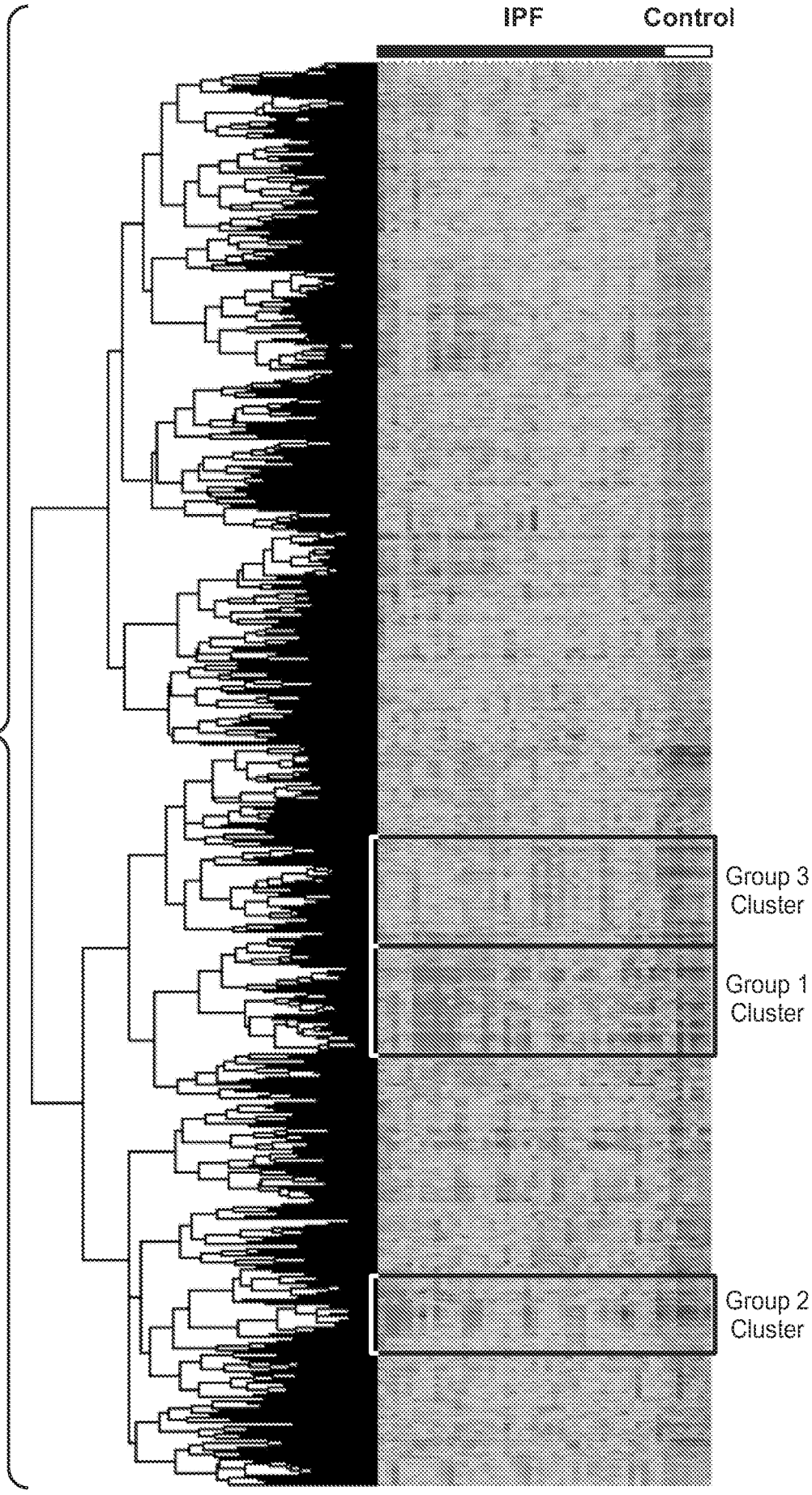
FIG. 1A



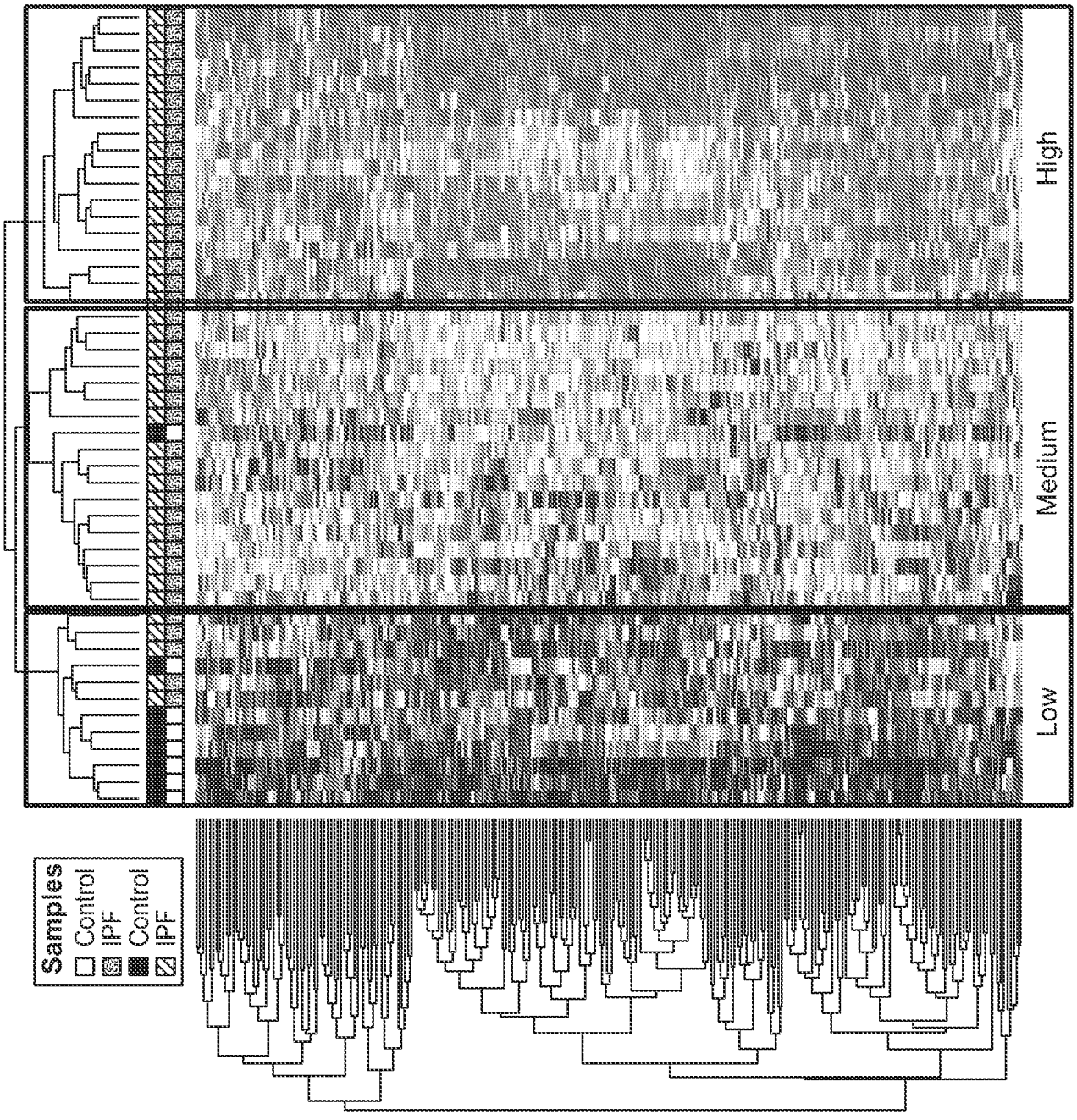


FIG. 1B

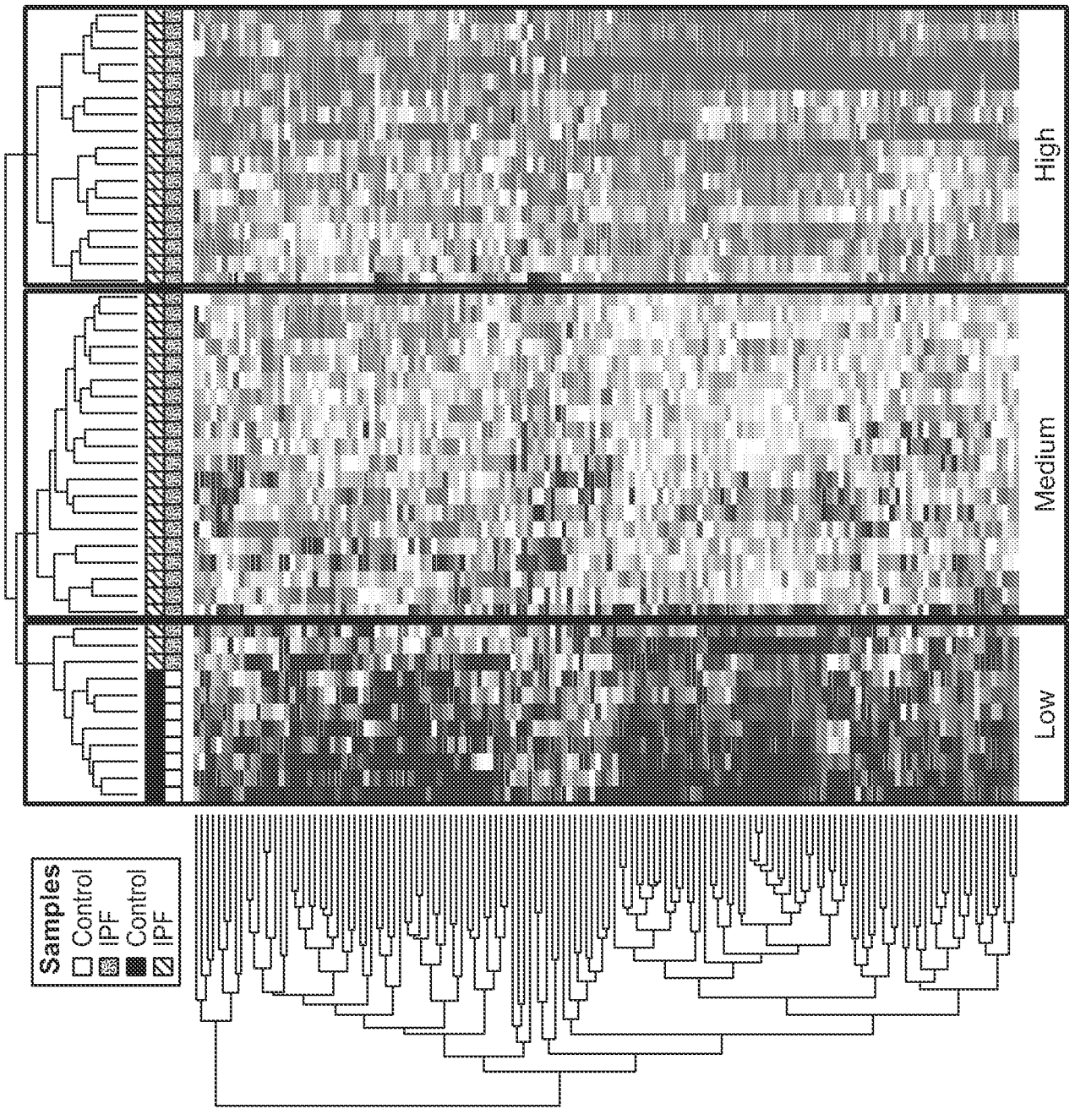


FIG. 1C

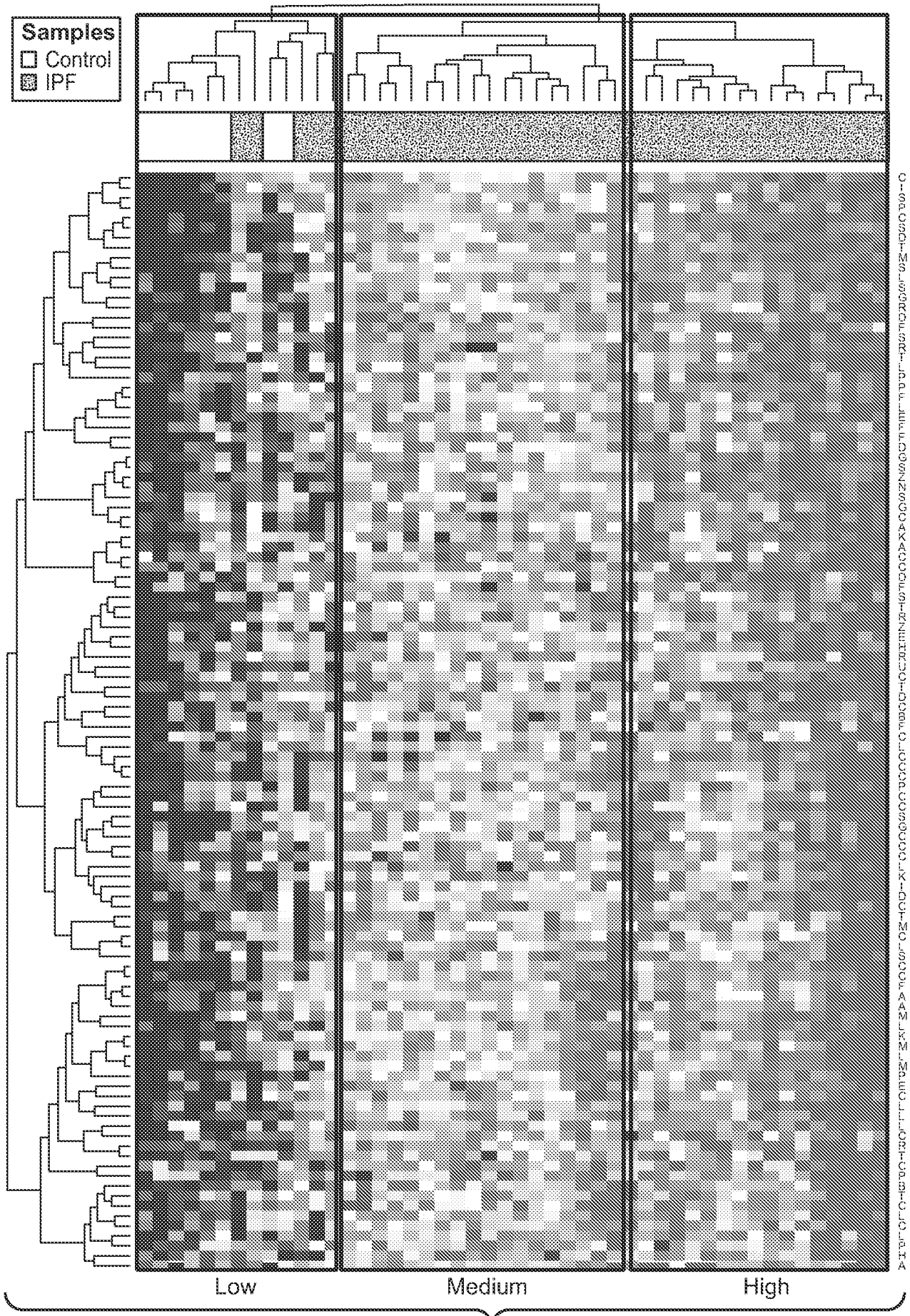


FIG. 1D

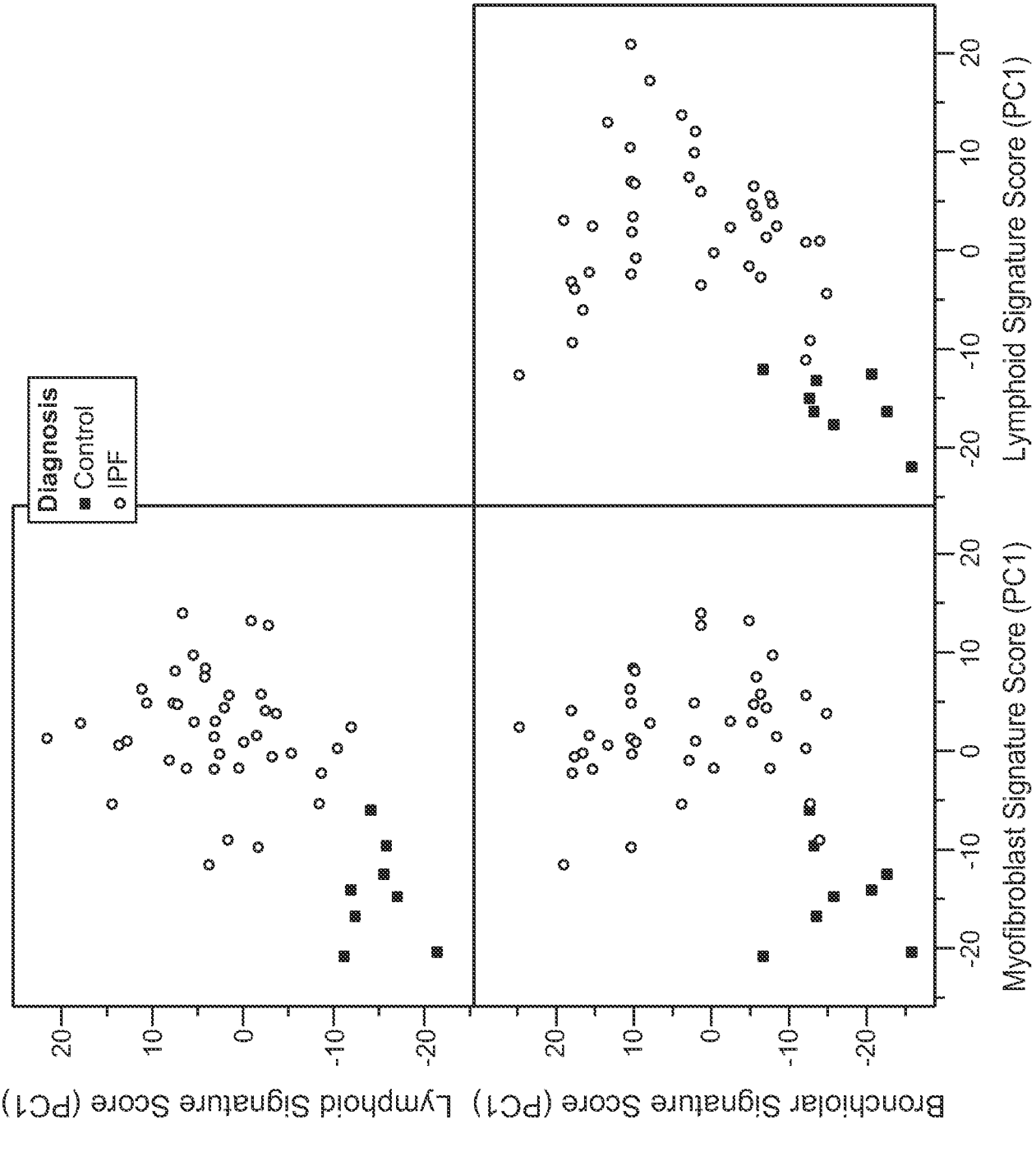

Diagnosis
Control
IPF
Bronchiolar Signature Score (PC1)
Lymphoid Signature Score (PC1)
Myofibroblast Signature Score (PC1)
Lymphoid Signature Score (PC1)
20
10
0
-10
-20

