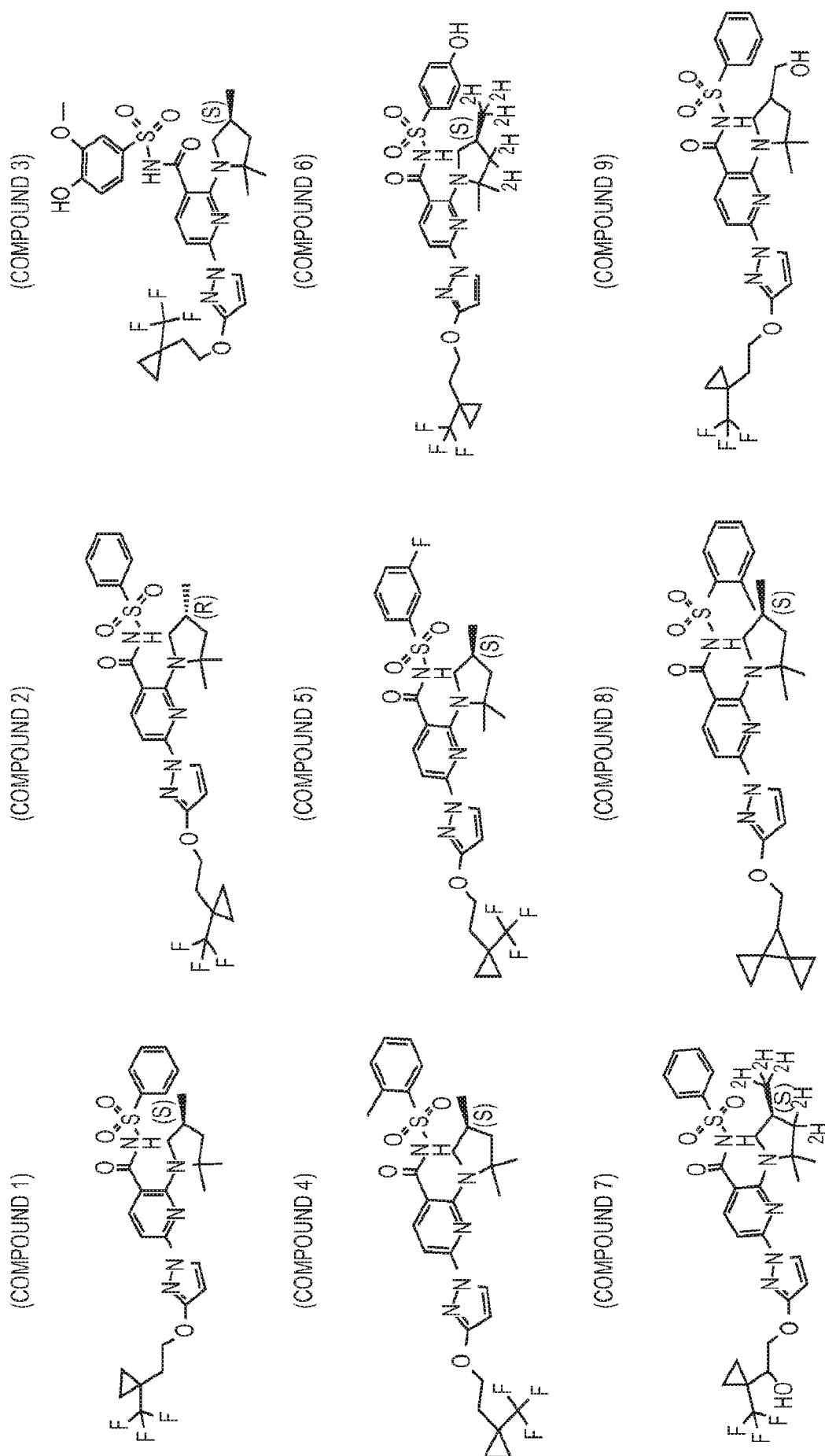
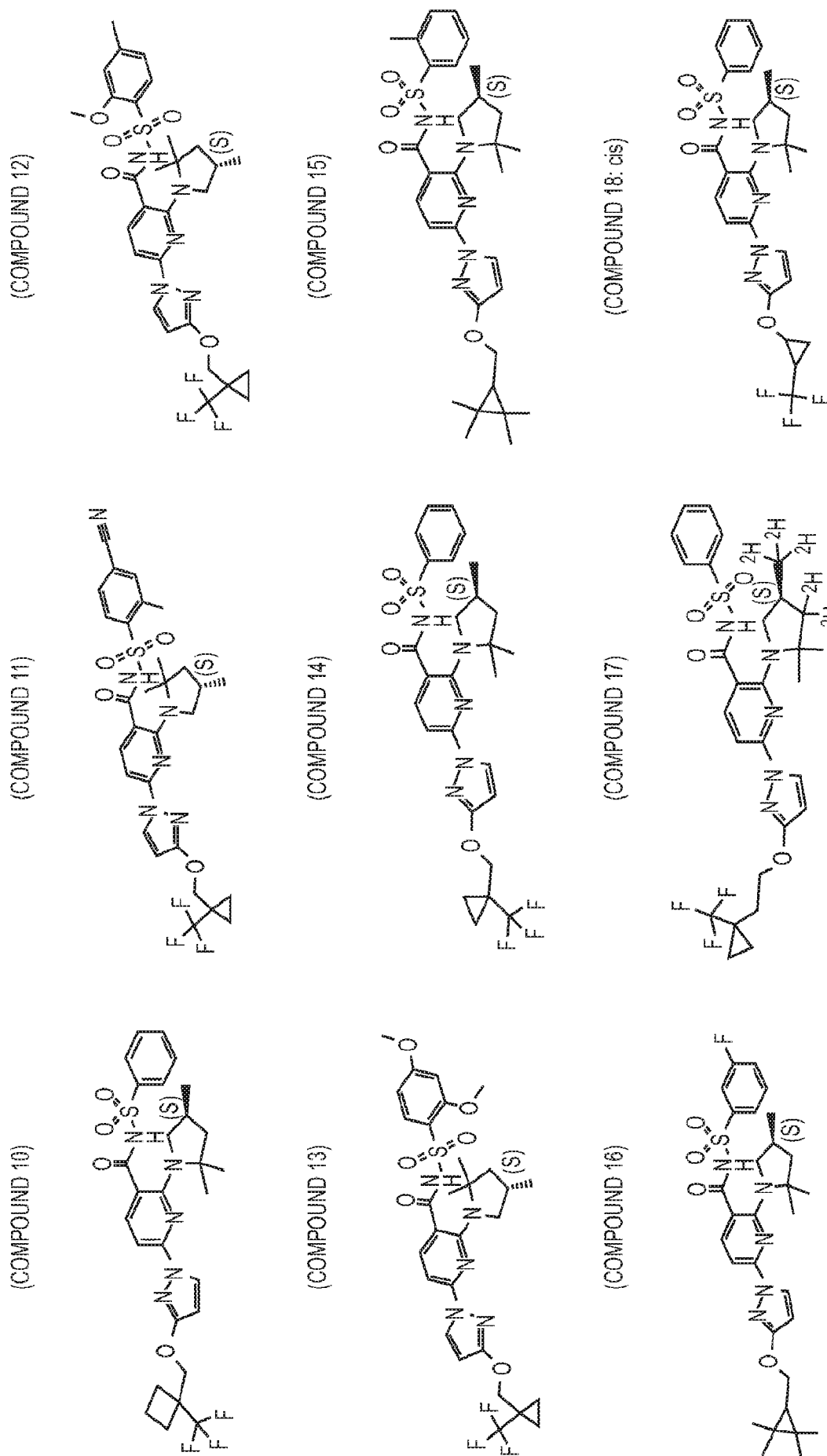
