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Alpha Dystroglycan Rabbit pAb

Catalog # AP99517

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q14118
Other Accession	Q14118
Reactivity	Rat, Mouse
Predicted	Pig, Human, Rabbit, Chicken, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	97441
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human DAG1
Epitope Specificity	451-550/895
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 1 peptidase S72 domain.
SUBUNIT	Monomer. Heterodimer of alpha- and beta-dystroglycan subunits which are the central components of the dystrophin-glycoprotein complex.
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-acety lamido)-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.
DISEASE	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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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]

Additional Information

Gene ID	1605
Other Names	156DAG; A3a; AGRNR; DAG; LGMDR16; MDDGA9; MDDGC7; MDDGC9; D9Wsu13e; DG; Dp427; Dp71; DAG1_HUMAN; DAG1; Dystroglycan; Dystrophin-associated glycoprotein 1; DAG1_MOUSE; dystroglycan 1; dystroglycan 1 (dystrophin-associated glycoprotein 1); alpha-dystroglycan; dystrophin-associated glycoprotein-1; beta-dystroglycan
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:200-800
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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 °C.

Protein Information

Name	DAG1 (HGNC:2666)
Function	The dystroglycan complex is involved in a number of signaling events and processes including laminin deposition and extracellular matrix assembly, acetylcholine receptor clustering, sarcolemmal stability, cell survival, peripheral nerve myelination, nodal structure, cell migration, epithelial polarization, and epithelium branching morphogenesis (By similarity). Required for the formation of photoreceptor ribbon synapses, and long-term maintenance of inhibitory synapses in cerebellar Purkinje cells (By similarity). Also involved in the positive regulation of cartilage formation through agrin (AGRN) binding and up-regulation of SOX9, a transcription factor that plays a key role in chondrocytes differentiation (PubMed:26290588). [Beta-dystroglycan]: 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.
Cellular Location	[Alpha-dystroglycan]: Secreted, extracellular space. Secreted, extracellular space, extracellular matrix, basement membrane {ECO:0000250|UniProtKB:Q62165} Synapse

	{ECO:0000250|UniProtKB:Q62165}. Note=Localized to inhibitory synapses in the cerebellar cortex. {ECO:0000250|UniProtKB:Q62165}
Tissue Location	Expressed in a variety of fetal and adult tissues. In epidermal tissue, located to the basement membrane. Also expressed in keratinocytes and fibroblasts.

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