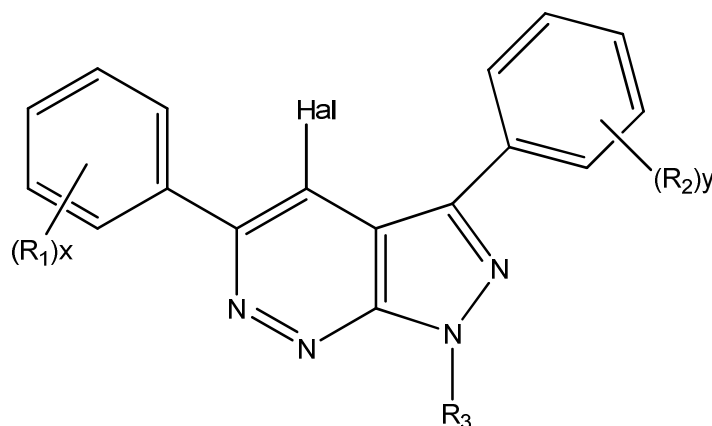


CLAIMS

1. An effective amount of a compound of Formula II:



Formula II

or a pharmaceutically acceptable salt thereof,

wherein Hal is -Cl, -F, -I, or -Br;

x is an integer ranging from 0 to 5;

each R₁ is independently -Cl, -F, -I, -Br, -C₁-C₃ alkyl, -O- C₁-C₃ alkyl, -CN, -CF₃, -C(O)NH(CH₃), or -C≡CCH₂OH;

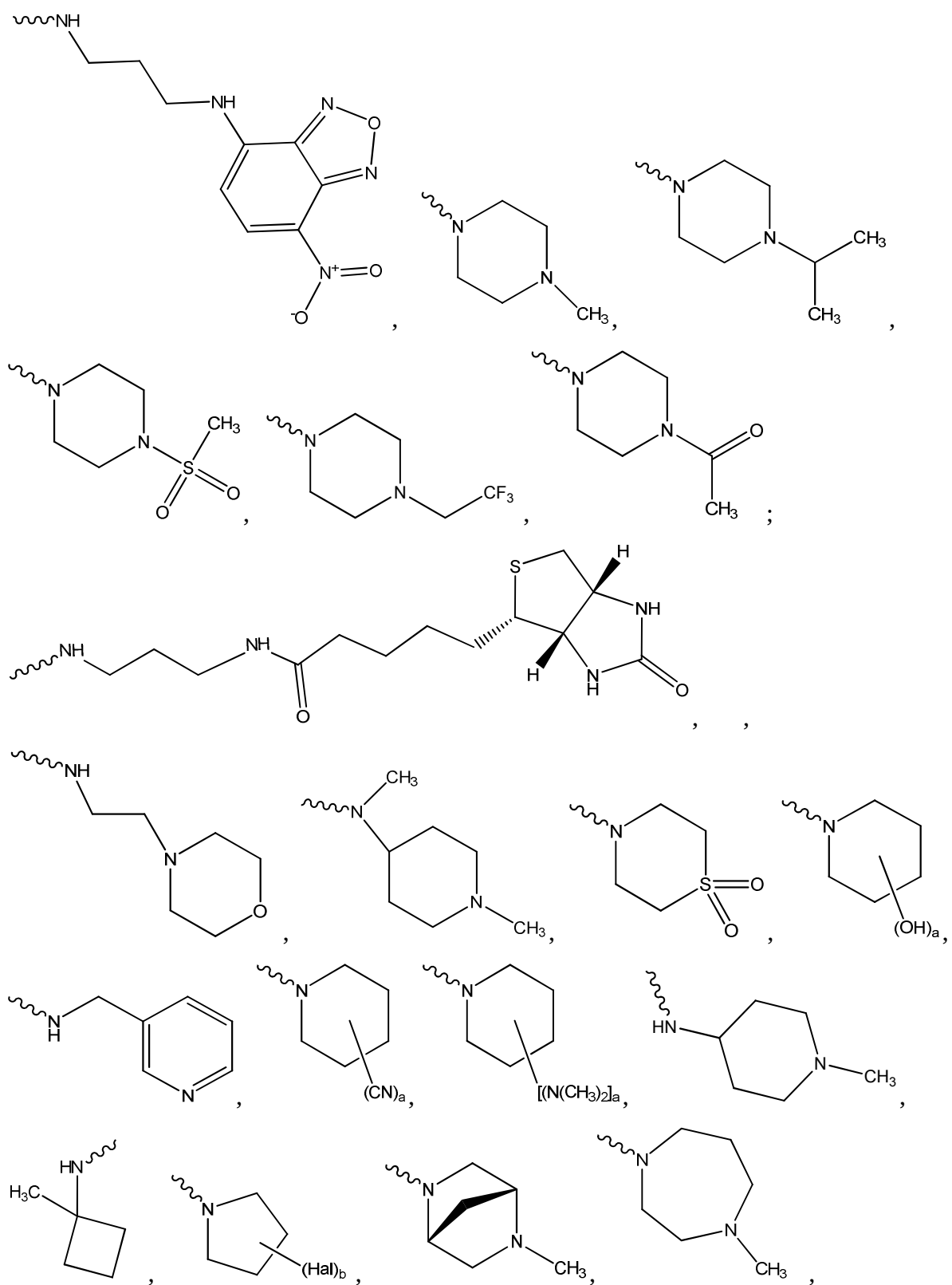
y is an integer ranging from 0 to 5;

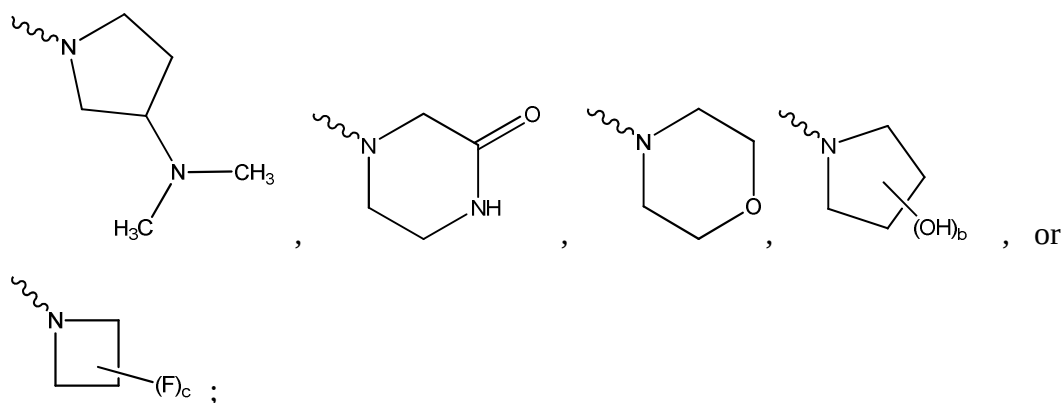
each R₂ is independently -Cl, -F, -Br, -C₁-C₃ alkyl, -O- C₁-C₃ alkyl, -CN, -CF₃, -C(O)NH(CH₃), or -C≡CCH₂OH;

R₃ is -H, -C₁-C₆ alkyl, -(C₁-C₆ alkylene)-OH, -(C₁-C₆ alkylene)-phenyl, -(C₁-C₆ alkylene)-O-(C₁-C₆ alkyl), -C₂-C₆ alkenyl, -(C₁-C₆ alkylene)-C(O)R₄,

-(C₁-C₆ alkylene)-R₅,  ;

R₄ is -OH, -O-(C₁-C₆ alkyl), -NH₂, -NH(C₁-C₆ alkyl), -NH((C₁-C₆ alkylene)-OH), -NH((C₁-C₆ alkylene)N(C₁-C₆ alkyl)₂), -N(C₁-C₆ alkyl)((C₁-C₆ alkylene)-CN), -N(C₁-C₆ alkyl)((C₁-C₆ alkylene)N(C₁-C₆ alkyl)₂), -NH(C₁-C₆ alkylene)-O-(C₁-C₆ alkyl),

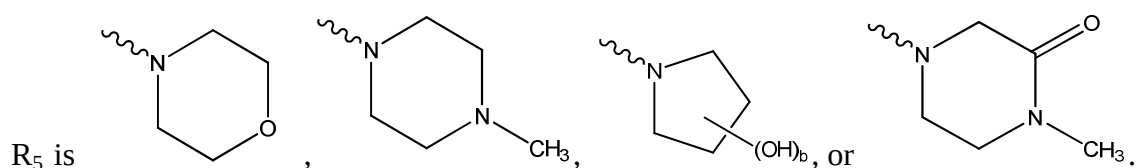




a is an integer ranging from 0 to 10;

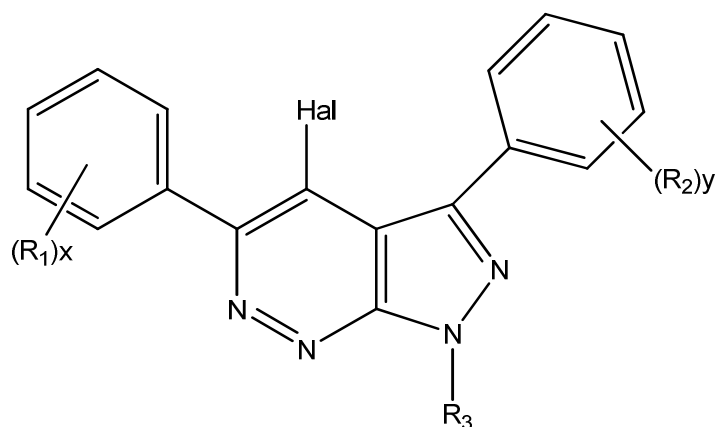
b is an integer ranging from 0 to 8;

c is an integer ranging from 0 to 6; and



for use in the treatment or prevention of a proteopathy.

2. Use of an effective amount of a compound of Formula II:



Formula II

or a pharmaceutically acceptable salt thereof,

wherein Hal is -Cl, -F, -I, or -Br;

x is an integer ranging from 0 to 5;

each R_1 is independently -Cl, -F, -I, -Br, -C₁-C₃ alkyl, -O- C₁-C₃ alkyl, -CN, -CF₃, -C(O)NH(CH₃), or -C≡CCH₂OH;

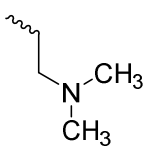
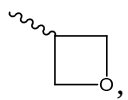
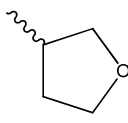
y is an integer ranging from 0 to 5;

each R_2 is independently -Cl, -F, -Br, -C₁-C₃ alkyl, -O- C₁-C₃ alkyl, -CN, -CF₃,

$-\text{C}(\text{O})\text{NH}(\text{CH}_3)$, or $-\text{C}\equiv\text{CCH}_2\text{OH}$;

R_3 is $-\text{H}$, $-\text{C}_1\text{-C}_6$ alkyl, $-(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-OH}$, $-(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-phenyl}$,

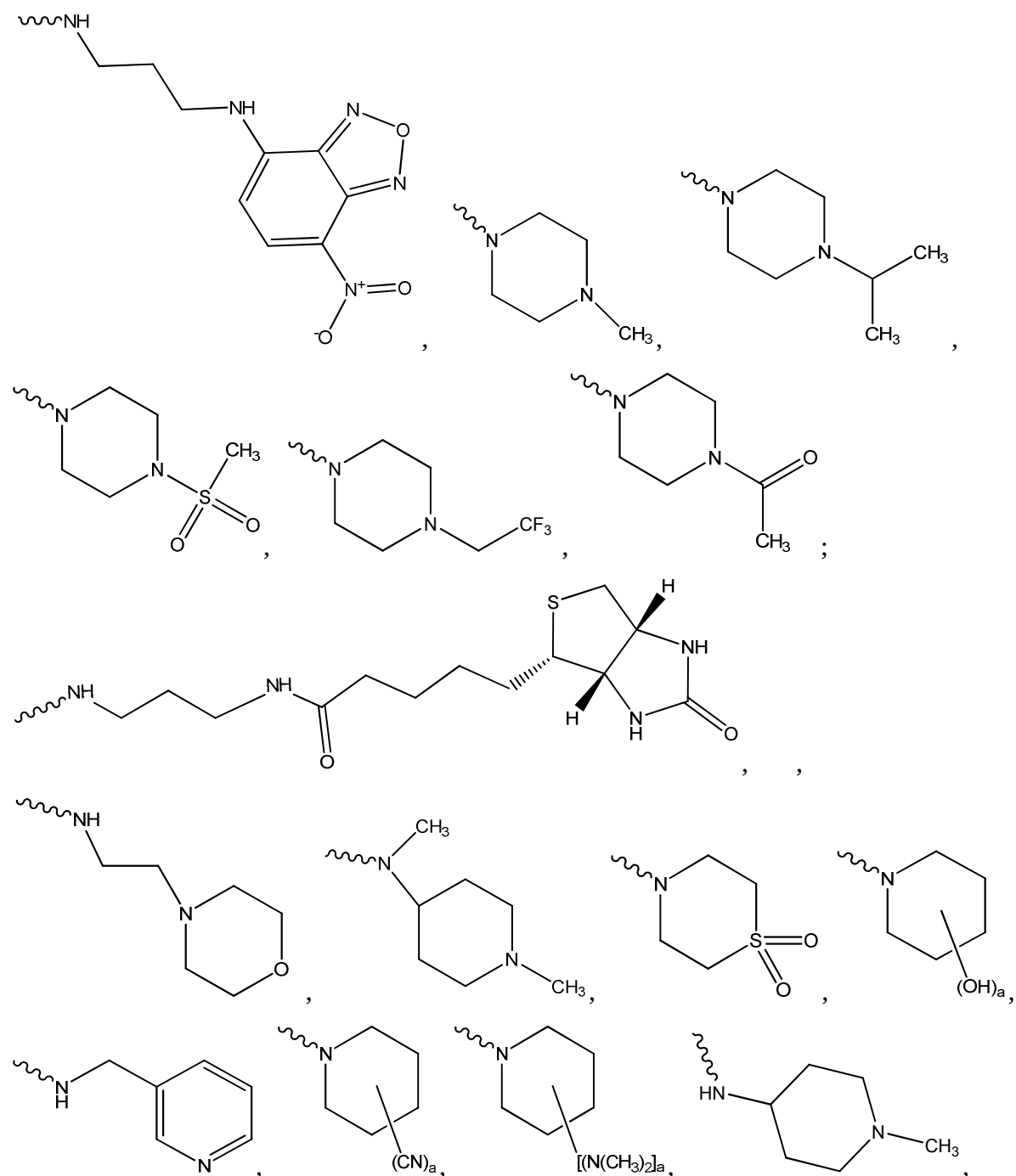
$-(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-O}(\text{C}_1\text{-C}_6 \text{ alkyl})$, $-\text{C}_2\text{-C}_6$ alkenyl, $-(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-C}(\text{O})\text{R}_4$,

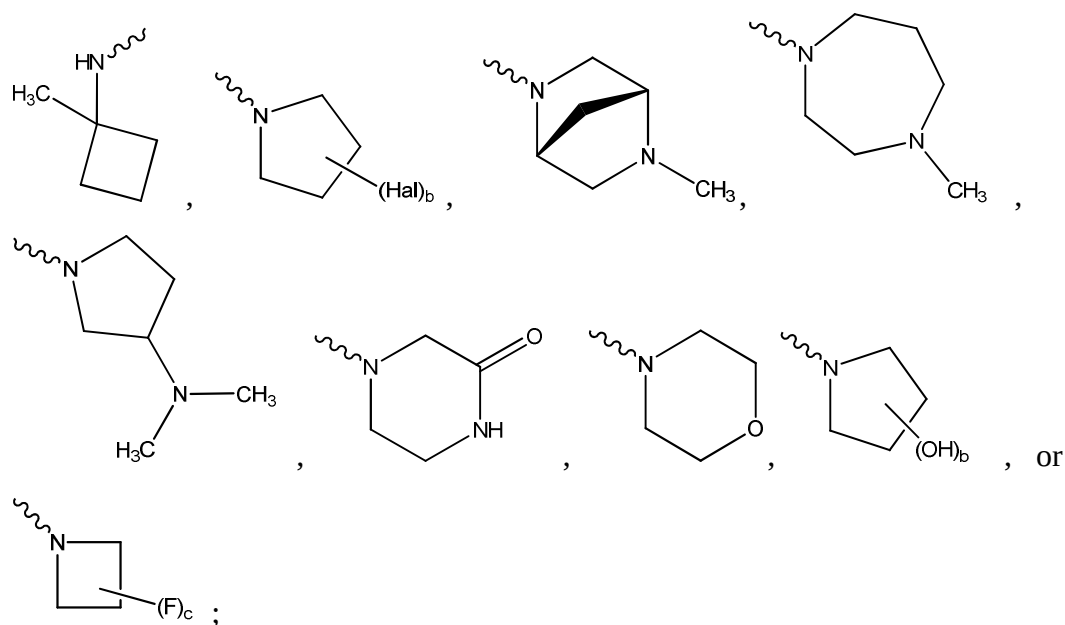
$-(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-R}_5$, , , or  ;

R_4 is $-\text{OH}$, $-\text{O}(\text{C}_1\text{-C}_6 \text{ alkyl})$, $-\text{NH}_2$, $-\text{NH}(\text{C}_1\text{-C}_6 \text{ alkyl})$, $-\text{NH}((\text{C}_1\text{-C}_6 \text{ alkylene})\text{-OH})$,

$-\text{NH}((\text{C}_1\text{-C}_6 \text{ alkylene})\text{N}(\text{C}_1\text{-C}_6 \text{ alkyl})_2)$, $-\text{N}(\text{C}_1\text{-C}_6 \text{ alkyl})((\text{C}_1\text{-C}_6 \text{ alkylene})\text{-CN})$,

$-\text{N}(\text{C}_1\text{-C}_6 \text{ alkyl})((\text{C}_1\text{-C}_6 \text{ alkylene})\text{N}(\text{C}_1\text{-C}_6 \text{ alkyl})_2)$, $-\text{NH}(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-O}(\text{C}_1\text{-C}_6 \text{ alkyl})$,

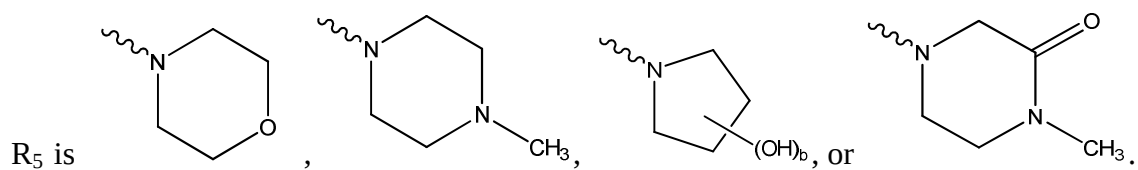




a is an integer ranging from 0 to 10;

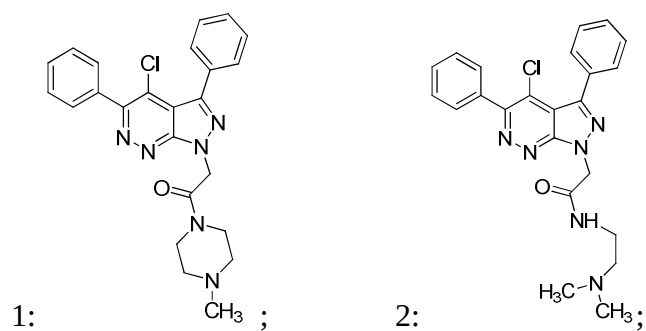
b is an integer ranging from 0 to 8;