FIG. 1E

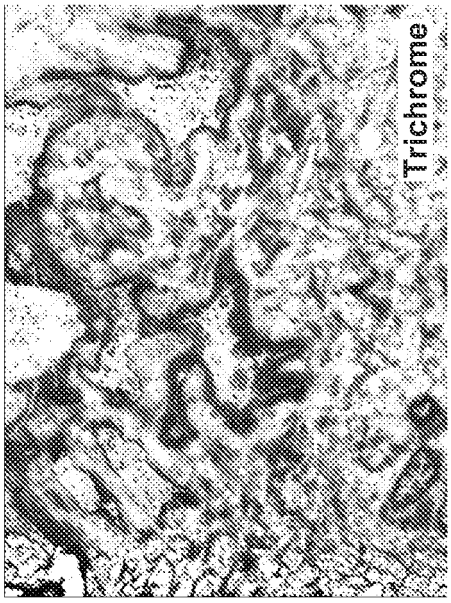


FIG. 2B

FIG. 2D

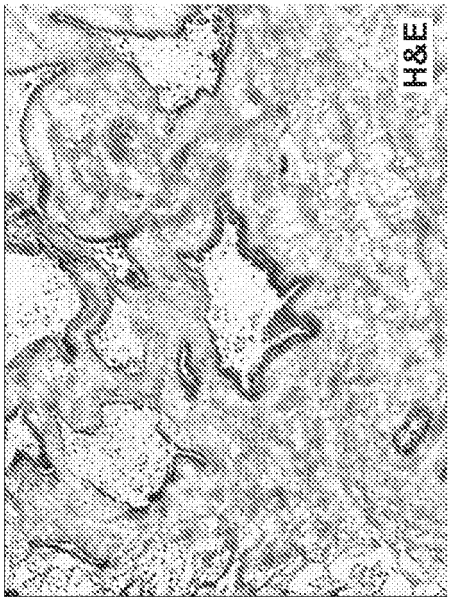


FIG. 2A

FIG. 2C

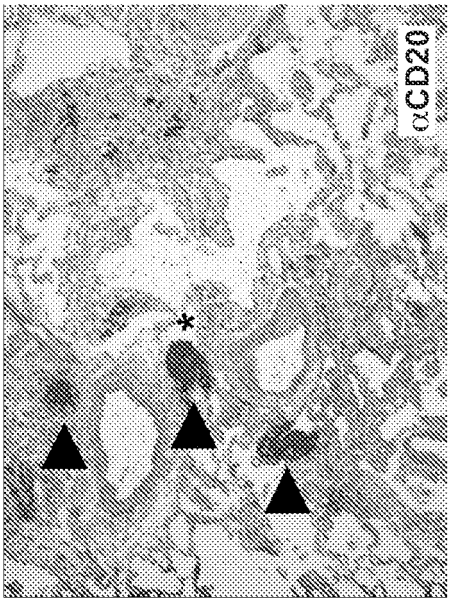


FIG. 2G

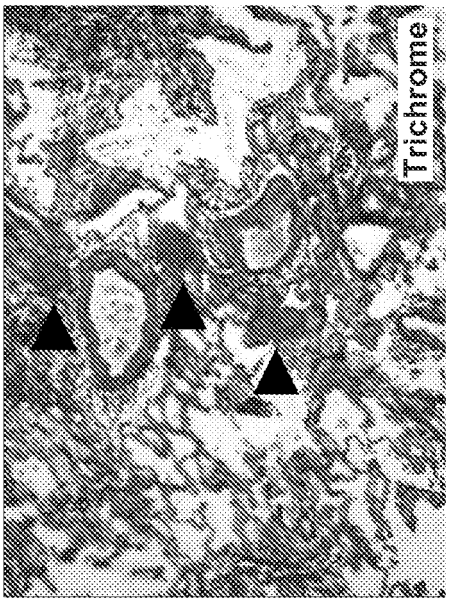


FIG. 2F

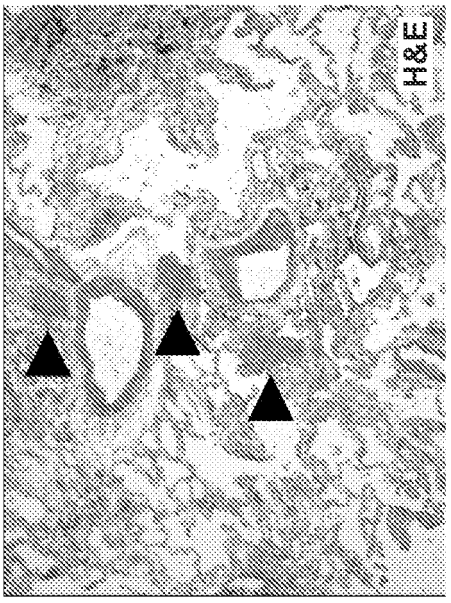


FIG. 2E

FIG. 2J

FIG. 2I

FIG. 2H

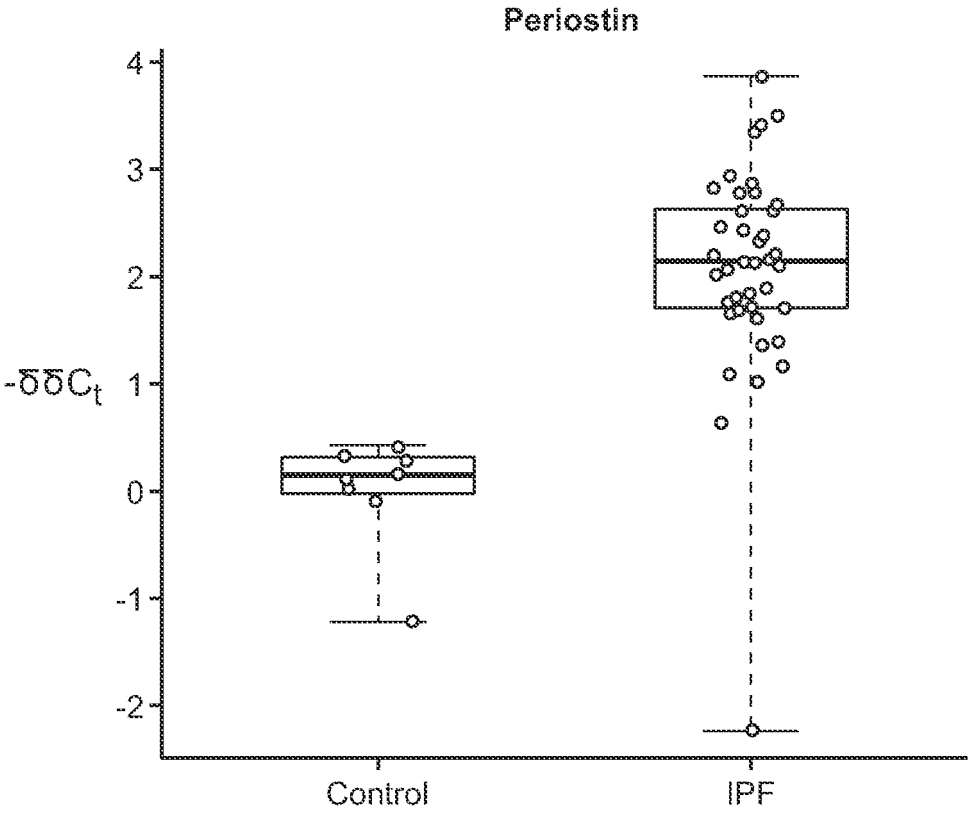

Periostin
4
3
2
1
0
-1
-2
-δδCt
Control
IPF

FIG. 3A

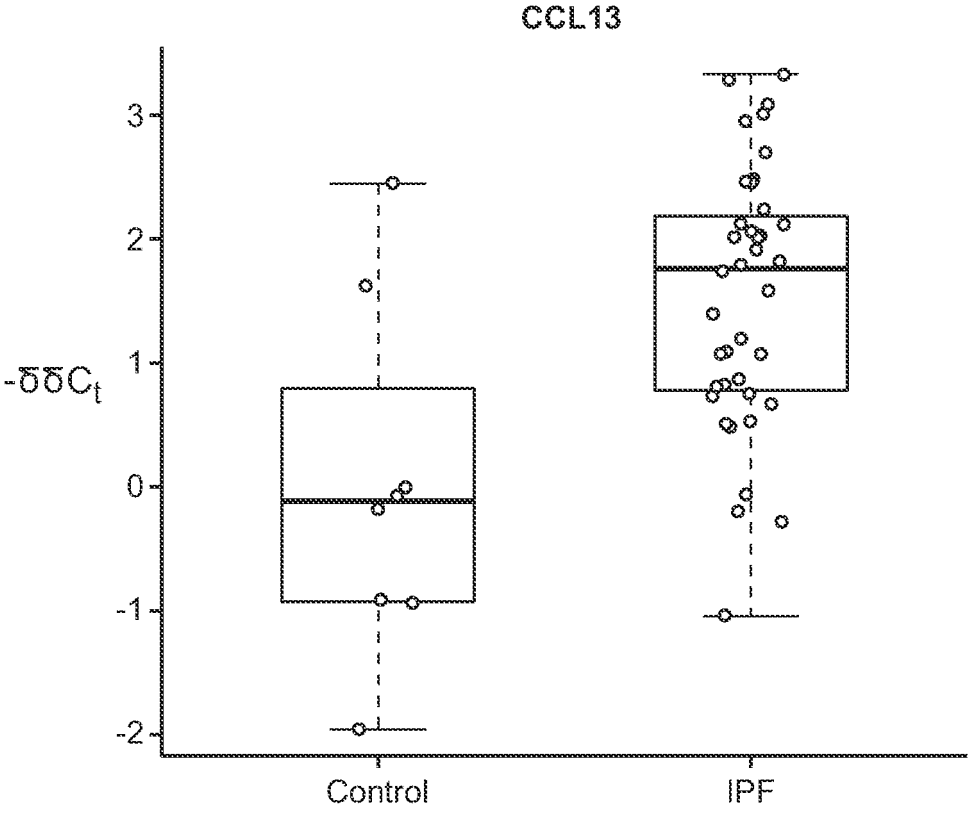

CCL13
3
2
1
0
-1
-2
-δδCt
Control
IPF

FIG. 3B

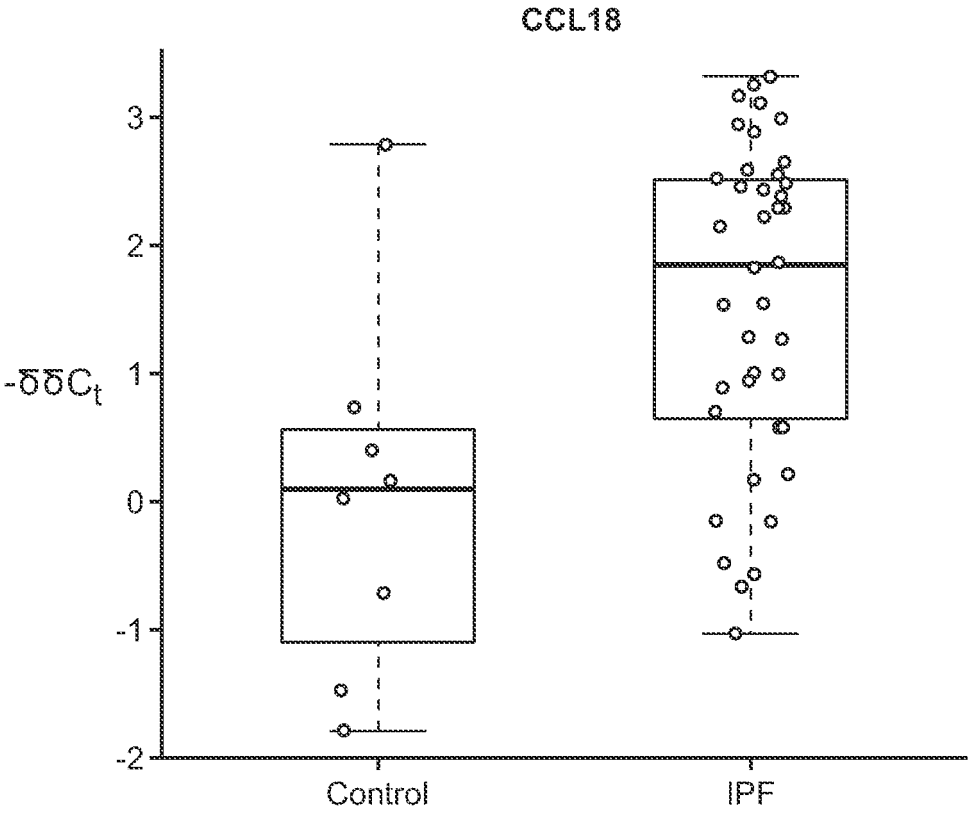

CCL18
-δδCt
3
2
1
0
-1
-2
Control
