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 claim 1 for the treatment of the mitochondria disease by repairing or restoring mitochondrial function.

~~2~~3. 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 (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)

~~34~~34. The use according to any one of the preceding claims for the treatment or prophylaxis of respiratory chain ~~disorders~~deficiencies.

~~34~~45. The use according to claim ~~34~~, wherein the respiratory chain ~~disorders~~ deficiencies involve Complex I, II, III, IV or V defects.

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

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

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

~~8~~9. The use according to any one of the preceding claims, wherein the mitochondria disease involves ~~for the treatment or prophylaxis of~~ damage or dysfunction of retina ~~or ocular diseases associated with mitochondrial defect(s) or dysfunction(s)~~.

~~9~~10. 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, or a drug-induced mitochondria side-effect ~~or an environmentally induced mitochondria effect~~.

~~10~~11. 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~~, wherein the mitochondrial disease involves a mitochondrial defect, a drug-induced mitochondria side-effect or an environmentally induced mitochondria effect in a subject.

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

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