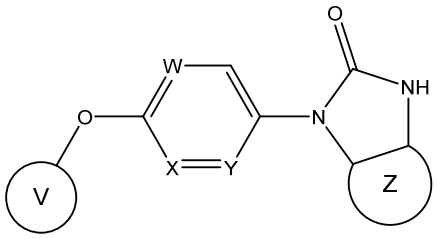
Claims

1. A compound of formula (I):

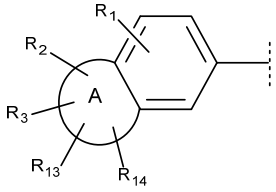


,

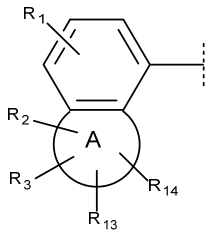
wherein:

V is group (Va), group (Vb) or group (Vc);

wherein group (Va) and group (Vb) are:



(Va);



(Vb);

wherein:

R_1 is H, $C_{1\text{-}4}$alkyl, halo, halo$C_{1\text{-}4}$alkyl, CN, $C_{1\text{-}4}$alkoxy, or halo$C_{1\text{-}4}$alkoxy;

R_2 is H, $C_{1\text{-}4}$alkyl, $C_{3\text{-}5}$ spiro carbocyclyl, halo$C_{1\text{-}4}$alkyl or halo;

R_3 is H, $C_{1\text{-}4}$alkyl, halo$C_{1\text{-}4}$alkyl, halo; or R_3 is absent;

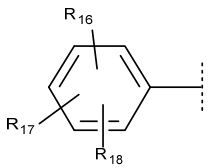
R_{13} is H, $C_{1\text{-}4}$alkyl, halo$C_{1\text{-}4}$alkyl, halo; or R_{13} is absent;

R_{14} is H, $C_{1\text{-}4}$alkyl, halo$C_{1\text{-}4}$alkyl, halo; or R_{14} is absent;

A is a 5 or 6 membered saturated or unsaturated heterocycle, with at least one O atom; which heterocycle is optionally fused with a cyclopropyl group, or a cyclobutyl group, or a cyclopentyl group to form a tricycle when considered together with the phenyl;

wherein R_2 and R_3 may be attached to the same or a different ring atom; R_2 may be attached to a fused ring atom; and wherein R_{13} and R_{14} may be attached to the same or a different ring atom;

wherein group (Vc) is:



(Vc);

wherein:

R_{16} is halo, C_{1-4}alkyl, C_{1-4}alkoxy, haloC_{1-4}alkyl, haloC_{1-4}alkoxy or CN;

R_{17} is H, halo, CN, C_{1-4}alkyl, C_{1-4}alkoxy or haloC_{1-4}alkoxy;

R_{18} is H, halo, CN, C_{1-4}alkyl or C_{1-4}alkoxy;

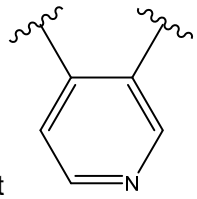
W is N or CH;

X is N or CH;

Y is N or CH;

wherein at least one of W, X and Y is CH, and when one of X and Y is N, the other is CH;

Z is a 5-membered heteroaryl comprising one or two nitrogen atoms, and wherein one of the nitrogen atoms and one of the carbon atoms may be independently optionally substituted by methyl; or Z is a 6-membered heteroaryl comprising one or two nitrogen atoms, wherein one of the carbon atoms may be optionally substituted by methyl; and

provided that Z is not 

;

or a salt and/or solvate and/or derivative thereof.

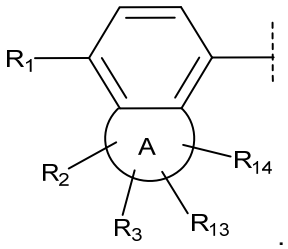
2. The pharmaceutically acceptable salt and/or solvate and/or derivative of the compound of formula (I) according to claim 1.

