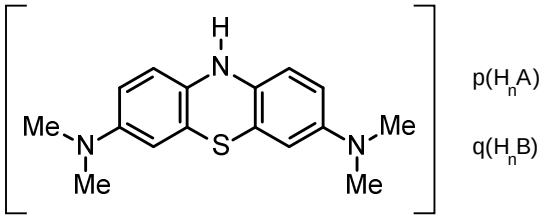
Claims

1 Use of an methylthioninium (“MT”)-containing compound in the manufacture of a medicament for use in a method of therapeutic or prophylactic treatment of a neurodegenerative disorder of protein aggregation in a human subject,

wherein the MT-containing compound is to be administered orally to the subject,

wherein a total daily dose of between 0.5 and 20mg of MT is to be administered to the subject per day, optionally split into 2 or 3 doses,

wherein the compound is a compound of the following formula (“LMTX”):



wherein each of H_nA and H_nB (where present) are protic acids which may be the same or different,

and wherein p = 1 or 2; q = 0 or 1; n = 1 or 2; (p + q) × n = 2,

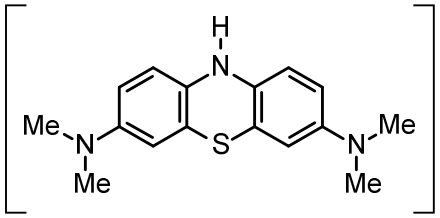
and wherein said neurodegenerative disorder is selected from the list consisting of: Alzheimer’s disease; Pick’s disease, progressive supranuclear palsy, frontotemporal dementia, FTD with parkinsonism linked to chromosome 17, frontotemporal lobar degeneration syndromes; disinhibition-dementia-parkinsonism-amyotrophy complex, pallido-ponto-nigral degeneration, Guam-ALS syndrome, pallido nigro luysian degeneration, cortico-basal degeneration, dementia with argyrophilic grains, dementia pugilistica or chronic traumatic encephalopathy, Down’s syndrome, subacute sclerosing panencephalitis, Niemann-Pick disease, type C, Sanfilippo syndrome type B, or a

myotonic dystrophy DM1 or DM2; Huntington's disease, spinal bulbar muscular atrophy, dentatorubropallidoluysian atrophy or spinocerebellar ataxias; A TDP-43 proteinopathy which is FTLD-TDP ; Parkinson's disease, dementia with Lewy bodies and multiple system atrophy; Hereditary cerebral angiopathy; Amyotrophic lateral sclerosis; familial encephalopathy with neuronal inclusion bodies.

2 Use as claimed in claim 1 wherein the total daily dosage is 2 to 15 mg; or 3 to 10 mg.

3 Use as claimed in claim 1 or claim 2 wherein:

(a) the compound has the following formula, where HA and HB are different mono-protic acids:

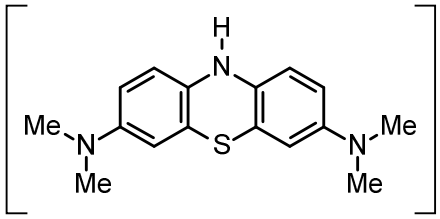


HA

HB

or

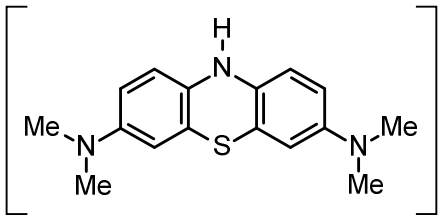
(b) the compound has the following formula:



$p(H_nX)$

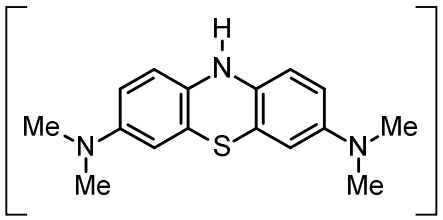
wherein each of H_nX is a protic acid, or

(c) the compound has the following formula and H_2A is a di-protic acid:



H_2A

4 Use as claimed in claim 3 wherein the compound has the following formula and is a bis-monoprotic acid:



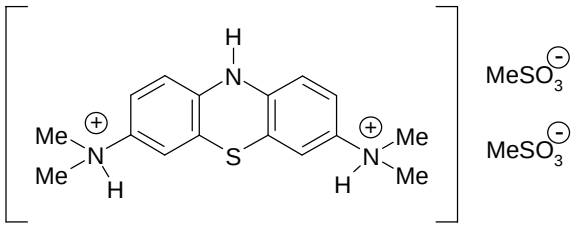
2(HA)

