

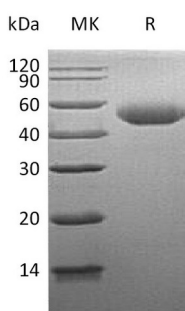
**Product Name: Recombinant Human GLA (C-6His)**  
**Catalog #: PHH0050**



## Summary

<b>Name</b>	alpha-Galactosidase A/GLA
<b>Purity</b>	Greater than 95% as determined by reducing SDS-PAGE
<b>Endotoxin level</b>	<1 EU/μg as determined by LAL test.
<b>Construction</b>	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.
<b>Accession #</b>	P06280
<b>Host</b>	Human Cells
<b>Species</b>	Human
<b>Predicted Molecular Mass</b>	46.39 KDa
<b>Formulation</b>	Supplied as a 0.2 μm filtered solution of 20mM Tris-HCl, 150mM NaCl, pH 8.0.
<b>Shipping</b>	The product is shipped on dry ice/polar packs. Upon receipt, store it immediately at the temperature listed below.
<b>Stability&amp;Storage</b>	Store at ≤-70°C, stable for 6 months after receipt. Store at ≤-70°C, stable for 3 months under sterile conditions after opening. Please minimize freeze-thaw cycles.
<b>Reconstitution</b>	

## SDS-PAGE image



## Background

<b>Alternative Names</b>	Alpha-Galactosidase A; Alpha-D-Galactosidase A; Alpha-D-Galactoside Galactohydrolase; Melibiase; Agalsidase; GLA
<b>Background</b>	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

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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.

### **Note**

For Research Use Only , Not for Diagnostic Use.