IPF

FIG. 3C

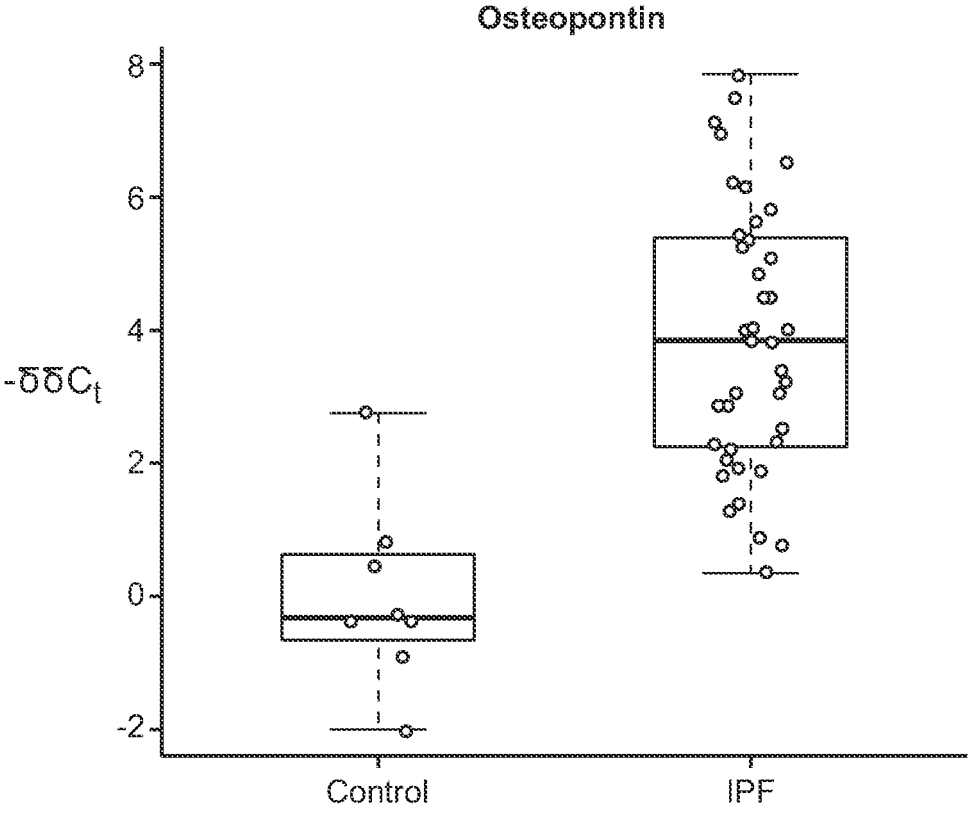

Osteopontin
-δδCt
8
6
4
2
0
-2
Control
IPF

FIG. 3D

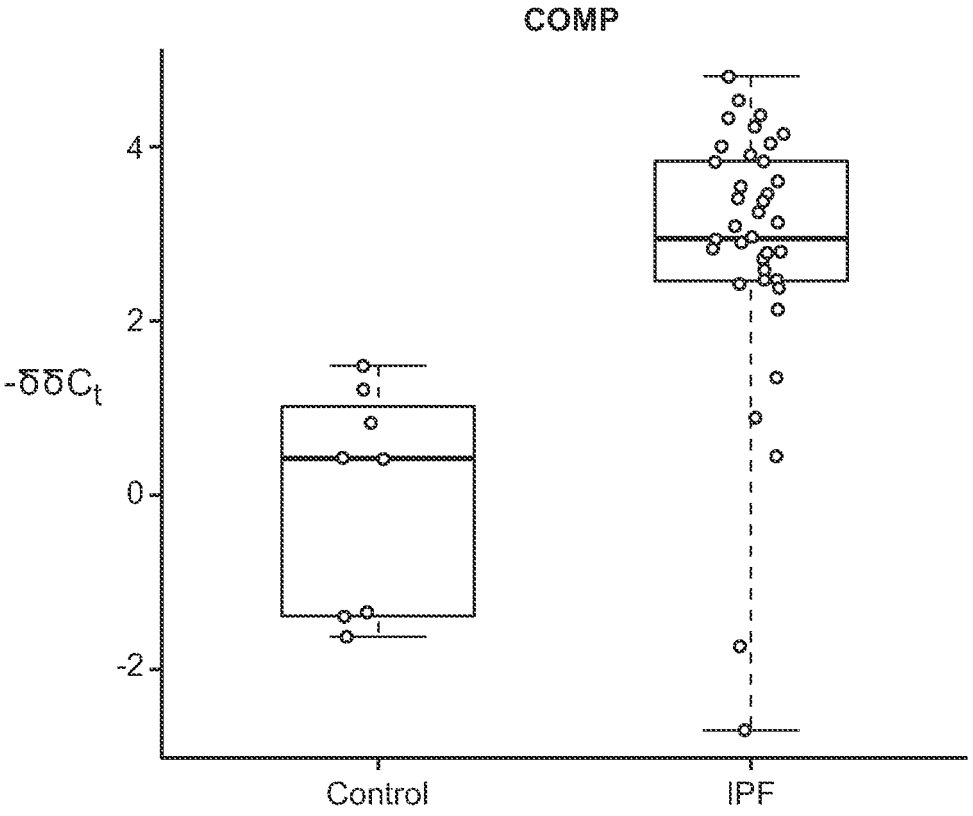


FIG. 3E

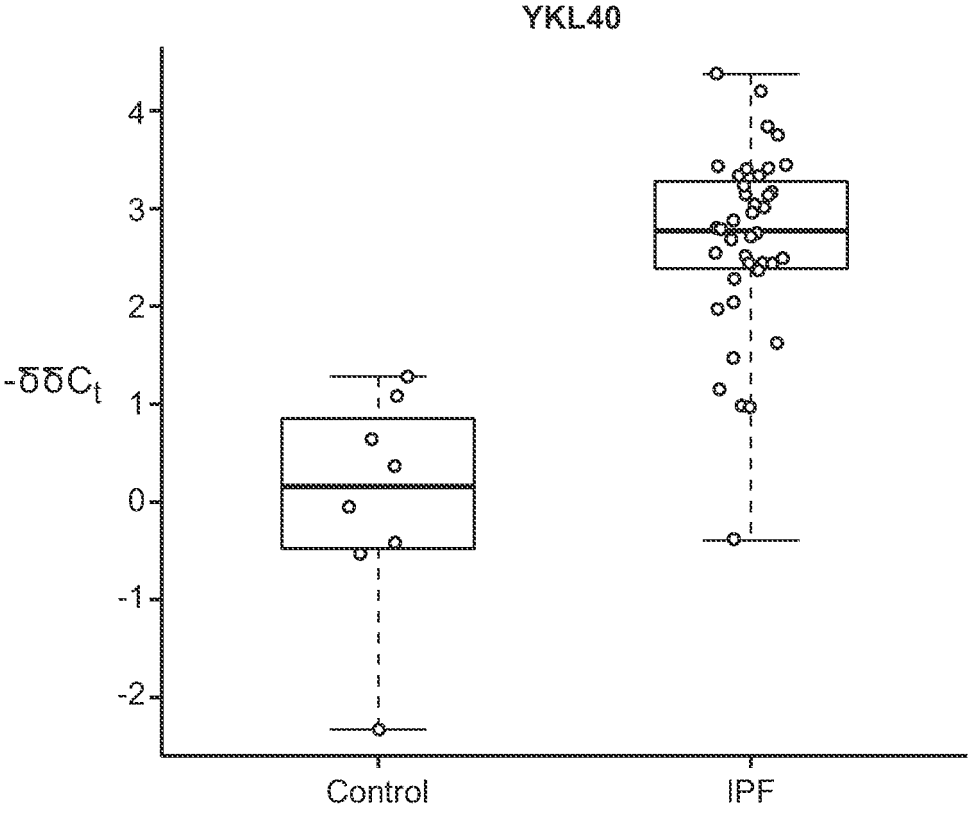


FIG. 3F

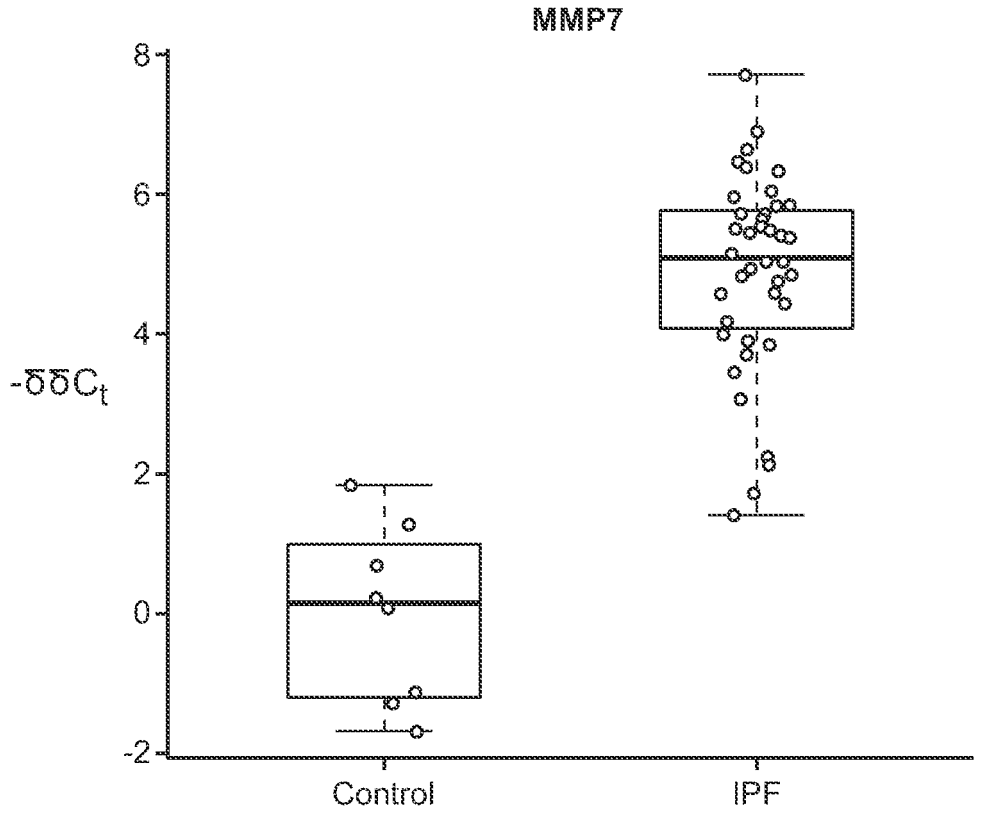

MMP7
8
6
4
2
0
-2
-δδCt
Control
IPF

FIG. 3G

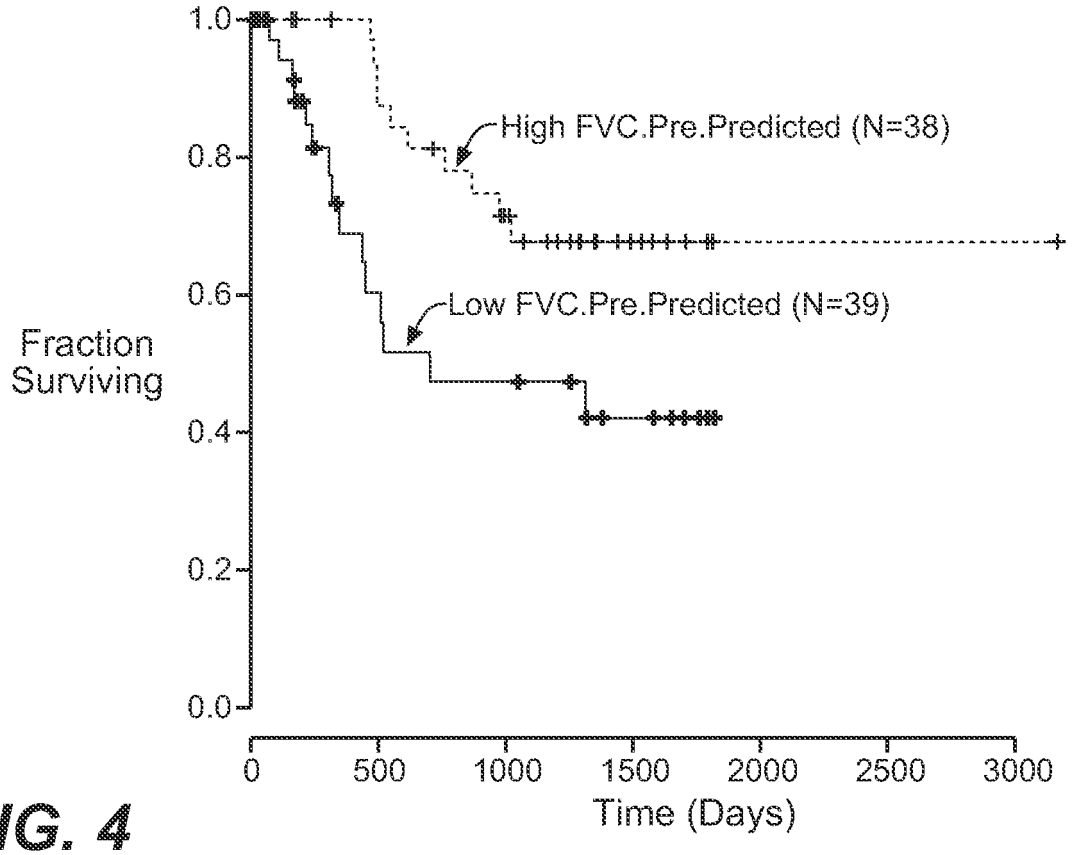

1.0
0.8
0.6
0.4
0.2
0.0
Fraction Surviving
High FVC.Pre.Predicted (N=38)
Low FVC.Pre.Predicted (N=39)
0
500
1000
1500
2000
2500
3000
Time (Days)

FIG. 4

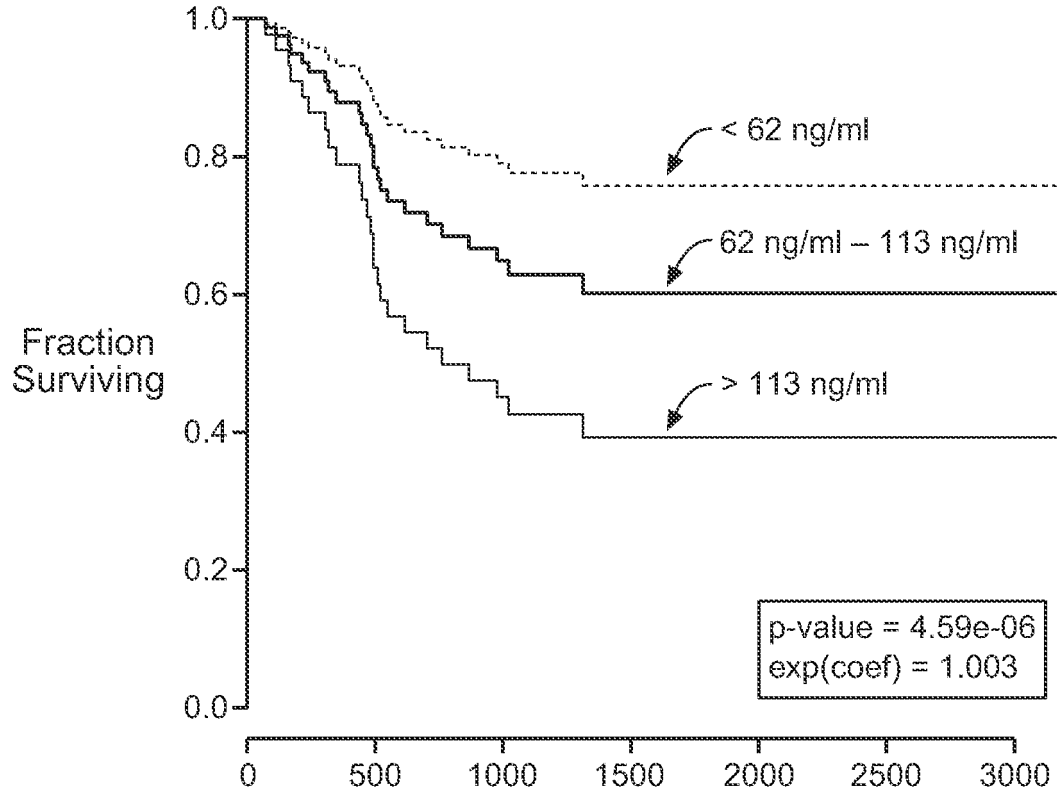

1.0
0.8
0.6
0.4
0.2
0.0
