

## Recombinant Human GLB1 (C-6His)

Catalog No: C473

Description Recombinant Human beta-Galactosidase is produced by our Mammalian expression system and the

target gene encoding Leu24-Val677 is expressed with a 6His tag at the C-terminus.

Source Human Cells

Alternative name Beta-Galactosidase; Acid Beta-Galactosidase; Lactase; Elastin Receptor 1; GLB1; ELNR1

Accession No. P16278

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

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

Endotoxin Less than 0.1 ng/µg (1 EU/µg)

**Shipping** The product is shipped on dry ice/polar packs.

Upon receipt, store it immediately at the temperature listed below.

Storage Store at < -20°C, stable for 6 months after receipt.

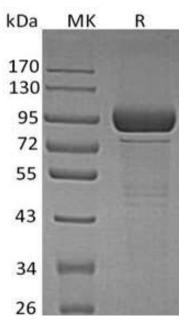
Please minimize freeze-thaw cycles.

Background  $\beta$  Galactosidase is a lysosomal  $\beta$  Galactosidase that hydrolyzes the terminal  $\beta$  Galactose from

Ganglioside and Keratan sulfate. In lysosome, the mature  $\beta$  Galactosidase protein associates with Cathepsin A and Neuraminidase 1 to form the lysosomal multienzyme complex. An alternative splicing at the RNA level of  $\beta$  Galactosidase results a catalytically inactive  $\beta$  Galactosidase that plays an important role in vascular development. Defects of  $\beta$ -galactosidase (GLB1) are the cause of diseases like GM1-gangliosidosis which is a lysosomal storage disease and Morquio Syndrome B that cause patients to have abnormal elastic fibers. More than 100 mutations have been identified for  $\beta$  Galactosidase, which result in different residual activities of the mutant enzymes and a spectrum of

symptoms in the two related diseases.

**SDS-PAGE** 



MK: Marker

R: Sample under reducing conditions

