SEQ ID NO: 1

1	Ser	Glu	Thr	Gln	Ala	Asn	Ser	Thr	Thr	Asp	Ala	Leu	Asn	Val	Leu	Leu	Ile	Ile	Val	Asp
21	Asp	Leu	Arg	Pro	Ser	Leu	Gly	Cys	Tyr	Gly	Asp	Lys	Leu	Val	Arg	Ser	Pro	Asn	Ile	Asp
41	Gln	Leu	Ala	Ser	His	Ser	Leu	Leu	Phe	Gln	Asn	Ala	Phe	Ala	Gln	Gln	Ala	Val	Cys	Ala
61	Pro	Ser	Arg	Bal	Ser	Phe	Leu	Thr	Gly	Arg	Arg	Pro	Asp	Thr	Thr	Arg	Leu	Tyr	Asp	Phe
81	Asn	Ser	Tyr	Trp	Arg	Val	His	Ala	Gly	Asn	Phe	Ser	Thr	Ile	Pro	Gln	Tyr	Phe	Lys	Glu
101	Asn	Gly	Tyr	Val	Thr	Met	Ser	Val	Gly	Lys	Val	Phe	His	Pro	Gly	Ile	Ser	Ser	Asn	His
121	Thr	Asp	Asp	Ser	Pro	Tyr	Ser	Trp	Ser	Phe	Pro	Pro	Tyr	His	Pro	Ser	Ser	Glu	Lys	Tyr
141	Glu	Asn	Thr	Lys	Thr	Cys	Arg	Gly	Pro	Asp	Gly	Glu	Leu	His	Ala	Asn	Leu	Leu	Cys	Pro
161	Val	Asp	Val	Leu	Asp	Val	Pro	Glu	Gly	Thr	Leu	Pro	Asp	Lys	Gln	Ser	Thr	Glu	Gln	Ala
181	Ile	Gln	Leu	Leu	Glu	Lys	Met	Lys	Thr	Ser	Ala	Ser	Pro	Phe	Phe	Leu	Ala	Val	Gly	Tyr
201	His	Lys	Pro	His	Ile	Pro	Phe	Arg	Tyr	Pro	Lys	Glu	Phe	Gln	Lys	Leu	Tyr	Pro	Leu	Glu
221	Asn	Ile	Thr	Leu	Ala	Pro	Asp	Pro	Glu	Val	Pro	Asp	Gly	Leu	Pro	Pro	Val	Ala	Tyr	Asn
241	Pro	Trp	Met	Asp	Ile	Arg	Gln	Arg	Glu	Asp	Val	Gln	Ala	Leu	Asn	Ile	Ser	Val	Pro	Tyr
261	Gly	Pro	Ile	Pro	Val	Asp	Phe	Gln	Arg	Lys	Ile	Arg	Gln	Ser	Tyr	Phe	Ala	Ser	Val	Ser

FIG. 1A

281 Tyr Leu Asp Thr Gln Val Gly Arg Leu Leu Ser Ala Leu Asp Asp Leu Gln Leu Ala Asn

301 Ser Thr Ile Ile Ala Phe Thr Ser Asp His Gly Trp Ala Leu Gly Glu His Gly Glu Trp

321 Ala Lys Tyr Ser Asn Phe Asp Val Ala Thr His Val Pro Leu Ile Phe Tyr Val Pro Gly

341 Arg Thr Ala Ser Leu Pro Glu Ala Gly Glu Lys Leu Phe Pro Tyr Leu Asp Pro Phe Asp

361 Ser Ala Ser Gln Leu Met Glu Pro Gly Arg Gln Ser Met Asp Leu Val Glu Leu Val Ser

381 Leu Phe Pro Thr Leu Ala Gly Leu Ala Gly Leu Gln Val Pro Pro Arg Cys Pro Val Pro

401 Ser Phe His Val Glu Leu Cys Arg Clu Gly Lys Asn Leu Leu Lys His Phe Arg Phe Arg

421 Asp Leu Glu Glu Asp Pro Tyr Leu Pro Gly Asn Pro Arg Glu Leu Ile Ala Tyr Ser Gln

441 Tyr Pro Arg Pro Ser Asp Ile Pro Gln Trp Asn Ser Asp Lys Pro Ser Leu Lys Asp Ile

461 Lys Ile Met Gly Tyr Ser Ile Arg Thr Ile Asp Tyr Arg Tyr Thr Val Trp Val Gly Phe

481 Asn Pro Asp Glu Phe Leu Ala Asn Phe Ser Asp Ile His Ala Gly Glu Leu Tyr Phe Val

501 Asp Ser Asp Pro Leu Gln Asp His Asn Met Tyr Asn Asp Ser Gln Gly Gly Asp Leu Phe

521 Gln Leu Leu Met Pro

Asn - marks sites of N-linked glycosylation

Cys - example site of cysteine conversion

FIG. 1B

FIG. 2A

I2S and SUMF1 co-expression options

Expression units on separate vectors (co-transfection or subsequent transfections)

