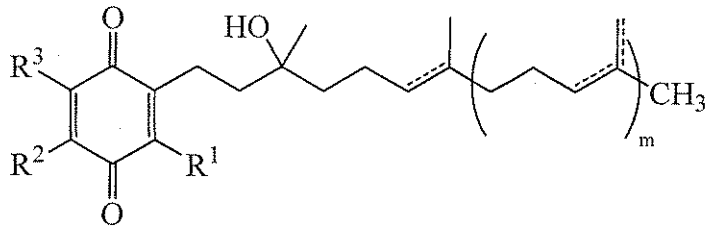
CLAIMS

What is claimed is:

1. A formulation for use in preventing, reducing, ameliorating or treating an ophthalmic disorder, or for stopping the progression of, or reversing, the loss of vision in a patient, wherein the formulation comprises an ophthalmically effective amount of one or more compounds selected from:

quinones of Formula I:



Formula I

and the hydroquinone forms thereof,

wherein:

the bonds indicated by a dashed line are independently of each other, at each occurrence, double or single with the proviso that at least one bond is a double bond, and with the proviso that they are not both double within the same unit;

R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6) alkoxy; and

m is an integer from 0-1 ;

or any stereoisomer, or mixture of stereoisomers thereof; and additionally comprises a pharmaceutically or ophthalmically acceptable vehicle;

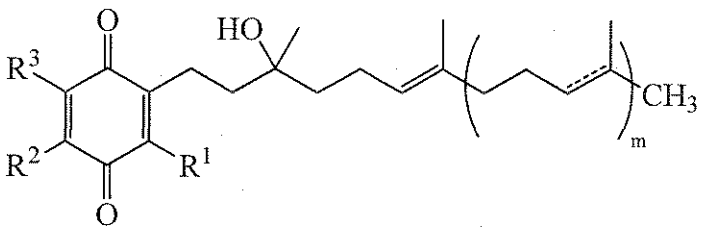
wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of Leber's Hereditary Optic Neuropathy (LHON); Dominant Optic Atrophy (DOA); Chronic Progressive External Ophthalmoplegia (CPEO); Spinocerebellar ataxia (SCA); Leigh's Syndrome; Friedreich's ataxia (FRDA); Mitochondrial Myopathy, Encephalopathy, Lactacidosis, and Stroke (MELAS); Myoclonic Epilepsy with Ragged Red Fibers (MERRF); Kearns-Sayre Syndrome (KSS); an overlap syndrome; Co-Enzyme Q10 (CoQ10) Deficiency; Complex I deficiency; Complex II deficiency; Complex III deficiency; Complex IV deficiency; Complex V deficiency; a neurodegenerative disease; Parkinson's disease; Alzheimer's disease; Amyotrophic Lateral Sclerosis (ALS); a motor neuron disease; Huntington's Disease; an age-associated disease; glaucoma; a disorder of the outer retina; macular degeneration; age related macular degeneration; juvenile

macular degeneration; diabetic retinopathy; Progressive Supranuclear Palsy (PSP); a Parkinson-like disease; Chacot-Marie-Tooth Disease; a Mucopolysaccharidosis; Adrenoleukodystrophy; Niemann-Pick disease; Krabbe's disease; Pelizaeus-Merzbacher disease; and Progressive Encephalopathy, Edema, Hypsarrhythmia and Optic Atrophy (PEHO); or

wherein the ophthalmic disorder is associated with a trauma selected from the group consisting of retinal ischemia, an acute retinopathy associated with trauma, a post-surgical complication, damage associated with laser therapy including photodynamic therapy (PDT), traumatic optic neuropathy (TON), damage associated with surgical light induced iatrogenic retinopathy, damage associated with a corneal transplant, and damage associated with stem cell transplant of eye cells.

2. The formulation of Claim 1, for use as defined in said claim, wherein the formulation comprises an ophthalmically effective amount of one or more compounds selected from:

quinones of Formula I-a:



Formula I-a

and the hydroquinone forms thereof,

wherein:

the bond indicated by a dashed line is double or single;

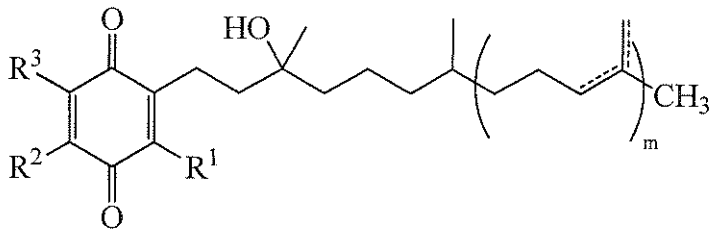
R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6)alkoxy; and

m is 0-1;

or any stereoisomer, or mixture of stereoisomers thereof.

