

Active Recombinant Mouse Glb1 protein, His-tagged

Cat. No. Glb1-1523R **Lot. No.** (See product label)

SPECIFICATION

Product Overview	Active Recombinant Mouse Glb1 protein(P23780)(25~647 aa), fused with N-terminal His tag, was expressed in HEK293.
Species	Mouse
Source	HEK293
ProteinLength	25-647 aa
Form	Lyophilized from sterile PBS, pH7.4, containing 0.01% SKL, 1mM DTT, 5% Trehalose and Proclin300.
Bio-activity	GLB1 is a lysosomal beta -galactosidase that hydrolyzes the terminal beta -galactose from ganglioside and keratan sulfate. Defects in this gene are the causes of lysosomal storage diseases for GM1-gangliosidosis and Morquio B syndrome (also known as mucopolysaccharidosis IVB). In GM1 gangliosidosis, GM1 ganglioside accumulates in the neurons of the central nervous system, because of the deficiency of lysosomal beta -galactosidase activity. GM1 gangliosidosis demonstrates varying degrees of clinical severity but is invariably fatal, and children with the most common and severe form of GM1 gangliosidosis usually die within 3 years of birth. Morquio B syndrome patients are neurologically normal, but display severe skeletal dysostosis multiplex because of an accumulation of keratan sulfate. The activity assay of GLB1 was measured by its ability to cleave a peptide substrate, 4-Methylumbelliferyl-beta -D-galactopyranoside. The reaction was performed in 50 mM Sodium Citrate, pH 3.5 (

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Assay Buffer), initiated by addition 50 μ L of 1.5 ug/ml uPA (diluted by Assay Buffer) to 50 μ of 1.2 mM Substrate. Read at excitation and emission wavelengths of 365 nm and 445 nm (top read), respectively, in kinetic mode for 5 minutes. The specific activity of recombinant mouse GLB1 is >17000 pmol/min/g.

Molecular Mass 72.2kDa

Purity > 90% as determined by SDS-PAGE.

Storage Avoid repeated freeze/thaw cycles. Store at -20°C for 12 months. Aliquot and store at -80°C for 12 months.

Reconstitution Reconstitute in 10mM PBS (pH7.4) to a concentration of 0.1-1.0 mg/mL. Do not vortex.

GENE INFORMATION

Gene Name Glb1 galactosidase, beta 1 [*Mus musculus*]

Official Symbol Glb1

Synonyms GLB1; galactosidase, beta 1; beta-galactosidase; lactase; acid beta-galactosidase; beta-galactosidase complex; beta-galactosidase temporal; beta-galactosidase electrophoretic; beta-galactosidase systemic regulator; Bge; Bgl; Bgs; Bgt; Bgl-e; Bgl-s; Bgl-t; AW125515; C130097A14Rik;

Gene ID 12091

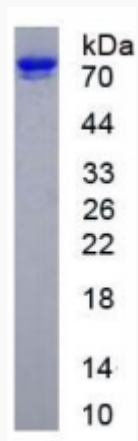
mRNA Refseq NM_009752

Protein Refseq NP_033882

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SDS-PAGE

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