

Recombinant Human GAA protein, GST-tagged

Cat. No. GAA-28090TH Lot. No. (See product label)

SPECIFICATION

Product Overview	Recombinant Human GAA(1 a.a. - 952 a.a.) fused with GST tag at N-terminal was expressed in Wheat Germ.
Species	Human
Source	Wheat Germ
ProteinLength	1-952 a.a.
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.
Form	50 mM Tris-HCl, 10 mM reduced Glutathione, pH=8.0 in the elution buffer.
Molecular Mass	131.7 kDa
AA Sequence	MGVRHPPCSHRLAVCALVSLATAALLGHILLHDFLLVPRELSGSSPVLEETHPAHQ QGASRPGPRDAQAHPGRP RAVPTQCDVPPNSRFDKAITQEQCEARGCCYIP AKQGLQGAQMGPWCFFPPSYPSYKLENLSSSEMGYTA TLTRTTPTFFPKDILTLR LDVMMETENRLHFTIKDPANRRYEVPLETPRVHSRAPSPLYSVEFSEEPFGVIVHRQ LDGRVLLNTTVAPLFFADQFLQLSTSLPSQYITGLAEHLSPLMLSTSWTRITLWNRDL

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APTPGANLYGSHPFYLA LEDGGS AHGVFLLSNAMDVVLQPSPALSWRSTGGILDV
 YIFLGPEPKSVVQQYLDVVGYPFMPYWG LGFHLCR WGYSSAITRQVVENMTRA
 HFPLDVQWNDLDYMDSRRDFTFNKDGFRDFPAMVQELHQGGRRYMMIVDPAISS
 G PAGSYRPYDEGLRRGVFITNETGQPLIGKVWPGSTAFPDTNPTALAWWEDMVA
 EFHDQVPFDGMWIDMNEPSNF IRGSEDGCPNNELENPPYVPGVVGGTLQAATICA
 SSHQFLSTHYNLHNLGLTEAIASHRALVKARGTRPFVISR STFAGHGRYAGHWGTG
 DVWSSWEQLASSVPEILQFNLLGVPLVGADVCGFLGNTSEELCVRWTQLGAFYPFM
 RNHN SLLSLPQEPYSFSEPAQQAMRKALTRYALLPHLYTLFHQAHVAGETVARPL
 FLEFPKDSSTWTVDHQLLWGEAL LITPVLQAGKAEVTGYFPLGTWYDLQTVPIEALG
 SLPPPPAAPREPAIHSEGQWVTL PQLDITINVHLRAGYIIP LQGPGLTTTESRQQPM
 ALAVALTKGGGEARGELFWDDGESLEVLERGAYTQVIFLARNTIVNELVRVTSEGAG
 LQ LQKVTVLGVATAPQQVLSNGVPSNFTYSPDTKVLDCVSLLMGEQFLVSWC

Applications Enzyme-linked Immunoabsorbent Assay; Western Blot (Recombinant protein);
 Antibody Production; Protein Array

Notes Best use within three months from the date of receipt of this protein.

Storage Store at -80 centigrade. Aliquot to avoid repeated freezing and thawing.

GENE INFORMATION

Gene Name [GAA glucosidase, alpha; acid \[Homo sapiens \]](#)

Official Symbol [GAA](#)

Synonyms GAA; glucosidase, alpha; acid; lysosomal alpha-glucosidase; glycogen storage
 disease type II; Pompe disease; acid maltase; aglucosidase alfa; LYAG;

Gene ID [2548](#)

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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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