Recombinant Human GAA therapeutic protein(Alglucosidase alfa)

Cat. No. GAA-P026H **Lot. No.** (See product label)

SPECIFICATION

Product Overview	Recombinant Human acid alpha-glucosidase therapeutic protein consists of the human enzyme acid alpha-glucosidase (GAA) which is essential for the degradation of glygogen to glucose in lysosomes. It is encoded by the most predominant of nine observed haplotypes of this gene. It is produced by recombinant DNA technology in a Chinese hamster ovary cell line. The protein degrades glycogen by catalyzing the hydrolysis of a-1,4- and a-1,6- glycosidic linkages of lysosomal glycogen. Structurally, it is a glycoprotein with a calculated mass of 98,008 daltons for the 883 residue mature polypeptide chain, and a total mass of approximately 109,000 daltons, including carbohydrates. It is used for the treatment of Pompe disease (GAA deficiency) in infants and pediatric patients.
Species	Human
Source	CHO
Description	This gene encodes lysosomal alpha-glucosidase, which is essential for the degradation of glycogen to glucose in lysosomes. The encoded preproprotein is proteolytically processed to generate multiple intermediate forms and the mature form of the enzyme. Defects in this gene are the cause of glycogen storage disease II, also known as Pompe's disease, which is an autosomal recessive disorder with a broad clinical spectrum. Alternative splicing results in multiple transcript variants.
Molecular Mass	105270.802

Tel: 1-631-559-9269 1-516-512-3133
Email: info@creative-biomart.com Fax: 1-631-938-8127
45-1 Ramsey Road, Shirley, NY 11967, USA

AA Sequence	AHPGRPRAVPTQCDVPPNSRFDCAPDKAITQEQCEARGCCYIPAKQGLQGAQMGQ PWCFFPPSYPSYKLEN LSSSEMGYTATLTRTTPTFFPKDILTLRLDVMMETENRLHF TIKDPANRRYEVPLETPHVHSRAPSPLYSV EFSEEPFGVIVRRQLDGRVLLNTTVAP LFFADQFLQLSTSLPSQYITGLAEHLSPLMLSTSWTRITLWNRD LAPTPGANLYGSH PFYLALEDGGSAHGVFLLNSNAMDVVLQPSPALSWRSTGGILDVYIFLGPEPKSVVQ Q YLDVVGYPFMPPYWGLGFHLCRWGYSSTAITRQVVENMTRAHFPLDVQWNDLD YMDSRRDFTFNKDGFRDF PAMVQELHQGGRRYMMIVDPAISSSGPAGSYRPYDE GLRRGVFITNETGQPLIGKVWPGSTAFPDFT NPTALAWWEDMVAEFHDQVPFDGM WIDMNEPSNFIRGSEDGCPNNELENPPYVPGVVGGTLQAATICASSH QFLSTHYNL HNLYGLTEAIASHRALVKARGTRPFVISRSTFAGHGRYAGHWTGDVWSSWEQLASS VPEILQ FNLLGVPLVGADVCGFLGNTSEELCVRWTQLGAFYPFMRNHNSLLSLPQE PYSFSEPAQQAMRKALTLRYA LLPHLYTLFHQAHVAGETVARPLFLEFPKDSSTWT VDHQLLWGEALLITPVLQAGKAEVTGYFPLGTWYDL QTVPVEALGSLPPPPAAPRE PAIHSEGQWVTLPAPLDTINVHLRAGYIIPLQGPGLTTTESRQQPMALAVA LTKGGE ARGELFWDDGESLEVLERGAYTQVIFLARNNTIVNELVRVTSEGAGLQLQKVTVLGV ATAP QQVLSNGVPVSNFTYSPDTKVLDICVSLLMGEQFLVSWC
Endotoxin	< 0.1 EU per μg of the protein
Purity	>95%
Alias	GAA; LYAG; Alglucosidase alfa

GENE INFORMATION

Gene Name	GAA glucosidase, alpha; acid [Homo sapiens]
Official Symbol	GAA
Synonyms	GAA; glucosidase, alpha; acid; lysosomal alpha-glucosidase; glycogen storage

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	disease type II; Pompe disease; acid maltase; aglucosidase alfa; LYAG;
Gene ID	2548
mRNA Refseq	NM_000152
Protein Refseq	NP_000143
MIM	606800
UniProt ID	P10253
Chromosome Location	17q25.2-q25.3
Pathway	Galactose metabolism, organism-specific biosystem; Galactose metabolism, conserved biosystem; Lysosome, organism-specific biosystem; Lysosome, conserved biosystem; Metabolic pathways, organism-specific biosystem; Notch-mediated HES/HEY network, organism-specific biosystem; Starch and sucrose metabolism, organism-specific biosystem;
Function	alpha-glucosidase activity; carbohydrate binding; hydrolase activity, hydrolyzing O-glycosyl compounds; maltose alpha-glucosidase activity;

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