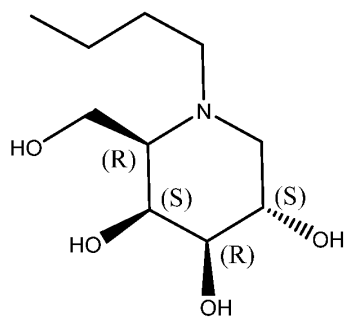


Claims

1. A crystalline form of the compound [(2R,3S,4R,5S)-1-butyl-2-(hydroxymethyl)piperidine-3,4,5-triol:



- 5 characterized by the presence of peaks in the X-ray powder diffraction diagram at the following angles of refraction 2θ : 5.6° , 17.7° , and 21.5° ; wherein said X-ray powder diffraction diagram is obtained by using combined Cu $K\alpha_1$ and $K\alpha_2$ radiation, without $K\alpha_2$ stripping; and the accuracy of the 2θ values is in the range of $2\theta \pm 0.2^\circ$.
2. A crystalline form of the compound according to claim 1, characterized by the presence of
- 10 peaks in the X-ray powder diffraction diagram at the following angles of refraction 2θ : 5.6° , 12.4° , 14.8° , 17.7° , and 21.5° ; wherein said X-ray powder diffraction diagram is obtained by using combined Cu $K\alpha_1$ and $K\alpha_2$ radiation, without $K\alpha_2$ stripping; and the accuracy of the 2θ values is in the range of $2\theta \pm 0.2^\circ$.
3. A crystalline form of the compound according to claim 1, characterized by the presence of
- 15 peaks in the X-ray powder diffraction diagram at the following angles of refraction 2θ : 5.6° , 12.4° , 13.4° , 14.8° , 16.8° , 17.7° , 19.4° , 21.5° , 22.1° , and 24.2° ; wherein said X-ray powder diffraction diagram is obtained by using combined Cu $K\alpha_1$ and $K\alpha_2$ radiation, without $K\alpha_2$ stripping; and the accuracy of the 2θ values is in the range of $2\theta \pm 0.2^\circ$.
4. A crystalline form of the compound according to claim 1, which essentially shows the X-ray
- 20 powder diffraction pattern as depicted in Figure 1.
5. A crystalline form of the compound according to any one of claims 1 to 4, wherein said crystalline form shows endothermal melting with a peak at about 130°C as determined by differential scanning calorimetry.
6. A crystalline form of the compound according to any one of claims 1 to 5, wherein said
- 25 crystalline form consists essentially of particles having a particle size distribution, wherein D_{50} is equal or greater than $50\text{ }\mu\text{m}$.

7. A crystalline form of the compound according to any one of claims 1 to 5, wherein said crystalline form consists essentially of particles having a particle size distribution, wherein
- D50 is from about 50 μm to about 150 μm ; and
 - D90 is from about 100 μm to about 400 μm .
- 5 8. A process for the preparation of the crystalline form of the compound according to any one of claims 1 to 7, wherein said process comprises:
- I) Step 1 comprising:
- i. Preparing a solution of COMPOUND in isopropanol, wherein preferably about 100 ww% to 140 ww% of isopropanol with respect to the weight of COMPOUND is used;
 - 10 ii. Warming up the solution to about 45 °C to 60 °C;
 - xii. Adding anhydrous acetone, wherein about 450 ww% to 550 ww% with respect to the weight of COMPOUND is added, and wherein said acetone is added during about 30 min;
 - iii. Cooling the obtained solution to about – 5 °C to + 5 °C at a constant rate;
 - 15 iv. Agitating at about 0 °C for 1 to 3 hours;
 - v. Isolating the product by solid-liquid separation;
 - vi. Washing the solid with anhydrous acetone, wherein about 250 ww% to 300 ww% of acetone with respect to the weight of COMPOUND is used; and
 - vii. Drying; and
- 20 II) Step 2 comprising:
- i. Dissolving the product of the first crystallization step at about 45 °C in about 200 ww% to 250 ww% methanol with respect to the weight of COMPOUND;
 - ii. Filtering over activated carbon;
 - iii. Washing the activated carbon with about 90 ww% of methanol with respect to the weight of COMPOUND;
 - 25 iv. Concentrating the solution under vacuum;
 - v. Dissolving the distillation residue of the previous step in about 60 ww% to 65 ww% methanol and about 4 ww% to 8 ww% water with respect to the weight of COMPOUND;
 - 30 vi. Warming up the solution to about 55 °C to 58 °C.

