Active Recombinant Human GAA protein, His-tagged

Cat. No. GAA-173H **Lot. No.** (See product label)

SPECIFICATION

Product Overview	Recombinant Human GAA(Ala70-Cys952) fused with His tag at N-terminal was expressed in HEK293.
Species	Human
Source	HEK293
ProteinLength	70-952 a.a.
Description	Acid alpha-glucosidase (GAA) is an enzyme that is essential in the degradation of glycogen to glucose in the lysosome. Defects in GAA are the cause of glycogen storage disease II, also known as Pompe's disease, which is a rare autosomal recessive metabolic disorder that damages muscle and nerve cells throughout the body, primarily due to the accumulation of glycogen in the lysosome. Pompe disease occurs in babies, children, and adults who inherit a defective GAA gene and affects an estimated 5,000 to 10,000 people worldwide. Enzyme replacement therapy (ERT) is used to treat patients with this disease.
Predicted N Terminal	His
Form	Supplied as a 0.2 μm filtered solution in Tris, NaCl and Glycerol.
Bio-activity	Measured by its ability to release glucose from starch. The specific activity is >7,500 pmol/min/ug.

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Molecular Mass	Predicted Molecular Mass: 99 kDa;SDS-PAGE: 95-105 kDa, reducing conditions
Endotoxin	<1.0 EU per 1 μg of the protein by the LAL method.
Purity	>95%, by SDS-PAGE under reducing conditions and visualized by Colloidal Coomassie® Blue stain at 5 μg per lane.
Storage	Avoid repeated freeze-thaw cycles.6 months from date of receipt, -20 to -70 centigrade as supplied.3 months, -20 to -70 centigrade under sterile conditions after opening.

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
mRNA Refseq	NM_000152
Protein Refseq	NP_000143
MIM	606800
UniProt ID	P10253
Chromosome	17q25.2-q25.3

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Location	
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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