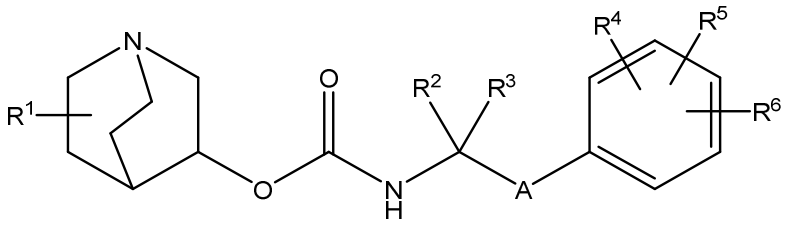
CLAIMS

1. Use of a compound in the manufacture of a medicament for treating or preventing dermatological disorders caused by GL-3 accumulation, in a subject in need thereof, wherein the compound is a compound of formula (I),



(I)

or a pharmaceutically acceptable salt thereof, wherein:

R^1 is selected from hydrogen, halogen (e.g., fluorine), cyano, nitro, hydroxy, thio, amino, C_{1-6}-alkyl (e.g., methyl or ethyl), C_{2-6}-alkenyl, C_{2-6}-alkynyl, C_{1-6}-alkyloxy, C_{2-6}-alkenyloxy, and C_{2-6}-alkynyloxy, wherein said alkyl, alkenyl, alkynyl, alkyloxy, alkenyloxy, or alkynyloxy is optionally substituted with one or more (e.g., 1, 2 or 3) groups selected from halogen, cyano, nitro, hydroxy, thio or amino;

R^2 and R^3 are independently selected from C_{1-3}-alkyl, optionally substituted by one or more (*e.g.* 1, 2 or 3) halogens, or R^2 and R^3 together form a cyclopropyl or cyclobutyl group, optionally substituted by one or more (*e.g.* 1 or 2) halogens;

R^4, R^5 and R^6 are each independently selected from hydrogen, halogen, nitro, hydroxy, thio, amino, C_{1-6}-alkyl, and C_{1-6}-alkyloxy, wherein said alkyl or alkyloxy is optionally substituted by one or more (*e.g.* 1, 2 or 3) groups selected from halogen, hydroxy, cyano, and C_{1-6}-alkyloxy; and

A is a 5- or 6-membered aryl or heteroaryl group (e.g., phenyl or thiazolyl), optionally substituted with 1, 2 or 3 groups independently selected from halogen, hydroxy, thio, amino, nitro, C_{1-6}alkoxy and C_{1-6}alkyl.

2. The use of claim 1, wherein:

R^1 is selected from hydrogen, fluorine, methyl and ethyl, wherein said methyl or ethyl is optionally substituted by 1 or 2 groups selected from halogen, hydroxy, thio or amino;

R^2 and R^3 are each independently selected from methyl and ethyl groups, optionally substituted with one or more fluorines;

R^4 is selected from a halogen (e.g., fluorine), C_{1-3}-alkyl (e.g., methyl) and C_{1-3}-alkyloxy (e.g., methoxy or ethoxy), wherein said alkyl or alkyoxy is optionally substituted by one or more (e.g., 1, 2 or 3) groups selected from a halogen and C_{1-3}-alkyloxy (e.g., methoxy or ethoxy);

R^5 and R^6 are each hydrogen;

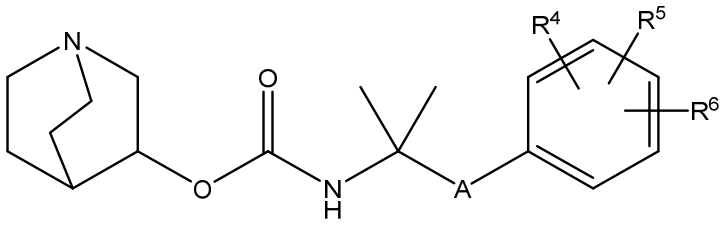
R^4 is fluorine or 2-methoxyethoxy, and R^5 and R^6 are hydrogen;

R^4 is positioned at the 4-position of the phenyl ring to which it is attached (i.e., *para* to the A substituent);