Fraction Surviving
< 62 ng/ml
62 ng/ml – 113 ng/ml
> 113 ng/ml
p-value = 4.59e-06
exp(coef) = 1.003
0
500
1000
1500
2000
2500
3000
Time (Days)

FIG. 5A

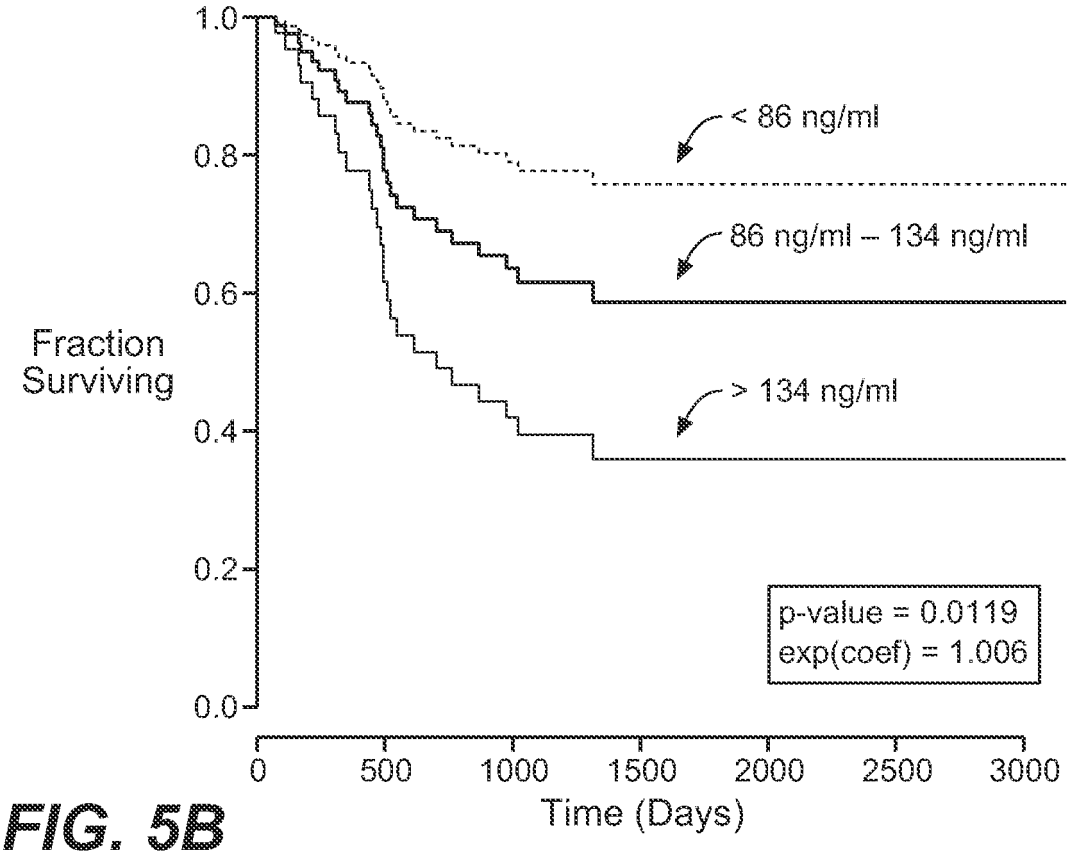

1.0
0.8
0.6
0.4
0.2
0.0
Fraction Surviving
< 86 ng/ml
86 ng/ml – 134 ng/ml
> 134 ng/ml
p-value = 0.0119
exp(coef) = 1.006
0
500
1000
1500
2000
2500
3000
Time (Days)

FIG. 5B

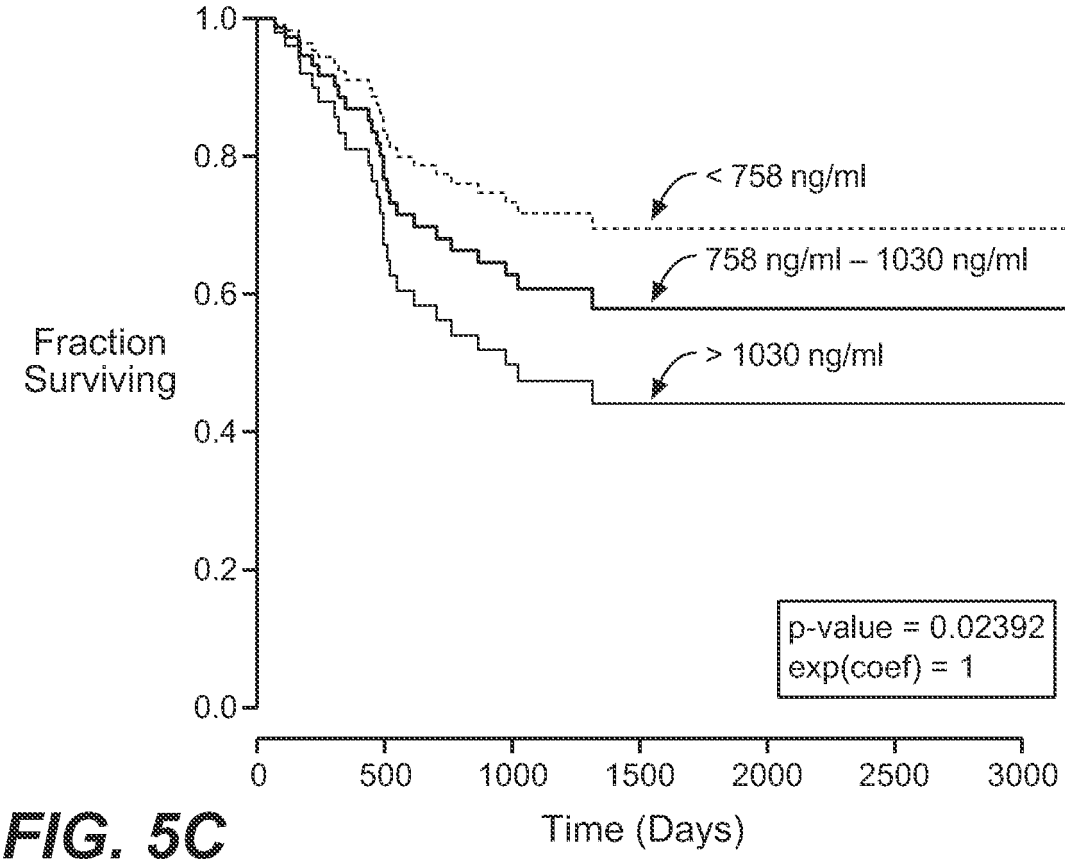

1.0
0.8
0.6
0.4
0.2
0.0
Fraction Surviving
< 758 ng/ml
758 ng/ml – 1030 ng/ml
> 1030 ng/ml
p-value = 0.02392
exp(coef) = 1
0
500
1000
1500
2000
2500
3000
Time (Days)

