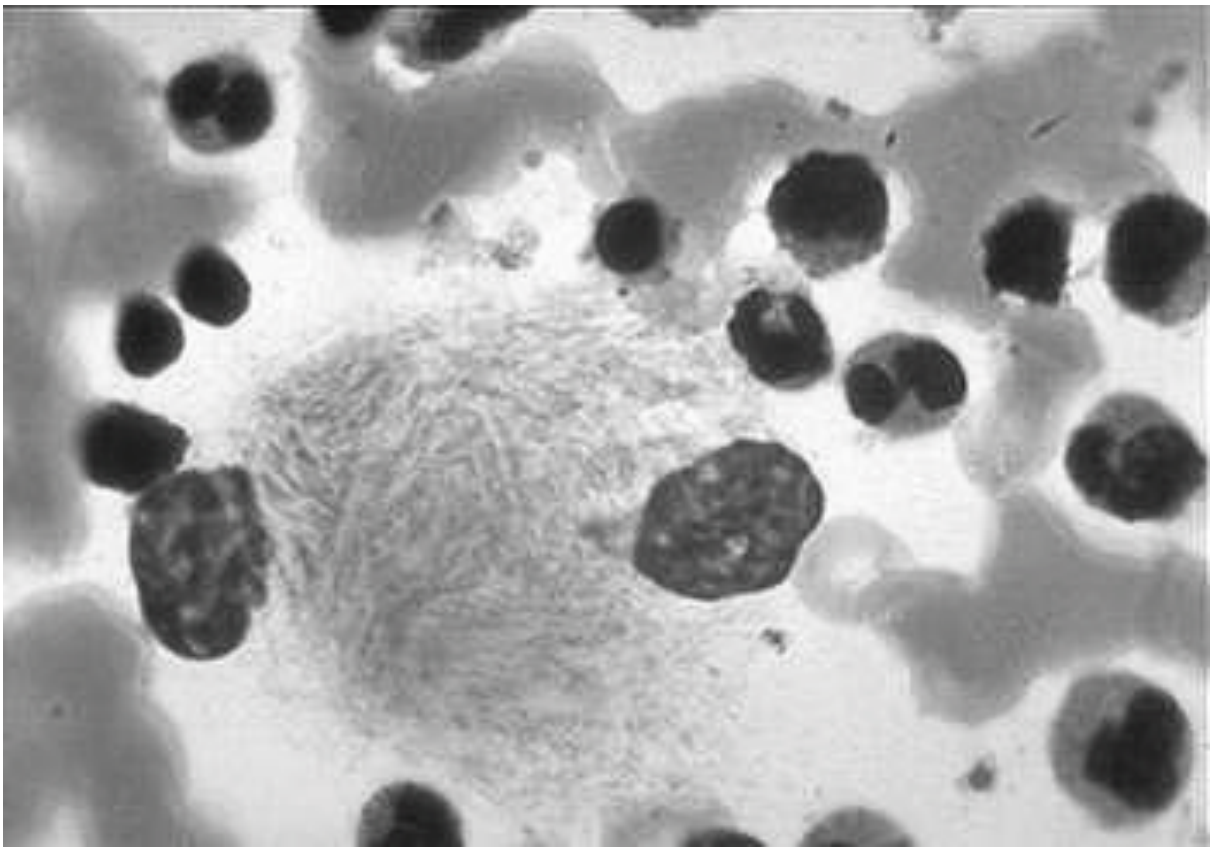


FIGURE 3



FIGURE 4

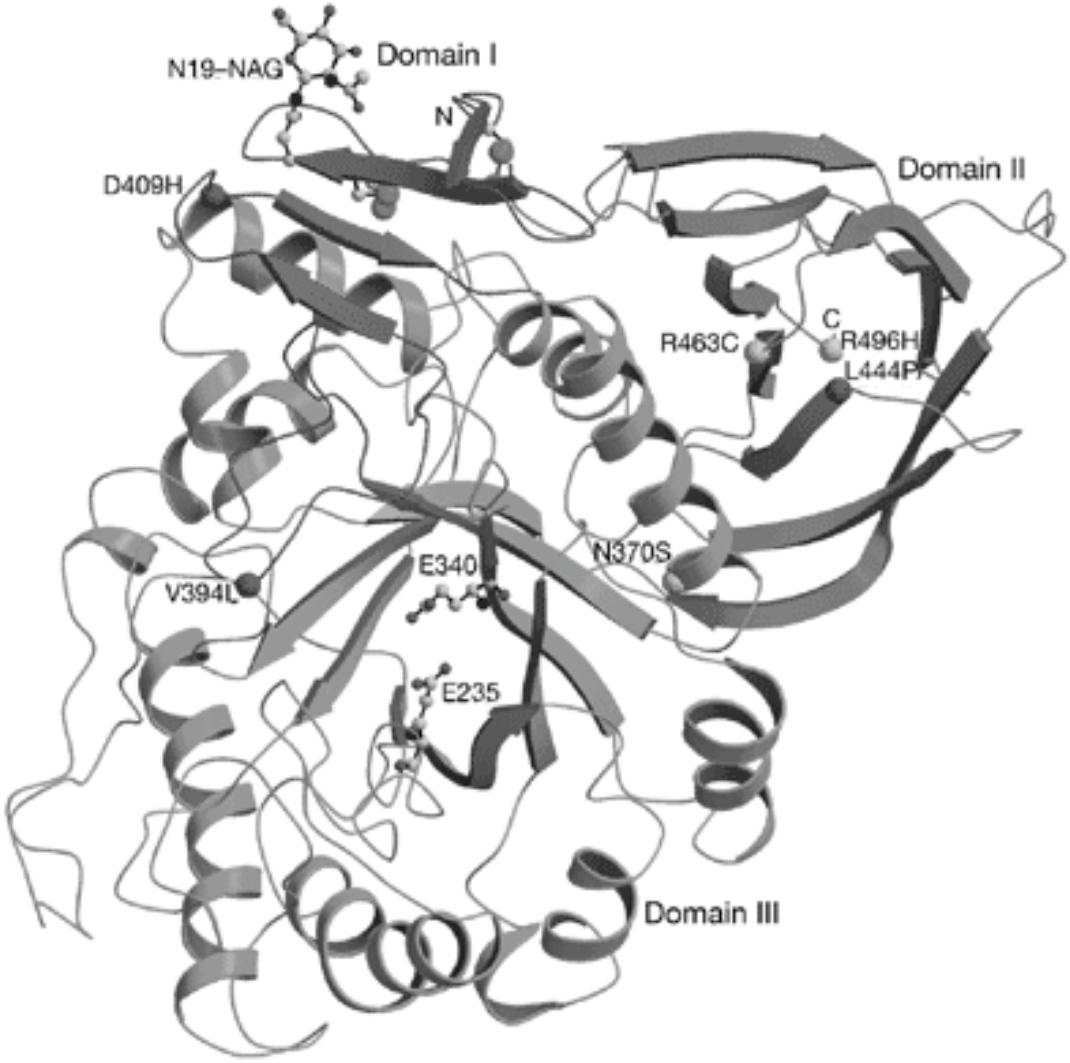

N19–NAG
Domain I
N
D409H
Domain II
C
R463C
R496H
L444P
N370S
E340
V394L
E235
Domain III