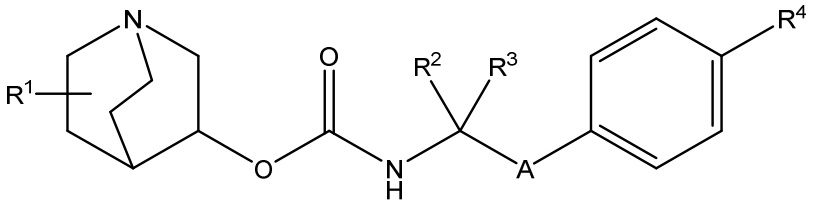
A is phenyl, optionally substituted with 1, 2 or 3 groups independently selected from halogen, hydroxy, thio, amino, nitro, C_{1-6}alkoxy and C_{1-6}alkyl (e.g., methyl), optionally wherein the two groups attached to the A substituent are positioned in a 1,3- or a 1,4-relationship to each other (i.e., meta or para); and/or

A is a 5-membered heteroaryl group which contains 1 or 2 heteroatoms selected from N and S, optionally wherein the two groups attached to the A substituent are positioned in a 1,3- relationship to each other (i.e., meta).

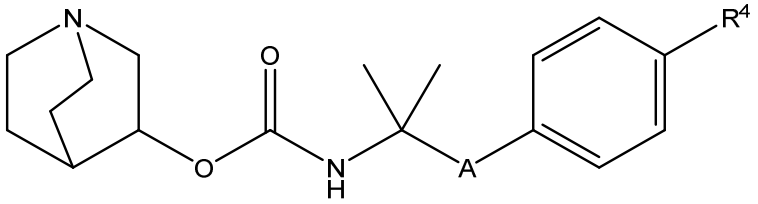
3. The use of claim 1 or claim 2, wherein said compound is a compound of formula (II), (III), (IV), (V), (VI), (VII), (VIII), (IX) or (XI),



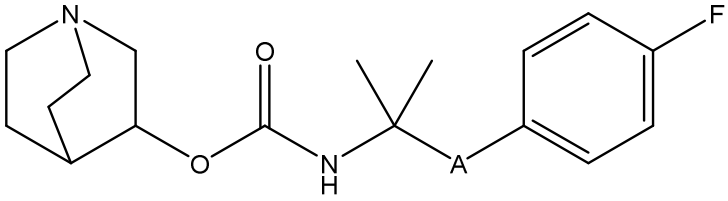
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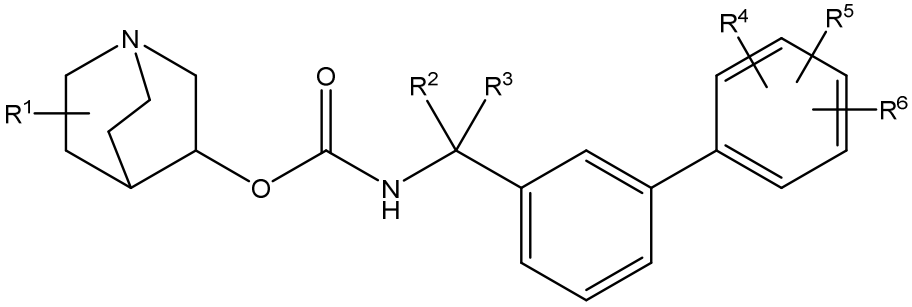
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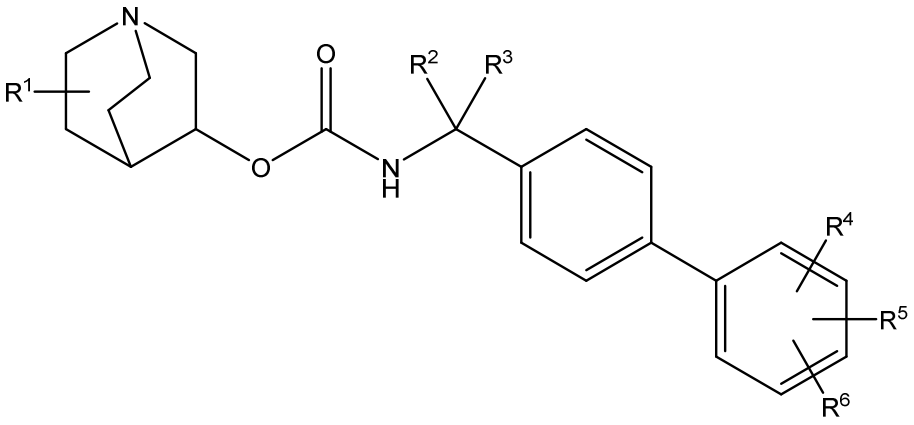
(IV)



