



(12) Translation of
European patent specification

(11) NO/EP 2977057 B1

NORWAY

(19) NO

(51) Int Cl.

C12N 9/18 (2006.01)

A61K 31/465 (2006.01)

A61K 31/135 (2006.01)

A61K 38/46 (2006.01)

A61K 31/366 (2006.01)

A61P 3/06 (2006.01)

A61K 31/397 (2006.01)

Norwegian Industrial Property Office

(45)	Translation Published	2020.02.17
(80)	Date of The European Patent Office Publication of the Granted Patent	2019.11.06
(86)	European Application Nr.	14200334.2
(86)	European Filing Date	2011.09.09
(87)	The European Application's Publication Date	2016.01.27
(30)	Priority	2010.09.09, US, 403011 P 2010.10.29, US, 456014 P 2011.01.13, US, 201161432372 P 2011.04.23, WO, PCT/US11/033699
(84)	Designated Contracting States:	AL ; AT ; BE ; BG ; CH ; CY ; CZ ; DE ; DK ; EE ; ES ; FI ; FR ; GB ; GR ; HR ; HU ; IE ; IS ; IT ; LI ; LT ; LU ; LV ; MC ; MK ; MT ; NL ; NO ; PL ; PT ; RO ; RS ; SE ; SI ; SK ; SM ; TR
(62)	Divided application	EP2613798, 2011.09.09
(73)	Proprietor	Alexion Pharmaceuticals, Inc., 121 Seaport Boulevard, Boston, MA 02210, USA
(72)	Inventor	Quinn, Anthony, 107 Manet Road, Chestnut Hill, MA Massachusetts 02467, USA
(74)	Agent or Attorney	ZACCO NORWAY AS, Postboks 2003 Vika, 0125 OSLO, Norge

(54)	Title	ISOLATED RECOMBINANT HUMAN N-GLYCOSYLATED LYSOSOMAL ACID LIPASE
(56)	References Cited:	US-A1- 2009 297 496, WO-A1-01/56596 IKEDA S ET AL: "Production of recombinant human lysosomal acid lipase in Schizosaccharomyces pombe: Development of a fed-batch fermentation and purification process", JOURNAL OF BIOSCIENCE AND BIOENGINEER, ELSEVIER, AMSTERDAM, NL, vol. 98, no. 5, 1 January 2004 (2004-01-01), pages 366-373, XP004695391, ISSN: 1389-1723 ANDERSON R ET AL: "Cloning and expression of cDNA encoding human lysosomal acid lipase/cholestreryl ester hydrolase", JOURNAL OF BIOLOGICAL CHEMISTRY, AMERICAN SOCIETY FOR BIOCHEMISTRY AND MOLECULAR BIOLOGY, US, vol. 266, no. 33, 25 November 1991 (1991-11-25), pages 22479-22484, XP002192752, ISSN: 0021-9258 Ag Quinn ET AL: "SBC-102, a recombinant enzyme replacement therapy, corrects key abnormalities due to lysosomal acid lipase deficiency.", , 2 November 2010 (2010-11-02), XP055399695, Washington DC Retrieved from the Internet: URL: http://www.ashg.org/2010meeting/abstracts/fulltext/f20205.htm [retrieved on 2017-08-18]

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

- 5 **1.** Isolert rekombinant human lysosomal sur lipase (rhLAL) som inneholder N-bundne glykaner knyttet til Asn15, Asn80, Asn140, Asn252 og Asn300, som angitt i SEQ ID NO: 1, hvori de N-bundne glykanene omfatter bi-, tri- og tetra-antennære strukturer med N-acetylglukosamin (GlcNAc), mannose og/eller manose-6-fosfat (M6P), og hvori rhLAL-en omfatter M6P-inneholdende glykaner ved Asn80, Asn140 og Asn252.
- 10 **2.** Den isolerte rhLAL-en ifølge krav 1, hvori rhLAL-en ikke inneholder sialinsyre.
- 3.** Den isolerte rhLAL-en ifølge krav 1 eller krav 2, hvori rhLAL-ens molekylvekt er omtrent 55 kD.
- 15 **4.** Den isolerte rhLAL-en ifølge et hvilket som helst av kravene 1 til 3, hvori rhLAL-ens spesifikke aktivitet er minst omtrent 100 U/mg.
- 5.** Den isolerte rhLAL-en ifølge et hvilket som helst av kravene 1 til 4, hvori rhLAL-en ikke omfatter O-bundne glykaner.
- 20 **6.** Den isolerte rhLAL-en ifølge et hvilket som helst av kravene 1 til 5, hvori rhLAL-ens Asn51 ikke er glykosylert.
- 7.** Den isolerte rhLAL-en ifølge et hvilket som helst av kravene 1 til 6, hvori rhLAL-en har halveringstid i serum ($t_{1/2}$) som er mindre enn 40 minutter.
- 25