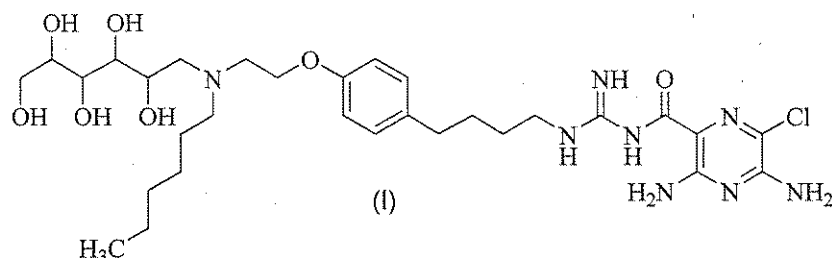


CLAIMS

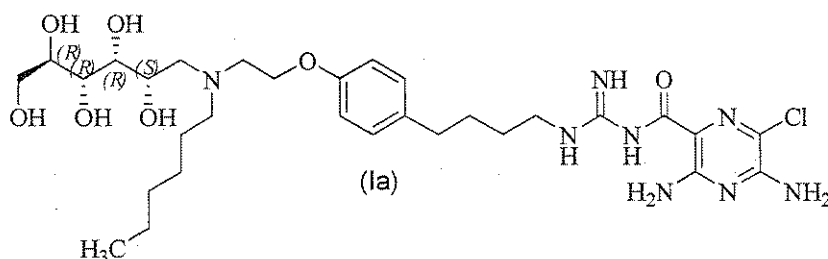
That Which is Claimed Is:

1. A compound of the formula:



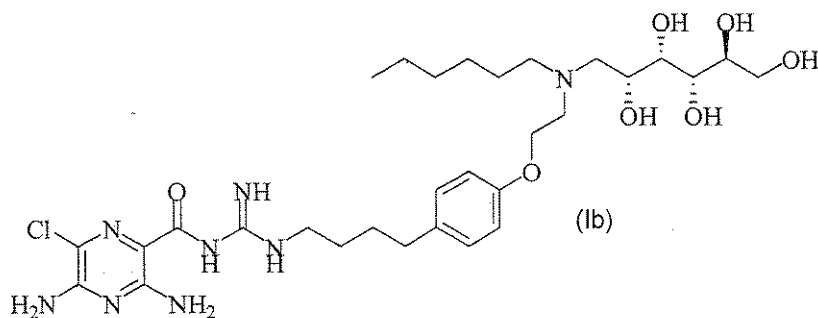
or a pharmaceutically acceptable salt thereof.

2. A compound of Claim 1 which is 3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3R,4R,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide, having the formula:

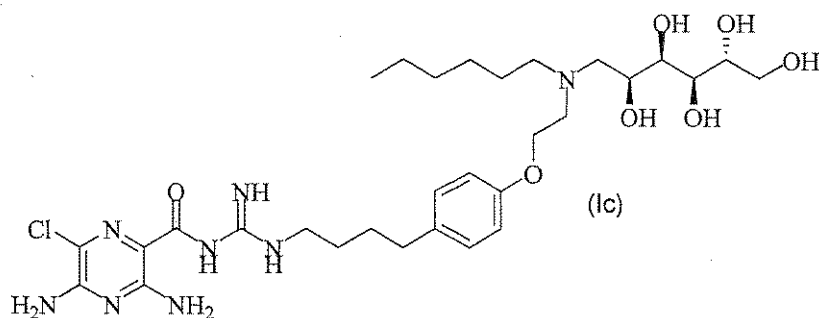


or a pharmaceutically acceptable salt thereof.

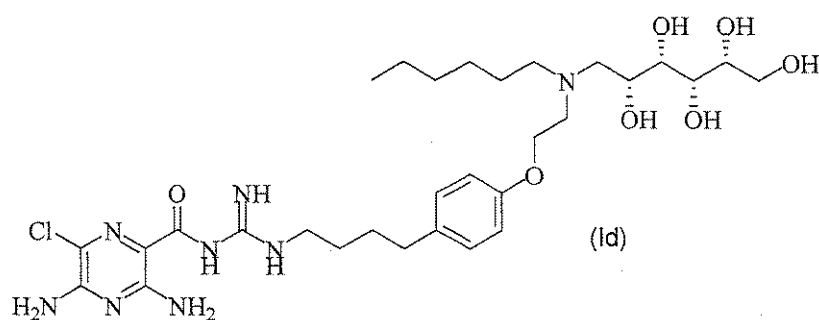
3. A compound of Claim 1 selected from the group of:
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3S,4S,5S)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide



