SEQ ID NO: 1

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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  Glu Leu Ala Ser His Ser Leu Leu Phe Glu Asn Ala Phe Ala Gln Gln Ala Val [Cys] Ala
61  Pro Ser Arg Val 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 Glu 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
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 Glu 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
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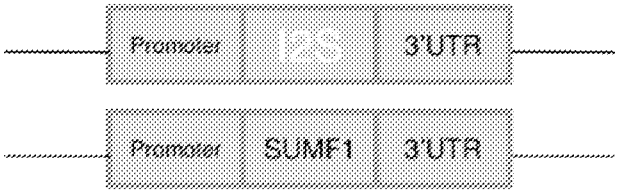
[Asn] - marks sites of N-linked glycosylation

[Cys] - example site of cysteine conversion

Fig. 1

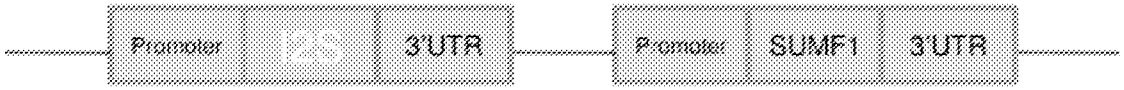
I2S and SUMF1 co-expression options

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

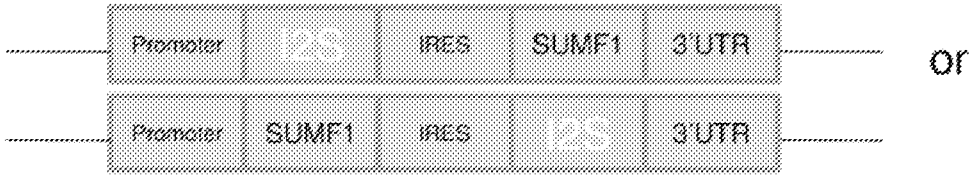


B) Expression units on the same vector (one transfection)

1) Separate cistrons

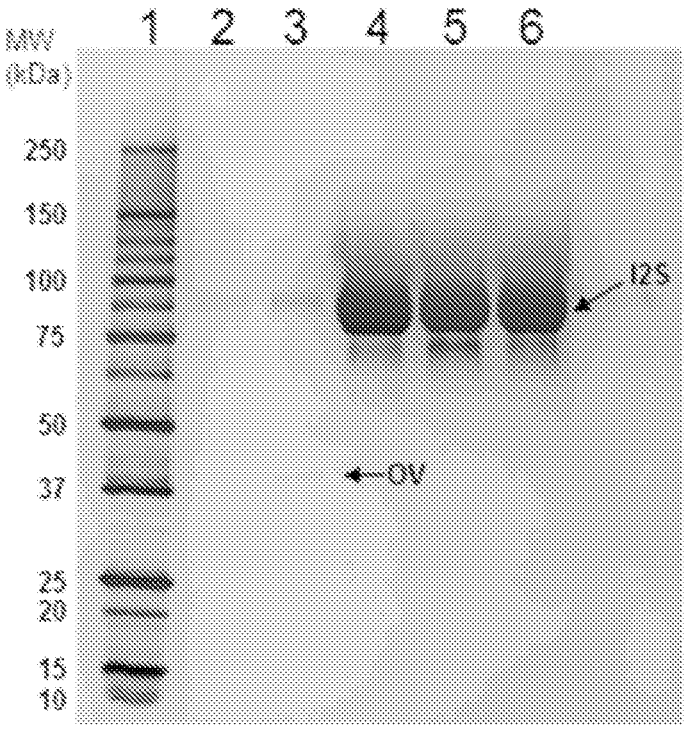


2) Transcriptionally linked cistrons



or

Fig. 2



Lane	Load Sample	Load Volume	Total µg
1	Protein Stds	3 µl	
2	Assay Control #1	20 µl	8 µg
3	Assay Control #2	20 µl	16 µg
4	I2S Reference Standard	20 µl	8 µg
5	I2S-AF 2D Serum-free Culture	20 µl	8 µg
6	I2S-AF 4D Serum-free Culture	20 µl	8 µg

