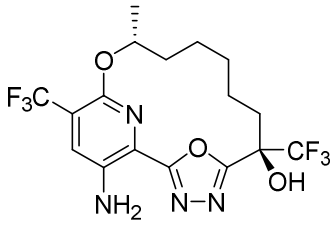
CLAIMS:

1. Compound **I**:



(Compound **I**),

or a deuterated derivative or pharmaceutically acceptable salt thereof, for use in a treatment of cystic fibrosis in a patient, wherein about 0.1 mg to about 50 mg of Compound **I** or an equivalent amount of a deuterated derivative or pharmaceutically acceptable salt thereof is to be administered to the patient daily.

2. The compound for use according to claim 1, wherein the treatment comprises about 0.25 mg to about 50 mg of Compound **I**, or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily.

3. The compound for use according to claim 1 or claim 2, wherein the treatment comprises about 0.25 mg to about 25 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily.

4. The compound for use according to any one of claims 1-3, wherein the treatment comprises about 1 mg to about 15 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily.

5. The compound for use according to any one of claims 1-4, wherein the treatment comprises about 1.5 mg to about 14 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily.

6. The compound for use according to any one of claims 1-5, wherein the treatment comprises about 4 mg to about 13 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily.

7. The compound for use according to any one of claims 1-6, wherein the treatment comprises about 8 mg to about 13 mg of Compound **I** or an equivalent amount of

a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily.

8. The compound for use according to claim 1, wherein the treatment comprises about 10 mg to about 40 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily.

9. The compound for use according to claim 1, wherein about 0.1 mg, about 0.25 mg, about 1 mg, about 1.5 mg, about 2.5 mg, about 4 mg, about 4.5 mg, about 5 mg, about 5.5 mg, about 6 mg, about 6.5 mg, about 7 mg, about 7.5 mg, about 8 mg, about 8.5 mg, about 9, about 9.5 mg, about 10 mg, about 10.5 mg, about 11 mg, about 11.5 mg, about 12 mg, about 12.5 mg, about 13 mg, about 14 mg, about 15 mg, about 17.5 mg, about 20 mg, about 22.5 mg, about 25 mg, about 27.5 mg, about 30 mg, about 32.5 mg, about 35 mg, about 37.5 mg, about 40 mg, about 42.5 mg, about 45 mg, about 47.5 mg, or about 50 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof is to be administered daily.

10. Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof for use in a treatment of cystic fibrosis in a patient comprising about 0.1 mg to about 50 mg of Compound **I** or an equivalent amount of a deuterated derivative or pharmaceutically acceptable salt thereof to be administered daily, and wherein the patient is heterozygous and has one F508del mutation.

11. The compound for use according to claim 10, wherein the treatment comprises about 0.25 mg to about 50 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily, and wherein the patient is heterozygous and has one F508del mutation.

12. The compound for use according to claim 10 or claim 11, wherein the treatment comprises about 0.25 mg to about 25 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily, and wherein the patient is heterozygous and has one F508del mutation.

13. The compound for use according to any one of claims 10-12, wherein the treatment comprises about 1 mg to about 15 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily, and wherein the patient is heterozygous and has one F508del mutation.
14. The compound for use according to any one of claims 10-13, wherein the treatment comprises about 1.5 mg to about 14 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily, and wherein the patient is heterozygous and has one F508del mutation.
15. The compound for use according to any one of claims 10-14, wherein the treatment comprises about 4 mg to about 13 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily, and wherein the patient is heterozygous and has one F508del mutation.
16. The compound for use according to any one of claims 10-15, wherein the treatment comprises about 8 mg to about 13 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily, and wherein the patient is heterozygous and has one F508del mutation.
17. The compound for use according to claim 10, wherein the treatment comprises about 10 mg to about 40 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof to be administered daily.
18. The compound for use according to claim 10, wherein about 0.1 mg, about 0.25 mg, about 1 mg, about 1.5 mg, about 2.5 mg, about 4 mg, about 4.5 mg, about 5 mg, about 5.5 mg, about 6 mg, about 6.5 mg, about 7 mg, about 7.5 mg, about 8 mg, about 8.5 mg, about 9, about 9.5 mg, about 10 mg, about 10.5 mg, about 11 mg, about 11.5 mg, about 12 mg, about 12.5 mg, about 13 mg, about 14 mg, about 15 mg, about 17.5 mg, about 20 mg, about 22.5 mg, about 25 mg, about 27.5 mg, about 30 mg, about 32.5 mg, about 35 mg, about 37.5 mg, about 40 mg, about 42.5 mg, about 45 mg, about 47.5 mg, or about 50 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable

salt thereof is to be administered daily, and wherein the patient is heterozygous and has one F508del mutation.

19. The compound for use according to any one of claims 1-18, wherein Compound **I** or a deuterated derivative or a pharmaceutically acceptable salt thereof is to be administered as a single dose, once daily.

20. The compound for use according to any one of claims 1-18, wherein Compound **I** or a deuterated derivative or a pharmaceutically acceptable salt thereof is to be administered in two doses daily.

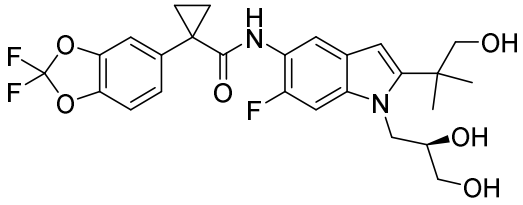
21. The compound for use according to any one of claims 1-20, wherein the treatment further comprises one or more additional therapeutic agent(s) to be administered.

22. The compound for use according to claim 21, wherein the one or more additional therapeutic agent(s) comprise(s) a compound with CFTR-modulating activity or a deuterated derivative or a pharmaceutically acceptable salt thereof.

23. The compound for use according to claim 21 or claim 22, wherein the one or more additional therapeutic agent(s) comprise(s) a CFTR corrector.

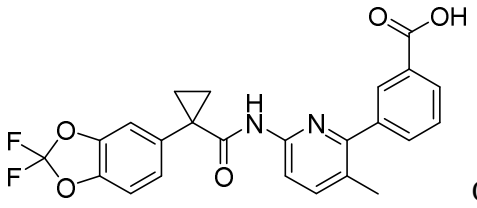
24. The compound for use according to any one of claims 21-23, wherein the one or more additional therapeutic agent(s) comprise(s) a compound selected from:

(a) (*R*)-1-(2,2-difluorobenzo[d][1,3]dioxol-5-yl)-*N*-(1-(2,3-dihydroxypropyl)-6-fluoro-2-(1-hydroxy-2-methylpropan-2-yl)-1*H*-indol-5-yl)cyclopropanecarboxamide (Compound **II**):



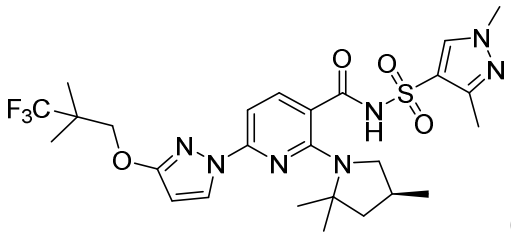
Compound **II**,

(b) 3-(6-(1-(2,2-difluorobenzo[d][1,3]dioxol-5-yl)cyclopropanecarboxamido)-3-methylpyridin-2-yl)benzoic acid (Compound **IV**):



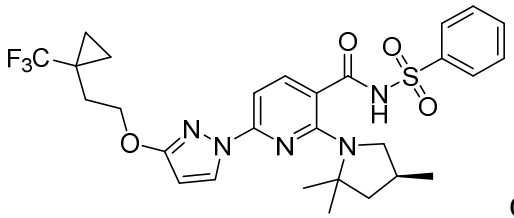
Compound **IV**,

(c) *N*-(1,3-dimethylpyrazol-4-yl)sulfonyl-6-[3-(3,3,3-trifluoro-2,2-dimethyl-propoxy)pyrazol-1-yl]-2-[(4*S*)-2,2,4-trimethylpyrrolidin-1-yl]pyridine-3-carboxamide (Compound **V**):