FIG. 5C

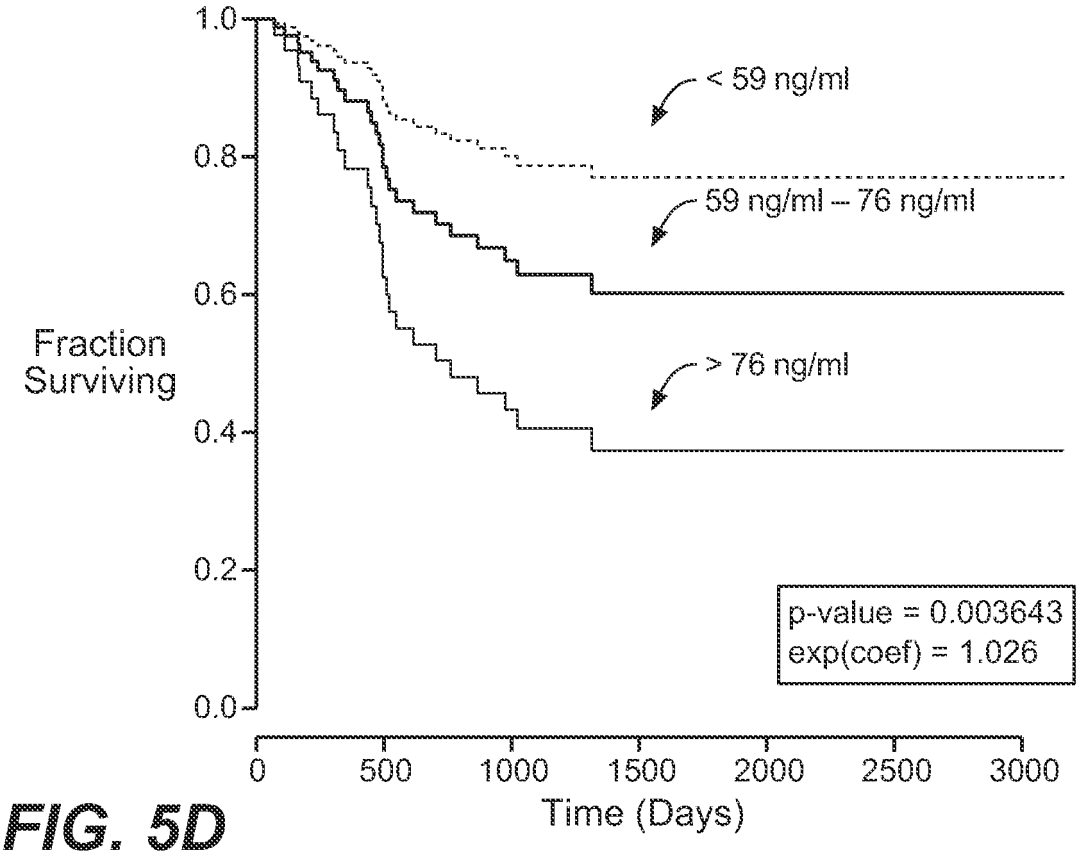

1.0
0.8
0.6
0.4
0.2
0.0
Fraction Surviving
< 59 ng/ml
59 ng/ml – 76 ng/ml
> 76 ng/ml
p-value = 0.003643
exp(coef) = 1.026
0
500
1000
1500
2000
2500
3000
Time (Days)

FIG. 5D

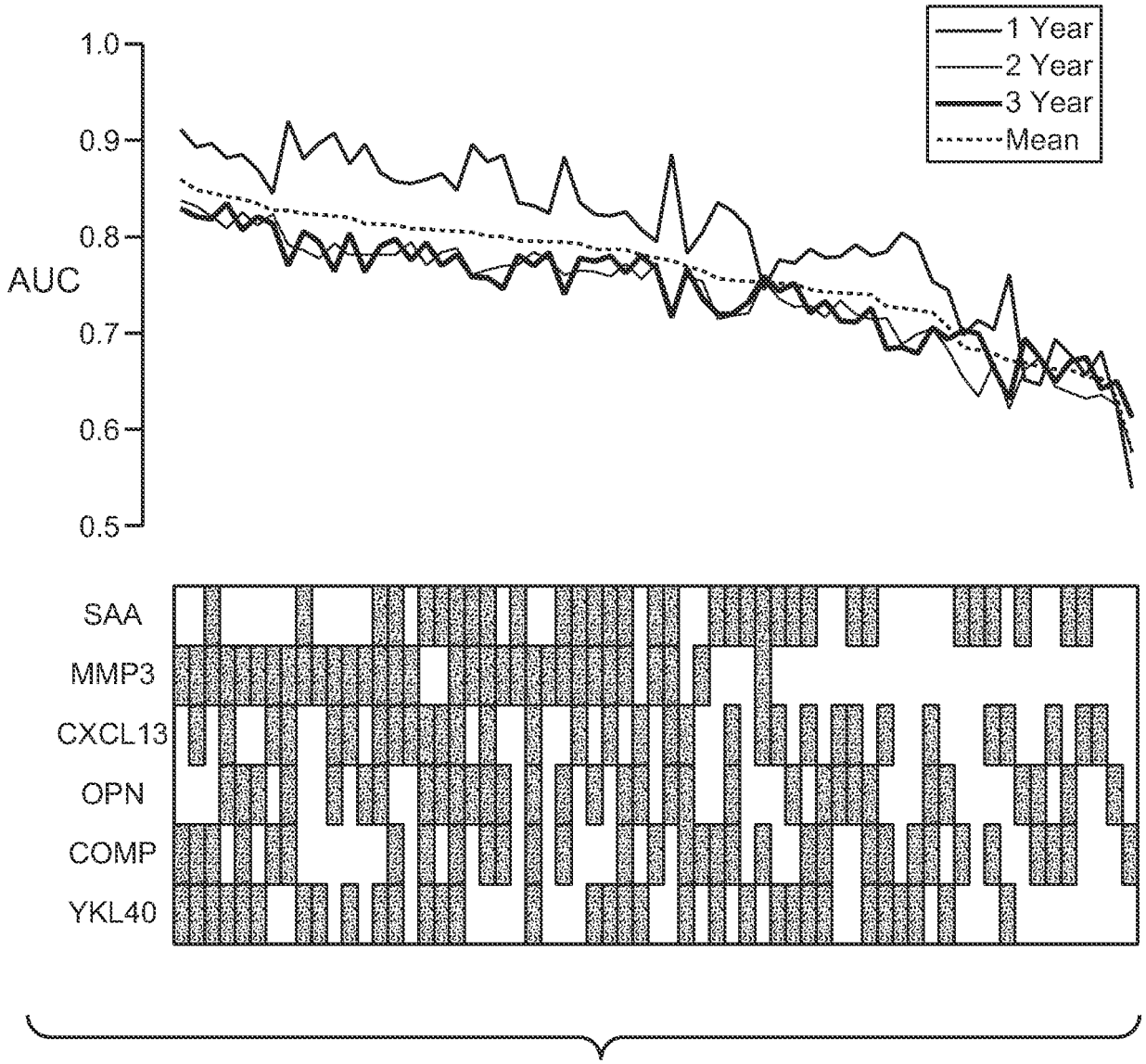

1 Year
2 Year
3 Year
Mean
AUC
1.0
0.9
0.8
0.7
0.6
0.5
SAA
MMP3
CXCL13
OPN
COMP
YKL40

FIG. 6A

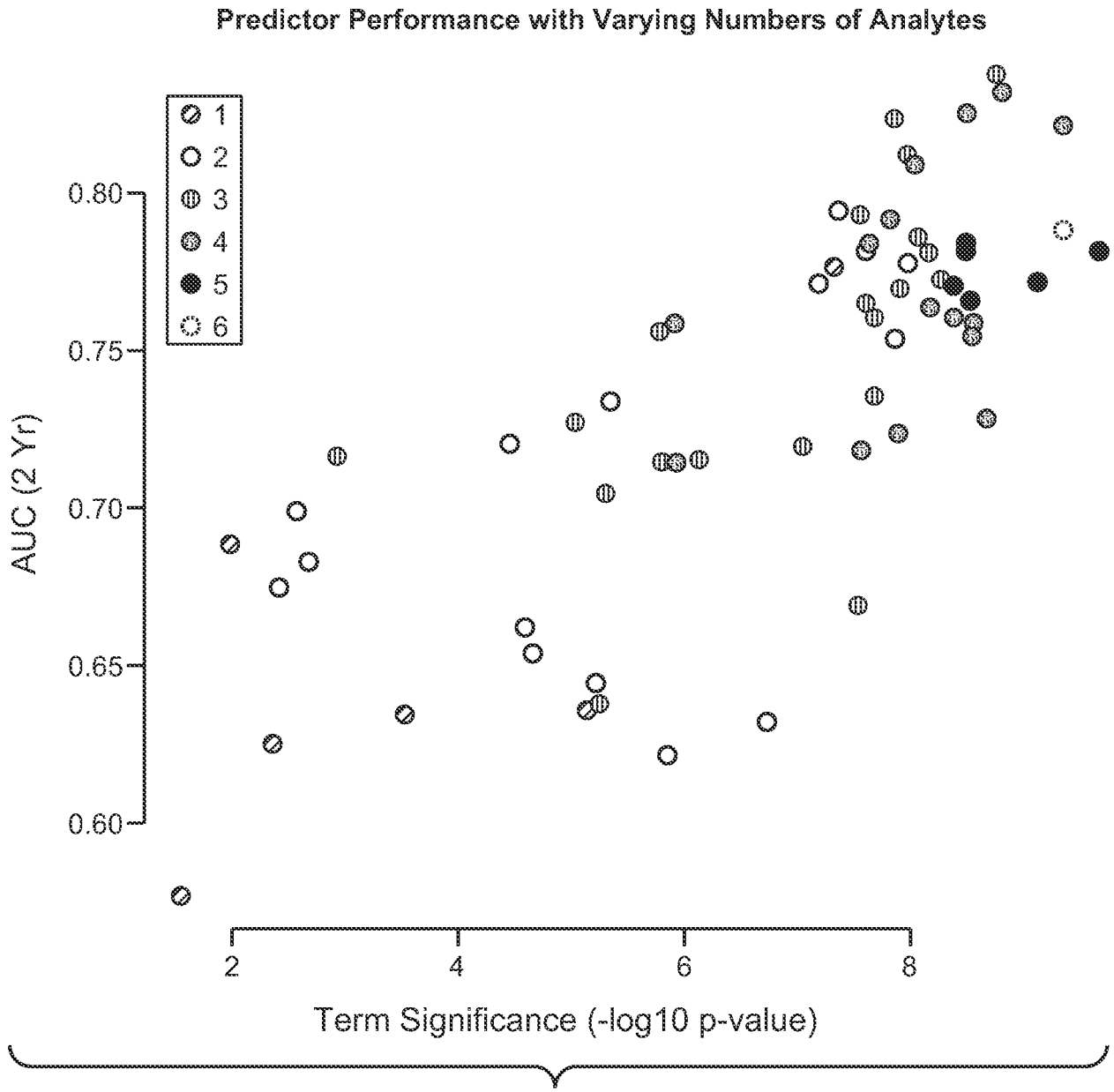

Predictor Performance with Varying Numbers of Analytes
1
2
3
4
5
6
0.80
0.75
0.70
0.65
0.60
AUC (2 Yr)
2
4
6
8
Term Significance (-log10 p-value)

FIG. 6B

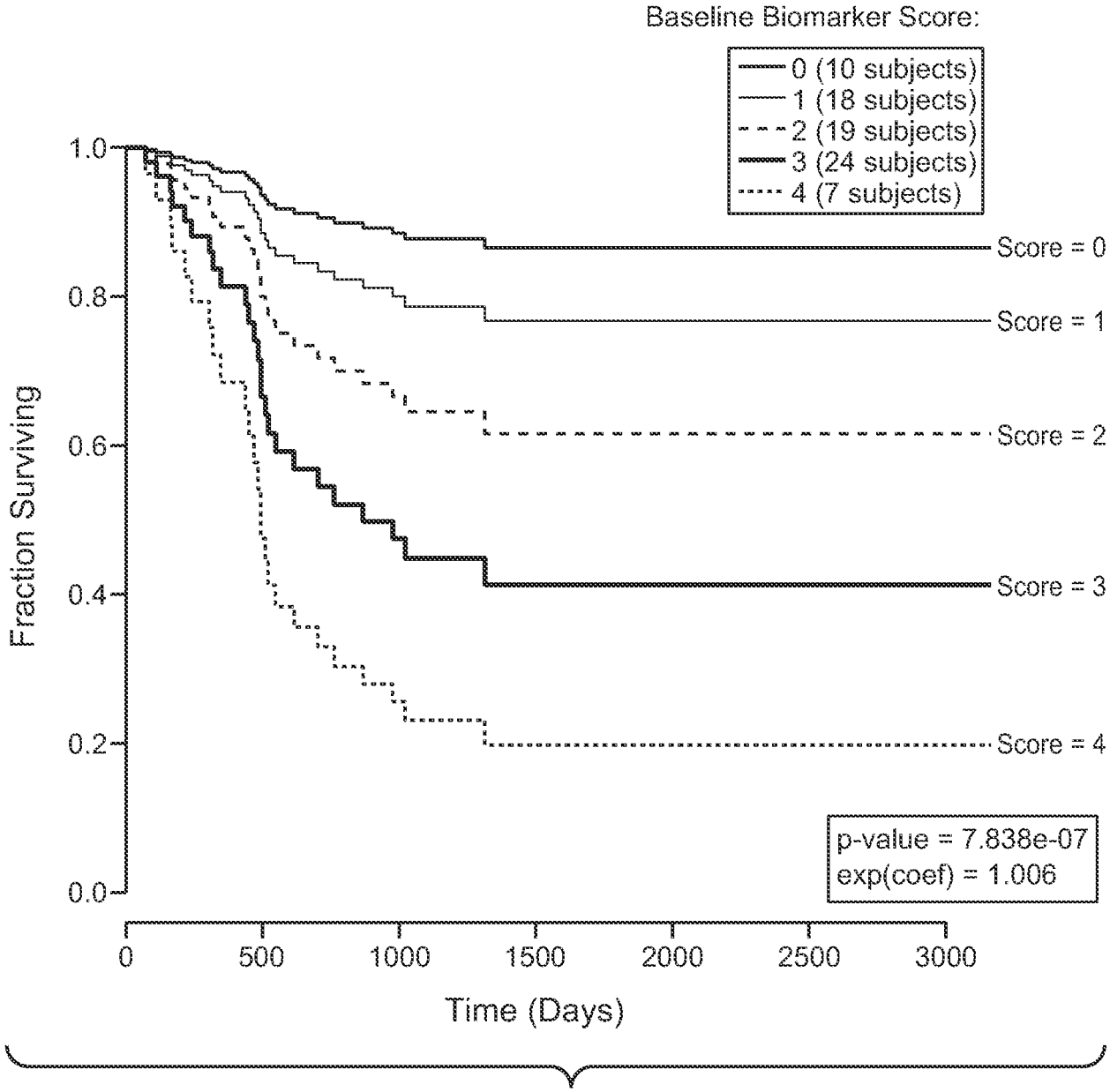

Baseline Biomarker Score:
0 (10 subjects)
1 (18 subjects)
2 (19 subjects)
3 (24 subjects)
4 (7 subjects)
Score = 0
Score = 1
Score = 2
Score = 3
Score = 4
p-value = 7.838e-07
exp(coef) = 1.006
Fraction Surviving
1.0
0.8
0.6
0.4
0.2
0.0
0
500
1000
1500
2000
2500
3000
Time (Days)