3. The formulation of Claim 1, for use as defined in said claim, wherein the formulation comprises an ophthalmically effective amount of one or more compounds selected from:

quinones of Formula I-c:



Formula I-c

and the hydroquinone forms thereof,

wherein:

the bonds indicated by a dashed line are double or single, with the proviso that they are not both double within the same unit; and the further proviso that at least one bond is a double bond;

R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6) alkoxy; and m is 1;

or any stereoisomer, or mixture of stereoisomers thereof.

4. The formulation according to Claim 1, for use as defined in said claim, wherein the one or more compounds are selected from the group consisting of:

2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodec-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodec-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

and the hydroquinone forms thereof;

or any stereoisomer, or mixture of stereoisomers thereof.

5. The formulation according to Claim 1, for use as defined in said claim, wherein the one or more compounds are selected from the group consisting of:

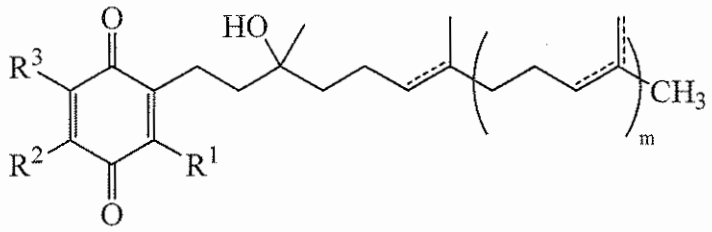
2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione; and

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

or any stereoisomer, or mixture of stereoisomers thereof.

6. The formulation according to any one of Claims 1-4, for use as defined in said claim, wherein the one or more compounds are quinones.

7. Use of a compound of Formula I



Formula I

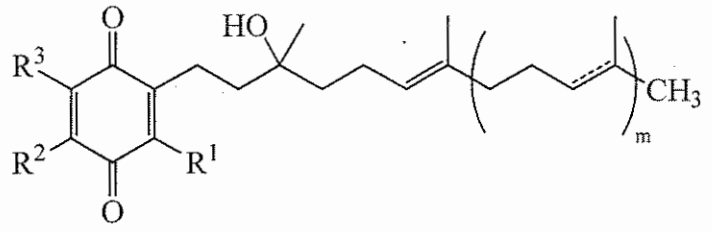
or the hydroquinone form thereof,

wherein:

the bonds indicated by a dashed line are independently of each other, at each occurrence, double or single with the proviso that at least one bond is a double bond, and with the proviso that they are not both double within the same unit;

R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6) alkoxy; and

m is an integer from 0-1; or any stereoisomer, or mixture of stereoisomers thereof; or a compound of Formula I-a



Formula I-a

or the hydroquinone form thereof,

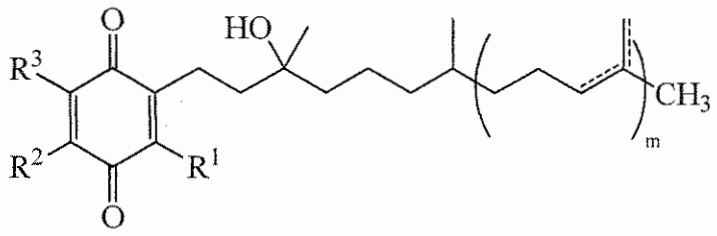
wherein:

the bond indicated by a dashed line is double or single;

R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6)alkoxy; and

m is 0-1; or any stereoisomer, or mixture of stereoisomers thereof; or

a compound of Formula I-c



Formula I-c

or the hydroquinone form thereof,

wherein:

the bonds indicated by a dashed line are double or single, with the proviso that they are not both double within the same unit; and the further proviso that at least one bond is a double bond;

R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6) alkoxy; and m is 1; or any stereoisomer, or mixture of stereoisomers thereof; or

a compound selected from the group consisting of:

2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodec-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodec-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione; and

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

or the hydroquinone form thereof, or any stereoisomer, or mixture of stereoisomers thereof;

in the manufacture of a medicament for preventing, reducing, ameliorating or treating an ophthalmic disorder, or for stopping the progression of, or reversing, the loss of vision in a patient; wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of Leber's Hereditary Optic Neuropathy (LHON); Dominant Optic