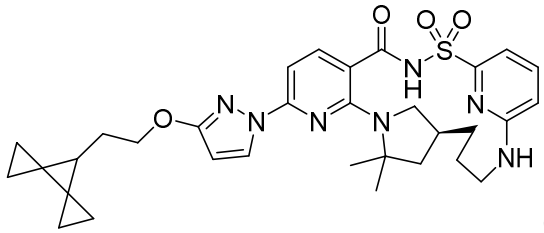
Compound **V**,

(d) *N*-(benzenesulfonyl)-6-[3-[2-[1-(trifluoromethyl) cyclopropyl]ethoxy]pyrazol-1-yl]-2-[(4*S*)-2,2,4-trimethylpyrrolidin-1-yl]pyridine-3-carboxamide (Compound **VI**):



Compound **VI**,

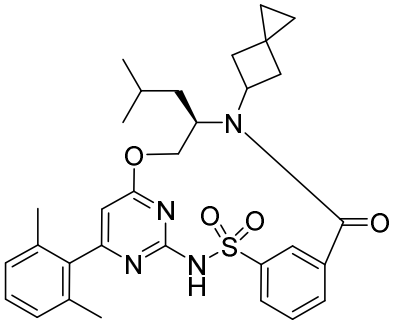
(e) (14*S*)-8-[3-(2-{dispiro[2.0.2.1]heptan-7-yl}ethoxy)-1*H*-pyrazol-1-yl]-12,12-dimethyl-2λ^6-thia-3,9,11,18,23-pentaazatetracyclo [17.3.1.111,14.05,10]tetracosa-1(22),5,7,9,19(23),20-hexaene-2,2,4-trione (Compound **VII**):



Compound **VII**,

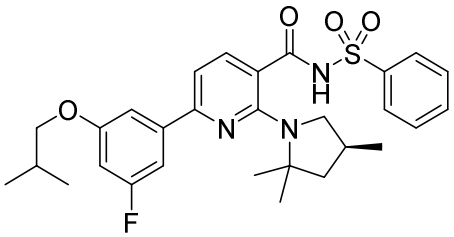
(f) (11*R*)-6-(2,6-dimethylphenyl)-11-(2-methylpropyl)-12-{spiro[2.3]hexan-5-yl}-9-oxa-2λ^6-thia-3,5,12,19-tetraazatricyclo[12.3.1.14,8]nonadeca-

1(17),4(19),5,7,14(18),15-hexaene-2,2,13-trione (Compound **VIII**):



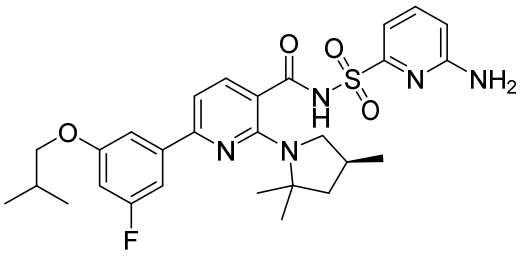
Compound **VIII**,

(g) *N*-(benzenesulfonyl)-6-(3-fluoro-5-isobutoxy-phenyl)-2-[(4*S*)-2,2,4-trimethylpyrrolidin-1-yl]pyridine-3-carboxamide (Compound **IX**):



Compound **IX**,

(h) *N*-[(6-amino-2-pyridyl)sulfonyl]-6-(3-fluoro-5-isobutoxy-phenyl)-2-[(4*S*)-2,2,4-trimethylpyrrolidin-1-yl]pyridine-3-carboxamide (Compound **X**):

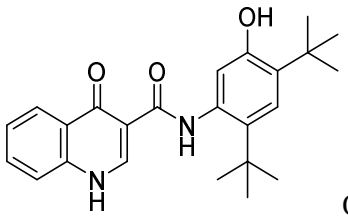


Compound **X**, and deuterated derivatives and pharmaceutically acceptable salts thereof.

