

## 小鼠抗结蛋白单克隆抗体

产品货号： mlR0195

英文名称： Desmin

中文名称： 小鼠抗结蛋白单克隆抗体

别名： CMD1i; CSM1; CSM2; DES; FLJ12025; FLJ39719; FLJ41013; FLJ41793; Intermediate filament protein; OTTHUMP00000064865; DESM\_HUMAN; Desmin; FLJ12025; FLJ39719; FLJ41013; FLJ41793.

研究领域： 肿瘤 心血管 免疫学 信号转导

抗体来源： Mouse

克隆类型： Monoclonal

克隆号： 4B12

交叉反应： Human,

产品应用： IHC-P=1:100-500 IHC-F=1:100-500 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分子量： 52kDa

细胞定位： 细胞浆

性状： Lyophilized or Liquid

浓度： 1mg/ml

免疫原： KLH conjugated synthetic peptide derived from human Desmin:

**亚 型 :** IgG1

**纯化方法 :** affinity purified by Protein G

**储 存 液 :** 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

**保存条件 :** 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 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

**PubMed :** PubMed

**产品介绍 :** 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 girdle 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.

**SWISS:**

P17661

**Gene ID:**

1674

**Important Note:**

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

产品图片

