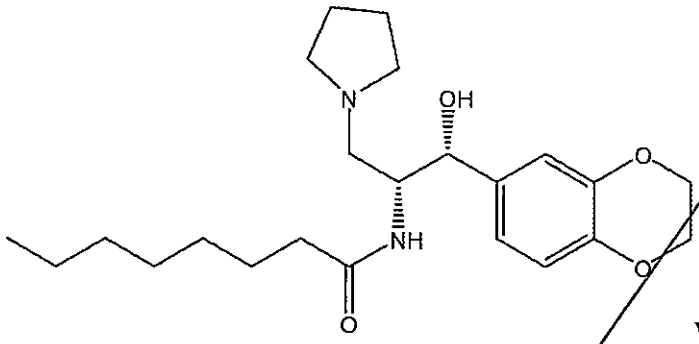
CLAIMS

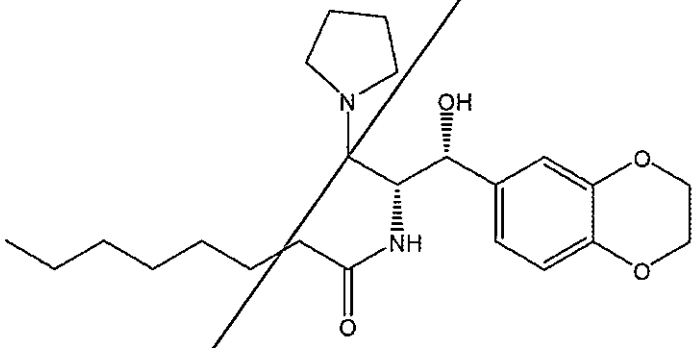
What is claimed is:

1. The hemitartrate salt of a compound represented by the following structural formula:



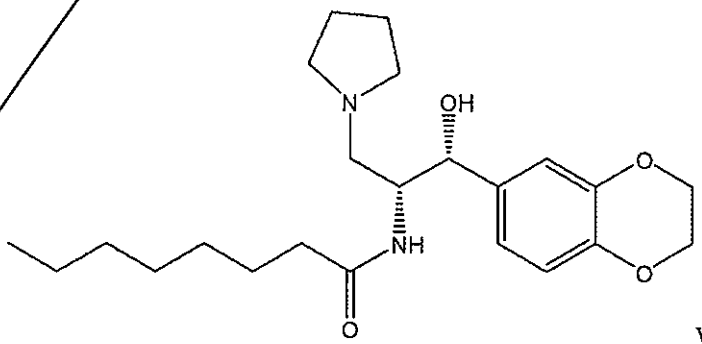
wherein the salt is an amorphous salt.

2. The hemitartrate salt of a compound represented by the following structural formula:



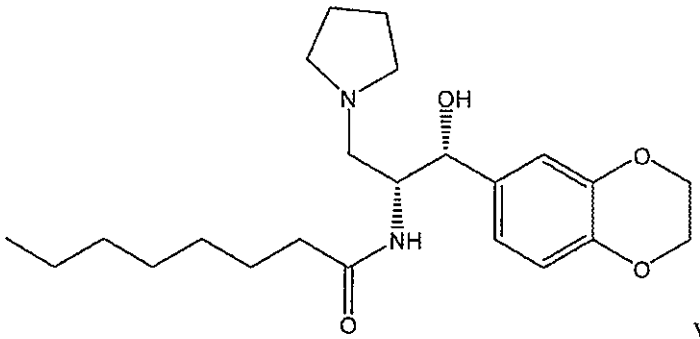
wherein at least 70% by weight of the salt is crystalline.

