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Rabbit Anti-Desmin/AP Conjugated antibody

SL1026R-AP

Product Name	Anti-Desmin/AP
Chinese Name	碱性磷酸酶（AP）标记的结蛋白抗体
Alias	CMD1I; CSM1; CSM2; DES; FLJ12025; FLJ39719; FLJ41013; FLJ41793; Intermediate filament protein; OTTHUMP00000064865; DESM_HUMAN; Desmin; FLJ12025; FLJ39719; FLJ41013; FLJ41793.
Research Area	Tumour Cardiovascular immunology Signal transduction Cell type markers
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Human,Mouse,Rat
Applications	IHC-P=1:50-200 IHC-F=1:50-200 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	52kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human Desmin
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol.
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Product Detail	**background:** filaments found in muscle cells. 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. Defects in Desmin are the cause of desmin related cardio skeletal myopathy (CSM) also known as desmin related myopathy (DRM). CSM is characterized by skeletal muscle weakness associated with cardiac conduction blocks, arrhythmias, restrictive heart

failure, and by intracytoplasmic accumulation of desmin reactive deposits in cardiac and skeletal muscle cells. A desmin related myopathy can have a distal onset, it is then known as hereditary distal myopathy (HDM). Defects in Desmin are also the cause of dilated cardiomyopathy type 1I (CMD1I). CMD1I is an autosomal form of dilated cardiomyopathy characterized by ventricular dilatation and impaired systolic function. Antidesmin antibodies are useful in identification of tumours of myogenic origin.

Function:
Desmin are class-III intermediate filaments found in muscle cells. 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.

Subunit:
Homopolymer. Interacts with DST. Interacts with MTM1.

Subcellular Location:
Cytoplasm.

Post-translational modifications:
ADP-ribosylation prevents ability to form intermediate filaments.

DISEASE:
Defects in DES are the cause of myopathy myofibrillar type 1 (MFM1) [MIM:601419]. A neuromuscular disorder characterized by skeletal muscle weakness associated with cardiac conduction blocks, arrhythmias, restrictive heart failure, and by myofibrillar destruction with intracytoplasmic accumulation of desmin-reactive deposits in cardiac and skeletal muscle cells. Note=Mutations in the DES gene are associated with a variable clinical phenotype which encompasses isolated myopathies, pure cardiac phenotypes (including dilated cardiomyopathy, restrictive cardiomyopathy and arrhythmogenic right ventricular cardiomyopathy), cardiac conduction disease, and combinations of these disorders. If both cardiologic and neurologic features occur, they can manifest in any order, as cardiologic features can precede, occur simultaneously with, or follow manifestation of generalized neuromuscular disease (PubMed:19879535).
Defects in DES are the cause of cardiomyopathy dilated type 1I (CMD1I) [MIM:604765]. Dilated cardiomyopathy is a disorder characterized by ventricular dilation and impaired systolic function, resulting in congestive heart failure and arrhythmia. Patients are at risk of premature death.
Defects in DES are the cause of neurogenic scapuloperoneal syndrome Kaeser type (Kaeser syndrome) [MIM:181400]. Kaeser syndrome is an autosomal dominant disorder with a peculiar scapuloperoneal distribution of weakness and atrophy. A large clinical variability is observed ranging from

scapuloperoneal, limb grindle and distal phenotypes with variable cardiac or respiratory involvement. Facial weakness, dysphagia and gynaecomastia are frequent additional symptoms. Affected men seemingly bear a higher risk of sudden, cardiac death as compared to affected women. Histological and immunohistochemical examination of muscle biopsy specimens reveal a wide spectrum of findings ranging from near normal or unspecific pathology to typical, myofibrillar changes with accumulation of desmin.

Similarity:
Belongs to the intermediate filament family.

Database links:

Entrez Gene: 1674 Human

Entrez Gene: 13346 Mouse

Entrez Gene: 64362 Rat

Omim: 125660 Human

SwissProt: P17661 Human

SwissProt: P31001 Mouse

SwissProt: P48675 Rat

Unigene: 594952 Human

Unigene: 6712 Mouse

Unigene: 39196 Rat

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Desmin 在很多哺乳动物中的横纹肌和各种平滑肌及其来源的 Tumour 组织中都有表达。结蛋白是一种中间丝蛋白，广泛分布于骨骼肌细胞、平滑肌细胞、心肌细胞和肌 epithelial cells 及其 Tumour 中，主要用于子宫、皮肤、胃肠道及其它横纹肌肉瘤和肌上皮瘤的诊断和鉴别诊断。