

January 25, 2012

## STATEMENT ON A NONPROPRIETARY NAME ADOPTED BY THE USAN COUNCIL:

USAN (bc-46) SEBELIPASE ALFA

PRONUNCIATION se bel' ip ase

THERAPEUTIC CLAIM Enzyme replacement therapy

## CHEMICAL NAMES

1. Esterase, cholesterol (human gene LIPA)
2. Human lysosomal acid lipase/cholesteryl ester hydrolase (cholesteryl esterase, lipase A, EC=3.1.1.13) glycosylated (produced in transgenic *Gallus*)

## STRUCTURAL FORMULA

SGGKLTAVDP	ETNMNVSEII	SYWGFPSSEY	LVETEDGYIL	CLNRIPHGRK	50
NHSDKGPKPV	VFLQHGLLAD	SSNWVTNLAN	SSLGFILADA	GFDVVMGNSR	100
GNTWSRKHKT	LSVSQDEFWA	FSYDEMAKYD	LPASINFILN	KTGQEQVYV	150
GHSQGTTFGF	IAFSQIPELA	KRIKMFALG	PVASVAFCTS	PMAKLGRLPD	200
HLIKDLFGDK	EFLPQSAFLK	WLGTHVCTHV	ILKELCGNLC	FLLCGFNERN	250
LNM <sup>U</sup> SRVDVYT	THSPAGTSVQ	NMLHWSQAVK	FQKFQAFDWG	SSAKNYFHYN	300
QSYPTYNVK	DMLVPTAVWS	GGHDWLADVY	DVNILLTQIT	NLVPHEIPE	350
WEHLDFIWGL	DAPWRLYNKI	INLMRKYQ			378

Glycosylation sites (N)

Asn-15 Asn-80 Asn-140 Asn-252 Asn-300

## MOLECULAR FORMULA

C<sub>1968</sub>H<sub>2945</sub>N<sub>507</sub>O<sub>551</sub>S<sub>15</sub>

## MOLECULAR WEIGHT

43.00 kDa (protein), 53 kDa (glycosylated)

## TRADEMARK

None as yet

## SPONSOR

Synageva BioPharma Corp.

## CODE DESIGNATION

SBC-102

CAS REGISTRY NUMBER

1276027-63-4

## UNII

K4YTU42T8G

## WHO NUMBER

9594

gbk