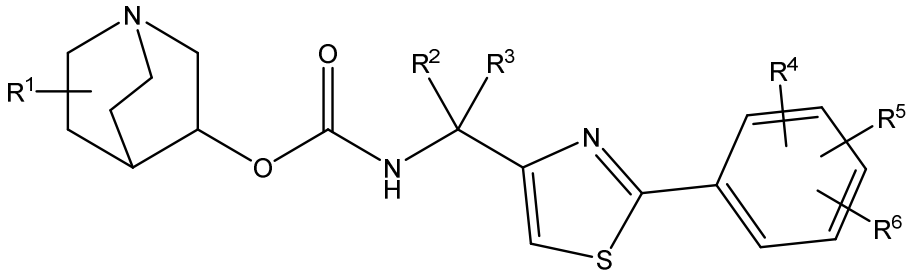
(V)



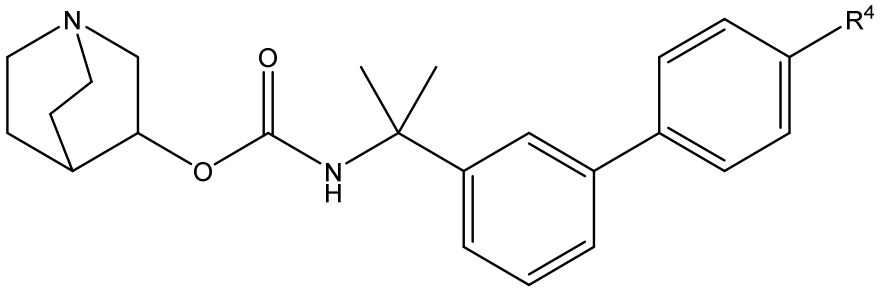
(VI)



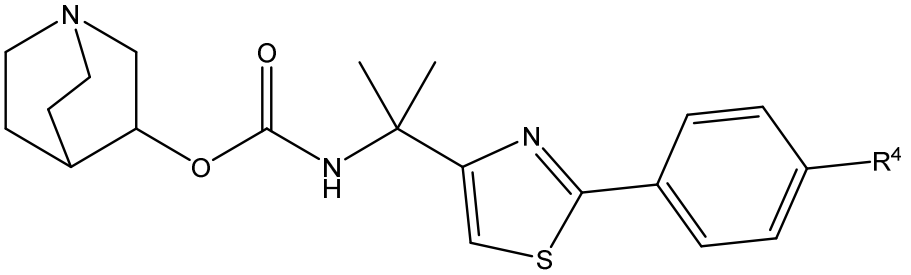
(VII)



(VIII)



(IX)



(XI)

or a pharmaceutically acceptable salt thereof.

4. The use of claim 3, wherein said compound is a compound of formula (IX) or (XI) and wherein R^4 is fluorine.

5. The use of claim 1, wherein said compound is selected from: quinuclidin-3-yl (2-(4'-fluoro-[1,1'-biphenyl]-3-yl)propan-2-yl)carbamate; (*S*)-quinuclidin-3-yl (2-(2-(4-fluorophenyl)thiazol-4-yl)propan-2-yl)carbamate; (*S*)-quinuclidin-3-yl (2-(4'-(2-methoxyethoxy)-[1,1'-biphenyl]-4-yl)propan-2-yl)carbamate; and the pharmaceutically acceptable salts thereof.