25. The compound for use according to any one of claims 21-24, wherein the one or more additional therapeutic agent(s) comprise(s) a CFTR potentiator.

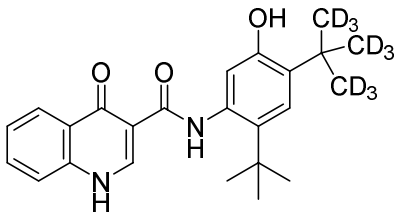
26. The compound for use according to any one of claims 21-25, wherein the one or more additional therapeutic agent(s) comprise(s) a compound selected from:

(a) *N*-(5-hydroxy-2,4-di-*tert*-butyl-phenyl)-4-oxo-1*H*-quinoline-3-carboxamide (Compound **III**):



Compound **III**,

(b) *N*-(2-(*tert*-butyl)-5-hydroxy-4-(2-(methyl-d3)propan-2-yl-1,1,1,3,3,3-d6)phenyl)-4-oxo-1,4-dihydroquinoline-3-carboxamide (Compound **III-d**):



Compound **III-d**, and deuterated derivatives and pharmaceutically acceptable salts thereof.

27. A pharmaceutical composition comprising about 0.1 mg to about 50 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof.

28. The pharmaceutical composition according to claim 27, wherein the composition comprises about 0.25 mg to about 50 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof.

29. The pharmaceutical composition according to claim 27 or claim 28, wherein the composition comprises about 0.25 mg to about 25 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof.

30. The pharmaceutical composition according to any one of claims 27-29, wherein the composition comprises about 1 mg to about 15 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof.

31. The pharmaceutical composition according to any one of claims 27-30, wherein the composition comprises about 1.5 mg to about 14 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof.

32. The pharmaceutical composition according to any one of claims 27-31, wherein the composition comprises about 4 mg to about 13 mg of Compound **I** or an

equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof.

33. The pharmaceutical composition according to any one of claims 27-32, wherein the composition comprises about 8 mg to about 13 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof.

34. The pharmaceutical composition according to claim 27, wherein the composition comprises about 10 mg to about 40 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof.

35. The pharmaceutical composition according to claim 27, wherein the composition comprises about 0.1 mg, about 0.25 mg, about 1 mg, about 1.5 mg, about 2.5 mg, about 4 mg, about 4.5 mg, about 5 mg, about 5.5 mg, about 6 mg, about 6.5 mg, about 7 mg, about 7.5 mg, about 8 mg, about 8.5 mg, about 9, about 9.5 mg, about 10 mg, about 10.5 mg, about 11 mg, about 11.5 mg, about 12 mg, about 12.5 mg, about 13 mg, about 14 mg, about 15 mg, about 17.5 mg, about 20 mg, about 22.5 mg, about 25 mg, about 27.5 mg, about 30 mg, about 32.5 mg, about 35 mg, about 37.5 mg, about 40 mg, about 42.5 mg, about 45 mg, about 47.5 mg, or about 50 mg of Compound **I** or an equivalent amount of a deuterated derivative or a pharmaceutically acceptable salt thereof.

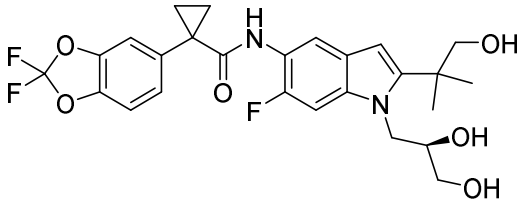
36. The pharmaceutical composition according to any one of claims 27-35, wherein the composition further comprises one or more additional therapeutic agent(s).

37. The pharmaceutical composition according to claim 36, wherein the one or more additional therapeutic agent(s) comprise(s) a compound with CFTR-modulating activity or a deuterated derivative or a pharmaceutically acceptable salt thereof.