FIGURE 5

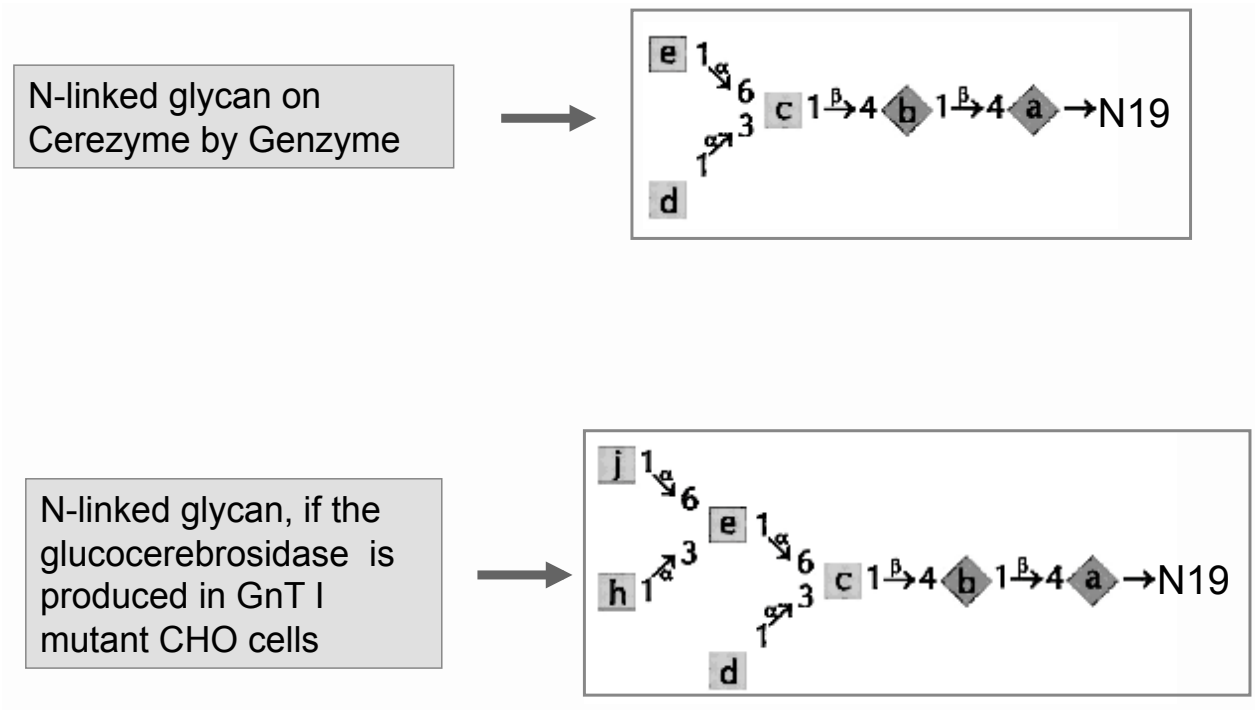

N-linked glycan on Cerezyme by Genzyme
e
1
α
6
3
c
1β→4
b
1β→4
a
→N19
d
N-linked glycan, if the glucocerebrosidase is produced in GnT I mutant CHO cells
j
1
α
6
e
h
1
α
3
c
1β→4
b
1β→4
a
→N19
d

