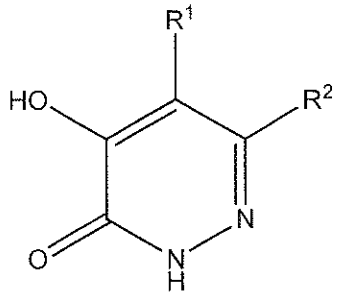
~~In one aspect of t~~The present ~~invention~~disclosure~~, there is~~ provide~~s~~d a compound of formula (I), or a pharmaceutically acceptable salt thereof,

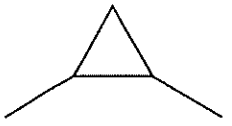
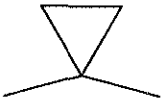


(I)

wherein

R^1 represents a hydrogen or fluorine atom or a trifluoromethyl group;

R^2 represents a group –X-Y-R^3;

X and Y each independently represent a bond, an oxygen atom or a group -C(O), -$S(O)_n$, -$C(O)NR^4$, -$S(O)_2NR^4$, -NR^4, ,  or -CR^4R^5-, provided that X and Y cannot both simultaneously represent a bond and provided that if X and Y are both other than a bond, then at least one of X and Y represents -CR^4R^5-;

n is 0, 1 or 2;

each R^4 independently represents a hydrogen atom or a C_1-C_6 alkyl or C_1-C_6 haloalkyl group;

each R^5 independently represents a hydrogen atom, a C_1-C_6 alkyl or C_1-C_6 haloalkyl group or =CH-;

R^3 represents a 3- to 10-membered saturated or unsaturated carbocyclic or heterocyclic ring system, the ring system itself being optionally substituted by at least one substituent selected from halogen, hydroxyl, cyano, oxo, C_1-C_6 alkyl, C_2-C_6 alkenyl, C_1-C_6 haloalkyl, C_1-C_6 hydroxyalkyl, C_1-C_6 alkoxy, C_1-C_6 haloalkoxy, C_1-C_6 alkylthio, C_1-C_6 alkylsulphinyl, C_1-C_6 alkylsulphonyl, C_1-C_6 alkylcarbonyl,

C_1-C_6 alkylcarbonyloxy, C_1-C_6 alkoxycarbonyl, amino (-NH_2), -$CON(R^6)_2$, C_1-C_6 alkylamino, di-(C_1-C_6 alkyl)amino, C_3-C_6 cycloalkyl, C_3-C_6 cycloalkyloxy, C_3-C_6 cycloalkylmethyl, -$[O]_p$-$(CH_2)_q$-O-R^7 and a 4- to 6-membered saturated or unsaturated heterocyclic ring (optionally substituted with at least one substituent selected from C_1-C_4 alkyl and C_1-C_4 alkoxy);

each R^6 independently represents a hydrogen atom or a C_1-C_6 alkyl group;

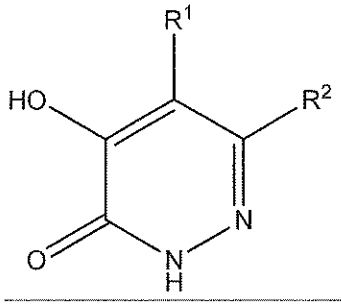
p is 0 or 1;

q is 1, 2, 3 or 4; and

R^7 represents a C_1-C_6 alkyl group;

for use in the prevention or treatment of an ataxic disorder, in particular a spinocerebellar ataxic disorder.

In one aspect of the present disclosure, there is provided a compound of formula (I), or a pharmaceutically acceptable salt thereof,

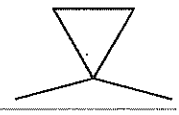


(I)

wherein

R^1 represents a hydrogen or fluorine atom or a trifluoromethyl group;

R^2 represents a group -X-Y-R^3;

X and Y each independently represent a bond, an oxygen atom or a group -C(O), -$S(O)_n$, -$C(O)NR^4$, -$S(O)_2NR^4$, -NR^4,  ,  or -CR^4R^5-, provided that X and Y cannot both simultaneously represent a bond and

provided that if X and Y are both other than a bond, then at least one of X and Y represents $-CR^4R^5-$;

n is 0, 1 or 2;

each R^4 independently represents a hydrogen atom or a C_1-C_6 alkyl or C_1-C_6 haloalkyl group;

each R^5 independently represents a hydrogen atom, a C_1-C_6 alkyl or C_1-C_6 haloalkyl group or =CH-;