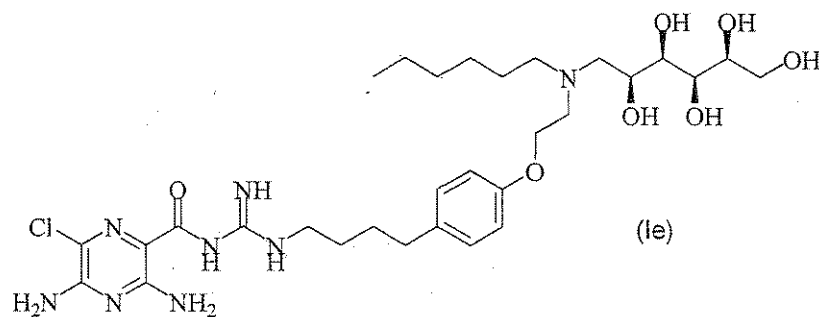
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3R,4R,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide



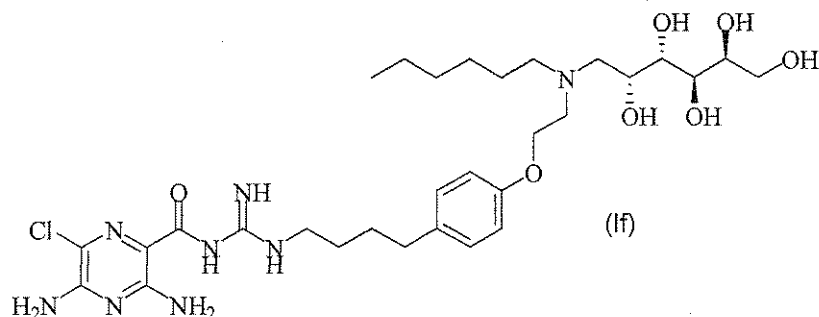
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3S,4S,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide



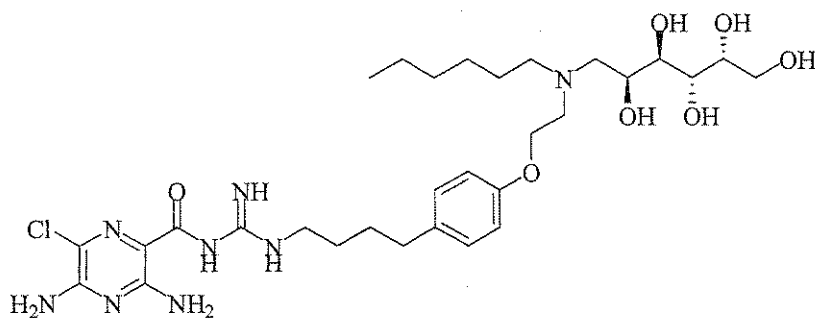
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3R,4R,5S)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide



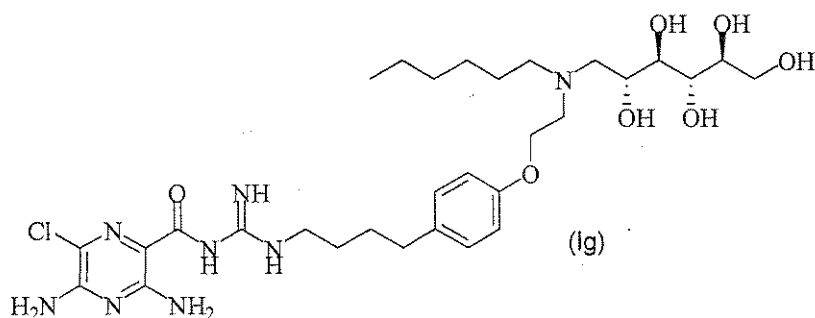
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3S,4R,5S)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide



