Galactosidase Alpha (GLa) Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	Galactosidase Alpha (GLa), Polyclonal Antibody
Full Product Name	APC Linked Polyclonal Antibody to Galactosidase Alpha (GLa)
Product Gene Name	anti-GLa antibody
Product Synonym Names	GLa/Galactosidase Alpha; GALA; GLA; AlphaDGalactoside Galactohydrolase; AlphaDGalactosidase A; Melibiase; Agalsidase
Clonality	Polyclonal
Host	Rabbit
Species Reactivity	Human
Form/Format	Liquid; 0.01M PBS, pH7.4, containing 0.05% Proclin300, 50% glycerol.
Purity/Purification	Antigenspecific affinity chromatography followed by Protein A affinity chromatography
Concentration	>=0.1mg/mL (Please refer to the vial label for the specific concentration.) (lot specific)
Specificity	The antibody is a rabbit polyclonal antibody raised against GLa. It has been selected for its ability to recognize GLa in immunohistochemical staining and western blotting.
Preparation and Storage	Store at 2 degree C to 8 degree C for frequent use, 20 degree C for 24 months The thermal stability is described by the loss rate. The loss rate was determined by accelerated thermal degradation test, that is, incubate the protein at 37 degree C for 48h, and no obvious degradation and precipitation were observed. The loss rate is less than 5% within the expiration date under appropriate storage condition.
Matching Pairs	Unconjugated Antibody: GLa (MBS2139460) APC Conjugated Antibody: GLa (MBS2128795)
Molecular Weight	48,767 Da
OMIM	300644
3D Structure	ModBase 3D Structure for P06280
Supply Chain Verification	Manufactured in a lab with traceable raw materials. Bulk orders can typically be prepared to the customer's specifications, please inquire.
Immunogen Type	Prokaryotic Recombinant proteins/RP
Sequence of Immunogen	Leu32~Leu429
Conjugation	APC
Product Categories/Family for anti-GLa antibody	Enzyme & Kinase; Metabolic pathway; Primary, APC labeled
ISO Certification	Manufactured in an ISO 9001:2015 and ISO 13485:2016 Certified Laboratory.
Applications Tested/Suitable for anti-GLa antibody	Western Blot (WB), Immunohistochemistry (IHC), Immunocytochemistry (ICC), Immunoprecipitation (IP)
Application Notes for anti-GLa antibody	WB: 0.52ug/mL IHC: 520ug/mL ICC: 520ug/mL

Related Product Information for anti-GLa antibody	MBS2128795 is an antibody targeting GLa. Also known as APC Linked Polyclonal Antibody to Galactosidase Alpha (GLa). Reactive in Human. Host: Rabbit.
Other Notes	Small volumes of anti-GLa antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.

ADDITIONAL DETAILS

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

NCBI GI #	4504009
NCBI GeneID	2717
NCBI Accession #	NP_000160.1
UniProt Primary Accession #	P06280
UniProt Secondary Accession #	Q6LER7
UniProt Related Accession #	P06280
GenBank Nucleotide #	NM_000169.2
NCBI Official Full Name	alpha-galactosidase A
NCBI Official Synonym Full Names	galactosidase, alpha
NCBI Official Symbol	GLA
NCBI Official Synonym Symbols	GALA
UniProt Protein Name	Alpha-galactosidase A
UniProt Synonym Protein Names	Alpha-D-galactosidase A; Alpha-D-galactoside galactohydrolase; Melibiase
Protein Family	Alpha-galactosidase
UniProt Gene Name	GLA
UniProt Entry Name	AGAL_HUMAN
NCBI Protein Information	alpha-galactosidase A; melibiase; alpha-gal A; agalsidase alfa; alpha-D-galactosidase A; alpha-D-galactoside galactohydrolase 1
NCBI Summary for GLa	This gene encodes a homodimeric glycoprotein that hydrolyses the terminal alpha-galactosyl moieties from glycolipids and glycoproteins. This enzyme predominantly hydrolyzes ceramide trihexoside, and it can catalyze the hydrolysis of melibiose into galactose and glucose. A variety of mutations in this gene affect the synthesis, processing, and stability of this enzyme, which causes Fabry disease, a rare lysosomal storage disorder that results from a failure to catabolize alpha-D-galactosyl glycolipid moieties. [provided by RefSeq, Jul 2008]

UniProt Comments for GLa	GLA: Defects in GLA are the cause of Fabry disease (FD). FD is a rare X-linked sphingolipidosis disease where glycolipid accumulates in many tissues. The disease consists of an inborn error of glycosphingolipid catabolism. FD patients show systemic accumulation of globotriaoslyceramide (Gb3) and related glycosphingolipids in the plasma and cellular lysosomes throughout the body. Clinical recognition in males results from characteristic skin lesions (angiokeratomas) over the lower trunk. Patients may show ocular deposits, febrile episodes, and burning pain in the extremities. Death results from renal failure, cardiac or cerebral complications of hypertension or other vascular disease. Heterozygous females may exhibit the disorder in an attenuated form, they are more likely to show corneal opacities. Belongs to the glycosyl hydrolase 27 family. Protein type: Carbohydrate Metabolism - galactose; Lipid Metabolism - sphingolipid; Glycan Metabolism - glycosphingolipid biosynthesis - globo series; Hydrolase; EC 3.2.1.22; Lipid Metabolism - glycerolipid Chromosomal Location of Human Ortholog: Xq22 Cellular Component: Golgi apparatus; lysosomal lumen; lysosome; cytoplasm; extracellular region Molecular Function: protein binding; protein homodimerization activity; hydrolase activity; alpha-galactosidase activity; galactoside binding; catalytic activity; receptor binding Biological Process: sphingolipid metabolic process; negative regulation of nitric-oxide synthase activity; glycoside catabolic process; negative regulation of nitric oxide biosynthetic process; glycosphingolipid catabolic process; glycosylceramide catabolic process; glycosphingolipid metabolic process; oligosaccharide metabolic process Disease: Fabry Disease
Research Articles on GLa	**1.** Missense mutation p.R112C ablates the activity of the alpha-gal A gene and results in the development of Fabry disease with renal damage in a Chinese family.
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