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## Rabbit Anti-Alpha Dystroglycan/AF350 Conjugated antibody

SL5152R-AF350

**Product Name** Anti-Alpha Dystroglycan/AF350

**Chinese Name** AF350 标记的肌萎缩相关蛋白 DAG1 抗体

**Alias** AGRNR; Alpha-DG; Beta-DG; Beta-dystroglycan; beta Dystroglycan; DAG; Dag1; DAG1\_HU  
Dystroglycan 1 (dystrophin-associated glycoprotein 1); Dystroglycan; Dystrophin-associated gly  
156DAG; A3a; Dystrophin-associated glycoprotein 1.

**Research Area** immunology Neurobiology Signal transduction transcriptional regulatory factor Cytoskeleton Ex  
matrix

**Immunogen Species** Rabbit

**Clonality** Polyclonal

**React Species** Mouse,Rat(predicted:Human,Chicken,Dog,Pig,Horse,Rabbit)  
IF=1:100-500

**Applications** not yet tested in other applications.  
optimal dilutions/concentrations should be determined by the end user.

**Molecular weight** 98kDa

**Form** Lyophilized or Liquid

**Concentration** 1mg/ml

**immunogen** KLH conjugated synthetic peptide derived from human DAG1 (501-550aa)

**Lsotype** IgG

**Purification** affinity purified by Protein A

**Storage Buffer** 1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol

**Storage** Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at  
temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in  
7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

### background:

**Product Detail** Dystroglycan is a laminin binding component of the dystrophin-glycoprotein complex which pro  
between the subsarcolemmal cytoskeleton and the extracellular matrix. Dystroglycan 1 is a candi  
site of the mutation in autosomal recessive muscular dystrophies. The dramatic reduction of dyst

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 laminin-2 in the Schwann cell membrane. The muscle and nonmuscle isoforms of dystroglycan differ in their carbohydrate moieties but not protein sequence. Alternative splicing results in multiple transcripts 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-cell 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 (LA2) in peripheral nerve Schwann cells. Also acts as a receptor for M.leprae in peripheral nerve Schwann cells. In the presence of the G-domain of LAMA2, and for lymphocytic choriomeningitis virus, Old World monkey 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 laminin-2 and UTRN and, through these interactions, scaffolds axin to the cytoskeleton. Also functions in cell-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 > nucleolus. The monomeric form translocates to the nucleus via the action of importins and depends on RAN. Nuclear import is inhibited by Tyr-892 phosphorylation. In skeletal muscle, this phosphorylated form localizes to a vesicular membrane compartment. In peripheral nerves, localizes to the Schwann cell membrane. Colocalizes with other 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 thousand glycan structures and the carbohydrate composition differs depending on tissue type. Mucin-type O-glycosylation is essential 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-GlcNAc-beta-1-2-Man. In muscle, phosphorylation 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-phosphate 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 La

and clade C New World arenaviruses. The O-glycosyl hexose on Thr-367, Thr-369, Thr-372, Thr-388 is probably mannose. O-glycosylated in the N-terminal region with a core 1 or possibly The beta subunit is N-glycosylated. Autolytic cleavage produces the alpha and beta subunits. In addition, as well as in certain pathological conditions, shedding of beta-dystroglycan can occur releasing a peptide of approximately 100 kDa.

SRC-mediated phosphorylation of the PPXY motif of the beta subunit recruits SH2 domain-containing proteins, but inhibits binding to WW domain-containing proteins, DMD and UTRN. This phosphorylation is required for nuclear entry.

**Similarity:**

Contains 1 peptidase S72 domain.

**Database links:**

[Entrez Gene: 1605](#) Human

[Entrez Gene: 13138](#) Mouse

[Entrez Gene: 100009278](#) Rabbit

[Entrez Gene: 114489](#) Rat

[Omim: 128239](#) Human

[SwissProt: Q14118](#) Human

[SwissProt: Q62165](#) Mouse

[Unigene: 76111](#) Human

[Unigene: 7524](#) Mouse

[Unigene: 36260](#) Rat

**Important Note:**

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