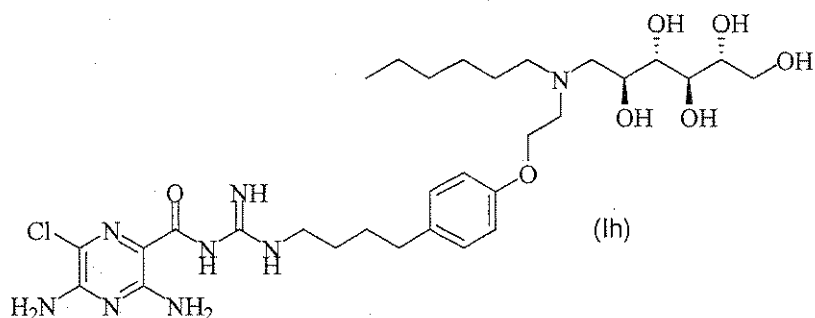
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3R,4S,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide



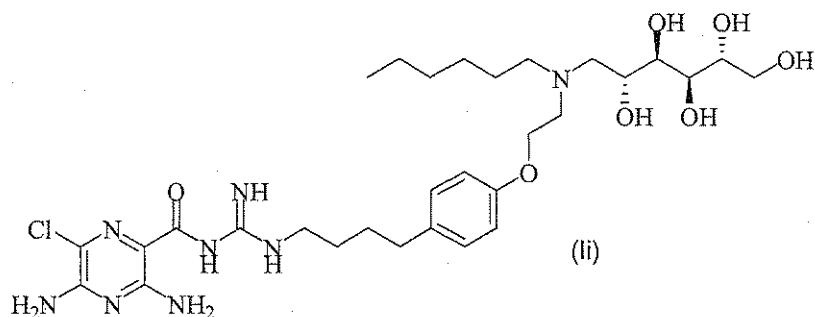
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3R,4S,5S)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide



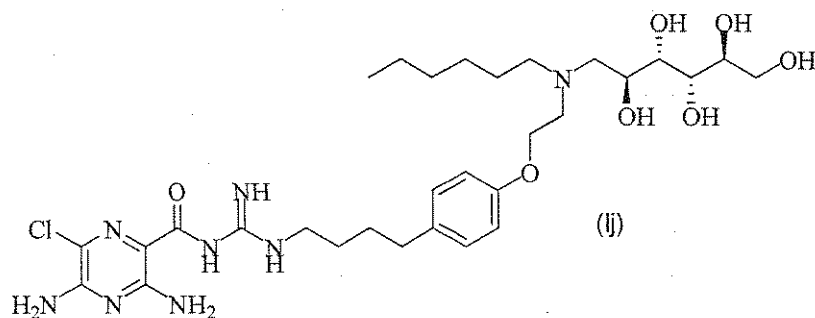
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3S,4R,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide



3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3R,4R,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide



3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3S,4S,5S)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide



and pharmaceutically acceptable salts thereof.

4. A pharmaceutical composition comprising a pharmaceutically effective amount of a compound of Claims 1, 2, or 3, or a pharmaceutically acceptable salt thereof, and a pharmaceutically acceptable carrier or excipient.

5. The pharmaceutical composition of Claim 4 wherein the compound is 3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3R,4R,5R)-2,3,4,5,6-pentahydroxyhexyl) amino)ethoxy)phenyl)butyl)carbamidoyl)pyrazine-2-carboxamide, or a pharmaceutically acceptable salt thereof.
6. The composition according to either of Claims 4 or 5, wherein said composition is suitable for inhalation.
7. The composition according to either of Claims 4 or 5, wherein said composition is a solution for aerosolization and administration by nebulizer.
8. The composition according to either of Claims 4 or 5, wherein said composition is suitable for administration by metered dose inhaler.
9. The composition according to either of Claims 4 or 5, wherein said composition is a dry powder suitable for administration by dry powder inhaler.
10. The composition according to either of Claims 4 or 5 further comprising a pharmaceutically effective amount of a therapeutically active agent selected from anti-inflammatory agents, anticholinergic agents, β -agonists, P2Y2 receptor agonists, peroxisome proliferator-activated receptor agonists, kinase inhibitors, antiinfective agents and antihistamines.
11. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for treating a pulmonary disease in a human in need thereof.
12. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for blocking sodium channels in a human.
13. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for promoting hydration of mucosal surfaces or restoring mucosal defense in a human.

14. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for treating a disease selected from the group of reversible or irreversible airway obstruction, chronic obstructive pulmonary disease (COPD), asthma, bronchiectasis (including bronchiectasis due to conditions other than cystic fibrosis), acute bronchitis, chronic bronchitis, post-viral cough, cystic fibrosis, emphysema, pneumonia, panbronchiolitis, transplant-associated bronchiolitis, and ventilator-associated tracheobronchitis or preventing ventilator-associated pneumonia in a human in need thereof.
15. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for treating chronic obstructive pulmonary disease (COPD) in a human in need thereof.
16. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for treating cystic fibrosis in a human in need thereof.
17. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for treating dry mouth (xerostomia), dry skin, vaginal dryness, sinusitis, rhinosinusitis, or nasal dehydration, including nasal dehydration brought on by administering dry oxygen, dry eye or Sjogren's disease, promoting ocular or corneal hydration, treating distal intestinal obstruction syndrome, treating otitis media, primary ciliary dyskinesia, distal intestinal obstruction syndrome, esophagitis, constipation, or chronic diverticulitis in a human in need thereof.
18. A compound according to any of claims 1-3, or a pharmaceutically acceptable salt thereof for use as a medicament.
19. A compound according to any of claims 1-3, or a pharmaceutically acceptable salt thereof for use in treating a pulmonary disease.
20. A compound according to any of claims 1-3 or a pharmaceutically acceptable salt thereof for use in treating a disease associated with reversible or irreversible airway obstruction, chronic obstructive pulmonary disease (COPD), asthma, bronchiectasis (including

bronchiectasis due to conditions other than cystic fibrosis), acute bronchitis, chronic bronchitis, post-viral cough, cystic fibrosis, emphysema, pneumonia, panbronchiolitis, transplant-associated bronchiolitis, and ventilator-associated tracheobronchitis or preventing ventilator-associated pneumonia in a human in need thereof.

21. A compound according to any of claims 1-3, or a pharmaceutically acceptable salt thereof, for use in treating dry mouth (xerostomia), dry skin, vaginal dryness, sinusitis, rhinosinusitis, or nasal dehydration, including nasal dehydration brought on by administering dry oxygen, dry eye or Sjogren's disease, promoting ocular or corneal hydration, treating distal intestinal obstruction syndrome, treating otitis media, primary ciliary dyskinesia, distal intestinal obstruction syndrome, esophagitis, constipation, or chronic diverticulitis in a human in need thereof.

22. A composition comprising a compound according to any one of claims 1-3, or a pharmaceutically acceptable salt thereof, for use in the preparation of a medicament for treating a disease associated with reversible or irreversible airway obstruction, chronic obstructive pulmonary disease (COPD), asthma, bronchiectasis (including bronchiectasis due to conditions other than cystic fibrosis), acute bronchitis, chronic bronchitis, post-viral cough, cystic fibrosis, emphysema, pneumonia, panbronchiolitis, transplant-associated bronchiolitis, and ventilator-associated tracheobronchitis or preventing ventilator-associated pneumonia.

23. A composition comprising a compound according to any one of claims 1-3, or a pharmaceutically acceptable salt thereof, for use in the preparation of a medicament for treating dry mouth (xerostomia), dry skin, vaginal dryness, sinusitis, rhinosinusitis, or nasal dehydration, including nasal dehydration brought on by administering dry oxygen, dry eye or Sjogren's disease, promoting ocular or corneal hydration, treating distal intestinal obstruction syndrome, treating otitis media, primary ciliary dyskinesia, distal intestinal obstruction syndrome, esophagitis, constipation, or chronic diverticulitis.

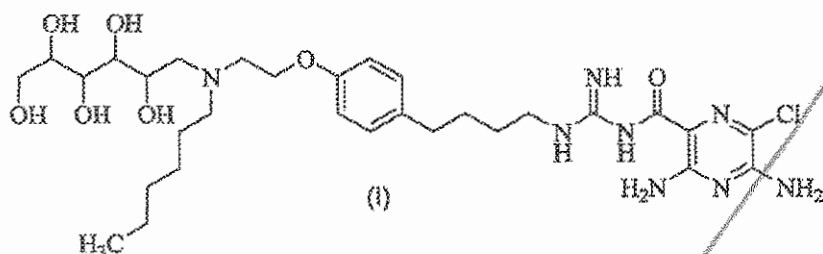
24. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for preventing, mitigating, and/or treating deterministic health effects to the respiratory tract and/or other bodily organs caused by respirable aerosols containing radionuclides in a human in need thereof.

