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| (54) | Title             | <b>NITROGEN SCAVENGING DRUG FOR USE IN A METHOD OF TREATING A NITROGEN RETENTION DISORDER</b>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                    |
| (56) | References Cited: | US-A1- 2008 119 554<br>US-A1- 2010 008 859<br>US-B1- 6 219 567<br>UTA LICHTER-KONECKI ET AL: "Ammonia control in children with urea cycle disorders (UCDs); Phase 2 comparison of sodium phenylbutyrate and glycerol phenylbutyrate", MOLECULAR GENETICS AND METABOLISM, ACADEMIC PRESS, AMSTERDAM, NL, vol. 103, no. 4, 21 April 2011 (2011-04-21), pages 323-329, XP028249731, ISSN: 1096-7192, DOI: 10.1016/J.YMGME.2011.04.013 [retrieved on 2011-05-05]<br>ENNS ET AL.: 'Survival after Treatment with Phenylacetate and Benzoate for Urea-Cycle Disorders' N ENGL J MED vol. 356, no. 22, 31 May 2007, XP055148817<br>LEE ET AL.: 'Phase 2 Comparison of A Novel Ammonia Scavenging Agent with Sodium Phenylbutyrate in Patients with Urea Cycle Disorders: Safety, Pharmacokinetics, and Ammonia Control.' MOL. GENET. METAB. vol. 100, no. 3, July 2010, XP027079433<br>LICHTER-KONECKI ET AL.: 'Ammonia Control with Urea Cycle Disorders (UCDs); Phase 2 comparison of sodium phenylbutyrate and glycerol phenylbutyrate.' MOL. GENET. METAB. vol. 103, no. 4, 05 May 2011, pages 323-329, XP028249731 |

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.** Glyseryl tri-[4-fenylbutyrat] (HPN-100) for anvendelse i en fremgangsmåte til behandling av en nitrogenretensjonssykdom i et individ,
- hvor fremgangsmåten omfatter å justere dosen av glyseryl tri-[4-fenylbutyrat] (HPN-10) for å oppnå et fastende blodammoniakknivå som er mindre enn eller lik halvdelen av den øvre grense av normal (ULN) for blodammoniakk, hvor dosejusteringen omfatter:
- (a) å måle fastende blodammoniakk for individet;
- (b) å sammenligne det fastende blodammoniakknivå med ULN for blodammoniakk; og
- (c) å administrere til individet en øket dose av glyseryl tri-[4-fenylbutyrat] (HPN-100) dersom det fastende blodammoniakknivå har en verdi som er større enn halvparten av ULN.
- 2.** Glyseryl tri-[4-fenylbutyrat] (HPN-100) for anvendelse ifølge krav 1, hvor dosejusteringen ytterligere omfatter trinnet å bestemme den øvre normalgrensen for blodammoniakknivået for individet før trinn (b).
- 3.** Glyseryl tri-[4-fenylbutyrat] (HPN-100) for anvendelse ifølge krav 1, hvor den øvre normalgrense for blodammoniakknivået er 35 µmol/l.
- 4.** Glyseryl tri-[4-fenylbutyrat] (HPN-100) for anvendelse ifølge krav 1, hvor dosejusteringen ytterligere omfatter:
- (d) å måle fenylacetylglutamin (PAGN)-ekskresjon i urin; og
- (e) å bestemme en effektiv dose av glyseryl tri-[4-fenylbutyrat] (HPN-100) basert på en gjennomsnittlig omdannelse av glyseryl tri-[4-fenylbutyrat] (HPN-100) til PAGN i urin på 60-75%.