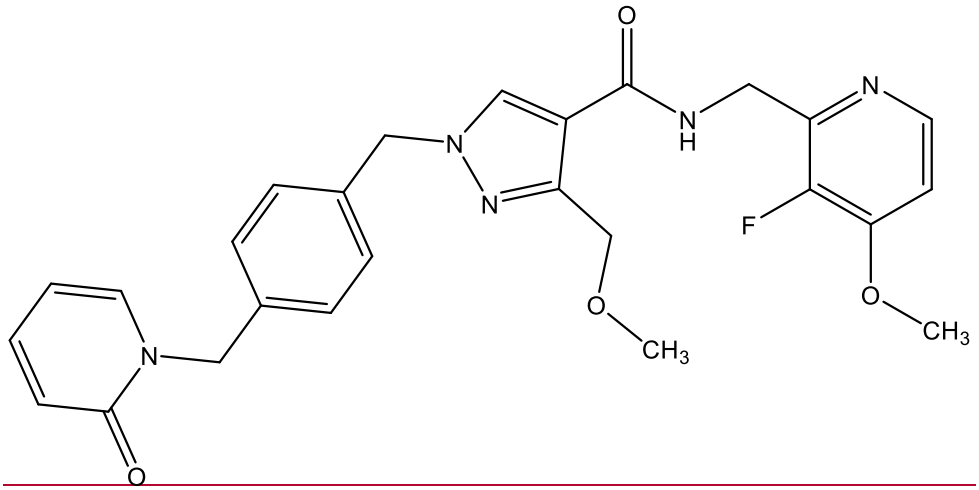
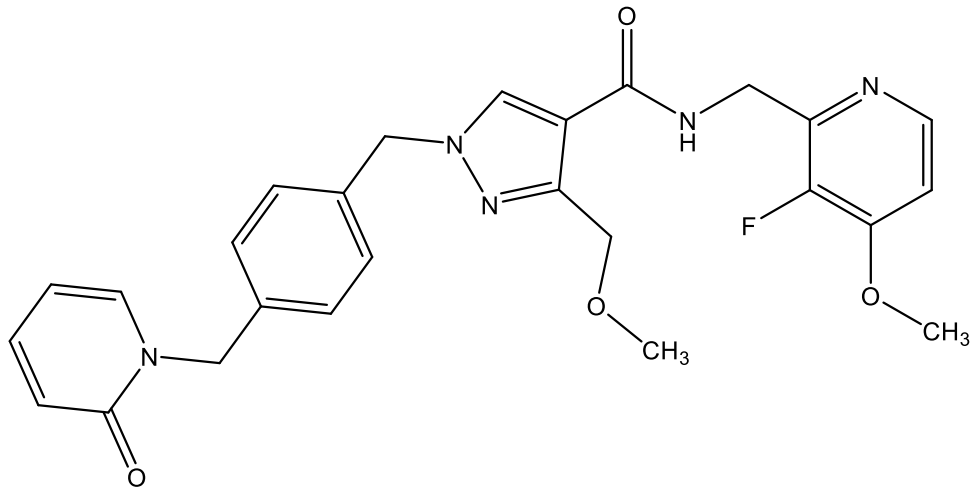
Claims

~~1. A method for treating hereditary angioedema (HAE) on-demand comprising: orally administering the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) to a patient in need thereof on-demand,~~



~~Formula A.~~

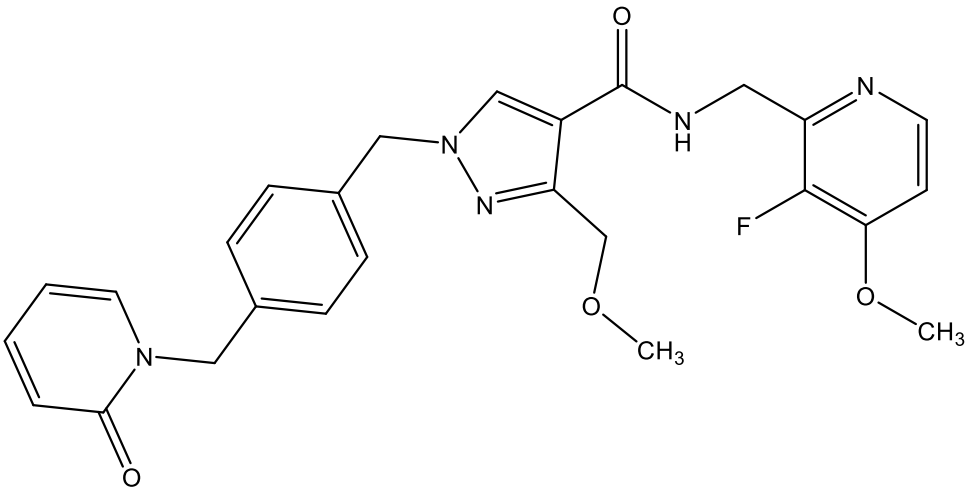
~~1~~2. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use in treating hereditary angioedema (HAE) on-demand comprising: orally administering the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) to a patient in need thereof on-demand,



Formula A.

