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(54) Title: PHARMACEUTICAL COMPOSITION FOR THE TREATMENT OF PULMONARY VASCULAR DISEASE AND/OR CARDIAC DYSFUNCTION IN FONTAN-PALLIATED PATIENTS

(57) Abstract: The present invention relates to high doses of macitentan (INN), i.e. propylsulfamic acid [5-(4-bromo-phenyl)-6-[2-(5-bromo-pyrimidin-2-ylloxy)-ethoxy]-pyrimidin-4-yl]-amide or pharmaceutically acceptable salts, solvates, hydrates or morphological forms thereof for use in the treatment of pulmonary vascular disease and/or cardiac dysfunction in functional single ventricular heart disease patients, especially in Fontan-palliated patients. Moreover, the present invention relates to the use of high doses of macitentan for the manufacture of a medicament as well as to a method for the treatment of said diseases. Further, the present invention relates to a dosage regimen as well as to a combination of macitentan with one or more phosphodiesterase type 5 (PDE5) inhibitors, prostacyclin analogues, prostacyclin receptor agonists or soluble guanylate cyclase stimulators. Besides, the present invention relates to a pharmaceutical composition for the treatment of pulmonary vascular disease and/or cardiac dysfunction in functional single ventricular heart disease patients, especially in Fontan-palliated patients comprising a high dose of macitentan. Moreover, the present invention relates to the use of high doses aprocitentan for the same purpose.



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From the INTERNATIONAL BUREAU

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1. The following indications appeared on record concerning: ☐ the applicant ☒ the inventor ☐ the agent ☐ the common representative			
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