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Product datasheet

Anti-Alpha Dystroglycan antibody [2238] ab106110

3 References

概述

产品名称	Anti-Alpha Dystroglycan抗体[2238]
描述	**小鼠单克隆抗体**[2238] to Alpha Dystroglycan
宿主	Mouse
特异性	ab106110 is specific for a glycoepitope on brain bovine Alpha Dystroglycan, which is absent on Alpha Dystroglycan expressed in all other tissues.
经测试应用	**适用于:** WB, IHC-Fr, ELISA
种属反应性	**与反应:** Mouse, Rat, Rabbit, Cow, Human
免疫原	Other Immunogen Type corresponding to Cow Alpha Dystroglycan.

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
存储溶液	Preservative: 0.02% Sodium Azide Constituents: 0.1% BSA, PBS
纯度	Protein A purified
纯化说明	0.2 µm filtered antibody solution
克隆	单克隆
克隆编号	2238
同种型	IgG2b

应用

Our Abpromise guarantee covers the use of **ab106110** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

应用	Ab评论	说明
WB		1/50. Predicted molecular weight: 97 kDa. Recognizes Alpha-Dystroglycans as protein of ~130 kD, especially after enrichment of the lysates for dystroglycans.

应用	Ab评论	说明
IHC-Fr		1/50.
ELISA		Use at an assay dependent concentration.

靶标

功能

The dystroglycan complex is involved in a number of processes including laminin and basement membrane assembly, sarcolemmal stability, cell survival, peripheral nerve myelination, nodal structure, cell migration, and epithelial polarization.
Alpha-dystroglycan is an extracellular peripheral glycoprotein that acts as a receptor for both extracellular matrix proteins containing laminin-G domains, and for certain adenoviruses. Receptor for laminin-2 (LAMA2) and agrin in peripheral nerve Schwann cells. Also acts as a receptor for M.leprae in peripheral nerve Schwann cells but only in the presence of the G-domain of LAMA2, and for lymphocytic choriomeningitis virus, Old World Lassa fever virus, and clade C New World arenaviruses.
Beta-dystroglycan is a transmembrane protein that plays important roles in connecting the extracellular matrix to the cytoskeleton. Acts as a cell adhesion receptor in both muscle and non-muscle tissues. Receptor for both DMD and UTRN and, through these interactions, scaffolds axin to the cytoskeleton. Also functions in cell adhesion-mediated signaling and implicated in cell polarity.

组织特异性

Expressed in a variety of fetal and adult tissues. In epidermal tissue, located to the basement membrane. Also expressed in keratinocytes and fibroblasts.

疾病相关

Defects in DAG1 are the cause of muscular dystrophy-dystroglycanopathy limb-girdle type C7 (MDDGC7) [MIM:613818]. An autosomal recessive muscular dystrophy showing onset in early childhood, and associated with mental retardation without structural brain anomalies.
Note=MDDGC7 is caused by DAG1 mutations that interfere with normal post-translational processing, resulting in defective DAG1 glycosylation and impaired interactions with extracellular-matrix components. Other muscular dystrophy-dystroglycanopathies are caused by defects in enzymes involved in protein O-glycosylation.

序列相似性

Contains 1 peptidase S72 domain.

翻译后修饰

O- and N-glycosylated. Alpha-dystroglycan is heavily O-glycosylated comprising of up to two thirds of its mass and the carbohydrate composition differs depending on tissue type. Mucin-type O-glycosylation is important for ligand binding activity. O-mannosylation of alpha-DAG1 is found in high abundance in both brain and muscle where the most abundant glycan is Sia-alpha-2-3-Gal-beta-1-4-Glc-NAc-beta-1-2-Man. In muscle, glycosylation on Thr-379 by a phosphorylated O-mannosyl glycan with the structure 2-(N-acetylamido)-2-deoxygalactosyl-beta-1,3-2-(N-acetylamido)-2-deoxyglucosyl-beta-1,4-6-phosphomannose is mediated by like-acetylglucosaminyltransferase (LARGE) protein and is required for laminin binding. O-mannosylation is also required for binding lymphocytic choriomeningitis virus, Old World Lassa fever virus, and clade C New World arenaviruses. The O-glycosyl hexose on Thr-367, Thr-369, Thr-372, Thr-381 and Thr-388 is probably mannose. O-glycosylated in the N-terminal region with a core 1 or possibly core 8 glycan. The beta subunit is N-glycosylated.
Autolytic cleavage produces the alpha and beta subunits. In cutaneous cells, as well as in certain pathological conditions, shedding of beta-dystroglcan can occur releasing a peptide of about 30 kDa.

	SRC-mediated phosphorylation of the PPXY motif of the beta subunit recruits SH2 domain-containing proteins, but inhibits binding to WWW domain-containing proteins, DMD and UTRN. This phosphorylation also inhibits nuclear entry.
细胞定位	Secreted > extracellular space and Cell membrane. Cytoplasm > cytoskeleton. Nucleus > nucleoplasm. The monomeric form translocates to the nucleus via the action of importins and depends on RAN. Nuclear transport is inhibited by Tyr-892 phosphorylation. In skeletal muscle, this phosphorylated form locates to a vesicular internal membrane compartment. In peripheral nerves, localizes to the Schwann cell membrane. Colocalizes with ERM proteins in Schwann-cell microvilli.

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