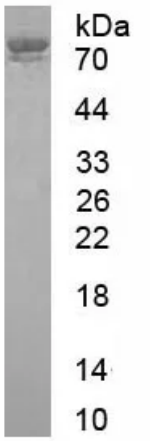
Active Galactosidase Beta (GLb)

Catalog#:PHM12272

概述（Summary）

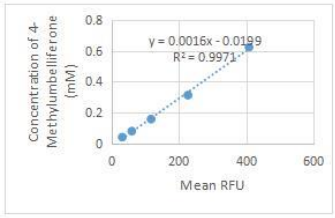
别称（Alternative Names）	GL-B; b-GAL; bGAL; ELNR1; EBP; GLB1; Lactase; Elastin Receptor 1,67kDa; Elastin Receptor 1(67kD); Acid Beta-Galactosidase
纯度（Purity）	Greater than 90% as determined by reducing SDS-PAGE.
内毒素（Endotoxin level）	Please contact with the lab for this information
蛋白构建（Construction）	【Ile25-Ser647】
Accession #	P23780
表达宿主（Host）	293F
种属（Species）	Mus musculus (Mouse)
预测分子量（Predicted MW）	72.2 kDa
制剂（Form）	Lyophilized from a 0.2 μm filtered solution of PBS, pH7.4, containing 0.01% SKL, 1mM DTT, 5% Trehalose and Proclin300.
运输方式（Shipping）	The product is shipped with ice bag. Upon receipt, store it immediately at the temperature listed below.
稳定性&储存（Stability &Storage）	12 months at -20°C to -80°C in lyophilized state.6 months at -20°C to -80°C under sterile conditions after reconstitution.7-10 days at 2°C to 8°C under sterile conditions after reconstitution.Please minimize freeze-thaw cycles.
复溶（Reconstitution）	Reconstitute in 10mM PBS (pH7.4) to a concentration of 0.1-1.0 mg/mL. Do not vortex.
背景介绍（References）	Cleaves beta-linked terminal galactosyl residues from gangliosides, glycoproteins, and glycosaminoglycans.

电泳图（SDS-PAGE image）



Greater than 90% as determined by reducing SDS-PAGE.

活性图（Bioactivity image）



RFU	4-Methylumbelliferone (mM)
407.3912	0.625
226.2912	0.3125
117.1912	0.15625
59.9312	0.078125
31.6012	0.0390625

Figure 1. The standard curve of 4-Methylumbelliferone

One unit of enzyme activity is defined as the 1 µg of enzyme required to convert 1 pmol of 4-Methylumbelliferyl-beta -D-galactopyranoside to 4-Methylumbelliferone in 1 min.

Specific Activity (pmol/min/µg)= $\frac{\Delta RFU * F}{T * N}$

△RFU=Adjusted for Substrate Blank

F=Conversion Factor (convert from standard curve of 4-Methylumbelliferone)

T= Time

N=Amount of enzyme

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 (Assay Buffer), ainitiated by addition 50 µL of 1.5 ug/ml uPA (diluted by Assay Buffer) to 50 µL 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/µg.

注意事项（Note）

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