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Product datasheet

Anti-Desmin antibody [RD301] ab8976

3 References 2 Images

Overview

Product name	Anti-Desmin antibody [RD301]
Description	Mouse monoclonal [RD301] to Desmin
Host species	Mouse
Specificity	This antibody reacts exclusively with desmin, which is expressed in smooth and striated muscle cells and their tumors e.g. rhabdomyosarcoma and leiomyosarcoma.
Tested applications	**Suitable for:** ICC/IF, IHC-Fr, WB
Species reactivity	**Reacts with:** Mouse, Rat, Rabbit, Chicken, Hamster, Dog, Human, Pig **Does not react with:** Zebrafish
Immunogen	Cytoskeletal desmin extract of chicken gizzard.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Storage buffer	Preservative: 0.09% Sodium azide Constituent: PBS
Purity	Protein A purified
Clonality	Monoclonal
Clone number	RD301
Myeloma	Sp2/0-Ag14
Isotype	IgG2b
Light chain type	unknown

Applications

Our Abpromise guarantee covers the use of **ab8976** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
ICC/IF		Use at an assay dependent concentration. Permeabilised cells
IHC-Fr		1/100 - 1/2000. with avidin-biotinylated horseradish peroxidase complex (ABC) as detection reagent.
WB		1/100 - 1/1000.

Target

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.

Involvement in disease

Defects in DES are the cause of myopathy myofibrillar desmin-related (MFM-DES) [MIM:601419]; also known as desmin-related myopathy (DRM). 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.
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.

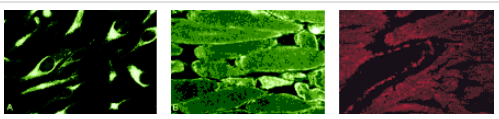
Sequence similarities

Belongs to the intermediate filament family.

Cellular localization

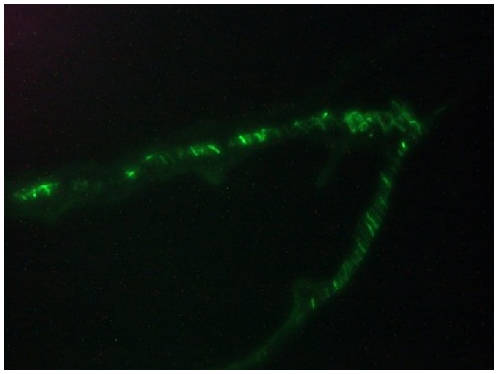
Cytoplasm.

Images



Immunohistochemistry (Frozen sections) - Anti-Desmin antibody [RD301] (ab8976)

Fluorescence microscopy of (a) cultured BHK-21 cells, (b) human skeletal muscle and (c) humanmyocardium stained with the monoclonal desmin antibody ab8976.



Immunofluorescence staining of a 7 days old zebrafish embryo

Immunohistochemistry (Frozen sections) - Anti-Desmin antibody [RD301] (ab8976)

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