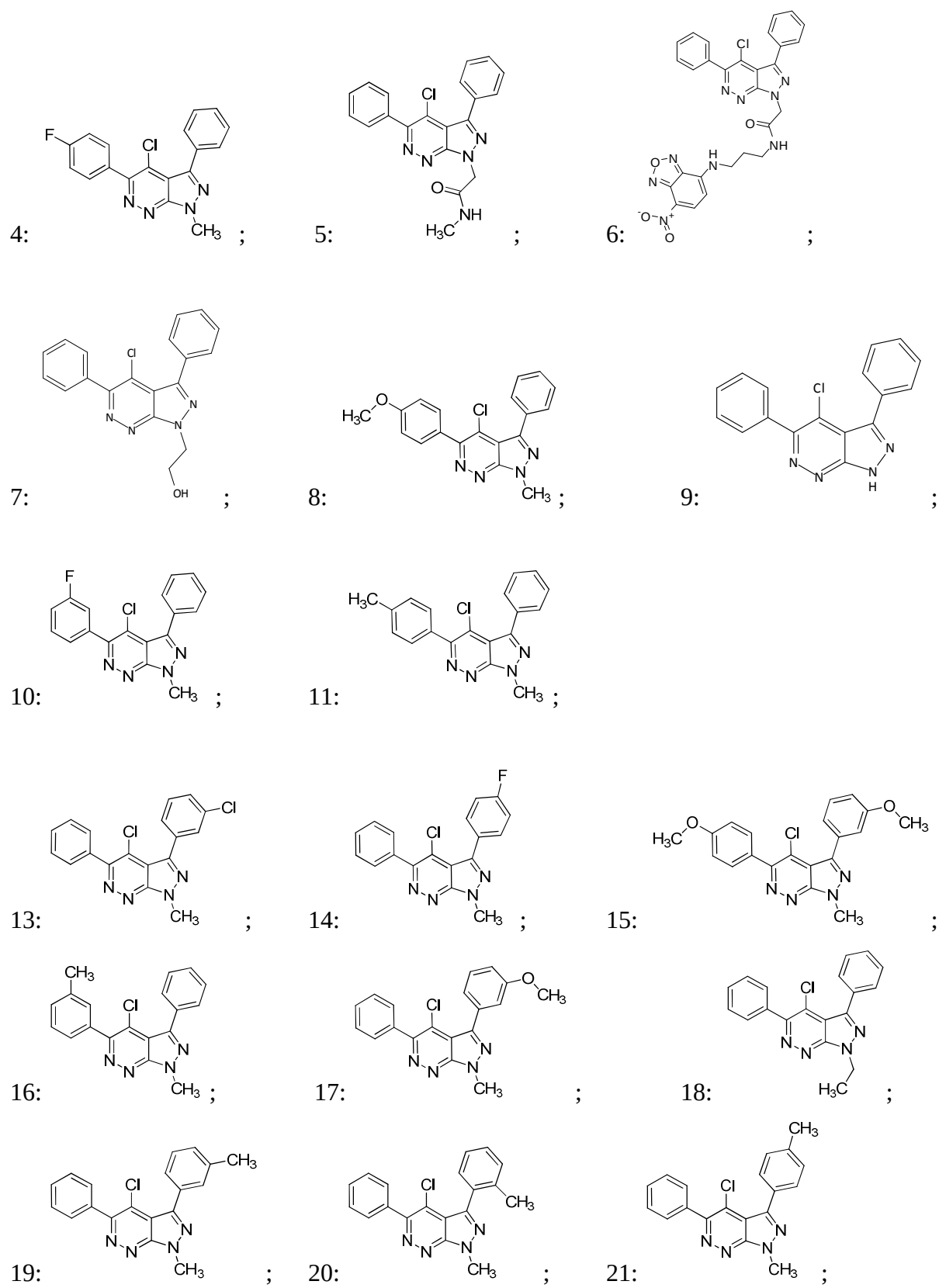
c is an integer ranging from 0 to 6; and

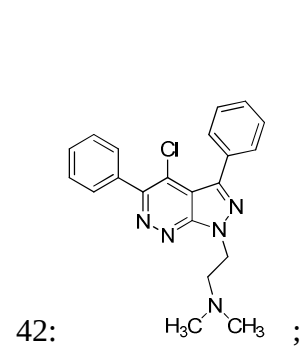
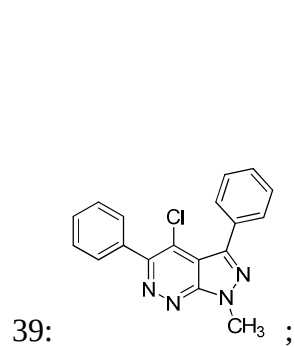
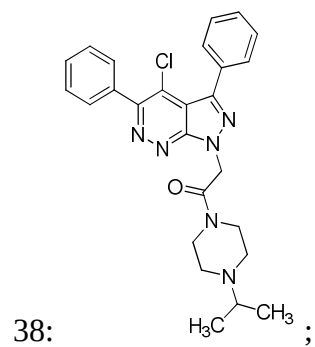
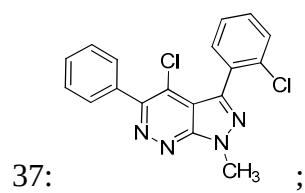
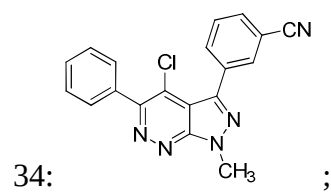
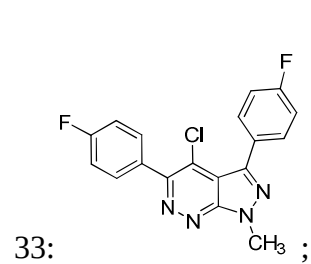
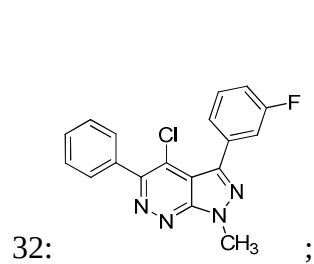
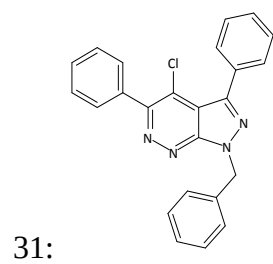
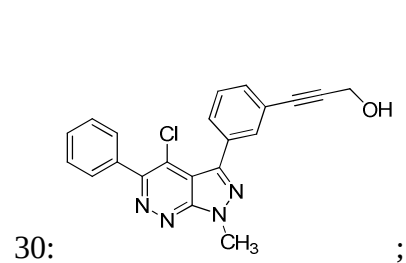
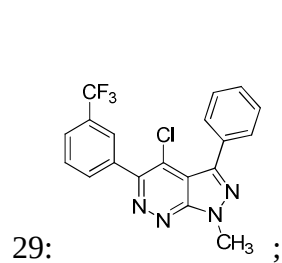
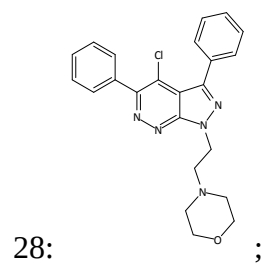
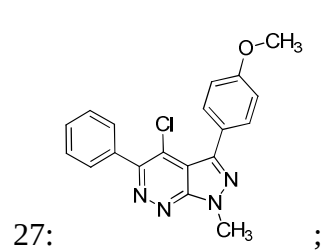
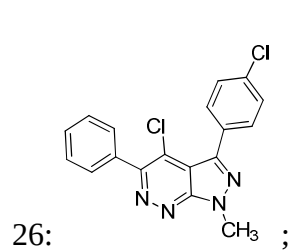
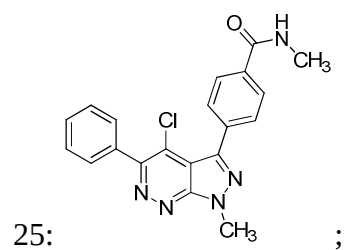
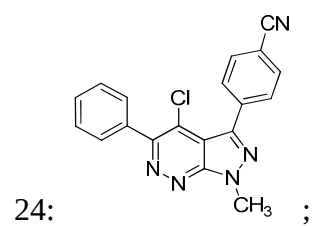
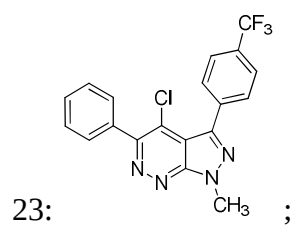
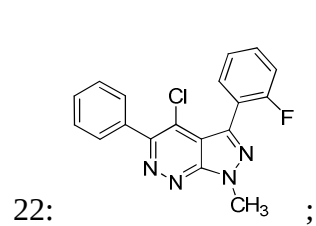


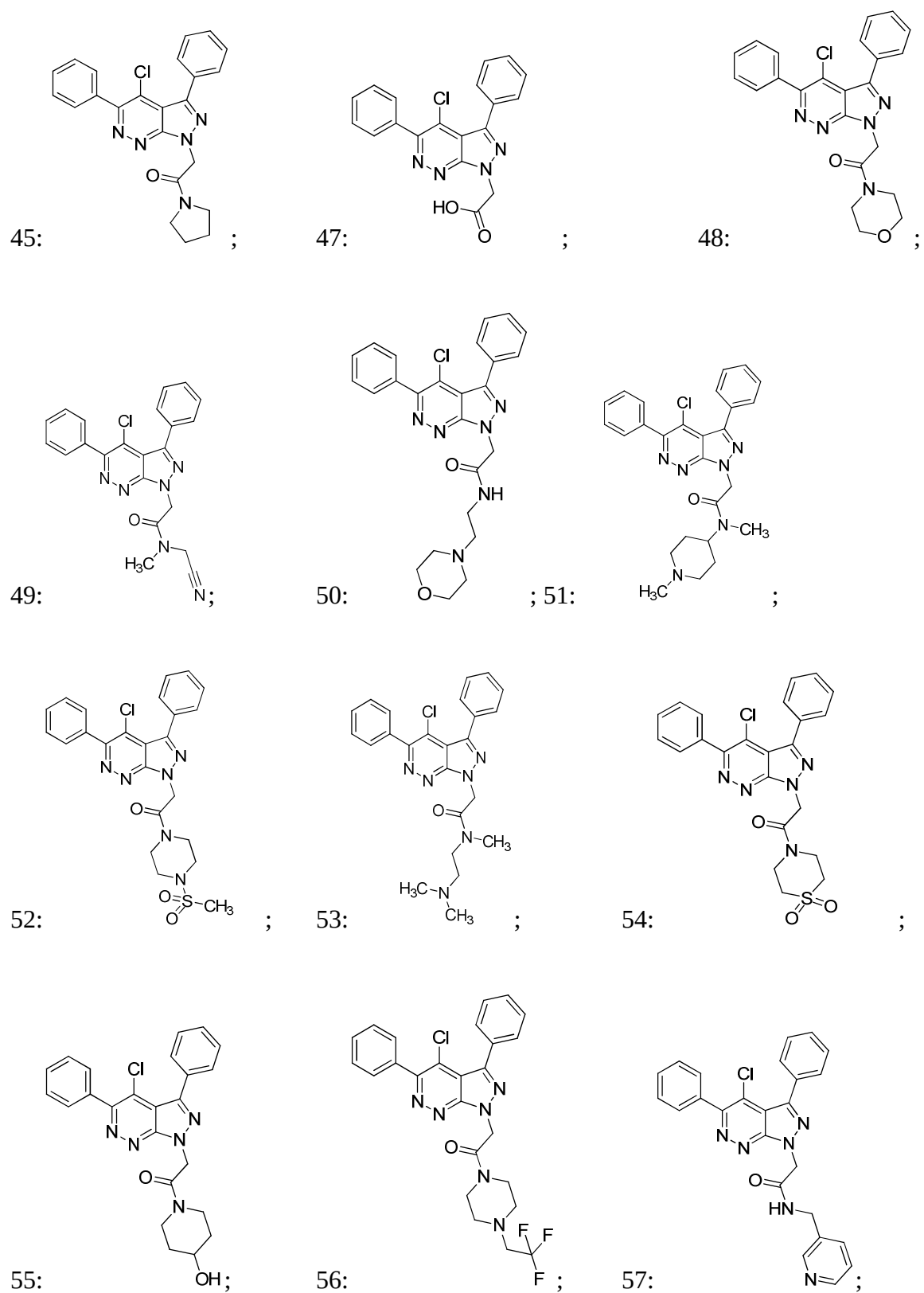
in the manufacture of a medicament for the treatment or prevention of a proteopathy .

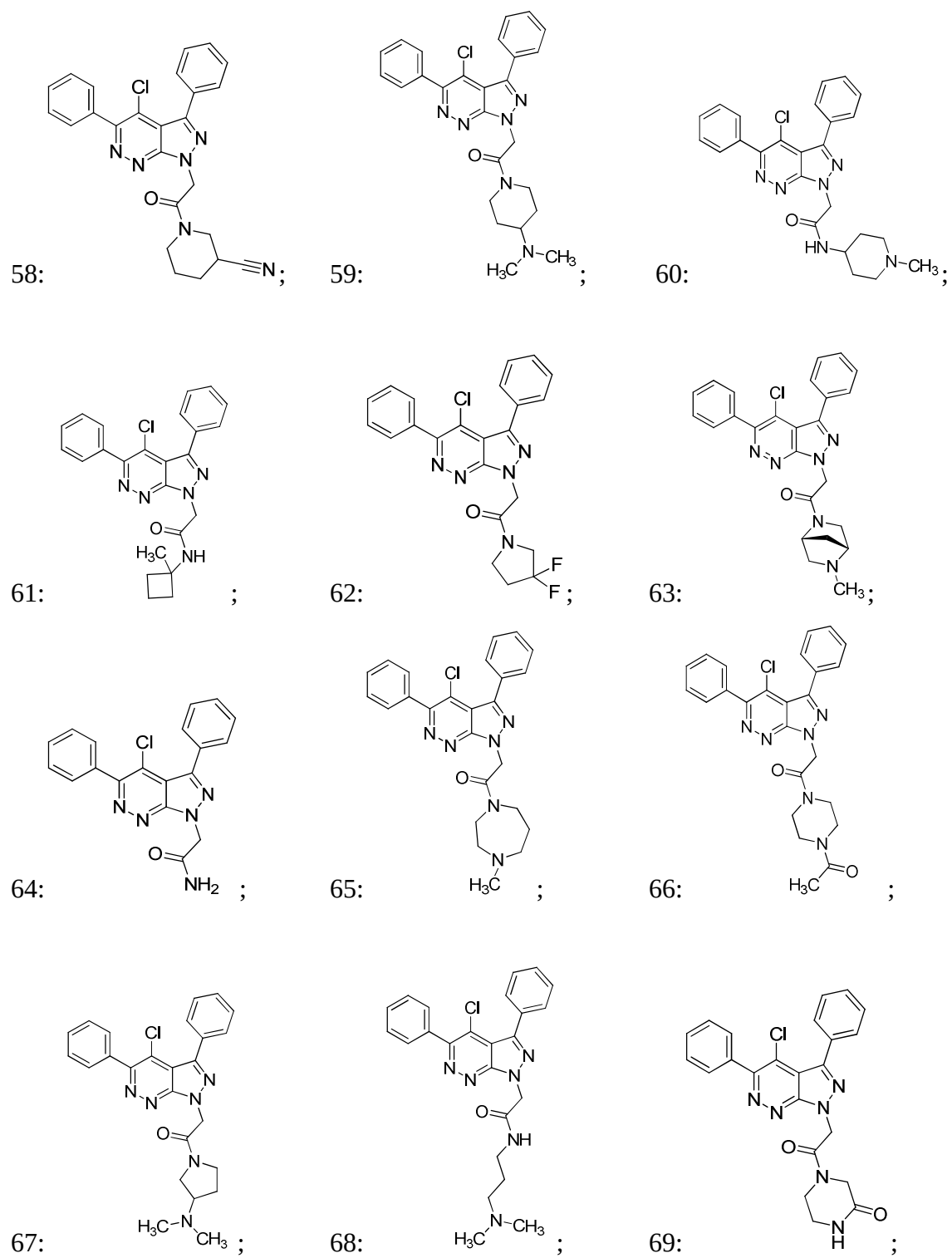
3. The use of a compound or pharmaceutically acceptable salt of claim 1 or claim 2, wherein the compound has the structure:

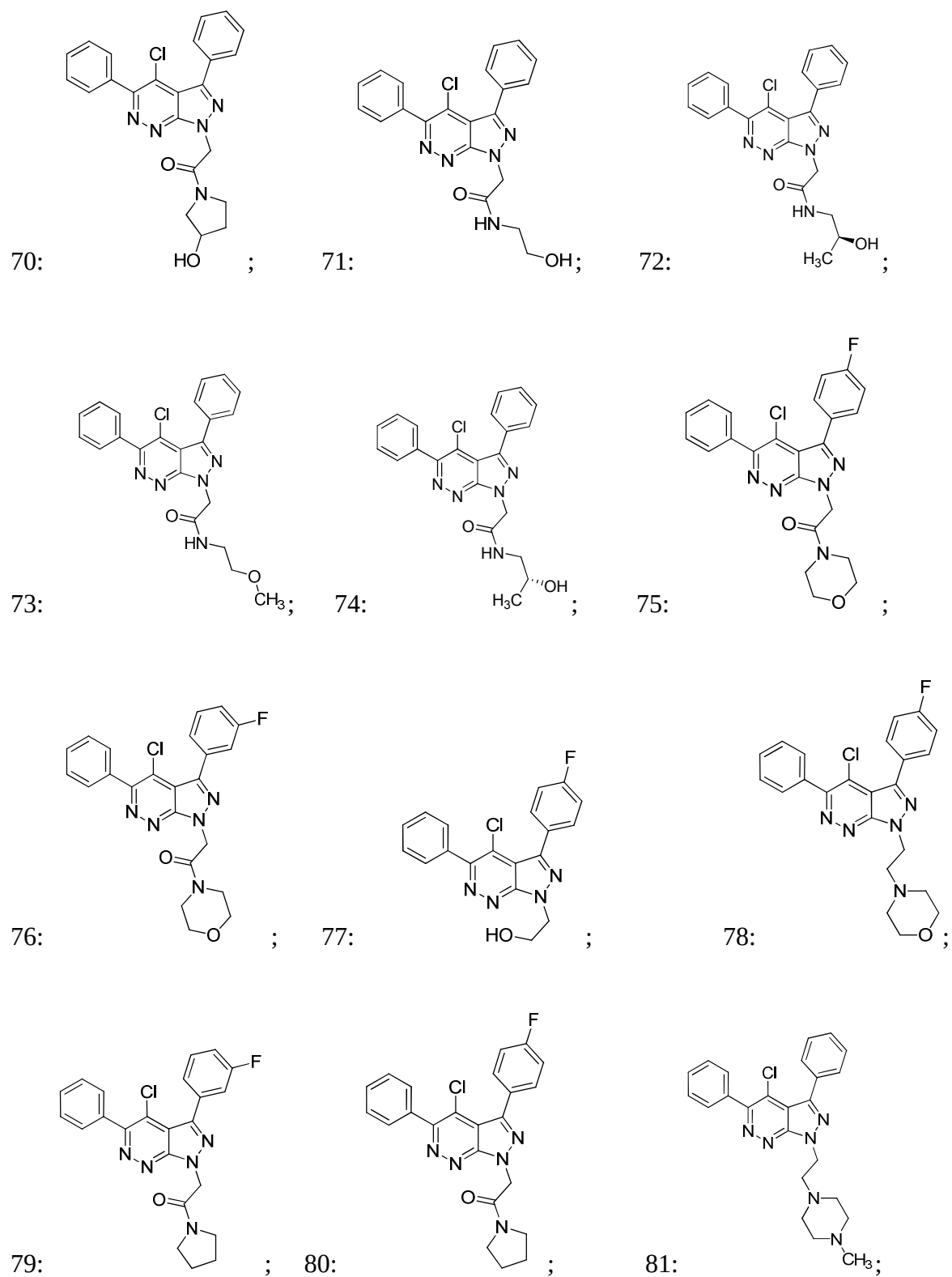


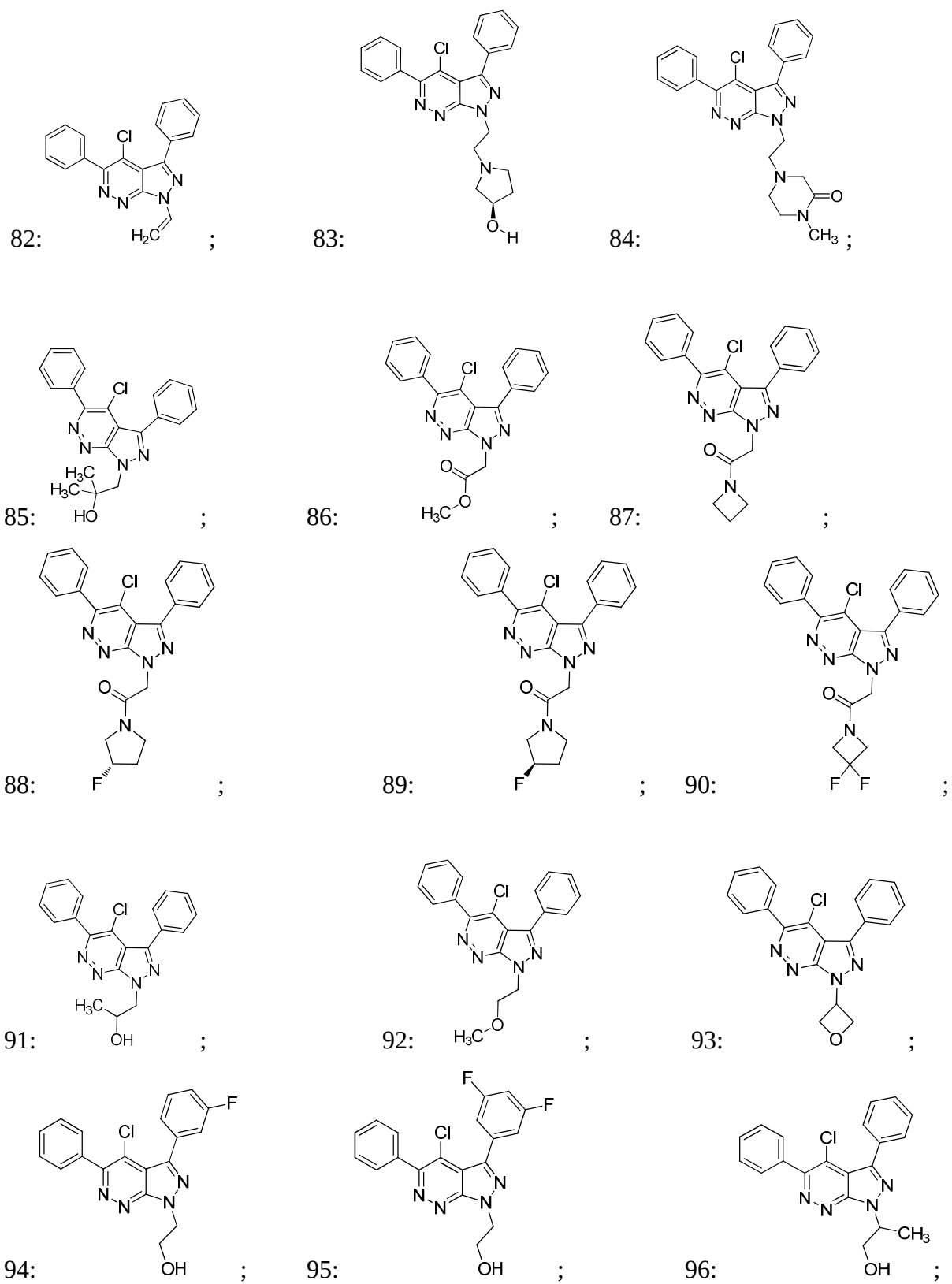


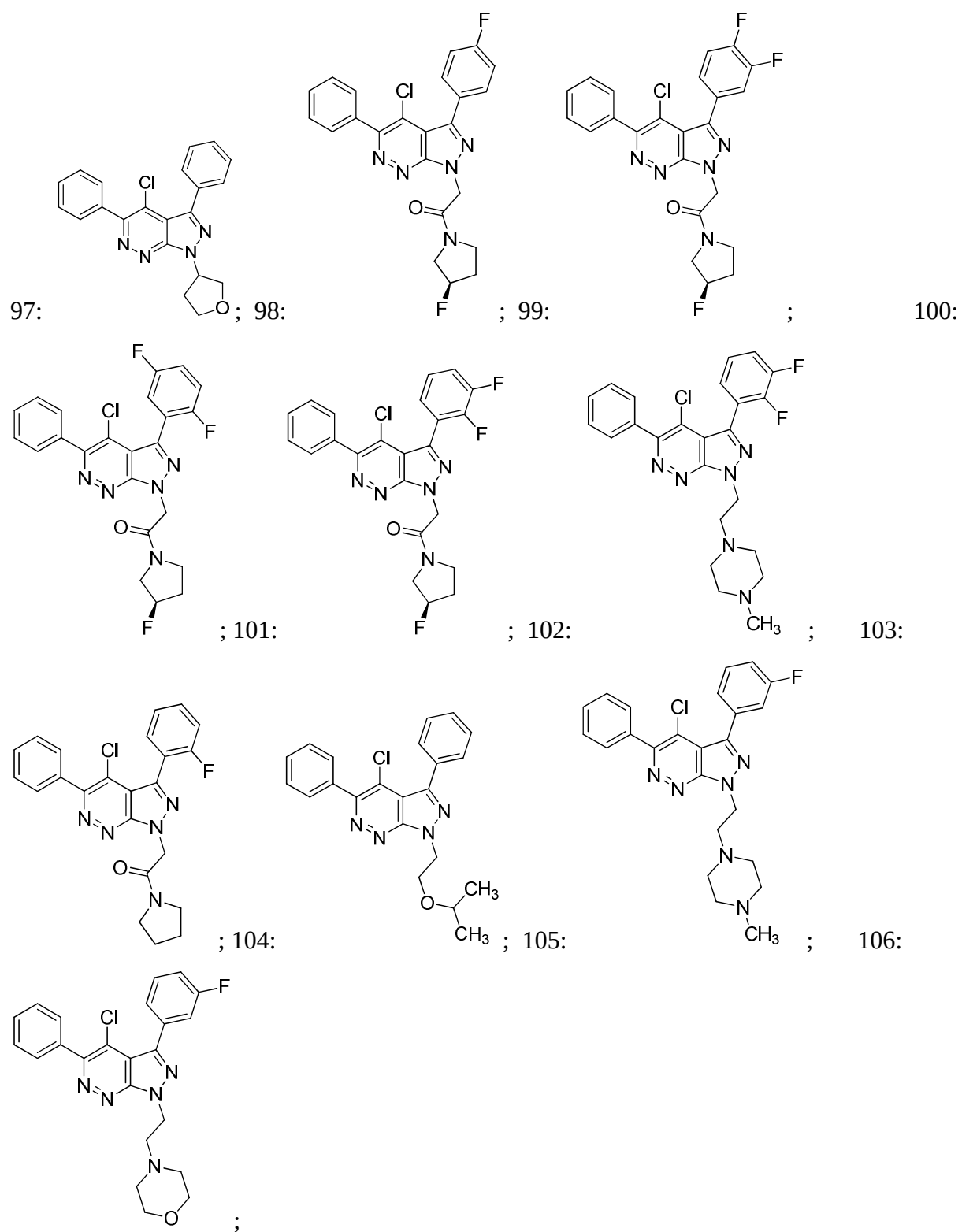


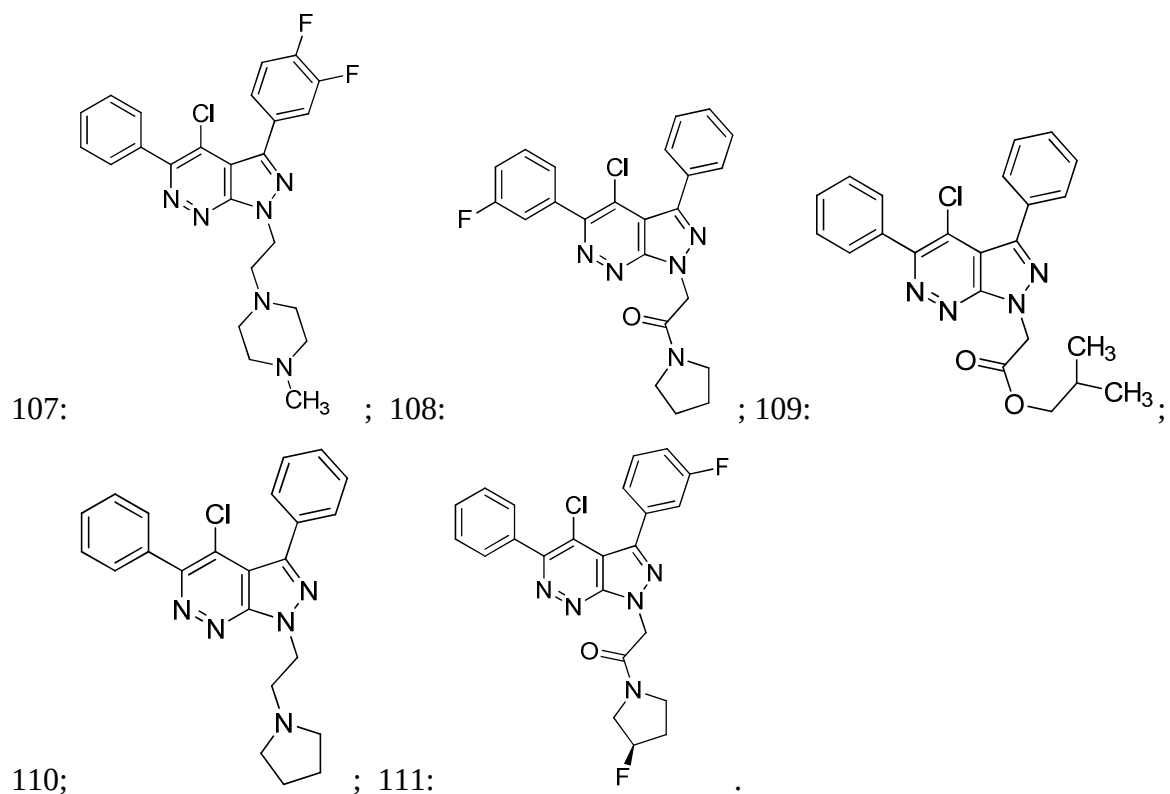




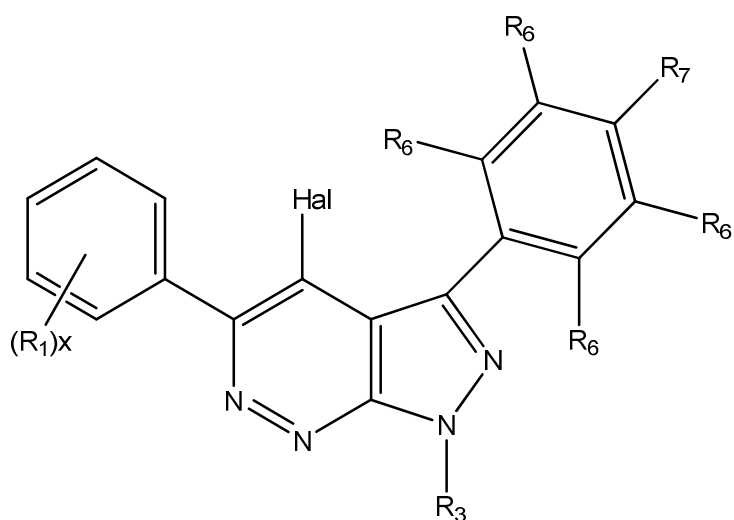








4. An effective amount of a compound of Formula III:



Formula III

or a pharmaceutically acceptable salt thereof,

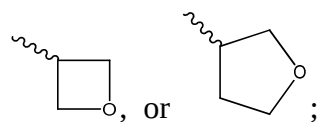
wherein Hal is -Cl, -F, -I, or -Br;

x is an integer ranging from 0 to 5;

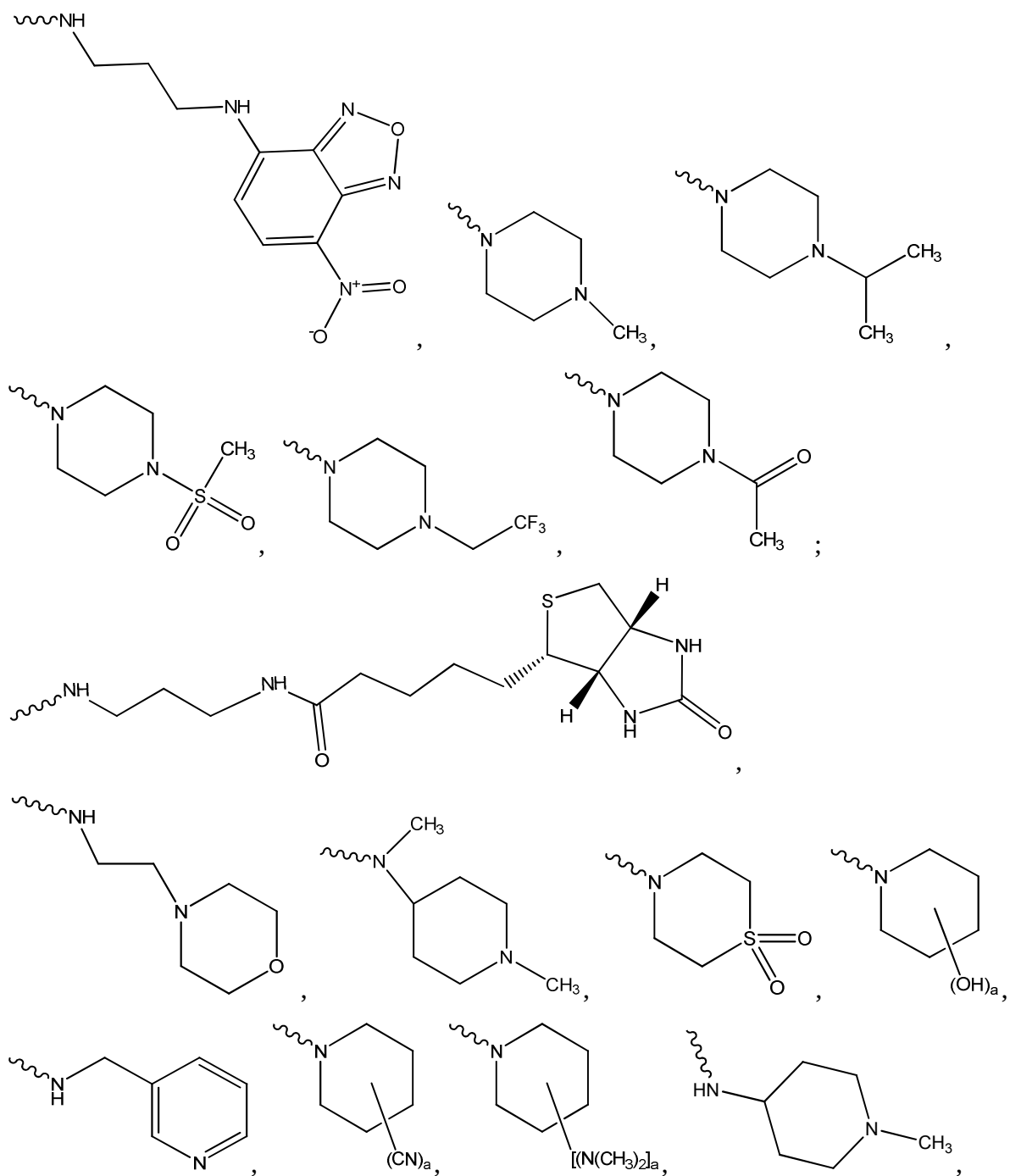
each R₁ is independently -Cl, -F, -I, -Br, -C₁-C₃ alkyl, -O- C₁-C₃ alkyl, -CN, -CF₃, -C(O)NH(CH₃), or -C≡CCH₂OH;

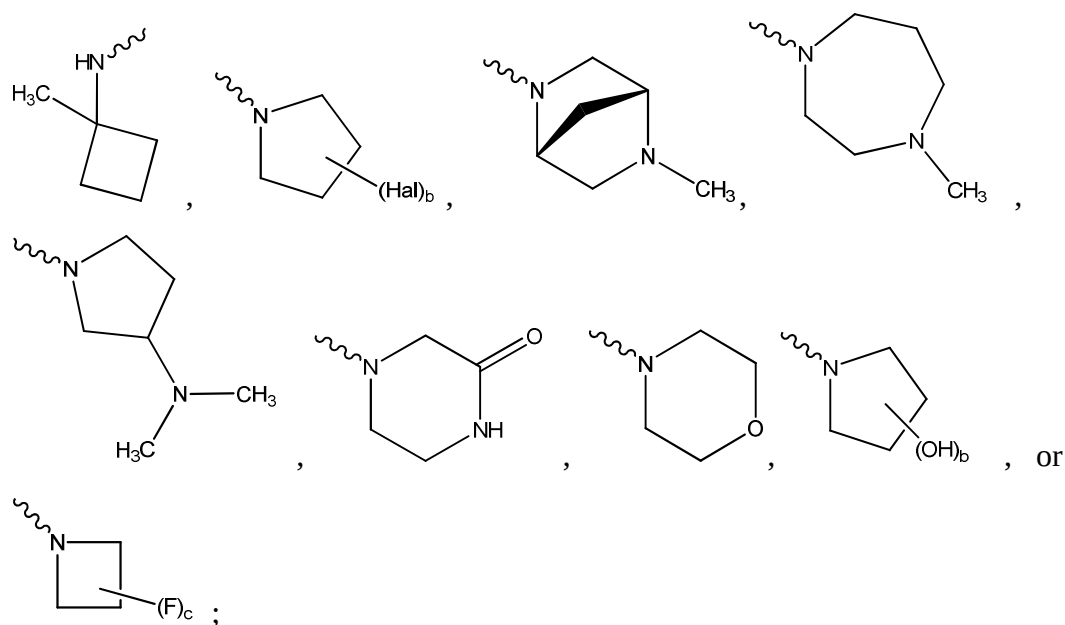
R₃ is -H, -C₁-C₆ alkyl, -(C₁-C₆ alkylene)-OH, -(C₁-C₆ alkylene)-phenyl, -(C₁-C₆

alkylene)-O-(C₁-C₆ alkyl), -C₂-C₆ alkenyl, -(C₁-C₆ alkylene)-C(O)R₄, -(C₁-C₆ alkylene)-R₅,



R₄ is -OH, -O-(C₁-C₆ alkyl), -NH₂, -NH(C₁-C₆ alkyl), -NH((C₁-C₆ alkylene)-OH),
-NH((C₁-C₆ alkylene)N(C₁-C₆ alkyl)₂), -N(C₁-C₆ alkyl)((C₁-C₆ alkylene)-CN), -N(C₁-C₆
alkyl)((C₁-C₆ alkylene)N(C₁-C₆ alkyl)₂), -NH(C₁-C₆ alkylene)-O-(C₁-C₆ alkyl),

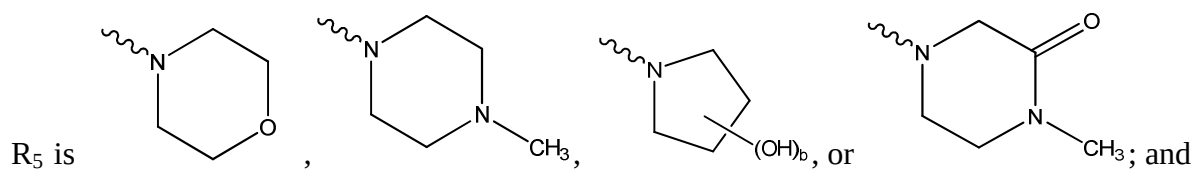




a is an integer ranging from 0 to 10;

b is an integer ranging from 0 to 8;

c is an integer ranging from 0 to 6;

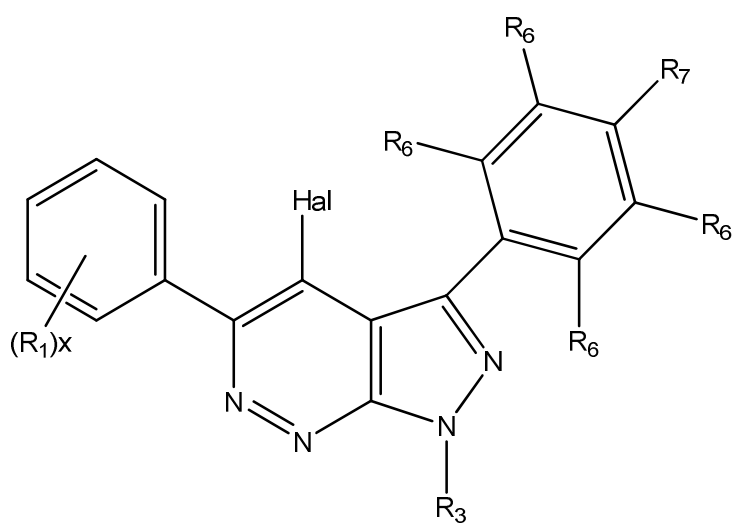


each R₆ and R₇ is independently -H or -I, wherein at least one of R₆ and R₇ is -I,

and wherein when R₃ is -C₁-C₃ alkyl, then R₇ is -H.

for use in the treatment or prevention of a proteopathy.