FIG. 7

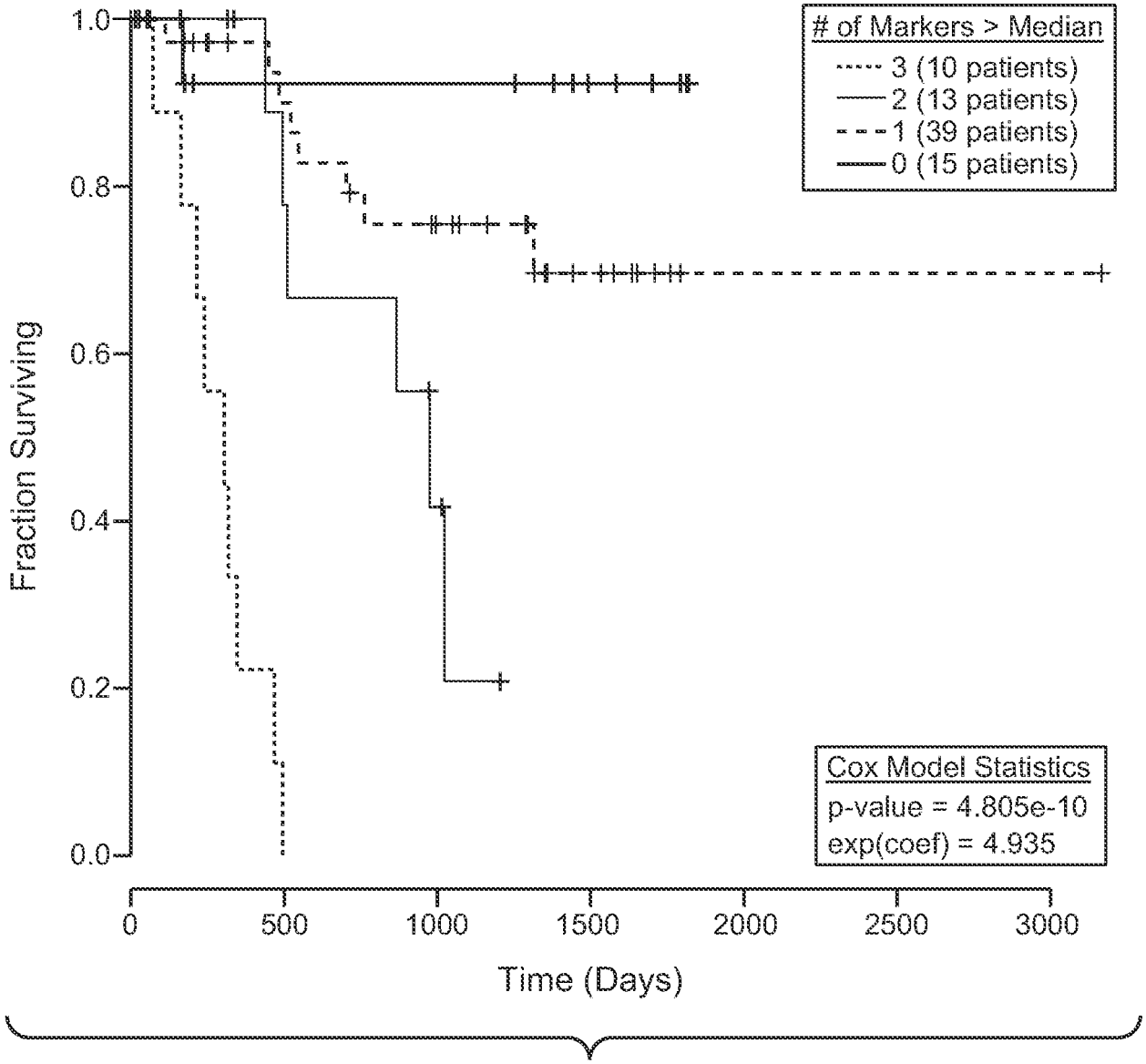

1.0
0.8
0.6
0.4
0.2
0.0
Fraction Surviving
0
500
1000
1500
2000
2500
3000
Time (Days)
# of Markers > Median
3 (10 patients)
2 (13 patients)
1 (39 patients)
0 (15 patients)
Cox Model Statistics
p-value = 4.805e-10
exp(coef) = 4.935

FIG. 8

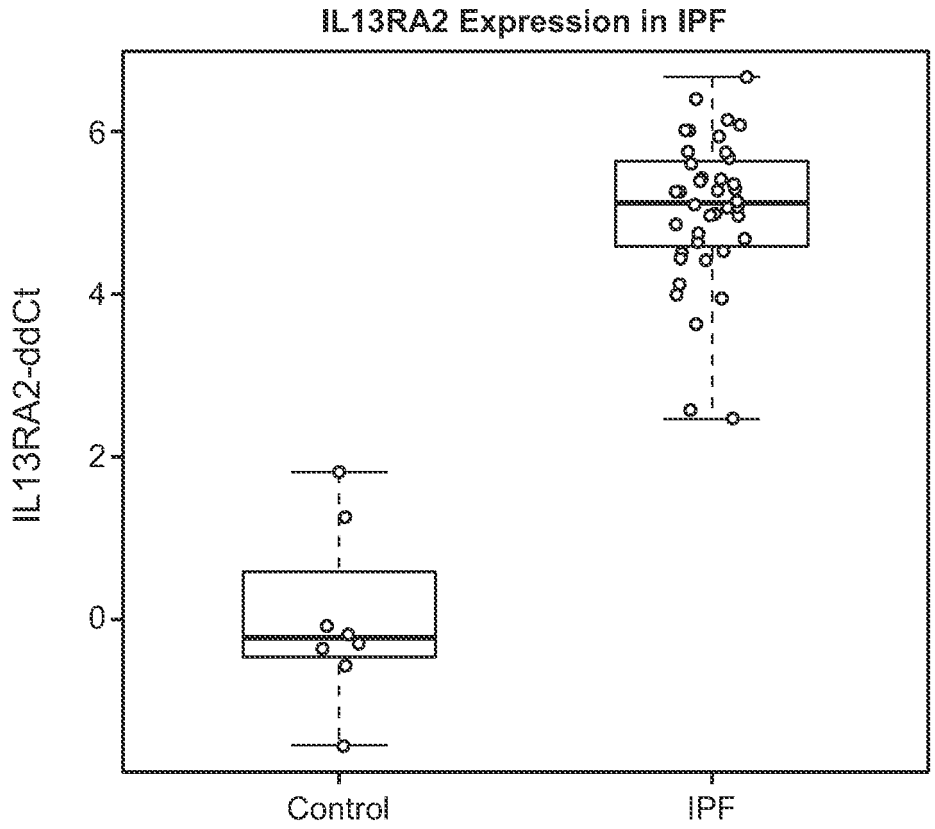

IL13RA2 Expression in IPF
IL13RA2-ddCt
6
4
2
0
Control
IPF

FIG. 9

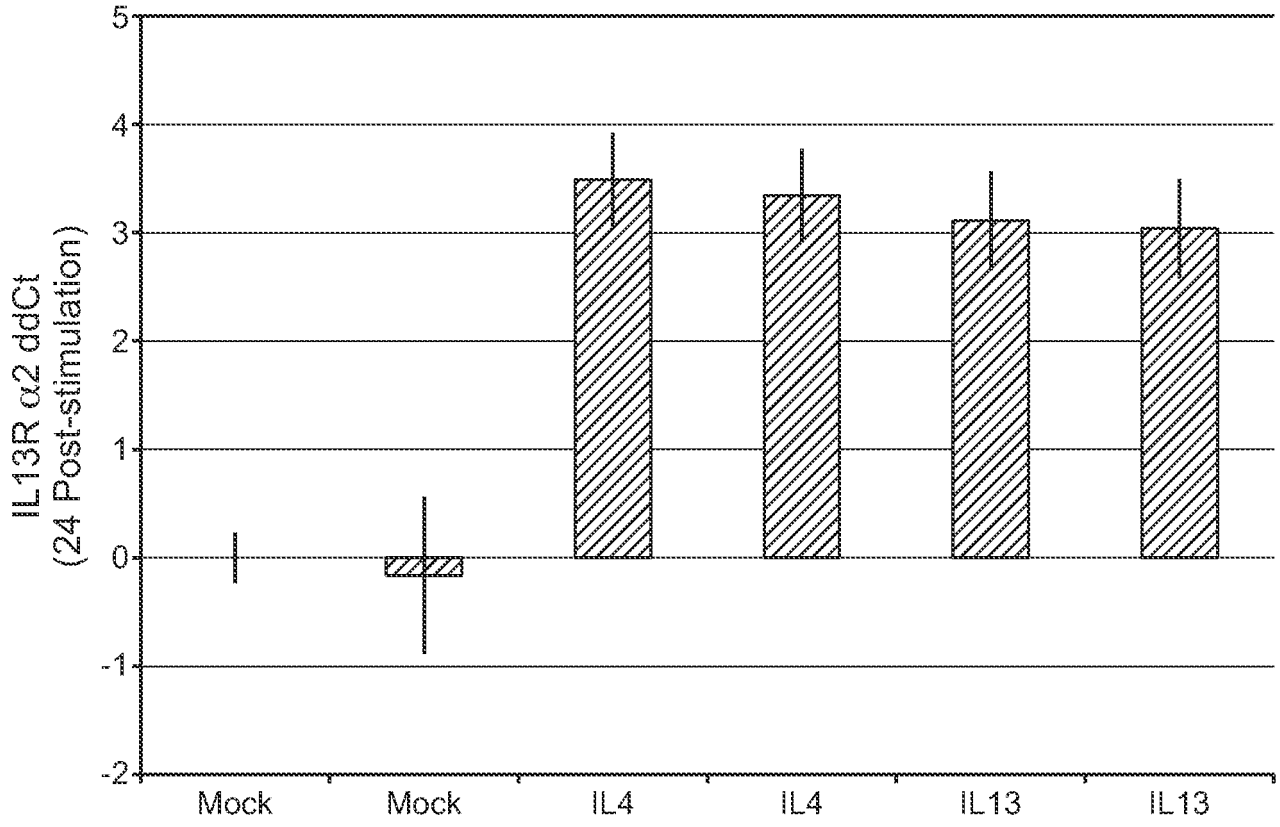

IL13R α2 ddCt
(24 Post-stimulation)
5
4
3
2
1
0
-1
-2
Mock
Mock
IL4
IL4
IL13
IL13