3. The hemitartrate salt of a compound represented by the following structural formula:



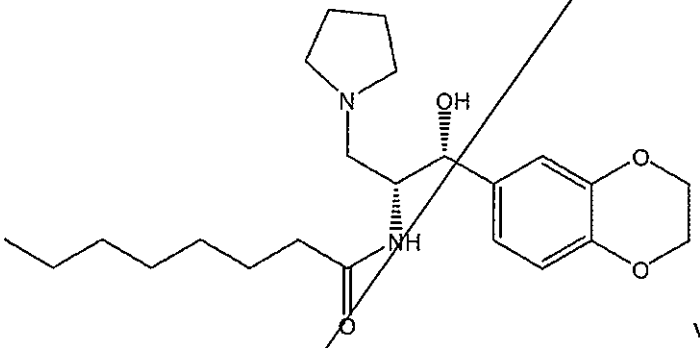
wherein at least 70% by weight of the salt is in a single crystalline form.

4. The hemitartrate salt of a compound represented by the following structural formula:



wherein at least 99% by weight of the salt is crystalline.

5. The hemitartrate salt of a compound represented by the following structural formula:



wherein at least 99% by weight of the salt is in a single crystalline form.

6. The hemitartrate salt of any one of Claims 1-5, wherein the hemitartrate salt is selected from D-hemitartrate, L- hemitartrate, hemimesotartaric acid or racemic D,L-hemitartrate.

7. The hemitartrate salt hemitartrate salt of any one of Claims 1-5, wherein the hemitartrate salt is L-hemitartrate.

8. The salt of Claims 3 or 5, wherein at least 70 % by weight of the salt is the single crystalline form Form A.

9. The salt of Claims 3 or 5, wherein the single crystalline form is characterized by at least one major x-ray powder diffraction peak at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 15.9°, and 21.7°.

10. The salt of Claims 3 or 5, wherein the single crystalline form is characterized by at least two major x-ray powder diffraction peaks at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 15.9°, and 21.7°.

11. The salt of Claims 3 or 5, wherein the single crystalline form is characterized by at least three major x-ray powder diffraction peaks at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 15.9°, and 21.7°.

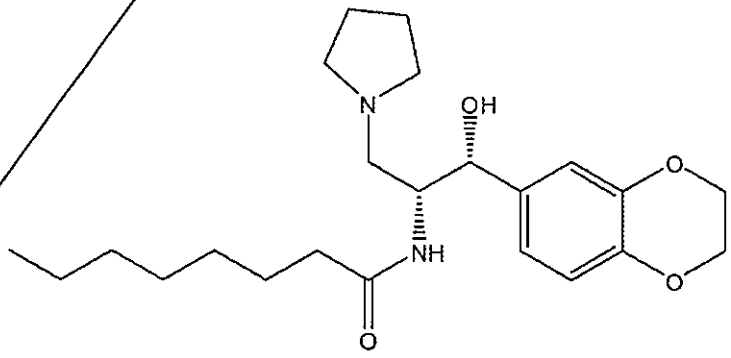
12. The salt of Claims 3 or 5, wherein the single crystalline form is characterized by at least four major x-ray powder diffraction peaks at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 15.9°, and 21.7°.

13. The salt of Claims 3 or 5, wherein the single crystalline form is characterized by major x-ray powder diffraction peaks at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 15.9°, and 21.7°.

14. The salt of Claims 3 or 5, wherein the single crystalline form is characterized by x-ray powder diffraction peaks at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 13.3°, 15.1°, 15.9°, 16.5°, 17.6°, 18.6°, 18.7°, 19.0°, 20.2°, 21.7° and 23.5°.

15. The salt of Claims 3 or 5, wherein the single crystalline form is characterized by x-ray powder diffraction pattern of FIG. 1.

16. A pharmaceutical composition comprising the hemitartrate salt of a compound represented by the following structural formula:



and a pharmaceutically acceptable carrier or diluent.

17. The pharmaceutical composition of Claim 16, wherein the salt is an amorphous salt.

18. The pharmaceutical composition of Claim 16, wherein at least 70% by weight of the salt is crystalline.

19. The pharmaceutical composition of Claim 16, wherein at least 70% by weight of the salt is in a single crystalline form.

20. The pharmaceutical composition of Claim 16, wherein at least 99% by weight of the salt is crystalline.

21. The pharmaceutical composition of Claim 16, wherein at least 99% by weight of the salt is in a single crystalline form.