3. The pharmaceutically acceptable salt of the compound of formula (I) according to claim 1.

4. The pharmaceutically acceptable solvate of the compound of formula (I) according to claim 1.

5. The compound of formula (I) according to claim 1.

6. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to any one of claims 1 to 5, wherein V is group (Vb) and wherein (Vb) is:



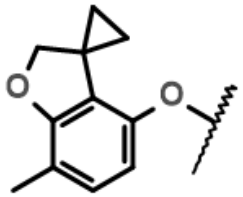
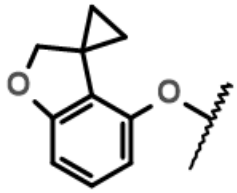
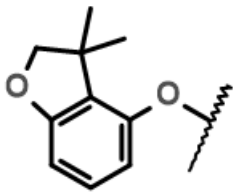
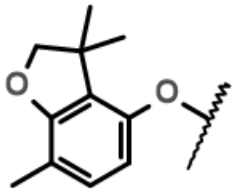
.

7. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to claim 6, wherein R_1 is H or methyl.

8. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to either claim 6 or 7, wherein R_2 is C_3 spiro carbocycle.

9. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to either claim 6 or 7, wherein R_2 is methyl and R_3 is methyl.

10. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to any one of claims 1 to 6, wherein V is selected from the group consisting of:


; 
; 
; and 
.

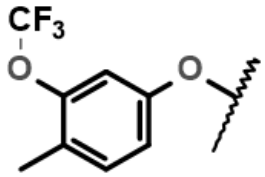
11. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to any one of claims 1 to 5, wherein V is group (Vc).

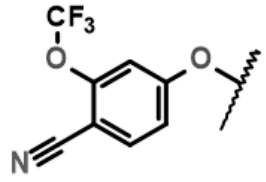
12. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to claim 11, wherein R_{16} is methyl, ethyl, propyl, butyl, cyclopropyl, chloro, fluoro, methoxy, ethoxy, propoxy, trifluoromethyl, trifluoromethoxy or CN, in particular R_{16} is trifluoromethoxy or methoxy, and R_{17} is H, methyl, ethyl, propyl, butyl, cyclopropyl, chloro, fluoro, methoxy, ethoxy, propoxy, trifluoromethoxy or CN, in particular R_{17} is H or methyl or CN.

13. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to claim 11, wherein R_{17} is H and R_{18} is H.

14. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to clause 13, wherein R_{16} is at the meta-position.

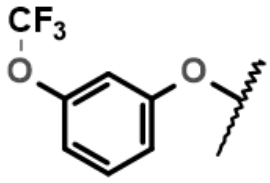
15. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to claim 11, wherein V is selected from the group consisting of:



and


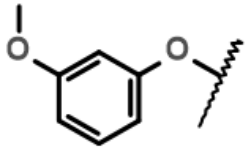
.

16. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to claim 11, wherein V is:



.

17. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to claim 11, wherein V is:

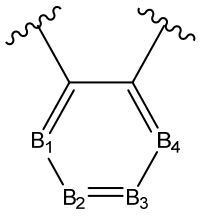


.

18. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to any one of claims 1 to 17, wherein W is N, and X and Y are CH.

19. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to any one of claims 1 to 17, wherein W is N, X is N and Y is CH.

20. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to any one of claims 1 to 19, wherein Z is group (Za):



(Za),

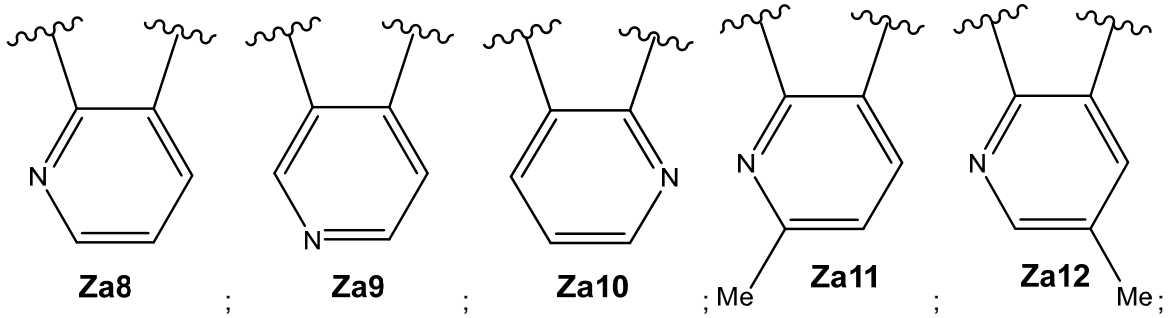
wherein:

B_1, B_2, B_3 and B_4 are each independently selected from N, CH and C(Me);

