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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                  | <a href="#">P06280</a>  |
| 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