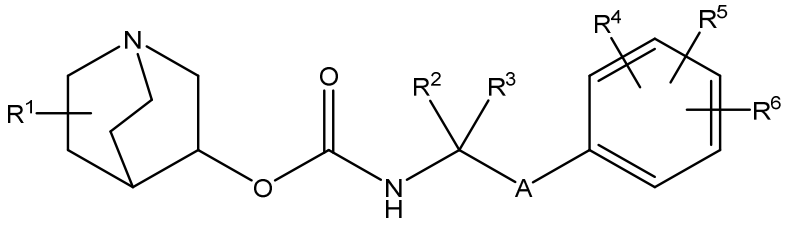
6. The use of any one of claims 1 to 5, wherein the dermatological disorder is one or more of angiokeratoma, hypohidrosis, anhidrosis, hyperhidrosis, lymphedema, and acroparesthesia.

7. The use of any one of claims 1 to 6, wherein the subject has marked accumulation of GL-3 in one or more of endothelial cells of superficial skin vessels, endothelial cells of deep skin vessels, smooth muscle cells of deep skin vessels, and perineurium cells, optionally wherein the reduction in skin GL-3 levels (e.g., GL-3 inclusions) is found in one or more of endothelial cells of superficial skin vessels, endothelial cells of deep skin vessels, smooth muscle cells of deep skin vessels, and perineurium cells, within 26 weeks or 52 weeks or 156 weeks.

8. The use of claim 7, wherein the subject has a GL-3 skin score (measured on a scale 0 to 4, as described in the Examples) of 2 or more prior to treatment, and the treatment results in a drop of at least one point on the scale, e.g., a drop of one point or two points.

9. The use of claim 7 or claim 8, wherein the subject has GL-3 inclusions in skin cells (e.g. endothelial cells) showing a cytoplasmic volume fraction of GL-3 inclusion of at least 0.25 (e.g., at least 0.27 or 0.3) prior to treatment, and the treatment results in a drop in the cytoplasmic volume fraction of GL-3 inclusions to less than 0.25 (e.g., less than 0.23 or less than 0.20, or less than 0.19).

10. Use of a compound in the manufacture of a medicament for treating or preventing pain, including neuropathic pain, gastrointestinal pain (e.g., abdominal pain), and peripheral neuropathy, in a subject in need thereof, wherein the compound is a compound of formula (I),



(I)

or a pharmaceutically acceptable salt thereof, wherein:

R^1 is selected from hydrogen, halogen (e.g., fluorine), cyano, nitro, hydroxy, thio, amino, C_{1-6}-alkyl (e.g., methyl or ethyl), C_{2-6}-alkenyl, C_{2-6}-alkynyl, C_{1-6}-alkyloxy, C_{2-6}-alkenyloxy, and C_{2-6}-alkynyloxy, wherein said alkyl, alkenyl, alkynyl, alkyloxy, alkenyloxy, or alkynyloxy is optionally substituted with one or more (e.g., 1, 2 or 3) groups selected from halogen, cyano, nitro, hydroxy, thio or amino;

R^2 and R^3 are independently selected from C_{1-3}-alkyl, optionally substituted by one or more (*e.g.* 1, 2 or 3) halogens, or R^2 and R^3 together form a cyclopropyl or cyclobutyl group, optionally substituted by one or more (*e.g.* 1 or 2) halogens;

R^4, R^5 and R^6 are each independently selected from hydrogen, halogen, nitro, hydroxy, thio, amino, C_{1-6}-alkyl, and C_{1-6}-alkyloxy, wherein said alkyl or alkyloxy is optionally substituted by one or more (*e.g.* 1, 2 or 3) groups selected from halogen, hydroxy, cyano, and C_{1-6}-alkyloxy; and

A is a 5- or 6-membered aryl or heteroaryl group (e.g., phenyl or thiazolyl), optionally substituted with 1, 2 or 3 groups independently selected from halogen, hydroxy, thio, amino, nitro, C_{1-6}alkoxy and C_{1-6}alkyl.