~~2~~3. The use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) in the manufacture of a medicament for the treatment of hereditary angioedema (HAE) on-demand, ~~comprising:~~ wherein ~~orally administering~~ the compound of Formula A (or a pharmaceutically

acceptable salt and/or solvate thereof) is to be orally administered to a patient in need thereof on-demand,



Formula A.

34. ~~The method according to claim 1, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 12, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 23,

wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is for use in treating an acute attack of hereditary angioedema (HAE) on-demand and is to be orally administered on-demand upon recognition of a symptom of an acute HAE attack.

45. ~~The method according to claim 4, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 34, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 34,

wherein the symptom of an acute HAE attack recognised is at least one of: swelling of tissues; fatigue; headache; muscle aches; skin tingling; abdominal pain; nausea; vomiting; diarrhoea; difficulty swallowing; hoarseness; shortness of breath; and/or mood changes.

56. ~~The method according to any of claims 4 or 5, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 34 or 45, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 34 to 45,

wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is to be orally administered on-demand within 1 hour of the symptom of an acute HAE attack being recognised.

67. ~~The method according to any of claims 4 to 6, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 34 to 56, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 34 to 56,

wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is to be orally administered on-demand within 30 minutes, within 20 minutes, within 10 minutes, or within 5 minutes of the symptom of an acute HAE attack being recognised.

78. ~~The method according to any of claims 4 to 7, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 34 to 67, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 34 to 67,

wherein the compound of Formula A (or a pharmaceutically acceptable salt or solvate thereof) is to be orally administered on-demand in the prodromal phase of an acute HAE attack.

89. ~~The method according to claim 8, t~~The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 78, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 78,

wherein the symptom recognised is at least one of: a slight swelling, abdominal pain or reddening of the skin.

910. ~~The method according to claim 9, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 89, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 89,

wherein the symptom recognised is erythema marginatum.

1011. ~~The method according to any of claims 1 and 4 to 10, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 12 and 34 to 910, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 23 to 910,

wherein the treatment shortens the duration of the acute HAE attack.

1112. ~~The method according to claim 8, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 78, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 78,

wherein the treatment prevents the acute HAE attack from progressing to the swelling stage of an acute HAE attack.

1213. ~~The method according to claim 1, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 12, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 23,

wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is to be orally administered on-demand to prophylactically reduce the likelihood of an acute HAE attack.

1314. ~~The method according to claim 13, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim ~~13~~12, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim ~~13~~12,

Wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is to be orally administered on-demand when it is anticipated that an acute HAE attack will be induced.

1415. ~~The method according to claim 14, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 1314, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 1314,

wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is to be orally administered on-demand when it is anticipated that an acute HAE attack will be induced by physical traumata and/or stress.

1516. ~~The method according to claim 15, t~~The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 1415, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 1415,

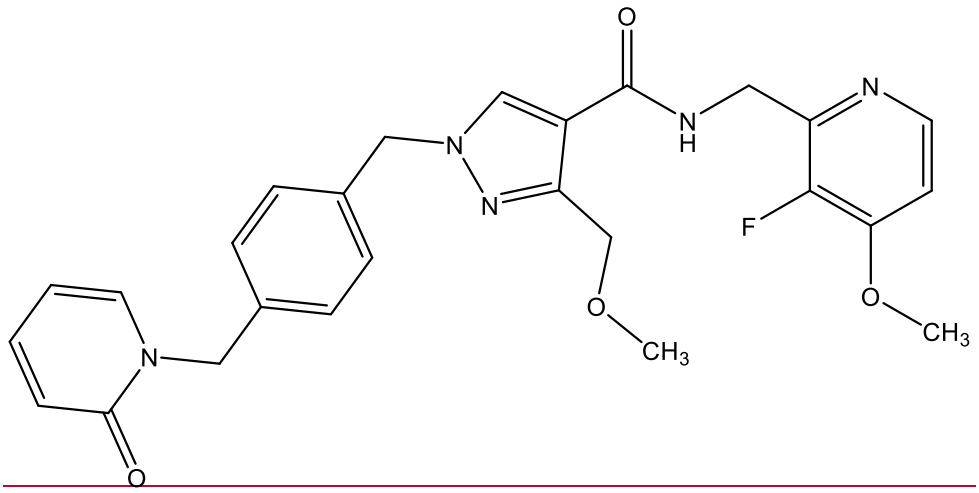
wherein it is anticipated that an acute HAE attack will be induced by the physical traumata of a dental procedure and/or the mental stress associated with a dental procedure.

