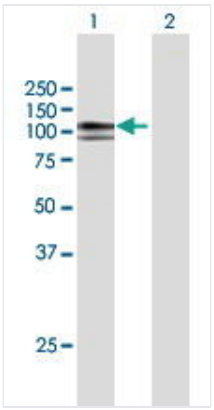
GAA Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	GAA, Polyclonal Antibody
Full Product Name	GAA (Lysosomal alpha-glucosidase, Acid Maltase, Aglucosidase alfa) (Biotin)
Product Gene Name	anti-GAA antibody
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Species Reactivity	Human, Mouse
Form/Format	Supplied as a liquid in PBS, pH 7.4. No preservative added. Labeled with Biotin.
Purity/Purification	Purified by Protein A Affinity Chromatography.
Specificity	Recognizes human GAA. Species Crossreactivity: mouse
Preparation and Storage	Store product at 4 degree C if to be used immediately within two weeks. For long-term storage, aliquot to avoid repeated freezing and thawing and stor
Molecular Weight	105,324 Da
OMIM	Y00839 mRNA
3D Structure	ModBase 3D Structure for P10253
Immunogen	Full length protein corresponding to aa1-952 from human GAA.
Immunogen Sequence	MGVRHPPCSHRLLAVCALVSLATAALLGHILLHDFLLVPRELSGSSPVLEETHPAHQQGASRPGPRDAQAHPGRPRAVPTQCDVPPNSRFDCAPDKAIT
Conjugate	Biotin
Note	Preservative Free
Product Categories/Family for anti-GAA antibody	Antibodies; Enzymes
Applications Tested/Suitable for anti-GAA antibody	Western Blot (WB)
Application Notes for anti-GAA antibody	Applications are based on unconjugated antibody.
Related Product Information for anti-GAA antibody	MBS6379122 is an antibody targeting GAA. Also known as GAA (Lysosomal alpha-glucosidase, Acid Maltase, Aglucosidase alfa) (Biotin). Reactive in H
Other Notes	Small volumes of anti-GAA antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessa



Western Blot (WB) of anti-GAA antibody
Western Blot analysis of GAA expression in transfected 293T cell line using 127076. Lane 1: GAA transfected lysate (105.3kD). Lane 2: Non-transfected lysate.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for GAA. It may not necessarily be applicable to this product.

NCBI GI #	119393891
NCBI GeneID	2548
NCBI Accession #	NP_000143.2
UniProt Primary Accession #	P10253
UniProt Secondary Accession #	Q09GN4; Q14351; Q16302; Q8IWE7
UniProt Related Accession #	P10253
GenBank Nucleotide #	NM_000152.2
NCBI Official Full Name	lysosomal alpha-glucosidase preproprotein
NCBI Official Synonym Full Names	glucosidase alpha, acid
NCBI Official Symbol	GAA
NCBI Official Synonym Symbols	LYAG
UniProt Protein Name	Lysosomal alpha-glucosidase
UniProt Synonym Protein Names	Acid maltase; Aglucosidase alfa
Protein Family	Lysosomal alpha-glucosidase
UniProt Gene Name	GAA
NCBI Protein Information	lysosomal alpha-glucosidase
NCBI Summary for GAA	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. [provided by RefSeq, Jan 2016]

UniProt Comments for GAA	GAA: Essential for the degradation of glygogen to glucose in lysosomes. Defects in GAA are the cause of glycogen storage disease type 2 (GSD2); also called acid alpha-glucosidase (GAA) deficiency or acid maltase deficiency (AMD). GSD2 is a metabolic disorder with a broad clinical spectrum. The severe infantile form, or Pompe disease, presents at birth with massive accumulation of glycogen in muscle, heart and liver. Cardiomyopathy and muscular hypotonia are the cardinal features of this form whose life expectancy is less than two years. The juvenile and adult forms present as limb-girdle muscular dystrophy beginning in the lower limbs. Final outcome depends on respiratory muscle failure. Patients with the adult form can be free of clinical symptoms for most of their life but finally develop a slowly progressive myopathy. Belongs to the glycosyl hydrolase 31 family. Protein type: Carbohydrate Metabolism - galactose; Carbohydrate Metabolism - starch and sucrose; Contractile; EC 3.2.1.20; Hydrolase Chromosomal Location of Human Ortholog: 17q25.3 Cellular Component: lysosomal lumen; lysosomal membrane; lysosome; membrane Molecular Function: alpha-glucosidase activity; oligo-1,6-glucosidase activity Biological Process: cardiac muscle contraction; diaphragm contraction; glucose metabolic process; glycogen catabolic process; lysosome organization and biogenesis; maltose metabolic process; sucrose metabolic process; vacuolar sequestering Disease: Glycogen Storage Disease Ii
Research Articles on GAA	**1.** Compared with controls, GAA gene expression levels in coronary artery disease (CAD) patients were significantly increased, suggesting that GAA may be involved in the CAD development.
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