

## GLA/alpha-Galactosidase A Protein, Human, Recombinant (His)

### General Information

Synonyms:	GLA/ $\alpha$ -Galactosidase A Protein;galactosidase, $\alpha$ ;galactosidase, alpha;GALA
Protein Construction:	A DNA sequence encoding the human GLA (NP_000160.1) (Met 1-Leu 429) was fused with a polyhistidine tag at the C-terminus. Predicted N terminal: Leu 32
Species:	Human
Expression Host:	HEK293 Cells
Accession:	P06280
Molecular Weight:	46.8 kDa (predicted)

### QC Testing

Biological Activity:	Activity testing is in progress. It is theoretically active, but we cannot guarantee it. If you require protein activity, we recommend choosing the eukaryotic expression version first.
Purity:	> 97 % as determined by SDS-PAGE
Endotoxin:	< 1.0 EU/ $\mu$ g of the protein as determined by the LAL method.
Formulation:	Lyophilized from a solution filtered through a 0.22 $\mu$ m filter, containing 50 mM Tris, 150 mM NaCl, pH 7.5. Typically, a mixture containing 5% to 8% trehalose, mannitol, and 0.01% Tween 80 is incorporated as a protective agent before lyophilization.

### Preparation and Storage

Reconstitution:	Reconstituted with sterile deionized water to 0.25 mg/mL. Reconstitution conditions may vary depending on the lot.
Stability & Storage:	It is recommended to store recombinant proteins at -20°C to -80°C for future use. Lyophilized powders can be stably stored for over 12 months, while liquid products can be stored for 6-12 months at -80°C. For reconstituted protein solutions, the solution can be stored at -20°C to -80°C for at least 3 months. Please avoid multiple freeze-thaw cycles and store products in aliquots. <small>Actual storage temperature shall be subject to the COA.</small>
Shipping:	In general, lyophilized powders are shipped with blue ice, while solutions are shipped with dry ice.

### Protein Background

Alpha-galactosidase A, also known as Alpha-D-galactoside galactohydrolase, Alpha-D-galactosidase A, Melibiase and GLA, is a member of the glycosyl hydrolase 27 family. GLA is used as a long-term enzyme replacement therapy in patients with a confirmed diagnosis of Fabry disease. Defects in GLA are the cause of Fabry disease (FD) which 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. Deficiency of GLA leads to the accumulation of glycosphingolipids in the vasculature leading to multiorgan pathology. In addition to well-described microvascular disease, deficiency of GLA is also characterized by premature macrovascular events such as stroke and possibly myocardial infarction.

### Reference

- Koide T. et al., 1990, FEBS Lett. 259:353-356.  
Yang C.-C. et al., 2003, Clin. Genet. 63:205-209.  
Verovnik F. et al., 2004, Eur. J. Hum. Genet. 12:678-681.  
Nance C.S. et al., 2006, Arch. Neurol. 63:453-457.

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