Human β-Galactosidase-1/GLB1 Antibody

Monoclonal Mouse IgG_{2A} Clone # 810226

Catalog Number: MAB6464

DESCRIPTION

Species Reactivity	Human
Specificity	Detects human β-Galactosidase-1/GLB1 in direct ELISAs.
Source	Monoclonal Mouse IgG_{2A} Clone # 810226
Purification	Protein A or G purified from hybridoma culture supernatant
Immunogen	Chinese hamster ovary cell line CHO-derived recombinant human β-Galactosidase-1/GLB1 Met1-Val677 Accession # P16278
Formulation	Lyophilized from a 0.2 μm filtered solution in PBS with Trehalose. See Certificate of Analysis for details. *Small pack size (-SP) is supplied either lyophilized or as a 0.2 μm filtered solution in PBS.

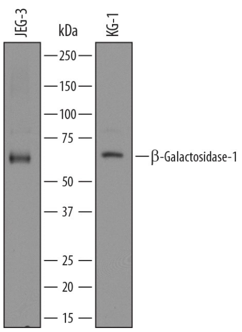
APPLICATIONS

Please Note: *Optimal dilutions should be determined by each laboratory for each application. General Protocols are available in the Technical Information section on our website.*

	Recommended Concentration	Sample
Western Blot	1 µg/mL	See Below

DATA

Western Blot



Detection of Human β-Galactosidase-1/GLB1 by Western Blot. Western blot shows lysates of JEG-3 human epithelial choriocarcinoma cell line and KG-1 human acute myelogenous leukemia cell line. PVDF membrane was probed with 1 µg/mL of Mouse Anti-Human β-Galactosidase-1/GLB1 Monoclonal Antibody (Catalog # MAB6464) followed by HRP-conjugated Anti-Mouse IgG Secondary Antibody (Catalog # HAF007). A specific band was detected for β-Galactosidase-1/GLB1 at approximately 64 kDa (as indicated). This experiment was conducted under reducing conditions and using Immunoblot Buffer Group 1.

PREPARATION AND STORAGE

Reconstitution	Sterile PBS to a final concentration of 0.5 mg/mL.
Shipping	The product is shipped at ambient temperature. Upon receipt, store it immediately at the temperature recommended below. *Small pack size (-SP) is shipped with polar packs. Upon receipt, store it immediately at -20 to -70 °C
Stability & Storage	**Use a manual defrost freezer and avoid repeated freeze-thaw cycles.** ● 12 months from date of receipt, -20 to -70 °C as supplied. ● 1 month, 2 to 8 °C under sterile conditions after reconstitution. ● 6 months, -20 to -70 °C under sterile conditions after reconstitution.

BACKGROUND

GLB1, a 60-76 kDa (predicted) glycoprotein, is a lysosomal β-galactosidase that hydrolyzes the terminal β-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) (1, 2, 3). In GM1 gangliosidosis, GM1 ganglioside accumulates in the neurons of the central nervous system, because of the deficiency (0±3% of normal) of lysosomal β-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 (4). More than 100 mutations have been identified for GLB1, which result in different residual activities of the mutant enzymes and a spectrum of symptoms in the two related diseases (5). In lysosome, the mature β-galactosidase protein associates with cathepsin A and neuraminidase 1 to form the lysosomal multienzyme complex (6). An alternative splicing at the RNA level of GLB1 results a catalytically inactive β-galactosidase (also called elastin-binding protein) that plays an important role in vascular development (7).

References:

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2. Brunetti-Pierri N, and Scaglia F. (2008) Mol. Genet. Metab. **94**:391.
3. Santamaria, R. *et al*. (2007) J. Lipid Res. **48**:2275.
4. Prat, C. (2008) Joint Bone Spine, **75**:495.
5. Zhang, S. *et al*. (2000) Biochem. J. **348**:621.
6. Pshezhetsky, A.V. and Ashmarina, M. (2001) Prog. Nucleic Acid Res. Mol. Biol. **69**:81.
7. Salvatore P, *et al*. (1998) J. Biol. Chem. **273**:6319.