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

产品货号: TP07972

第三版

别名:GALA

**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:**46.8 kDa

**序列:**Met 1-Leu 429

**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.