Atrophy (DOA); Chronic Progressive External Ophthalmoplegia (CPEO); Spinocerebellar ataxia (SCA); Leigh's Syndrome; Friedreich's ataxia (FRDA); Mitochondrial Myopathy, Encephalopathy, Lactacidosis, and Stroke (MELAS); Myoclonic Epilepsy with Ragged Red Fibers (MERRF); Kearns-Sayre Syndrome (KSS); an overlap syndrome; Co-Enzyme Q10 (CoQ10) Deficiency; Complex I deficiency; Complex II deficiency; Complex III deficiency; Complex IV deficiency; Complex V deficiency; a neurodegenerative disease; Parkinson's disease; Alzheimer's disease; Amyotrophic Lateral Sclerosis (ALS); a motor neuron disease; Huntington's Disease; an age-associated disease; glaucoma; a disorder of the outer retina; macular degeneration; age related macular degeneration; juvenile macular degeneration; diabetic retinopathy; Progressive Supranuclear Palsy (PSP); a Parkinson-like disease; Chacot-Marie-Tooth Disease; a Mucopolysaccharidosis; Adrenoleukodystrophy; Niemann-Pick disease; Krabbe's disease; Pelizaeus-Merzbacher disease; and Progressive Encephalopathy, Edema, Hypsarrhythmia and Optic Atrophy (PEHO); or
wherein the ophthalmic disorder is associated with a trauma selected from the group consisting of retinal ischemia, an acute retinopathy associated with trauma, a post-surgical complication, damage associated with laser therapy including photodynamic therapy (PDT), traumatic optic neuropathy (TON), damage associated with surgical light induced iatrogenic retinopathy, damage associated with a corneal transplant, and damage associated with stem cell transplant of eye cells.

8. The formulation of any one of Claims 1-6, for use as defined in said claim, wherein the formulation is administered orally.

9. The formulation of any one of Claims 1-6, for use as defined in said claim, wherein the formulation is administered topically.

10. The formulation of any one of Claims 1-6, for use as defined in said claim, wherein the formulation is administered topically in eye drops or an irrigating solution, periocularly, or intraocularly.

11. The formulation of any one of Claims 1-6 and 8-10, for use as defined in said claim, wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of Leber's Hereditary Optic Neuropathy (LHON); Dominant Optic Atrophy

(DOA); Chronic Progressive External Ophthalmoplegia (CPEO); Spinocerebellar ataxia (SCA); Leigh's Syndrome; Friedreich's ataxia (FRDA); Mitochondrial Myopathy, Encephalopathy, Lactacidosis, and Stroke (MELAS); Myoclonic Epilepsy with Ragged Red Fibers (MERRF); Kearns-Sayre Syndrome (KSS); an overlap syndrome; Co-Enzyme Q10 (CoQ10) Deficiency; Complex I deficiency; Complex II deficiency; Complex III deficiency; Complex IV deficiency; and Complex V deficiency.

12. The formulation of any one of Claims 1-6 and 8-10, for use as defined in said claim, wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of Leber's Hereditary Optic Neuropathy (LHON); Dominant Optic Atrophy (DOA); and Chronic Progressive External Ophthalmoplegia (CPEO).

13. The formulation of any one of Claims 1-6 and 8-10, for use as defined in said claim, wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of Friedreich's ataxia (FRDA); Mitochondrial Myopathy, Encephalopathy, Lactacidosis, and Stroke (MELAS); Myoclonic Epilepsy with Ragged Red Fibers (MERRF); Leigh's syndrome; Kearns-Sayre Syndrome (KSS); and an overlap syndrome .

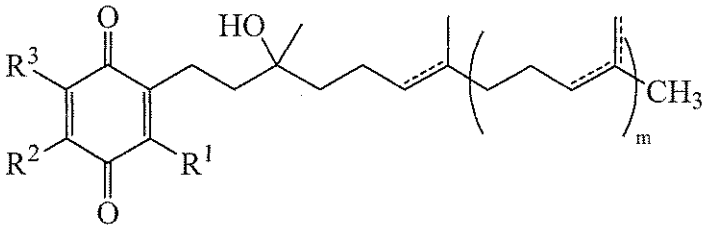
14. The formulation of any one of Claims 1-6 and 8-10, for use as defined in said claim, wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of a neurodegenerative disease; Parkinson's disease; Alzheimer's disease; Amyotrophic Lateral Sclerosis (ALS); a motor neuron disease; Huntington's Disease; an age-associated disease; glaucoma; a disorder of the outer retina; macular degeneration; age related macular degeneration; and juvenile macular degeneration.

15. The formulation of any one of Claims 1-6 and 8-10, for use as defined in said claim, wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of diabetic retinopathy; Progressive Supranuclear Palsy (PSP); a Parkinson-like disease; Chacot-Marie-Tooth Disease; a Mucopolysaccharidosis; Adrenoleukodystrophy; Niemann-Pick disease; Krabbe's disease; Pelizaeus-Merzbacher disease; and Progressive Encephalopathy, Edema, Hypsarrhythmia and Optic Atrophy (PEHO).

