Claims

1) Macitentan for use in the treatment and/or prevention of chronic thromboembolic pulmonary hypertension (CTEPH) in human, wherein the dosage of macitentan is from 60 to 90 mg per day.

2) Macitentan for use in the treatment and/or prevention of chronic thromboembolic pulmonary hypertension (CTEPH) according to claim 1, wherein the dosage of macitentan is from 70 to 80 mg per day.

3) Macitentan for use in the treatment and/or prevention of CTEPH according to claim 1 or 2, wherein the dosage of macitentan is 75 mg per day.

4) Macitentan for use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 3, wherein the dosages are given once per day.

5) Macitentan for use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 3, wherein the dosage of macitentan is 35 to 40 mg twice per day.

6) Macitentan for use in the treatment and/or prevention of CTEPH according to claim 5, wherein the dosage of macitentan is 37.5 mg twice per day.

7) Macitentan for use in the treatment and/or prevention of CTEPH according to claim 1, wherein the dosage of macitentan is escalated from 10 mg per day, followed by 25 to 50 mg per day, and followed by 60 to 90 mg per day.

8) Macitentan for use in the treatment and/or prevention of CTEPH according to claim 7, wherein the dosage of macitentan is escalated from 10 mg per day, followed by 37.5 mg per day, and followed by 75 mg per day.

9) Macitentan for use in the treatment and/or prevention of CTEPH according to claim 1 or 2, wherein the dosage of macitentan is escalated from 10 mg per day, followed by 37.5 mg per day, followed by 75 mg per day.

10) Macitentan for use in the treatment and/or prevention of CTEPH according to claim 9, wherein the dosage of macitentan is escalated from 10 mg per day, followed by 75 mg once per day or 37.5 mg twice a day.

11) Macitentan for use in the treatment and/or prevention of CTEPH according to any one of claims 7 to 10, wherein the lower dosage of macitentan is applied for 15 to 45 days in case it is followed by a higher dose.

12) Macitentan for use in the treatment and/or prevention of CTEPH according to claim 7, wherein the dosage of macitentan is escalated from 10 mg once per day, for 15 to 45 days; followed by 25 to 50 mg per day, for 15 to 45 days; and followed by 60 to 90 mg per day.

13) Macitentan for use in the treatment and/or prevention of CTEPH according to claim 12, wherein the dosage of macitentan is escalated from 10 mg once per day for 15 to 45 days; followed by 37.5 mg once per day for 15 to 45 days; and followed by 75 mg once per day or 37.5 mg twice per day.

14) Macitentan for use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 3, provided that the patient is already treated with an endothelin receptor antagonist preferably selected from bosentan and ambrisentan.

15) Macitentan for use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 14, wherein macitentan is combined with a PDE5 inhibitor and/or a prostacyclin analogue, and/or a prostacyclin receptor agonist and/or a soluble guanylate cyclase stimulator.

16) Macitentan for the use in the treatment and/or prevention of CTEPH according to claim 15, wherein the PDE5 inhibitor is selected from sildenafil, tadalafil, vardenafil, and udenafil; the prostacyclin analogue is selected from epoprostenol, treprostinil, iloprost, and beraprost; the prostacyclin receptor agonist is selected from selexipag and ralinepag; and the soluble guanylate cyclase stimulator is selected from riociguat, olinciguat, praliciguat and vericiguat.

17) Macitentan for the use in the treatment and/or prevention of CTEPH according to claim 15 or 16, wherein macitentan is combined with tadalafil and/or selexipag and/or riociguat, preferably with riociguat.

18) Macitentan for the use in the treatment and/or prevention of CTEPH according to claim 17, wherein macitentan is combined with riociguat and riociguat has a dosage of 0.5 mg three times per day to 2.5 mg three times per day, preferably 2.5 mg three times per day.

19) Macitentan for the use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 18, wherein macitentan is for patients before PEA surgery.

20) Macitentan for the use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 18, wherein the patient to be treated is inoperable with PEA surgery.

21) Macitentan for the use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 18, wherein the patient to be treated suffers from persistent or recurrent PH after PEA surgery.

22) Macitentan for the use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 18, wherein the patient to be treated has been treated with BPA and PEA surgery.

23) Macitentan for the use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 18, wherein the patient to be treated is excluded from BPA.

24) Macitentan for the use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 18, wherein the patient to be treated suffers from persistent or recurrent PH after BPA.

25) Macitentan for use in the treatment and/or prevention of CTEPH according to any one of claims 1 to 24, wherein the treatment and/or prevention means the reduction of morbidity and/or mortality risk of CTEPH.

26) A pharmaceutical composition for use in the treatment of CTEPH comprising macitentan and at least a pharmaceutically acceptable excipient, containing macitentan in an amount of 25 to 50 mg or 60 to 90 mg.

27) A pharmaceutical composition according to claim 26, which contains macitentan in an amount of 37.5 mg or 75 mg.

28) The pharmaceutical composition according to claim 26 or 27, which comprises

(i) macitentan in a total amount of 10 to 50% in weight based on the total weight of the pharmaceutical composition,

(ii) a filler, consisting of lactose monohydrate with microcrystalline cellulose, in a total amount of 10 to 85% in weight based on the total weight of the pharmaceutical composition,

(iii) a disintegrant, consisting of sodium starch glycolate or a combination of sodium starch glycolate and polyvinylpyrrolidone, in a total amount of 1 to 10% in weight based on the total weight of the pharmaceutical composition,

(iv) a surfactant, consisting of a polysorbate, in a total amount of 0.1 to 1% in weight based on the total weight of the pharmaceutical composition, and

(v) a lubricant, consisting of magnesium stearate, in a total amount of 0.05 to 5% in weight based on the total weight of the pharmaceutical composition.

29) The pharmaceutical composition according to claim 26 or 28, which is in the form of a capsule or a tablet.