

Mouse alpha-Galactosidase A / GLA Protein (His Tag)

Catalog Number: 50964-M08H



Sino Biological Inc.

Biological Solution Specialist

General Information

Gene Name Synonym:

Gla

Protein Construction:

A DNA sequence encoding the mouse Gla (Q8BGZ6) (Met1-Arg421) was expressed with a C-terminal polyhistidine tag.

Source: Mouse

Expression Host: Human Cells

QC Testing

Purity: > 95 % as determined by SDS-PAGE

Bio-Activity

Measured by its ability to hydrolyze 4-methylumbelliferyl- α -D-galactopyranoside.

The specific activity is > 400 pmoles/min/ μ g.

Endotoxin:

< 1.0 EU per μ g of the protein as determined by the LAL method

Stability:

Samples are stable for up to twelve months from date of receipt at -70 °C

Predicted N terminal: Leu 34

Molecular Mass:

The recombinant mouse Gla comprises 399 amino acids and has a predicted molecular mass of 45.6 kDa. The apparent molecular mass of the protein is approximately 46-52 kDa in SDS-PAGE under reducing conditions due to glycosylation.

Formulation:

Lyophilized from sterile PBS, pH 7.4

Normally 5 % - 8 % trehalose and mannitol are added as protectants before lyophilization. Specific concentrations are included in the hardcopy of COA. Please contact us for any concerns or special requirements.

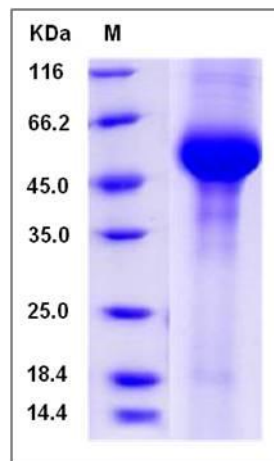
Usage Guide

Storage:

Store it under sterile conditions at -20°C to -80°C upon receiving. Recommend to aliquot the protein into smaller quantities for optimal storage.

Avoid repeated freeze-thaw cycles.

SDS-PAGE:



Reconstitution:

Detailed reconstitution instructions are sent along with the products.

Protein Description

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.

References

1. Koide T. et al., 1990, FEBS Lett. 259: 353-6.
2. Yang C-C. et al., 2003, Clin. Genet. 63: 205-9.
3. Verovnik F. et al., 2004, Eur J Hum Genet. 12: 678-81.
4. Nance CS. et al., 2006, Arch Neurol. 63: 453-7.

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