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Recombinant Mouse alpha-Galactosidase A / GLA Protein (His tag)

产品货号: TP08532

第三版

别名:Ags

描述: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.

配方:PBS

分子量:45.6 kDa

序列:Met1-Arg421

纯度:> 95% by HPLC

浓度:

内毒素:<1.0 EU per 1 ug of protein (determined by LAL method)

存储: +4 ° C 保存 (1-2 周). 长期保存在-20 ° C或者-70 ° C. 避免反复冻融.