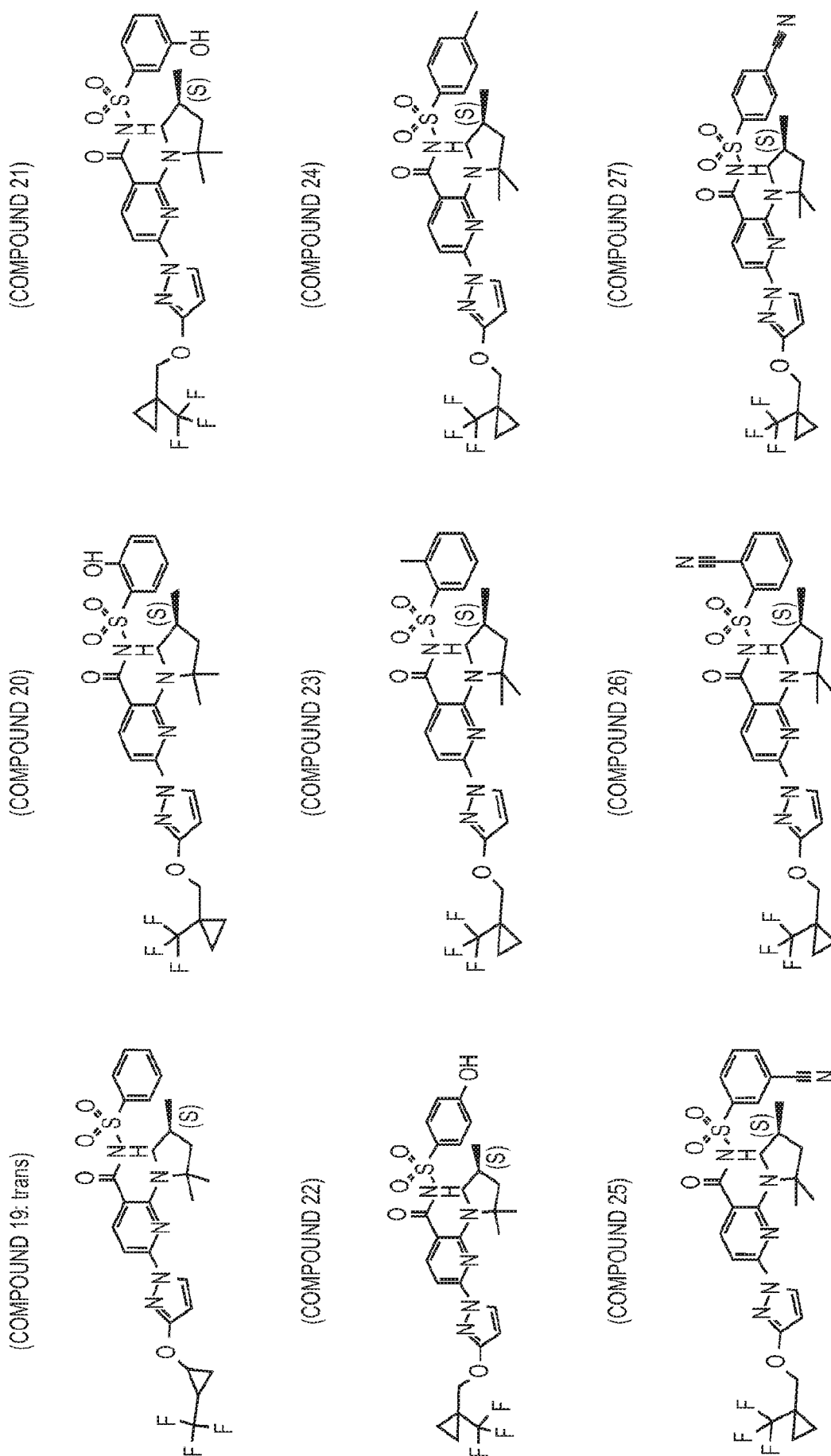
