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Product Datasheet

Recombinant Human GLA (C-6His) EBT-EPT291

Artikelname	Recombinant Human GLA (C-6His)
Artikelnummer	EBT-EPT291
Hersteller Artikelnummer	EPT291
Alternativnummer	EBT-EPT291-1
Hersteller	ELK Biotechnology
Kategorie	Proteine/Peptide
Produktbeschreibung	Recombinant Human Alpha-Galactosidase is produced by our Mammalian expression system and the target gene encoding Leu32-Leu429 is expressed with a 6His tag at the C-terminus....
Molekulargewicht	Molecular weight: 46.39 KDa. Apparent molecular weight: 50-60 KDa, reducing conditions
UniProt	P06280
Reinheit	Greater than 95% as determined by reducing SDS-PAGE.

Anwendungsbeschreibung	Endotoxin: Less than 0.1 ng/μg (1 EU/μg) as determined by LAL test. Biological activity: Measured by its ability to hydrolyze 4-Nitrophenyl - alpha -D-galactopyranoside. The specific activity is 2835 pmol/min/μg. Background: alpha-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. alpha-Galactosidase A can hydrolyze terminal alpha-galactosyl moieties from glycolipids and glycoproteins and catalyze the hydrolysis of melibiose into galactose and glucose. Defects alpha-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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