Recombinant Human GLA (C-6His)

货号: RB91EPT291

gui_ge

1mg

biao_da_su_zhu

Human Cells

biao_qian

C-6His

fen_zi_liang

46.39 KDa

biao_guan_fen_zi_liang

50-60 KDa, reducing conditions

xing_tai

Supplied as a 0.2 μm filtered solution of 20mM Tris-HCl, 150mM NaCl, pH 8.0.

chun_du

Greater than 95% as determined by reducing SDS-PAGE.

nei_du_su_shui_ping

Less than 0.1 ng/μg (1 EU/μg) as determined by LAL test.

huo_xing

Measured by its ability to hydrolyze 4-Nitrophenyl -alpha -D-galactopyranoside. The specific activity is 2835 pmol/min/μg.

chu_cun_fang_fa

Store at ≤-70° C, stable for 6 months after receipt.
Store at ≤-70° C, stable for 3 months under sterile conditions after opening.
Please minimize freeze-thaw cycles.

yun_shu_tiao_jian

The product is shipped on dry ice/polar packs.
Upon receipt, store it immediately at the temperature listed below.

bei_jing_shuo_ming

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 globotriaoslyceramide (Gb3) and related glycosphingolipids in the plasma and cellular lysosomes throughout the body. 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.

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