β-Galactosidase Rabbit mAb

Catalog No	YP-rAb-17877
Isotype	IgG
Reactivity	Human,Mouse,Rat
Applications	WB,IHC,IF,ELISA
Gene Name	GLB1
Protein Name	Beta-galactosidase
Purification Process	Protein A
Specificity	Endogenous
Formulation	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA
Source	Monoclonal, Rabbit,IgG
Dilution	IHC 1:2000-1:10000; WB 1:1000-1:5000; IF 1:200-1:1000; ELISA 1:5000-1:20000; Note: For IHC, we suggest antigen retrieval with TE buffer pH 9.0
Concentration	0.5 mg/ml
Purity	≥90%
Storage Stability	-15° C to -25° C/1 year(Do not lower than -25° C)
Synonyms	GLB1 ; ELNR1 ; Beta-galactosidase ; Acid beta-galactosidase ; Lactase ; Elastin receptor 1
Observed Band	100kD
Calculated Molecular Weight	76kD
Cell Pathway	Lysosome
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 dysmorphology reminiscent of Hurler syndrome, and rapidly progressive psychomotor deterioration. Urinary oligosaccharide levels are high. It leads to 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]; also known as late infantile/juvenile GM1-gangliosidosis. GM1G2 is characterized by onset between ages 1 and 5. The main symptom is locomotor ataxia, ultimately leading to a state of decerebration with epileptic seizures. Patients do not display the skeletal changes associated with the infantile form, but they nonetheless excrete elevated amounts of beta-linked galactose-terminal oligosaccharides. Inheritance is autosomal recessive.,Disease:Defects in GLB1 are the cause of GM1-gangliosidosis type 3 (GM1G3) [MIM:230650]; also known as adult or chronic GM1-gangliosidosis. GM1G3 is characterized by a variable phenotype. Patients show mild skeletal abnormalities, dysarthria, gait disturbance, dystonia and visual impairment. Visceromegaly is absent. Intellectual deficit can initially be mild or absent but progresses over time. Inheritance is autosomal recessive.,Disease:Defects in GLB1 are the cause of mucopolysaccharidosis type 4B (MPS4B) [MIM:253010]; also known as Morquio syndrome B. MPS4B is a form of mucopolysaccharidosis type 4, an autosomal recessive lysosomal storage disease characterized by intracellular accumulation of keratan sulfate and chondroitin-6-sulfate. Key clinical features include short stature, skeletal dysplasia, dental anomalies, and corneal clouding. Intelligence is normal and there is no direct central nervous system involvement, although the skeletal changes may result in neurologic complications. There is variable severity, but patients with the severe phenotype usually do not survive past the second or third decade of life.,Function:Cleaves beta-linked terminal galactosyl residues from gangliosides, glycoproteins, and glycosaminoglycans.,Function:Isoform 2 has no beta-galactosidase catalytic activity, but plays functional roles in the formation of extracellular elastic fibers (elastogenesis) and in the development of connective tissue. Seems to be identical to the elastin-binding protein (EBP), a major component of the non-integrin cell surface receptor expressed on fibroblasts, smooth muscle cells, chondroblasts, leukocytes, and certain cancer cell types. In elastin producing cells, associates with tropoelastin intracellularly and functions as a recycling molecular chaperone which facilitates the secretions of tropoelastin and its assembly into elastic fibers.,online information:Beta-galactosidase entry,similarity:Belongs to the glycosyl hydrolase 35 family.,subcellular location:Localized to the perinuclear area of the cytoplasm but not to lysosomes.,

Background

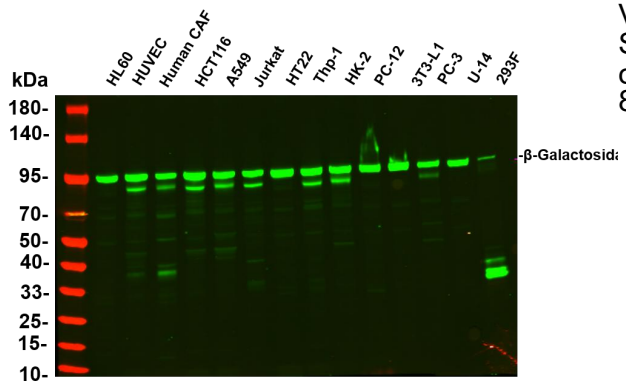
This gene encodes a member of the glycosyl hydrolase 35 family of proteins. Alternative splicing results in multiple transcript variants, at least one of which encodes a preproprotein that is proteolytically processed to generate the mature lysosomal enzyme. This enzyme catalyzes the hydrolysis of a 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

