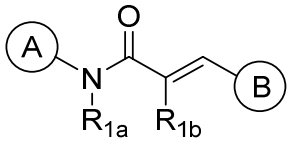
CLAIMS

1. A compound of formula (I):



(I)

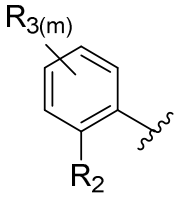
wherein:

R_{1a} is H or methyl;

R_{1b} is H or fluoro;

A is group (Aa), (Ab), (Ac) or (Ad):

wherein group (Aa) is:



(Aa);

wherein:

R_2 is H, C_{1-4}alkyl, C_{1-4}alkylene(aryl), C_{1-4}alkylene(OH), C_{1-4}alkylene(C_{3-6}cycloalkyl), C_{1-4}alkylene(4-7 membered heterocycloalkyl), C_{1-4}alkoxy, OC_{1-4}alkylene(aryl), C_{1-4}alkyleneOC_{1-4}alkyl, C_{1-4}alkyleneOC_{3-6}cycloalkyl, C_{1-4}alkyleneO(4-7 membered heterocycloalkyl), C_{1-4}alkyleneO(aryl), C_{3-6}alkynyl and C_{1-4}alkyleneO(C_{3-6}alkynyl);

wherein said aryl, heterocycloalkyl or cycloalkyl are optionally substituted by 1, 2 or 3 substituents each independently selected from C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, halo, CN, OH, $NR_{2a}R_{2b}$, SO_2R_{2c} and $NHSO_2R_{2c}$;

R_{2a} is selected from H and C_{1-4}alkyl;

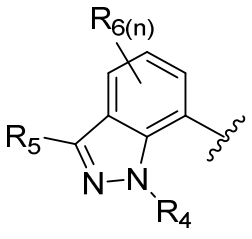
R_{2b} is selected from H, C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, aryl and 4-7 membered heterocycloalkyl;

R_{2c} is selected from C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, aryl and 4-7 membered heterocycloalkyl;

each R_3 is independently halo, methyl, ethyl or n-propyl;

m is 0, 1, 2, 3 or 4;

wherein group (Ab) is:



(Ab);

wherein:

R_4 is H, C_{1-4}alkyl or C_{1-4}alkylene(aryl); wherein said aryl is optionally substituted by 1, 2 or 3 substituents each independently selected from C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, halo, CN, OH, $NR_{4a}R_{4b}$, SO_2R_{4c} and $NHSO_2R_{4c}$;

R_{4a} is selected from H and C_{1-4}alkyl;

R_{4b} is selected from H, C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, aryl and 4-7 membered heterocycloalkyl;

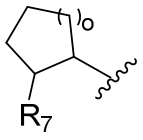
R_{4c} is selected from C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, aryl and 4-7 membered heterocycloalkyl;

R_5 is H or C_{1-4}alkyl;

each R_6 is independently C_{1-4}alkyl or halo;

n is 0, 1, 2 or 3;

wherein group (Ac) is:



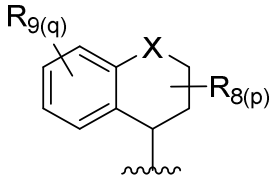
(Ac);

wherein:

R_7 is C_{1-4}alkyl, C_{1-4}alkylene(OH) or C_{1-4}alkyleneOC_{1-4}alkyl;

o is 1 or 2;

wherein group (Ad) is:



(Ad);

wherein:

X is a bond, O or CH_2;

each R_8 is independently halo, C_{1-4}alkyl, C_{1-4}alkoxy OC_{1-4}haloalkyl, OC_{1-4}alkylene(C_{3-6}cycloalkyl), OC_{1-4}alkylene(4-7 membered heterocycloalkyl) or OH; wherein said heterocycloalkyl and cycloalkyl are optionally substituted by 1, 2 or 3 substituents independently selected from C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, halo, CN, OH, $NR_{8a}R_{8b}$, SO_2R_{8c} and $NHSO_2R_{8c}$;

R_{8a} is selected from H and C_{1-4}alkyl;