22. The pharmaceutical composition of Claim 16, wherein the hemitartrate salt is selected from D-hemitartrate, L- hemitartrate, hemimesotartaric acid or racemic D,L-hemitartrate.

23. The pharmaceutical composition of Claim 16, wherein the hemitartrate salt is L-hemitartrate.

24. The pharmaceutical composition of Claim 19 or 21, wherein at least 70 % by weight of the salt is the single crystalline form Form A.

25. The pharmaceutical composition of Claim 19 or 21, wherein the single crystalline form is characterized by at least one major x-ray powder diffraction peak at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 15.9°, and 21.7°.

26. The pharmaceutical composition of Claim 19 or 21, wherein the single crystalline form is characterized by at least two major x-ray powder diffraction peaks at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 15.9°, and 21.7°.

27. The pharmaceutical composition of Claim 19 or 21, wherein the single crystalline form is characterized by at least three major x-ray powder diffraction peaks at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 15.9°, and 21.7°.

28. The pharmaceutical composition of Claim 19 or 21, wherein the single crystalline form is characterized by at least four major x-ray powder diffraction peaks at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 15.9°, and 21.7°.

29. The pharmaceutical composition of Claim 19 or 21, wherein the single crystalline form is characterized by major x-ray powder diffraction peaks at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 15.9°, and 21.7°.

30. The pharmaceutical composition of Claim 19 or 21, wherein the single crystalline form is characterized by x-ray powder diffraction peaks at 2θ angles of 5.1°, 6.6°, 10.7°, 11.0°, 13.3°, 15.1°, 15.9°, 16.5°, 17.6°, 18.6°, 18.7°, 19.0°, 20.2°, 21.7° and 23.5°.

31. The pharmaceutical composition of Claim 19 or 21, wherein the single crystalline form is characterized by x-ray powder diffraction pattern of FIG. 1.

32. A method of treating a subject with Gaucher disease, comprising administering to the subject an effective amount of the hemitartrate salt of any one of Claims 1-15.

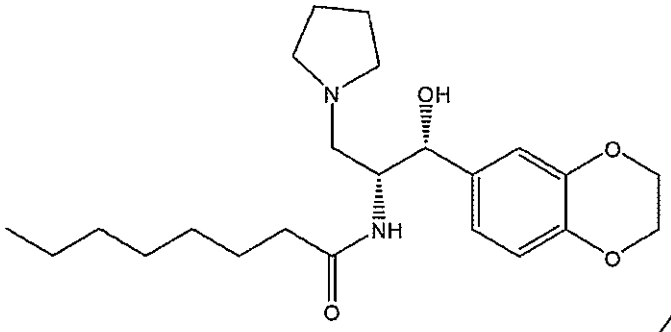
33. A method of treating a subject with Fabry disease, comprising administering to the subject an effective amount of the hemitartrate salt of any one of Claims 1-15.

34. A method of inhibiting glucosylceramide synthase or lowering glycosphingolipid concentrations in a subject in need thereof by administering to the subject an effective amount of the hemitartrate salt of any one of Claims 1-15.

35. The method of Claim 32, 33 or 34, comprising administering the hemitartrate salt at a twice daily dose of from 25 milligrams to 200 milligrams.

36. The method of Claim 32, 33 or 34, comprising administering the hemitartrate salt at a twice daily dose of 50 milligrams.

37. A method of treating a subject with Gaucher disease, comprising administering to the subject an effective amount of a first compound represented by the following structural formula:



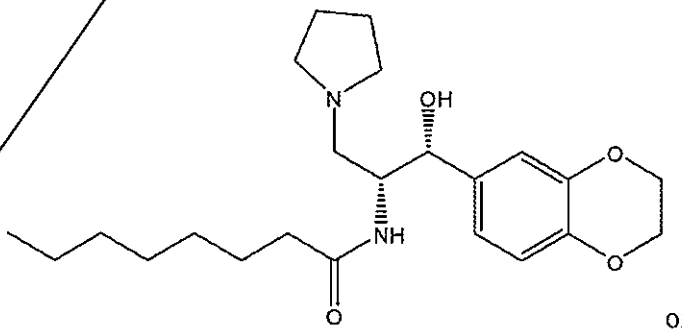
, or a pharmaceutically acceptable salt thereof in combination with an effective amount of a second therapeutic agent that is effective for treating Gaucher disease.