Fig. 3

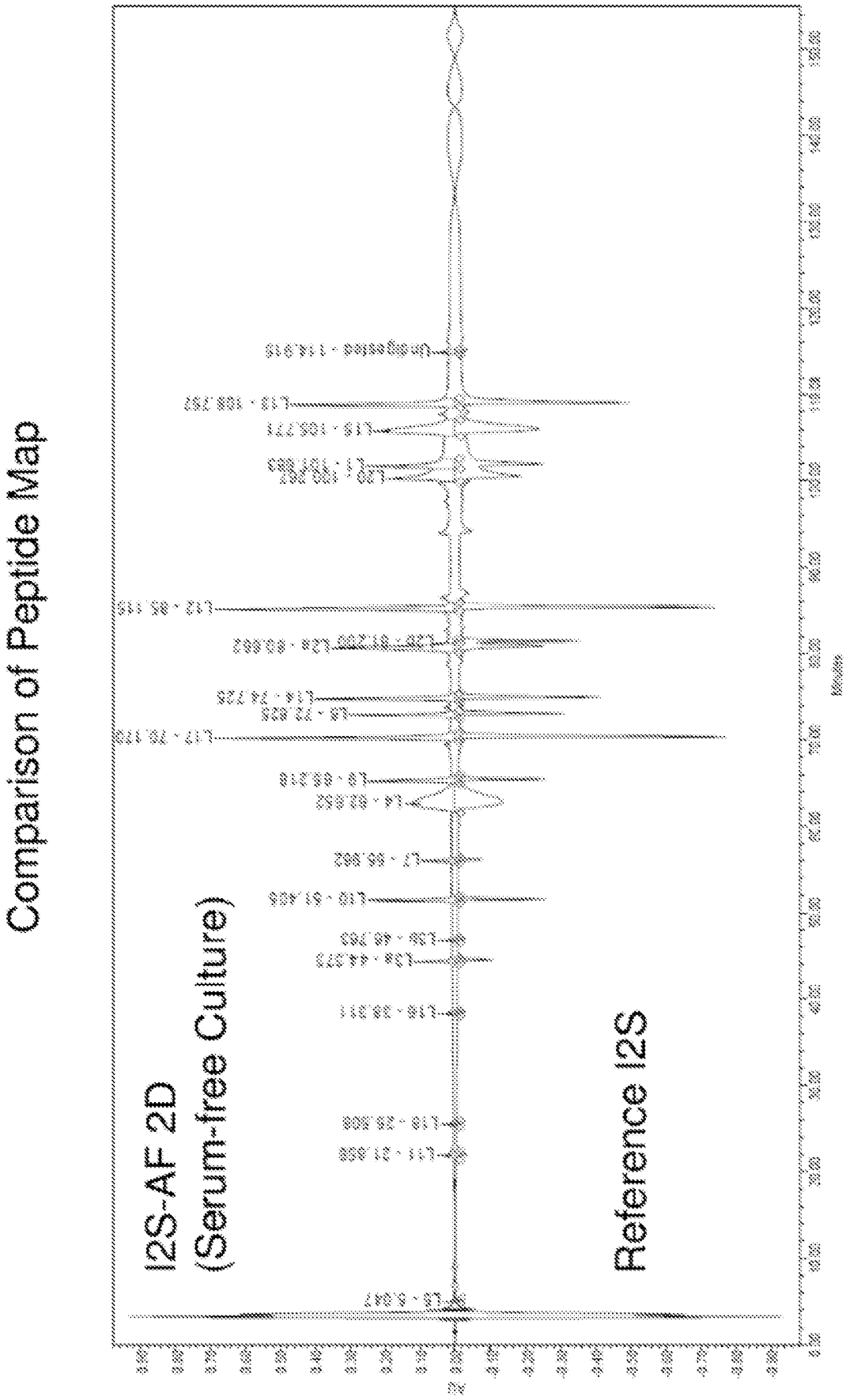

Comparison of Peptide Map
I2S-AF 2D
(Serum-free Culture)
Reference I2S
Minutes

Fig. 4

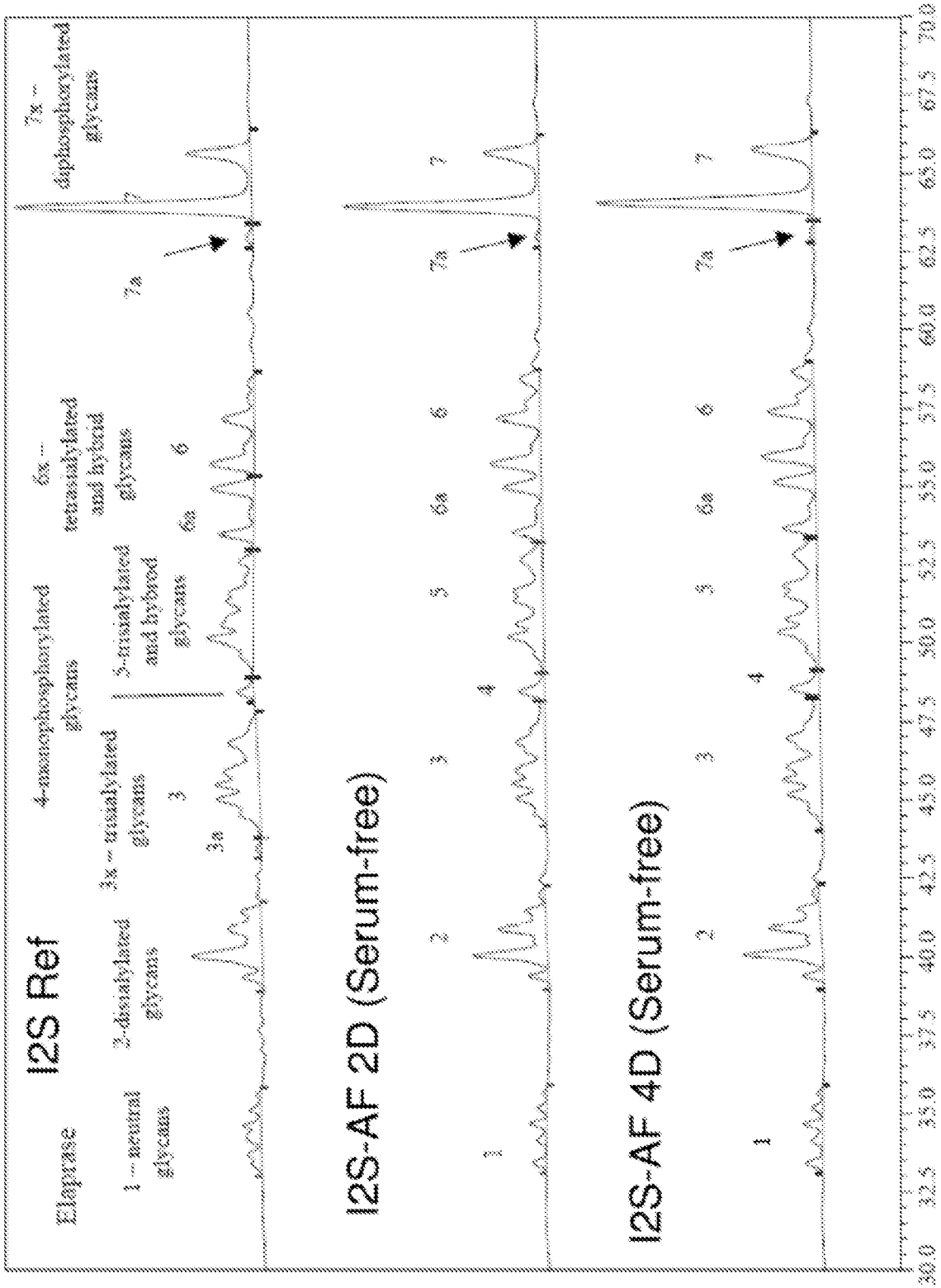

I2S Ref
