abcam

Product datasheet

Recombinant Human DAG1 protein ab175460

1 Image

Description

Product name	Recombinant Human DAG1 protein
Purity	> 90 % Densitometry. ab175460 is purifed using Ni-NTA chromatography and filtered (0.4 µm).
Endotoxin level	< 1.000 Eu/µg
Expression system	Escherichia coli
Accession	Q14118
Protein length	Protein fragment
Animal free	No
Nature	Recombinant
Species	Human
Sequence	MKHHHHHHASHWPSEPSEAVRDWENQLEASMHSVLSD LHEAVPTVVGIPD GTAVVGRSFRVTIPTDLIASSGDIIKVSAAGKEALPSWLHW DSQSHTLEG LPLDTDKGVHYISVSATRLGANGSHIPQTSSVFSIEVYPED HSELQSVRT ASPDPGEVVSSACAADEPVTVLTVILDADLTKMTPKQRID LLHRMRSFSE VELHNMKLVPVVNNRLFDMSAFMAGPGNAKKVVENGAL LSWKLGCSLNQN SVPDIHGVEAPAREGAMSAQLGYPVVGWHIANKKPPLPK RVRR
Predicted molecular weight	32 kDa including tags
Amino acids	30 to 312
Tags	His tag N-Terminus

Specifications

Our **Abpromise guarantee** covers the use of **ab175460** in the following tested applications.

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

Applications	Western blot
	Mass Spectrometry

	ELISA
	SDS-PAGE
Mass spectrometry	LC-MS/MS
Form	Lyophilized

Preparation and Storage

Stability and Storage	Shipped at 4°C. Store at -80°C. Avoid freeze / thaw cycle.
	Constituents: 0.44% Sodium chloride, 99% Phosphate Buffer
Reconstitution	Add 200ul of deionized water to prepare a working stock solution of 0.5 mg/mL and let the lyophilized pellet dissolve completely.

General Info

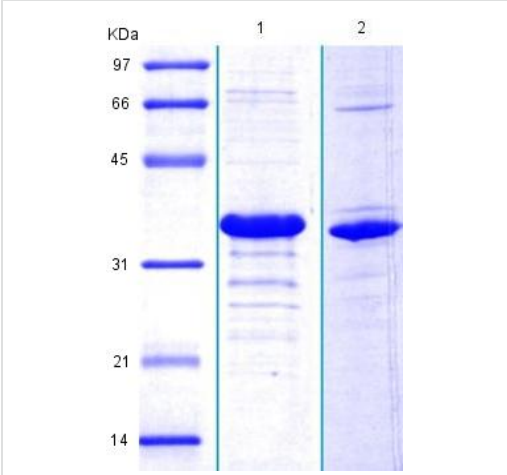
Function	The dystroglycan complex is involved in a number of processes including laminin and basement membrane assembly, sarcolemmal stability, cell survival, peripheral nerve myelination, nodal structure, cell migration, and epithelial polarization. Alpha-dystroglycan is an extracellular peripheral 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.
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.
Involvement in 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.
Sequence similarities	Contains 1 peptidase S72 domain.
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.

Cellular localization

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.

Images



SDS-PAGE - Recombinant Human DAG1 protein (ab175460)

14% SDS-PAGE analysis of ab175460.

Lane 1: Reduced and heated sample, 2.5 µg/lane

Lane 2: Non-reduced and non-heated sample, 25 µg/lane.

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