R_{8b} is selected from H, C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, aryl and 4-7 membered heterocycloalkyl;

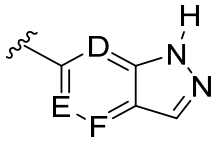
R_{8c} is selected from C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, aryl and 4-7 membered heterocycloalkyl;

each R_9 is independently halo or C_{1-4}alkyl;

p is 0, 1 or 2;

q is 0, 1, 2, 3 or 4;

wherein B is:



;

wherein:

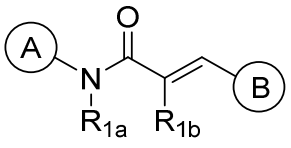
R_{10} is H, halo or C_{1-4}alkyl;

D, E and F are each independently C(R_{10}); or one of D, E and F is N and the two remaining D, E and F groups are independently C(R_{10}); and

provided that the compound of formula (I) is not (E)-*N*-(3-fluoro-2-methylphenyl)-3-(1H-indazol-6-yl)acrylamide;

or a pharmaceutically acceptable salt and/or solvate thereof.

2. The compound, pharmaceutically acceptable salt and/or solvate thereof according to claim 1, which is a compound of formula (I):



(I)

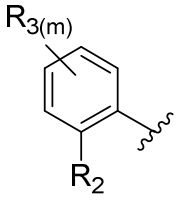
wherein:

R_{1a} is H or methyl;

R_{1b} is H or fluoro;

A is group (Aa), (Ab), (Ac) or (Ad):

wherein group (Aa) is:



(Aa);

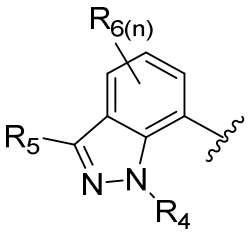
wherein:

R_2 is H, C_{1-4}alkyl, C_{1-4}alkylene(aryl), C_{1-4}alkylene(OH), C_{1-4}alkylene(C_{3-6}cycloalkyl), C_{1-4}alkylene(4-7 membered heterocycloalkyl), C_{1-4}alkoxy, OC_{1-4}alkylene(aryl), C_{1-4}alkyleneOC_{1-4}alkyl, C_{1-4}alkyleneOC_{3-6}cycloalkyl, C_{1-4}alkyleneO(4-7 membered heterocycloalkyl), C_{1-4}alkyleneO(aryl), C_{3-6}alkynyl or C_{1-4}alkyleneO(C_{3-6}alkynyl); wherein said aryl, heterocycloalkyl or cycloalkyl may be optionally substituted by up to 3 substituents each independently selected from C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, halo and CN;

each R_3 is independently halo, methyl, ethyl or n-propyl;

m is 0, 1, 2, 3 or 4;

wherein group (Ab) is:



(Ab);

wherein:

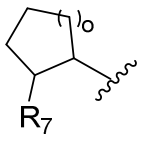
R_4 is H, C_{1-4}alkyl or C_{1-4}alkylene(aryl); wherein said aryl may be optionally substituted by up to 3 substituents each independently selected from C_{1-4}alkyl, C_{3-6}cycloalkyl, C_{1-4}alkoxy, C_{1-4}haloalkyl, halo and CN;

R_5 is H or C_{1-4}alkyl;

each R_6 is independently C_{1-4}alkyl or halo;

n is 0, 1, 2 or 3;

wherein group (Ac) is:



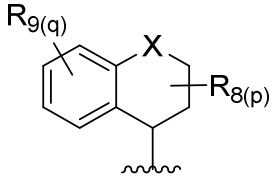
(Ac);

wherein:

R_7 is C_{1-4}alkyl, C_{1-4}alkylene(OH) or C_{1-4}alkyleneOC_{1-4}alkyl;

o is 1 or 2;

wherein group (Ad) is:



(Ad);

wherein:

X is a bond, O or CH_2;

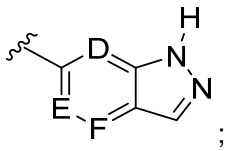
each R_8 is independently halo, C_{1-4}alkyl, C_{1-4}alkoxy or OH;

each R_9 is independently halo or C_{1-4}alkyl;

p is 0, 1 or 2;

q is 0, 1, 2, 3 or 4;

wherein B is:



;

wherein:

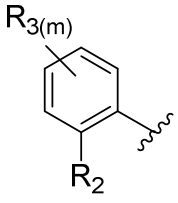
R_{10} is H, halo or C_{1-4}alkyl; and

D, E and F are each independently C(R_{10}); or one of D, E and F is N and the two remaining D, E and F groups are independently C(R_{10});

provided that the compound of formula (I) is not (E)-*N*-(3-fluoro-2-methylphenyl)-3-(1H-indazol-6-yl)acrylamide;

or a pharmaceutically acceptable salt and/or solvate thereof.

3. The compound, pharmaceutically acceptable salt and/or solvate thereof according to claim 1 or 2, wherein A is group (Aa):



(Aa),

.

~~4. The compound according to any one of claims 1 to 3,~~ wherein:

R_2 is C_{1-4}alkyl, C_{1-4}alkylene(aryl), C_{1-4}alkylene(OH), C_{1-4}alkyleneOC_{1-4}alkyl, C_{1-4}alkyleneOC_{3-6}cycloalkyl, C_{1-4}alkyleneO(aryl), C_{1-4}alkylene(4-7 membered heterocycloalkyl), C_{1-4}alkyleneO(4-7 membered heterocycloalkyl) and C_{1-4}alkyleneO(C_{3-6}alkynyl); wherein said aryl, heterocycloalkyl or cycloalkyl may be optionally substituted by up to 3 substituents each independently selected from C_{1-4}alkyl, C_{3-6}cycloalkyl; C_{1-4}alkoxy, C_{1-4}haloalkyl, halo and CN; .

~~5. The compound according to claim 4, wherein the aryl is unsubstituted.~~

~~6. The compound according to claim 4, wherein the aryl is substituted by 1, 2 or 3 substituents independently selected from methyl, chloro and fluoro.~~

~~7. The compound according to claim 4, wherein the heterocycloalkyl is unsubstituted.~~

~~8. The compound according to claim 4, wherein the heterocycloalkyl is substituted by 1, 2 or 3 substituents independently selected from CH_2CH_2F and fluoro.~~

~~9. The compound according to any one of claims 1 to 8, wherein~~ each R_3 is independently fluoro or methyl; and~~.~~

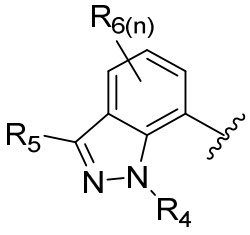
~~10. The compound according to any one of claims 1 to 9, wherein~~ m is 1 or 2.

~~11. The compound according to any one of claims 1 to 10, wherein m is 1 and R_3 is in the 3-position.~~

~~12. The compound according to any one of claims 1 to 10, wherein m is 1 and R_3 is in the 6-position.~~

~~13. The compound according to any one of claims 1 to 10, wherein m is 2, one R_3 is in the 3-position, and the other R_3 is in the 6-position.~~

~~1~~4. The compound, pharmaceutically acceptable salt and/or solvate thereof according to claim 1 or 2, wherein A is group (Ab):



(Ab).

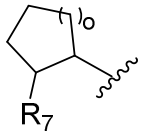
~~1~~5. The compound, pharmaceutically acceptable salt and/or solvate thereof according to claim ~~14~~4, wherein R_4 is H, methyl or benzyl, ~~.~~

~~16. The compound according to claim 14 or 15, wherein~~ R^5 is H and~~.~~

~~17. The compound according to any one of claims 14 to 16, wherein each R_6 is independently fluoro or methyl.~~

~~18. The compound according to any one of claims 14 to 17, wherein~~ n is 0.

~~19~~6. The compound, pharmaceutically acceptable salt and/or solvate thereof according to claim 1 or 2, wherein A is group (Ac):

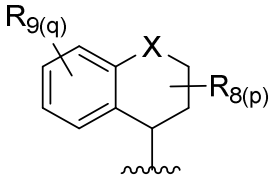


(Ac).

