CLAIMS

1. A pharmaceutical composition comprising

the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);

or any pharmaceutically acceptable salt thereof; wherein

OctG is (*S*)-2-aminodecanoic acid;

^{D}Pro is D-proline;

sodium chloride;

and optionally one or more pharmaceutically acceptable diluents, excipients or carriers;

for use in the prevention, management or treatment of diseases or conditions of the lungs being mediated by or resulting from human neutrophil elastase activity in a subject,

wherein the diseases or conditions of the lungs being mediated by or resulting from human neutrophil elastase activity are pulmonary diseases selected from alpha-1 antitrypsin deficiency (AATD), cystic fibrosis (CF), non-cystic fibrosis bronchiactasis (NCFB), or chronic obstructive pulmonary disease (COPD), or infections of the lungs causing diseases or conditions of the lungs, being mediated by human neutrophil elastase activity.

2. A pharmaceutical composition for use according to claim 1, wherein the pulmonary disease is non-cystic fibrosis bronchiactasis (NCFB) or cystic fibrosis (CF).

3. A pharmaceutical composition for use according to claim 1, wherein the pulmonary disease is cystic fibrosis (CF).

4. A pharmaceutical composition for use according to any one of claims 1-3, wherein the counter ion of the active compound is acetate.

5. A pharmaceutical composition for use according to any one of claims 1-4, wherein the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);

or any pharmaceutically acceptable salt thereof; is administered to the subject as pharmaceutical aerosol for pulmonary administration comprising a dispersed liquid phase and a continuous gas phase, wherein the dispersed liquid phase

(a) comprises aqueous droplets comprising the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);

or any pharmaceutically acceptable salt thereof;

(b) has a mass median diameter from 1.5 µm to 5 µm; and

(c) has a droplet size distribution having a geometrical standard deviation from 1.2 to 1.7.

6. A pharmaceutical composition for use according to claim 5, wherein the aerosol being emitted from an aerosol generator at a rate of at least 0.1 mL dispersed liquid phase per minute.

7. A pharmaceutical composition for use according to claim 5, wherein the aerosol being emitted from an aerosol generator at a mean delivery rate of at least 0.8 mg of the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; per minute.

8. A pharmaceutical composition for use according to any one of claims 1 to 7, wherein the pharmaceutical composition is a liquid pharmaceutical composition for preparing an aerosol comprising the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; in a concentration within a range from 4 mg/mL to 100 mg/mL.

9. A pharmaceutical composition for use according to any one of claims 1 to 8, wherein the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);

or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose between 0.1 and 10000 mg/day.

10. A pharmaceutical composition for use according to any one of claims 1 to 8, wherein the active compound
cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);
or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose between 0.001 and 100 mg/kg.

11. A pharmaceutical composition for use according to any one of claims 1 to 8, wherein the active compound
cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);
or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose between 5 and 1000 mg/day.

12. A pharmaceutical composition for use according to any one of claims 1 to 8, wherein the active compound
cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);
or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose of 20, 60, 120, 240, 480 or 960 mg/day.

13. A pharmaceutical composition for use according to any one of claims 1 to 8, wherein the active compound
cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);
or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose between 80 and 320 mg/day.

14. A pharmaceutical composition for use according to any one of claims 1 to 8, wherein the active compound
cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);

or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose of 80, 160, or 320 mg/day.

15. A pharmaceutical composition for use according to any one of claims 1 to 14, wherein the active compound
cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);
or any pharmaceutically acceptable salt thereof is administered to the subject by oral inhalation.

16. A pharmaceutical aerosol for pulmonary administration comprising a dispersed liquid phase and a continuous gas phase, wherein the dispersed liquid phase
(a) comprises aqueous droplets comprising the active compound
cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);
or any pharmaceutically acceptable salt thereof; wherein
OctG is (*S*)-2-aminodecanoic acid;
^{D}Pro is D-proline;
(b) has a mass median diameter from 1.5 µm to 5 µm; and
(c) has a droplet size distribution having a geometrical standard deviation from 1.2 to 1.7, as measured as described in the description;
for use in the prevention, management or treatment of diseases or conditions of the lungs being mediated by or resulting from human neutrophil elastase activity in a subject, wherein the diseases or conditions of the lungs being mediated by or resulting from human neutrophil elastase activity are pulmonary diseases selected from alpha-1 antitrypsin deficiency (AATD), cystic fibrosis (CF), non-cystic fibrosis bronchiactasis (NCFB), or chronic obstructive pulmonary disease (COPD), or infections of the lungs causing diseases or conditions of the lungs, being mediated by human neutrophil elastase activity.

17. An aerosol for use according to claim 16, wherein the pulmonary disease is non-cystic fibrosis bronchiactasis (NCFB) or cystic fibrosis (CF).

18. An aerosol for use according to claim 16, wherein the pulmonary disease is cystic fibrosis (CF).

19. An aerosol for use according to any one of claims 16 to 18, wherein the counter ion of the active compound is acetate.

20. An aerosol for use according to any one of claims 16 to 19, wherein the aerosol being emitted from an aerosol generator at a rate of at least 0.1 mL dispersed liquid phase per minute.

21. An aerosol for use according to any one of claims 16to 19, wherein the aerosol being emitted from an aerosol generator at a mean delivery rate of at least 0.8 mg of the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; per minute.

22. An aerosol for use according to any one of claims 16 to 21, wherein the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose between 0.1 and 10000 mg/day.

23. An aerosol for use according to any one of claims 16 to 21, wherein the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose between 0.001 and 100 mg/kg.

24. An aerosol for use according to any one of claims 16 to 21, wherein the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);

or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose between 5 and 1000 mg/day.

25. An aerosol for use according to any one of claims 16 to 21, wherein the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose of 20, 60, 120, 240, 480 or 960 mg/day.

26. An aerosol for use according to any one of claims 16 to 21, wherein the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose between 80 and 320 mg/day.

27. An aerosol for use according to any one of claims 16 to 21, wherein the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; is administered to the subject at a dose of 80, 160, or 320 mg/day.

28. An aerosol for use according to any one of claims 16 to 27, wherein the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; is administered to the subject by oral inhalation.

29. A kit comprising

a pharmaceutical composition comprising

the active compound

cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-);

or any pharmaceutically acceptable salt thereof; wherein

OctG is (*S*)-2-aminodecanoic acid;

^{D}Pro is D-proline; and

sodium chloride

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30. A kit according to claim 29, wherein the kit comprises the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; as aerosol for pulmonary administration comprising a dispersed liquid phase and a continuous gas phase, wherein the dispersed liquid phase

(a) comprises aqueous droplets comprising the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof;

(b) has a mass median diameter from 1.5 µm to 5 µm; and

(c) has a droplet size distribution having a geometrical standard deviation from 1.2 to 1.7.

31. A kit according to any one of claims 29 to 30, wherein at least 70 wt.-% of the loaded dose of the active compound cyclo(-OctG-Glu-Thr-Ala-Ser-Ile-Pro-Pro-Gln-Lys-Tyr-^{D}Pro-Pro-); or any pharmaceutically acceptable salt thereof; is comprised of droplets having a mass median diameter of not more than 5 µm.

32. A kit according to any one of claims 29 to 31, wherein the counter ion of the active compound is acetate.