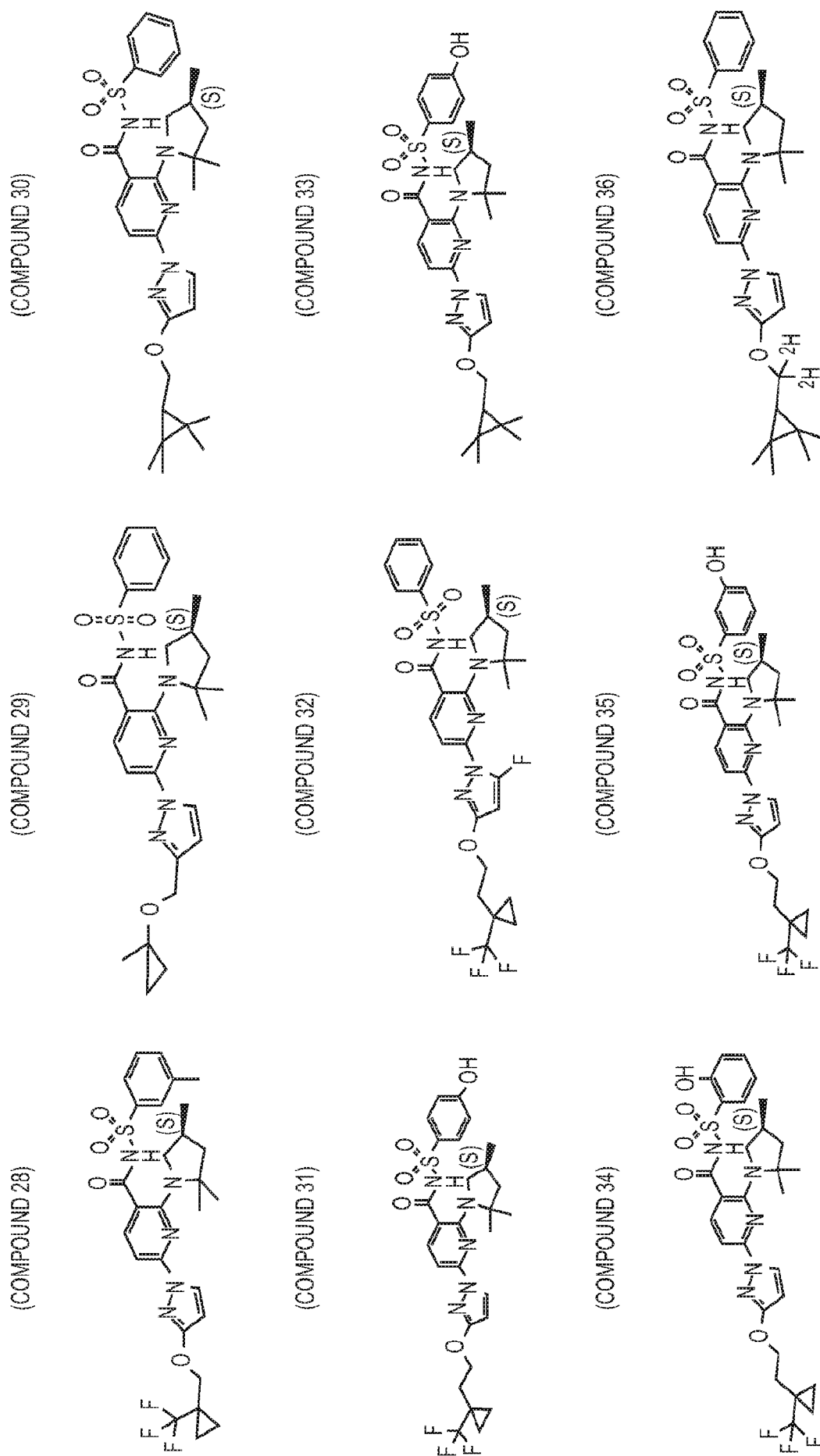
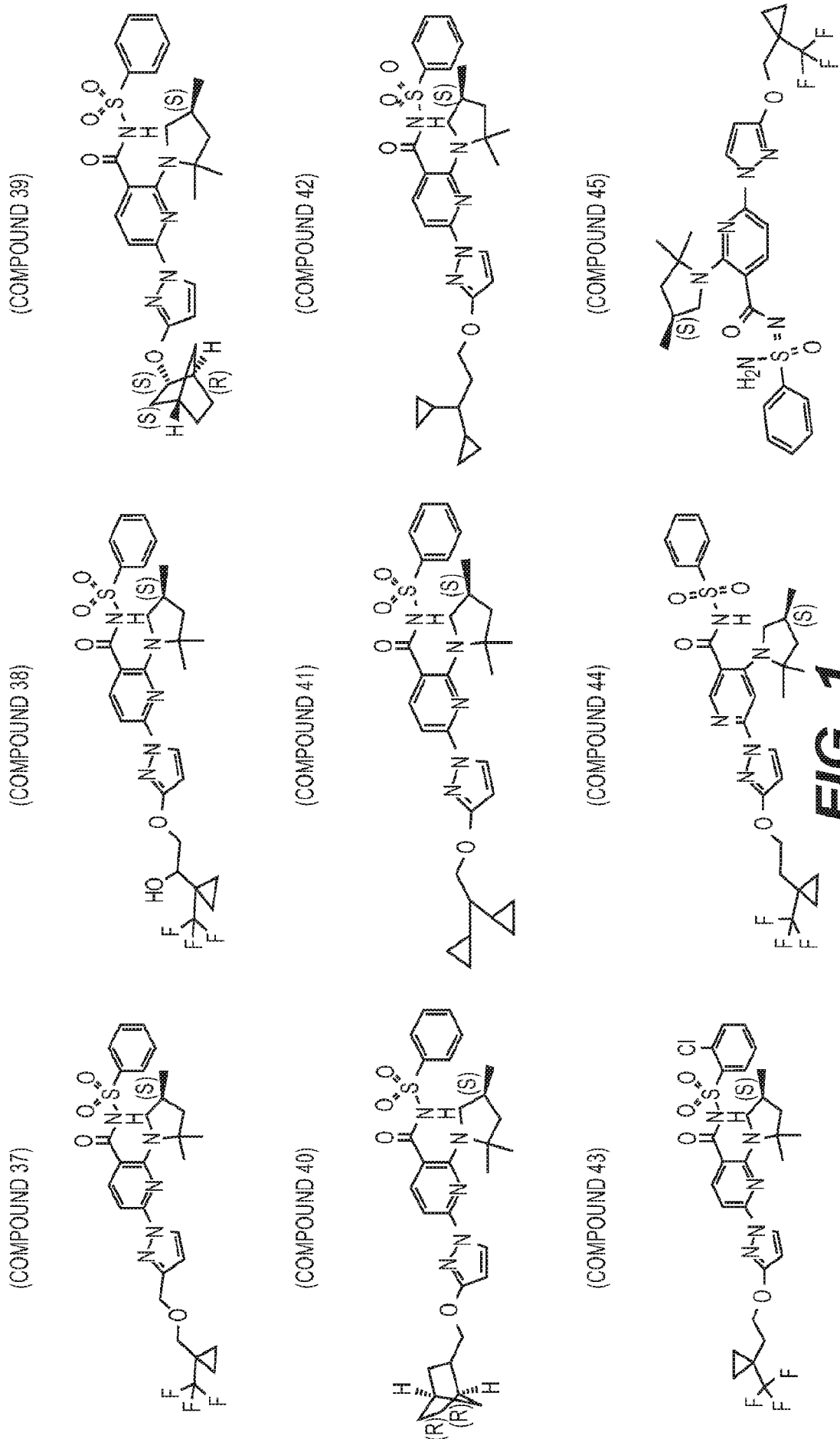