FIG. 10

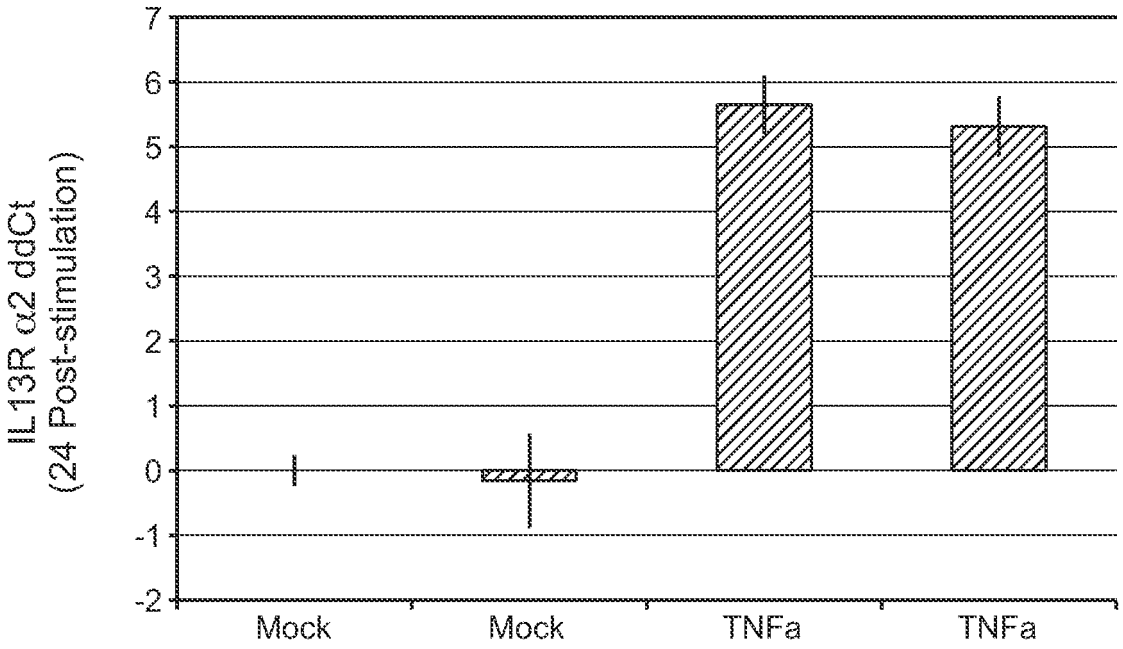

IL13R α2 ddCt
(24 Post-stimulation)
7
6
5
4
3
2
1
0
-1
-2
Mock
Mock
TNFa
TNFa

FIG. 11A

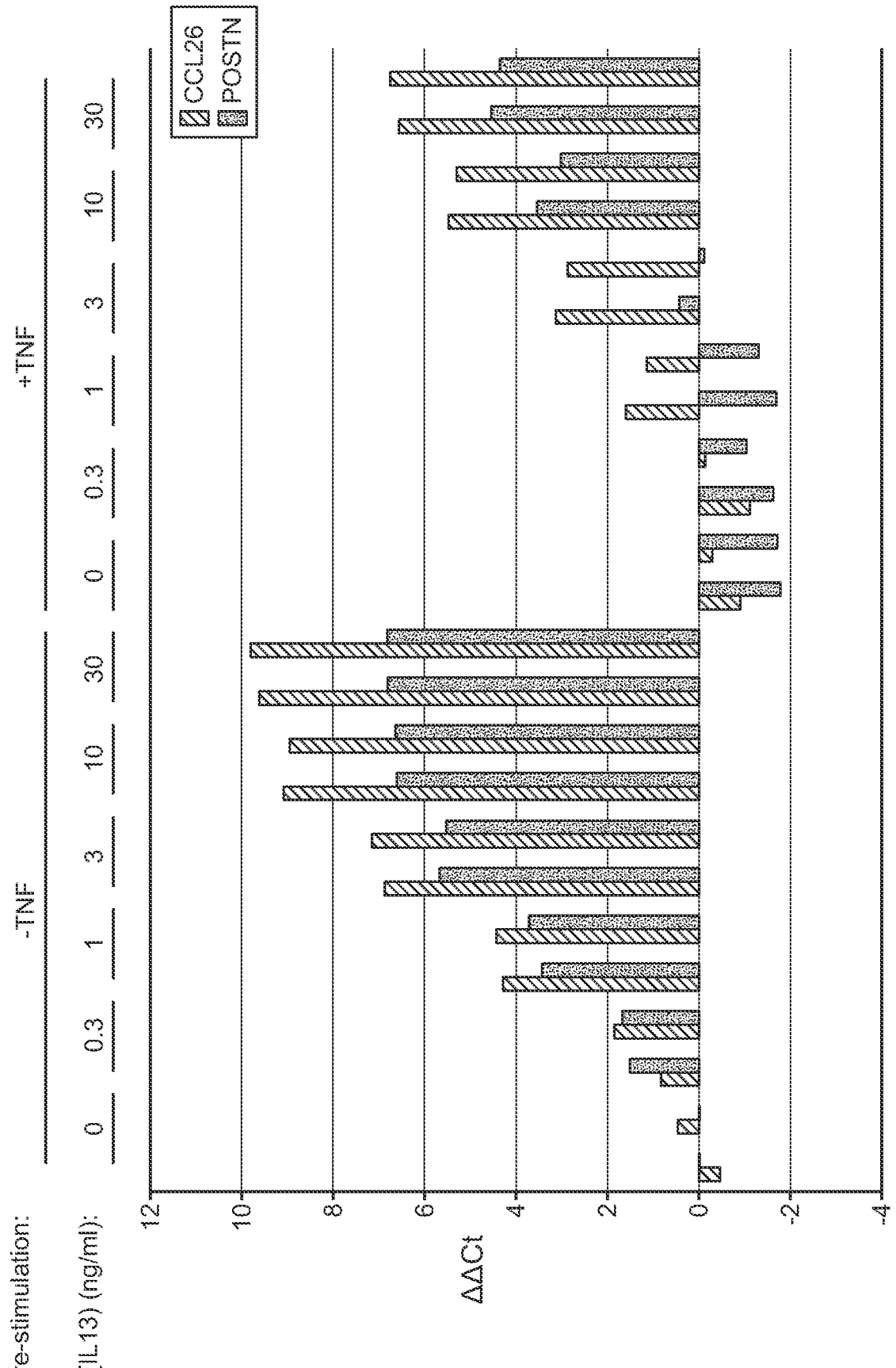

Pre-stimulation:
-TNF
+TNF
[IL13) (ng/ml):
0
0.3
1
3
10
30
0
0.3
1
3
10
30
CCL26
POSTN
ΔΔCt
12
10
8
6
4
2
0
-2
-4

FIG. 11B

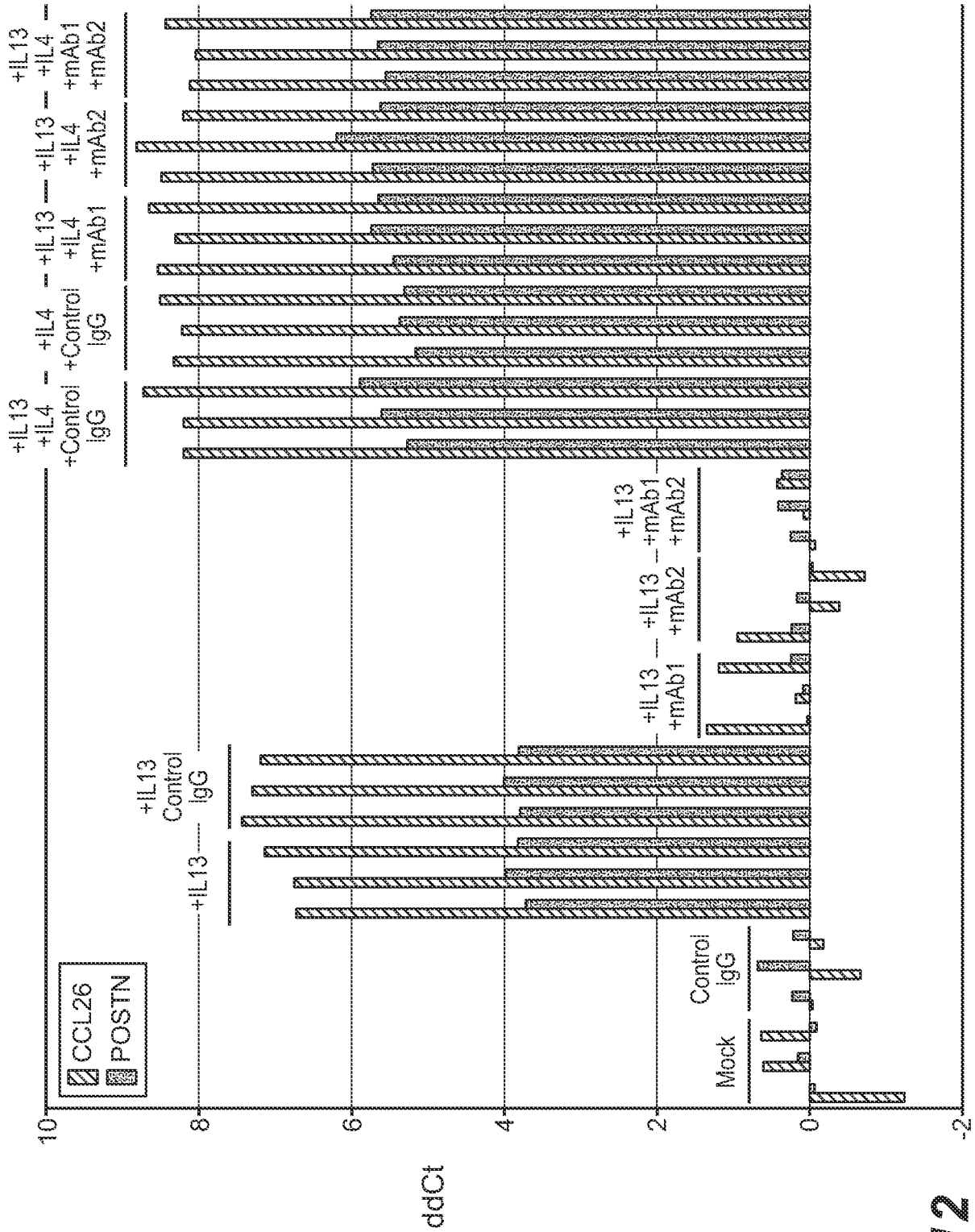

CCL26
POSTN
ddCt
10
8
6
4
2
0
-2
Mock
Control IgG
+IL13
+IL13 Control IgG
+IL13 +mAb1
+IL13 +mAb2
+IL13 +mAb1 +mAb2
+IL13 +IL4 +Control IgG
+IL13 +IL4 +mAb1
+IL13 +IL4 +mAb2
+IL13 +IL4 +mAb1 +mAb2

