

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 QGASRPGPRDAQAHGPRP RAVPTQCDVPPNSRFDCAPDKAITQEQCEARGCCYIP AKQGLQGAQMGPWCFFPPSYPSYKLENLSSEMGYTA TLTRTTPTFFPKDILTLR LDVMMETENRLHFTIKDPANRRYEVPLETPRVHSRAPSPLYSVEFSEEPFGVIVHRQ LDGRVLLNTTVAPLFFADQFLQLSTSLPSQYITGLAEHLSPLMLSTSWTRITLWNRDL

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APTPGANLYGSHPFYLA LEDGGS AHGVFLLSNAMDVVLQPSALSWRSTGGILDV
 YIFLGPEPKSVVQQYLDVVGYPFMPPIYWGLGFHLGR WGYSSTAITRQVVENMTRA
 HFPLDVQWNLDDYMDSRDFTFNKDGFRDFPAMVQELHQGGRRYMMIVDPAISS
 G PAGSYRPYDEGLRRGVFITNETGQPLIGKVWPGSTAFPDTNPTALAWWEDMVA
 EFHDQVPFDGMWIDMNEPSNF IRGSEDGCPNNELENPPYVPGVVGGTLQAATICA
 SSHQFLSTHYNLHNLGLTEAIAASHRALVKARGTRPFVISR STFAGHGRYAGHWGTG
 DVWSSWEQLASSVPEILQFNLLGVPLVGADVCGFLGNTSEELCVRWTQLGAFYPPM
 RNHN SLLSLPQEPYSFSEPAQQAMRKALTTRYALLPHLYTLFHQAHVAGETVARPL
 FLEFPKDSSTWTVDHQLLWGEAL LITPVLQAGKAEVTGYFPLGTWYDLQTVPIEALG
 SLPPPPAAPREPAIHSEGQWVTLPAPLDTINVHLRAGYIIP LQGPGLTTTESRQQPM
 ALAVALTKGGGEARGELFWDDGESLEVLERGAYTQVIFLARNTIVNELVRVTSEGAG
 LQ LQKVTVLGVATAPQQVLSNGVPVSNFTYSPDTKVLDICVSLLMGEQFLVSWC

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;