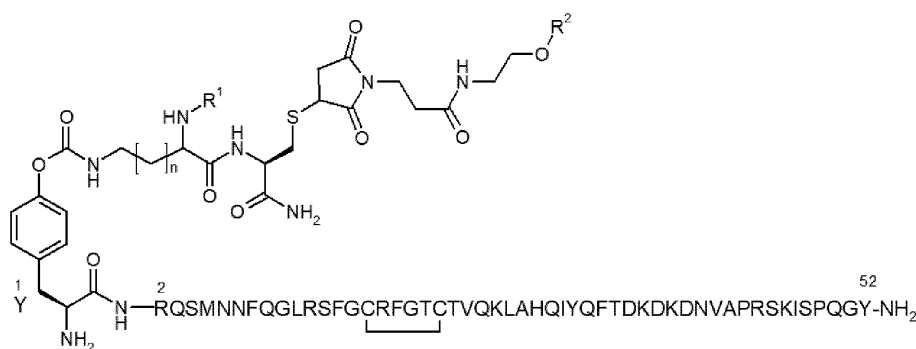


Claims

1. A pharmaceutical formulation comprising:

- PEG-ADM, wherein the PEG-ADM is a compound according to the general formula (I),



(I)

in which

n represents the number 0, 1, 2 or 3,

R¹ represents hydrogen, methyl, ethyl, n-propyl or isopropyl,

R² represents linear or branched PEG 20kDa to 80kDa endcapped with a methoxy-group,

or a hydrate thereof, solvate thereof, salt thereof, pharmaceutically acceptable salt thereof, or the solvates of salts thereof;

- a pH regulator; and

- trehalose or a hydrate thereof, solvate thereof, salt thereof, pharmaceutically acceptable salt thereof, or the solvates of salts thereof.

2. The pharmaceutical formulation according to claim 1, wherein the pharmaceutical formulation comprises 60 wt.-% to 98 wt.-% trehalose, wherein the concentrations of components are based on the total weight of the pharmaceutical formulation.

3. The pharmaceutical formulation according to claim 1 or 2, wherein the pharmaceutical formulation comprises

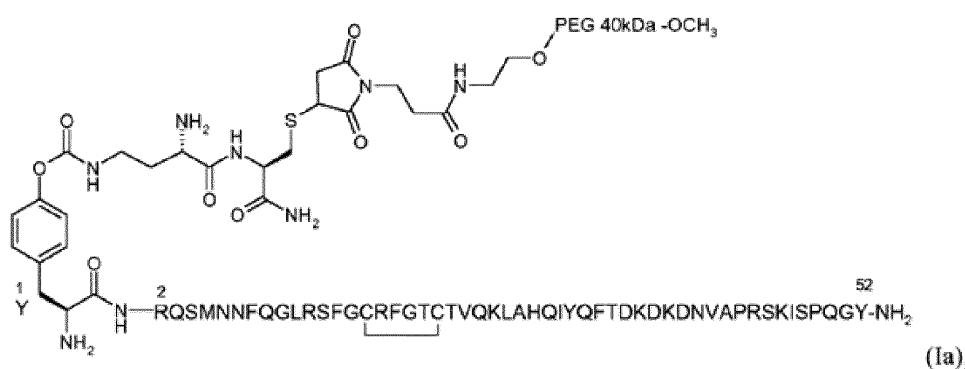
- 1 wt.-% to 15 wt.-% of PEG-ADM or a hydrate thereof, solvate thereof, salt thereof, pharmaceutically acceptable salt thereof, or the solvates of salts thereof;