R^3 represents a 3- to 10-membered saturated or unsaturated carbocyclic or heterocyclic ring system, the ring system itself being optionally substituted by at least one substituent selected from halogen, hydroxyl, cyano, oxo, C_1-C_6 alkyl, C_2-C_6 alkenyl, C_1-C_6 haloalkyl, C_1-C_6 hydroxyalkyl, C_1-C_6 alkoxy, C_1-C_6 haloalkoxy, C_1-C_6 alkylthio, C_1-C_6 alkylsulphinyl, C_1-C_6 alkylsulphonyl, C_1-C_6 alkylcarbonyl, C_1-C_6 alkylcarbonyloxy, C_1-C_6 alkoxycarbonyl, amino, $-CON(R^6)_2$, C_1-C_6 alkylamino, di-(C_1-C_6 alkyl)amino, C_3-C_6 cycloalkyl, C_3-C_6 cycloalkyloxy, C_3-C_6 cycloalkylmethyl, $-[O]_p-(CH_2)_q-O-R^7$ and a 4- to 6-membered saturated or unsaturated heterocyclic ring (optionally substituted with at least one substituent selected from C_1-C_4 alkyl and C_1-C_4 alkoxy);

each R^6 independently represents a hydrogen atom or a C_1-C_6 alkyl group;

p is 0 or 1;

q is 1, 2, 3 or 4; and

R^7 represents a C_1-C_6 alkyl group;

for use in the prevention or treatment of an ataxic disorder.

~~In another aspect, t~~The present ~~invention~~ disclosure also provides the use of a compound of formula (I) or a pharmaceutically acceptable salt thereof as hereinbefore defined in the manufacture of a medicament for use in the prevention or treatment of an ataxic disorder.

In another aspect of the present disclosure, there is provided the use of a compound as hereinbefore defined in the manufacture of a medicament for the prevention or treatment of a spinocerebellar ataxic disorder or Friedrich's ataxia.

~~In still another aspect, t~~The present ~~invention~~ disclosure also provides a method of treating, or reducing the risk of, an ataxic disorder comprising administering to a patient in need thereof a therapeutically effective amount of a compound of formula (I) or a pharmaceutically acceptable salt thereof as hereinbefore defined.

In a further aspect of the present disclosure, there is provided a use of a compound of formula (I) or a pharmaceutically acceptable salt thereof as hereinbefore defined in the manufacture of a medicament for the prevention or treatment of an ataxic disorder.

In another aspect of the present disclosure, there is provided a compound of formula (I) or a pharmaceutically acceptable salt thereof as hereinbefore defined, for use in treating, or reducing the risk of, an ataxic disorder.

In a further aspect of the present disclosure, there is provided a pharmaceutical composition comprising a compound of formula (I) or a pharmaceutically acceptable salt thereof as hereinbefore defined in association with a pharmaceutically acceptable adjuvant, diluent or carrier, for use in the prevention or treatment of an ataxic disorder.

In a further aspect of the present disclosure, there is provided a use of a pharmaceutical composition comprising a compound of formula (I) or a pharmaceutically acceptable salt thereof as hereinbefore defined in association with a pharmaceutically acceptable adjuvant, diluent or carrier, in the manufacture of a medicament for the prevention or treatment of an ataxic disorder.

The compounds of formula (I) and pharmaceutically acceptable salts thereof are known from WO 2013/027000, the entire contents of which are incorporated herein by reference.

Compounds of formula (I) have been found from radio-imaging assays to be concentrated in the cerebellar region of the brain and, consequently, to exert their effects primarily in the cerebellum and spinal cord. Therefore, the compounds of formula (I) have the potential to be useful in the treatment of ataxic disorders.

Pharmaceutically acceptable salts of the compounds of formula (I) include acid addition salts which may be formed with inorganic acids and organic acids, e.g., acetate,

C_1-C_6 alkylcarbonyloxy, C_1-C_6 alkoxycarbonyl, amino, -CON(R^6)$_2$, C_1-C_6 alkylamino, di-(C_1-C_6 alkyl)amino, C_3-C_6 cycloalkyl, C_3-C_6 cycloalkyloxy, C_3-C_6 cycloalkylmethyl, -[O]$_p$-(CH_2)$_q$-O-R^7 and a 4- to 6-membered saturated or unsaturated heterocyclic ring (optionally substituted with at least one substituent selected from C_1-C_4 alkyl and C_1-C_4 alkoxy);

each R^6 independently represents a hydrogen atom or a C_1-C_6 alkyl group;

p is 0 or 1;

q is 1, 2, 3 or 4; and

R^7 represents a C_1-C_6 alkyl group;

for use in the prevention or treatment of an ataxic disorder.

2. A compound according to claim 1 ~~which is used~~ for use in the prevention or treatment of a spinocerebellar ataxic disorder or Friedrich's ataxia.

