GLalpha Human Recombinant Protein

PRODUCT OVERVIEW

Product Name	GLalpha, Recombinant Protein
Full Product Name	Recombinant Human GLalpha protein (His tag)
Product Gene Name	GLalpha recombinant protein
Product Synonym Names	Agalsidase; Alpha-D-Galactosidase A; Alpha-D-Galactoside; Alpha-Galactosidase A; GLA; GLAL; Galactohydrolase; Melibiase
Sequence	Leu32-Leu4 29
Host	E coli
Form/Format	Lyophilized from sterile PBS, pH 7.4. Normally 5%-8% trehalose, mannitol and 0.01% Tween 80 are added as protectants before lyophilization. Please refer to the specific buffer information in the printed manual.
Purity/Purification	>95% as determined by reducing SDS-PAGE.
Preparation and Storage	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80 degree C. Reconstituted protein solution can be stored at 4-8 degree C for 2-7 days. Aliquots of reconstituted samples are stable at < -20 degree C for 3 months.
Molecular Weight	48,767 Da
OMIM	300644
3D Structure	ModBase 3D Structure for P06280
Species	Human
Tag	N-His
Endotoxin	<10EU/mg of the protein as determined by the LAL method.
Reconstitution	It is recommended that sterile water be added to the vial to prepare a stock solution of 0.5mg/mL. Concentration is measured by UV-Vis.
Product Categories/Family for GLalpha recombinant protein	Proteins; Recombinant Proteins; Others; Cardiovascular
ISO Certification	Manufactured in an ISO 9001:2015 Certified Laboratory.
Related Product Information for GLalpha recombinant protein	alpha-Galactosidase A is a homodimeric glycoprotein that belongs to the glycosyl hydrolase 27 family. It is a lysosomal enzyme and used as a long-term enzyme replacement therapy in patients with a confirmed identification of Fabry disease. alpha-Galactosidase A can hydrolyze terminal alpha-galactosyl moieties from glycolipids and glycoproteins and catalyze the hydrolysis of melibiose into galactose and glucose. Defects alpha-Galactosidase A are the cause of Fabry disease (FD) which is a rare X-linked sphingolipidosis disease with 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. 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.
Other Notes	Small volumes of GLalpha recombinant protein 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.

Searchable Terms for GLalpha purchase	MBS2580477 is a recombinant protein targeting GLalpha. Also known as Recombinant Human GLalpha protein (His tag). Host: E coli.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for GLalpha. 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
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 GLalpha	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 GLalpha	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 GLalpha	**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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