

Recombinant Human GLA (C-6His)

Catalog #	EPT291
Expression Host	Human Cells
DESCRIPTION	Recombinant Human Alpha-Galactosidase is
	produced by our Mammalian expression system and
	the target gene encoding Leu32-Leu429 is expressed
	with a 6His tag at the C-terminus.
Accession	P06280
Synonyms	Alpha-Galactosidase A; Alpha-D-Galactosidase A;
	Alpha-D-Galactoside Galactohydrolase; Melibiase;
	Agalsidase; GLA
Mol Mass	46.39 KDa
AP Mol Mass	50-60 KDa, reducing conditions
Purity	Greater than 95% as determined by reducing
	SDS-PAGE.
Endotoxin	Less than 0.1 ng/ μ g (1 EU/ μ g) as determined by LAL
	test.
FORMULATION	Supplied as a 0.2 μ m filtered solution of 20mM
	Tris-HCl, 150mM NaCl, pH 8.0.



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RECONSTITUTION

SHIPPING

The product is shipped on dry ice/polar packs. Upon receipt, store it immediately at the temperature listed below.

STORAGEStore at \leq -70°C, stable for 6 months after receipt.Store at \leq -70°C, stable for 3 months under sterile
conditions after opening.

Please minimize freeze-thaw cycles.

BACKGROUND α -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. α -Galactosidase A can hydrolyze terminal α -galactosyl moieties from glycolipids and glycoproteins and catalyze the hydrolysis of melibiose into galactose and glucose. Defects α -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)



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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.



SDS-PAGE



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