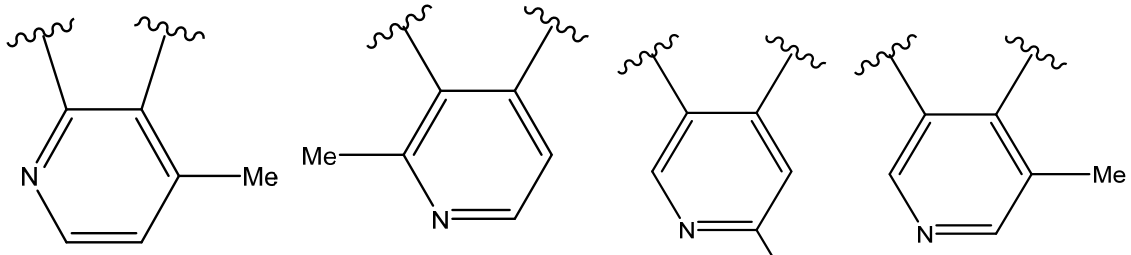
wherein one or two of B_1, B_2, B_3 and B_4 are N, and only one of B_1, B_2, B_3 and B_4 may be C(Me);

and wherein when B_1, B_2 and B_4 are CH or C(Me), B_3 is not N;

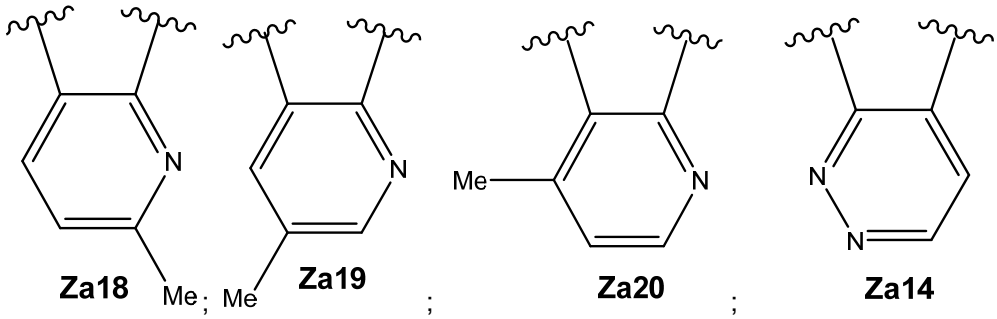
and wherein (Za) is selected from the group consisting of:



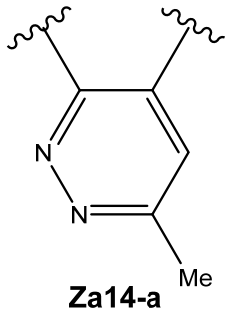
Za8 ; **Za9** ; **Za10** ; **Za11** ; **Za12** ;



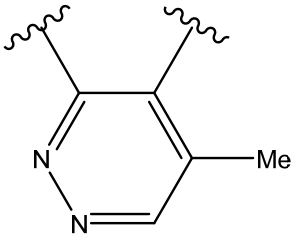
Za13 ; **Za9-a** ; **Za9-b** ; **Za9-c** ;



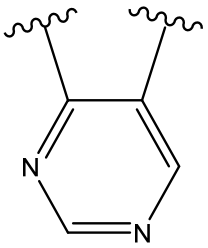
Za18 ; **Za19** ; **Za20** ; **Za14** ;



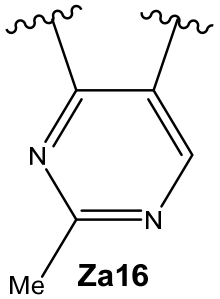
Za14-a ;



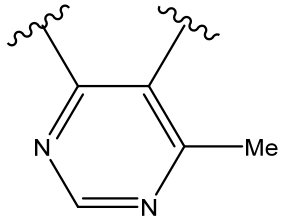
Za14-b ;



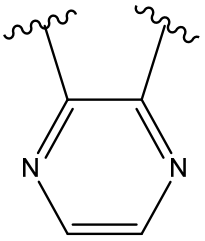
Za15 ;



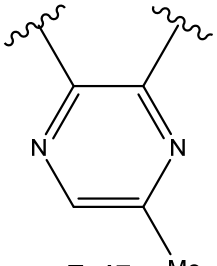
Za16 ;



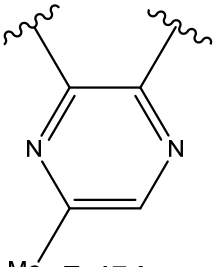
Za16-a ;