38. The method of Claim 37 wherein the second therapeutic agent is imiglucerase.

39. The method of Claim 37 wherein the second therapeutic agent is isofagomine.

40. The method of Claim 37 wherein the second therapeutic agent is miglustat.

41. A method of treating a subject with Fabry disease, the method comprising administering to the subject an effective amount of a first therapeutic agent represented by the following structural formula:



or a pharmaceutically acceptable salt thereof in combination with an effective amount of a second therapeutic agent that is effective for treating Fabry disease.

42. The method of Claim 41 where the second therapeutic agent is migalastat.

43. The method of Claim 41 where the second therapeutic agent is agalsidase β.

44. The method of any one of Claims 37-43 wherein the first therapeutic agent is administered as the hemitartrate of any one of Claims 1-15.

45. The method of any of Claims 37-43, wherein treatment with the first therapeutic agent is begun after treatment over a period of at least ten weeks with the second therapeutic agent.

46. The method of any one of Claims 37-43, wherein treatment with the first therapeutic agent is begun after treatment with the second therapeutic agent and wherein treatment with the first therapeutic agent is initiated after the subject's platelet count is equal to or greater than 100,000 mm^3 ; hemoglobin concentration is equal to or greater than 11 g/dl (female) or 12 g/dl (male); and/or the subject's spleen volume is less than or equal to 10 multiples of normal and liver volumes are less than or equal to 1.5 multiples of normal.

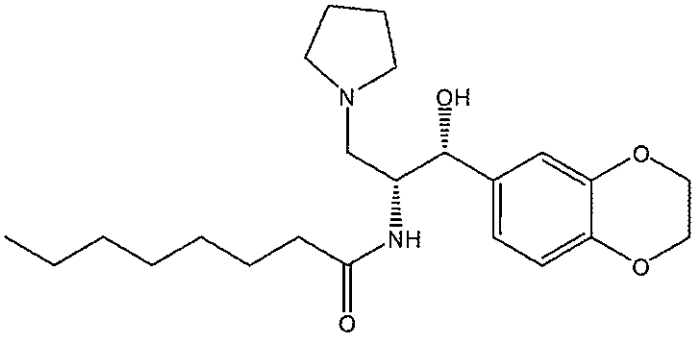
47. The method of Claim 45 or 46 wherein treatment with the second therapeutic agent is terminated after initiation of treatment with the first therapeutic agent.

48. The method of any one of Claims 37-47, comprising administering the first therapeutic agent at a twice daily dose of from 25 milligrams to 200 milligrams.

49. The method of any one of Claims 37-47, comprising administering the first therapeutic agent at a twice daily dose of 50 milligrams.

50. A pharmaceutical composition comprising:

the hemitartrate salt of a compound represented by the following structural formula:



at least one water-soluble filler;

at least one water-insoluble filler;

at least one binder; and

at least one lubricant.

51. The pharmaceutical composition of claim 50, wherein the hemitartrate salt is the hemitartrate salt of any one of Claims 1-15.

52. The pharmaceutical composition of claim 50, wherein the water-soluble filler is selected from the group consisting of anhydrous lactose, lactose monohydrate, mannitol, sodium chloride, powdered sugar, sorbitol, sucrose, inositol and pregelatinized starch.

53. The pharmaceutical composition of claim 50, wherein the water-insoluble filler is selected from the group consisting of microcrystalline cellulose, calcium phosphate and starch.