3. A compound according to claim 1 or claim 2 selected from

4-Hydroxy-6-(2-phenylethyl)pyridazin-3(2H)-one;

6-[2-(4-Fluorophenyl)ethyl]-4-hydroxypyridazin-3(2H)-one;

4-Hydroxy-6-{2-[5-(trifluoromethyl)pyridin-2-yl]ethyl}pyridazin-3(2H)-one;

6-[(4-Chlorobenzyl)sulfanyl]-4-hydroxypyridazin-3(2H)-one;

4-Hydroxy-6-{2-[6-(trifluoromethyl)pyridin-3-yl]ethyl}pyridazin-3(2H)-one;

6-[2-(3-Fluorophenyl)ethyl]-4-hydroxypyridazin-3(2H)-one;

6-[2-(2-Fluorophenyl)ethyl]-4-hydroxypyridazin-3(2H)-one;

6-[2-(3,5-Difluorophenyl)ethyl]-4-hydroxypyridazin-3(2H)-one;

6-[2-(3,4-Difluorophenyl)ethyl]-4-hydroxypyridazin-3(2H)-one;

4-Hydroxy-6-{2-[3-(trifluoromethoxy)phenyl]ethyl}pyridazin-3(2H)-one;

4-Hydroxy-6-{2-[3-(trifluoromethyl)phenyl]ethyl}pyridazin-3(2H)-one;

4-Hydroxy-6-{2-[5-(trifluoromethyl)pyridin-3-yl]ethyl}pyridazin-3(2H)-one;

6-(2-Cyclohexylethyl)-4-hydroxypyridazin-3(2H)-one;

6-(2-Cyclopropylethyl)-4-hydroxypyridazin-3(2H)-one;

6-(2-Cyclopentylethyl)-4-hydroxypyridazin-3(2H)-one;

4-Hydroxy-6-[2-(4-methoxycyclohexyl)ethyl]pyridazin-3(2H)-one;

7. A compound of formula (I) according to any one of claims 1 to 3 which is 6-(2-Fluorobenzyl)-4-hydroxypyridazin-3(2H)-one.

8. Use of a compound according to any one of claims 1 to 7 in the manufacture of a medicament for the prevention or treatment of a spinocerebellar ataxic disorder or Friedrich's ataxia.

~~8~~9. Use of a compound of formula (I) or a pharmaceutically acceptable salt thereof as defined in claim 1 in the manufacture of a medicament for ~~use in~~ the prevention or treatment of an ataxic disorder.

~~9~~10. A compound of formula (I) or a pharmaceutically acceptable salt thereof as defined in claim 1, for use in ~~method of~~ treating, or reducing the risk of, an ataxic disorder ~~comprising administering to a patient in need thereof a therapeutically effective amount of a compound of formula (I) or a pharmaceutically acceptable salt thereof as defined in claim 1~~.

~~10~~11. A pharmaceutical composition comprising a compound of formula (I) or a pharmaceutically acceptable salt thereof as defined in claim 1 in association with a pharmaceutically acceptable adjuvant, diluent or carrier, for use in the prevention or treatment of an ataxic disorder.

~~11~~12. A pharmaceutical composition according to claim ~~10~~11 which further comprises D-serine, D-serine ethyl ester, D-cycloserine, amantadine, amantadine hydrochloride, buspirone, acetazolamide, topiramate, divalproex sodium, L-dopa, propranolol, primidone, clonazepam, levetiracetam, carbamazepine, gabapentin, baclofen, ondansetron, tizanidine or pramipexole.

~~12~~13. Use of a pharmaceutical composition comprising a compound of formula (I) or a pharmaceutically acceptable salt thereof as defined in claim 1 in association with a pharmaceutically acceptable adjuvant, diluent or carrier, in the manufacture of a medicament for ~~use in~~ the prevention or treatment of an ataxic disorder.

~~13~~14. Use according to claim ~~12~~13 wherein the pharmaceutical composition further comprises D-serine, D-serine ethyl ester, D-cycloserine, amantadine, amantadine hydrochloride, buspirone, acetazolamide, topiramate, divalproex sodium, L-dopa, propranolol, primidone, clonazepam, levetiracetam, carbamazepine, gabapentin, baclofen, ondansetron, tizanidine or pramipexole.

14. A method of treating, or reducing the risk of, an ataxic disorder comprising administering to a patient in need thereof a therapeutically effective amount of a pharmaceutical composition comprising a compound of formula (I) or a pharmaceutically acceptable salt thereof as defined in claim 1 in association with a pharmaceutically acceptable adjuvant, diluent or carrier.

15. A method according to claim 14 wherein the pharmaceutical composition further comprises D-serine, D-serine ethyl ester, D-cycloserine, amantadine, amantadine hydrochloride, buspirone, acetazolamide, topiramate, divalproex sodium, L-dopa, propranolol, primidone, clonazepam, levetiracetam, carbamazepine, gabapentin, baclofen, ondansetron, tizanidine or pramipexole.