

Recombinant Human GL₃ protein (His tag)

Catalog Number:PDEH100177



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

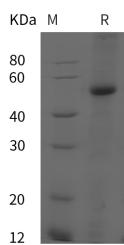
Description

| | |
|------------------------------------|--|
| Synonyms | Alpha-Galactosidase A;Alpha-D-Galactosidase A;Alpha-D-Galactoside;Galactohydrolase;Melibiase;Agalsidase;GLA;GLAL |
| Species | Human |
| Expression Host | E.coli |
| Sequence | Leu 32-Leu 429 |
| Accession | P06280 |
| Calculated Molecular Weight | 43.7 kDa |
| Observed molecular weight | 48 kDa |
| Tag | N-His |

Properties

| | |
|-----------------------|---|
| Purity | > 95 % as determined by reducing SDS-PAGE. |
| Endotoxin | Please contact us for more information. |
| 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 sterile PBS, pH 7.4. Normally 5 % - 8 % trehalose, mannitol and 0.01% Tween80 are added as protectants before lyophilization. Please refer to the specific buffer information in the printed manual. |
| Reconstitution | Please refer to the printed manual for detailed information. |

Data



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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 globotriaosylceramide (Gb3) and related glycosphingolipids in the plasma and cellular lysosomes throughout the body. Patients may show ocular deposits,

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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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