54. The pharmaceutical composition of claim 50, wherein the binder is selected from the group consisting of pre-gelatinized starch, sodium carboxymethyl cellulose, hydroxypropyl cellulose, hydroxypropyl methyl cellulose, polyvinyl pyrrolidone, copolyvidone, gelatin, natural gums, starch paste, sucrose, corn syrup, polyethylene glycols and sodium alginate.

55. The pharmaceutical composition of claim 50, wherein the lubricant is selected from the group consisting of hydrogenated vegetable oil, calcium stearate, and glyceryl behenate.

56. The pharmaceutical composition of claim 50, wherein the water-soluble filler is selected from the group consisting of anhydrous lactose, lactose monohydrate, mannitol, sodium chloride, powdered sugar, sorbitol, sucrose, inositol and pregelatinized starch; the water-insoluble filler is selected from the group consisting of microcrystalline cellulose, calcium phosphate and starch; the binder is selected from the group consisting of pre-gelatinized starch, sodium carboxymethyl cellulose, hydroxypropyl cellulose, hydroxypropyl methyl cellulose, polyvinyl pyrrolidone, copolyvidone, gelatin, natural gums, starch paste, sucrose, corn syrup, polyethylene glycols and sodium alginate; and the lubricant is selected from the group consisting of hydrogenated vegetable oil, calcium stearate, and glyceryl behenate.

57. The pharmaceutical composition of claim 56, wherein the composition comprises 26 wt% to 50 wt% of the water-soluble filler on a dry solids basis.

58. The pharmaceutical composition of claim 56, wherein the composition comprises 8 wt% to 32 wt% of the water-insoluble filler on a dry solids basis.

59. The pharmaceutical composition of claim 56, wherein the composition comprises 8 wt% to 24 wt% of the water-insoluble filler on a dry solids basis.

60. The pharmaceutical composition of claim 56, wherein the composition comprises 12 wt% to 20 wt% of the water-insoluble filler on a dry solids basis.

61. The pharmaceutical composition of claim 56, wherein the composition comprises 14 wt% to 18 wt% of the water-insoluble filler on a dry solids basis.

62. The pharmaceutical composition of claim 56, wherein the composition comprises 2 wt% to 6 wt% of the binder on a dry solids basis.

63. The pharmaceutical composition of claim 56, wherein the composition comprises 0.1 wt% to 2 wt% of a lubricant on a dry solids basis.

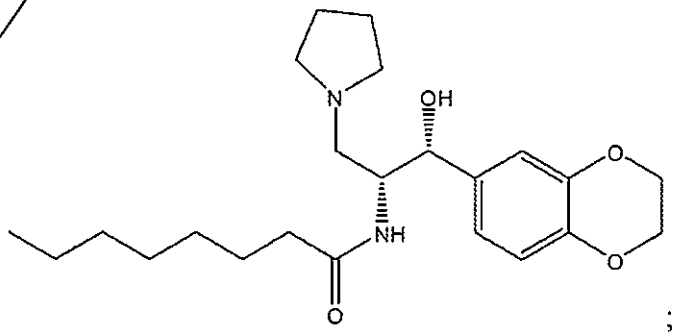
64. The pharmaceutical composition of claim 56, wherein the composition comprises 35 wt% to 40 wt% of the hemitartrate salt, 26 wt% to 50 wt% of the water-soluble filler; 8 wt% to 32 wt% of the water-insoluble filler; 2 wt% to 6 wt% of the binder; and 0.1 wt% to 2 wt% of the lubricant, all on a dry solids basis.

65. The pharmaceutical composition of claim 56, wherein the water-soluble filler is lactose monohydrate; the water-insoluble filler is microcrystalline cellulose; the binder is hydroxypropyl methylcellulose; and the lubricant is glyceryl behenate.

66. The pharmaceutical composition of claim 65, wherein the composition comprises 35 wt% to 40 wt% of the hemitartrate salt, 26 wt% to 50 wt% of the lactose monohydrate; 8 wt% to 32 wt% of the microcrystalline cellulose; 2 wt% to 6 wt% of the hydroxypropyl methylcellulose; and 0.1 wt% to 2 wt% of the glyceryl behenate, all on a dry solids basis..

