

Recombinant Human GLA Protein(His Tag)

Catalog Number: PDMH100451



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

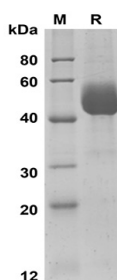
Description

| | |
|---------------------|---|
| Species | Human |
| Source | Mammalian-derived Human GLA protein Met1-Leu429, with an C-terminal His |
| Mol_Mass | 47.2 kDa |
| Accession | P06280 |
| Bio-activity | Not validated for activity |

Properties

| | |
|-----------------------|--|
| Purity | > 95% as determined by reducing SDS-PAGE. |
| Endotoxin | < 1.0 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 GLA proteins, 2µg/lane of Recombinant Human GLA proteins was resolved with SDS-PAGE under reducing conditions, showing bands at 42-60

kDa

Background

For Research Use Only

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Rev. V1.0

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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 globotriaosylceramide (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.

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