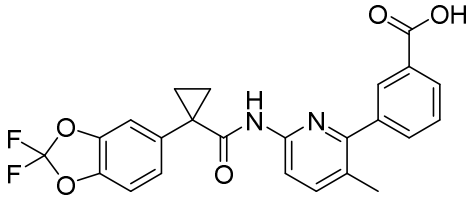
38. The pharmaceutical composition according to claim 36 or claim 37, wherein the one or more additional therapeutic agent(s) comprise(s) a CFTR corrector.

39. The pharmaceutical composition according to any one of claims 36-38, wherein the one or more additional therapeutic agent(s) comprise(s) a compound selected from:

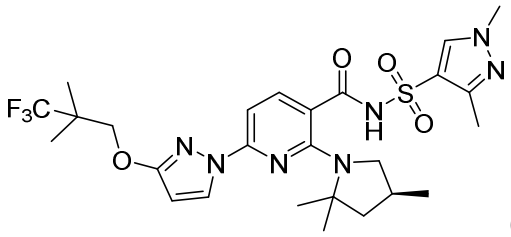
(a) (*R*)-1-(2,2-difluorobenzo[d][1,3]dioxol-5-yl)-*N*-(1-(2,3-dihydroxypropyl)-6-fluoro-2-(1-hydroxy-2-methylpropan-2-yl)-1*H*-indol-5-yl)cyclopropanecarboxamide (Compound **II**):


Compound **II**,

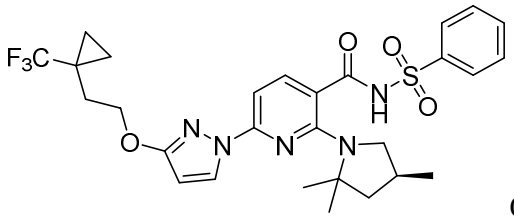
(b) 3-(6-(1-(2,2-difluorobenzo[d][1,3]dioxol-5-yl)cyclopropanecarboxamido)-3-methylpyridin-2-yl)benzoic acid (Compound **IV**):


Compound **IV**,

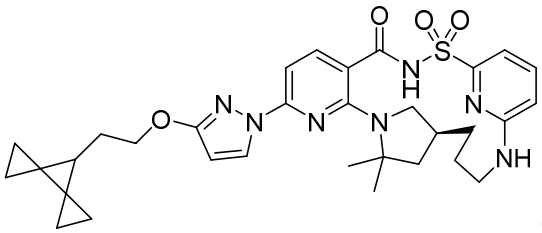
(c) *N*-(1,3-dimethylpyrazol-4-yl)sulfonyl-6-[3-(3,3,3-trifluoro-2,2-dimethyl-propoxy)pyrazol-1-yl]-2-[(4*S*)-2,2,4-trimethylpyrrolidin-1-yl]pyridine-3-carboxamide (Compound **V**):


Compound **V**,

(d) *N*-(benzenesulfonyl)-6-[3-[2-[1-(trifluoromethyl) cyclopropyl]ethoxy]pyrazol-1-yl]-2-[(4*S*)-2,2,4-trimethylpyrrolidin-1-yl]pyridine-3-carboxamide (Compound **VI**):

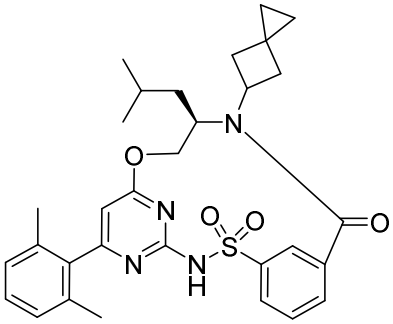

Compound **VI**,

(e) (14*S*)-8-[3-(2-{dispiro[2.0.2.1]heptan-7-yl}ethoxy)-1*H*-pyrazol-1-yl]-12,12-dimethyl-2λ^6-thia-3,9,11,18,23-pentaazatetracyclo [17.3.1.111,14.05,10]tetracosa-1(22),5,7,9,19(23),20-hexaene-2,2,4-trione (Compound **VII**):



