Recombinant Human GLα Protein (His Tag)

Catalog Number: PDEH100833

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

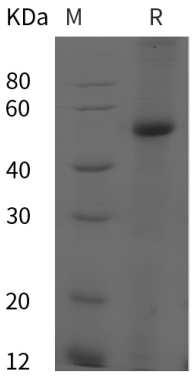
Description

Species	Human
Source	E.coli-derived Human GLα protein Leu32-Leu429, with an N-terminal His
Calculated MW	43.7 kDa
Observed MW	48 kDa
Accession	P06280
Bio-activity	Not validated for activity

Properties

Purity	> 95% as determined by reducing SDS-PAGE.
Endotoxin	< 10 EU/mg of the protein as determined by the LAL method
Storage	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80°C. Reconstituted protein solution can be stored at 4-8°C for 2-7 days. Aliquots of reconstituted samples are stable at < -20°C for 3 months.
Shipping	This product is provided as lyophilized powder which is shipped with ice packs.
Formulation	Lyophilized from a 0.2 μm filtered solution in PBS with 5% Trehalose and 5% Mannitol.
Reconstitution	It is recommended that sterile water be added to the vial to prepare a stock solution of 0.5 mg/mL. Concentration is measured by UV-Vis.

Data



SDS-PAGE analysis of Human GLα proteins, 2 μg/lane of Recombinant Human GLα proteins was resolved with SDS-PAGE under reducing conditions, showing bands at 48 kDa.

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) 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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