16. The formulation of any one of Claims 1-6 and 8-10, for use as defined in said claim, wherein the ophthalmic disorder is associated with a trauma selected from the group

consisting of retinal ischemia, an acute retinopathy associated with trauma, a post-surgical complication , damage associated with laser therapy including photodynamic therapy (PDT), traumatic optic neuropathy (TON), damage associated with surgical light induced iatrogenic retinopathy, damage associated with a corneal transplant , and damage associated with stem cell transplant of eye cells.

17. A compound of the Formula I:



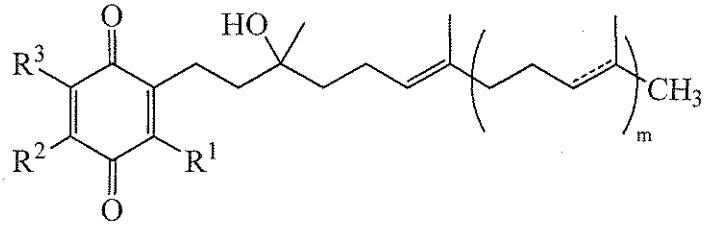
Formula I

or the hydroquinone form thereof,

wherein the bond indicated by a dashed line can be double or single; and

wherein the compound is selected from the group consisting of:

(a)



or the hydroquinone form thereof,

wherein:

the bond indicated by a dashed line is double or single;

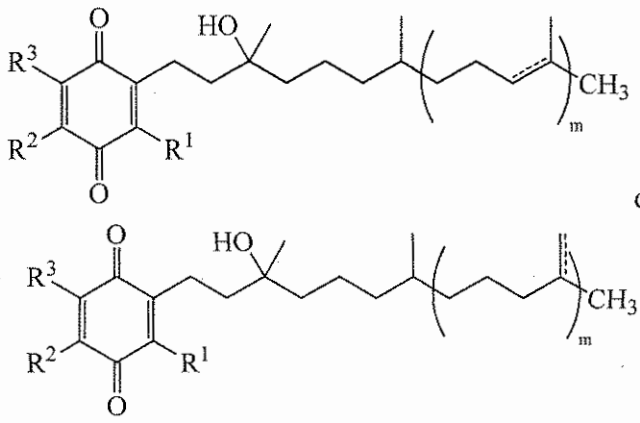
either R^2 and R^3 are independently of each other (C_1-C_6) alkoxy and R^1 is (C_1-C_6) alkyl or hydrogen, or R^1, R^2, and R^3 are independently of each other (C_1-C_6) alkyl; and

m is an integer from 0-1;

or any stereoisomer, mixture of stereoisomers, or salt, thereof;

with the proviso that the compound is not 2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione, , or the corresponding hydroquinone form thereof; and

(b)



or

or the hydroquinone form thereof,

wherein:

the bond indicated by a dashed line is a double bond;

R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6) alkoxy; and m is 1,

or any stereoisomer, mixture of stereoisomers, or salt thereof.

18. The compound according to claim 17, wherein m is 0 in the compound in (a).

19. The compound according to claim 17, wherein m is 1.

20. A compound according to claim 17, wherein the compound is selected from the group consisting of:

2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodec-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodec-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

and the hydroquinone forms thereof;

or any stereoisomer, mixture of stereoisomers, or salt thereof.

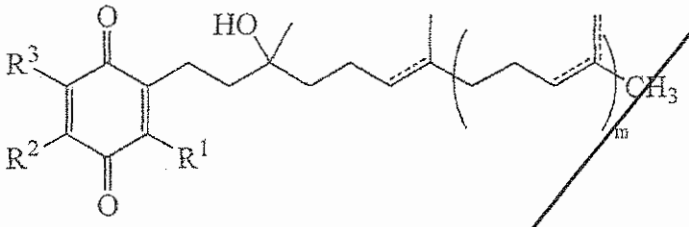
21. The compound according to any one of claims 17-20, wherein the compound is a quinone.

22. The compound of claim 20, wherein the compound is 2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione; or any stereoisomer, mixture of stereoisomers, or salt thereof.