67. A method of treating a subject with Fabry disease, the method comprising:

a) administering to the subject an effective amount of a compound of Formula (I):



; or a pharmaceutically acceptable salt thereof;

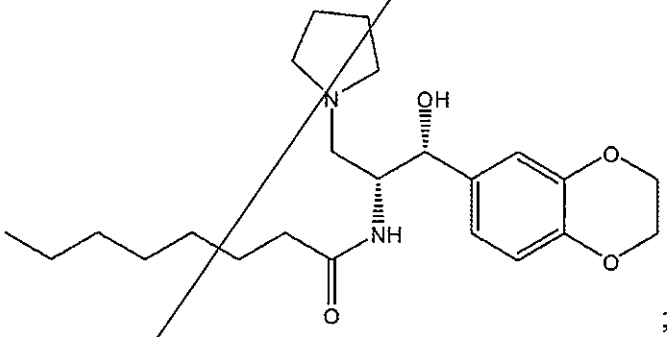
b) testing the subject to determine whether the subject is a poor, intermediate or extensive/ultra rapid P450 metabolizer;

c) if the subject is an intermediate or extensive/ultra rapid P450 metabolizer,
determining an adjusted effective amount of the compound; and

d) administering to the subject an adjusted effective amount of the compound of Formula (I) if the subject is an intermediate or extensive/ultra rapid P450 metabolizer and administering to the subject an effective amount of the compound of Formula (I) if the subject is a poor P450 metabolizer.

68. A method of treating a subject with Gaucher disease, the method comprising:

a) administering to the subject an effective amount of a compound of Formula (I):



; or a pharmaceutically acceptable salt thereof;

b) testing the subject to determine whether the subject is a poor, intermediate or extensive/ultra rapid P450 metabolizer;

c) if the subject is an intermediate or extensive/ultra rapid P450 metabolizer, determining an adjusted effective amount of the compound; and

d) administering to the subject an adjusted effective amount of the compound of Formula (I) if the subject is an intermediate or extensive/ultra rapid P450 metabolizer and administering to the subject an effective amount of the compound of Formula (I) if the subject is a poor P450 metabolizer.

69. The method of Claim 67 or 68, wherein the compound is the hemitartrate salt of any one of Claims 1-15.

70. The method of any one of Claims 67-69, wherein testing comprises monitoring trough plasma levels of the compound in the subject for a period of at least one week.

71. The method of any one of Claims 67-69, wherein testing comprises monitoring trough plasma levels of the compound in the subject for a period of at least ten weeks.

72. The method of any one of Claims 67-71, wherein the subject is an intermediate or extensive/ultra rapid P450 metabolizer enzyme if plasma trough levels of the compound in the subject are below 5 ng/ml and the adjusted effective amount of the compound is selected to result in trough levels of the compound in the subject of at least 5 ng/mg.

73. The method of any one of Claims 67-71, wherein the subject is a poor P450 enzyme if plasma trough levels of the compound are at least 5 ng/ml after being administered an effective amount of the compound.

74. The method of any one of Claims 67 or 68, wherein testing in step b) occurs before or after treatment begins.

75. The method of any one of Claims 67-74, wherein the P450 enzyme is CYP2D6 enzyme and/or CYP3A4 enzyme.

76. The method of any one of Claims 67-75, wherein the subject is a poor P450 metabolizer as a result of being co-administered an inhibitor of CYP2D6 enzyme or CYP3A4 enzyme.

77. The method of claim 76, wherein the inhibitor is selected from the group consisting of paroxetine, fluoxetine, quinidine, and ketoconazole.

