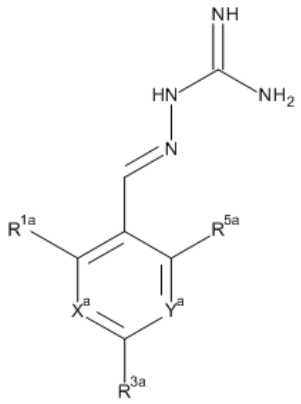
CLAIMS

1. A compound of formula IA:



(IA)

or a pharmaceutically acceptable salt thereof, wherein:

X^a is CR^{2a};

Y^a is CR^{4a};

R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} each independently represent H, F or Cl;

wherein:

two of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represent Cl, one of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represents F, and two of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represent H;

two of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represent F, one of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represents Cl, and two of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represent H; or

the compound is selected from:

2-(3,4,5-trichlorobenzylidene)hydrazine-1-carboximidamide;

2-(2,4,5-trichlorobenzylidene)hydrazine-1-carboximidamide;

2-(2,3,4-trichlorobenzylidene)hydrazine-1-carboximidamide,

or a pharmaceutically acceptable salt of any one thereof.

2. The compound according to claim 1, wherein R^{1a} and R^{3a} both represent Cl.

3. The compound according to claim 1, wherein R^{1a} is F or Cl.

4. The compound according to any preceding claim, wherein the compound is in the E-isomer form.

5. The compound according to claim 1, wherein the compound is selected from

(E)-2-(3,4,5-trichlorobenzylidene)hydrazine-1-carboximidamide,
(E)-2-(2,4,5-trichlorobenzylidene)hydrazine-1-carboximidamide,
(E)-2-(3,5-dichloro-4-fluorobenzylidene)hydrazine-1-carboximidamide,
(E)-2-(2,3,4-trichlorobenzylidene)hydrazine-1-carboximidamide,
(E)-2-(2,4-dichloro-5-fluorobenzylidene)hydrazine-1-carboximidamide,
(E)-2-(2,4-dichloro-3-fluorobenzylidene)hydrazine-1-carboximidamide,
(E)-2-(2,3-dichloro-4-fluorobenzylidene)hydrazine-1- carboximidamide,
(E)-2-(4,5-dichloro-2-fluorobenzylidene)hydrazine-1-carboximidamide,
(E)-2-(2-chloro-3,4-difluorobenzylidene)hydrazine-1-carboximidamide,
(E)-2-(2-chloro-3,5-difluorobenzylidene)hydrazine-1-carboximidamide,
(E)-2-(2-chloro-3,5-difluorobenzylidene)hydrazine-1-carboximidamide,
or a pharmaceutically acceptable salt of any one thereof.

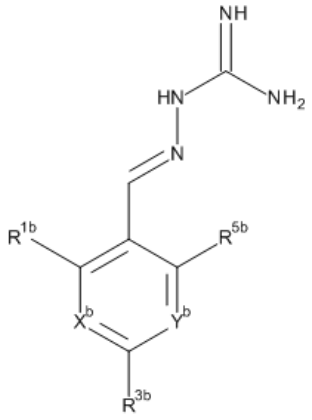
6. The compound according to claim 5, which is (E)-2-(3,4,5-trichlorobenzylidene)hydrazine-1-carboximidamide or a pharmaceutically acceptable salt thereof.

7. The compound according to claim 5, which is (E)-2-(2,3,4-trichlorobenzylidene)hydrazine-1-carboximidamide or a pharmaceutically acceptable salt thereof.

8. The compound according to claim 5, which is (E)-2-(2,3-dichloro-4-fluorobenzylidene)hydrazine-1- carboximidamide or a pharmaceutically acceptable salt thereof.

9. A pharmaceutical composition comprising a compound according to any one of claims 1 to 8, in combination with a pharmaceutically acceptable excipient.

10. A compound of formula IA:



(IA)

or a pharmaceutically acceptable salt thereof, for use in therapy wherein:

X^a is CR^{2a};

Y^a is CR^{4a};

R^{1a}, R^{2a} , R^{3a}, R^{4a} and R^{5a} each independently represent H, F or Cl;

wherein:

two of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represent Cl, one of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represents F, and two of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represent H;

two of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represent F, one of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represents Cl, and two of R^{1a}, R^{2a}, R^{3a}, R^{4a} and R^{5a} represent H; or

the compound is selected from:

2-(3,4,5-trichlorobenzylidene)hydrazine-1-carboximidamide;

2-(2,4,5-trichlorobenzylidene)hydrazine-1-carboximidamide;

2-(2,3,4-trichlorobenzylidene)hydrazine-1-carboximidamide,

or a pharmaceutically acceptable salt of any one thereof.

11. A pharmaceutical composition comprising a compound according to claim 10 in combination with a pharmaceutically acceptable excipient, for use in therapy.

12. Use of the compound according to claim 10 or pharmaceutical composition according to claim 11, in the preparation of a medicament for the treatment of a disorder associated with accumulation of misfolded proteins or a proteostasis disorder.

13. Use of the compound or pharmaceutical composition according to claim 12, wherein the disorder is selected from Alzheimer’s disease, Parkinson’s disease, Huntington’s disease, other polyglutamine disorders and tauopathies.

14. The compound or pharmaceutical composition for use according to any one of claims 10 to 13 in combination with one or more other active agents.