11. The use of claim 10, wherein:

R^1 is selected from hydrogen, fluorine, methyl and ethyl, wherein said methyl or ethyl is optionally substituted by 1 or 2 groups selected from halogen, hydroxy, thio or amino;

R^2 and R^3 are each independently selected from methyl and ethyl groups, optionally substituted with one or more fluorines;

R^4 is selected from a halogen (e.g., fluorine), C_{1-3}-alkyl (e.g., methyl) and C_{1-3}-alkyloxy (e.g., methoxy or ethoxy), wherein said alkyl or alkyoxy is optionally substituted by

one or more (e.g., 1, 2 or 3) groups selected from a halogen and C_{1-3}-alkyloxy (e.g., methoxy or ethoxy);

R^5 and R^6 are each hydrogen;

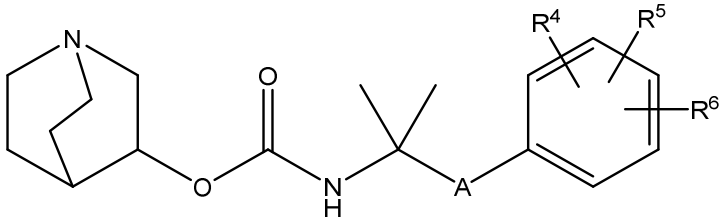
R^4 is fluorine or 2-methoxyethoxy, and R^5 and R^6 are hydrogen;

R^4 is positioned at the 4-position of the phenyl ring to which it is attached (i.e., *para* to the A substituent);

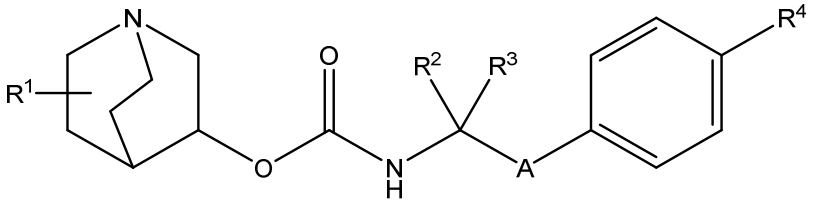
A is phenyl, optionally substituted with 1, 2 or 3 groups independently selected from halogen, hydroxy, thio, amino, nitro, C_{1-6}alkoxy and C_{1-6}alkyl (e.g., methyl), optionally wherein the two groups attached to the A substituent are positioned in a 1,3- or a 1,4-relationship to each other (i.e., meta or para); and/or

A is a 5-membered heteroaryl group which contains 1 or 2 heteroatoms selected from N and S, optionally wherein the two groups attached to the A substituent are positioned in a 1,3- relationship to each other (i.e., meta).

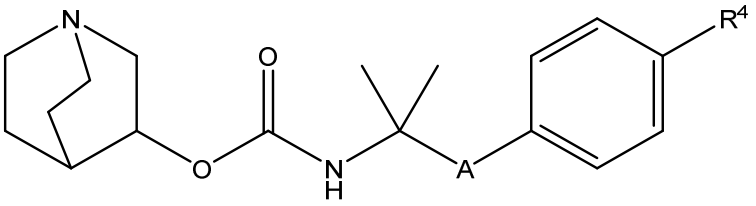
12. The use of claim 10 or claim 11, wherein said compound is a compound of formula (II), (III), (IV), (V), (VI), (VII), (VIII), (IX) or (XI),



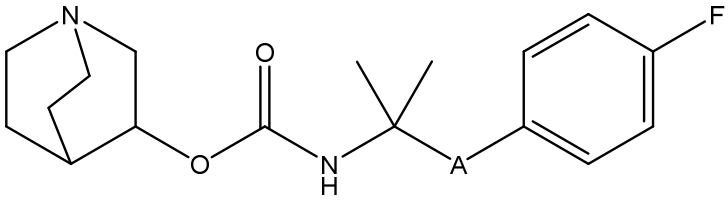
(II)