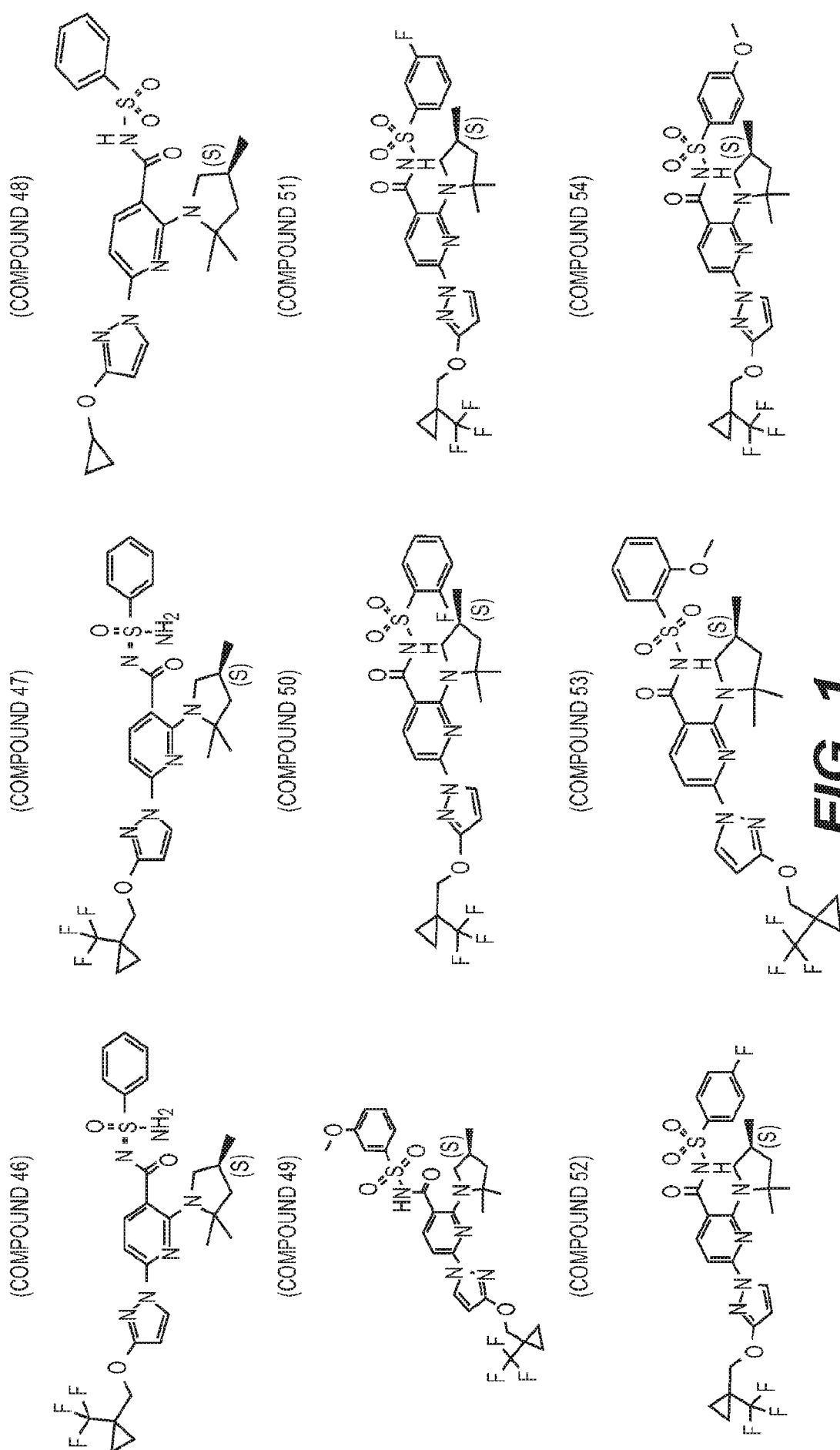


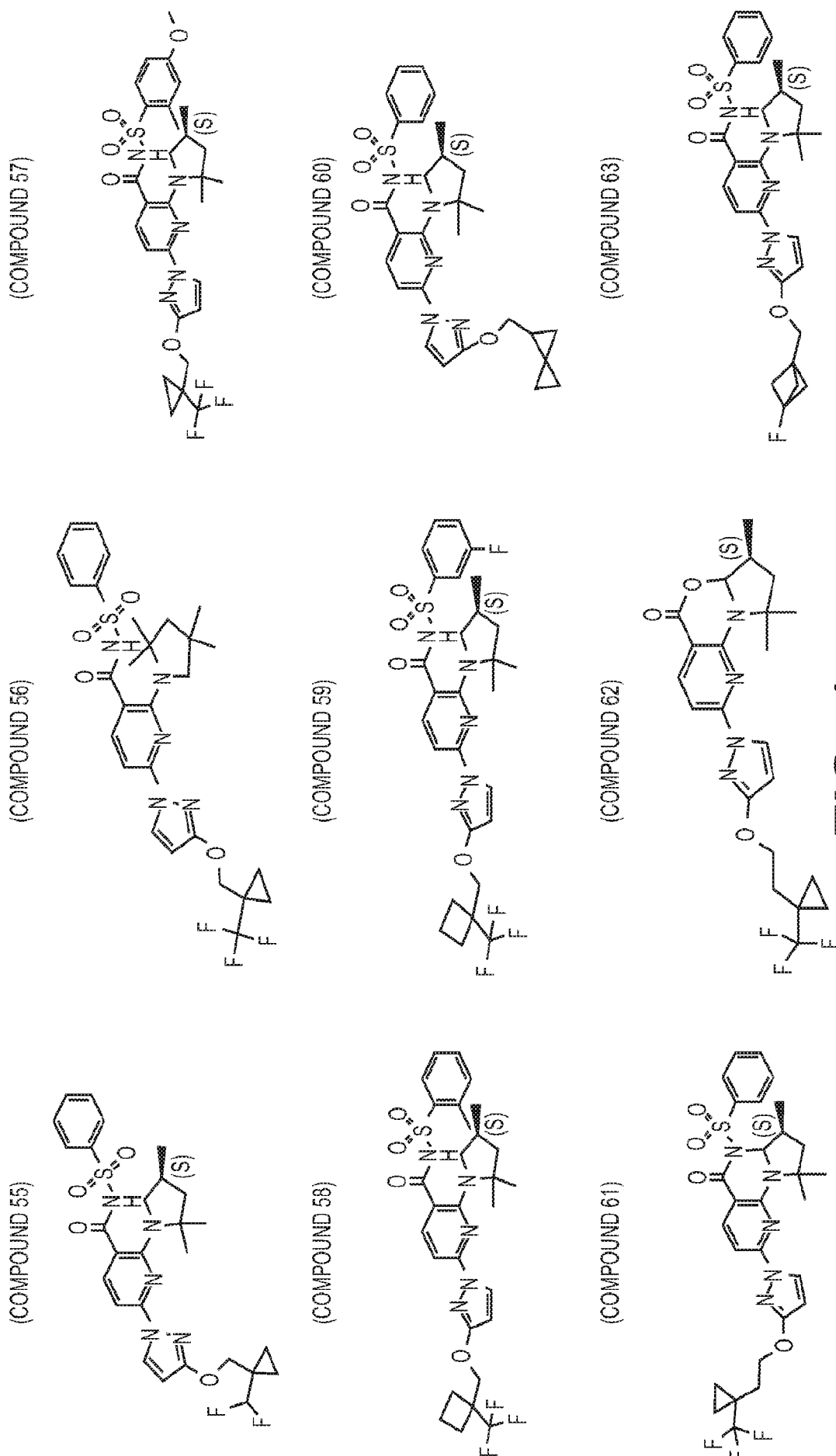
**FIG. 1**

**FIG. 1**

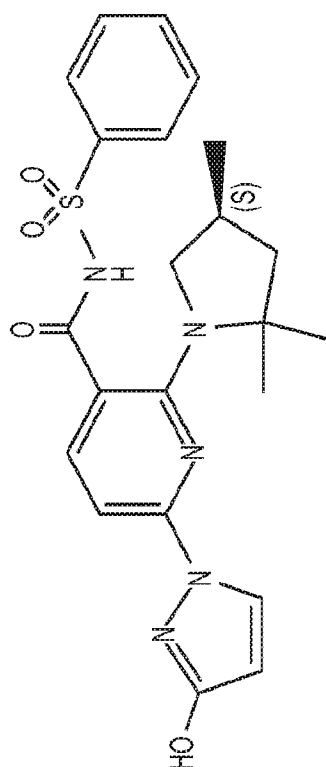
**FIG. 1**

**FIG. 1**

**FIG. 1**



(COMPOUND 65)



(COMPOUND 64)

