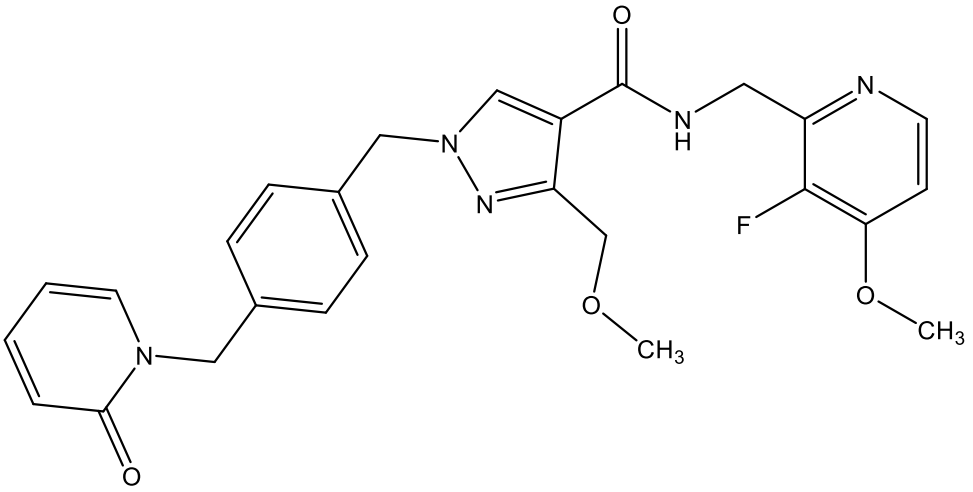
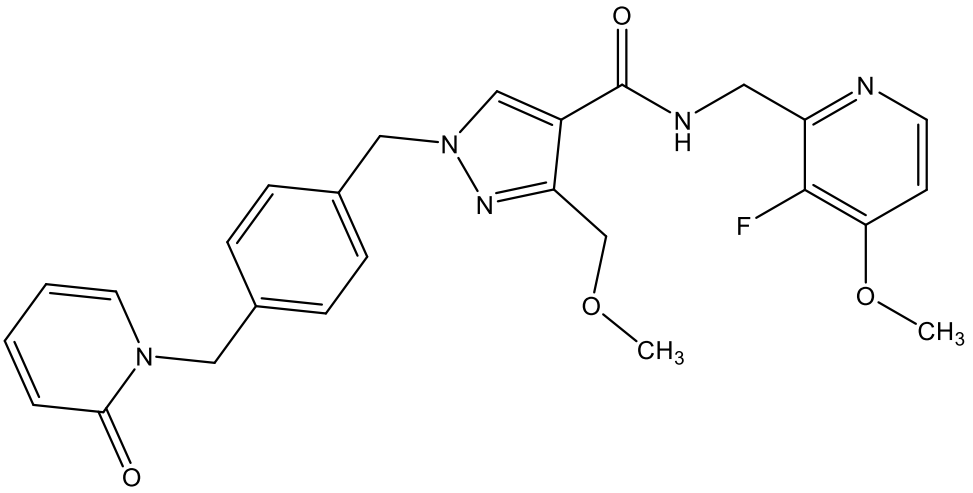
Claims

1. 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. 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, wherein 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.

3. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 1, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 2,

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.

4. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 3, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 3,

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.

5. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 3 or 4, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 3 to 4,

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.

6. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 3 to 5, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 3 to 5,

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.

7. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 3 to 6, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 3 to 6,

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.

8. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 7, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 7,

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

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

wherein the symptom recognised is erythema marginatum.

10. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 1 and 3 to 9, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 2 to 9,

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

11. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 7, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 7,

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

12. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 1, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 2,

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.

13. 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 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.

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

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.

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

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.

16. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 12 to 15, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 12 to 15,

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.

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

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.

19. 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.

20. 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 19,

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.

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

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).

22. 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.

23. 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.

24. 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.

25. 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.

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

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

27. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 25 or 26, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 25 or 26,

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.

28. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 25 or 26, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 25 or 26,

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

29. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 1, 3 to 24, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 2 to 24,

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.

30. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 29, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to claim 29,

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

31. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 29 or 30, or the use of the compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 29 or 30,

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

32. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to any of claims 22 to 31, or the use of a compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) according to any of claims 22 to 31,

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

33. The compound of Formula A (or a pharmaceutically acceptable salt and/or solvate thereof) for use according to claim 32, or the use according to any claim 32,

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