- vii. Adding about 200 ww% to 220 ww% ethyl acetate during about 25 min to 45 min while keeping the temperature between about 55 °C to 58 °C;
 - viii. Cooling the resulting suspension down to about -5 to + 5 °C at a constant rate.
 - ix. Agitating at about 0 °C for 1.5 to 3 hours;
 - 5 x. Isolating the product by solid-liquid separation;
 - xi. Washing the solid twice with ethyl acetate; wherein especially about 35 ww% to 55 ww% of ethyl acetate with respect to the weight of COMPOUND is used; and
 - xii. Drying.
9. A pharmaceutical composition comprising:
- 10
 - from about 55 ww% to about 75 ww% of the crystalline form of the compound according to any one of claims 1 to 7; and
 - from about 25 ww% to about 45 ww% of one or more diluents selected from a group consisting of lactose anhydrous, silicified microcrystalline cellulose, microcrystalline cellulose, calcium hydrogen phosphate and isomalt.
 - 15 10. A pharmaceutical composition according to claim 9, comprising:
 - from about 55 ww% to about 75 ww% of the crystalline form of the compound according to any one of claims 1 to 7;
 - from about 24 ww% to about 44 ww% of lactose anhydrous; and
 - 20 • from 0 ww% to about 6 ww% of a component selected from a group consisting of talc, magnesium stearate, sodium steryl fumarate, silicon dioxide, or a combination thereof.
 11. A capsule comprising the pharmaceutical composition according to claim 9 or 10.
 12. A crystalline form of the compound according to any one of claims 1 to 7, or a pharmaceutical composition according to claim 9 or 10, or a capsule according to claim 11,
25 for use as a medicament.
 13. A crystalline form of the compound according to any one of claims 1 to 7, or a pharmaceutical composition according to claim 9 or 10, or a capsule according to claim 11,
for use in the prevention/prophylaxis or treatment of Niemann-Pick type C disease; Gaucher disease types 1, 2 and 3; GM2 gangliosidoses including Tay-Sachs disease, Sandhoff
30 disease, and GM2 gangliosidosis AB variant; GM1 gangliosidosis; Fabry disease; Schindler disease; Smith-Lemly-Opitz syndrome; Tangier disease; mucopolidosis IV; mucopolysaccharidoses; GbA1-synucleopathies and syndromes thereof; Huntington's

- disease; polycystic kidney disease; Darier's disease; Guillain-Barré syndrome; the inflammatory components of human malignancies; infectious diseases including urinary tract infection; neuronal disorders and neurodegenerative processes including Alzheimer's disease, epilepsy, Parkinson's disease and syndromes thereof, stroke, spinal cord injuries,
- 5 motor neuron disease (ALS), multiple sclerosis, and Lewy-Body dementia; inflammatory diseases and other disorders associated with macrophage recruitment and activation including rheumatoid arthritis, Crohn's disease, asthma and sepsis; Krabbe disease; or cystic fibrosis.
14. A crystalline form of the compound according to any one of claims 1 to 7, or a
- 10 pharmaceutical composition according to claim 9 or 10, or a capsule according to claim 11, for use in the prevention/prophylaxis or treatment of Fabry disease.
15. A crystalline form of the compound according to any one of claims 1 to 7, or a pharmaceutical composition according to claim 9 or 10, or a capsule according to claim 11, for use as glycolipid biosynthesis inhibitor in a substrate reduction method for reducing the
- 15 amount of glycolipids in a patient having a glycolipid storage disease or related disorder.
16. Use of a crystalline form of the compound according to any one of claims 1 to 7, or a pharmaceutical composition according to claim 9 or 10, or a capsule according to claim 11, for the preparation of a medicament for the prevention/prophylaxis or treatment of Fabry disease.
- 20 17. A method of treatment or prophylaxis of Fabry disease, comprising administering to a patient an effective amount of a crystalline form of the compound according to any one of claims 1 to 7, or a pharmaceutical composition according to claim 9 or 10, or a capsule according to claim 11.