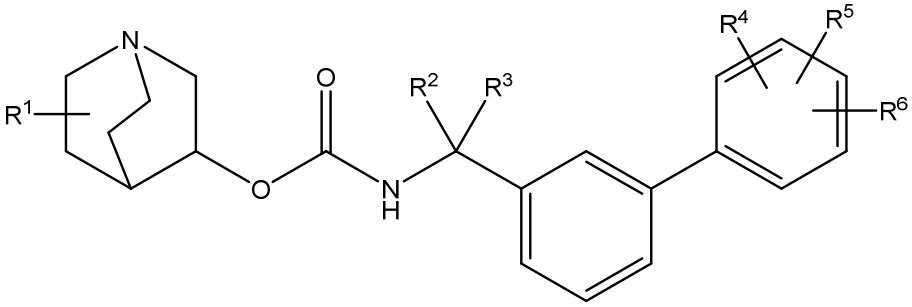
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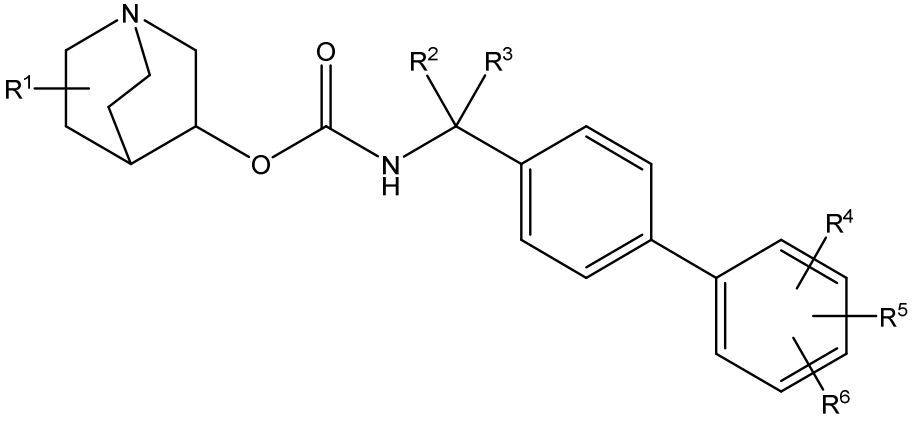
(IV)



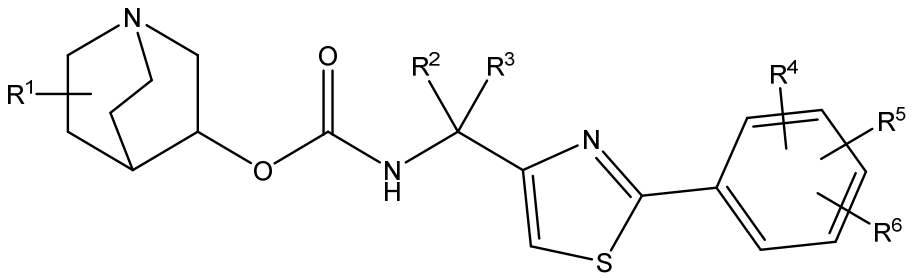
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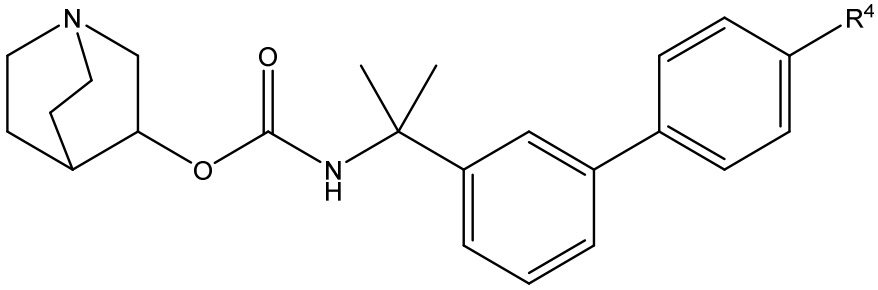
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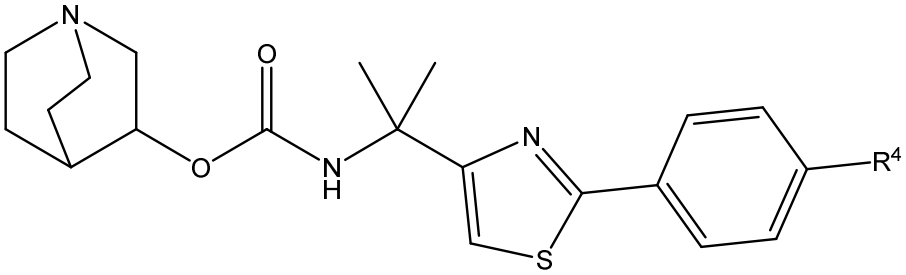
(VII)



(VIII)



(IX)



(XI)

or a pharmaceutically acceptable salt thereof.