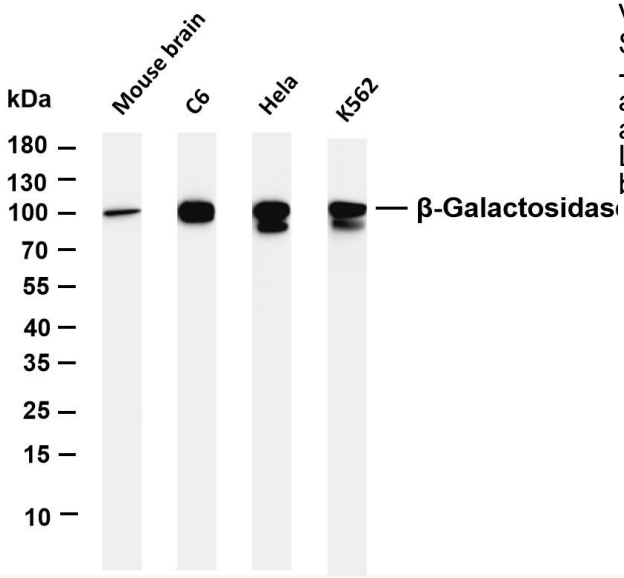
Avoid repeated freezing and thawing!

Usage suggestions

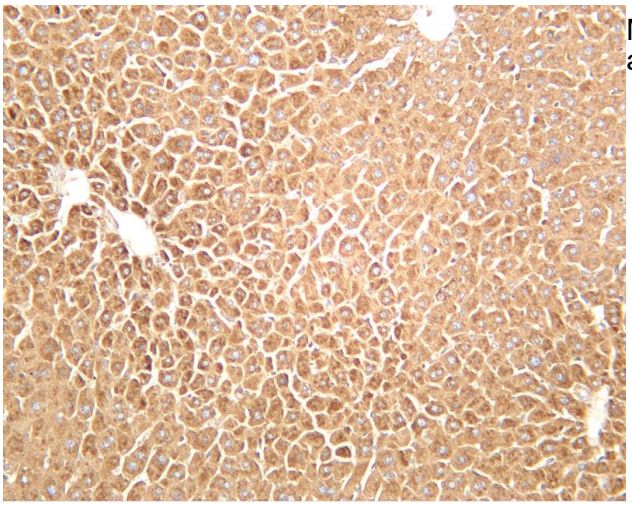
This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.



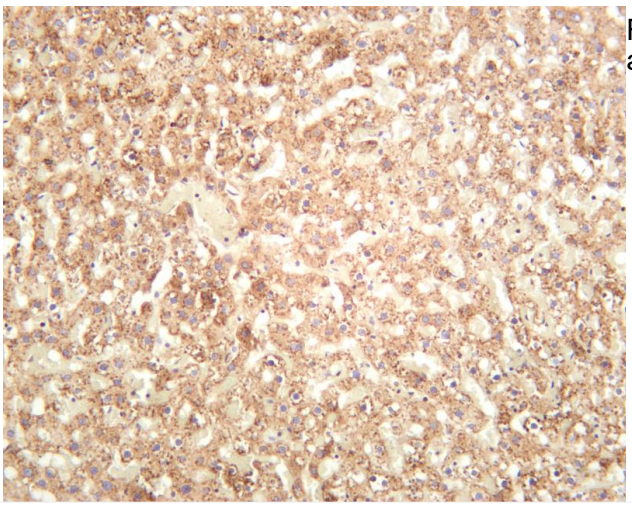
Various whole cell lysates were separated by 4-20% SDS-PAGE, and the primary antibody was used at 4℃, over night with a 1:2500 dilution . The Dylight 800-conjugated Goat anti-Rabbit antibody



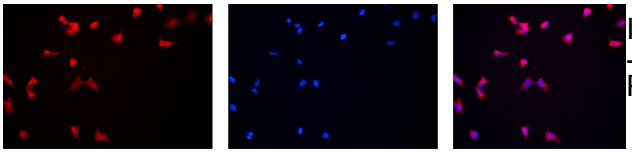
Various whole cell lysates were separated by 4-20% SDS-PAGE, and the membrane was blotted with anti- β -Galactosidase antibody. The HRP-conjugated Goat anti-Rabbit IgG(H + L) antibody was used to detect the antibody. Lane 1: Mouse brain Lane 2: C6 Lane 3: Hela Lane 4: K562 Predicted band size: 76kDa Observed band size: 100kDa



Mouse liver was stained with anti- β -Galactosidase rabbit antibody



Rat liver was stained with anti- β -Galactosidase rabbit antibody



Immunofluorescence analysis of HEK293. Picture A: β -Galactosidase antibody (red). Picture B: DAPI (blue). Picture C: Merge of A+B