CLAIMS

That Which Is Claimed Is:

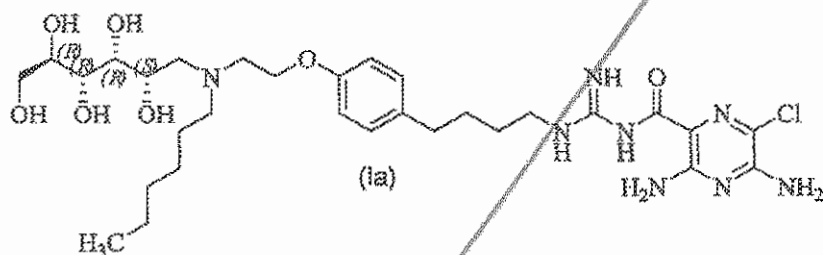
1. A compound of the formula:

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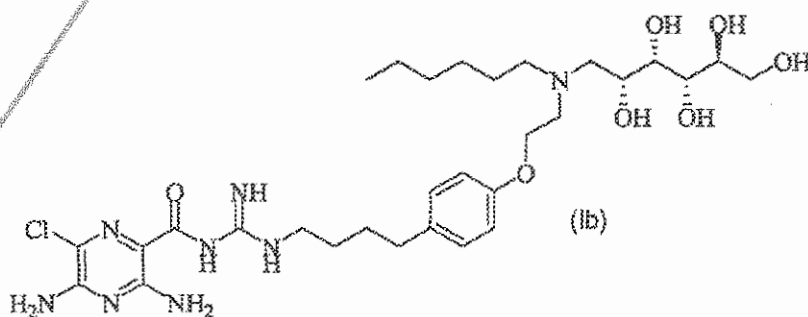
or a pharmaceutically acceptable salt thereof.

2. A compound of Claim 1 which is 3,5-diamino-6-chloro-N-(N-(4-(4-(2-
 10 (hexyl((2S,3R,4R,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)
 phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide, having the formula:



or a pharmaceutically acceptable salt thereof.

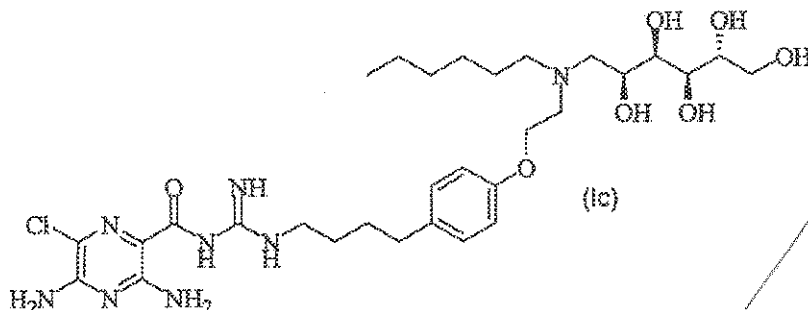
3. A compound of Claim 1 selected from the group of:
 3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3S,4S,5S)-2,3,4,5,6-
 15 pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-
 carboxamide



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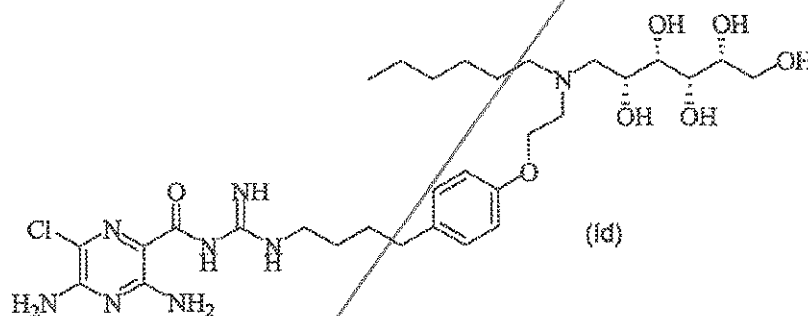
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3R,4R,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamidoyl)pyrazine-2-carboxamide

