



Galactosidase β Monoclonal Antibody

Catalog No	BYmab-04347
Isotype	IgG
Reactivity	Human;Rat;Mouse;
Applications	WB
Gene Name	GLB1
Protein Name	Beta-galactosidase
Immunogen	Synthesized peptide derived from the Internal region of human Galactosidase β .
Specificity	Galactosidase β Monoclonal Antibody detects endogenous levels of Galactosidase β protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	$\geq 90\%$
Storage Stability	-20°C/1 year
Synonyms	GLB1; ELNR1; Beta-galactosidase; Acid beta-galactosidase; Lactase; Elastin receptor 1
Observed Band	76kD
Cell Pathway	[Isoform 1]: Lysosome .; [Isoform 2]: Cytoplasm, perinuclear region . Localized to the perinuclear area of the cytoplasm but not to lysosomes. .
Tissue Specificity	Detected in placenta (at protein level) (PubMed:8383699). Detected in fibroblasts and testis (PubMed:2511208).
Function	catalytic activity:Hydrolysis of terminal non-reducing beta-D-galactose residues in beta-D-galactosides.,disease:Defects in GLB1 are the cause of GM1-gangliosidosis type 1 (GM1G1) [MIM:230500]; also known as infantile GM1-gangliosidosis. GM1-gangliosidosis is an autosomal recessive lysosomal storage disease marked by the accumulation of GM1 gangliosides, glycoproteins and keratan sulfate primarily in neurons of the central nervous system. GM1G1 is characterized by onset within the first three months of life, central nervous system degeneration, coarse facial features, hepatosplenomegaly, skeletal dysmorphism reminiscent of Hurler syndrome, and rapidly progressive psychomotor deterioration. Urinary oligosaccharide levels are high. It leads to

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death usually between the first and second year of life.,disease:Defects in GLB1 are the cause of GM1-gangliosidosis type 2 (GM1G2) [MIM:230600];

Background

This gene encodes a member of the glycosyl hydrolase 35 family of proteins. Alternative splicing results in multiple transcript variants, at least one of which encodes a preproprotein that is proteolytically processed to generate the mature lysosomal enzyme. This enzyme catalyzes the hydrolysis of a terminal beta-linked galactose residue from ganglioside substrates and other glycoconjugates. Mutations in this gene may result in GM1-gangliosidosis and Morquio B syndrome. [provided by RefSeq, Nov 2015],

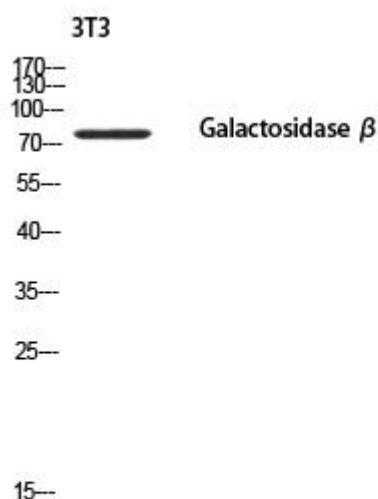
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using Galactosidase β Monoclonal Antibody