Recombinant Human Alpha-Galactosidase A/GLA Protein (His Tag)

Catalog Number: PKSH033249

Note: *Centrifuge before opening to ensure complete recovery of vial contents.*

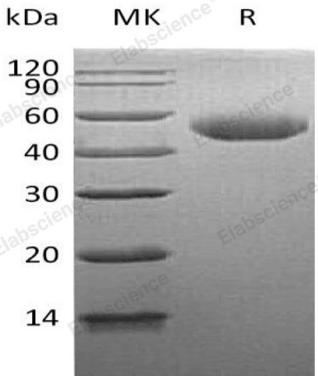
Description

Species	Human
Source	HEK293 Cells-derived Human Alpha-Galactosidase A/GLA protein Leu32-Leu429, with an C-terminal His
Calculated MW	46.4 kDa
Observed MW	50-60 kDa
Accession	P06280
Bio-activity	Not validated for activity

Properties

Purity	> 95 % as determined by reducing SDS-PAGE.
Concentration	Subject to label value.
Endotoxin	< 1.0 EU per μg of the protein as determined by the LAL method.
Storage	Store at < -20°C, stable for 6 months. Please minimize freeze-thaw cycles.
Shipping	This product is provided as liquid. It is shipped at frozen temperature with blue ice/gel packs. Upon receipt, store it immediately at < - 20°C.
Formulation	Supplied as a 0.2 μm filtered solution of 20mM Tris-HCl, 150mM NaCl, pH 8.0.

Data



> 95 % as determined by reducing SDS-PAGE.

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 (Gb 3) 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.

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