~~20~~7. The compound, pharmaceutically acceptable salt and/or solvate thereof according to claim ~~19~~6, wherein R_7 is methyl, CH_2OH or CH_2OMe and ~~.~~

~~21. The compound according to claim 19 or 20, wherein~~ o is 2.

~~22~~8. The compound, pharmaceutically acceptable salt and/or solvate thereof according to claim 1 or 2, wherein A is group (Ad):



(Ad).

~~23~~9. The compound, pharmaceutically acceptable salt and/or solvate thereof according to claim ~~22~~8, wherein X is a bond or O~~.~~.

~~24. The compound according to claim 22 or 23, wherein~~ p is 1 and ~~each~~ R_8 is ~~independently~~selected from methyl, OMe or fluoro~~.~~.

~~25. The compound according to any one of claims 22 to 24, wherein p is 0 or 1.~~

~~26~~10. The compound, pharmaceutically acceptable salt and/or solvate thereof according to ~~any one of~~ claim~~s~~ ~~22~~8 ~~to~~or ~~25~~9, wherein each R_9 is independently fluoro and~~.~~

~~27. The compound according to any one of claims 22 to 26, wherein~~ q is 1 or 2.

~~28~~11. The compound, pharmaceutically acceptable salt and/or solvate thereof according to any one of claims 1 to ~~27~~10, wherein D, E and F are $C(R_{10})$ and ~~.~~

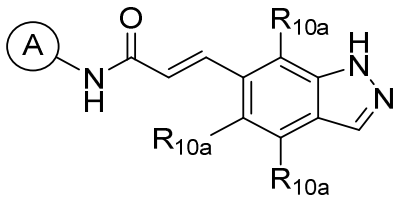
~~29. The compound according to any one of claims 1 to 27, wherein D is N, and E and F are C(R_{10}).~~

~~30. The compound according to any one of claims 1 to 27, wherein E is N, and D and F are C(R_{10}).~~

~~31. The compound according to any one of claims 1 to 27, wherein F is N, and D and E are C(R_{10}).~~

~~32. The compound according to any one of claims 1 to 31, wherein~~ each R_{10} is independently H, fluoro, chloro or methyl.

12. The compound, pharmaceutically acceptable salt and/or solvate thereof according to claim 1, which is a compound of formula (Ia):

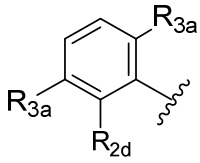


(Ia),

wherein A is group (Aa'), (AdI') or (AdII');

R_{10a} is H, fluoro, chloro or methyl;

wherein group (Aa') is:



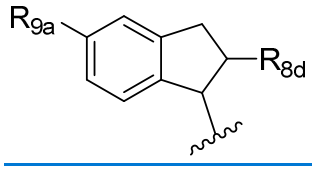
(Aa');

wherein:

R_{2d} is methyl, CH_2OMe or $CH_2OCH_2C{\equiv}CH$;

each R_{3a} is independently H, methyl or fluoro; and

wherein group (AdI') is:



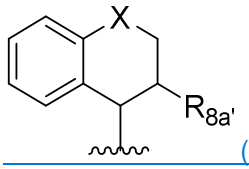
(AdI');

wherein:

R_{8d} is H, methyl or OMe;

R_{9a} is H or F;

wherein group (AdII') is:



(AdII'),

wherein:

X is O; and

$R_{8a'}$ is methyl;

provided that when A is group (AdI') and one of R_{10a} is fluoro or chloro, the two remaining R_{10a} groups are independently H;

or a pharmaceutically acceptable salt and/or solvate thereof.