78. The method of claim 67 or 69, wherein the testing in step b) is by genotyping for P450 enzyme.

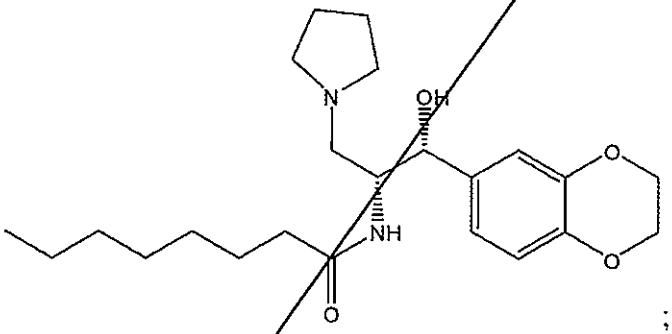
79. The method of Claim 78 wherein the P450 enzyme is CYP2D6.

80. The method of any one of Claims 67-79, wherein the effective amount of the compound is a twice daily dose of from 25 milligrams to 200 milligrams.

81. The method of any one of Claims 67-79, wherein the effective amount of the compound is a twice daily dose of 50 milligrams.

82. A method of treating a subject with Fabry disease comprising the steps of:

a) administering to the subject an effective amount of a compound represented by the following structural formula:



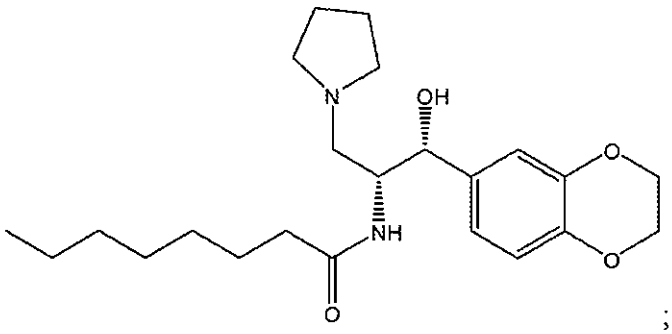
; or a pharmaceutically acceptable salt thereof;

b) assessing trough plasma levels of the compound in the subject;

c) adjusting the amount of compound administered to the subject so that the trough plasma levels of the compound in the subject are at least 5 ng/ml.

83. A method of treating a subject with Gaucher disease comprising the steps of:

a) administering to the subject an effective amount of a compound represented by the following structural formula:



; or a pharmaceutically acceptable salt thereof;

b) assessing trough plasma levels of the compound in the subject;

c) adjusting the amount of compound administered to the subject so that the trough plasma levels of the compound in the subject are at least 5 ng/ml.

84. The method of Claim 82 or 83, wherein the subject is tested before treatment with the compound begins to assess whether the subject is a poor, intermediate or extensive/ultra rapid P450 metabolizer, and wherein the effective amount is determined based on whether the subject is a poor, intermediate or extensive/ultra rapid P450 metabolizer.

85. The method of Claim 82, 83 or 84, wherein the compound is the hemitartrate salt of any one of Claims 1-15.

86. The method of Claim 82 or 83, wherein in step b) the C_{max} of the compound in the subject is also assessed and wherein in step c) the amount of compound administered to the subject is adjusted so that the trough plasma levels of the compound in the subject are at least 5 ng/ml and the C_{max} of the compound in the subject are below 100 ng/ml.

87. The method of any one of Claims 82-86, wherein the P450 enzyme is CYP2D6 enzyme and/or CYP3A4 enzyme.

88. The method of any one of Claims 82-86, wherein the subject is a poor P450 enzyme metabolizer as a result of co-administration of a P450 enzyme inhibitor.

89. The method of Claim 88, wherein the inhibitor is selected from the group consisting of paroxetine, fluoxetine, quinidine, and ketoconazole.

90. The method of any one of Claims 82-86, wherein P450 enzyme expression in the subject is assessed to determine whether the subject is a poor, intermediate or extensive/ultra rapid P450 metabolizer.

91. The method of Claim 90 wherein the p450 enzyme is CYP2D6 or CYP3A4.

92. The method of any of Claims 82-90, wherein the effective amount of the compound is a twice daily dose of from 25 milligrams to 200 milligrams.

93. The method of any of Claims 82-90, wherein the effective amount of the compound is a twice daily dose of 50 milligrams.

Claims:

1. An invention not claimed in any divisional application.