

DESM Antibody

Mouse Monoclonal Antibody (Mab)

Catalog # AM1926b

Product Information

Application	WB, E
Primary Accession	P17661
Other Accession	NP_001918.3
Reactivity	Human
Host	Mouse
Clonality	Monoclonal
Isotype	IgM,k
Clone Names	290CT7.1.1
Calculated MW	53536

Additional Information

Gene ID	1674
Other Names	Desmin, DES
Target/Specificity	This DESM monoclonal antibody is generated from mouse immunized with DESM recombinant protein.
Dilution	WB~~1:500~1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.05% (V/V) Proclin 300. This antibody is prepared by Saturated Ammonium Sulfate (SAS) precipitation followed by dialysis against PBS.
Storage	Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	DESM Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name	DES
Function	Muscle-specific type III intermediate filament essential for proper muscular structure and function. Plays a crucial role in maintaining the structure of sarcomeres, inter-connecting the Z-disks and forming the myofibrils, linking them not only to the sarcolemmal cytoskeleton, but also to the nucleus and mitochondria, thus providing strength for the muscle fiber during activity (PubMed: 25358400). In adult striated muscle they form a fibrous network

connecting myofibrils to each other and to the plasma membrane from the periphery of the Z-line structures (PubMed:[24200904](#), PubMed:[25394388](#), PubMed:[26724190](#)). May act as a sarcomeric microtubule-anchoring protein: specifically associates with dephosphorylated tubulin- α chains, leading to buckled microtubules and mechanical resistance to contraction. Required for nuclear membrane integrity, via anchoring at the cell tip and nuclear envelope, resulting in maintenance of microtubule-derived intracellular mechanical forces (By similarity). Contributes to the transcriptional regulation of the NKX2-5 gene in cardiac progenitor cells during a short period of cardiomyogenesis and in cardiac side population stem cells in the adult. Plays a role in maintaining an optimal conformation of nebulin (NEB) on heart muscle sarcomeres to bind and recruit cardiac α -actin (By similarity).

Cellular Location

Cytoplasm, myofibril, sarcomere, Z line. Cytoplasm Cell membrane, sarcolemma. Nucleus {ECO:0000250|UniProtKB:P31001}. Cell tip {ECO:0000250|UniProtKB:P31001}. Nucleus envelope {ECO:0000250|UniProtKB:P31001}. Note=Localizes in the intercalated disks which occur at the Z line of cardiomyocytes (PubMed:24200904, PubMed:26724190). Localizes in the nucleus exclusively in differentiating cardiac progenitor cells and premature cardiomyocytes (By similarity). PKP2 is required for correct anchoring of DES at the cell tip and nuclear envelope (By similarity) {ECO:0000250|UniProtKB:P31001, ECO:0000269|PubMed:24200904, ECO:0000269|PubMed:26724190}

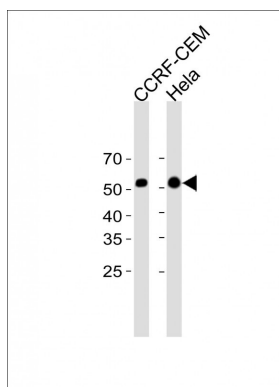
Background

This gene encodes a muscle-specific class III intermediate filament. Homopolymers of this protein form a stable intracytoplasmic filamentous network connecting myofibrils to each other and to the plasma membrane. Mutations in this gene are associated with desmin-related myopathy, a familial cardiac and skeletal myopathy (CSM), and with distal myopathies. [provided by RefSeq].

References

Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010)
van Spaendonck-Zwarts, K., et al. Clin. Genet. (2010) In press :
Zimmerman, R.S., et al. Genet. Med. 12(5):268-278(2010)
Bar, H., et al. J. Mol. Biol. 397(5):1188-1198(2010)
Levin, J., et al. J. Neuropathol. Exp. Neurol. 69(4):415-424(2010)

Images



All lanes: Anti-DESM Antibody at 1:1000 dilution Lane 1: CCRF-CEM whole cell lysate Lane 2: HeLa whole cell lysate Lysates/proteins at 20 μ g per lane. Secondary: Goat Anti-Mouse IgM, (H+L), Peroxidase conjugated (ASP1613) at 1/8000 dilution. Observed band size: 54 kDa Blocking/Dilution buffer: 5% NFDN/TBST.