- 0.01 wt.-% to 25 wt.-% of the pH regulator; and

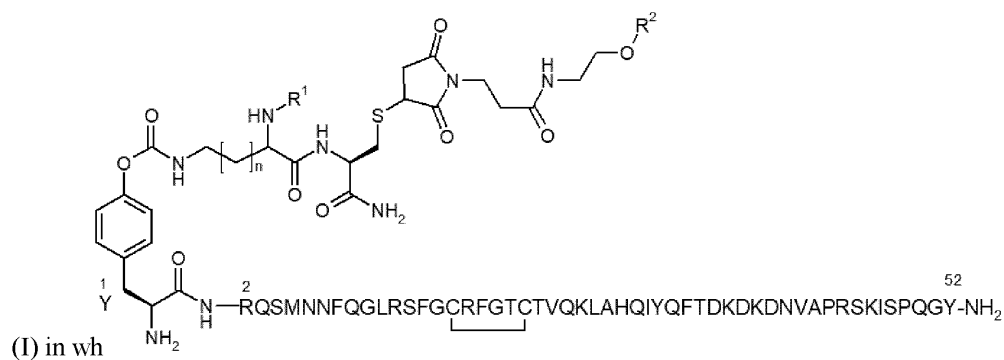
- 60 wt.-% to 98 wt.-% trehalose or a hydrate thereof, solvate thereof, salt thereof, pharmaceutically acceptable salt thereof, or the solvates of salts thereof;

wherein the concentrations of components are based on the total weight of the pharmaceutical formulation.

- 5 4. The pharmaceutical formulation according to any one of the preceding claims 1 to 3, wherein the PEG-ADM is the compound according to formula (Ia)



5. The pharmaceutical formulation according to any one of the preceding claims 1 to 4, wherein the pharmaceutical formulation is a lyophilizate.
- 10 6. A liquid pharmaceutical formulation comprising:
 - a. 0.04 mg/mL to 145 mg/mL of PEG-ADM, wherein the PEG-ADM is a compound according to the general formula (I),



- 15 in which

n represents the number 0, 1, 2 or 3,

R¹ represents hydrogen, methyl, ethyl, n-propyl or isopropyl,

R² represents linear or branched PEG 20kDa to 80kDa endcapped with a methoxy-group,
or a hydrate thereof, solvate thereof, salt thereof, pharmaceutically acceptable salt thereof,
or the solvates of salts thereof;

b. a solvent;

5 c. a pH regulator;

d. an osmolarity regulator; and

e. trehalose;

wherein the presence of the osmolarity regulator (component d) is optional;

wherein the pharmaceutical formulation has a pH between 3 and 5; and

10 wherein the concentrations of components are based on the total volume of the liquid pharmaceutical formulation.

7. The pharmaceutical formulation according to claim 6, wherein the liquid pharmaceutical formulation comprises

15 - 0.385 mg/mL to 77 mg/mL PEG-ADM, wherein the PEG-ADM is a compound according to the general formula (I) or formula (Ia), a hydrate thereof, solvate thereof, salt thereof, pharmaceutically acceptable salt thereof, or the solvates of salts thereof; and

- 10 mg/mL to 100 mg/mL trehalose.

8. The pharmaceutical formulation according to claim 6 or 7, wherein the liquid pharmaceutical formulation comprises the lyophilizate according to claim 5 and a solvent.

20 9. Pharmaceutical formulation according to any of the preceding claims 1 to 8 for inhalation.

10. Medicament comprising the pharmaceutical formulation according to any one of claims 1 to 9 or a medicament comprising the pharmaceutical formulation according to any one of claims 1 to 9 in combination with an inert nontoxic pharmaceutically suitable excipient, optionally in combination with a further active ingredient.

25 11. Combined pharmaceutical dose form comprising components (1) and (2), wherein

- component (1) comprises a pharmaceutical formulation according to any one of the preceding claims 1 to 9; and

