Claims

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

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

3) Macitentan for use in the treatment and/or prevention of pulmonary arterial hypertension (PAH) 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 pulmonary arterial hypertension (PAH) according to one of claims 1 to 3, wherein the PAH is mild or moderate PAH.

5) Macitentan for use in the treatment and/or prevention of PAH according to any one of claims 1 to 4, wherein the dosage of macitentan is 30 to 45 mg twice per day.

6) Macitentan for use in the treatment and/or prevention of PAH 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 PAH according to claim, 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 PAH 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 PAH according to any one of claims 1 to 4, wherein the dosage of macitentan is escalated from 10 mg per day, followed by 60 to 90 mg per day.

10) Macitentan for use in the treatment and/or prevention of PAH 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 PAH 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.

12) Macitentan for use in the treatment and/or prevention of PAH according to claim 11, 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.

13) Macitentan for use in the treatment and/or prevention of PAH according to any one of claims 1 to 4, provided that the patient is already treated with an endothelin receptor antagonist preferably selected from bosentan and ambrisentan.

14) Macitentan for use in the treatment and/or prevention of PAH according to any one of claims 1 to 13, 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.

15) Macitentan for the use in the treatment and/or prevention of PAH according to claim 14, 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 and vericiguat.

16) Macitentan for the use in the treatment and/or prevention of PAH according to claim 14 or 15, wherein macitentan is combined with tadalafil and/or selexipag or ralinepag, preferably with tadalafil and/or selexipag.

17) Macitentan for the use in the treatment and/or prevention of PAH according to claim 16, wherein tadalafil, if applicable, has a dose of 20 to 40 mg per day, preferably 40 mg per day, selexipag, if applicable, has a dose of 0.2 to 1.6 mg twice per day and ralinepag, if applicable, has a dose of 0.05 to 1.45 mg per day.

18) Macitentan for use in the treatment and/or prevention of PAH according to any one of claims 1 to 17, wherein the treatment and/or prevention means the reduction of morbidity and/or mortality risk of PAH.

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

20) A pharmaceutical composition according to claim 19, which contains macitentan in amount of 37.5 mg or 75 mg.

21) The pharmaceutical composition according to claim 19 or 20, 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.

22) The pharmaceutical composition according to any one of claims 19 to 21, which is in the form of a capsule or a tablet.