5. Use of an effective amount of a compound of Formula III:



Formula III

or a pharmaceutically acceptable salt thereof,

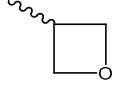
wherein Hal is $-\text{Cl}$, $-\text{F}$, $-\text{I}$, or $-\text{Br}$;

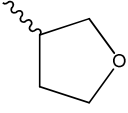
x is an integer ranging from 0 to 5;

each R_1 is independently $-\text{Cl}$, $-\text{F}$, $-\text{I}$, $-\text{Br}$, $-\text{C}_1\text{-C}_3$ alkyl, $-\text{O}-\text{C}_1\text{-C}_3$ alkyl, $-\text{CN}$, $-\text{CF}_3$,

$-\text{C}(\text{O})\text{NH}(\text{CH}_3)$, or $-\text{C}\equiv\text{CCH}_2\text{OH}$;

R_3 is $-\text{H}$, $-\text{C}_1\text{-C}_6$ alkyl, $-(\text{C}_1\text{-C}_6 \text{ alkylene})-\text{OH}$, $-(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-phenyl}$, $-(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-}$

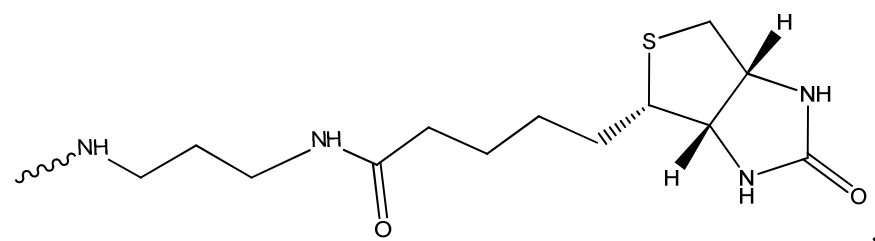
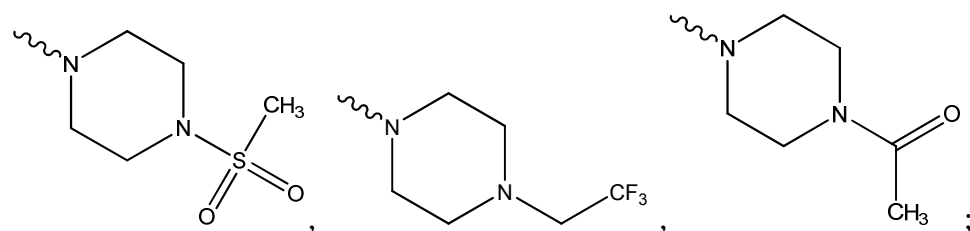
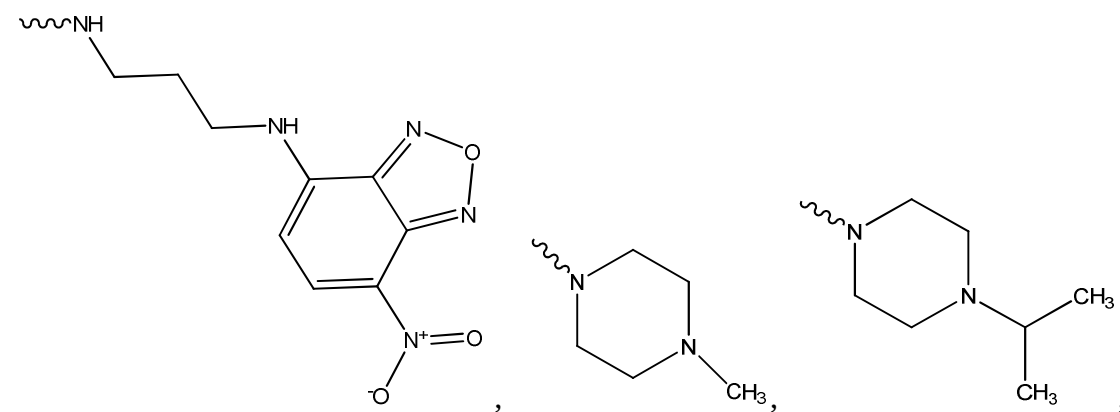
$\text{O}-(\text{C}_1\text{-C}_6 \text{ alkyl})$, $-\text{C}_2\text{-C}_6$ alkenyl, $-(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-C}(\text{O})\text{R}_4$, $-(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-R}_5$, ,

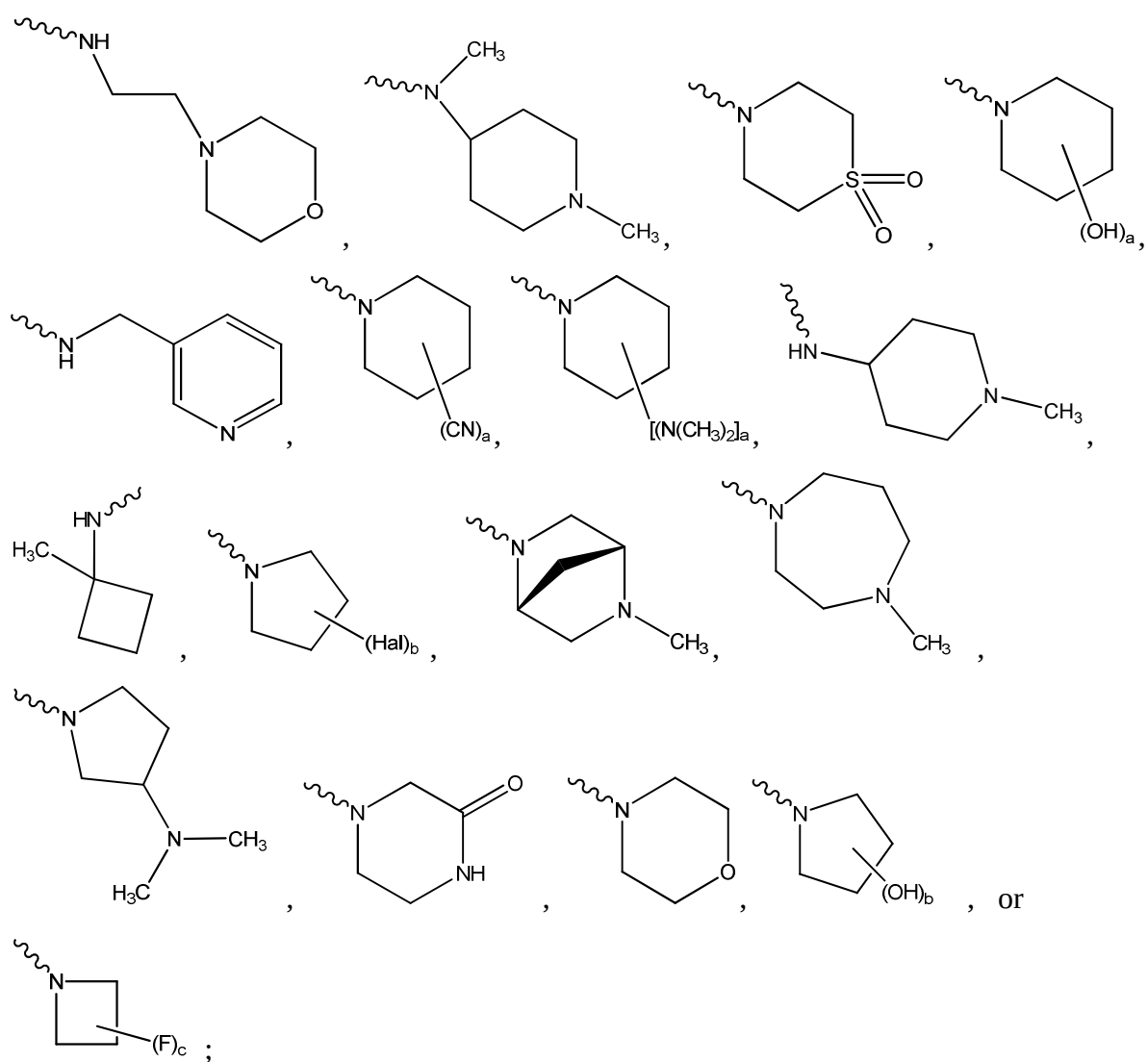
or  ;

R_4 is $-\text{OH}$, $-\text{O}-(\text{C}_1\text{-C}_6 \text{ alkyl})$, $-\text{NH}_2$, $-\text{NH}(\text{C}_1\text{-C}_6 \text{ alkyl})$, $-\text{NH}((\text{C}_1\text{-C}_6 \text{ alkylene})-\text{OH})$,

$-\text{NH}((\text{C}_1\text{-C}_6 \text{ alkylene})\text{N}(\text{C}_1\text{-C}_6 \text{ alkyl})_2)$, $-\text{N}(\text{C}_1\text{-C}_6 \text{ alkyl})((\text{C}_1\text{-C}_6 \text{ alkylene})-\text{CN})$, $-\text{N}(\text{C}_1\text{-C}_6$

$\text{alkyl})((\text{C}_1\text{-C}_6 \text{ alkylene})\text{N}(\text{C}_1\text{-C}_6 \text{ alkyl})_2)$, $-\text{NH}(\text{C}_1\text{-C}_6 \text{ alkylene})\text{-O}-(\text{C}_1\text{-C}_6 \text{ alkyl})$,

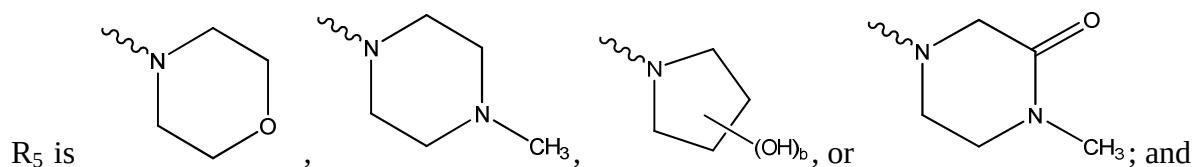




a is an integer ranging from 0 to 10;

b is an integer ranging from 0 to 8;

c is an integer ranging from 0 to 6;

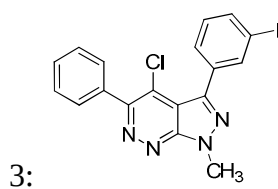


each R₆ and R₇ is independently -H or -I, wherein at least one of R₆ and R₇ is -I,

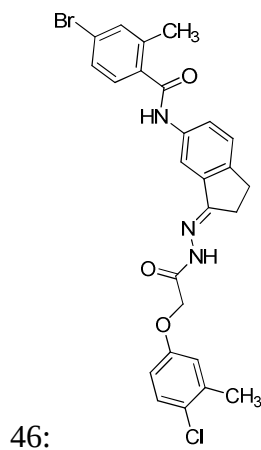
and wherein when R₃ is -C₁-C₃ alkyl, then R₇ is -H.

in the manufacture of a medicament for the treatment or prevention of a proteopathy.

6. The use of a compound or pharmaceutically acceptable salt of claim 4 or claim 5, wherein the compound has the structure:

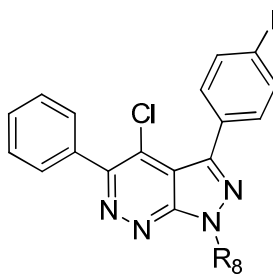


7. An effective amount of Compound 46 having the structure:



or a pharmaceutically acceptable salt thereof,
for use in the treatment or prevention of a proteopathy.

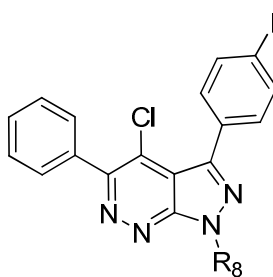
8. An effective amount of a compound of Formula IV:



Formula IV

or a pharmaceutically acceptable salt thereof,
for use in the treatment or prevention of a proteopathy.

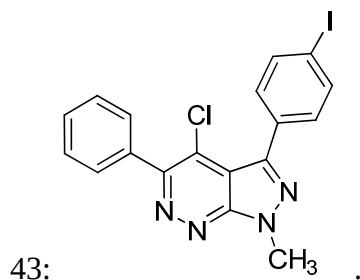
9. Use of an effective amount of a compound of Formula IV:



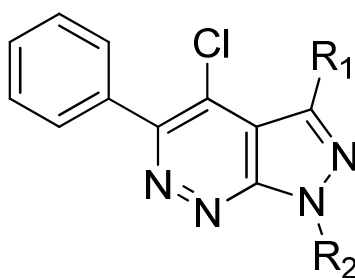
Formula IV

or a pharmaceutically acceptable salt thereof,
in the manufacture of a medicament for the treatment or prevention of a proteopathy.

10. The use of a compound or pharmaceutically acceptable salt of claim 8 or claim 9, wherein the compound has the structure:



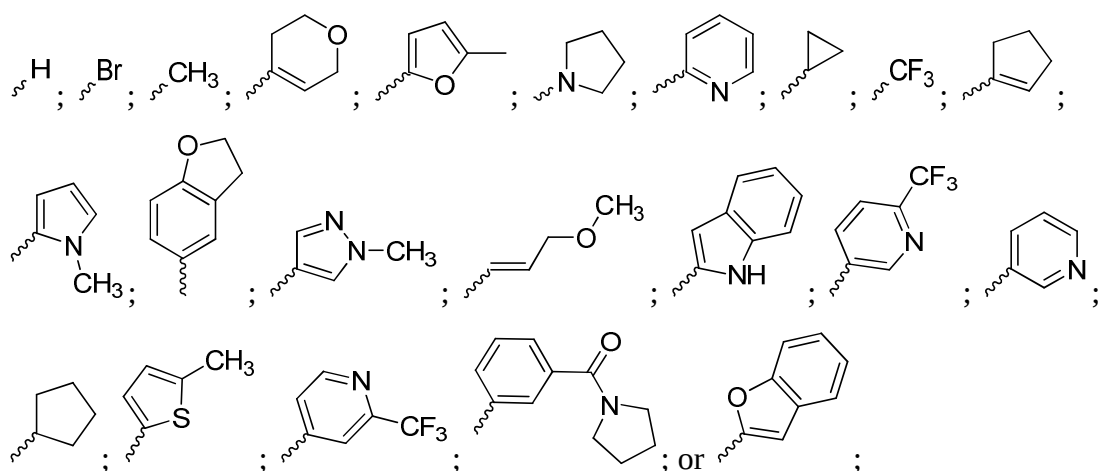
11. An effective amount of a compound of Formula V:



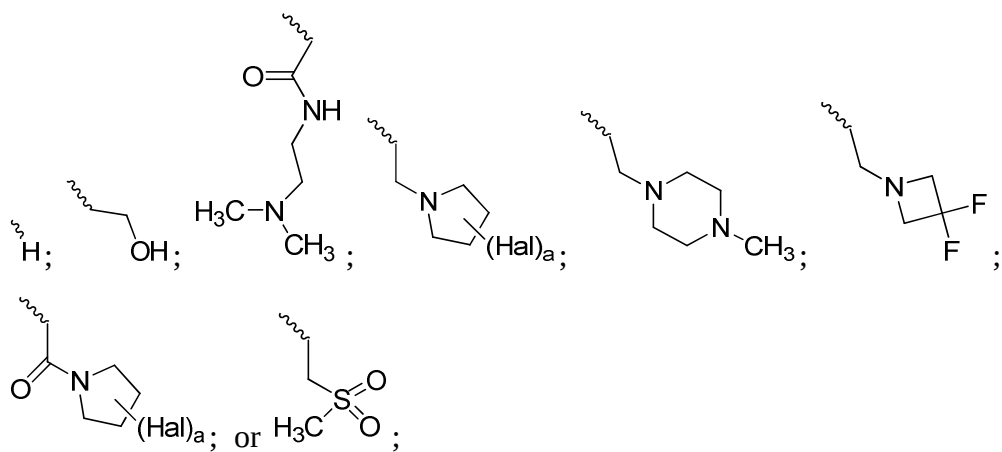
Formula V

or a pharmaceutically acceptable salt thereof,

wherein R₁ is:



R₂ is:

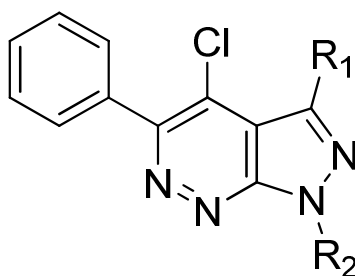


Hal is -Cl, -F, -I, or -Br;

and a is 0, 1, or 2,

for use in the treatment or prevention of a proteopathy.

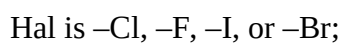
12. Use of an effective amount of a compound of Formula V:



Formula V

or a pharmaceutically acceptable salt thereof,

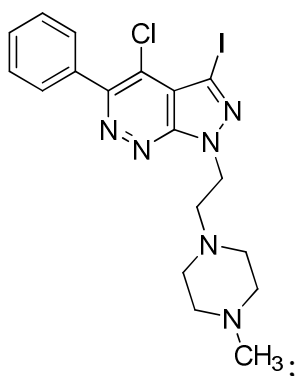
wherein R₁ is:



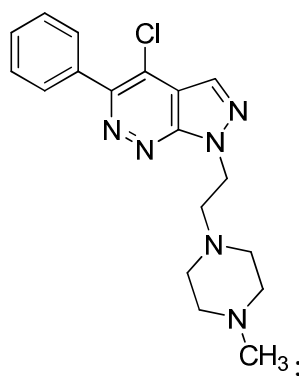
and a is 0, 1, or 2,

in the manufacture of a medicament for the treatment or prevention of a proteopathy.

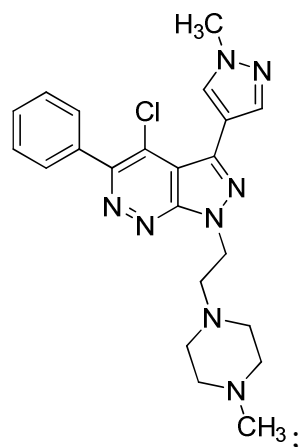
13. The use of a compound or pharmaceutically acceptable salt of claim 11 or claim 12, wherein the compound has the structure:



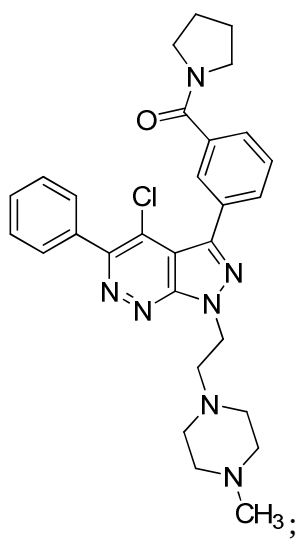
Va:



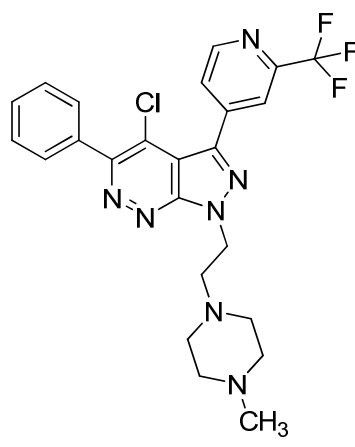
Vb:



Vc:

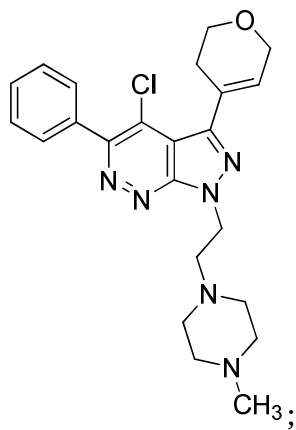


Vd:

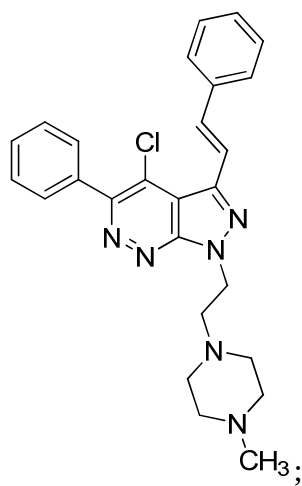


Ve:

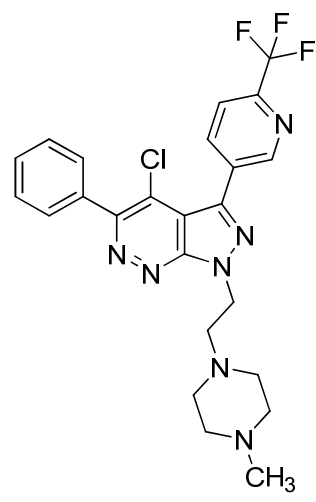
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Vf:

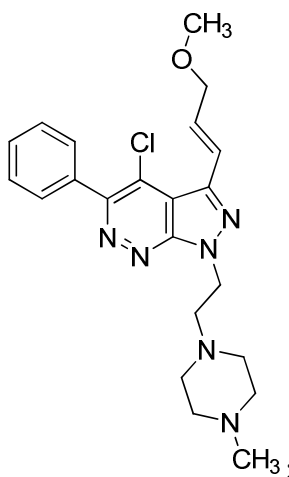


Vg:

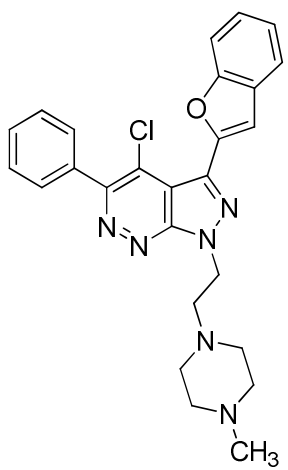


Vh:

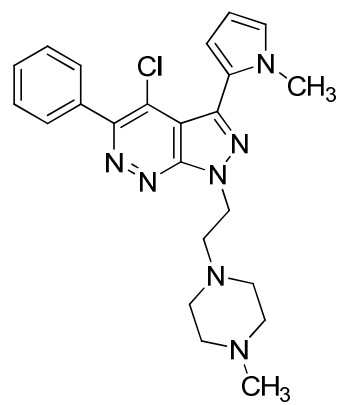
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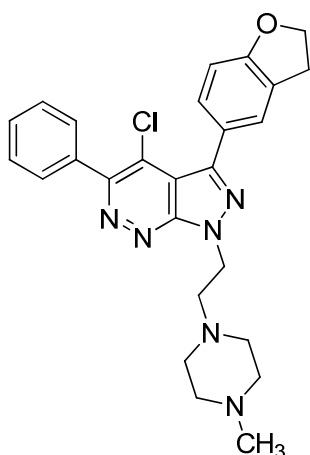
Vi:



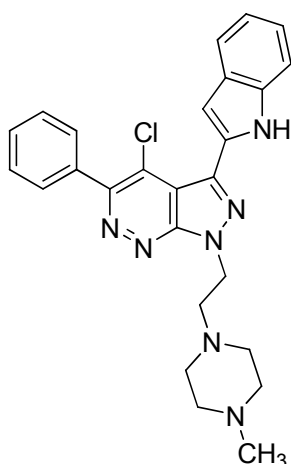
Vj:



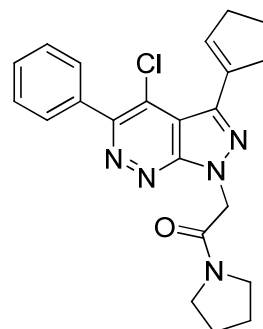
Vk:



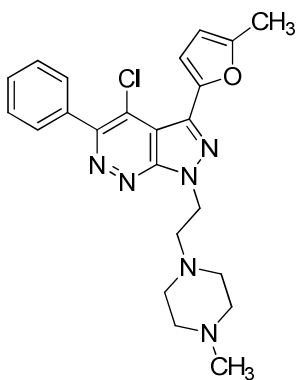
Vl:



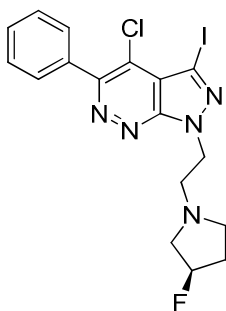
Vm:



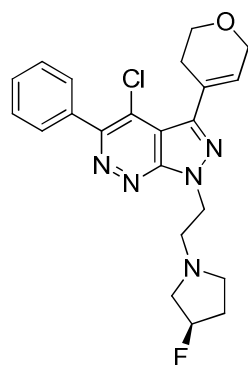
Vn:



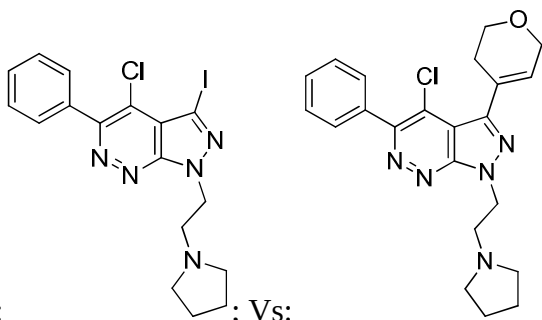
Vo:



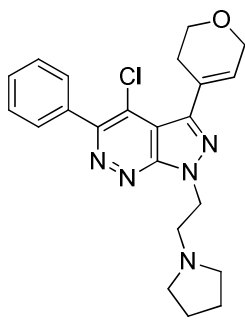
Vp:



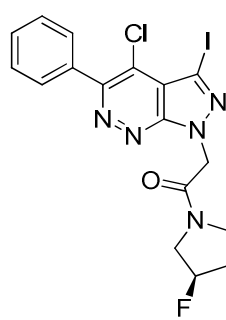
Vq:



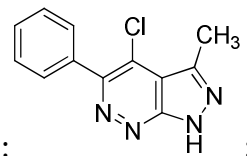
Vr:



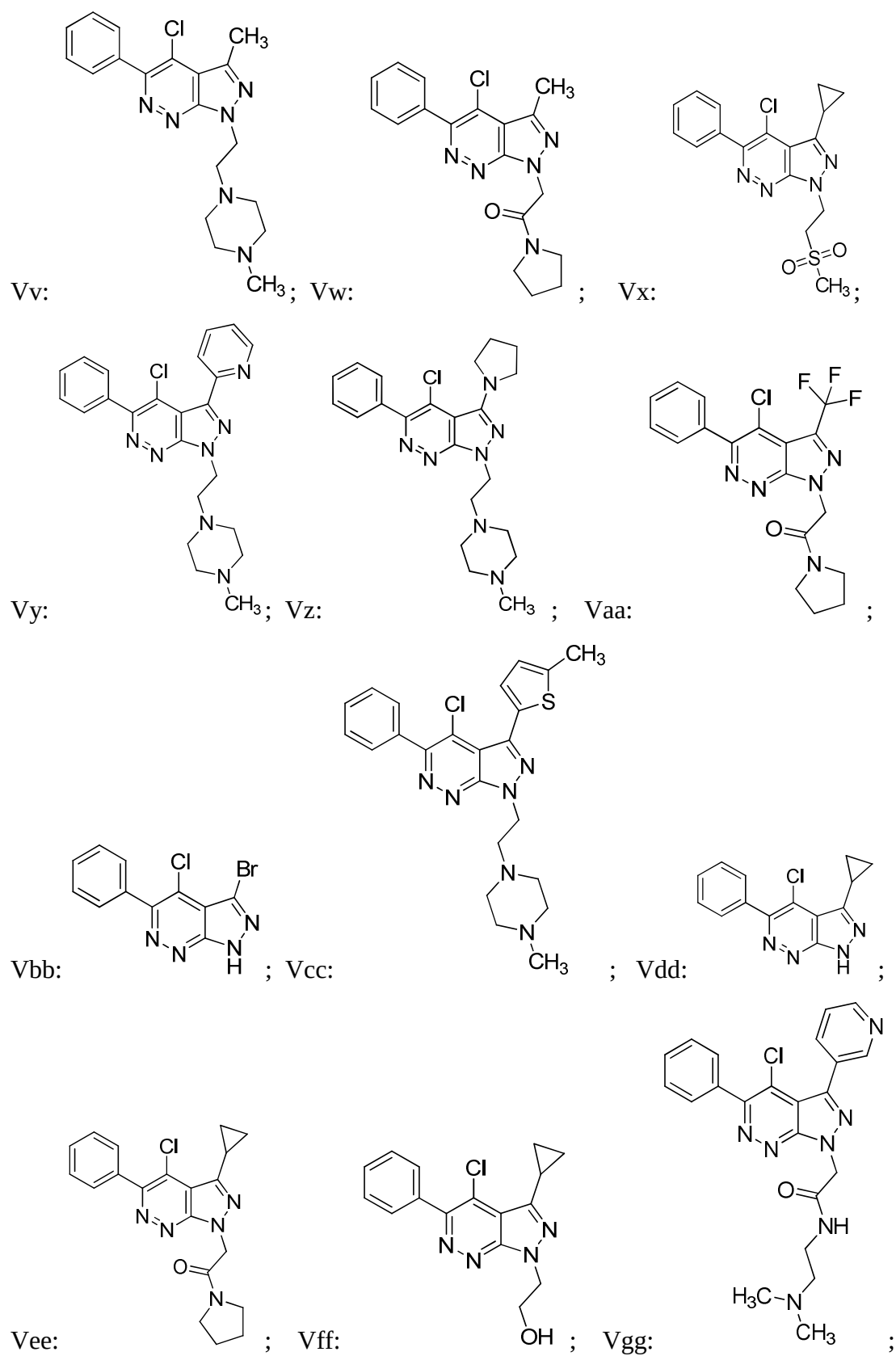
Vs:

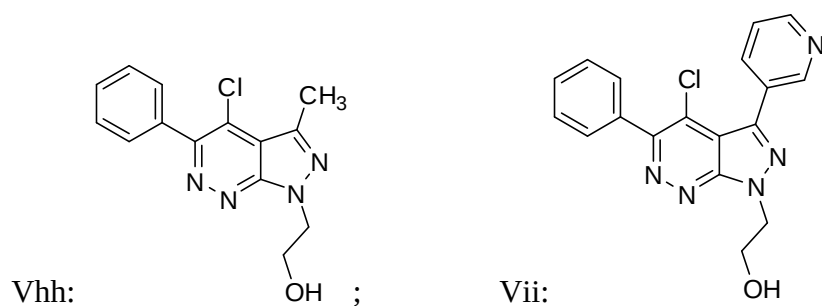


Vt:

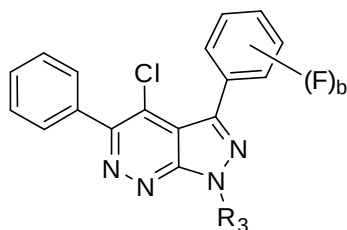


Vu:





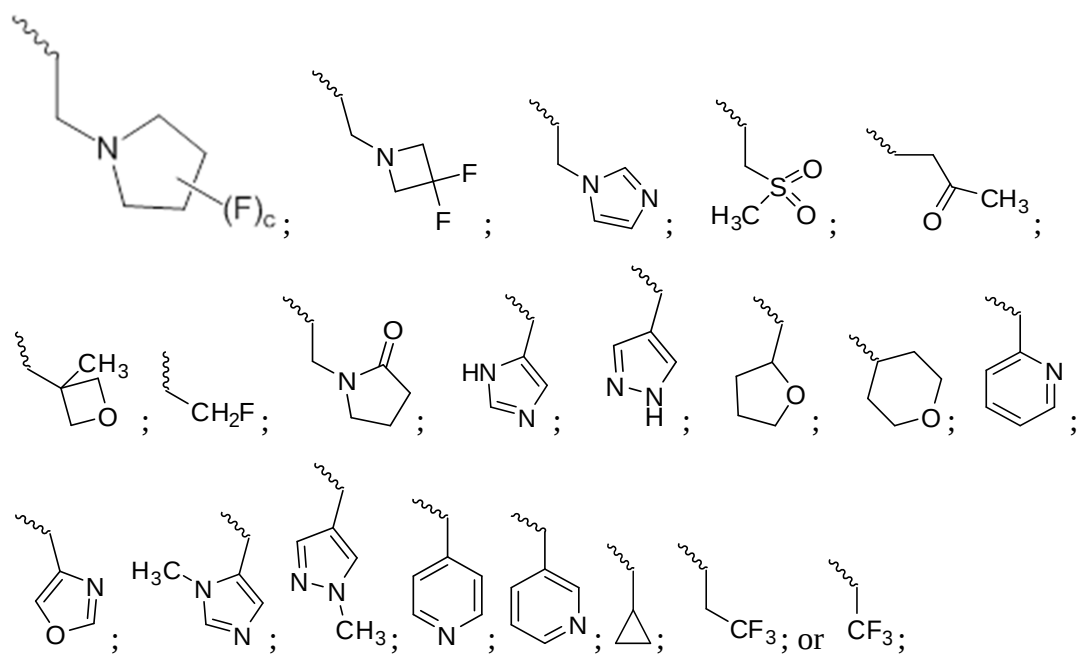
14. An effective amount of a compound of Formula VI:



Formula VI

or a pharmaceutically acceptable salt thereof,

wherein R₃ is:

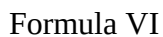


b is 0 or 1; and

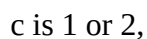
c is 1 or 2,

for use in the treatment or prevention of a proteopathy.

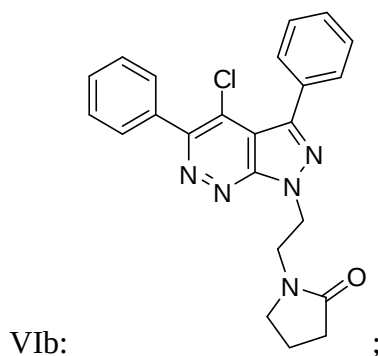
15. Use of an effective amount of a compound of Formula VI:

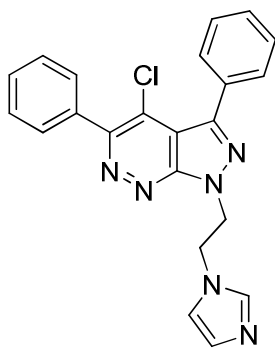


wherein R₃ is:

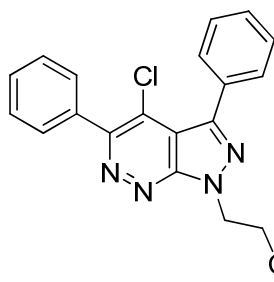


wherein the compound has the structure:

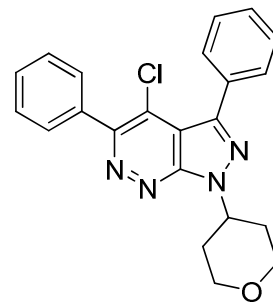




VIc:

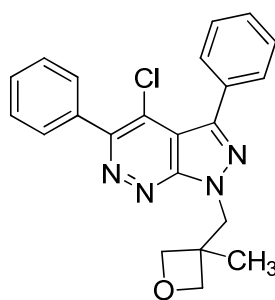


VIId:

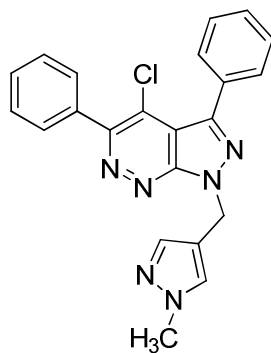


VIe:

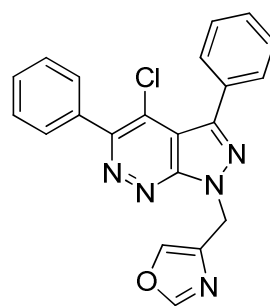
;



VIIf:

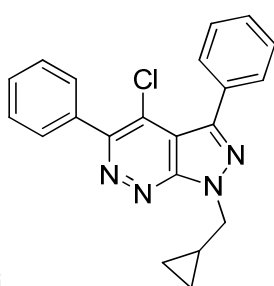


VIg:



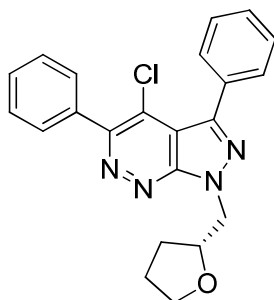
VIh:

;



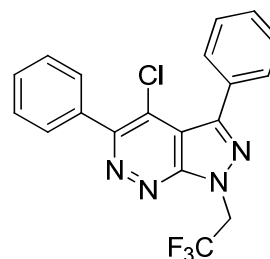
VIi

;



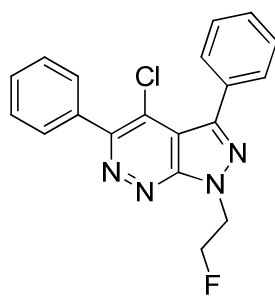
VIj:

;



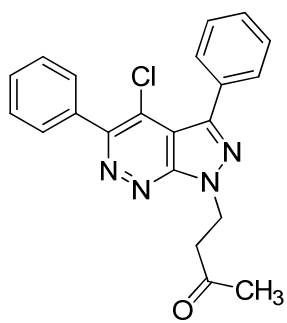
VIk:

;



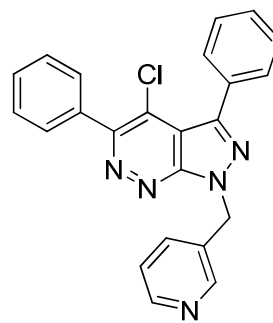
VII:

;



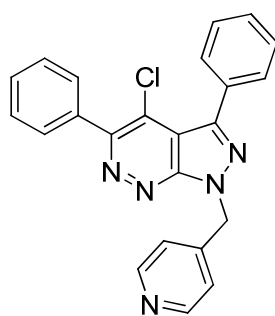
VIIm:

;



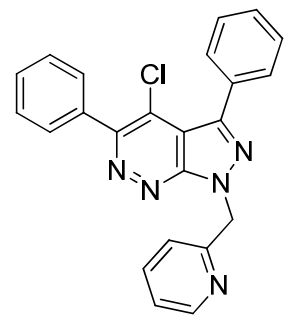
VIIn:

;



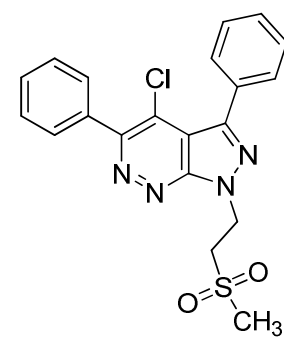
VIo:

;

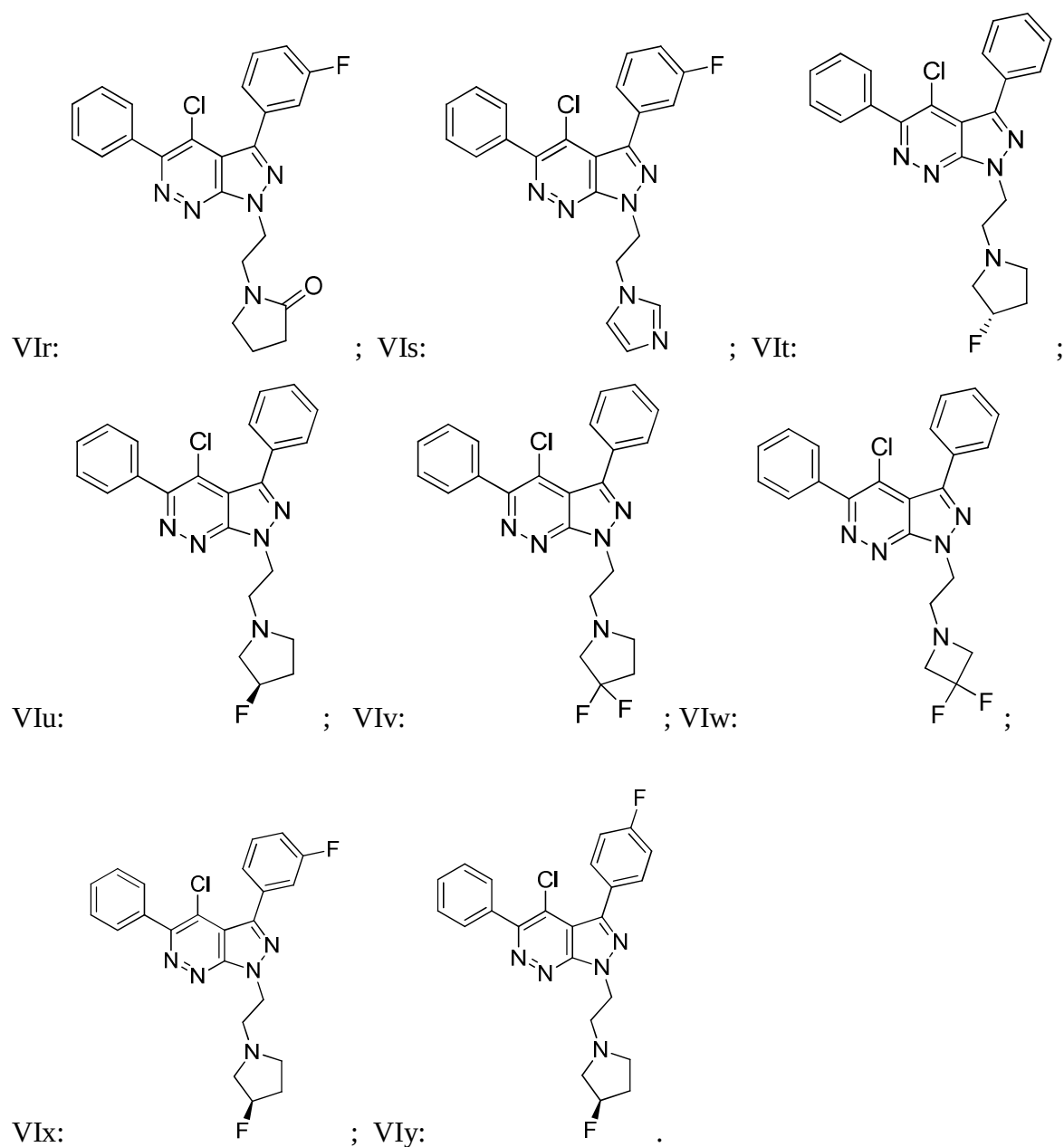


VIp:

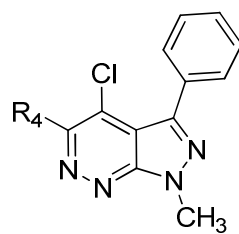
;



VIq:

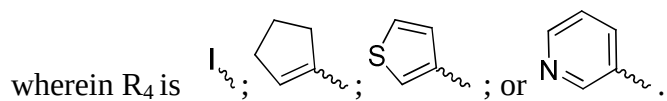


17. An effective amount of a compound of Formula VII:



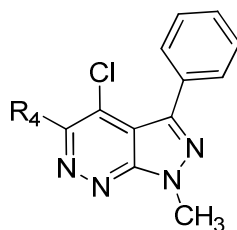
Formula VII

or a pharmaceutically acceptable salt thereof,



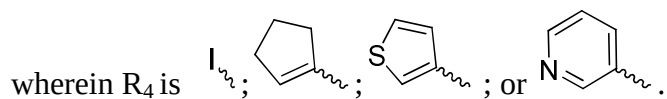
for use in the treatment or prevention of a proteopathy.