What is claimed is:

1. A formulation for use in preventing, reducing, ameliorating or treating an ophthalmic disorder, or for stopping the progression of, or reversing, the loss of vision in a patient, wherein the formulation comprises an ophthalmically effective amount of one or more compounds selected from:

quinones of Formula I:



Formula I

and the hydroquinone forms thereof,

wherein:

the bonds indicated by a dashed line are independently of each other, at each occurrence, double or single with the proviso that at least one bond is a double bond, and with the proviso that they are not both double within the same unit;

R^1, R^2, and R^3 are independently of each other hydrogen, $(C_1\text{-}C_6)$ alkyl, or $(C_1\text{-}C_6)$ alkoxy; and

m is an integer from 0-1 or 3-12 inclusive, wherein each unit is the same or different;

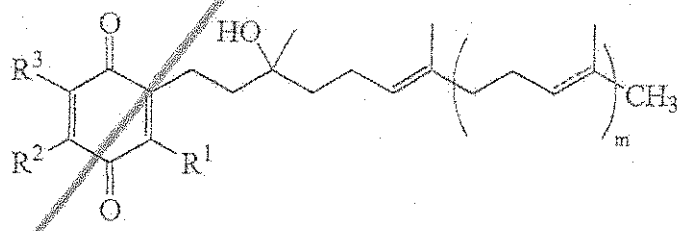
or any stereoisomer, mixture of stereoisomers, hydrate or solvate thereof;

wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of an inherited mitochondrial disease; Leber's Hereditary Optic Neuropathy (LHON); Dominant Optic Atrophy (DOA); Chronic Progressive External Ophthalmoplegia (CPEO); Spinocerebellar ataxia (SCA); Leigh's Syndrome; Friedreich's ataxia (FRDA); Mitochondrial Myopathy, Encephalopathy, Lactacidosis, and Stroke (MELAS); Myoclonic Epilepsy with Ragged Red Fibers (MERRF); Kearns-Sayre Syndrome (KSS); an overlap syndrome; Co-Enzyme Q10 (CoQ10) Deficiency; Complex I deficiency; Complex II deficiency; Complex III deficiency; Complex IV deficiency; Complex V deficiency; a neurodegenerative disease; Parkinson's disease; Alzheimer's

disease; Amyotrophic Lateral Sclerosis (ALS); a motor neuron disease; Huntington's Disease; an age-associated disease; glaucoma; a disorder of the outer retina; macular degeneration; age related macular degeneration; juvenile macular degeneration; diabetic retinopathy; Progressive Supranuclear Palsy (PSP); a Parkinson-like disease; Charcot-Marie-Tooth Disease; a Mucopolysaccharidosis; Adrenoleukodystrophy; Niemann-Pick disease; Krabbe's disease; Pelizaeus-Merzbacher disease; and Progressive Encephalopathy, Edema, Hypsarrhythmia and Optic Atrophy (PEHO); or wherein the ophthalmic disorder is associated with a trauma selected from the group consisting of retinal ischemia, an acute retinopathy associated with trauma, a post-surgical complication, damage associated with laser therapy including photodynamic therapy (PDT), traumatic optic neuropathy (TON), damage associated with surgical light induced iatrogenic retinopathy, damage associated with a corneal transplant, and damage associated with stem cell transplant of eye cells.

2. The formulation of Claim 1, for use as defined in said claim, wherein the formulation comprises an ophthalmically effective amount of one or more compounds selected from:

quinones of Formula I-a:



Formula I-a

and the hydroquinone forms thereof,

wherein:

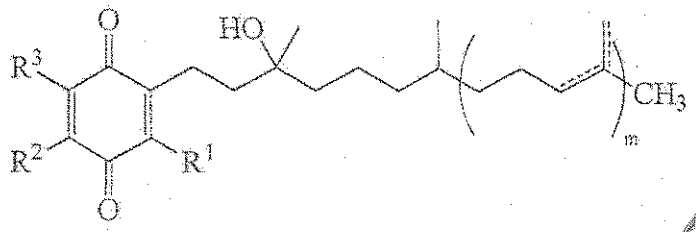
the bond indicated by a dashed line is double or single;

R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6)alkoxy; and

m is 0-1 or 3-12 inclusive, wherein each unit is the same or different;

or any stereoisomer, mixture of stereoisomers, hydrate or solvate thereof.

3. The formulation of Claim 1, for use as defined in said claim, wherein the formulation comprises an ophthalmically effective amount of one or more compounds selected from quinones of Formula I-c:



Formula I-c

and the hydroquinone forms thereof,

wherein:

the bonds indicated by a dashed line are double or single, with the proviso that they are not both double within the same unit; and the further proviso that at least one bond is a double bond;

R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6) alkoxy; and m is 1 or an integer from 3-12 inclusive, wherein each unit is the same or different;

or any stereoisomer, mixture of stereoisomers, hydrate or solvate thereof.