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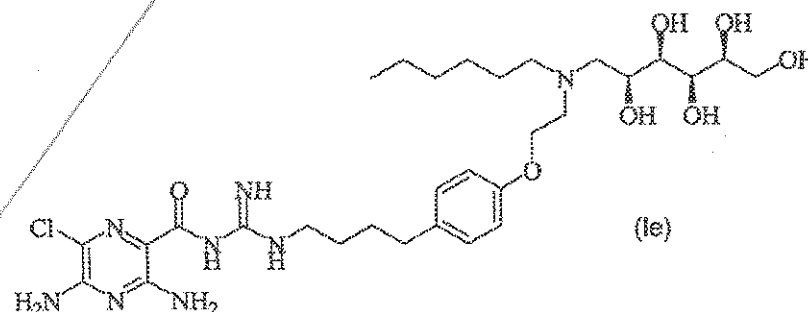
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3S,4S,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamidoyl)pyrazine-2-carboxamide

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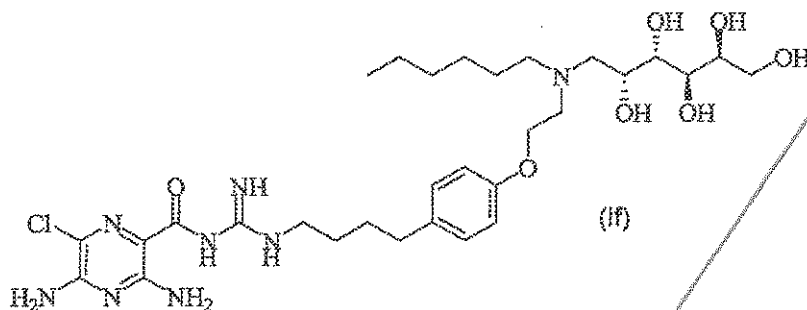
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3R,4R,5S)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamidoyl)pyrazine-2-carboxamide

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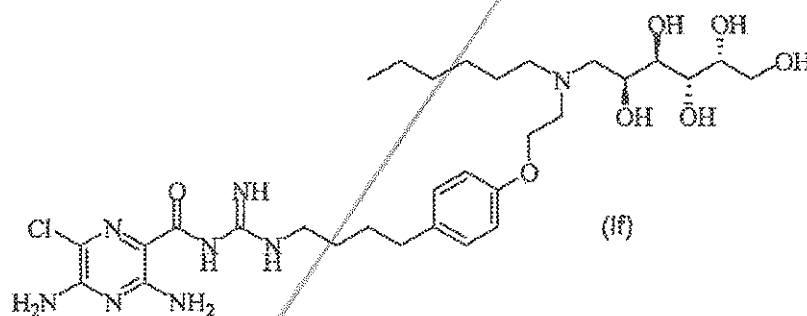
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3S,4R,5S)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide

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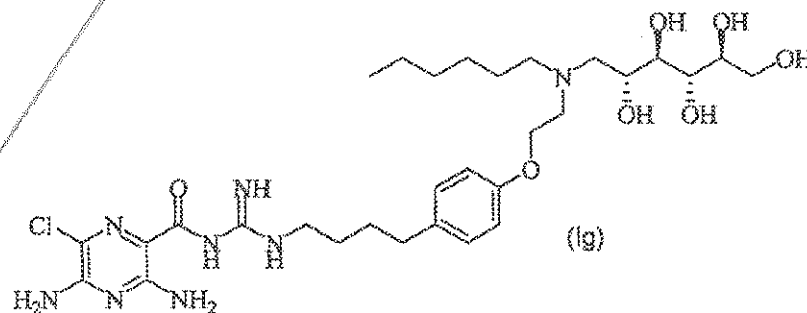
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3R,4S,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide

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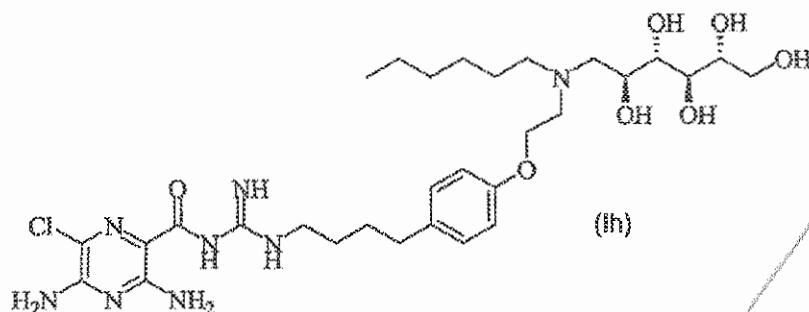
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3R,4S,5S)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide