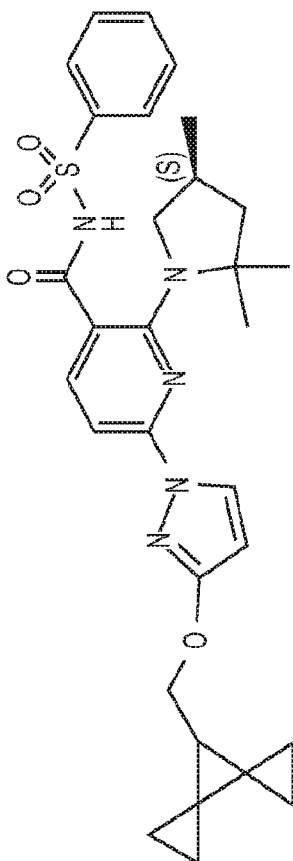
**FIG. 1**

FIG. 2

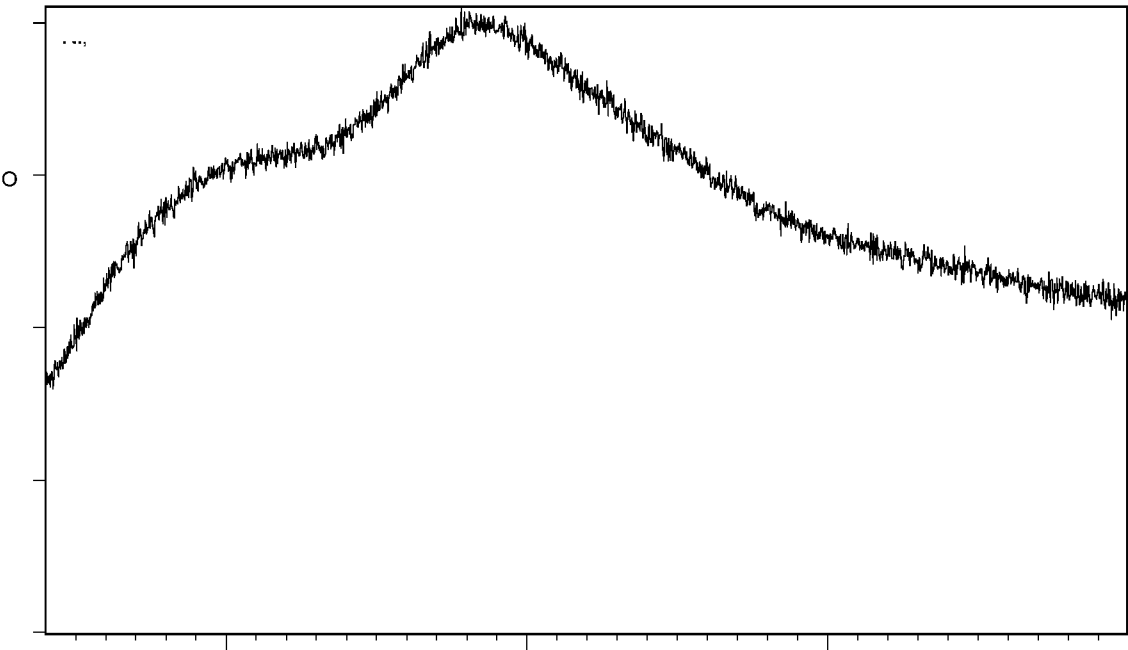


FIG. 3

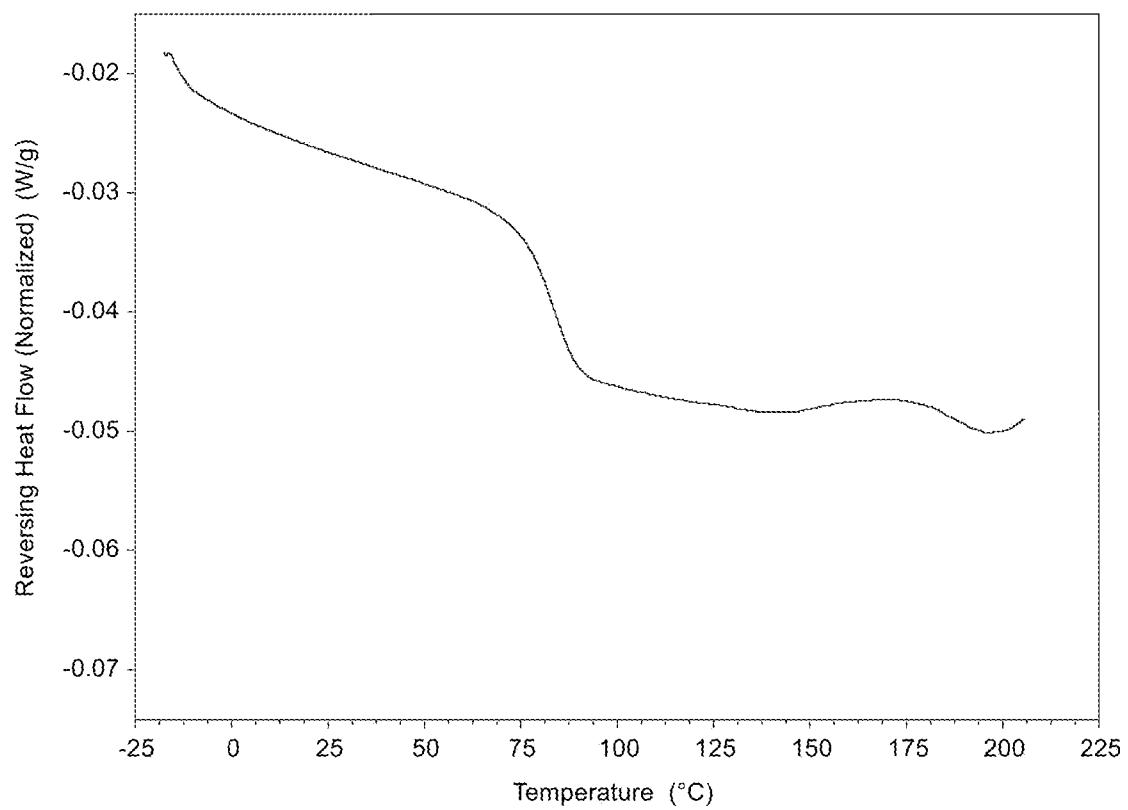


FIG. 4

Mutation cDNA Name	Mutation Protein Name	Mutation Legacy Name
c.1A>G	p.? (unknown)	M1V
c.54-5940_273+10250del21kb	p.Ser18ArgfsX16	CFTRdele2,3
c.91C>T	p.Arg31Cys	R31C
c.115C>T	p.Gln39X	Q39X
c.137C>A	p.Ala46Asp	A460
c.165-1G>A	No protein name	297-1G->A
c.166G>A	p.Glu56Lys	E56K
c.174_175insA	p.Arg59LysfsX10	306insA
c.178G>T	p.Glu60X	E60X
c.200C>T	p.Pro67Leu	P67L
c.220C>T	p.Arg74Trp	R74W
c.223C>T	p.Arg75X	R75X
c.224G>A	p.Arg75Gln	R75Q
c.254G>A	p.Gly85Glu	G85E
c.262_263delTT	p.Leu88IlefsX22	394delTT
c.273+1G>A	No protein name	405+1G->A
c.274-1G>A	No protein name	406-1G->A
c.274G>A	p.Glu92Lys	E92K
c.274G>T	p.Glu92X	E92X
c.292C>T	p.Gln98X	Q98X
c.313delA	p.Ile105SerfsX2	444delA
c.325_327delTATinsG	p.Tyr109GlyfsX4	457TAT->G
c.328G>C	p.Asp110His	D110H
c.349C>T	p.Arg117Cys	R117C
c.350G>A	p.Arg117His	R117H
c.366T>A	p.Tyr122X	Y122X
c.442delA	p.Ile148LeufsX5	574delA
c.443T>C	p.Ile148Thr	I148T
c.489+1G>T	No protein name	621+1G->T
c.531delT	p.Ile177MetfsX12	663delT