- component (2) comprises a solvent.
12. Combination pack comprising component (1) and (2), wherein
- component (1) comprises the pharmaceutical formulation, the medicament, or the combined pharmaceutical dose form according to any one of claims 1 to 11; and
- 5 component (2) comprises a nebulizer, preferably a mesh nebulizer.
13. The pharmaceutical formulation according to any one of claims 1 to 9, the medicament according to claim 10, the combined pharmaceutical dose form according to claim 11, or the combination pack according to claim 12 for use in the treatment and/or prevention of diseases.
14. The pharmaceutical formulation, the combined pharmaceutical dose form, or the combination pack
- 10 according to claim 13 for use in the treatment and/or prevention of diseases, wherein the disease is selected from
- pulmonary disorders, such as pulmonary hypertension; secondary pulmonary hypertension; pulmonary hypertension following pulmonary embolism with and without acute cor pulmonale; primary pulmonary hypertension; chronic obstructive pulmonary disease; asthma; acute
 - 15 pulmonary edema; chronic pulmonary edema; allergic alveolitis; pneumonitis due to inhaled organic dust; pneumonitis due to inhaled particles of fungal, actinomycetic or other origin; acute chemical bronchitis; acute chemical pulmonary edema and/or chronic chemical pulmonary edema (e.g. after inhalation of phosgene, nitrogen oxide); neurogenic pulmonary edema; acute pulmonary manifestations due to radiation; chronic pulmonary manifestations due to radiation;
 - 20 acute and/or chronic interstitial lung disorders (such as but not restricted to drug-induced interstitial lung disorders, e.g. secondary to Bleomycin treatment); acute lung injury (ALI); acute lung injury (ALI) in adult or child including newborn; acute respiratory distress syndrome (ARDS); acute respiratory distress syndrome (ARDS) in adult or child including newborn; ALI/ARDS secondary to pneumonia and sepsis, aspiration pneumonia and ALI/ARDS secondary
 - 25 to aspiration (such as but not restricted to aspiration pneumonia due to regurgitated gastric content); ALI/ARDS secondary to smoke gas inhalation; transfusion-related acute lung injury (TRALI), ALI/ARDS or acute pulmonary insufficiency following surgery; trauma or burns, ventilator induced lung injury (VILI); lung injury following meconium aspiration; pulmonary fibrosis; and mountain sickness;
- 30 - ALI/ARDS secondary to pneumonia caused by bacterial infection of the lungs, such as, but not restricted to, bacterial pneumonia caused by Pneumococci, Haemophilus Influenzae, Mycoplasma Pneumoniae, Chlamydia species, Enterococci, beta-hemolytic Streptococci, Staphylococci, Gram-negative Enterobacteriaceae, Pseudomonas species, Klebsiella species, Acinetobacter species,

Legionella species, and Mycobacteria;

- 5 - ALI/ARDS secondary to pneumonia caused by viral infections such as, but not restricted to, Influenza viruses (e.g. caused by strains of serotypes H1N1, H5N1, H7N9), Corona viruses (e.g. SARS-CoV, the pathogen of severe acute respiratory syndrome (SARS), MERS-CoV, the pathogen of Middle East respiratory syndrome (MERS), and SARS-CoV-2 the pathogen of COVID-19 pandemic), Respiratory-Syncytial-Virus (RSV), and Cytomegalovirus (CMV);
- ALI/ARDS secondary to pneumonia caused by fungal infections such as, but not restricted to, fungal pneumonia caused by Pneumocystis Jirovecii;
- 10 - ALI/ARDS secondary to pneumonia irrespective of the context of pneumonia origin such as for community acquired pneumonia (CAP) as well as for hospital acquired pneumonia (HAP), in particular for HAP acquired in the context of artificial ventilation (VAP);
- ALI/ARDS secondary to pneumonia irrespective of the diverse pathoanatomical appearances of pneumonias such as, but not restricted to, lobar (i.e. affecting an entire lung lobe), lobular (i.e. affecting smaller lung lobules), interstitial (i.e. diffuse affection of the lung tissue);
- 15 - ALI/ARDS secondary to pneumonia occurring in consequence of bacterial and/or virus infection;
- ALI/ARDS secondary to pneumonia occurring in consequence of a bacterial superinfection of a primary lung affection by viruses; and
- prevention and/or treatment of lung dysfunction after lung transplantations.
- 15. Lyophilizate according to claim 5 obtainable by freeze-drying of the liquid pharmaceutical
20 formulation as defined in any one of claims 6 to 8.
- 16. Liquid Pharmaceutical formulation according to any one of claims 6 to 8 obtainable by mixing the lyophilizate according to claim 5 with a solvent.
- 17. A method for the preparation of the pharmaceutical formulation according to claim 5, comprising the following steps:
- 25 step 1. Providing at least components a, c and e; and
- step 2. Mixing the components provided in step 1;
- step 3: freeze-drying the pharmaceutical formulation obtained after any one of steps 1 and/or 2
- whereby the following pharmaceutical formulation according to claim 5 is obtained.