BIOZOL
Diagnostica Vertrieb GmbH, Oehleckerring 11-13
22419 Hamburg, Germany
Telephone: +49 (0)89 3799666-6 | **Fax:** +49 (0)89 3799666-99
E-Mail: info@biozol.de

Product Datasheet

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

Article Name	Recombinant Human GLA (C-6His)
Biozol Catalog Number	EBT-EPT291
Supplier Catalog Number	EPT291
Alternative Catalog Number	EBT-EPT291-1
Manufacturer	ELK Biotechnology
Category	Proteine/Peptide
Product Description	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....
Molecular Weight	Molecular weight: 46.39 KDa. Apparent molecular weight: 50-60 KDa, reducing conditions
UniProt	P06280
Purity	Greater than 95% as determined by reducing SDS-PAGE.

Application Notes	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