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(54) Title **TREATMENT OF MITOCHONDRIAL DISEASES**

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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 sammensetning omfattende deoksyadenosin, deoksyguanosin, deoksycytidin, og deoksytymidin til anvendelse for å behandle et mitokondrielt DNA-deplesjons- og/eller
5 delesjonssyndrom, der syndromet skyldes en defekt i DNA polymerase subenheten gamma-1 (POLG1)-protein.
2. Sammensetningen til anvendelse ifølge krav 1, der det mitokondrielle DNA deplesjons- og delesjonssyndromet er forårsaket av en defekt i den mitokondrielle replikasjonsveien,
10 nevnte defekt skyldes en eller flere av følgende mutasjoner i POLG1-proteinet: R309C, W748S, V1177L, G848S, E1143, og E1143G, posisjonene er referert til med hensyn til SEQ ID NO:1.
3. Sammensetningen til anvendelse ifølge et hvilket som helst av kravene 1 eller 2, som
15 ikke inneholder noe annet ytterligere nukleosid.
4. Sammensetningen til anvendelse ifølge et hvilket som helst av de foregående kravene, der sammensetningen videre omfatter en eller flere farmasøytisk akseptable inhibitorer av nukleosid-nedbrytning.
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5. Sammensetningen til anvendelse ifølge krav 4, der den farmasøytisk akseptable inhibatoren er en inhibitor av deoksyadenosin-nedbrytning.
6. Sammensetningen til anvendelse ifølge krav 5, der inhibatoren er erytro-
25 9-(2-hydroksy-3-nonyl)adenin (EHNA).