c.532G>A	p.Gly178Glu	G178R

Mutation cDNA Name	Mutation Protein Name	Mutation Legacy Name
c.543_546delTAGT	p.Leu183PhefsX5	675del4
c.579+1G>T	No protein name	711+1G->T
c.579+3A>G	No protein name	711+3A->G
c.579+5G>A	No protein name	711+5G->A
c.580-1G>T	No protein name	712-1G->T
C.59SOT	p.His199Tyr	H199Y
C.613CM	p.Pro205Ser	P205S
c.617T>G	p.Leu20GTrp	L206W
C.6580T	p.Gln220X	Q220X
c.580T>G	p.Leu227Arg	L227R
c.720_741delAGGG AGAATGATGATGAA GTAC	p.Gly241GluX13	852del22
c.828C>A	p.Cys276X	C276X
c.948delT	p.Phe316LeufsX1 2	1078delT
c.988G>T	p.Gly330X	G330X
c.1000C>T	p.Arg334Trp	R334W
c.1007T>A	p.Ile336Lys	I336K
c.1013C>T	p.Thr338Ele	T338I
c.1021T>C	p.Ser341Pro	S341P
c.1022_1023insTC	p.Phe342HisfsX2 8	1154insTC
c.1040G>A	p.Arg347His	R347H
c.1040G>C	p.Arg347Pro	R347P
c.1055G>A	p.Arg352Gln	R352Q
c.[1075C>A; 10799C> A]	p.[Gln359Lys,Thr 360Lys]	Q359K/T360K
c.1081delT	p.Trp361GlyfsX8	1213delT
c.1116+1G>A	No protein name	1248+1G->A
c.1127_1128insA	p.Gln378AlafsX4	1259insA
c.1153_1154insAT	p.Asn386IlefsX3	1288insTA
c.1202G>A c.1203G>A	or p.Trp401X	W401X
c.1209+1G>A	No protein name	1341+1G->A
c.1210-12[5]	No protein name	5T

