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DESM Antibody

Mouse Monoclonal Antibody (Mab)
Catalog # AM1926b

Product Information

Application	WB, E
Primary Accession	P17661
Other Accession	NP_001918.3
Reactivity	Human
Predicted	Mouse, Rat, Bovine, Canine, Rabbit, Chicken
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~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified monoclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is prepared by Euglobin 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 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}

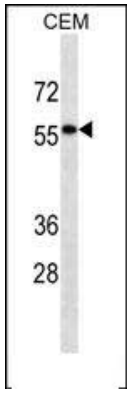
Background

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



DESM Antibody (Cat. #AM1926b) western blot analysis in CEM cell line lysates (35µg/lane).This demonstrates the DESM antibody detected the DESM protein (arrow).

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.