

June 29, 2016

STATEMENT ON A NONPROPRIETARY NAME ADOPTED BY THE USAN COUNCIL

USAN (DE-52) LESINIDASE ALFA

PRONUNCIATION le sin' i dase al fa

THERAPEUTIC CLAIM Treatment of Mucopolysaccharidosis IIIB (MPS IIIB; Sanfilippo B Syndrome)

CHEMICAL NAMES

1. α -*N*-Acetylglucosaminidase
2. recombinant human alpha-*N*-acetylglucosaminidase

STRUCTURAL FORMULA

Amino acid sequence

DEAREAAAVR	ALVARLLGPG	PAADFSVSVE	RALAAKPLD	TYSLGGGGAA	50
RVRVRGSTGV	AAAAGLHRYL	RDFCGCHVAW	SGSQLRLPRP	LPAVPGELTE	100
ATPNRYRYIQ	NVCTQSYSFV	WWDWARWERE	IDWMALNGIN	LALAWSGQEA	150
IWQRVYLALG	LTQAEINEFF	TGPAFLAWGR	MGNLHTWDGP	LPPSWHIKQL	200
YLQHRVLDQM	RSFGMTPVLP	AFAGHVPEAV	TRVFPQVNVN	KMGSWGHFNC	250
SYSCSFLAP	EDPIFPIIGS	LFLRELIKEF	GTDHIYGADT	FNEMQPPSSE	300
PSYLAATA	VYEAMTAVDT	EAVWLLQGWL	FQHQPQFWGP	AQIRAVLGAV	350
PRGRLVLDDL	FAESQPVYTR	TASFQGQPFI	WCMLHNFVGN	HGLFGALEAV	400
NGGPEEARLF	PNSTMVGTGM	APEGISQNEV	VYSLMAELGW	RKDPVVDLAA	450
WVTSFAARRY	GVSHPDAGAA	WRLLLRSVYN	CSGEACRGHN	RSPLVRRPSL	500
QMNTSIWYNR	SDVFEAWRL	LTSAPSLATS	PAFRYDLDL	TRQAVQELVS	550
LYYEEARSAY	LSKELASLLR	AGGVLAYELL	PALDEVLASD	SRFLLGSWLE	600
QARAAVSE	EADFYEQNSR	YQLTLWGPEG	NILDYANKQL	AGLVANYTTP	650
RWRLFLEALV	DSVAQGIPFQ	QHQFDKNVFQ	LEQAFVLSKQ	RYSQPRGDT	700
VDLAKKIFLK	YYPRWVAGSW				720

Disulfide bridges

250-254 481-486

Glycosylation sites (N)

Asn-238 Asn-249 Asn-412 Asn-480 Asn-503 Asn-509

MOLECULAR FORMULA $C_{3660}H_{5527}N_{991}O_{1018}S_{20}$ (peptide)

MOLECULAR WEIGHT 80.34 kDa (peptide)

TRADEMARK None as yet

SPONSOR Alexion Pharmaceuticals Inc.

CODE DESIGNATIONS SBC-103

CAS REGISTRY NUMBER 1522433-40-4

UNII 61A4G0P3IB

WHO NUMBER 10449

gbk