~~3~~13. The compound, pharmaceutically acceptable salt and/or solvate thereof according to claim 1, which is selected from the group consisting of:

(E)-*N*-(3-fluoro-2-methylphenyl)-3-(7-methyl-1H-indazol-6-yl)acrylamide;

(E)-*N*-(2,3-dihydro-1H-inden-1-yl)-3-(1H-indazol-6-yl)acrylamide;

(*R,E*)-*N*-(2,3-dihydro-1H-inden-1-yl)-3-(1H-indazol-6-yl)acrylamide;

(E)-3-(1H-indazol-6-yl)-*N*-((1R,2R)-2-methylcyclohexyl) acrylamide (racemic mixture);

(E)-3-(1H-indazol-6-yl)-*N*-((1R,2R)-2-methylcyclohexyl) acrylamide (enantiomer 1);

(E)-3-(1H-indazol-6-yl)-*N*-((1R,2R)-2-methylcyclohexyl) acrylamide (enantiomer 2);

(E)-*N*-(7-fluoro-2,3-dihydro-1H-inden-1-yl)-3-(1H-indazol-6-yl)acrylamide;

(E)-*N*-(6-fluoro-2,3-dihydro-1H-inden-1-yl)-3-(1H-indazol-6-yl)acrylamide;

(E)-*N*-(5-fluoro-2,3-dihydro-1H-inden-1-yl)-3-(1H-indazol-6-yl)acrylamide;

(E)-*N*-(4-fluoro-2,3-dihydro-1H-inden-1-yl)-3-(1H-indazol-6-yl)acrylamide;

(E)-*N*-(2,3-dihydro-1H-inden-1-yl)-3-(7-fluoro-1H-indazol-6-yl)acrylamide;

(E)-*N*-(2,3-dihydro-1H-inden-1-yl)-3-(5-fluoro-1H-indazol-6-yl)acrylamide;

(E)-*N*-(2,3-dihydro-1H-inden-1-yl)-3-(4-fluoro-1H-indazol-6-yl)acrylamide;

(E)-3-(5-chloro-1H-indazol-6-yl)-*N*-(2,3-dihydro-1H-inden-1-yl)acrylamide;

(E)-3-(4-chloro-1H-indazol-6-yl)-*N*-(2,3-dihydro-1H-inden-1-yl)acrylamide;

(E)-3-(1H-indazol-6-yl)-*N*-(2-methyl-2,3-dihydro-1H-inden-1-yl)acrylamide (mixture of stereoisomers);

(E)-3-(1H-indazol-6-yl)-*N*-(2-methyl-2,3-dihydro-1H-inden-1-yl)acrylamide (stereoisomer 1);

(E)-3-(1H-indazol-6-yl)-*N*-(2-methyl-2,3-dihydro-1H-inden-1-yl)acrylamide (stereoisomer 2);

(E)-3-(1H-indazol-6-yl)-*N*-(2-methyl-2,3-dihydro-1H-inden-1-yl)acrylamide (stereoisomer 3);

(E)-3-(1H-indazol-6-yl)-*N*-(2-methyl-2,3-dihydro-1H-inden-1-yl)acrylamide (stereoisomer 4);

(E)-3-(1H-indazol-6-yl)-*N*-(3-methylchroman-4-yl)acrylamide (mixture of stereoisomers);

