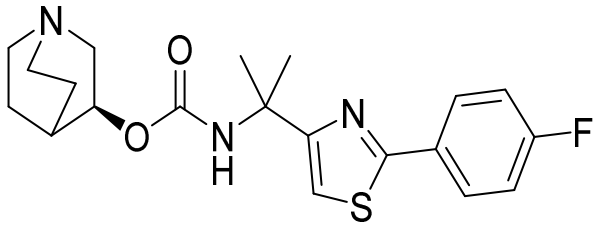
Claims

1. A compound represented by the following structural formula,



or a pharmaceutically acceptable salt thereof.

2. The compound according to claim 1 for use in the treatment of a lysosomal storage disease.

3. The compound for use according to claim 2, wherein the lysosomal storage disease results from a defect in the glycosphingolipid pathway.

4. The compound for use according to claim 3, wherein the lysosomal storage disease is selected from the group consisting of Gaucher, Fabry, G_{M1}-gangliosidosis, G_{M2} Activator deficiency, Tay-Sachs and Sandhoff.

5. The compound for use according to claim 4, wherein the lysosomal storage disease is Fabry.

6. The compound for use according to claim 4, wherein the lysosomal storage disease is Gaucher type 2 or type 3.

7. The compound for use according to any one of claims 2 to 6, wherein said compound is used in combination with a therapeutically effective amount of a lysosomal enzyme.

8. The compound for use according to claim 7, wherein the lysosomal enzyme is selected from the group consisting of glucocerebrosidase, alpha-galactosidase A, Hexosaminidase A, Hexosaminidase B and G_{M1}-ganglioside-β-galactosidase.

9. The compound for use according to claim 8, wherein the lysosomal enzyme is alpha-galactosidase A.

10. The compound for use according to claim 8, wherein the lysosomal enzyme is glucocerebrosidase.

11. The compound for use according to claim 7 in the treatment of a subject diagnosed as having a lysosomal storage disorder, wherein prior to treatment the subject undergoing treatment has elevated levels of a lysosomal substrate.

12. The compound for use according to claim 11, wherein the subject has lower combined amounts of the lysosomal substrate in the urine and plasma than a subject treated with either the lysosomal enzyme or compound alone.

13. The compound for use according to claim 12, wherein the substrate is selected from the group consisting of globotriaosylceramide and lyso-globotriaosylceramide, and combinations thereof.

14. The compound according to claim 1 for use in the treatment of a disease or disorder mediated by glucosylceramide synthase (GCS) or a disease or disorder in which GCS is implicated, wherein said disease or disorder is cancer, a metabolic disorder, a neuropathic disease, or a disease in which excessive glycolipid synthesis occurs selected from atherosclerosis, polycystic kidney disease, and renal hypertrophy.

15. The compound for use according to claim 14, wherein the neuropathic disease is Alzheimers disease or Parkinsons disease.