4. The formulation according to Claim 1, for use as defined in said claim, wherein the one or more compounds are selected from the group consisting of:

2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodec-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodec-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15,19,23-hexamethyltetracosa-6,10,14,18,22-pentaen-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;
5-(3-hydroxy-3,7,11,15,19,23-hexamethyltetracosa-6,10,14,18,22-pentaen-1-yl)-2,3-dimethylcyclohexa-2,5-diene-1,4-dione;
2-(3-hydroxy-3,7,11,15,19,23-hexamethyltetracos-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;
2-(3-hydroxy-3,7,11,15,19,23-hexamethyltetracosa-6,10,14,18,22-pentaen-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;
5-(3-hydroxy-3,7,11,15,19,23-hexamethyltetracosa-6,10,14,18,22-pentaen-1-yl)-2,3-dimethoxycyclohexa-2,5-diene-1,4-dione;
2-(3-hydroxy-3,7,11,15,19,23,27,31,35-nonamethylhexatriaconta-6,10,14,18,22,26,30,34-octaen-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;
2-(3-hydroxy-3,7,11,15,19,23,27,31,35-nonamethylhexatriaconta-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;
2-(3-hydroxy-3,7,11,15,19,23,27,31,35-nonamethylhexatriaconta-6,10,14,18,22,26,30,34-octaen-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;
2-(3-hydroxy-3,7,11,15,19,23,27,31,35-nonamethylhexatriaconta-6,10-dien-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;
5-(3-hydroxy-3,7,11,15,19,23,27,31,35-nonamethylhexatriaconta-6,10,14,18,22,26,30,34-octaen-1-yl)-2,3-dimethoxycyclohexa-2,5-diene-1,4-dione;
and the hydroquinone forms thereof;
or any stereoisomer, mixture of stereoisomers, hydrate or solvate thereof.

5. The formulation according to Claim 1, for use as defined in said claim, wherein the one or more compounds are selected from the group consisting of:
2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione; and
2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;
or any stereoisomer, mixture of stereoisomers, hydrate or solvate thereof.

6. The formulation according to any one of Claims 1-4, for use as defined in said claim, wherein the one or more compounds are quinones.

7. The formulation of any one of Claims 1-6, for use as defined in said claim, additionally comprising a pharmaceutically or ophthalmically acceptable vehicle.

8. Use of a compound of Formula I as defined in claim 1, Formula I-a as defined in claim 2, Formula I-c as defined in claim 3, or a compound according to claim 4, or the hydroquinone forms thereof, or any stereoisomer, mixture of stereoisomers, hydrate or solvate thereof, in the manufacture of a medicament for preventing, reducing, ameliorating or treating an ophthalmic disorder, or for stopping the progression of, or reversing, the loss of vision in a patient; wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of an inherited mitochondrial disease; Leber's Hereditary Optic Neuropathy (LHON); Dominant Optic Atrophy (DOA); Chronic Progressive External Ophthalmoplegia (CPEO); Spinocerebellar ataxia (SCA); Leigh's Syndrome; Friedreich's ataxia (FRDA); Mitochondrial Myopathy, Encephalopathy, Lactacidosis, and Stroke (MELAS); Myoclonic Epilepsy with Ragged Red Fibers (MERRF); Kearns-Sayre Syndrome (KSS); an overlap syndrome; Co-Enzyme Q10 (CoQ10) Deficiency; Complex I deficiency; Complex II deficiency; Complex III deficiency; Complex IV deficiency; Complex V deficiency; a neurodegenerative disease; Parkinson's disease; Alzheimer's disease; Amyotrophic Lateral Sclerosis (ALS); a motor neuron disease; Huntington's Disease; an age-associated disease; glaucoma; a disorder of the outer retina; macular degeneration; age related macular degeneration; juvenile macular degeneration; diabetic retinopathy; Progressive Supranuclear Palsy (PSP); a Parkinson-like disease; Chacot-Marie-Tooth Disease; a Mucopolysaccharidosis; Adrenoleukodystrophy; Niemann-Pick disease; Krabbe's disease; Pelizaeus-Merzbacher disease; and Progressive Encephalopathy, Edema, Hypsarrhythmia and Optic Atrophy (PEHO); or
wherein the ophthalmic disorder is associated with a trauma selected from the group consisting of retinal ischemia, an acute retinopathy associated with trauma, a post-surgical complication, damage associated with laser therapy including photodynamic therapy (PDT), traumatic optic neuropathy (TON), damage associated with surgical light

induced iatrogenic retinopathy, damage associated with a corneal transplant, and damage associated with stem cell transplant of eye cells.

9. The formulation of any one of Claims 1-7 , for use as defined in said claim, wherein the formulation is administered orally.

