abcam

Product datasheet

Anti-GLB1/Beta-galactosidase antibody ab96239

★★★★★ 1 Abreviews 4 References 画像数 2

製品の概要

製品名	Anti-GLB1/Beta-galactosidase antibody
製品の詳細	Rabbit polyclonal to GLB1/Beta-galactosidase
由来種	Rabbit
アプリケーション	**適用あり:** WB, IHC-P
種交差性	**交差種:** Human
免疫原	Recombinant fragment. This information is proprietary to Abcam and/or its suppliers.
ポジティブ・コントロール	H1299 cell lysates.
特記事項	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
バッファー	pH: 7.00 Preservative: 0.025% Proclin 300 Constituents: 78% PBS, 1% BSA, 20% Glycerol (glycerin, glycerine)
精製度	Immunogen affinity purified
ポリ/モノ	ポリクローナル
アイソタイプ	IgG

アプリケーション

The Abpromise guarantee	Abpromise保証は、次のテスト済みアプリケーションにおけるab96239の使用に適用されます

アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
WB		1/500 - 1/3000. Predicted molecular weight: 76 kDa.
IHC-P		1/1000. Perform heat mediated antigen retrieval via the pressure cooker method before commencing with IHC staining protocol.

ターゲット情報

機能

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.

関連疾患

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.
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.
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.
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.

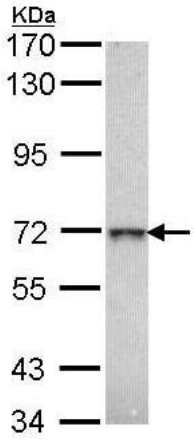
配列類似性

Belongs to the glycosyl hydrolase 35 family.

細胞内局在

Lysosome and Cytoplasm > perinuclear region. Localized to the perinuclear area of the cytoplasm but not to lysosomes.

画像

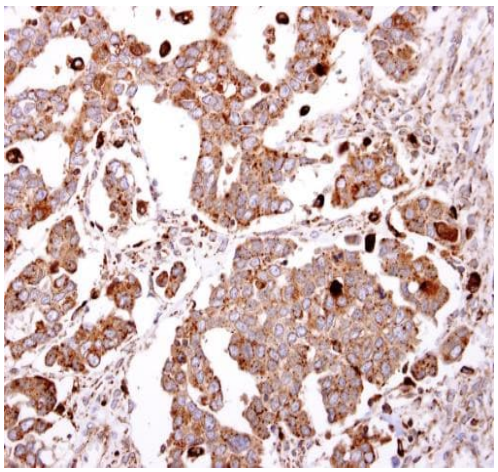


Western blot - Anti-GLB1/Beta-galactosidase antibody (ab96239)

Anti-GLB1/Beta-galactosidase antibody (ab96239) at 1/1000 dilution + H1299 whole cell lysate at 30 µg

Predicted band size: 76 kDa

7.5% SDS PAGE



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-GLB1/Beta-galactosidase antibody (ab96239)

ab96239 staining GLB1/Beta-galactosidase in human ovarian carcinoma by immunohistochemical analysis.

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