Claims:

1. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof for treating homozygous familial hypercholesterolemia; optionally in combination with an MTP inhibitor, an apoB-100 synthesis inhibitor, or a PCSK9 inhibitor.

2. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 1 for administration alone.

3. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 1 or 2 where the *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)acetic acid or a salt thereof is *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)acetic acid L-lysine dihydrate.

4. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of any one of claims 1-3 where the dose of *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)acetic acid or a salt thereof (when calculated as the free acid) is 20-200 mg/day, preferably 50-200 mg/day.

5. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of any one of claims 1-4 where the *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)acetic acid or a salt thereof is administered once/day.

6. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of any one of claims 1 and 3-5 where the *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)acetic acid or a salt thereof is administered in combination with an MTP inhibitor.

7. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 6 where the MTP inhibitor is administered once/day.

8. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of any one of claims 1 and 3-7 where the MTP inhibitor is

lomitapide or a salt thereof, SLx-4090, or JTT-130; such as lomitapide or a salt thereof, such as lomitapide mesylate.

9. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 8 where the MTP inhibitor is lomitapide or a salt thereof and the dose of the lomitapide or a salt thereof (when calculated as the mesylate salt) is 10-100 mg/day, preferably 20-80 mg/day, more preferably 30-60 mg/day.

10. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of any one of claims 1 and 3-9 where the MTP inhibitor is lomitapide or a salt thereof and the *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)-thio)-2-methylphenoxy)acetic acid or a salt thereof and the lomitapide or a salt thereof are administered in separate dosage forms.

11. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of any one of claims 1 and 3-9 where the MTP inhibitor is lomitapide or a salt thereof and the *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)-thio)-2-methylphenoxy)acetic acid or a salt thereof and the lomitapide or a salt thereof are administered in a single dosage form.

12. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of any one of claims 1 and 3-5 where the *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)acetic acid or a salt thereof is administered in combination with an apoB-100 synthesis inhibitor; such as mipomersen or a salt thereof; such as mipomersen sodium.

13. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of any one of claims 1, 3-5, and 12 where the apoB-100 synthesis inhibitor is mipomersen or a salt thereof and the dose of the mipomersen or a salt thereof (when calculated as the sodium salt) is 100-300 mg, preferably 200 mg, administered once/week.

14. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of any one of claims 1 and 3-5 where the *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)acetic acid or a salt thereof is administered in combination with a PCSK9 inhibitor.

15. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 14 where the PCSK9 inhibitor is evolocumab, alirocumab, bococizumab, RG7652, LGT-209, LY3015014, ALN-PCSsc, or BMS-962476.

16. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 15 where the PCSK9 inhibitor is evolocumab.

17. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 16 where the where the dose of evolocumab is 140 mg every 2 weeks or 420 mg every 4 weeks.

18. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 15 where the PCSK9 inhibitor is alirocumab.

19. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 18 where the where the dose of alirocumab is 150 mg every 2 weeks or 150, 200 or 300 mg every 4 weeks, such as 150 mg every 2 weeks.

20. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 15 where the PCSK9 inhibitor is bococizumab.

21. *(R)*-2-(4-((2-Ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)-acetic acid or a salt thereof of claim 20 where the where the dose of bococizumab is 50, 100, or 150 mg every 2 weeks or 200 or 300 mg every 4 weeks, such as 150 mg every 2 weeks.

22. A pharmaceutical formulation when used for the treatment of homozygous familial hypercholesterolemia, comprising *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)-thio)-2-methylphenoxy)acetic acid or a salt thereof; optionally in combination with an MTP inhibitor.

23. The use of *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)acetic acid or a salt thereof; optionally in combination with an MTP inhibitor, an apoB-100 synthesis inhibitor, or a PCSK9 inhibitor; in the manufacture of a medicament for the treatment of homozygous familial hypercholesterolemia.

24. A method of treating homozygous familial hypercholesterolemia by administering *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)propyl)thio)-2-methylphenoxy)acetic acid or a salt thereof; optionally in combination with an MTP inhibitor, an apoB-100 synthesis inhibitor, or a PCSK9 inhibitor.

5

CLAIMS

1. The use of a compound that is *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)phenoxy)-propyl)thio)-2-methylphenoxy)acetic acid or a salt thereof in the manufacture of a medicament for treating homozygous familial hypercholesterolemia in a subject.

2. The use of claim 1 where the compound is *(R)*-2-(4-((2-ethoxy-3-(4-(trifluoromethyl)-phenoxy)propyl)thio)-2-methylphenoxy)acetic acid L-lysine dihydrate.

3. The use of claim 1or 2 where the dose of the compound (when calculated as the free acid) is 20-200 mg/day.

4. The use of claim 3 where the dose of the compound (when calculated as the free acid) is 50-200 mg/day.

5. The use of any one of claims 1 to 4 where the compound is administered once/day.

6. The use of any one of claims 1 to 5, where the subject is administered only the medicament.

7. The use of any one of claims 1 to 5 where the subject is also administered an MTP inhibitor.

8. The use of claim 7 where the MTP inhibitor is lomitapide or a salt thereof, SLx-4090, or JTT-130.

9. The use of claim 8 where the MTP inhibitor is lomitapide or a salt thereof.

10. The use of claim 9 where the dose of the lomitapide or a salt thereof (when calculated as the mesylate salt) is 10-100 mg/day.

11. The use of any one of claims 7 to 10 where the MTP inhibitor is administered once/day.

12. The use of any one of claims 1 to 5 where the subject is also administered an apoB-100 synthesis inhibitor.

13. The use of claim 12 where the apoB-100 synthesis inhibitor is mipomersen or a salt thereof.

14. The use of claim 13 where the dose of the mipomersen or a salt thereof (when calculated as the sodium salt) is 100-300 mg, administered once/week.

15. The use of any one of claims 1 to 5 where the subject is also administered a PCSK9 inhibitor.

16. The use of claim 15 where the PCSK9 inhibitor is evolocumab, alirocumab, bococizumab, RG7652, LGT-209, LY3015014, ALN-PCSsc, or BMS-962476.

17. The use of claim 16 where the PCSK9 inhibitor is evolocumab.

18. The use of claim 17 where the dose of evolocumab is 140 mg every 2 weeks or 420 mg every 4 weeks.

19. The use of claim 15 where the PCSK9 inhibitor is alirocumab.

20. The use of claim 19 where the dose of alirocumab is 150 mg every 2 weeks or 150, 200 or 300 mg every 4 weeks.

21. The use of claim 15 where the PCSK9 inhibitor is bococizumab.

22. The use of claim 21 where the dose of bococizumab is 50, 100, or 150 mg every 2 weeks or 200 or 300 mg every 4 weeks.