abcam

Product datasheet

Anti-GLB1/Beta-galactosidase antibody [EPR8250] ab128993

リコンビナント RabMAb

4 References 画像数 3

製品の概要

製品名	Anti-GLB1/Beta-galactosidase antibody [EPR8250]
製品の詳細	Rabbit monoclonal [EPR8250] to GLB1/Beta-galactosidase
由来種	Rabbit
アプリケーション	**適用あり:** Flow Cyt (Intra), WB **適用なし:** ICC/IF,IHC-P or IP
種交差性	**交差種:** Human **交差が予測される動物種:** Mouse, Rat
免疫原	Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.
ポジティブ・コントロール	SH-SY5Y, HeLa, Human placenta, HepG2 or BxPC-3 lysate, permeabilized HeLa cells.
特記事項	This product is a recombinant monoclonal antibody, which offers several advantages including: - High batch-to-batch consistency and reproducibility - Improved sensitivity and specificity - Long-term security of supply - Animal-free production For more information **see here**. Our RabMAb® technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to **RabMAb® patents**.

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at -20°C. Stable for 12 months at -20°C.
バッファー	pH: 7.20 Preservative: 0.01% Sodium azide Constituents: 9% PBS, 40% Glycerol (glycerin, glycerine), 0.05% BSA, 50% Tissue culture supernatant
精製度	Protein A purified
ポリ/モノ	モノクローナル
クローン名	EPR8250

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ab128993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
Flow Cyt (Intra)		1/10 - 1/100. **ab172730** - Rabbit monoclonal IgG, is suitable for use as an isotype control with this antibody.
WB		1/10000 - 1/50000. Predicted molecular weight: 76 kDa.

追加情報 Is unsuitable for ICC/IF,IHC-P or IP.

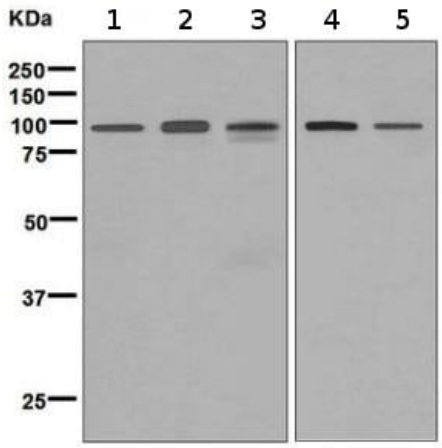
ターゲット情報

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 [EPR8250] (ab128993)

All lanes : Anti-GLB1/Beta-galactosidase antibody [EPR8250] (ab128993) at 1/10000 dilution

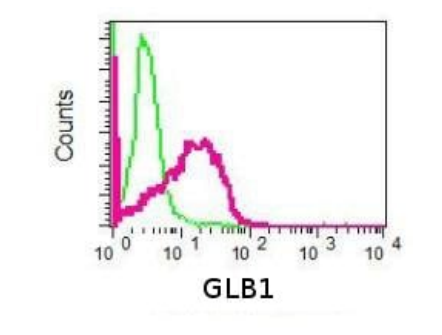
Lane 1 : SH-SY5Y cell lysate
Lane 2 : HeLa cell lysate
Lane 3 : Human placenta cell lysate
Lane 4 : HepG2 cell lysate
Lane 5 : BxPC-3 cell lysate

Lysates/proteins at 10 µg per lane.

Secondary
All lanes : HRP labelled goat

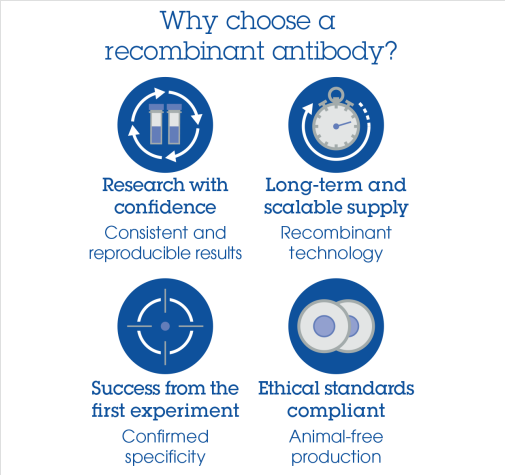
anti-rabbit at 1/2000 dilution

Predicted band size: 76 kDa



Flow Cytometry (Intracellular) - Anti-GLB1/Beta-galactosidase antibody [EPR8250] (ab128993)

Intracellular flow cytometric analysis of permeabilized HeLa cells using **ab128933** (red) or a rabbit IgG (negative) (green).



Anti-GLB1/Beta-galactosidase antibody [EPR8250]

(ab128993)

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