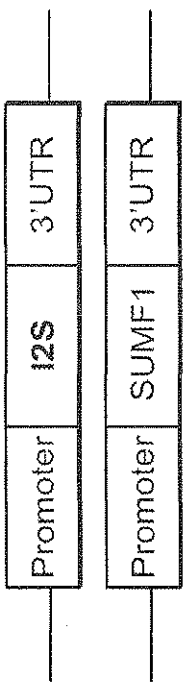


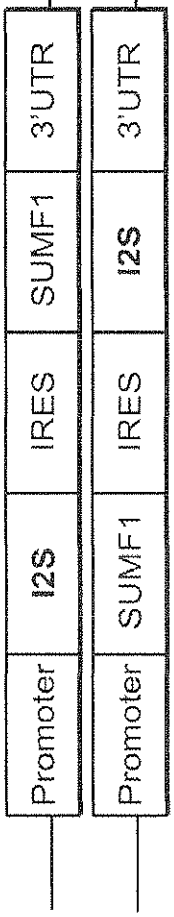
FIG. 2B

Expression units on the same vector (one transfection)

1) Separate cistrons



2) Transcriptionally linked cistrons



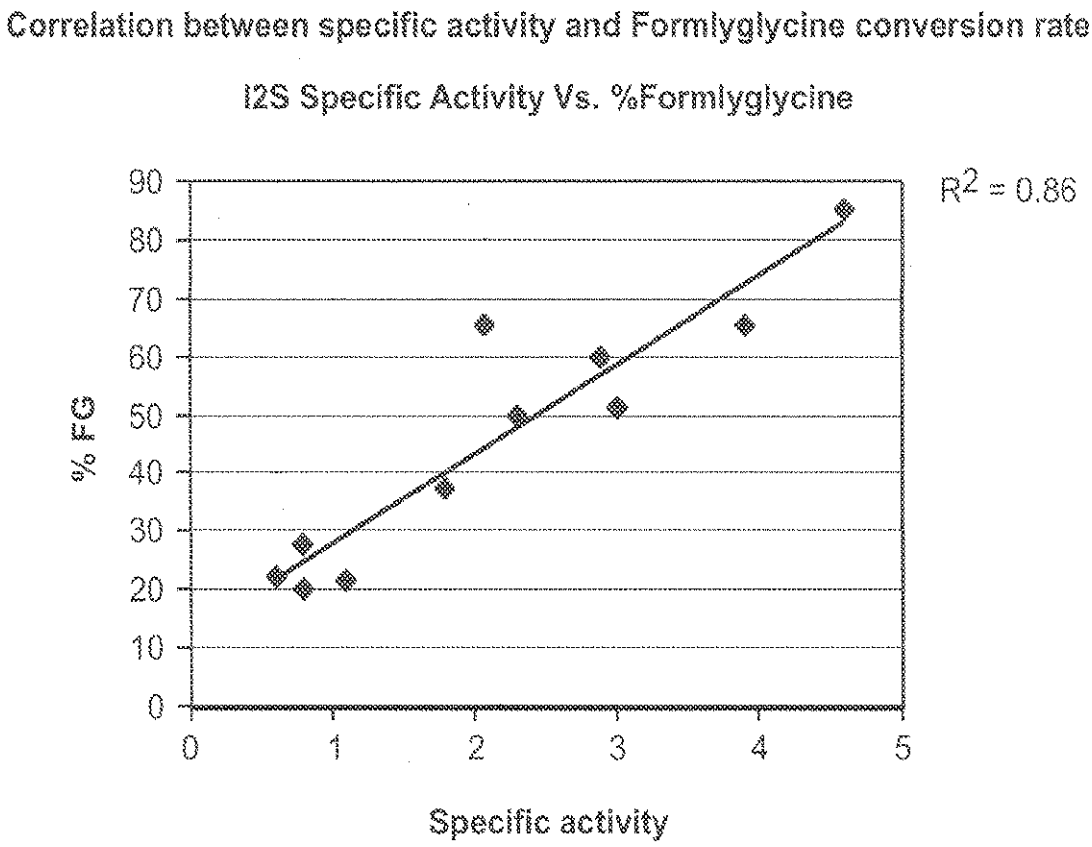

Correlation between specific activity and Formlyglycine conversion rate
I2S Specific Activity Vs. %Formlyglycine
R^2 = 0.86
% FG
90
80
70
60
50
40
30
20
10
0
0
1
2
3
4
5
Specific activity