Mutation cDNA Name	Mutation Protein Name	Mutation Legacy Name
c.1210-12(7]	No protein name	7T
c.1240C>T	p.Gln414X	Q414X
c.1329_1330insAGAT	p.Ile444ArgfsX3	1461ins4
c.1340delA	p.Lys447ArgfsX2	1471delA
c.1364C>A	p.Ala455Glu	A455E
c.1393-1G>A	No protein name	1525-1G->A
c.1397C>A c.1397C>G	or p.Ser466X	S466X
c.1400T>C	p.Leu467Pro	L467P
c.1408A>G	p.Met470Val	M470V
c.1418delG	p.Gly473Glu fsX54	1548delG
c.1466C>A	p.Ser489X	S489X
c.1475C>T	p.Ser492Phe	S492F
c.1477C>T	p.Gln493X	Q493X
c.1519_1521delATC	' p.Ile507del	I507del
c.1521_1523delCTT	p.Phe508del	F508del
c.1545_1546delTA	p.Tyr515X	1677delTA
c.1558G>T	p.Val520Phe	V520F
c.1573C>T	p.Gln525X	Q525X
c.1585-8G>A	No protein name	1717-8G->A
c.1585-1G>A	No protein name	1717-1G->A
c.1624G>T	p.Gly542X	G542X
c.1645A>C c.1G47T>G	or p.Ser549Arg	S549R
c.1646G>A	p.Ser549Asn	S549N
c.1650delA	p.Gly551Val fsX8	1782delA
c.1651G>A	p.Gly551Ser	G551S
c.1652G>A	p.Gly551Asp	G551D
c.1654C>T	p.Gln552X	Q552X
c.1657C>T	p.Arg553X	R553X
c.1673T>C	p.Leu558Ser	L558S
c.1675G>A	p.Ala559Thr	A559T

Mutation cDNA Name	Mutation Protein Name	Mutation Legacy Name
c.1679G>A	p.Arg560Lys	R560K
c.1679G>C	p.Arg560Thr	R560T
c.1679+1G>C	No protein name	1811+1G->C
c.1679+1.6kbA>G	No protein name	1811+1.6kbA->G
c.1680-1G>A	No protein name	1812-1G->A
c.1682C>A	p.Ala561Glu	A561E
c.1692delA	p.Asp565MetfsX7	1824delA
c.1705T>G	p.Tyr569Asp	Y569D
c.1727G>C	p.Gly576Ala	G576A
c.1736A>G	p.Asp579Gly	D579G
c.1753G>T	p.Glu585X	E585X
c.1766+1G>A	No protein name	1898+1G->A
c.1766+1G>C	No protein name	1898+1G->C
c.1766+3A>G	No protein name	1898+3A->G
c.1841A>G	p.Asp614Gly	D614G
c.1923_1931del9ins	p.Ser641ArgfsX5	2055del9->A
c.1973_1985del13insAGAAA	p.Arg658LysfsX4	2105-2117del13insAGAAA
c.1986_1989delAACT	p.Thr663ArgfsX8	2118del4
c.2002C>T	p.Arg668Cys	R668C
c.2012delT	p.Leu671X	2143delT
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G+
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183delAA->G#
c.2052_2053insA	p.Gln685ThrfsX4	2184insA
c.2052delA	p.Lys684AsnfsX38	2184delA
c.21250T	p.Arg709X	R709X
c.2128A>T	p.Lys710X	K710X
c.2175_2176insA	p.Glu726ArgfsX4	2307insA
c.2195T>G	p.Leu732X	L732X
c.2215delG	p.Val739TyrfsX16	2347delG

Mutation cDNA Name	Mutation Protein Name	Mutation Legacy Name
c.2260G>A	p.Val754Met	V754M
c.2290C>T	p.Arg764X	R764X
c.2353C>T	p.Arg785X	R785X
c.2374C>T	p.Arg792X	R792X
c.2424_2425insAT	p.Ser809IlefsX13	2556insAT
c.2453delT	p.Leu818TrpfsX3	2585delT
c.2462_2463delGT	p.Ser821ArgfsX4	No legacy name
c.2464G>T	p.Glu822X	E822X
c.2490+1G>A	No protein name	2622+1G->A
c.2491G>T	p.Glu831X	E831X
c.2537G>A c.2538G>A	or p.Trp846X	W846X
c.25470A	p.Tyr849X	Y849X
c.2551C>T	p.ArgSSIX	R851X
c.2583delT	p.Phe861LeufsX3	2711delT
c.2657+2_2657+3insA	No protein name	2789+2insA
c.2657+5G>A	No protein name	2789+5G->A
c.2658-1G>C	No protein name	2790-1G->C
c.2668C>T	p.Gln890X	Q890X
c.2735C>A	p.Ser912X	S912X
c.2737_2738insG		2869insG
c.2739T>A	p.Tyr913X	Y913X
c.2764_2765insAG	p.Val922GluX2	2896insAG
c.2780T>C	p.Leu927Pro	L927P
c.2834C>T	p.Ser945Leu	S945L
c.2875delG	p.Ala959HisfsX9	3007delG
c.2908G>C	p.Gly970Arg	G970R
c.2930C>T	p.Ser977Phe	S977F
c.2988G>A	No protein name	3120G->A
c.2988+1G>A	No protein name	3120+1G->A
c.2989-977_3367+248del	No protein name	3121-977_3499+248del2515
c.2989-1G>A	No protein name	3121-1G->A

