

Desmin Antibody

Purified Mouse Monoclonal Antibody Catalog # AO1075a

Product Information

Application Primary Accession Reactivity Host Clonality Clone Names Isotype Calculated MW Description	 WB, IHC, E P17661 Human Mouse Monoclonal 10H7D2 IgG1 53536 Desmin (DES), with 470-amino acid protein (about 52kDa), belongs to the intermediate filament family and Desmin is class III intermediate filaments found in muscle cells. Homopolymers of Desmin form a stable intracytoplasmic filamentous network connecting myofibrils to each other and to the plasma membrane.Mutations in Desmin are associated with desmin-related myopathy, a familial cardiac and skeletal myopathy (CSM), and with distal myopathies.Desmin is also expressed in smooth muscle cells of both airways and alveolar ducts and Desmin is a load-bearing protein that stiffens the airways and consequently the lung and modulates airway contractile response.
Immunogen	Purified recombinant fragment of Desmin expressed in E. Coli.
Formulation	Purified antibody in PBS containing 0.03% sodium azide.

Additional Information

Gene ID	1674
Other Names	Desmin, DES
Dilution	WB~~1/500 - 1/2000 IHC~~1/200 - 1/1000 E~~N/A
Storage	Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.
Precautions	Desmin Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Protein Information

Name

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: <u>25358400</u>). 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: <u>24200904</u> , PubMed: <u>25394388</u> , PubMed: <u>26724190</u>). May act as a sarcomeric microtubule-anchoring protein: specifically associates with detyrosinated tubulin-alpha 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 nebulette (NEB) on heart muscle sarcomeres to bind and recruit cardiac alpha-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}

References

1. Felix R. Shardonofsky, Yassemi Capetanaki, and Aladin M. Boriek. Am J Physiol Lung Cell Mol Physiol, May 2006; 290: L890 - L896. 2. Xupei Huang, Jian Li, Dalton Foster. Experimental Biology and Medicine, Dec 2002; 227: 1039.

Images

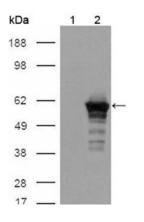
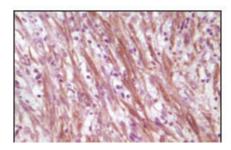


Figure 1: Western blot analysis using Desmin mouse mAb against HEK293T cells transfected with the pCMV6-ENTRY control (1) and pCMV6-ENTRY Desmin cDNA (2).

Figure 2: Immunohistochemical analysis of paraffin-embedded human smooth musde sarcoma, showing cytoplasmic localization using Desmin mouse mAb with DAB staining.



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