FIGURE 6

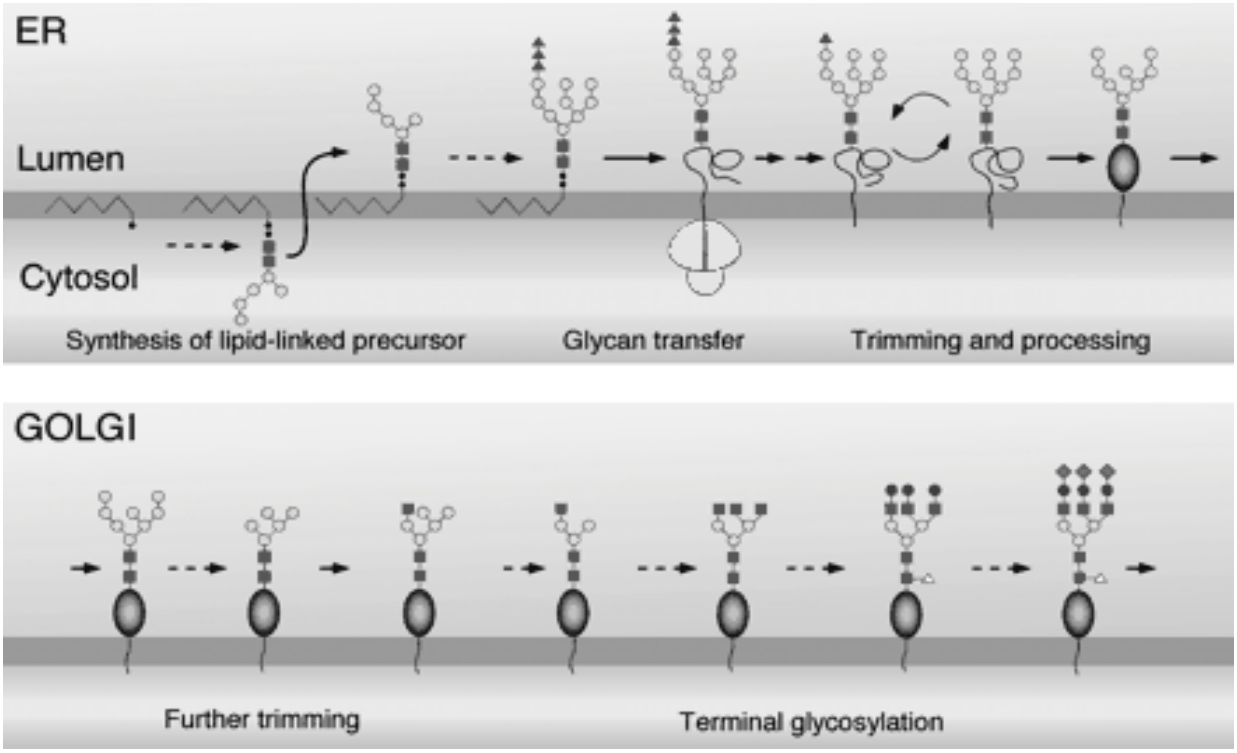

ER
Lumen
Cytosol
Synthesis of lipid-linked precursor
Glycan transfer
Trimming and processing
GOLGI
Further trimming
Terminal glycosylation

