肌萎缩相关蛋白 DAG1 抗体

产品货号： mIR5152

英文名称： Alpha Dystroglycan

中文名称： 肌萎缩相关蛋白 DAG1 抗体

别　　名： AGRNR; Alpha-DG; Beta-DG; Beta-dystroglycan; beta Dystroglycan; DAG; Dag1; DAG1_HUMAN; Dystroglycan 1 (dystrophin-associated glycoprotein 1); Dystroglycan; Dystrophin-associated glycoprotein 1; 156DAG; A3a; Dystrophin-associated glycoprotein 1.

研究领域： 免疫学　神经生物学　信号转导　转录调节因子　细胞骨架　细胞外基质

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Chicken, Dog, Pig, Horse, Rabbit,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 IF=1:200-800 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 98kDa

细胞定位： 细胞核 细胞浆 细胞膜 细胞外基质 分泌型蛋白

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human DAG1:451-550/895

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ℃ for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ℃.

PubMed：　PubMed

产品介绍 background:

Dystroglycan is a laminin binding component of the dystrophin-glycoprotein complex which provides a linkage between the subsarcolemmal cytoskeleton and the extracellular matrix. Dystroglycan 1 is a candidate gene for the site of the mutation in autosomal recessive muscular dystrophies. The dramatic reduction of dystroglycan 1 in Duchenne muscular dystrophy leads to a loss of linkage between the sarcolemma and extracellular matrix, rendering muscle fibers more susceptible to necrosis. Dystroglycan also functions as dual receptor for agrin and laminin-2 in the Schwann cell membrane. The muscle and nonmuscle isoforms of dystroglycan differ by carbohydrate moieties but not protein sequence. Alternative splicing results in multiple transcript variants all encoding the same protein.[provided by RefSeq, Apr 2010]

Function:

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.

Subunit:

Monomer. Heterodimer of alpha- and beta-dystroglycan subunits which are the central components of the dystrophin-glycoprotein complex.

Subcellular Location:

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.

Tissue Specificity:

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

Post-translational modifications:

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.

Similarity:

Contains 1 peptidase S72 domain.

SWISS:

Q14118

Gene ID:

1605

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

产品图片