(E)-3-(1H-indazol-6-yl)-*N*-(3-methylchroman-4-yl)acrylamide (stereoisomer 1);
(E)-3-(1H-indazol-6-yl)-*N*-(3-methylchroman-4-yl)acrylamide (stereoisomer 2);
(E)-3-(1H-indazol-6-yl)-*N*-(3-methylchroman-4-yl)acrylamide (stereoisomer 3);
(E)-3-(1H-indazol-6-yl)-*N*-(3-methylchroman-4-yl)acrylamide (stereoisomer 4);
(E)-*N*-(2-(benzyloxy)phenyl)-3-(1H-indazol-6-yl)acrylamide;
(E)-3-(1H-indazol-6-yl)-*N*-(2-(phenoxymethyl)phenyl)acrylamide;
(E)-*N*-(2-(cyclobutoxymethyl)phenyl)-3-(1H-indazol-6-yl)acrylamide;
(E)-*N*-(1-benzyl-1H-indazol-7-yl)-3-(1H-indazol-6-yl)acrylamide;
(E)-3-(1H-indazol-6-yl)-*N*-(1-methyl-1H-indazol-7-yl)acrylamide;
(E)-3-(1H-indazol-6-yl)-*N*-(m-tolyl)acrylamide;
(E)-*N*-(3-chlorophenyl)-3-(1H-indazol-6-yl)acrylamide;
(E)-*N*-(3-fluorophenyl)-3-(1H-indazol-6-yl)acrylamide;
(E)-*N*-(2,6-dimethylphenyl)-3-(1H-indazol-6-yl)acrylamide;
(E)-*N*-((1R,3R)-3-fluoro-2,3-dihydro-1H-inden-1-yl)-3-(1H-indazol-6-yl)acrylamide (mixture of stereoisomers);
(E)-*N*-((1R,3R)-3-fluoro-2,3-dihydro-1H-inden-1-yl)-3-(1H-indazol-6-yl)acrylamide (stereoisomer 1);
(E)-*N*-((1R,3S)-3-fluoro-2,3-dihydro-1H-inden-1-yl)-3-(1H-indazol-6-yl)acrylamide (stereoisomer 2);
(E)-*N*-(2-(hydroxymethyl)phenyl)-3-(1H-indazol-6-yl)acrylamide;
(E)-*N*-(3-fluoro-2,6-dimethylphenyl)-3-(1H-indazol-6-yl)acrylamide;
(E)-3-(1H-indazol-6-yl)-*N*-(2-(methoxymethyl)phenyl)acrylamide;
(E)-*N*-(2-(((1-(2-fluoroethyl)azetidin-3-yl)oxy)methyl)phenyl)-3-(1H-indazol-6-yl)acrylamide hydrochloride;
(E)-*N*-(2-((3-fluoroazetidin-1-yl)methyl)phenyl)-3-(1H-indazol-6-yl)acrylamide;
(E)-*N*-(2-(2-(3-fluoroazetidin-1-yl)ethyl)phenyl)-3-(1H-indazol-6-yl)acrylamide;
(E)-*N*-(2-fluoro-6-methylphenyl)-3-(1H-indazol-6-yl)acrylamide;
(E)-*N*-(3-fluoro-2-methylphenyl)-3-(1H-pyrazolo[4,3-*b*]pyridin-6-yl)acrylamide;
(E)-*N*-(2,3-dihydro-1H-inden-1-yl)-3-(1H-pyrazolo[4,3-*c*]pyridin-6-yl)acrylamide;
(E)-*N*-(2,3-dihydro-1H-inden-1-yl)-3-(1H-pyrazolo[3,4-*b*]pyridin-6-yl)acrylamide;
(E)-3-(1H-indazol-6-yl)-*N*-((1S,2S)-2-methoxy-2,3-dihydro-1H-inden-1-yl)acrylamide;
(E)-3-(1H-indazol-6-yl)-*N*-(7-methyl-2,3-dihydro-1H-inden-1-yl)acrylamide;
(E)-3-(1H-indazol-6-yl)-*N*-(3-methyl-2,3-dihydro-1H-inden-1-yl)acrylamide; and
(E)-3-(1H-indazol-6-yl)-*N*-(2-((prop-2-yn-1-yloxy)methyl)phenyl)acrylamide;
or a pharmaceutically acceptable salt and/or solvate of any one thereof.

~~34~~14. The compound, pharmaceutically acceptable salt and/or solvate thereof according to any one of claims 1 to ~~33~~13, for use as a pharmaceutical.

15. The compound of formula (I), or a pharmaceutically acceptable salt and/or solvate thereof according to any one of claims 1 to 13, for use in the manufacture of a medicament for the treatment or prophylaxis of a disease or disorder in which inhibition of mPTP provides a therapeutic or prophylactic effect.

~~35. The compound according to any one of claims 1 to 33, for use in the treatment or prophylaxis of a disease or disorder in which inhibition of mPTP provides a therapeutic or prophylactic effect.~~

~~36. The compound according to any one of claims 1 to 33, for use in the treatment of a disease or disorder in which inhibition of mPTP provides a therapeutic effect.~~

~~37. The compound according to any one of claims 1 to 33, for use in the prophylaxis of a disease or disorder in which inhibition of mPTP provides a prophylactic effect.~~