18. Use of an effective amount of a compound of Formula VII:



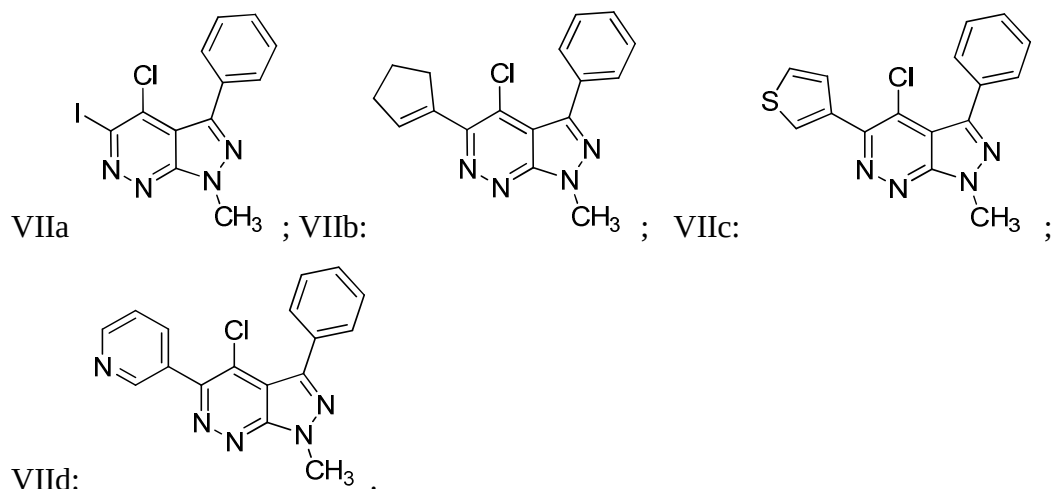
Formula VII

or a pharmaceutically acceptable salt thereof,

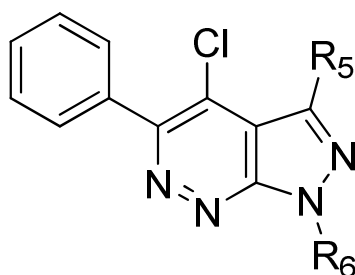


in the manufacture of a medicament for the treatment or prevention of a proteopathy.

19. The use of a compound or pharmaceutically acceptable salt of claim 17 or claim 18, wherein the compound has the structure:



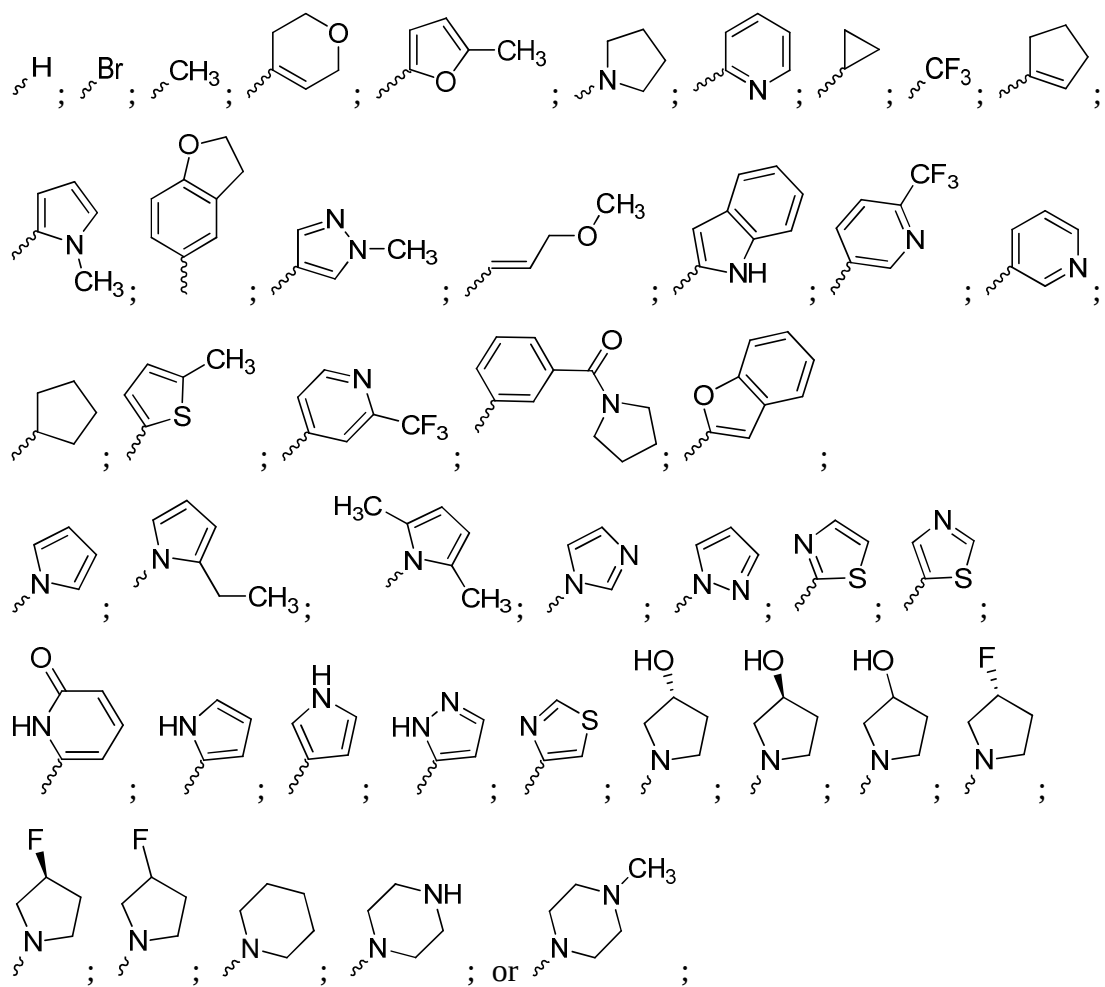
20. An effective amount of a compound of Formula XIII:



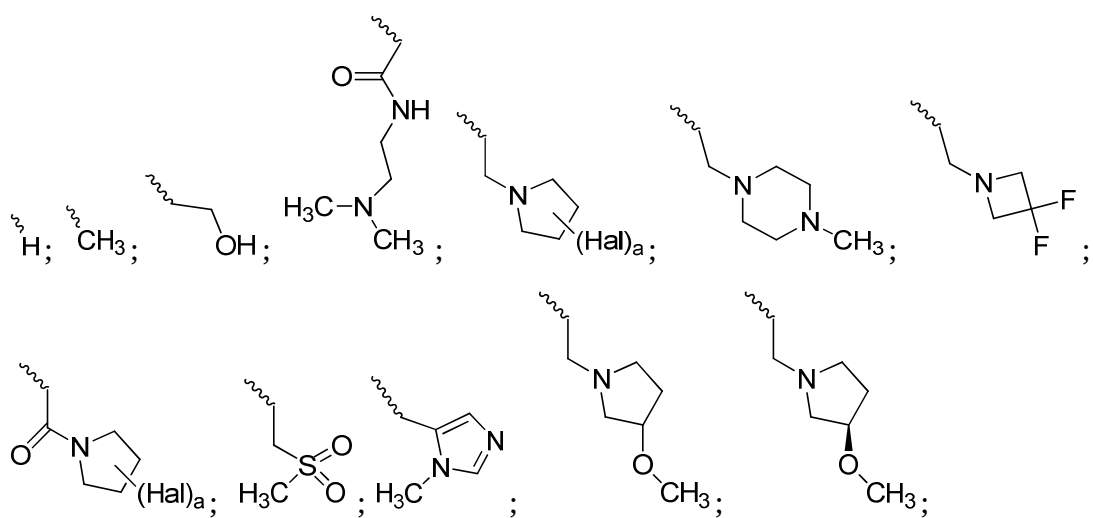
Formula XIII

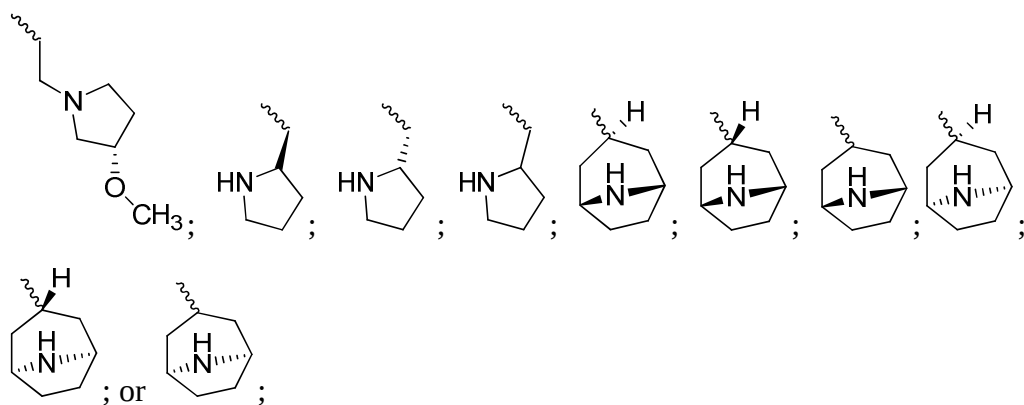
or pharmaceutically acceptable salt thereof,

wherein R₅ is:



R_6 is:



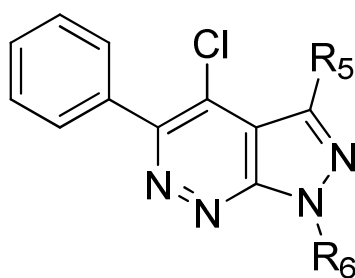


Hal is -Cl, -F, -I, or -Br; and

a is 0, 1, or 2,

for use in the treatment or prevention of a proteopathy.

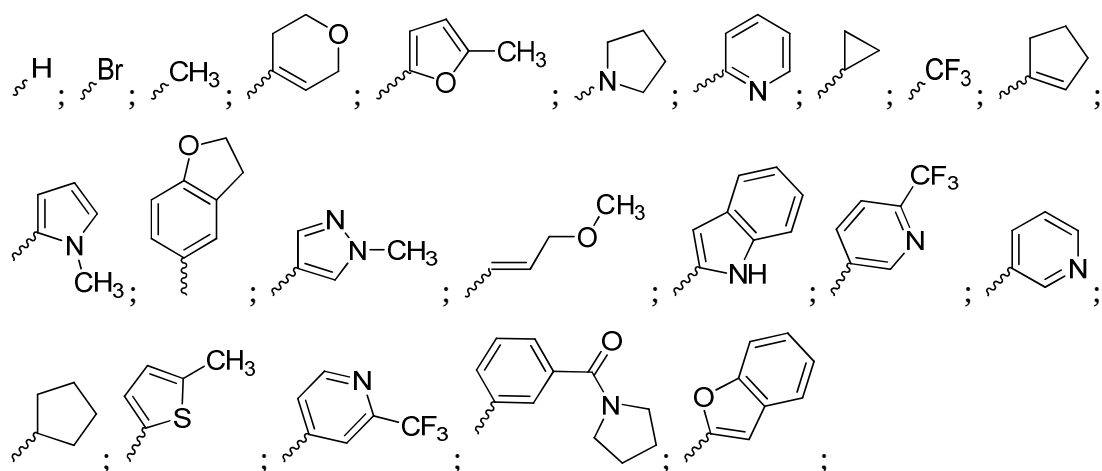
21. Use of an effective amount of a compound of Formula XIII:

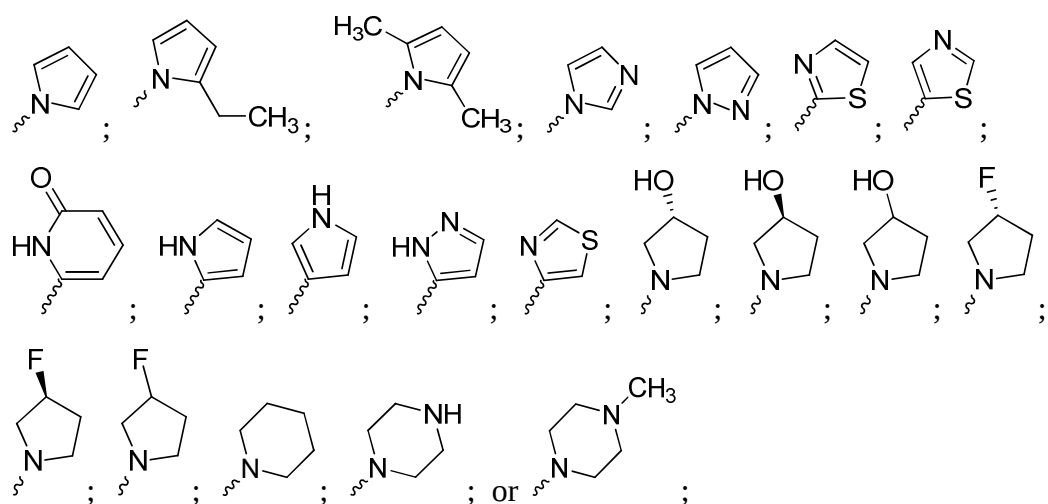


Formula XIII

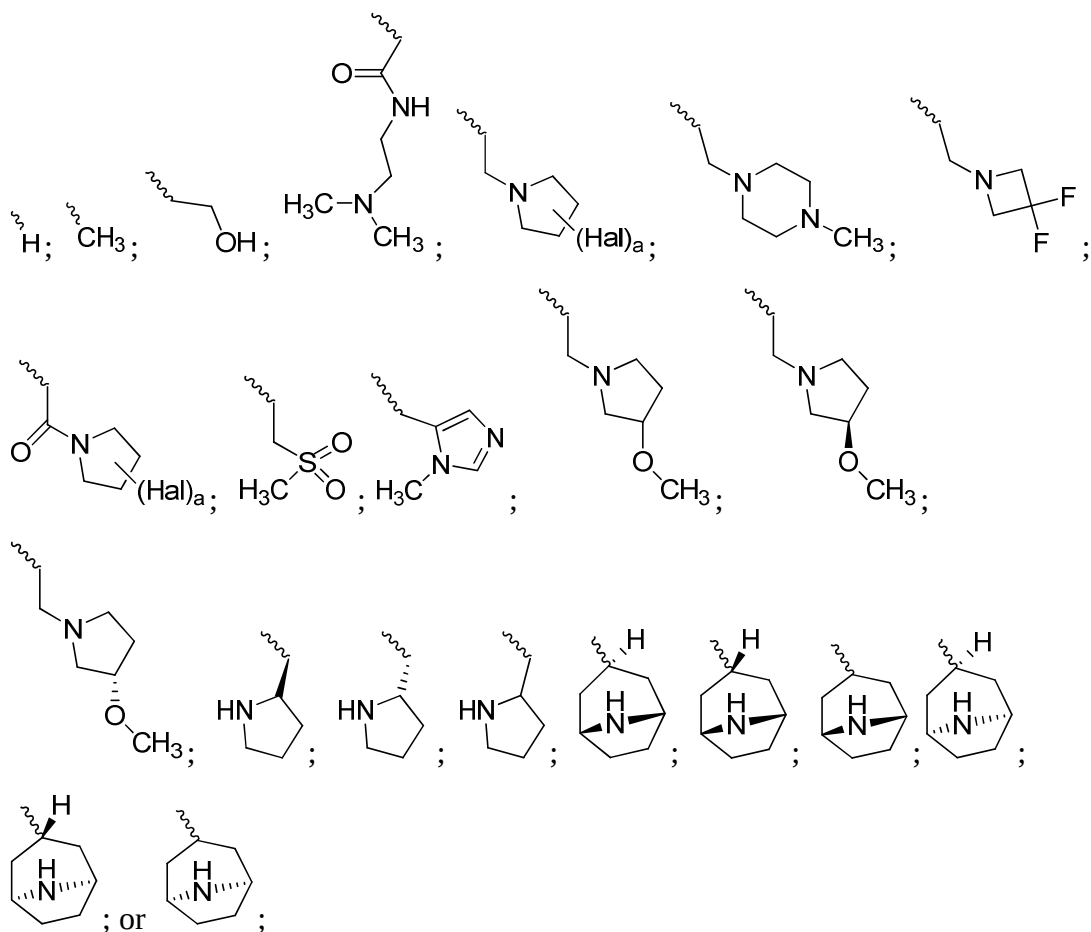
or pharmaceutically acceptable salt thereof,

wherein R₅ is:





R₆ is:



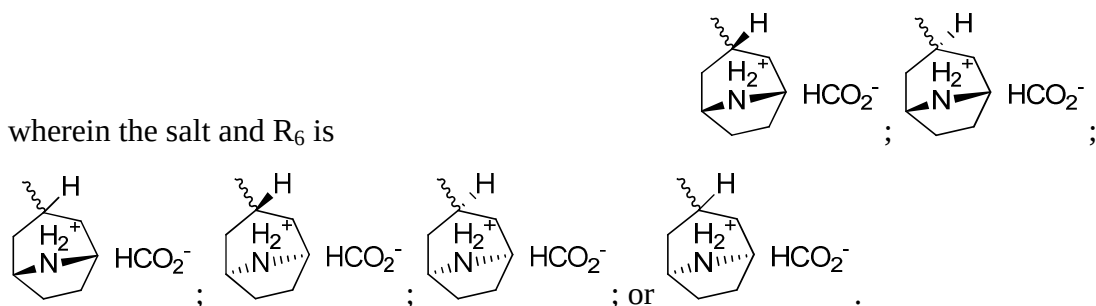
Hal is -Cl, -F, -I, or -Br; and

a is 0, 1, or 2,

in the manufacture of a medicament for the treatment or prevention of a proteopathy.

