Claims

1. Alpha-1-microglobulin for use in the treatment of a mitochondria disease or for use in repairing, restoring or maintaining mitochondrial function.

2. Alpha-1-microglobulin for use according to any one of the preceding claims, wherein the mitochondria-related disease or disorder is one or more of the diseases or disorders from the following list or a selection of diseases or disorder from the following list:

- Alpers Disease (Progressive Infantile Poliodystrophy)
- Barth syndrome (Lethal Infantile Cardiomyopathy)
- Beta-oxidation Defects
- Cardiomyopathy
- Carnitine-Acyl-Carnitine Deficiency
- Carnitine Deficiency
- Creatine Deficiency Syndromes (Cerebral Creatine Deficiency Syndromes (CCDS) includes: Guanidinoaceteate Methyltransferase Deficiency (GAMT Deficiency), L-Arginine:Glycine Amidinotransferase Deficiency (AGAT Deficiency), and SLC6A8-Related Creatine Transporter Deficiency (SLC6A8 Deficiency).
- Co-Enzyme Q10 Deficiency
- Complex I Deficiency (NADH dehydrogenase (NADH-CoQ reductase) deficiency)
- Complex II Deficiency (Succinate dehydrogenase deficiency)
- Complex III Deficiency (Ubiquinone-cytochrome c oxidoreductase deficiency)
- Complex IV Deficiency/COX Deficiency (Cytochrome c oxidase deficiency is caused by a defect in Complex IV of the respiratory chain)
- Complex V Deficiency (ATP synthase deficiency)
- COX Deficiency
- CPEO (Chronic Progressive External Ophthalmoplegia Syndrome)
- CPT I Deficiency
- CPT II Deficiency
- Friedreich's ataxia (FRDA or FA)
- Encephalomyopathy
- Glutaric Aciduria Type II
- KSS (Kearns-Sayre Syndrome)
- Lactic Acidosis

- LCAD (Long-Chain Acyl-CoA Dehydrongenase Deficiency)
- LCHAD
- Leigh Disease or Syndrome (Subacute Necrotizing Encephalomyelopathy)
- LHON (Leber's hereditary optic neuropathy)
- Luft Disease
- MCAD (Medium-Chain Acyl-CoA Dehydrongenase Deficiency)
- MELAS (Mitochondrial Encephalomyopathy Lactic Acidosis and Strokelike Episodes)
- MERRF (Myoclonic Epilepsy and Ragged-Red Fiber Disease)
- MIRAS (Mitochondrial Recessive Ataxia Syndrome)
- Mitochondrial Cytopathy
- Mitochondrial DNA Depletion
- Mitochondrial Encephalopathy includes: Encephalomyopathy, Encephalomyelopathy
- Mitochondrial Myopathy
- MNGIE (Myoneurogastointestinal Disorder and Encephalopathy)
- NARP (Neuropathy, Ataxia, and Retinitis Pigmentosa)
- Pearson Syndrome
- Pyruvate Carboxylase Deficiency
- Pyruvate Dehydrogenase Deficiency
- POLG Mutations
- Respiratory Chain Deficiencies
- SCAD (Short-Chain Acyl-CoA Dehydrogenase Deficiency)
- SCHAD
- VLCAD (Very Long-Chain Acyl-CoA Dehydrongenase Deficiency)

3. Alpha-1-microglobulin for use according to any one of the preceding claims in the treatment or prophylaxis of respiratory chain disorders.

4. Alpha-1-microglobulin for use according to claim 3, wherein the respiratory chain disorders involve Complex I, II, III, IV or V defects.

5. Alpha-1-microglobulin for use according to any one of the preceding claim in the treatment or prophylaxis of mitochondrial dysfunctions in children or young adults.

6. Alpha-1-microglobulin for use according to any one of the preceding claim for the treatment of prevention of Alpers disease, Barth syndrome, Fridreich's ataxia, KSS, Leigh Disease or Syndrome, LHON, MELAS, MERRF, MIRAS and NARP

7. Alpha-1-microglobulin for use according to any one of the preceding claim for the treatment or prophylaxis of mitochondrial dysfunctions in women.

8. Alpha-1-microglobulin for use according to any one of the preceding claims in the treatment or prophylaxis of damage or dysfunction of retina or ocular diseases associated with mitochondrial defect(s) or dysfunction(s).

9. A method for treating a subject suffering from one or more of: a mitochondrial defect, a mitochondria disease or disorder, a drug-induced mitochondria side-effect or an environmentally induced mitochondria effect, the method comprising administering alpha-1-microglobulin to the subject

10 A method for preventing one or more of: a mitochondrial defect, a mitochondria disease or disorder, a drug-induced mitochondria side-effect or an environmentally induced mitochondria effect, the method comprising administering alpha-1-microglobulin to the subject.

11. A method according to claim 9 or 10, wherein the subject is a mammal including a human.

12. A method according to any one of claim 9-11, wherein the mitochondria disease or disorder is as described in any one of claims 2-8.

