

July 29, 2015

STATEMENT ON A NONPROPRIETARY NAME ADOPTED BY THE USAN COUNCIL

USAN (CD-177) REVEGLUCOSIDASE ALFA

PRONUNCIATION rev" e gloo koe' si dase

THERAPEUTIC CLAIM Treatment of Pompe disease

CHEMICAL NAMES

1. 7-67-Insulin-like growth factor II [7-alanine] (human) fusion protein with peptide (synthetic spacer GAP) fusion protein with 70-952- α -glucosidase (human)
2. des-(2-7)-human insulin-like growth factor II fusion protein with glycyl-L-alanyl-L-prolyl-human lysosomal alpha-glucosidase (acid maltase, aglucosidase alfa) produced in CHO cells, glycoform alfa

STRUCTURAL FORMULA

Sequence

ALCGGELVDT	LQFVCGDRGF	YFSRPASRVS	RRSRGIVEEC	CFRSCDLALL	50
ETYCATPAKS	EGAPAHGPRP	RAVPTQCDVP	PNSRFDCAPD	KAITQEQCEA	100
RGCCYIPAKQ	GLQGAQMGP	WCFFPPSYPS	YKLENLSSE	MGYTATLTRT	150
TPTFPPKDIL	TLRLDVMET	ENRLHFTIKD	PANRRYEVL	ETPHVHSRAP	200
SPLYSVEFSE	EPFGVIVHRQ	LDGRVLLNTT	VAPLFFADQF	LQLSTSLPSQ	250
YITGLAEHLS	PLMLSTSWTR	ITLWNRDLAP	TPGANLYGSH	PFYLALEDGG	300
SAHGVFLLNS	NAMDVVLQPS	PALSWRSTGG	ILDVYIFLGP	EPKSVVQQYL	350
DVVGYPFMP	YWGLGFHLR	WGYSSAITR	QVVENMTRAH	FPLDVQWNDL	400
DYMDSRRDFT	FNKDGFRDFP	AMVQELHGG	RRYMMIVDPA	ISSSGPAGSY	450
RPYDEGLRRG	VFITNETGQP	LIGKVWPGST	AFPDTNPTA	LAWWEDMVAE	500
FHDQVPFDGM	WIDMNEPSNF	IRGSEDGCPN	NELENPPYVP	GVVGGTLQAA	550
TTCASSHQFL	STHYNLHNL	GLTEAIASHR	ALVKARGTRP	FVISRSTFAG	600
HGRYAGHWG	DVWSSWEQLA	SSVPEILQFN	LLGVPLVGAD	VCGFLGNTSE	650
ELCVRWTQLG	AFYPPFMRNHN	SLLSLPQEPY	SFSEPAQQAM	RKALTLYAL	700
LPHLYTLFHQ	AHVAGETVAR	PLFLEFPKDS	STWTVDHQLL	WGEALLITPV	750
LQAGKAEVTG	YFPLGTWYDL	QTVPIEALGS	LPPPPAAPRE	PAIHSEGQWV	800
TLPAPLDTIN	VHLRAGYIIP	LQGPGLTTTE	SRQQPMALAV	ALTKGGEARG	850
ELFWDDGESL	EVLERGAYTQ	VIFLARNTI	VNELVRVTSE	GAGLQLQKVT	900
VLGVATAPQQ	VLSNGVPVSN	FTYSPDTKVL	DICVSLLMGE	QFLVSWC	947

Disulfide bridges

3-41 15-54 40-45 77-103 87-104 98-122 528-553 642-653 933-947

Glycosylation sites (N)

Asn-135 Asn-228 Asn-465 Asn-487 Asn-647 Asn-877 Asn-920

MOLECULAR FORMULA $C_{4735}H_{7189}N_{1261}O_{1371}S_{38}$ MOLECULAR WEIGHT 104.93 kDa (peptide)
118 kDa (glycosylated protein)

TRADEMARK None as yet

SPONSOR	BioMarin Pharmaceutical, Inc.
CODE DESIGNATIONS	BMN 701
<u>CAS</u> REGISTRY NUMBER	1446198-96-4
UNII	E86R97M8XQ
WHO NUMBER	9994

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