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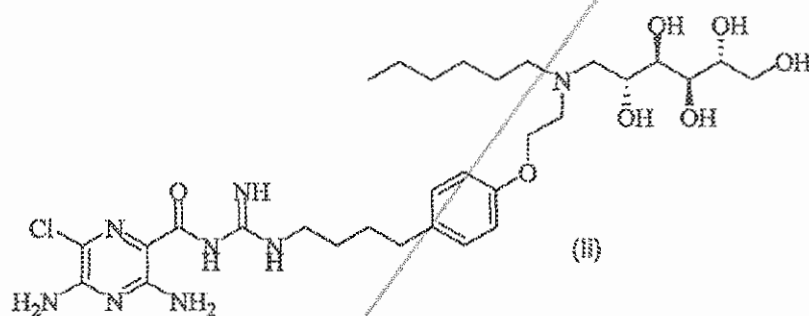
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3S,4R,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide

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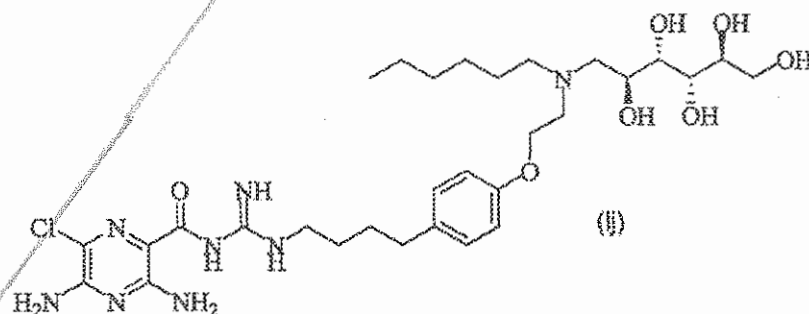
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3R,4R,5R)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide

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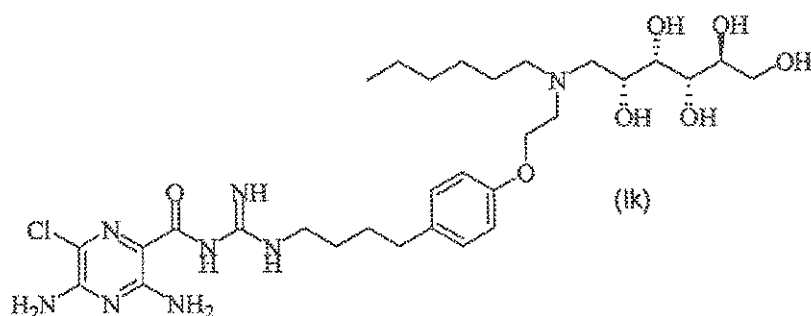
3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3S,4S,5S)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide

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3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2R,3S,4S,5S)-2,3,4,5,6-pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbamimidoyl)pyrazine-2-carboxamide

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; and

3,5-diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3R,4R,5R)-2,3,4,5,6-
 pentahydroxyhexyl)amino)ethoxy)phenyl)butyl)carbarnimidoyl)pyrazine-2-
 5 carboxamide

or a pharmaceutically acceptable salt thereof.

4. A pharmaceutical composition comprising a pharmaceutically effective
 10 amount of a compound of Claims 1, 2, or 3, or a pharmaceutically acceptable salt
 thereof, and a pharmaceutically acceptable carrier or excipient.

5. The pharmaceutical composition of Claim 4 wherein the compound is 3,5-
 diamino-6-chloro-N-(N-(4-(4-(2-(hexyl((2S,3R,4R,5R)-2,3,4,5,6- pentahydroxyhexyl)
 15 amino)ethoxy) phenyl)butyl)carbarnimidoyl)pyrazine-2-carboxamide, or a
 pharmaceutically acceptable salt thereof.

6. The composition according to either of Claims 4 or 5, wherein said
 composition is suitable for inhalation.

7. The composition according to either of Claims 4 or 5, wherein said
 composition is a solution for aerosolization and administration by nebulizer.

8. The composition according to either of Claims 4 or 5, wherein said
 25 composition is suitable for administration by metered dose inhaler.

9. The composition according to either of Claims 4 or 5, wherein said
 composition is a dry powder suitable for administration by dry powder inhaler.