Claims

1. Use of alpha-1-microglobulin in the manufacture of a medicament for the treatment of a mitochondria disease or for repairing, restoring or maintaining mitochondrial function.

2. The use according to any one of the preceding claims, wherein the mitochondria-related disease or disorder is one or more of the diseases or disorders from the following list or a selection of diseases or disorder from the following list:

- Alpers Disease (Progressive Infantile Poliodystrophy)
- Barth syndrome (Lethal Infantile Cardiomyopathy)
- Beta-oxidation Defects
- Cardiomyopathy
- Carnitine-Acyl-Carnitine Deficiency
- Carnitine Deficiency
- Creatine Deficiency Syndromes (Cerebral Creatine Deficiency Syndromes (CCDS) includes: Guanidinoaceteate Methyltransferase Deficiency (GAMT Deficiency), L-Arginine:Glycine Amidinotransferase Deficiency (AGAT Deficiency), and SLC6A8-Related Creatine Transporter Deficiency (SLC6A8 Deficiency).
- Co-Enzyme Q10 Deficiency
- Complex I Deficiency (NADH dehydrogenase (NADH-CoQ reductase) deficiency)
- Complex II Deficiency (Succinate dehydrogenase deficiency)
- Complex III Deficiency (Ubiquinone-cytochrome c oxidoreductase deficiency)
- Complex IV Deficiency/COX Deficiency (Cytochrome c oxidase deficiency is caused by a defect in Complex IV of the respiratory chain)
- Complex V Deficiency (ATP synthase deficiency)
- COX Deficiency
- CPEO (Chronic Progressive External Ophthalmoplegia Syndrome)
- CPT I Deficiency
- CPT II Deficiency
- Friedreich's ataxia (FRDA or FA)
- Encephalomyopathy
- Glutaric Aciduria Type II
- KSS (Kearns-Sayre Syndrome)

- Lactic Acidosis
- LCAD (Long-Chain Acyl-CoA Dehydrongenase Deficiency)
- LCHAD
- Leigh Disease or Syndrome (Subacute Necrotizing Encephalomyelopathy)
- LHON (Leber's hereditary optic neuropathy)
- Luft Disease
- MCAD (Medium-Chain Acyl-CoA Dehydrongenase Deficiency)
- MELAS (Mitochondrial Encephalomyopathy Lactic Acidosis and Strokelike Episodes)
- MERRF (Myoclonic Epilepsy and Ragged-Red Fiber Disease)
- MIRAS (Mitochondrial Recessive Ataxia Syndrome)
- Mitochondrial Cytopathy
- Mitochondrial DNA Depletion
- Mitochondrial Encephalopathy includes: Encephalomyopathy, Encephalomyelopathy
- Mitochondrial Myopathy
- MNGIE (Myoneurogastointestinal Disorder and Encephalopathy)
- NARP (Neuropathy, Ataxia, and Retinitis Pigmentosa)
- Pearson Syndrome
- Pyruvate Carboxylase Deficiency
- Pyruvate Dehydrogenase Deficiency
- POLG Mutations
- Respiratory Chain Deficiencies
- SCAD (Short-Chain Acyl-CoA Dehydrogenase Deficiency)
- SCHAD
- VLCAD (Very Long-Chain Acyl-CoA Dehydrongenase Deficiency)

3. The use according to any one of the preceding claims for the treatment or prophylaxis of respiratory chain disorders.

4. The use according to claim 3, wherein the respiratory chain disorders involve Complex I, II, III, IV or V defects.

5. The use according to any one of the preceding claim for the treatment or prophylaxis of mitochondrial dysfunctions in children or young adults.

6. The use according to any one of the preceding claim for the treatment of prevention of Alpers disease, Barth syndrome, Fridreich's ataxia, KSS, Leigh Disease or Syndrome, LHON, MELAS, MERRF, MIRAS and NARP

7. The use according to any one of the preceding claim for the treatment or prophylaxis of mitochondrial dysfunctions in women.

8. The use according to any one of the preceding claims for the treatment or prophylaxis of damage or dysfunction of retina or ocular diseases associated with mitochondrial defect(s) or dysfunction(s).

9. Use of alpha-1-microglobulin in the manufacture of a medicament for the treatment of a mitochondria disease or disorder in a subject suffering from one or more of: a mitochondrial defect, a drug-induced mitochondria side-effect or an environmentally induced mitochondria effect.

10. Use of alpha-1-microglobulin in the manufacture of a medicament for the prevention of one or more of: a mitochondrial defect, a mitochondria disease or disorder, a drug-induced mitochondria side-effect or an environmentally induced mitochondria effect in a subject.

11. The use according to claim 9 or 10, wherein the subject is a mammal including a human.

12. The use according to any one of claim 9-11, wherein the mitochondria disease or disorder is as described in any one of claims 2-8.