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(54) Title **USE OF RILUZOLE PRODRUGS TO TREAT ATAXIAS**

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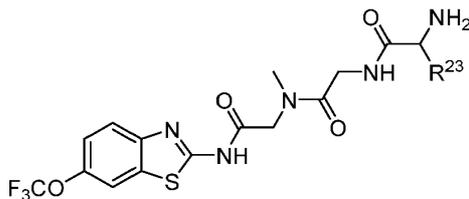
Enclosed is a translation of the patent claims in Norwegian. Please note that as per the Norwegian Patents Acts, section 66i the patent will receive protection in Norway only as far as there is agreement between the translation and the language of the application/patent granted at the EPO. In matters concerning the validity of the patent, language of the application/patent granted at the EPO will be used as the basis for the decision. The patent documents published by the EPO are available through Espacenet (<http://worldwide.espacenet.com>) or via the search engine on our website here: <https://search.patentstyret.no/>

**Patentkrav**

**1.** En riluzol-prodrome for anvendelse i behandlingen av ataksi i en pasient med behov for det, hvor

5 ataksien er spinocerebellær ataksi valgt fra SCA1, SCA2, SCA3, SCA6, SCA7, SCA8 og SCA10; og

riluzol-prodromen er i henhold til den følgende formelen:



10 og et farmasøytisk akseptabelt salt derav, hvor:

R<sub>23</sub> er valgt fra gruppen som består av H, CH<sub>3</sub>, CH<sub>2</sub>CH<sub>3</sub>, CH<sub>2</sub>CH<sub>2</sub>CH<sub>3</sub>, CH<sub>2</sub>CCH<sub>3</sub>, CH(CH<sub>3</sub>)<sub>2</sub>, CH<sub>2</sub>CH(CH<sub>3</sub>)<sub>2</sub>, CH(CH<sub>3</sub>)CH<sub>2</sub>CH<sub>3</sub>, CH<sub>2</sub>OH, CH<sub>2</sub>OCH<sub>2</sub>Ph, CH<sub>2</sub>CH<sub>2</sub>OCH<sub>2</sub>Ph, CH(OH)CH<sub>3</sub>, CH<sub>2</sub>Ph, CH<sub>2</sub>(sykloheksyl), CH<sub>2</sub>(4-OH-Ph), (CH<sub>2</sub>)<sub>4</sub>NH<sub>2</sub>, (CH<sub>2</sub>)<sub>3</sub>NHC(NH<sub>2</sub>)NH<sub>2</sub>, CH<sub>2</sub>(3-indol), CH<sub>2</sub>(5-imidazol), CH<sub>2</sub>CO<sub>2</sub>H, CH<sub>2</sub>CH<sub>2</sub>CO<sub>2</sub>H, CH<sub>2</sub>CONH<sub>2</sub>, og CH<sub>2</sub>CH<sub>2</sub>CONH<sub>2</sub>.

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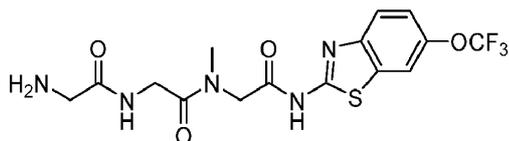
**2.** Riluzol-prodromen for anvendelse i henhold til krav 1, hvor ataksien er en ataksi assosiert med translaterede GAG-gjentatte ekspansjoner.

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**3.** Riluzol-prodromen for anvendelse i henhold til krav 1, hvor ataksien er en ataksi assosiert med ikke-translaterte gjentatte ekspansjoner i ikke-kodende områder.

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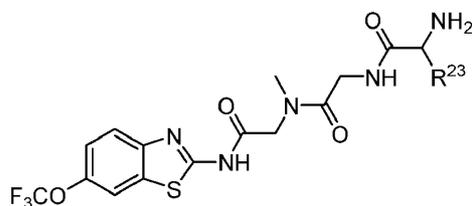
**4.** Riluzol-prodromen for anvendelse i henhold til et hvilket som helst av de foregående kravene, hvor riluzol-prodromen har den følgende formelen:



**5.** Et sett for anvendelse i behandlingen av en pasient rammet av ataksi, hvor ataksien er spinocerebellær ataksi valgt fra SCA1, SCA2, SCA3, SCA6, SCA7, SCA8 og SCA10, hvor settet omfatter:

(a) en riluzol-prodroge som har den følgende formelen:

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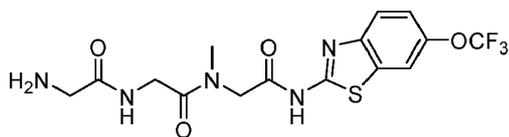
eller et farmasøytisk akseptabelt salt derav, hvor:

$R_{23}$  er valgt fra gruppen som består av H,  $CH_3$ ,  $CH_2CH_3$ ,  
 10  $CH_2CH_2CH_3$ ,  $CH_2CCH_3$ ,  $CH(CH_3)_2$ ,  $CH_2CH(CH_3)_2$ ,  $CH(CH_3)CH_2CH_3$ ,  
 $CH_2OH$ ,  $CH_2OCH_2Ph$ ,  $CH_2CH_2OCH_2Ph$ ,  $CH(OH)CH_3$ ,  $CH_2Ph$ ,  
 $CH_2$ (sykloheksyl),  $CH_2$ (4-OH-Ph),  $(CH_2)_4NH_2$ ,  $(CH_2)_3NHC(NH_2)NH$ ,  
 $CH_2$ (3-indol),  $CH_2$ (5-imidazol),  $CH_2CO_2H$ ,  $CH_2CH_2CO_2H$ ,  $CH_2CONH_2$ ,  
 og  $CH_2CH_2CONH_2$ ; og

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(b) et annet terapeutisk middel.

**6.** Settet for anvendelse ifølge krav 5, hvor riluzol-prodrogen har den følgende formelen:



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