Elaprase
1 -- neutral glycans
2 -- disialylated glycans
3x -- trisialylated glycans
4 -- monophosphorylated glycans
5 -- trisialylated and hybrid glycans
6x -- tetrasialylated and hybrid glycans
7x -- diphosphorylated glycans
I2S-AF 2D (Serum-free)
I2S-AF 4D (Serum-free)

Fig. 5

I2S Derived Using I2S-AF 2D Serum-free Cell Culture

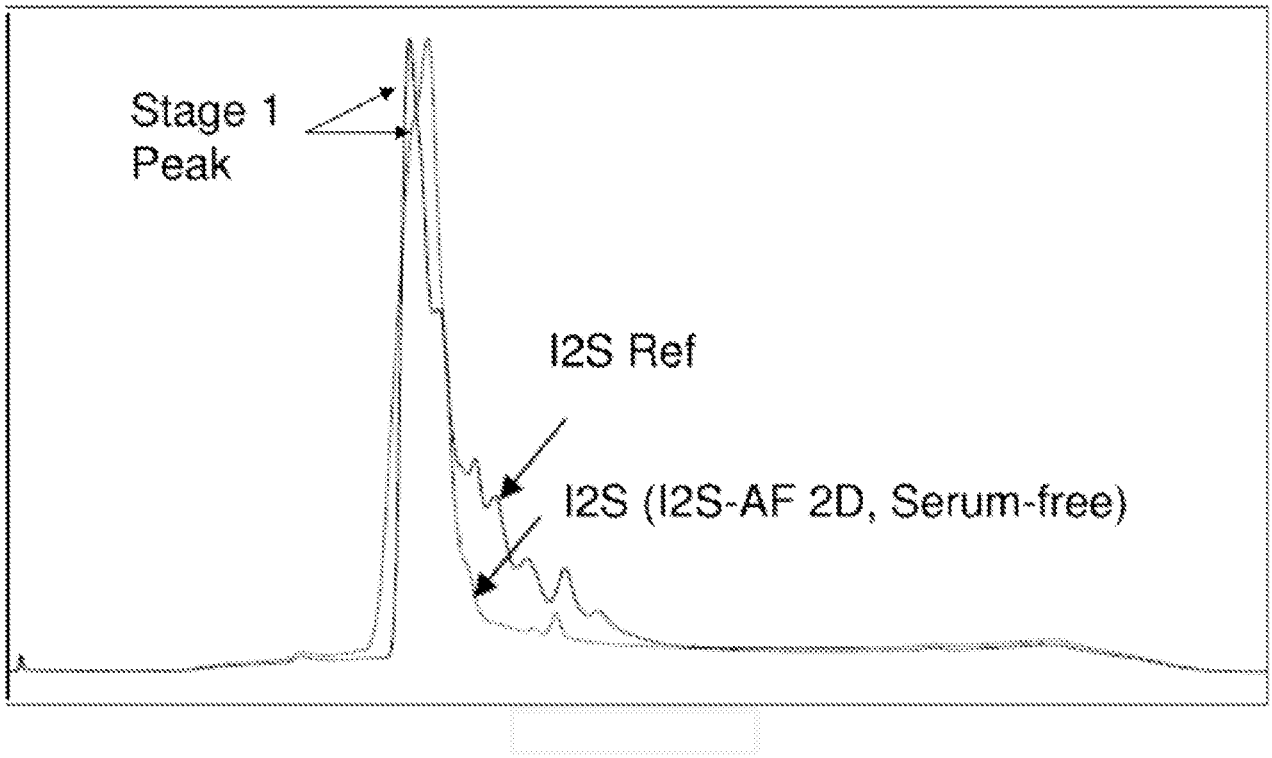


Fig. 6