10. The composition according to either of Claims 4 or 5 further comprising a
 30 pharmaceutically effective amount of a therapeutically active agent selected from

anti-inflammatory agents, anticholinergic agents, β -agonists, P2Y2 receptor agonists, peroxisome proliferator-activated receptor agonists, kinase inhibitors, antiinfective agents and antihistamines.

- 5 11. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for administration to a human.
12. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for blocking sodium channels in a human.
- 10 13. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for promoting hydration of mucosal surfaces or restoring mucosal defense in a human.
- 15 14. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for treating a disease selected from the group of reversible or irreversible airway obstruction, chronic obstructive pulmonary disease (COPD), asthma, bronchiectasis (including bronchiectasis due to conditions other than cystic fibrosis), acute bronchitis, chronic bronchitis, post-viral cough, cystic fibrosis, emphysema, pneumonia,
- 20 panbronchiolitis, transplant-associate bronchiolitis, and ventilator-associated tracheobronchitis or preventing ventilator-associated pneumonia in a human in need thereof.
15. Use of a

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compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for treating chronic obstructive pulmonary disease (COPD) in a human in need thereof.

- 5 16. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for treating cystic fibrosis in a human in need thereof.

17. Use of a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for treating dry mouth (xerostomia), dry skin, vaginal dryness, sinusitis, rhinosinusitis, or nasal dehydration, including nasal dehydration brought on by administering dry oxygen, dry eye or Sjogren's disease, promoting ocular or corneal hydration, treating distal intestinal obstruction syndrome, treating otitis media, primary ciliary dyskinesia, distal intestinal obstruction syndrome, esophagitis, constipation, or chronic diverticulitis in a human in need thereof.

18. A compound according to any of claims 1-3, or a pharmaceutically acceptable salt thereof for use as a medicament.

19. A compound according to any of claims 1-3 or a pharmaceutically acceptable salt thereof for use in treating a disease associated with reversible or irreversible airway obstruction, chronic obstructive pulmonary disease (COPD), asthma, bronchiectasis (including bronchiectasis due to conditions other than cystic fibrosis), acute bronchitis, chronic bronchitis, post-viral cough, cystic fibrosis, emphysema, pneumonia, panbronchiolitis, transplant-associated bronchiolitis, and ventilator-associated tracheobronchitis or preventing ventilator-associated pneumonia in a human in need thereof.

20. A compound according to any of claims 1-3, or a pharmaceutically acceptable salt thereof, for use in treating dry mouth (xerostomia), dry skin, vaginal dryness, sinusitis, rhinosinusitis, or nasal dehydration, including nasal dehydration brought on by administering dry oxygen, dry eye or Sjogren's disease, promoting ocular or corneal hydration, treating distal intestinal obstruction syndrome, treating otitis media, primary ciliary dyskinesia, distal intestinal obstruction syndrome, esophagitis, constipation, or chronic diverticulitis in a human in need thereof.

21. A composition comprising a compound according to any one of claims 1-3, or a pharmaceutically acceptable salt thereof, for use in the preparation of a medicament for treating a disease associated with reversible or irreversible airway obstruction, chronic obstructive pulmonary disease (COPD), asthma, bronchiectasis (including bronchiectasis due to conditions other than cystic fibrosis), acute bronchitis, chronic bronchitis, post-viral cough, cystic fibrosis, emphysema, pneumonia, panbronchiolitis, transplant-associated bronchiolitis, and ventilator-associated tracheobronchitis or preventing ventilator-associated pneumonia.

22. A composition comprising a compound according to any one of claims 1-3, or a pharmaceutically acceptable salt thereof, for use in the preparation of a medicament for treating dry mouth (xerostomia), dry skin, vaginal dryness, sinusitis, rhinosinusitis, or nasal dehydration, including nasal dehydration brought on by administering dry oxygen, dry eye or Sjogren's disease, promoting ocular or corneal hydration, treating distal intestinal obstruction syndrome, treating otitis media, primary ciliary dyskinesia, distal intestinal obstruction syndrome, esophagitis, constipation, or chronic diverticulitis.

23. Use a compound according to any of Claims 1-3, or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for preventing, mitigating, and/or treating deterministic health effects to the respiratory tract and/or other bodily organs caused by respirable aerosols containing radionuclides in a human in need thereof.