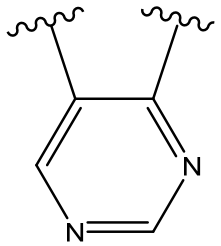
Za17 ;



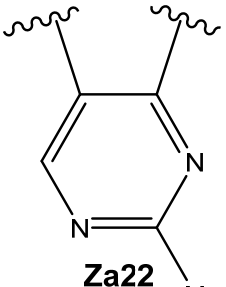
Za17-a ;



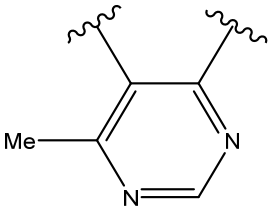
Za17-b



Za21 ;

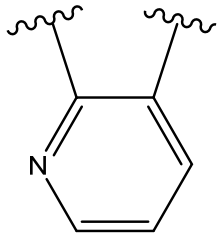


Za22 and



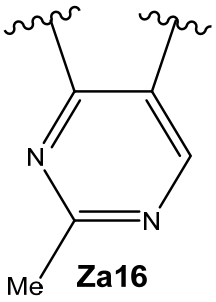
Za22-a .

21. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to claim 20, wherein (Za) is:



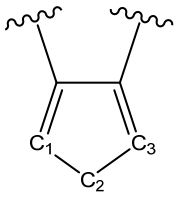
Za8 .

22. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to claim 20, wherein (Za) is:



Za16 .

23. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to any one of claims 1 to 19, wherein Z is group (Zb):

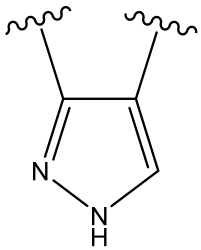


(Zb),

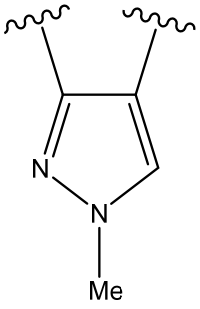
wherein:

C_1 and C_3 are each independently selected from CH, C(Me) and N;

C_2 is NH or N(Me); and wherein when one of C_1 and C_3 is N, the other is CH or C(Me); suitably wherein Zb is selected from the group consisting of:



Zb9 and



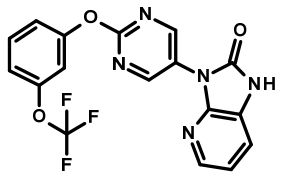
Zb10 .

24. The compound claim 5, which is selected from the group consisting of:
3-[6-(7-methylspiro[2H-benzofuran-3,1'-cyclopropane]-4-yl)oxy-3-pyridyl]-1H-imidazo[4,5-b]pyridin-2-one;
7-methyl-3-[6-(7-methylspiro[2H-benzofuran-3,1'-cyclopropane]-4-yl)oxy-3-pyridyl]-1H-imidazo[4,5-b]pyridin-2-one;
3-(6-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxy-3-pyridyl)-1H-imidazo[4,5-b]pyridin-2-one;
3-[6-[4-methyl-3-(trifluoromethoxy)phenoxy]-3-pyridyl]-1H-imidazo[4,5-b]pyridin-2-one;
7-methyl-3-(6-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxy-3-pyridyl)-1H-imidazo[4,5-b]pyridin-2-one;