10. The formulation of any one of Claims 1-7 , for use as defined in said claim, wherein the formulation is administered topically.

11. The formulation of any one of Claims 1-7 , for use as defined in said claim, wherein the formulation is administered topically in eye drops or an irrigating solution, periocularly, or intraocularly.

12. The formulation of any one of Claims 1-7 and 9-11, for use as defined in said claim, wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of an inherited mitochondrial disease; Leber's Hereditary Optic Neuropathy (LHON); Dominant Optic Atrophy (DOA); Chronic Progressive External Ophthalmoplegia (CPEO); Spinocerebellar ataxia (SCA); Leigh's Syndrome; Friedreich's ataxia (FRDA); Mitochondrial Myopathy, Encephalopathy, Lactacidosis, and Stroke (MELAS); Myoclonic Epilepsy with Ragged Red Fibers (MERRF); Kearns-Sayre Syndrome (KSS); an overlap syndrome; Co-Enzyme Q10 (CoQ10) Deficiency; Complex I deficiency; Complex II deficiency; Complex III deficiency; Complex IV deficiency; and Complex V deficiency.

13. The formulation of any one of Claims 1-7 and 9-11, for use as defined in said claim, wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of Leber's Hereditary Optic Neuropathy (LHON); Dominant Optic Atrophy (DOA); and Chronic Progressive External Ophthalmoplegia (CPEO).

14. The formulation of any one of Claims 1-7 and 9-11, for use as defined in said claim, wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of Friedreich's ataxia (FRDA); Mitochondrial Myopathy, Encephalopathy,

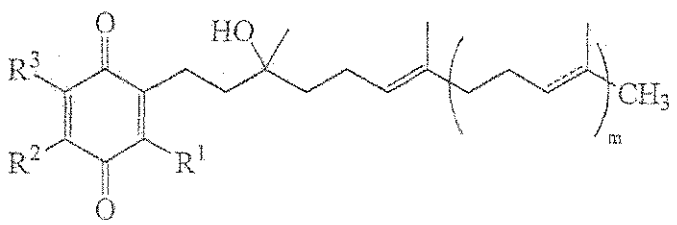
Lactacidosis, and Stroke (MELAS); Myoclonic Epilepsy with Ragged Red Fibers (MERRF); Leigh's syndrome; Kearns-Sayre Syndrome (KSS); and an overlap syndrome .

15. The formulation of any one of Claims 1-7 and 9-11, for use as defined in said claim, wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of a neurodegenerative disease; Parkinson's disease; Alzheimer's disease; Amyotrophic Lateral Sclerosis (ALS); a motor neuron disease; Huntington's Disease; an age-associated disease; glaucoma; a disorder of the outer retina; macular degeneration; age related macular degeneration; and juvenile macular degeneration.

16. The formulation of any one of Claims 1-7 and 9-11, for use as defined in said claim, wherein the ophthalmic disorder is associated with a disorder selected from the group consisting of diabetic retinopathy; Progressive Supranuclear Palsy (PSP); a Parkinson-like disease; Chacot-Marie-Tooth Disease; a Mucopolysaccharidosis; Adrenoleukodystrophy; Niemann-Pick disease; Krabbe's disease; Pelizaeus-Merzbacher disease; and Progressive Encephalopathy, Edema, Hypsarrhythmia and Optic Atrophy (PEHO).

17. The formulation of any one of Claims 1-7 and 9-11, for use as defined in said claim, wherein the ophthalmic disorder is associated with a trauma selected from the group consisting of retinal ischemia, an acute retinopathy associated with trauma, a post-surgical complication , damage associated with laser therapy including photodynamic therapy (PDT), traumatic optic neuropathy (TON), damage associated with surgical light induced iatrogenic retinopathy, damage associated with a corneal transplant , and damage associated with stem cell transplant of eye cells.

18. A compound of the formula:



or the hydroquinone form thereof,

wherein:

the bond indicated by a dashed line is double or single;

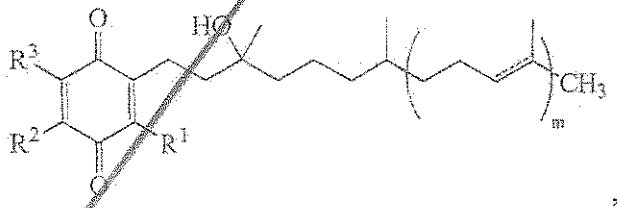
R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6) alkoxy; and

m is an integer from 0-1 or 3-12 inclusive, wherein each unit is the same or different;