13. The use of claim 12, wherein said compound is a compound of formula (IX) or (XI) and wherein R^4 is fluorine.

14. The use of claim 10, wherein said compound is selected from: quinuclidin-3-yl (2-(4'-fluoro-[1,1'-biphenyl]-3-yl)propan-2-yl)carbamate; (*S*)-quinuclidin-3-yl (2-(2-(4-fluorophenyl)thiazol-4-yl)propan-2-yl)carbamate; (*S*)-quinuclidin-3-yl (2-(4'-(2-methoxyethoxy)-[1,1'-biphenyl]-4-yl)propan-2-yl)carbamate; and the pharmaceutically acceptable salts thereof.

15. The use of any one of claims 10 to 14, wherein the treatment further comprises concurrently administering a second active agent, e.g., a second compound capable of treating or preventing pain in a patient in need thereof.

16. The use of any one of claims 10 to 15, wherein the pain is gastrointestinal pain (e.g., abdominal pain).

17. The use of any one of claims 1 to 16, wherein the subject has marked accumulation of GL-3 in tissues (e.g., in skin capillary endothelial cells or in plasma).

18. The use of any one of claims 1 to 17, wherein the treatment results in:

(a) reduction in GL-3 concentration in plasma of at least 20% after 2 weeks or 4 weeks or 8 weeks of treatment, e.g., at least 30%, at least 40%, or at least 50%, and/or reduction in GL-3 concentration in plasma of at least 40% after 26 weeks or 52 weeks or 104 weeks, e.g., at least 50%, at least 60% or at least 70%;

(b) a reduction in severity of pain (e.g., in bodily pain and/or gastrointestinal pain (e.g., abdominal pain)) within 26 weeks or 52 weeks or 156 weeks; and/or

(c) reduction in GL-3 levels in skin (e.g., skin capillary endothelial cells) within 26 weeks or 52 weeks or 156 weeks, e.g., as shown by the extent of GL-3 inclusions.

19. The use of any one of claims 1 to 18, wherein said medicament is for administration by systemic administration, *e.g.* via a non-parenteral route, optionally wherein said medicament is for oral administration; and/or wherein the medicament is for administration of a daily dose of about 1 mg to about 50 mg of the compound, e.g., from 5 to 50 mg, or from 10 to 40 mg, or from 10 to 30 mg, or from 10 to 20 mg, or from 20 to 30 mg, or from 30 to 40 mg, or from 40 to 50 mg, or from 5 to 25 mg, or from 20 to 50 mg, or from 5 to 15 mg, or from 15 to 30 mg, or about 15 mg.

20. The use of any one of claims 1 to 19, wherein the compound is in a pharmaceutically acceptable acid addition salt form selected from the hydrochloride, hydrobromide, hydroiodide, nitrate, sulfate, bisulfate, phosphate, acid phosphate, acetate, lactate, citrate, acid citrate, tartrate, bitartrate, succinate, hydroxysuccinate, malate, maleate, fumarate, gluconate, saccharate, benzoate, methanesulfonate, and pamoate.

21. The use of any one of claims 1 to 20, wherein the compound is (S)-quinuclidin-3-yl (2-(2-(4-fluorophenyl)thiazol-4-yl)propan-2-yl)carbamate in malate salt form.

22. The use of any one of claims 1 to 21, wherein the subject has Fabry disease.

23. The use of any one of claim 1 to 22, wherein the subject undergoes concurrent treatment with enzyme replacement therapy (ERT), e.g., using agalsidase alpha or agalsidase beta, or using a glucocerebrosidase (e.g., imiglucerase, velaglucerase, or taliglucerase), or an alpha-galactosidase inhibitor (e.g., migalastat).