1617. ~~The method according to any of claims 13 to 16, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 1213 to 1516,

or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 1213 to 1516,

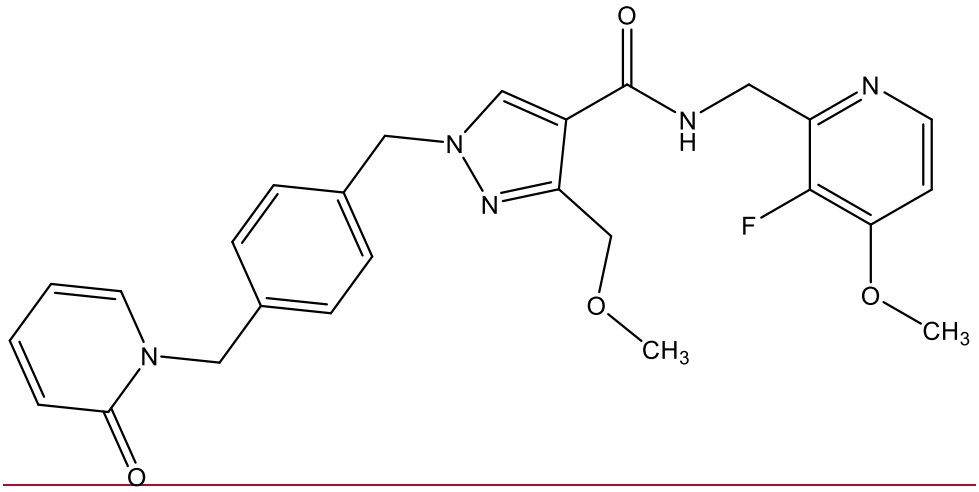
wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is to be orally administered on-demand to prevent an acute HAE attack.

~~18. A method for treating hereditary angioedema (HAE) comprising: orally administering the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) to a patient in need thereof, wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is orally administered prophylactically to reduce the likelihood of an acute HAE attack, wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is administered regularly to the patient,~~



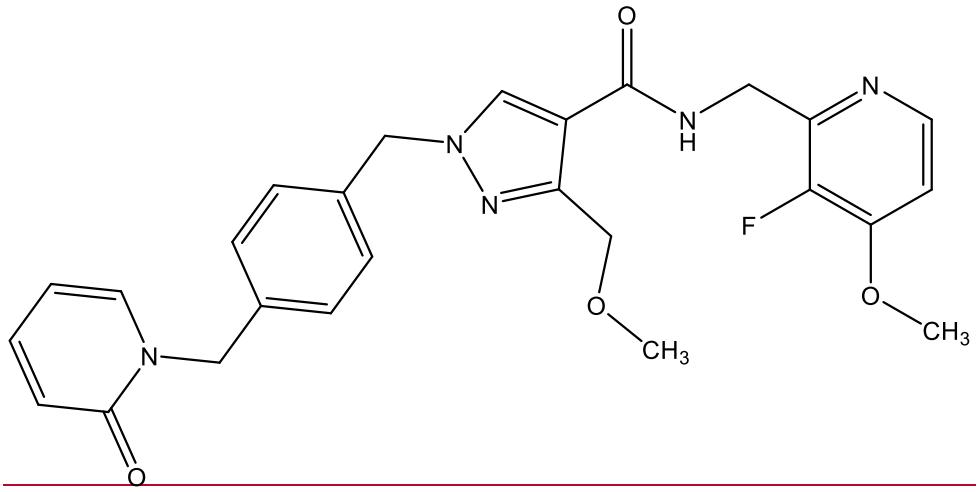
~~Formula A.~~

~~19. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use in treating hereditary angioedema (HAE) comprising: orally administering the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) to a patient in need thereof, wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is orally administered prophylactically to reduce the likelihood of an acute HAE attack, wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is administered regularly to the patient,~~



~~Formula A.~~

~~20. The use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) in the manufacture of a medicament for the treatment of hereditary angioedema (HAE) comprising: orally administering the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) to a patient in need thereof, wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is orally administered prophylactically to reduce the likelihood of an acute HAE attack, wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is administered regularly to the patient,~~