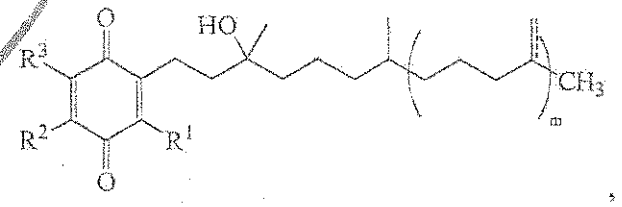
or any stereoisomer, mixture of stereoisomers, prodrug, metabolite, salt, crystalline form, non-crystalline form, hydrate or solvate thereof;

with the proviso that the compound is not 2-[(6E,10E,14E,18E,22E,26E,30E,34E)-3-hydroxy-3,7,11,15,19,23,27,31,35,39-decamethyl-6,10,14,18,22,26,30,34,38-tetracontanonaen-1-yl]-5,6-dimethoxy-3-methyl-2,5-cyclohexadiene-1,4-dione, 2-(3-hydroxy-3,7,11,15,19,23,27-heptamethyl-6,10,14,18,22,26-octacosahexaenyl)-5,6-dimethoxy-3-methyl-p-benzoquinone or an isotopologue thereof, 2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione, 5-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-2,3-dimethylcyclohexa-2,5-diene-1,4-dione, or the corresponding hydroquinone forms thereof.

19. A compound having a formula selected from the group consisting of:



,



,

and the hydroquinone forms thereof,

wherein:

the bond indicated by a dashed line is in every occurrence double or single; with the proviso that at least one bond is a double bond;

R^1, R^2, and R^3 are independently of each other hydrogen, (C_1-C_6) alkyl, or (C_1-C_6) alkoxy; and

m is 1 or an integer from 3-12 inclusive, wherein each unit is the same or different,

or any stereoisomer, mixture of stereoisomers, prodrug, metabolite, salt, crystalline form, non-crystalline form, hydrate or solvate thereof.

20. The compound according to claim 18, wherein m is 0.

21. The compound according to any one of claims 18-19, wherein m is 1.

22. A compound selected from the group consisting of:

2-(3-hydroxy-3,7-dimethyloct-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodec-6-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11-trimethyldodec-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15-tetramethylhexadeca-6,10,14-trien-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15-tetramethylhexadec-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

5-(3-hydroxy-3,7,11,15-tetramethylhexadeca-6,10,14-trien-1-yl)-2,3-dimethoxycyclohexa-2,5-diene-1,4-dione;

2,3-diethyl-5-(3-hydroxy-3,7,11,15-tetramethylhexadeca-6,10,14-trien-1-yl)-6-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15-tetramethylhexadeca-6,10,14-trien-1-yl)-5,6-diisopropyl-3-methylcyclohexa-2,5-diene-1,4-dione;

5-(3-hydroxy-3,7,11,15,19,23-hexamethyltetracosa-6,10,14,18,22-pentaen-1-yl)-2,3-dimethoxycyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15,19,23-hexamethyltetracosa-6,10,14,18,22-pentaen-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15,19,23-hexamethyltetracos-6-en-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15,19,23-hexamethyltetracosa-6,10,14,18,22-pentaen-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

5-(3-hydroxy-3,7,11,15,19,23-hexamethyltetracosa-6,10,14,18,22-pentaen-1-yl)-2,3-dimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15,19,23,27,31,35-nonamethylhexatriaconta-6,10,14,18,22,26,30,34-octaen-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15,19,23,27,31,35-nonamethylhexatriaconta-6,10,14,18,22,26,30,34-octaen-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

5-(3-hydroxy-3,7,11,15,19,23,27,31,35-nonamethylhexatriaconta-6,10,14,18,22,26,30,34-octaen-1-yl)-2,3-dimethoxycyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15,19,23,27,31,35-nonamethylhexatriaconta-6,10-dien-1-yl)-5,6-dimethoxy-3-methylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15,19,23,27,31,35-nonamethylhexatriaconta-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15-tetramethylhexadec-14-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

2-(3-hydroxy-3,7,11,15-tetramethylhexadec-15-en-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione;

and the hydroquinone forms thereof;

or any stereoisomer, mixture of stereoisomers, prodrug, metabolite, salt, crystalline form, non-crystalline form, hydrate or solvate thereof.

23. The compound according to any one of claims 18-22, wherein the compound is a quinone.

24. The compound of claim 22, wherein the compound is 2-(3-hydroxy-3,7,11-trimethyldodeca-6,10-dien-1-yl)-3,5,6-trimethylcyclohexa-2,5-diene-1,4-dione; or any stereoisomer, mixture of stereoisomers, salt, crystalline form, non-crystalline form, hydrate or solvate thereof.