3-(5-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxy-2-pyridyl)-1H-imidazo[4,5-b]pyridin-2-one;
3-[6-[3-(trifluoromethoxy)phenoxy]-3-pyridyl]-1H-imidazo[4,5-b]pyridin-2-one;
3-(5-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxypyrazin-2-yl)-1H-imidazo[4,5-b]pyridin-2-one;
3-[5-[(3,3-dimethyl-2H-benzofuran-4-yl)oxy]pyrazin-2-yl]-1H-imidazo[4,5-b]pyridin-2-one;
3-[5-[3-(trifluoromethoxy)phenoxy]pyrazin-2-yl]-1H-imidazo[4,5-b]pyridin-2-one;
3-[5-(7-methylspiro[2H-benzofuran-3,1'-cyclopropane]-4-yl)oxypyrazin-2-yl]-1H-imidazo[4,5-b]pyridin-2-one;
3-[6-[(3,3-dimethyl-2H-benzofuran-4-yl)oxy]-3-pyridyl]-1H-imidazo[4,5-b]pyridin-2-one;
3-(2-{2H-spiro[1-benzofuran-3,1'-cyclopropane]oxy}pyrimidin-5-yl)-1H,2H,3H-imidazo[4,5-b]pyridin-2-one;
4-[[5-(2-oxo-1H-imidazo[4,5-b]pyridin-3-yl)-2-pyridyl]oxy]-2-(trifluoromethoxy)benzonitrile;
7-methyl-3-(2-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxypyrimidin-5-yl)-1H-imidazo[4,5-b]pyridin-2-one;
3-[2-(3-methoxyphenoxy)pyrimidin-5-yl]-1H-imidazo[4,5-b]pyridin-2-one;
3-[2-[3-(trifluoromethoxy)phenoxy]pyrimidin-5-yl]-1H-imidazo[4,5-b]pyridin-2-one;
2-methyl-6-(6-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxy-3-pyridyl)-4H-imidazo[4,5-c]pyrazol-5-one;
2-methyl-6-[6-(7-methylspiro[2Hbenzofuran-3,1'-cyclopropane]-4-yl)oxy-3-pyridyl]-4H-imidazo[4,5-c]pyrazol-5-one;
6-[6-[(3,3-dimethyl-2H-benzofuran-4-yl)oxy]-3-pyridyl]-2-methyl-4H-imidazo[4,5-c]pyrazol-5-one;
2-methyl-6-(5-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxypyrazin-2-yl)-4H-imidazo[4,5-c]pyrazol-5-one;
2-methyl-6-(2-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxypyrimidin-5-yl)-4H-imidazo[4,5-c]pyrazol-5-one;
3-[2-[(3,3-dimethyl-2H-benzofuran-4-yl)oxy]pyrimidin-5-yl]-1H-imidazo[4,5-c]pyridin-2-one;
2-methyl-9-[6-[3-(trifluoromethoxy)phenoxy]-3-pyridyl]-7H-purin-8-one;
2-methyl-9-[6-(7-methylspiro[2H-benzofuran-3,1'-cyclopropane]-4-yl)oxy-3-pyridyl]-7H-purin-8-one;
2-methyl-9-(6-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxy-3-pyridyl)-7H-purin-8-one;
2-methyl-9-[6-[4-methyl-3-(trifluoromethoxy)phenoxy]-3-pyridyl]-7H-purin-8-one;
9-[6-(3-methoxyphenoxy)-3-pyridyl]-2-methyl-7H-purin-8-one;
9-[6-(7-methylspiro[2H-benzofuran-3,1'-cyclopropane]-4-yl)oxy-3-pyridyl]-7H-purin-8-one;
9-[6-[(3,3-dimethyl-2H-benzofuran-4-yl)oxy]-3-pyridyl]-2-methyl-7H-purin-8-one;
3-[2-(7-methylspiro[2H-benzofuran-3,1’-cyclopropane]-4-yl)oxypyrimidin-5-yl]-1H-imidazo[4,5-b]pyridine-2-one;
3-[2-[4-methyl-3-(trifluoromethoxy)phenoxy]pyrimidin-5-yl]-1H-imidazo[4,5-b]pyridin-2-one;

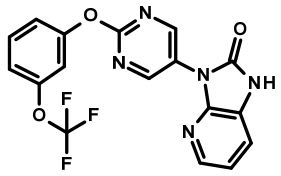
3-[2-[(3,3-dimethyl-2H-benzofuran-4-yl)oxy]pyrimidin-5-yl]-1H-imidazo[4,5-b]pyridin-2-one;
6-(6-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxy-3-pyridyl)-2,4-dihydroimidazo[4,5-c]pyrazol-5-one;
3-[6-(7-methylspiro[2H-benzofuran-3,1'-cyclopropane]-4-yl)oxy-3-pyridyl]-1H-imidazo[4,5-c]pyridin-2-one;
1-(6-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxy-3-pyridyl)-3H-imidazo[4,5-b]pyridin-2-one;
5-methyl-3-(5-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxypyrazin-2-yl)-1H-imidazo[4,5-b]pyridin-2-one;
6-methyl-3-(5-spiro[2H-benzofuran-3,1'-cyclopropane]-4-yloxypyrazin-2-yl)-1H-imidazo[4,5-b]pyridin-2-one; and
3-[2-[(3,3,7-trimethyl-2H-benzofuran-4-yl)oxy]pyrimidin-5-yl]-1H-imidazo[4,5-b]pyridin-2-one.

