

April 29, 2015

STATEMENT ON A NONPROPRIETARY NAME ADOPTED BY THE USAN COUNCIL

USAN (CD-91)

CERLIPONASE ALFA

PRONUNCIATION

ser lip' oh nase al fa

THERAPEUTIC CLAIM

Treatment of neurological genetic disorders

CHEMICAL NAMES

1. Aminopeptidase, tripeptidyl, I
2. immature human tripeptidyl-peptidase 1 (cell growth-inhibiting gene 1 protein, lysosomal pepstatin-insensitive protease, TPP-1, EC 3.4.14.9), 544 residues protein, produced in Chinese hamster ovary (CHO) cells, glycoform alfa

STRUCTURAL FORMULA

Sequence

SYSPEPDQRR	TLPPGWVSLG	RADPEEELS	TFALRQQNVE	RLSELVQAVS	50
DPSSPQYGKY	LTLNVADLV	RPSPLTLHTV	QKWLAAAGAQ	KCHSVITQDF	100
LTCWLSIRQA	ELLLPGAEPH	HYVGGPTETH	VVRSPHPYQL	PQALAPHVDF	150
VGGLHRFPPT	SSLRQRPEPQ	VTGTVGLHLG	VTPSVIRKRY	NLTSQDVGSG	200
TSNNSQACAQ	FLEQYFHDS	LAQFMRLF	NFAHQASVAR	VVGQQGRGRA	250
GIEASLDVQY	LMSAGANIST	WVYSSPGRHE	GQEPFLQWLM	LLSNESALPH	300
VHTVSYGDDE	DSLSSAYIQR	VNTELMKAAA	RGLTLLFASG	DSGAGCWSVS	350
GRHQFRPTFP	ASSPYVTTVG	GTSFQEPFLI	TNEIVDYISG	GGFSNVFPRP	400
SYQEEAVTKF	LSSSPHLPPS	SYFNASGRAY	PDVAALSDGY	WVVSNRVPIP	450
WVSGTSASTP	VFGGILSLIN	EHRILSGRPP	LGFLNRLYQ	QHGAGLFDVT	500
RGCHESCLDE	EVEGQGFCSG	PGWDPVTGWG	TPNFPALLKT	LLNP	544

Disulfide bridges location

92-103 346-507 503-518

Glycosylation sites

Asn-191 Asn-203 Asn-267 Asn-294 Asn-424

MOLECULAR FORMULA

 $C_{2657}H_{4042}N_{734}O_{793}S_{11}$

MOLECULAR WEIGHT

59.31 kDa (protein)

TRADEMARK

None as yet

SPONSOR

BioMarin Pharmaceutical Inc.

CODE DESIGNATIONS

BMN 190

CAS REGISTRY NUMBER

151662-36-1

UNII

X8R2D92QP1

WHO NUMBER 9970

SCS