abcam

Product datasheet

Anti-GLB1/Beta-galactosidase antibody ab96239

★★★★★ 1 Abreviews 4 References 2 图像

概述

产品名称	Anti-GLB1/Beta-galactosidase**抗体**
描述	**兔多克隆抗体**to GLB1/Beta-galactosidase
宿主	Rabbit
经测试应用	**适用于:** WB, IHC-P
种属反应性	**与反应:** Human
免疫原	Recombinant fragment. This information is proprietary to Abcam and/or its suppliers.
阳性对照	H1299 cell lysates.
常规说明	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

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	pH: 7.00 Preservative: 0.025% Proclin 300 Constituents: 78% PBS, 1% BSA, 20% Glycerol (glycerin, glycerine)
纯度	Immunogen affinity purified
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee	**Abpromise™承诺保证使用**ab96239**于以下的经测试应用**

“应用说明”部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
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.

靶标

功能

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.

疾病相关

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.

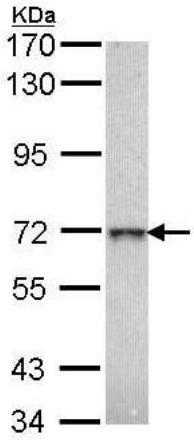
序列相似性

Belongs to the glycosyl hydrolase 35 family.

细胞定位

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

图片

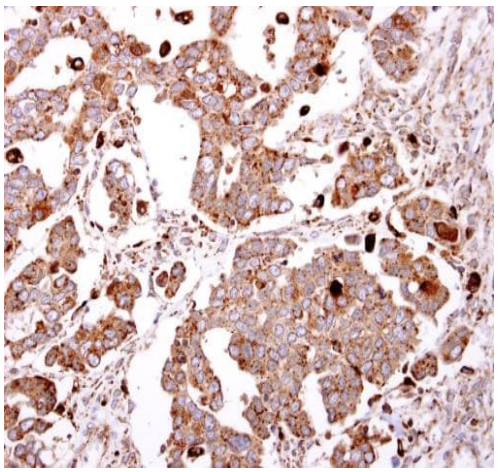


Western blot - Anti-GLB1/Beta-galactosidase antibody (ab96239)

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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