5 Use as claimed in any one of claims 1 to 4 wherein the or each protic acid is an inorganic acid.

6 Use as claimed in any one of claims 1 to 4 wherein the or each protic acid is an organic acid.

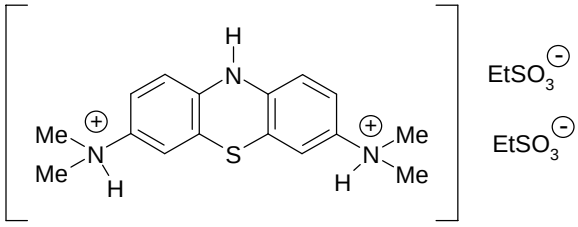
7 Use as claimed in claim 5 or claim 6 wherein the or each protic acid is selected from H_2CO_3; CH_3COOH; methanesulfonic acid, 1,2-ethanedisulfonic acid, ethansulfonic acid, naphthalenedisulfonic acid, p-toluenesulfonic acid.

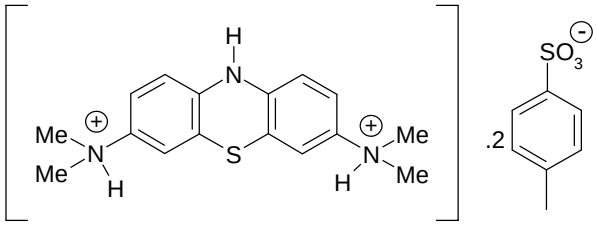
8 Use as claimed in claim 7 wherein the compound is LMTM:

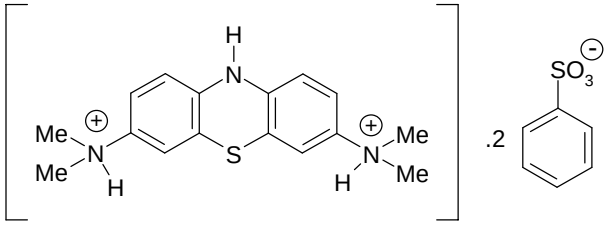


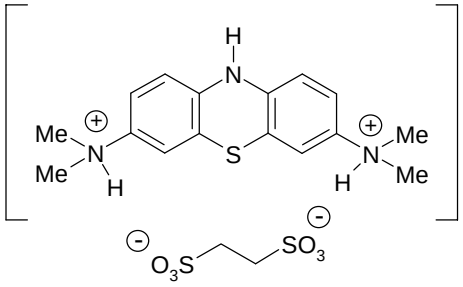
9 Use as claimed in claim 8 wherein the total daily dose of LMTM is around 0.8 to 33 mg/day, more preferably 6 to 12 mg/day of LMTM total.

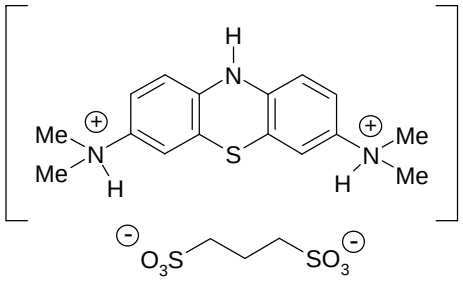
10 Use as claimed in claim 7 wherein the compound is selected from the list consisting of:

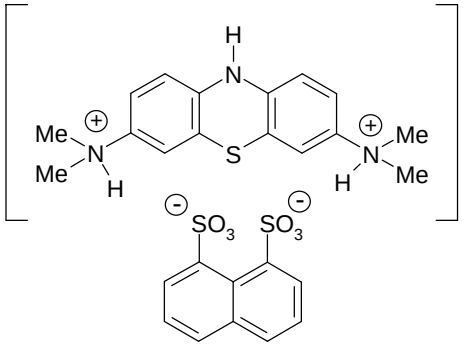












11 Use as claimed in any one of claims 1 to 10 wherein the subject has not historically received treatment with an acetylcholinesterase inhibitor or an N-methyl-D-aspartate receptor antagonist.

12 Use as claimed in any one of claims 1 to 10 wherein the subject has historically received treatment with an acetylcholinesterase inhibitor and\or an N-methyl-D-aspartate

receptor antagonist, but ceased that medication at least 1, 2, 3, 4, 5, 6, 7, or 8 weeks prior to treatment with the MT containing compound.

13 Use as claimed in any one of claims 1 to 10 wherein the subject is selected as one who is receiving treatment with an acetylcholinesterase inhibitor and\or an N-methyl-D-aspartate receptor antagonist,

wherein said treatment with the acetylcholinesterase inhibitor and\or an N-methyl-D-aspartate receptor antagonist is discontinued prior to treatment with the MT containing compound.

14. Use as claimed in any one of claims 1 to 10, wherein the medicament is to be administered once a day.