FIG. 3

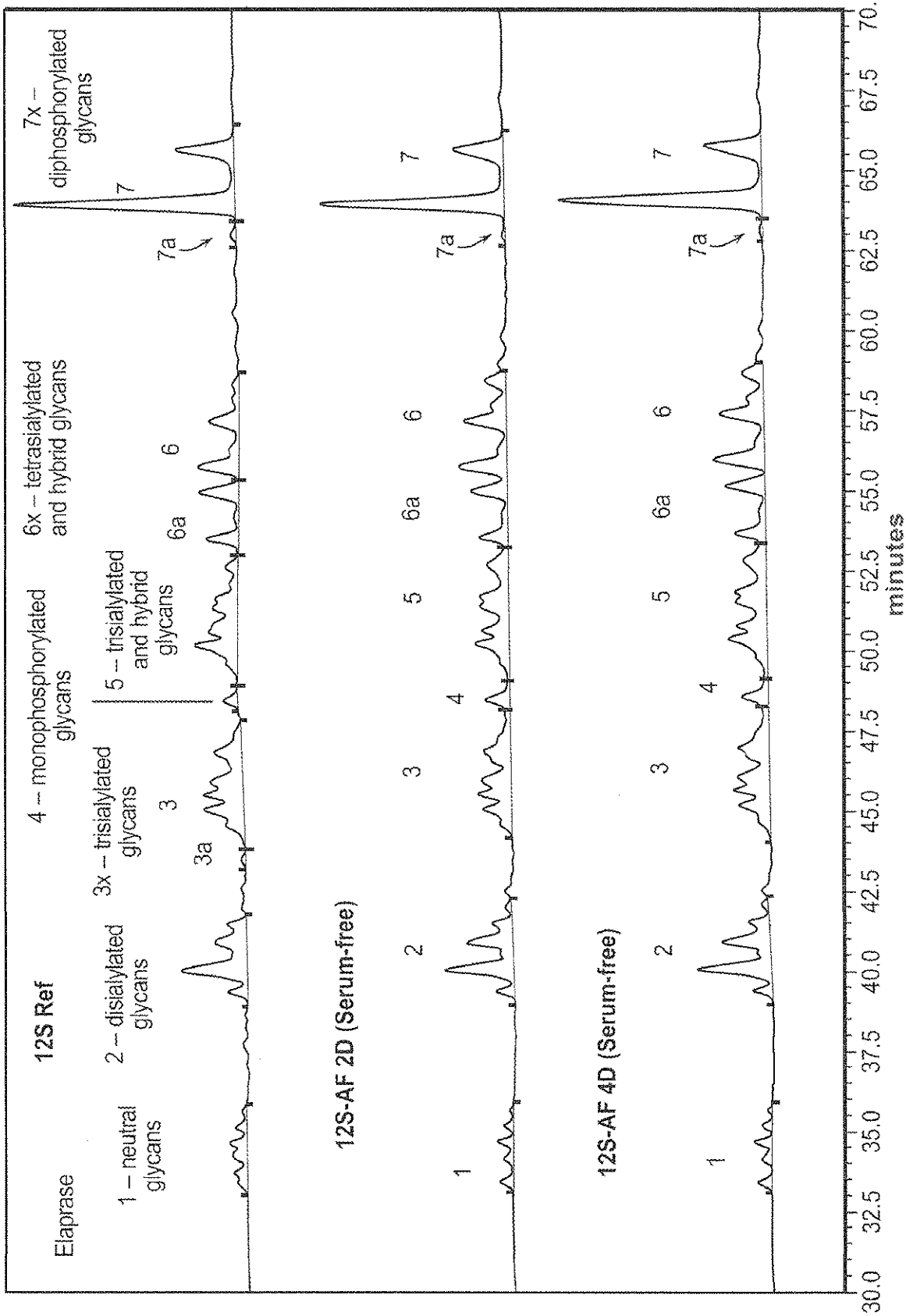

12S Ref
Elaprase
1 – neutral glycans
2 – disialylated glycans
3x – trisialylated glycans
4 – monophosphorylated glycans
5 – trisialylated and hybrid glycans
6x – tetrasialylated and hybrid glycans
7x – diphosphorylated glycans
12S-AF 2D (Serum-free)
12S-AF 4D (Serum-free)
minutes
30.0
32.5
35.0
37.5
40.0
42.5
45.0
47.5
50.0
52.5
55.0
57.5
60.0
62.5
65.0
67.5
70.0

FIG. 4