Claims

1. Use of alpha-1-microglobulin in the manufacture of a medicament for the treatment of a mitochondria disease.

~~2. The use according to claim 1 for the treatment of the mitochondria disease by repairing or restoring mitochondrial function.~~

~~3~~2. The use according to claim 1~~any one of the preceding claims~~, wherein the mitochondria~~-related~~ disease is selected ~~one or more of the diseases from the following list or a selection of diseases~~ 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 (Cytochrome c oxidase deficiency)
- CPEO (Chronic Progressive External Ophthalmoplegia Syndrome)
- CPT I Deficiency (Carnitine Palmitoyltransferase I Deficiency)
- CPT II Deficiency (Carnitine Palmitoyltransferase 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 (Long-Chain 3-hydroxyacyl-CoA Dehydrogenase Deficiency)
- 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 (DNA polymerase subunit gamma mutations)
- Respiratory Chain Deficiencies
- SCAD (Short-Chain Acyl-CoA Dehydrogenase Deficiency)
- SCHAD (Short Chain Hydroxyl Acyl-CoA Dehydrogenase Deficiency)
- VLCAD (Very Long-Chain Acyl-CoA Dehydrongenase Deficiency)

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

~~54~~. The use according to claim 43, wherein the respiratory chain deficiencies involve Complex I, II, III, IV or V defects.

~~6~~5. The use according to any one of the preceding claim for the treatment or prophylaxis of mitochondrial dysfunctions in children or young adults.

~~7~~6. The use according to any one of the preceding claim for the treatment or prevention of Alpers disease, Barth syndrome, Fridreich's ataxia, KSS, Leigh Disease or Syndrome, LHON, MELAS, MERRF, MIRAS and NARP

~~8~~7. The use according to any one of the preceding claim for the treatment or prophylaxis of mitochondrial dysfunctions in women.

~~9~~8. The use according to any one of the preceding claims, wherein the mitochondria disease involves damage or dysfunction of retina.

~~10~~9. Use of alpha-1-microglobulin in the manufacture of a medicament for the treatment of a mitochondria disease in a subject suffering from one or more of: a mitochondrial defect, or a drug-induced mitochondria side-effect.

~~11~~10. Use of alpha-1-microglobulin in the manufacture of a medicament for the prevention of a mitochondria disease, wherein the mitochondrial disease involves a mitochondrial defect, a drug-induced mitochondria side-effect or an environmentally induced mitochondria effect in a subject.

~~12~~11. The use according to claim ~~10~~ 9 ~~or11~~10, wherein the subject is a mammal including a human.

~~13~~12. The use according to any one of claim ~~10~~9-~~12~~11, wherein the mitochondria disease is as described in any one of claims ~~3~~2-~~9~~8.