Recombinant Human GAA

Cat. No. GAA-28087TH **Lot. No.** (See product label)

SPECIFICATION

Product Overview	Recombinant fragment Human GAA with N-terminal proprietary tag. Predicted MW 36.85kDa.
Species	Human
Source	Wheat Germ
ProteinLength	102 amino acids
Description	This gene encodes acid alpha-glucosidase, which is essential for the degradation of glycogen to glucose in lysosomes. Different forms of acid alpha-glucosidase are obtained by proteolytic processing. Defects in this gene are the cause of glycogen storage disease II, also known as Pompes disease, which is an autosomal recessive disorder with a broad clinical spectrum. Three transcript variants encoding the same protein have been found for this gene.
Molecular Weight	36.850kDa inclusive of tags
Form	Liquid
Purity	Proprietary Purification
Storage buffer	pH: 8.00Constituents:0.3% Glutathione, 0.79% Tris HCl
Storage	Shipped on dry ice. Upon delivery aliquot and store at -80oC. Avoid freeze / thaw

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	cycles.
Sequence Similarities	Belongs to the glycosyl hydrolase 31 family.Contains 1 P-type (trefoil) domain.

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;
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;
Function	alpha-glucosidase activity; carbohydrate binding; hydrolase activity, hydrolyzing O-

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	glycosyl compounds; maltose alpha-glucosidase activity;

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