FIG. 12

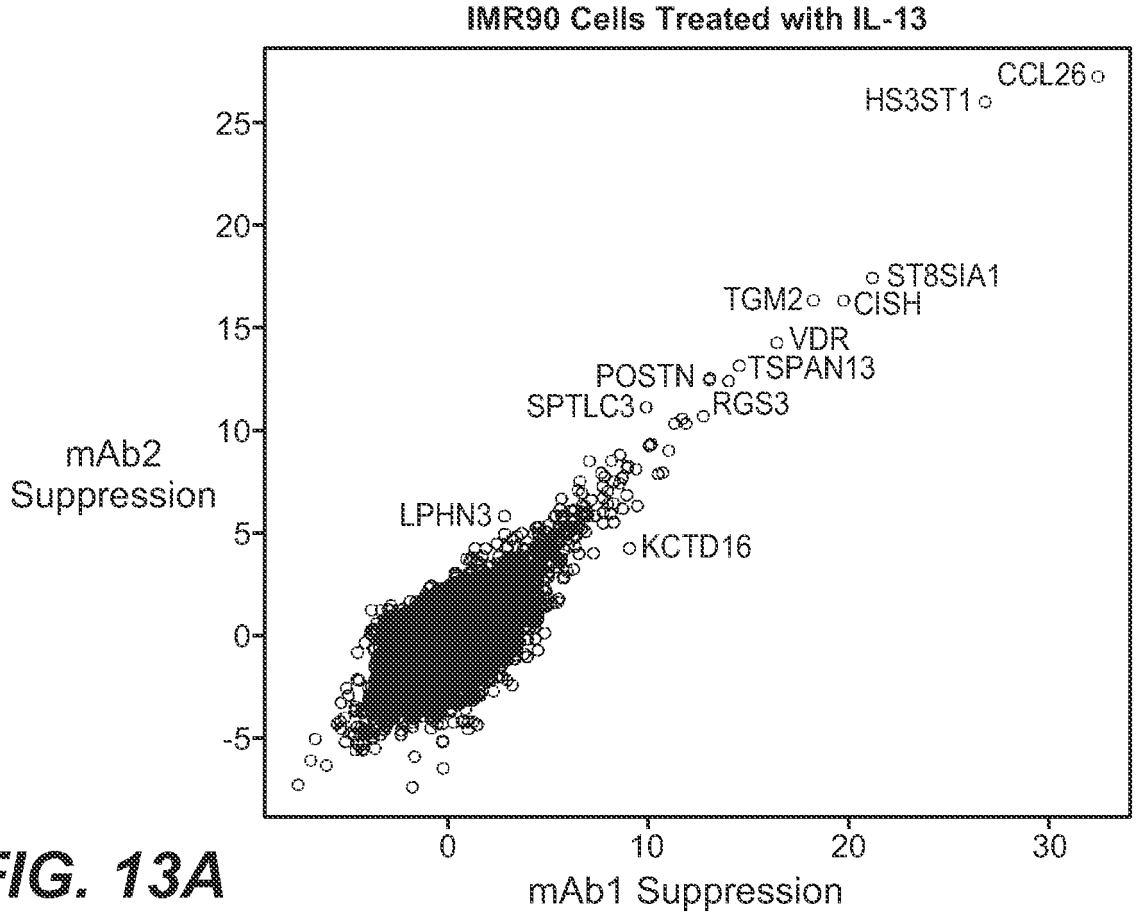


FIG. 13A

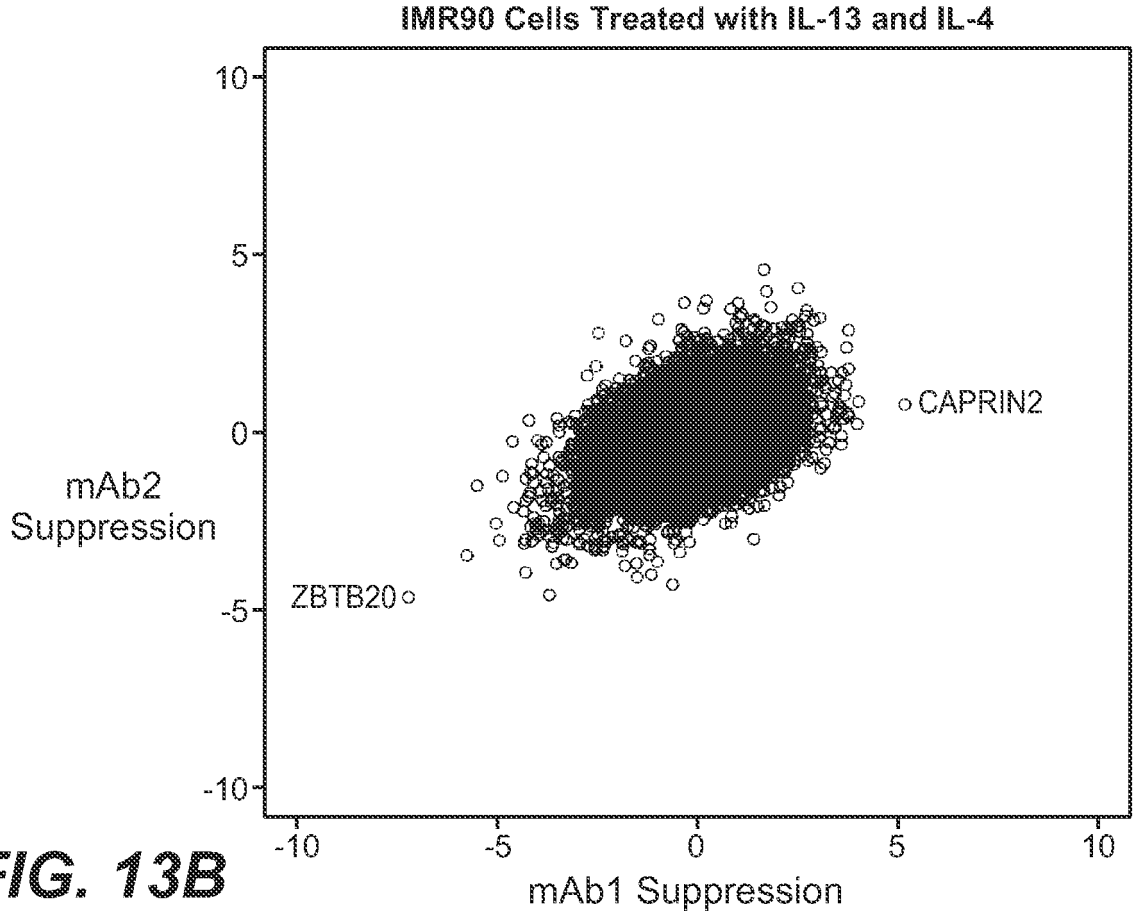


FIG. 13B

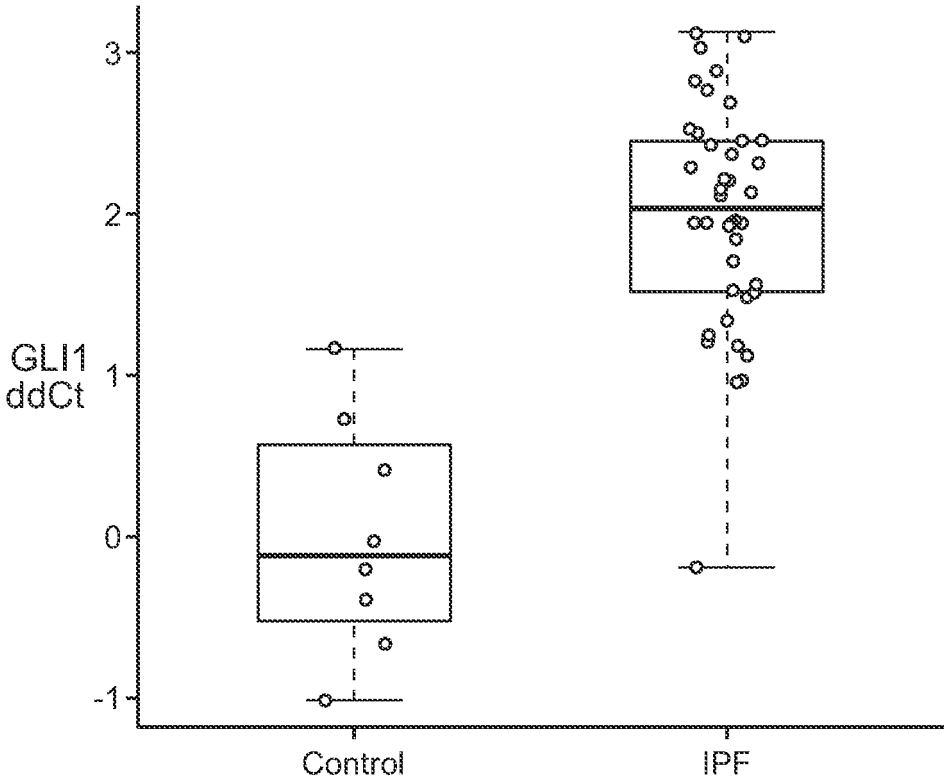

GLI1
ddCt
3
2
1
0
-1
Control
IPF

FIG. 14

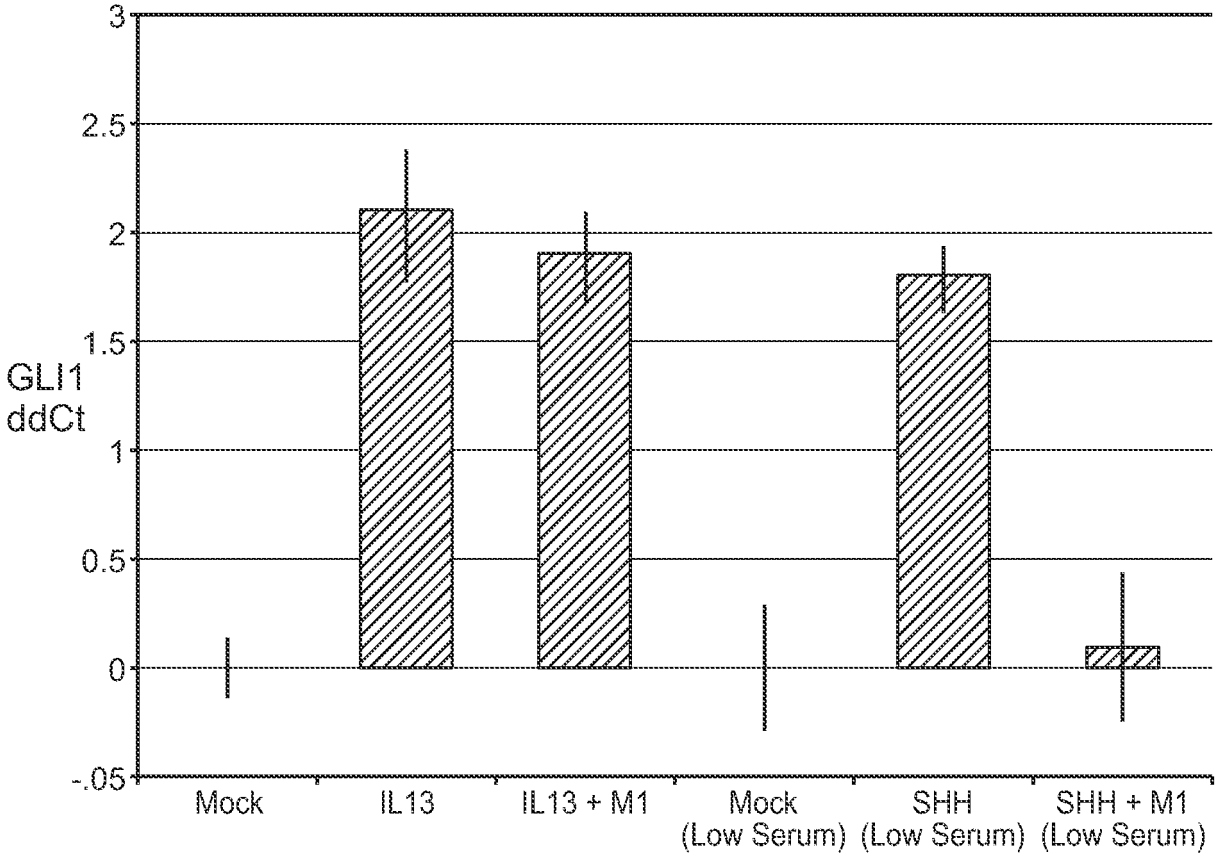

GLI1
ddCt
3
2.5
2
1.5
1
0.5
0
-.05
Mock
IL13
IL13 + M1
Mock
(Low Serum)
SHH
(Low Serum)
SHH + M1
(Low Serum)

FIG. 15

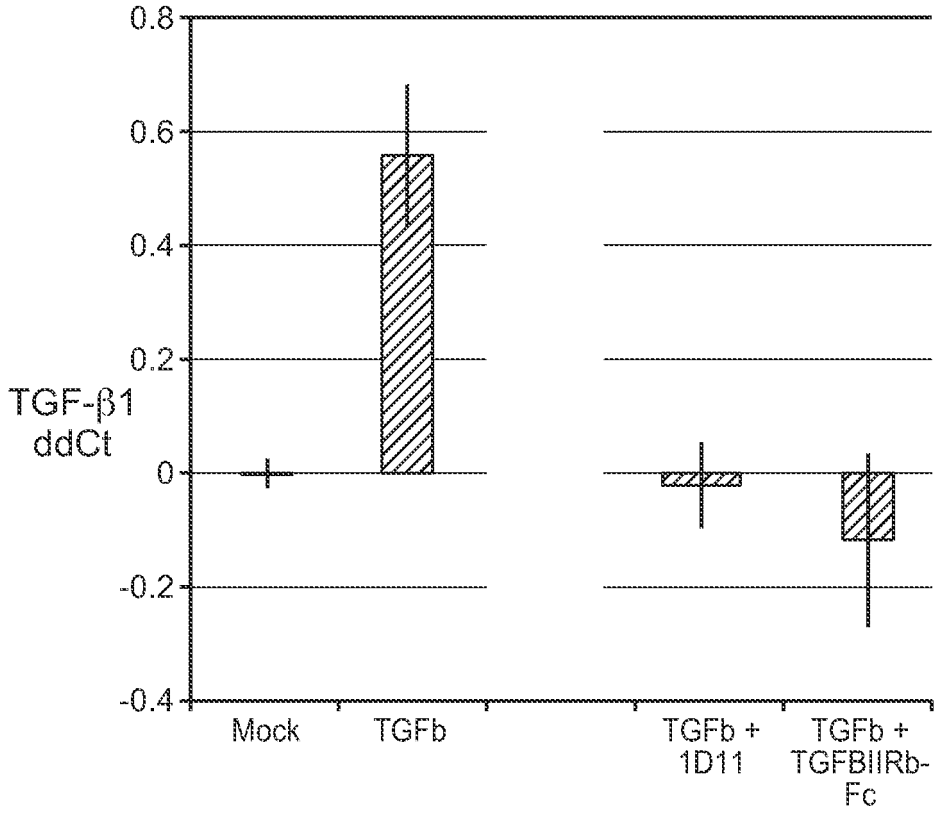


FIG. 16A

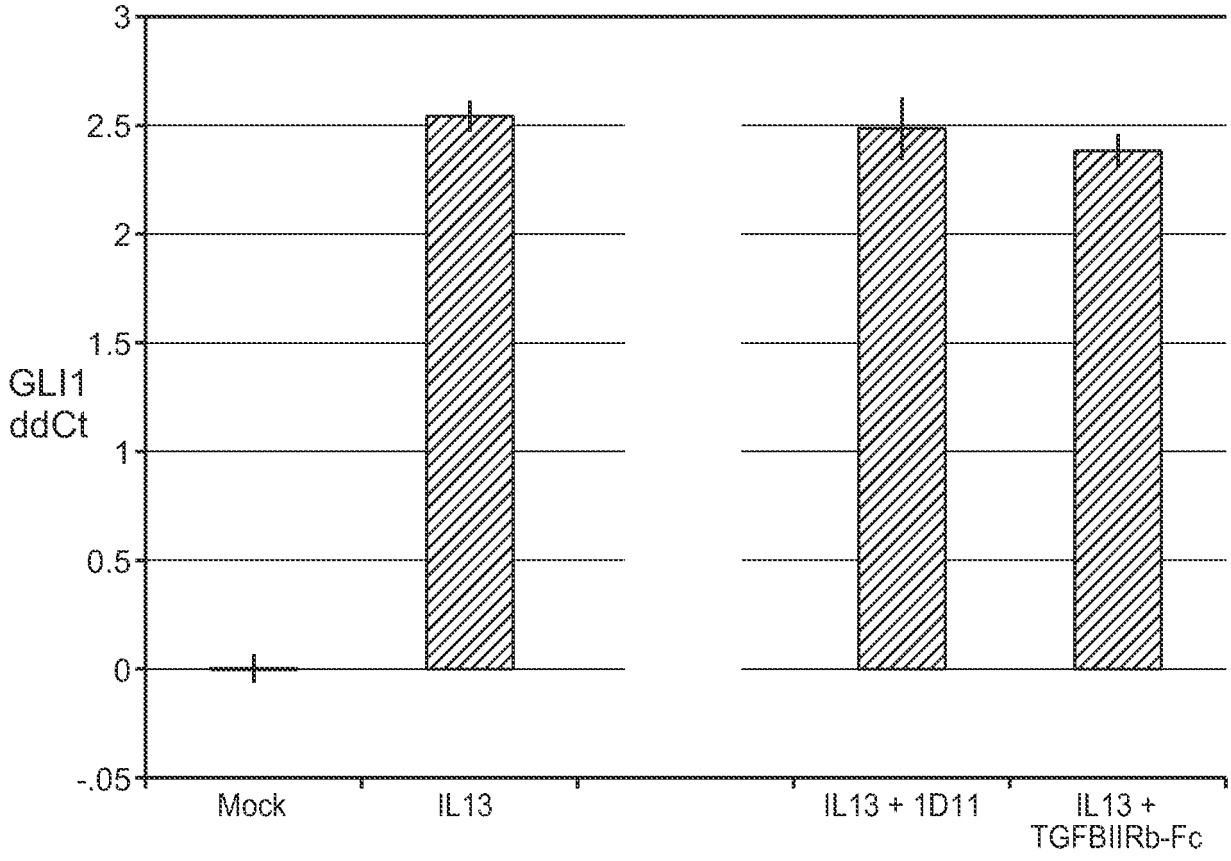


FIG. 16B