~~Formula A.~~

~~21. The method according to claim 18, the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 19, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 20,~~

~~wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is administered to prevent an acute HAE attack.~~

~~22. The method according to any of claims 18 or 21, the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 19 or 21, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 20 to 21,~~

~~wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is orally administered once daily.~~

~~23. The method according to any of claims 18 or 21, the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 19 or 21, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 20 to 21,~~

~~wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is orally administered twice daily.~~

~~24. The method according to any of claims 18 or 21, the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 19 or 21, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 20 to 21,~~

~~wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is orally administered three times daily.~~

17~~25~~. ~~The method according to any preceding claim, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any preceding claim, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any preceding claim,

wherein the compound (or a pharmaceutically acceptable salt and/or solvate thereof) is to be administered as an oral dosage form comprising: (i) the compound (or a pharmaceutically acceptable salt and/or solvate thereof), and (ii) pharmaceutically acceptable excipients.

18~~26~~. ~~The method according to claim 25, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 17~~25~~, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 17~~25~~,

wherein the oral dosage form is a tablet comprising microcrystalline cellulose as a diluent, croscarmellose sodium as a disintegrant, polyvinyl pyrrolidone as a binder, and optionally magnesium stearate as a lubricant.

1927. ~~The method according to any preceding claim, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any preceding claim, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any preceding claim,

wherein the compound (or a pharmaceutically acceptable salt and/or solvate thereof) (i) inhibits plasma kallikrein, (ii) reduces cleavage of plasma prekallikrein, and/or (iii) reduces the generation of Factor XIIa from Factor XII.

2028. ~~The method according to claim 27, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 1927, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 1927,

wherein the patient is to be administered a dose of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) such that the patient's plasma has a concentration of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) of at least 500 ng/mL.

2129. ~~The method according to claim 28, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 2028, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 2028,

wherein the patient is to be administered at least 60 mg of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof).

2230. ~~The method according to any preceding claim, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any preceding claim, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any preceding claim,

wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) blocks contact system activation for up to six hours.

2331. ~~The method according to any preceding claim, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any preceding claim, or the

use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any preceding claim,

wherein the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) is to be administered at a daily dosage amount of between 5 mg and 2000 mg.

2432. ~~The method according to any preceding claim, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any preceding claim, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any preceding claim,

Wherein the compound of Formula A is to be administered at a daily dosage amount of between 100 mg and 1500 mg, between 300 mg to 1800 mg, between 100 mg and 1400 mg per day, between 200 mg and 1200 mg, between 300 mg and 1200 mg, between 600 mg and 1200 mg, between 450 mg and 900 mg, between 500 mg and 1000 mg, between 450 mg and 600 mg, between 500 mg and 700 mg, between 800 mg and 1000 mg per day, between 900 mg and 1400 mg, or between 900 mg and 1200 mg.

2533. ~~The method according to any preceding claim, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any preceding claim, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any preceding claim,

wherein the patient is to be administered the daily dosage amount in two dosage amounts within a 24 hour period starting from the time of taking the first dosage amount.

2634. ~~The method according to claim 33, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 2533, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 2533,

wherein the two dosage amounts are to be administered simultaneously, separately or sequentially.

2735. ~~The method according to any of claims 33 or 34, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 2533 or 2634, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 2533 or 2634,

wherein the second dosage amount is to be administered between 2 and 6 hours of the first, preferably between about 3 and 6 hours of the first dosage amount.

2836. ~~The method according to any of claims 33 or 34, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 2533 or 2634, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 2533 or 2634,

wherein the second dosage amount ~~can~~ is to be administered at least about 6 hours after the first dosage amount.

2937. ~~The method according to any of claims 1, 4 to 18, or 21 to 32, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 12, 34 to 2417, ~~19, or 21 to 32,~~ or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 23 to 2417, ~~or 20 to 32~~,

wherein the patient is to be administered the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) as three dosage amounts per day.

3038. ~~The method according to claim 37, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 2937, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 2936,

wherein the three dosage amounts are to be administered simultaneously, separately or sequentially.

3139. ~~The method according to any of claims 37 or 38, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 2937 or 3038, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 2937 or 3038,

wherein the second and third dosage amounts ~~can~~ are to be administered at least about 6 hours after the preceding dosage amount.

3240. ~~The method according to any of claims 30 to 39, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 2230 to 3139, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 2230 to 3139,

wherein each dosage amount comprises about 600 mg of the compound of formula A.

3341. ~~The method according to claim 40, T~~the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 3240, or the use according to any claim 3240,

wherein each dosage amount is to be administered as two tablets each comprising about 300 mg of the compound of formula A.