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alpha-Galactosidase A Recombinant Protein

Cat. No.: 91-323

NO IMAGE AVAILABLE

alpha-Galactosidase A Recombinant Protein

Specifications

SPECIES:	Human
SOURCE SPECIES:	Human Cells
SEQUENCE:	Leu32-Leu429
FUSION TAG:	C-6 His tag
TESTED APPLICATIONS:	
APPLICATIONS:	This recombinant protein can be used for biological assays. For research use only.
PREDICTED MOLECULAR WEIGHT:	46.39 kD

Properties

PURITY:	Greater than 95% as determined by reducing SDS-PAGE. Endotoxin level less than 0.1 ng/ug (1 IEU/ug) as determined by LAL test.
PHYSICAL STATE:	Liquid
BUFFER:	Supplied as a 0.2 um filtered solution of 20mM TrisHCl, 150mM NaCl, pH 8.0. It is not recommended to reconstitute to a concentration less than 100 ug/ml.

STORAGE CONDITIONS:	Store at -20˚C, stable for 6 months after receipt. Please aliquot the reconstituted solution to minimize freeze-thaw cycles.

Additional Info

OFFICIAL SYMBOL:	GLA
ALTERNATE NAMES:	Alpha-Galactosidase A, Alpha-D-Galactosidase A, Alpha-D-Galactoside Galactohydrolase, Melibiase, Agalsidase, GLA
ACCESSION NO.:	P06280
GENE ID:	2717

Background and References

BACKGROUND:	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 diagnosis 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.