25. 3-[2-[3-(trifluoromethoxy)phenoxy]pyrimidin-5-yl]-1H-imidazo[4,5-b]pyridin-2-one:



, or a salt and/or solvate thereof.

26. 3-[2-[3-(trifluoromethoxy)phenoxy]pyrimidin-5-yl]-1H-imidazo[4,5-b]pyridin-2-one:



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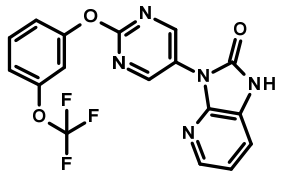
27. A pharmaceutically acceptable salt of 3-[2-[3-(trifluoromethoxy)phenoxy]pyrimidin-5-yl]-1H-imidazo[4,5-b]pyridin-2-one:



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28. The derivative according to any one of claims 1 or 6 to 23, functionalised at the secondary nitrogen of the urea with a group L, wherein L is selected from:
$–PO(OH)O^- \bullet M^+$, wherein M^+ is a pharmaceutically acceptable monovalent counterion,
$–PO(O^-)_2 \bullet 2M^+$,
$–PO(O^-)_2 \bullet D^{2+}$, wherein D^{2+} is a pharmaceutically acceptable divalent counterion,
$–CH(R^X)–PO(OH)O^- \bullet M^+$, wherein R^X is hydrogen or C_{1-3} alkyl,
$–CH(R^X)–PO(O^-)_2 \bullet 2M^+$,
$–CH(R^X)–PO(O^-)_2 \bullet D^{2+}$, and
$–CO–CH_2CH_2–CO_2 \bullet M^+$.

29. The compound, salt thereof and/or solvate thereof, and/or derivative thereof according to any one of claims 1 to 28 for use as a medicament.

30. Use of a compound, salt thereof and/or solvate thereof, and/or derivative thereof according to any one of claims 1 to 28 in the manufacture of a medicament.

31. The compound, salt thereof and/or solvate thereof, and/or derivative thereof or use according to any one of claims 1 to 30 for use in the treatment or prophylaxis of progressive myoclonic epilepsy.

32. The compound, salt thereof and/or solvate thereof, and/or derivative thereof or use according to claim 31 for use in the treatment or prophylaxis of progressive myoclonic epilepsy.

33. The compound, salt thereof and/or solvate thereof, and/or derivative thereof or use according to claim 31 for use in the treatment or prophylaxis of juvenile progressive myoclonic epilepsy.

34. The compound, salt thereof and/or solvate thereof, and/or derivative thereof or use according to any one of claims 1 to 33 for use as a human medicament.

35. The compound, salt thereof and/or solvate thereof, and/or derivative thereof or use according to claim 34, for use in a human of less than 18 years of age.

36. The compound, salt thereof and/or solvate thereof, and/or derivative thereof or use according to claim 35, for use in a human 4 to 17 years old.

37. The compound, salt thereof and/or solvate thereof, and/or derivative thereof or use according to claim 34, for use in a human aged 18 to 65.

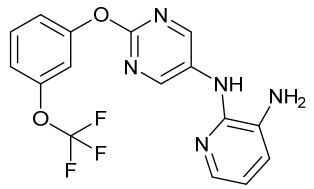
38. The compound, salt thereof and/or solvate thereof, and/or derivative thereof or use according to claim 34, for use in a human aged 66 years old or older.

39. The compound, salt thereof and/or solvate thereof, and/or derivative thereof or use according to any one of claims 29 to 38, for administration at 5 to 250 mg per day.

40. The compound, salt thereof and/or solvate thereof, and/or derivative thereof or use according to any one of claims 29 to 39, for administration for a period of at least three months.

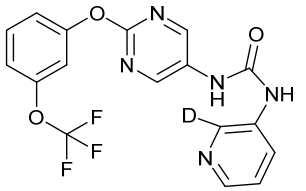
41. A compound or salt thereof selected from the group consisting of:

- a compound of formula (II):



(II) ;

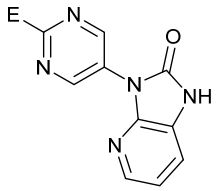
- a compound of formula (III):



(III) ,

wherein D is halo, such as Cl, Br or I; and

- a compound of formula (IV):



(IV) ,

wherein E is halo, such as F or Cl;

or a salt thereof, such as a pharmaceutically acceptable salt thereof.