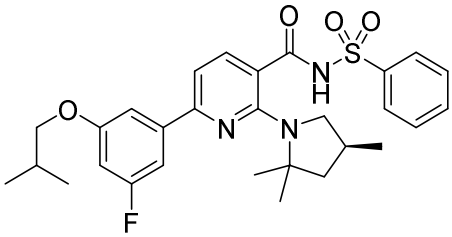
Compound **VII**,

(f) (11*R*)-6-(2,6-dimethylphenyl)-11-(2-methylpropyl)-12-{spiro[2.3]hexan-5-yl}-9-oxa-2λ^6-thia-3,5,12,19-tetraazatricyclo[12.3.1.14,8]nonadeca-1(17),4(19),5,7,14(18),15-hexaene-2,2,13-trione (Compound **VIII**):



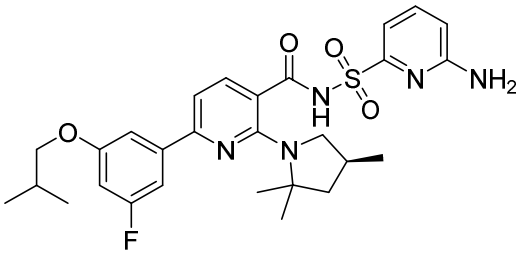
Compound **VIII**,

(g) *N*-(benzenesulfonyl)-6-(3-fluoro-5-isobutoxy-phenyl)-2-[(4*S*)-2,2,4-trimethylpyrrolidin-1-yl]pyridine-3-carboxamide (Compound **IX**):



Compound **IX**,

(h) *N*-[(6-amino-2-pyridyl)sulfonyl]-6-(3-fluoro-5-isobutoxy-phenyl)-2-[(4*S*)-2,2,4-trimethylpyrrolidin-1-yl]pyridine-3-carboxamide (Compound **X**):



Compound **X**, and deuterated derivatives and pharmaceutically acceptable salts thereof.

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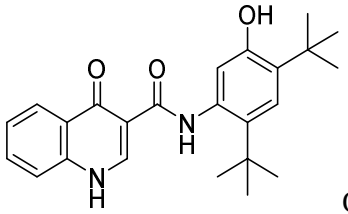
40. The pharmaceutical composition according to any one of claims 36-39, wherein the one or more additional therapeutic agent(s) comprise(s) a CFTR potentiator.

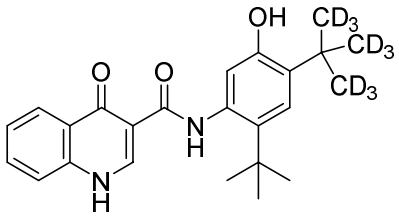
41. The pharmaceutical composition according to any one of claims 36-40, wherein the one or more additional therapeutic agent(s) comprise(s) a compound selected from:

(a) *N*-(5-hydroxy-2,4-di-*tert*-butyl-phenyl)-4-oxo-1*H*-quinoline-3-carboxamide (Compound **III**):



Compound **III**,

(b) *N*-(2-(*tert*-butyl)-5-hydroxy-4-(2-(methyl-d3)propan-2-yl-1,1,1,3,3,3-d6)phenyl)-4-oxo-1,4-dihydroquinoline-3-carboxamide (Compound **III-d**):



Compound **III-d**, and deuterated derivatives and pharmaceutically acceptable salts thereof.

42. Use of Compound **I** or a deuterated derivative or pharmaceutically acceptable salt thereof, or a pharmaceutical composition according to any one of claims 27-41, in the manufacture of a medicament for treating cystic fibrosis, wherein the medicament comprises about 0.1 mg to about 50 mg Compound **I** or an equivalent amount of a deuterated derivative or pharmaceutically acceptable salt thereof.