FIGURE 7

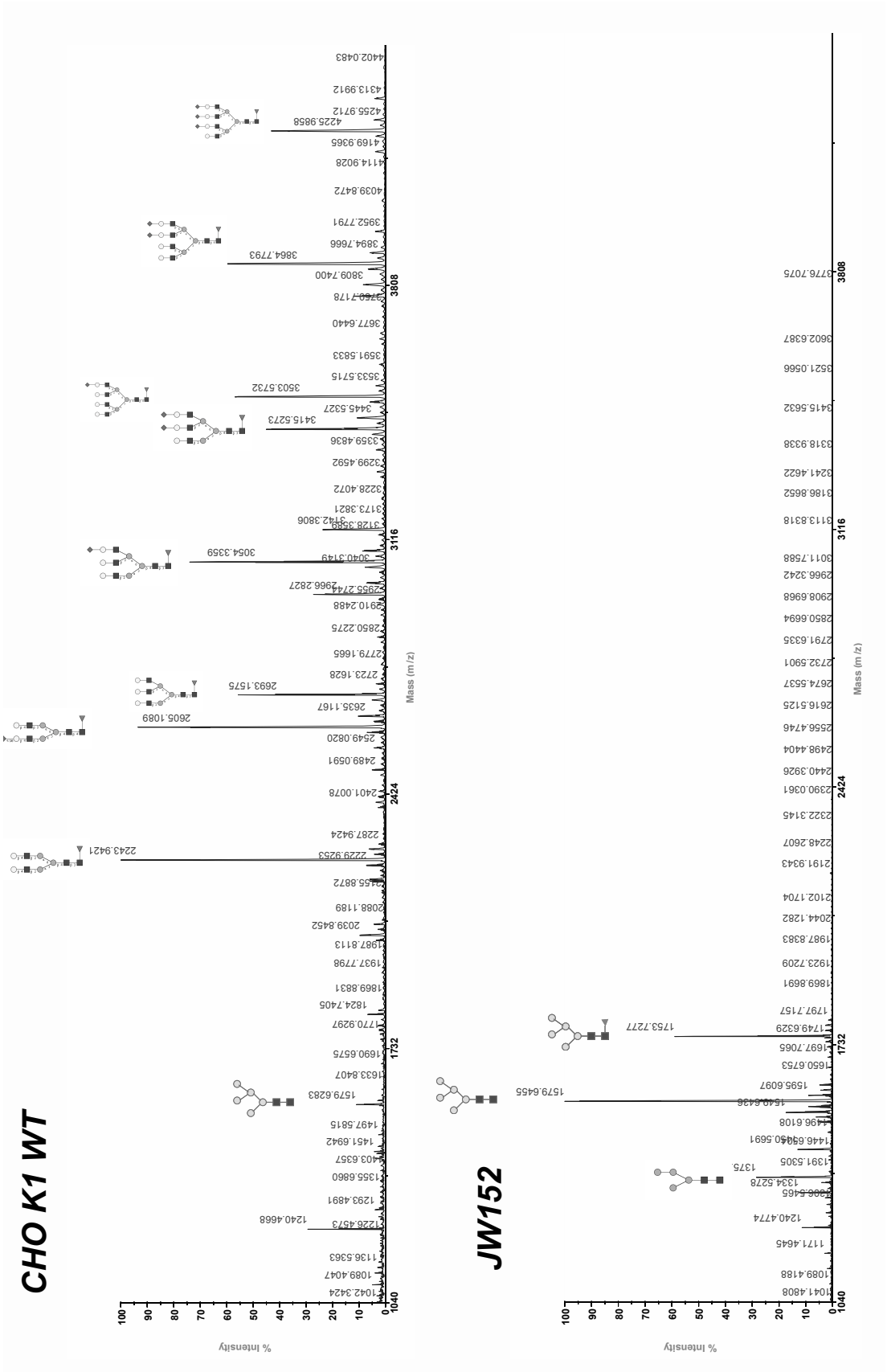
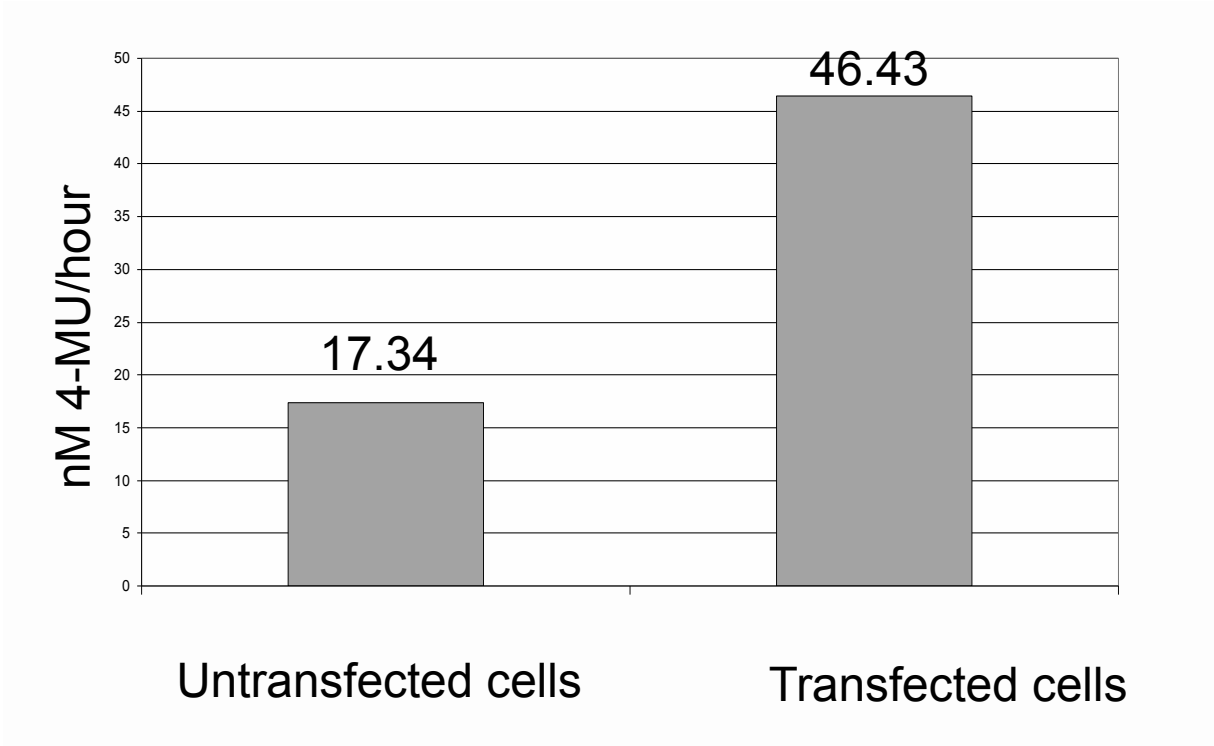

CHO K1 WT
JW152
% Intensity
Mass (m/z)
1040
1732
2424
3116
3808
1579.6283
1753.7277
2243.9421
2605.1089
3054.3359
3384.7793

FIGURE 8


nM 4-MU/hour
50
45
40
35
30
25
20
15
10
5
0
17.34
46.43
Untransfected cells
Transfected cells