Anti-GLB1 / Beta-galactosidase Antibody [JE40-77]

HA721562

Product Type:	Recombinant Rabbit monoclonal IgG, primary antibodies
Species reactivity:	Human, Mouse, Rat
Applications:	WB, FC, IHC-P
Molecular Wt:	Predicted band size: 76 kDa
Clone number:	JE40-77

Description: Cleaves beta-linked terminal galactosyl residues from gangliosides, glycoproteins, and glycosaminoglycans. 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. 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.

Immunogen: Synthetic peptide within Human GLB1 aa 628-677 / 677.

Positive control: SK-Br-3 cell lysate, Saos-2 cell lysate, U-87 MG cell lysate, MCF7 cell lysate, HepG2 cell lysate, K-562 cell lysate, Daudi cell lysate, NIH/3T3 cell lysate, Rat brain tissue lysate, mouse placenta tissue, SK-Br-3.

Subcellular location: Lysosome and Cytoplasm.

Database links: SwissProt: P16278 Human | P23780 Mouse
Entrez Gene: 316033 Rat

Recommended Dilutions:

WB	1:1,000
FC	1:500-1:1,000
IHC-P	1:1,000

Storage Buffer: 1*TBS (pH7.4), 0.05% BSA, 40% Glycerol. Preservative: 0.05% Sodium Azide.

Storage Instruction: Store at +4℃ after thawing. Aliquot store at -20℃. Avoid repeated freeze / thaw cycles.

Purity: Protein A affinity purified.

Images

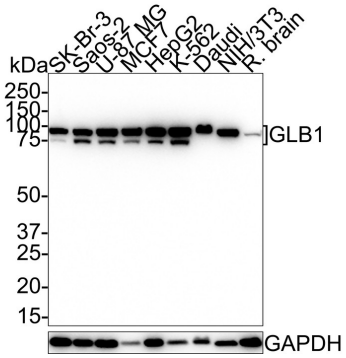

Fig1: Western blot analysis of GLB1 / Beta-galactosidase on different lysates with Rabbit anti-GLB1 / Beta-galactosidase antibody (HA721562) at 1/1,000 dilution.

Lane 1: SK-Br-3 cell lysate (20 µg/Lane)
Lane 2: Saos-2 cell lysate (20 µg/Lane)
Lane 3: U-87 MG cell lysate (20 µg/Lane)
Lane 4: MCF7 cell lysate (20 µg/Lane)
Lane 5: HepG2 cell lysate (20 µg/Lane)
Lane 6: K-562 cell lysate (20 µg/Lane)
Lane 7: Daudi cell lysate (20 µg/Lane)
Lane 8: NIH/3T3 cell lysate (20 µg/Lane)
Lane 9: Rat brain tissue lysate (40 µg/Lane)

Predicted band size: 76 kDa
Observed band size: 76/100 kDa

Exposure time: 3 minutes; ECL: K1801;

4-20% SDS-PAGE gel.

Proteins were transferred to a PVDF membrane and blocked with 5% NFDM/TBST for 1 hour at room temperature. The primary antibody (HA721562) at 1/1,000 dilution was used in 5% NFDM/TBST at 4℃ overnight. Goat Anti-Rabbit IgG - HRP Secondary Antibody (HA1001) at 1:100,000 dilution was used for 1 hour at room temperature.

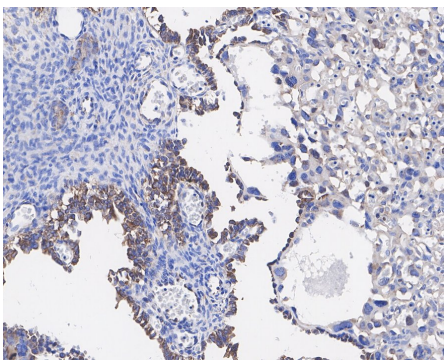

Fig2: Immunohistochemical analysis of paraffin-embedded mouse placenta tissue with Rabbit anti-GLB1 / Beta-galactosidase antibody (HA721562) at 1/1,000 dilution.

The section was pre-treated using heat mediated antigen retrieval with Tris-EDTA buffer (pH 9.0) for 20 minutes. The tissues were blocked in 1% BSA for 20 minutes at room temperature, washed with ddH_2O and PBS, and then probed with the primary antibody (HA721562) at 1/1,000 dilution for 1 hour at room temperature. The detection was performed using an HRP conjugated compact polymer system. DAB was used as the chromogen. Tissues were counterstained with hematoxylin and mounted with DPX.

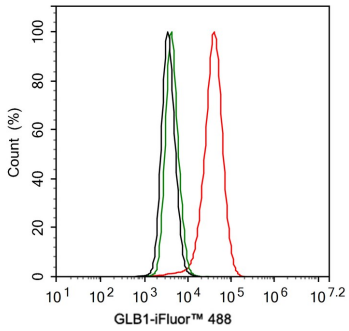

Fig3: Flow cytometric analysis of SK-Br-3 cells labeling GLB1 / Beta-galactosidase.

Cells were fixed and permeabilized. Then stained with the primary antibody (HA721562, 1ug/ml) (red) compared with Rabbit IgG Isotype Control (green). After incubation of the primary antibody at +4℃ for an hour, the cells were stained with a iFluor™ 488 conjugate-Goat anti-Rabbit IgG Secondary antibody (HA1121) at 1/1,000 dilution for 30 minutes at +4℃. Unlabelled sample was used as a control (cells without incubation with primary antibody; black).

Background References

1. Hinek A., Zhang S., Smith A.C., Callahan J.W. Impaired elastic-fiber assembly by fibroblasts from patients with either Morquio B disease or infantile GM1-gangliosidosis is linked to deficiency in the 67-kD spliced variant of beta-galactosidase. Am. J. Hum. Genet. 67:23-36 (2000).
2. Hinek A. Biological roles of the non-integrin elastin/laminin receptor. Biol. Chem. 377:471-480 (1996).