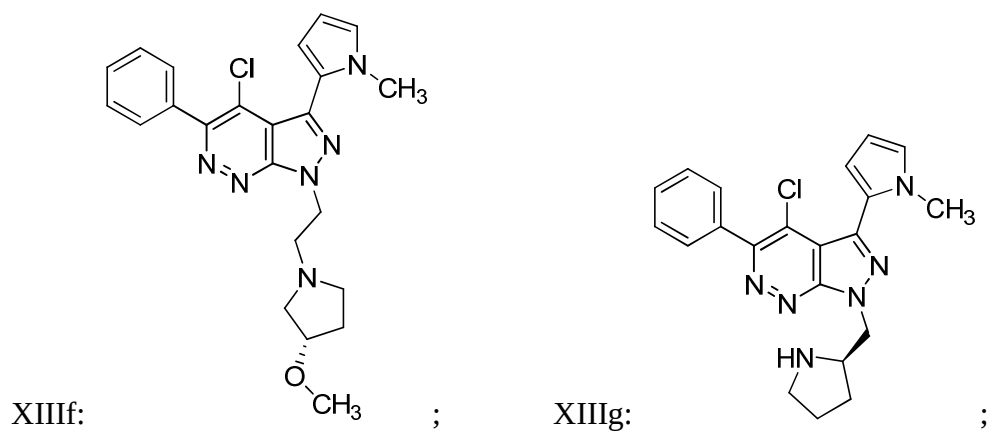
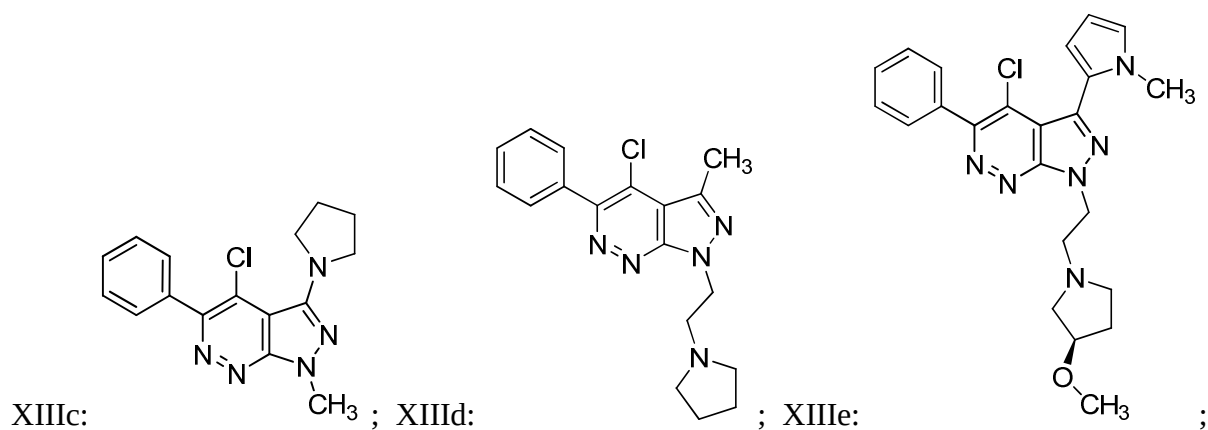
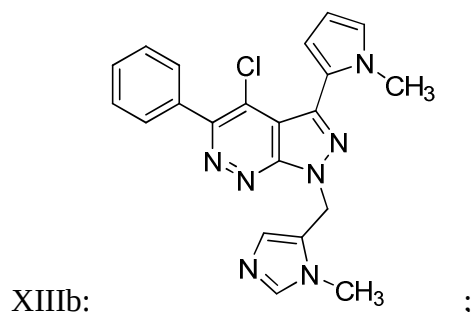
22. The use of a compound or pharmaceutically acceptable salt of claim 20 or claim 21,

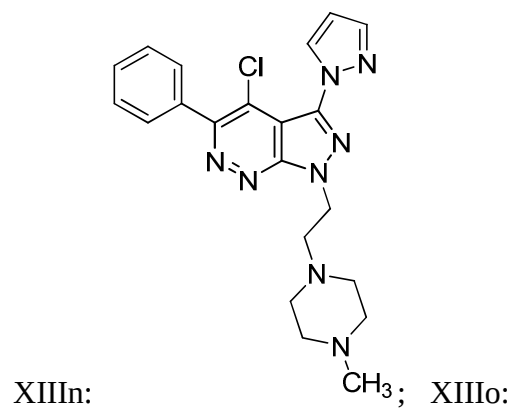
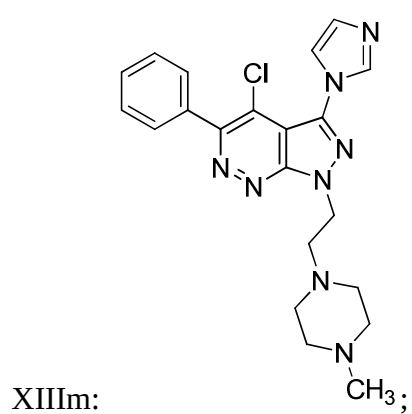
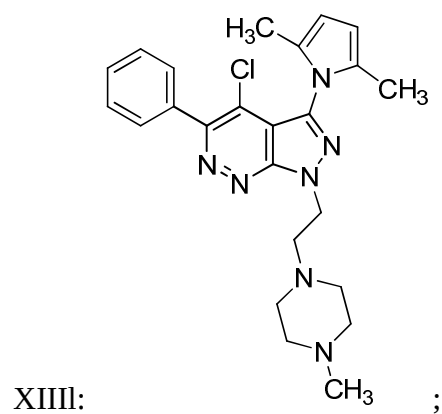
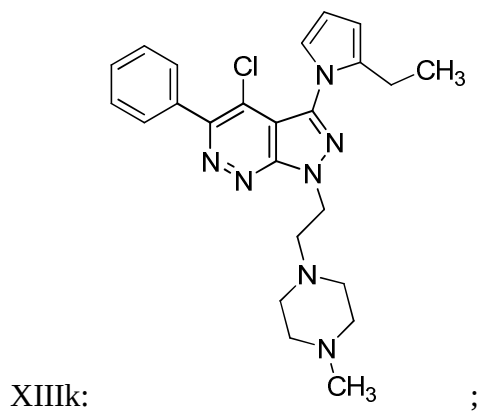
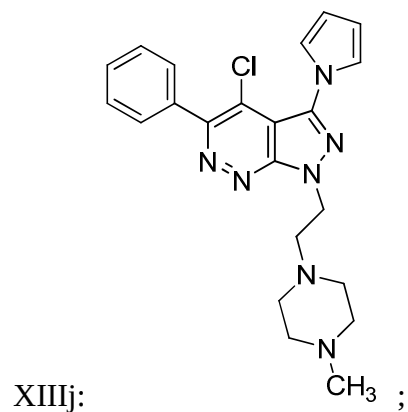
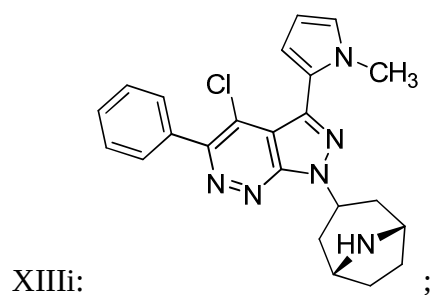
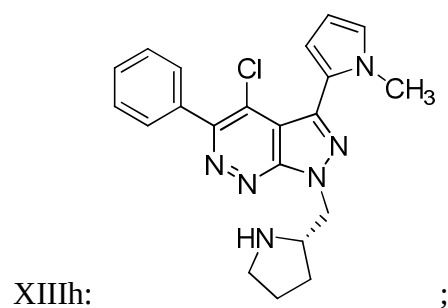
wherein the salt and R₆ is

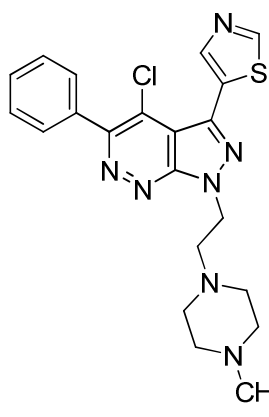


23. The use of a compound or pharmaceutically acceptable salt of claim 20 or claim 21,

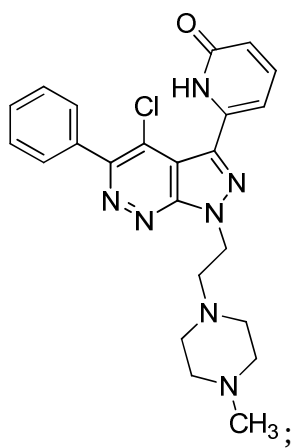
wherein the compound has the structure:



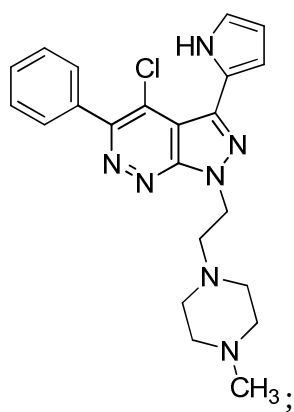




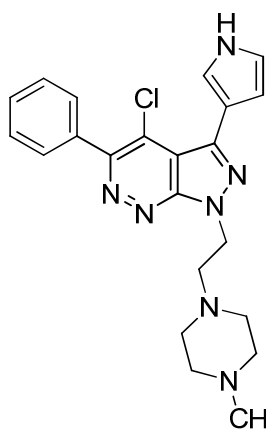
XIIIp:



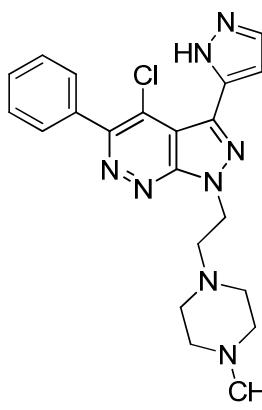
XIIIq:



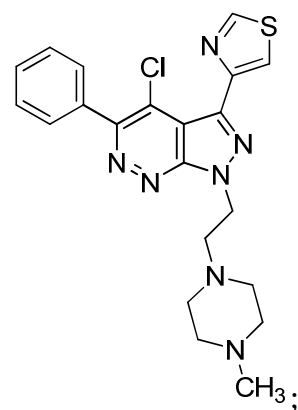
XIIIr:



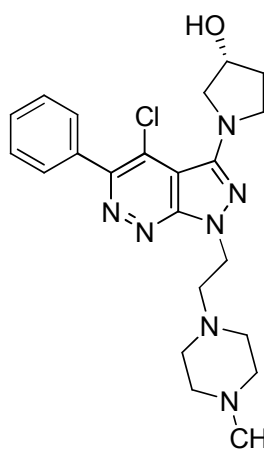
XIIIIs:



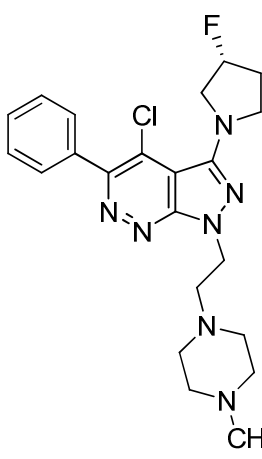
XIIIIt:



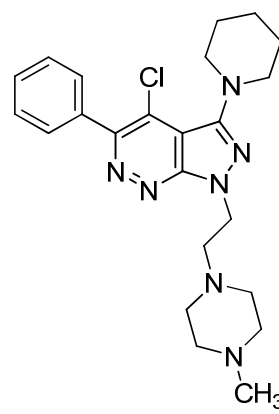
XIIIlu:



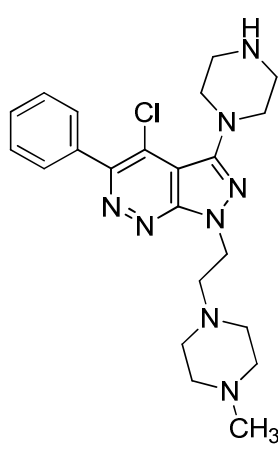
XIIIv:



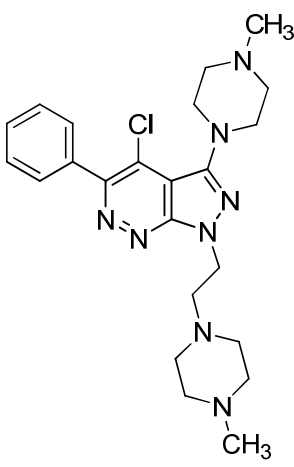
XIIIw:



XIIIx:

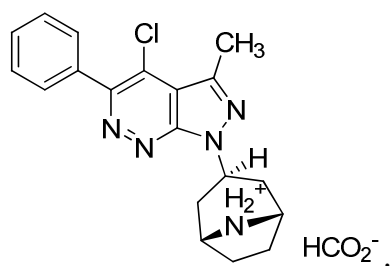


XIIIy:



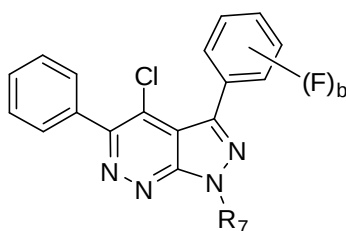
XIIIz:

24. The use of a compound or pharmaceutically acceptable salt of claim 20 or claim 21, wherein the pharmaceutically acceptable salt has the structure:



XIIIa:

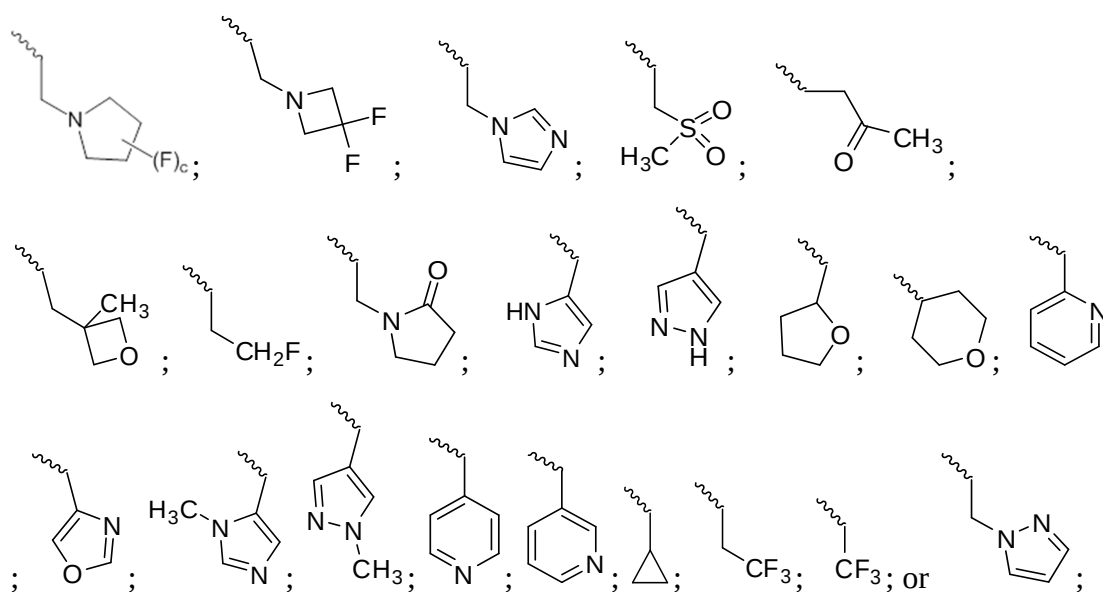
25. An effective amount of a compound of Formula XIV:



Formula XIV

or a pharmaceutically acceptable salt thereof,

wherein R₇ is:

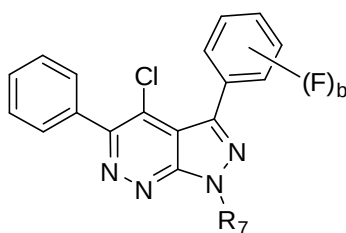


b is 0 or 1; and

c is 1 or 2,

for use in the treatment or prevention of a proteopathy.

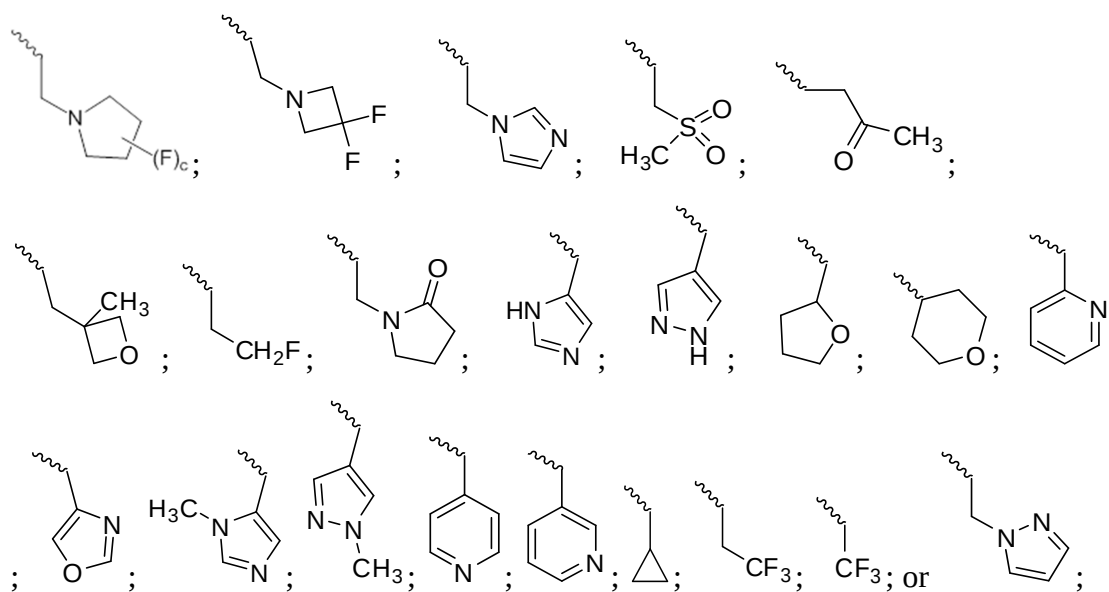
26. Use of an effective amount of a compound of Formula XIV:



Formula XIV

or a pharmaceutically acceptable salt thereof,

wherein R₇ is:

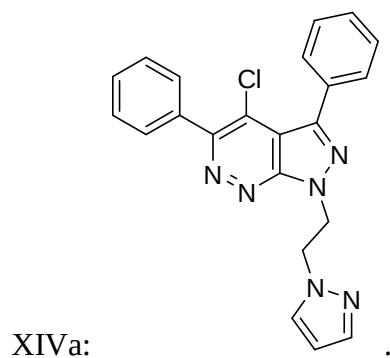


b is 0 or 1; and

c is 1 or 2,

in the manufacture of a medicament for the treatment or prevention of a proteopathy.

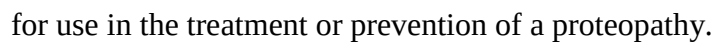
27. The use of a compound or pharmaceutically acceptable salt of claim 25 or claim 26, wherein the compound has the structure:



28. An effective amount of a compound of Formula XV:

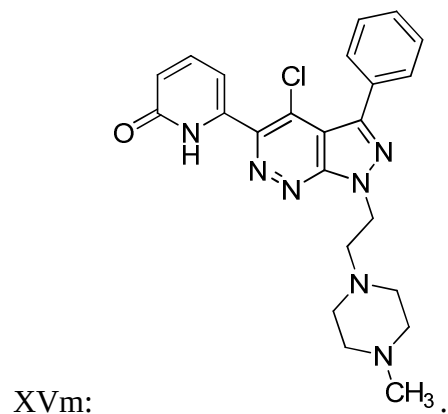
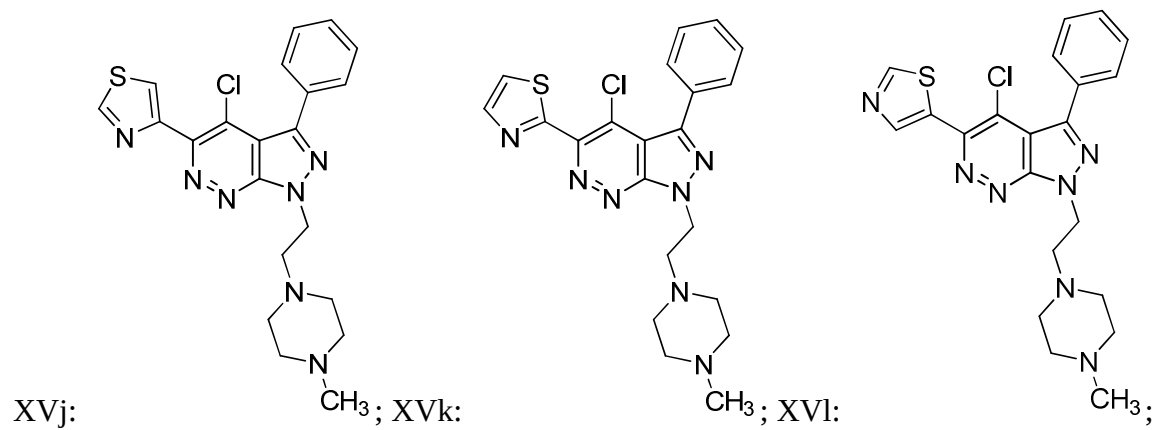


wherein R₈ is:

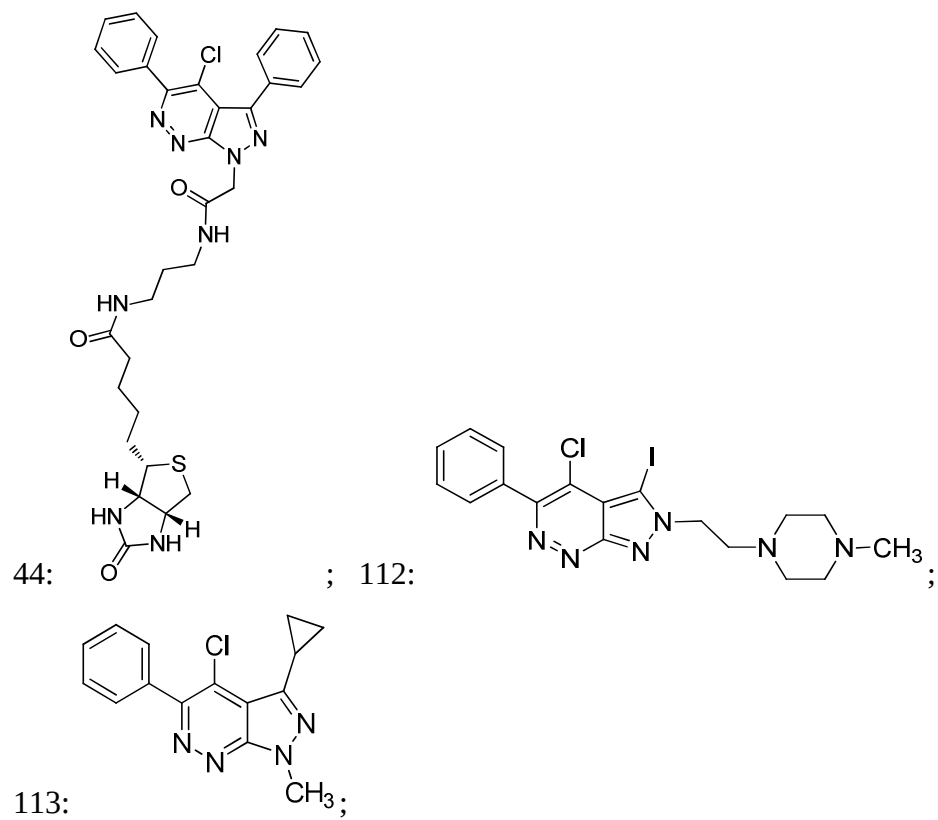


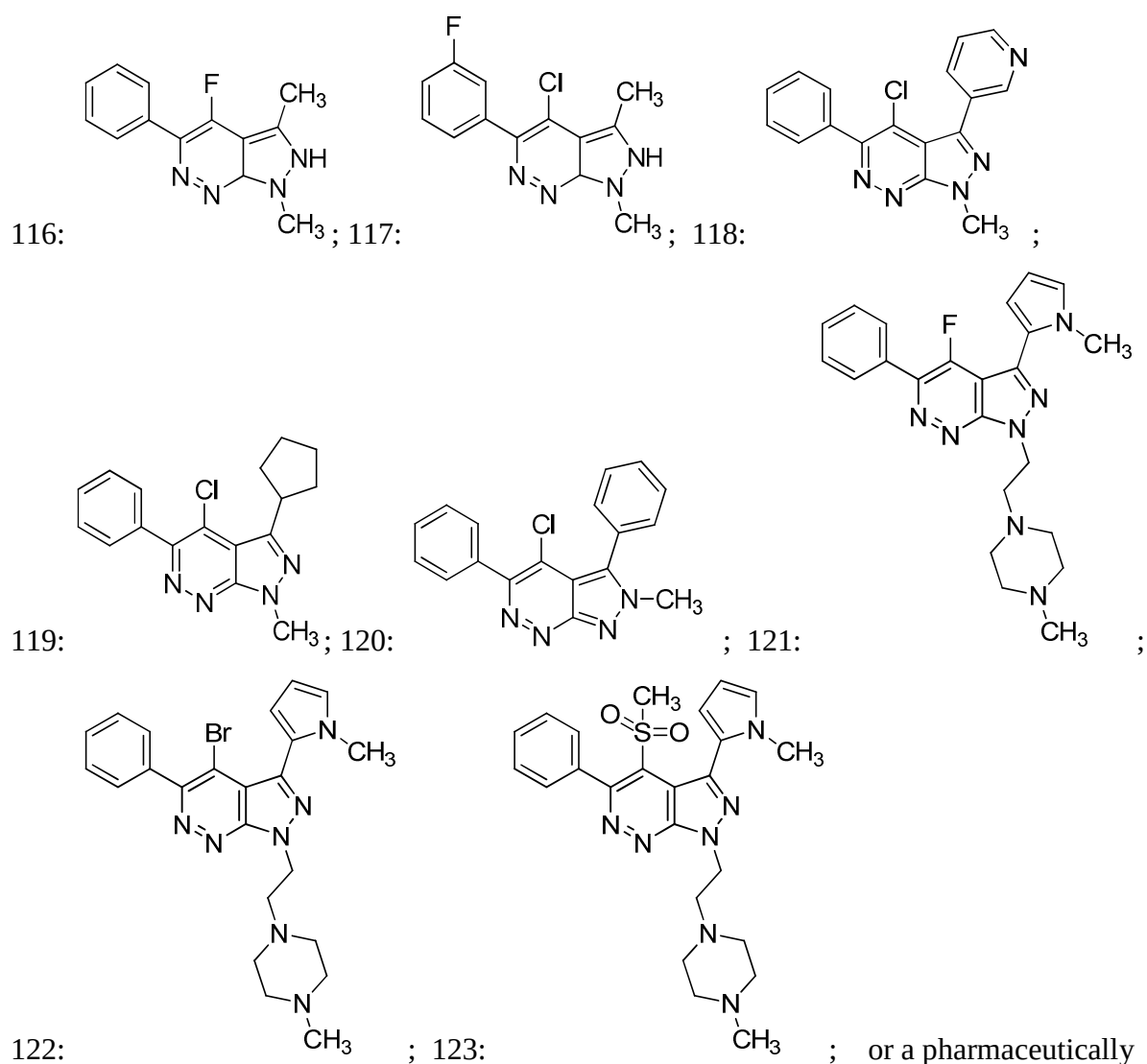
Formula XV

wherein R_8 is:



31. An effective amount of a compound having the structure:

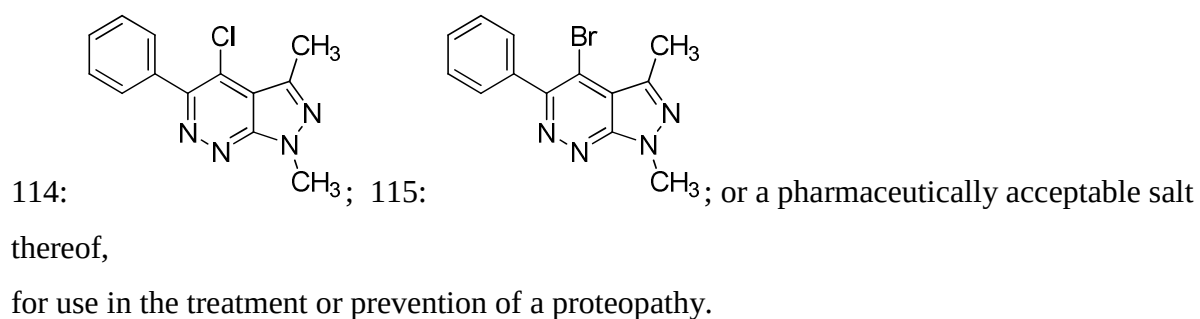




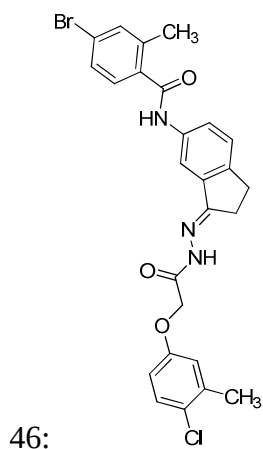
acceptable salt thereof

for use in the treatment or prevention of a proteopathy.

32. An effective amount of a compound having the structure:

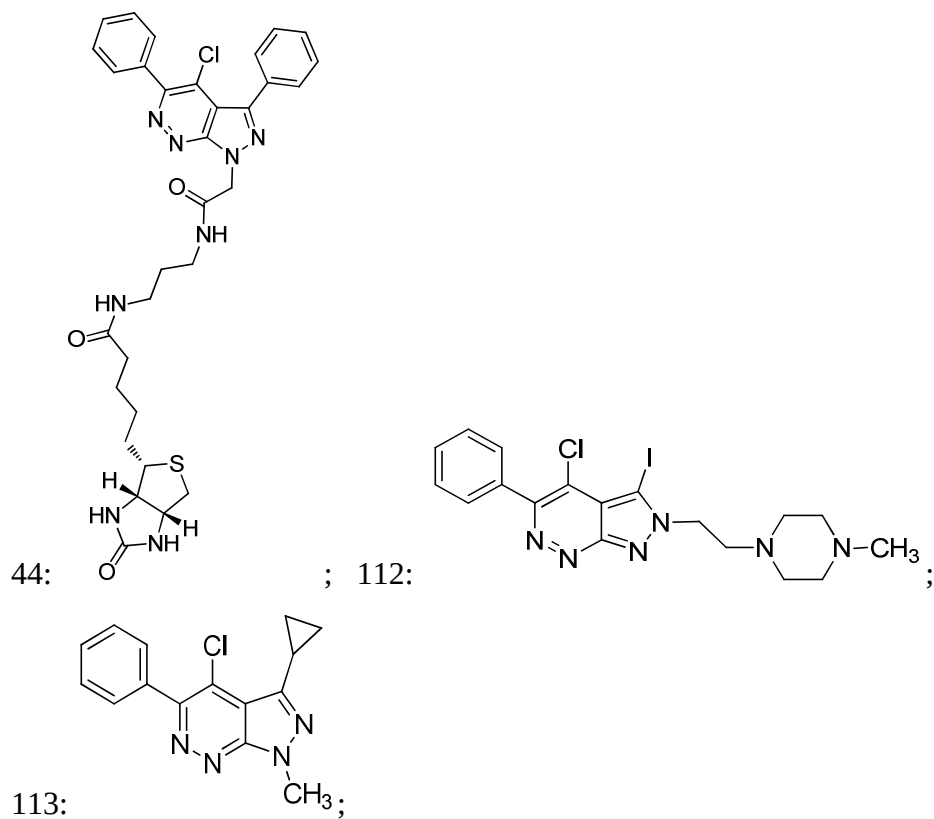


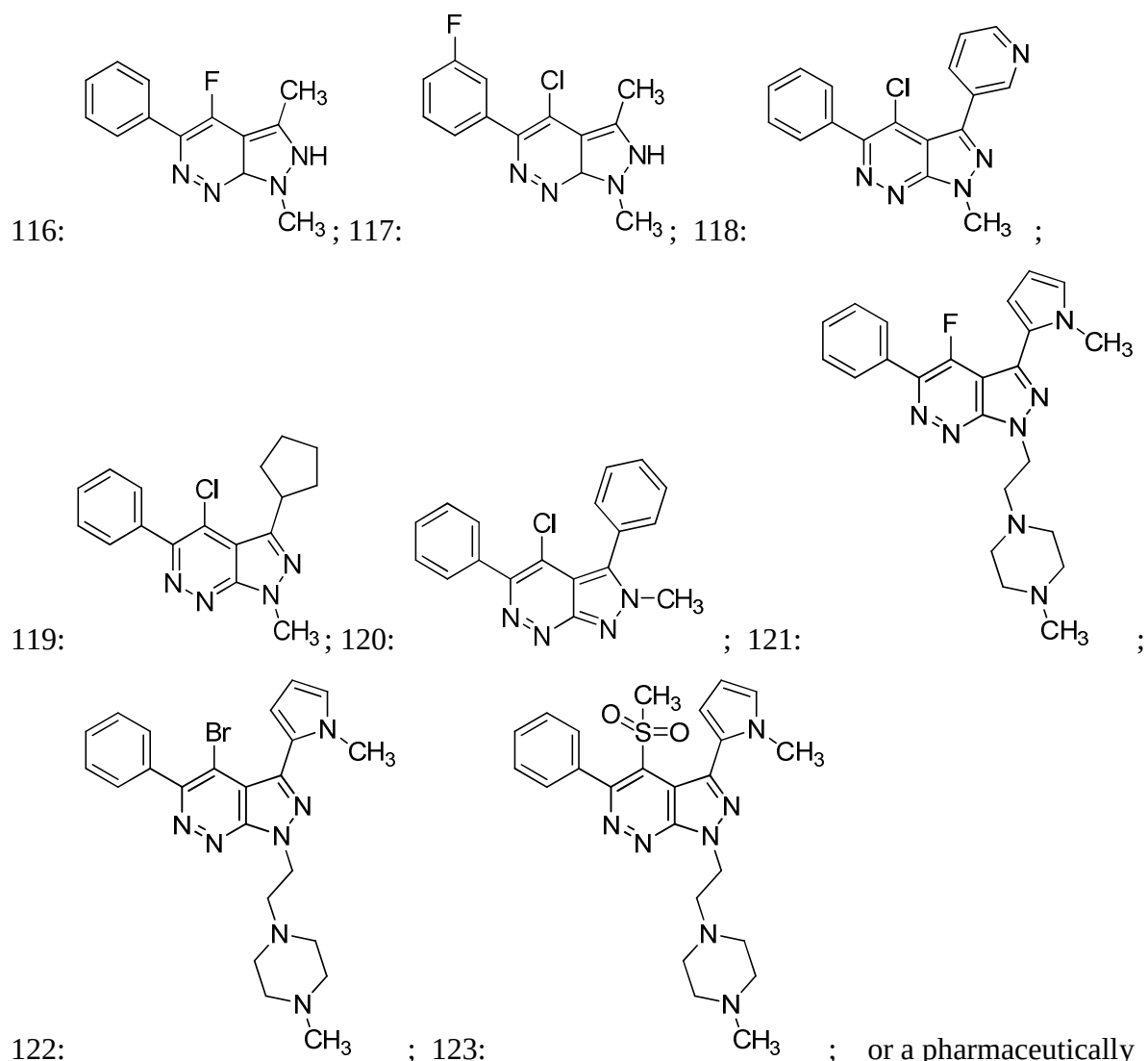
33. The use of an effective amount of Compound 46 having the structure:



or a pharmaceutically acceptable salt thereof in the manufacture of a medicament for the treatment or prevention of a proteopathy.

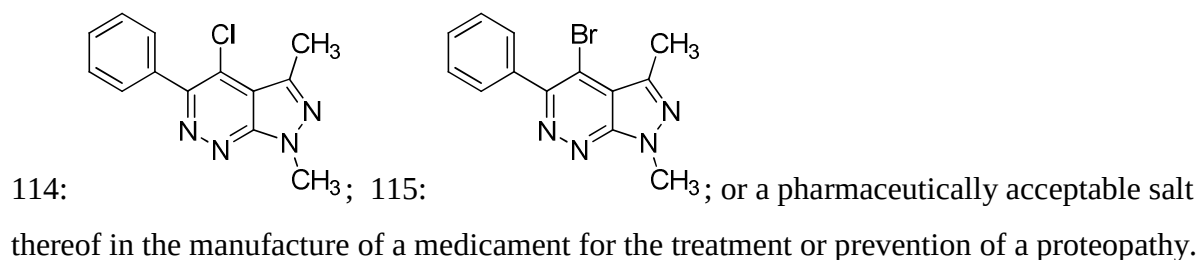
34. The use of an effective amount of a compound having the structure:





acceptable salt thereof in the manufacture of a medicament for the treatment or prevention of a proteopathy.

35. The use of an effective amount of a compound having the structure:



36. The use of a compound or a pharmaceutically acceptable salt of any one of claims 2, 3, 5, 6, 9, 10, 12, 13, 15, 16, 18, 19, 21, 22, 23, 24, 26, 27, 29, 30, 33, 34 or 35, wherein the proteopathy is a neurodegenerative disease.

37. The use of a compound or a pharmaceutically acceptable salt of claim 36, wherein the neurodegenerative disease is Alzheimer's disease, progressive supranuclear palsy, dementia pugilistica, frontotemporal dementia and parkinsonism linked to chromosome 17 (FTDP-17), Lytico-Bodig disease, tangle-predominant dementia, ganglioma, gangliocytoma, meningioangiomas, subacute sclerosing panencephalitis, lead encephalopathy, tuberous sclerosis, Hallervorden-Spatz disease, lipofuscinosis, Pick's disease, corticobasal degeneration, argyrophilic grain disease, Huntington's disease, Parkinson's disease, dementia with Lewy bodies, multiple system atrophy, neuroaxonal dystrophies, dentatorubralpallidoluysian atrophy (DRPLA), spinal-bulbar muscular atrophy (SBMA), spinocerebellar ataxia 1 (SCA 1), SCA 2, SCA 3, SCA 6, SCA 7, SCA 17, prion disease, amyotrophic lateral sclerosis, frontotemporal lobar degeneration (FTLD) or familial encephalopathy with neuroserpin inclusion bodies (FENIB).

38. The use of a compound or a pharmaceutically acceptable salt of any one of claims 2, 3, 5, 6, 9, 10, 12, 13, 15, 16, 18, 19, 21, 22, 23, 24, 26, 27, 29, 30, 33, 34 or 35, wherein the proteopathy is an amyloidosis.

39. The use of claim 38, wherein the amyloidosis is familial British dementia (ABri), familial Danish dementia (ADan), hereditary cerebral haemorrhage with amyloidosis-Icelandic (HCHWA-I), familial amyloidotic neuropathy (ATTR), AL (light chain) primary systemic amyloidosis, AH (heavy chain) amyloidosis, AA secondary amyloidosis, A β amyloidosis, aortic medial amyloidosis, LECT2 amyloidosis, AIAPP amyloidosis, apolipoprotein AI amyloidosis (AApoAI), apolipoprotein AII amyloidosis (AApoAII), apolipoprotein AIV amyloidosis (AApoAIV), familial amyloidosis of the Finnish type (FAF), fibrinogen amyloidosis (AFib), lysozyme amyloidosis (ALys), dialysis amyloidosis (A β_2 M), medullary thyroid carcinoma (ACal), cardiac atrial amyloidosis (AANF), pituitary prolactinoma (APro), hereditary lattice corneal dystrophy, cutaneous lichen amyloidosis (AKer), Mallory bodies, primary cutaneous amyloidosis, corneal lactoferrin amyloidosis, odontogenic (Pindborg) tumor amyloid or seminal vesicle amyloid.

40. The use of a compound or a pharmaceutically acceptable salt of any one of claims 2, 3, 5, 6, 9, 10, 12, 13, 15, 16, 18, 19, 21, 22, 23, 24, 26, 27, 29, 30, 33, 34 or 35, wherein the proteopathy is a lysosomal storage disease.

41. The use of claim 40, wherein the lysosomal storage disease is a mucopolysaccharidosis disorder.

42. The use of claim 41, wherein the mucopolysaccharidosis disorder is Pseudo-Hurler polydystrophy/Mucopolidosis IIIA, MPS I Hurler Syndrome, MPS I Scheie Syndrome, MPS I Hurler-Scheie Syndrome, MPS II Hunter syndrome, Sanfilippo syndrome Type A/MPS III A, Sanfilippo syndrome Type B/MPS III B, Sanfilippo syndrome Type C/MPS III C, Sanfilippo syndrome Type D/MPS III D, Morquio Type A/MPS IVA, Morquio Type B/MPS IVB, MPS IX Hyaluronidase Deficiency, MPS VI Maroteaux-Lamy, MPS VII Sly Syndrome, Mucopolidosis I/Sialidosis, Mucopolidosis IIIC or Mucopolidosis type IV.

43. The use of claim 40, wherein the lysosomal storage disease is Pompe disease/glycogen storage disease type II.

44. The use of claim 40, wherein the lysosomal storage disease is activator deficiency/GM2 gangliosidosis, alpha-mannosidosis, aspartylglucosaminuria, cholesteryl ester storage disease, chronic Hexosaminidase A Deficiency, cystinosis, Danon disease, Fabry disease, Farber disease, fucosidosis, galactosialidosis, Gaucher Disease Type I, Gaucher Disease Type II, Gaucher Disease Type III, GM1 gangliosidosis infantile, GM1 gangliosidosis late infantile/juvenile, GM1 gangliosidosis adult/chronic, I-Cell disease/Mucopolidosis II, Infantile Free Sialic Acid Storage Disease/ISSD, Juvenile Hexosaminidase A Deficiency, Krabbe disease infantile onset, Krabbe disease late onset, lysosomal acid lipase deficiency early onset, lysosomal acid lipase deficiency Late onset, Metachromatic Leukodystrophy, Pseudo-Hurler polydystrophy/Mucopolidosis IIIA, MPS I Hurler Syndrome, MPS I Scheie Syndrome, MPS I Hurler-Scheie Syndrome, MPS II Hunter syndrome, Sanfilippo syndrome Type A/MPS III A, Sanfilippo syndrome Type B/MPS III B, Sanfilippo syndrome Type C/MPS III C, Sanfilippo syndrome Type D/MPS III D, Morquio Type A/MPS IVA, Morquio Type B/MPS IVB, MPS IX Hyaluronidase Deficiency, MPS VI Maroteaux-Lamy, MPS VII Sly Syndrome, Mucopolidosis I/Sialidosis, Mucopolidosis IIIC, Mucopolidosis type IV, Multiple sulfatase deficiency, Niemann-Pick Disease Type A, Niemann-Pick Disease Type B, Niemann-Pick Disease Type C, CLN6 disease-atypical late infantile, CLN6 disease-late onset variant, CLN6 disease-early juvenile, Batten-Spielmeyer-Vogt/Juvenile NCL/CLN3 disease, Finnish Variant Late Infantile CLN5, Jansky-Bielschowsky disease/Late infantile CLN2/TPP1 Disease, Kufs/Adult-onset NCL/CLN4 disease, Northern Epilepsy/variant late

infantile CLN8, Santavuori-Haltia/Infantile CLN1/PPT disease, Beta-mannosidosis, Pompe disease/glycogen storage disease type II, Pycnodysostosis, Sandhoff disease/Adult Onset/GM2 Gangliosidosis, Sandhoff disease/GM2 gangliosidosis – Infantile, Sandhoff disease/GM2 gangliosidosis – Juvenile, Schindler disease, Salla disease/Sialic Acid Storage Disease, Tay-Sachs/GM2 gangliosidosis or Wolman disease.

45. The use of a compound or a pharmaceutically acceptable salt of any one of claims 2, 3, 5, 6, 9, 10, 12, 13, 15, 16, 18, 19, 21, 22, 23, 24, 26, 27, 29, 30, 33 or 34 to 44, wherein the compound or pharmaceutically acceptable salt binds to a molecular chaperone.

46. The use of claim 45, wherein the molecular chaperone is a member of the Hsp10 family, Hsp40 family, Hsp60 family, Hsp70 family, Hsp90 family, or Hsp100 family.