

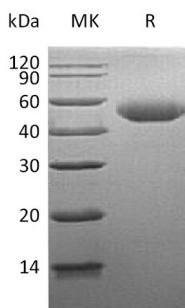
Product Name: Recombinant Human GLA (C-6His)
Catalog #: PHH0050



概述 (Summary)

英文全称	alpha-Galactosidase A/GLA
纯度 (Purity)	Greater than 95% as determined by reducing SDS-PAGE
内毒素 (Endotoxin level)	<1 EU/μg as determined by LAL test.
蛋白构建 (Construction)	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
蛋白标签 (Tag)	
表达宿主 (Host)	Human Cells
种属 (Species)	Human
预测分子量 (Predicted MW)	46.39 KDa
蛋白形态 (Form)	Supplied as a 0.2 μm filtered solution of 20mM Tris-HCl, 150mM NaCl, pH 8.0.
储存缓冲液 (Buffer)	
运输方式 (Shipping)	The product is shipped on dry ice/polar packs. Upon receipt, store it immediately at the temperature listed below.
稳定性&储存 (Stability &Storage)	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.
复溶 (Reconstitution)	

电泳图 (SDS-PAGE image)



背景 (Background)

分子别名 (Alternative Names) Alpha-Galactosidase A; Alpha-D-Galactosidase A; Alpha-D-Galactoside

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背景介绍 (References)

Galactohydrolase; Melibiase; Agalsidase; GLA

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 globotriaosylceramide (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.

注意事项 (Note)

For Research Use Only , Not for Diagnostic Use.