Mutation cDNA Name	Mutation Protein Name	Mutation Legacy Name
c.2991G>C	p.Leu997Phe	L997F
c.3002_3003delTG	p.Val1001AspfsX4 5	3132delTG
c.3080T>C	p.Ile1027Thr	I1027T
c.3140-26A>G	No protein name	3272-26A->G
c.3154T>G	p.Phe1052Val	F1052V
c.3160C>G	p.His1054Asp	H1054D
c.3181G>C	p.Gly1061Arg	G1061R
c.3194T>C	p.Leu1065Pro	L1065P
c.3196C>T	p.Arg1066Cys	R1066C
c.3197G>A	p.Arg1066His	R1066H
c.3205G>A	p.Gly1069Arg	G1069R
c.3208C>T	p.Arg1070Trp	R1070W
c.3209G>A	p.Arg1070Gln	R1070Q
c.3222T>A	p.Phe1074Leu	F1074L
c.3230T>C	p.Leu1077Pro	L1077P
c.3266G>A	p.Trp1089X	W1089X
c.3276C>A c.3276C>G	or p.Tyr1092X	Y1092X
c.3302T>A	p.Met1101Lys	M1101K
c.3310G>T	p.Glu1104X	E1104X
c.3454G>C	p.Asp1152His	D1152H
c.3472C>T	p.Arg1158X	R1158X
c.3484C>T	p.Arg1162X	R1162X
c.3485G>T	p.Arg1162Leu	R1162L
c.3528delC	p.Lys1177SerfsX1 5	3659delC
c.3535_3536insTCA A	p.Thr1179IlefsX1 7	3667ins4
c.3587C>G	p.Ser1196X	S1196X
c.3605delA	p.Asp1202AlafsX 9	3737delA
c.3611G>A c.3612G>A	or p.Trp1204X	W1204X
c.3659delC	p.Thr1220LysfsX8	3791delC

Mutation cDNA Name	Mutation Protein Name	Mutation Legacy Name
c.3691delT	p.Ser1231ProfsX4	3821delT
c.3700A>G	p.Ile1234Val	11234V
c.3705T>G	p.Ser1235Arg	S1235R
c.3717+12191C>T	No protein name	3849+10kbC->T
c.371S-1G>A	No protein name	3850-1G->A
c.3731G>A	p.Gly1244Glu	G1244E
c.3744delA	p.Lys1250ArgfsX9	3876delA
c.3752G>A	p.Ser1251Asn	S1251N
c.3763T>C	p.Ser1255Pro	S1255P
c.37640A	p.Ser1255X	S1255X
c.3773_3774insT	p.Leu1258PhefsX7	3905insT
c.3808G>A	p.Asp1270Asn	D1270N
c.3346G>A	p.Trp1282X	W1282X
c.3873+1G>A	No protein name	4005+1G->A
c.3883delA	p.Ile1295PhefsX3	4015delA
c.3884_3885insT	p.Ser1297PhefsX5	4016insT
c.3909C>G	p.Asn1303Lys	N1303K
c.3937C>T	p.Gln1313X	Q1313X
c.3964-78_4242+577del	NULL	CFTRdele22,23
c.4046G>A	p.Gly1349Asp	G1349D
c.4077_4080delTGT TinsAA	No protein name	4209TGTT->AA
c.4111G>T	p.Glu1371X	E1371X
c.4196_4197delTC	p.Cys1400X	4326delTC
c.4234C>T	p.Gln1412X	Q1412X
c.4242+1G>T	No protein name	4374+1G->T
c.4251delA	p.Glu1418ArgfsX14	4382delA
c.4296_4297insGA	p.Ser1435GlyfsX14	4428insGA

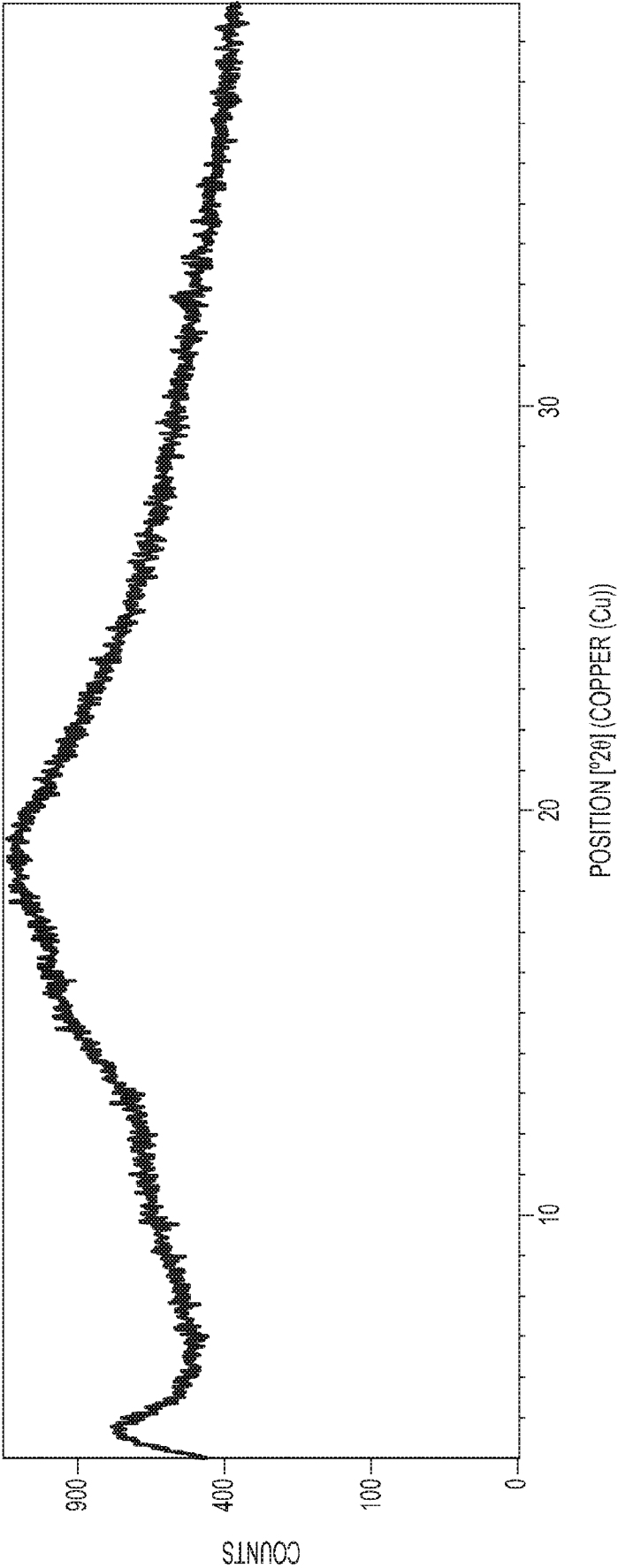


FIG. 5