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Desmin Antibody (C-term)

Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP14284b

Product Information

Application	WB, IHC-P, E
Primary Accession	P17661
Other Accession	P48675, P02540, P31001, O62654, NP_001918.3
Reactivity	Human
Predicted	Bovine, Mouse, Pig, Rat
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB35066
Calculated MW	53536
Antigen Region	332-359

Additional Information

Gene ID	1674
Other Names	Desmin, DES
Target/Specificity	This Desmin antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 332-359 amino acids from the C-terminal region of human Desmin.
Dilution	WB~~1:1000 IHC-P~~1:100~500 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.
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	Desmin Antibody (C-term) 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}

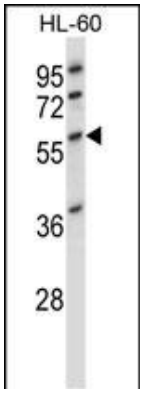
Background

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].

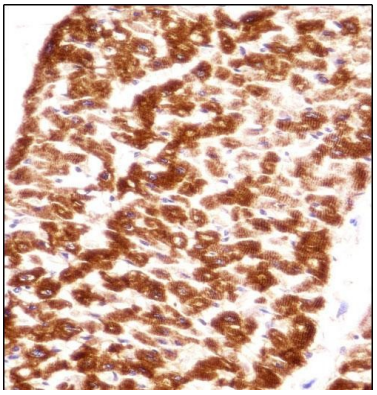
References

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



DES/Desmin (Muscle Cell Marker) Antibody (C-term) (Cat. #AP14284b) western blot analysis in HL-60 cell line lysates (35ug/lane).This demonstrates the DES antibody detected the DES protein (arrow).



DES/Desmin (Muscle Cell Marker) Antibody (C-term) (AP14284b)immunohistochemistry analysis in formalin fixed and paraffin embedded human heart tissue followed by peroxidase conjugation of the secondary antibody and DAB staining.This data demonstrates the use of DES/Desmin (Muscle Cell Marker) Antibody (C-term) for immunohistochemistry. Clinical relevance has not been evaluated.