~~38~~16. Use of ~~T~~the compound, pharmaceutically acceptable salt and/or solvate thereof ~~for use~~ according to ~~any one of~~ claim~~s 35 to 37~~14, in the treatment or prophylaxis of ~~wherein the~~a disease or disorder ~~is~~ selected from degenerative or neurodegenerative diseases, disorders of the central nervous system, ischemia and re-perfusion injury, metabolic diseases, inflammatory or autoimmune diseases, diseases of aging and renal diseases, such as ~~.~~

~~39. The compound for use according to claim 38, wherein the disease or disorder is~~ a degenerative or neurodegenerative disease~~,~~ selected from ~~such as~~ Parkinson's disease, dementia with Lewy bodies, Alzheimer's disease, amyotrophic lateral sclerosis, multiple sclerosis, frontal temporal dementia, chemotherapy induced neuropathy, Huntington's disease, spinocerebellar ataxias, progressive supranuclear palsy, hereditary spastic paraplegia, Duchenne muscular dystrophy, congenital muscular dystrophy, traumatic brain injury and Friedreich's ataxia.

~~40. The compound for use according to claim 38, wherein the disease or disorder is a disease of the central nervous system, such as AIDS dementia complex, depressive disorders, schizophrenia and epilepsy.~~

41. The compound for use according to claim 38, wherein the disease or disorder is ischemia or re-perfusion injury, such as acute myocardial infarction, stroke, kidney ischemia reperfusion injury, and organ damage during transplantation.

42. The compound for use according to claim 38, wherein the disease or disorder is a metabolic disease, such as hepatic steatosis, diabetes, diabetic retinopathy, cognitive decline and other diabetes associated conditions, obesity and feeding behaviours, and non-alcoholic fatty liver disease.

43. The compound for use according to claim 38, wherein the disease or disorder is an inflammatory or autoimmune disease, such as acute pancreatitis, systemic lupus, organ failure in sepsis and hepatitis.

44. The compound for use according to claim 38, wherein the disease or disorder is a disease of aging, such as bone repair, bone weakness in aging in osteoporosis and sarcopenia.

45. The compound for use according to claim 38, wherein the disease or disorder is a renal disease, such as chronic kidney disease associated with APOL1 genetic variants and chronic kidney disease.

46. The compound according to any one of claims 1 to 33, for use in the treatment or prophylaxis of a mitochondrial disease.

47. The compound according to any one of claims 1 to 33, for use in the treatment of a mitochondrial disease.

48. The compound according to any one of claims 1 to 33, for use in the prophylaxis of a mitochondrial disease.

49. The compound according to any one of claims 1 to 33, for use in the treatment or prophylaxis of a disease or disorder associated with TPD-43 proteinopathy such as TDP-43 associated neurodegeneration.

50. The compound for use according to claim 49, wherein the disease or disorder is selected from Amyotrophic Lateral Sclerosis, Frontotemporal dementia, Facial onset sensory and motor neuronopathy, Primary lateral sclerosis, Progressive muscular atrophy, Inclusion

~~body myopathy associated with early-onset Paget disease of the bone and Frontotemporal lobar degeneration dementia, Perry disease, Chronic traumatic encephalopathy, Severe traumatic brain injury, Alzheimer's disease, Hippocampal sclerosis dementia, Limbic-predominant age-related TDP-43 encephalopathy, and Cerebral age-related TDP-43 with sclerosis.~~

~~51. The compound according to any one of claims 1 to 33, for use in the treatment or prophylaxis of a disease or disorder associated with fibrosis.~~

~~52. The compound according to claim 51, wherein the disease or disorder is selected from chronic kidney disease, idiopathic pulmonary fibrosis, non-alcoholic steatohepatitis, primary biliary cholangitis and systemic sclerosis.~~

~~53~~17. A pharmaceutical composition comprising a compound according to any one of claims 1 to ~~33~~13, or a pharmaceutically acceptable salt and/or solvate thereof, and a pharmaceutically acceptable carrier or excipient.

~~54. The compound or solvate thereof according to any one of claims 1 to 53.~~

~~55. The compound or pharmaceutically acceptable salt thereof according to any one of claims 1 to 53.~~

~~56. The compound according to any one of claims 1 to 53.~~