

Product datasheet

Anti-Desmin antibody [RD301] ab8976

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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: IHC-Fr
Species reactivity	Reacts with: Human, Zebrafish
Immunogen	Tissue, cells or virus corresponding to Chicken Desmin. Cytoskeletal desmin extract of chicken gizzard.
General notes	<p>The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.</p> <p>If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As</p>

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

The Abpromise guarantee

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
IHC-Fr		1/100 - 1/2000. with avidin-biotinylated horseradish peroxidase complex (ABC) as detection reagent.

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 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.

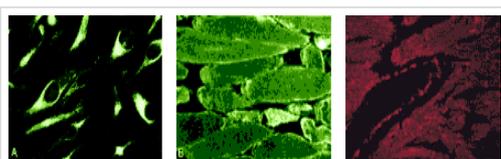
Sequence similarities

Belongs to the intermediate filament family.

Cellular localization

Cytoplasm.

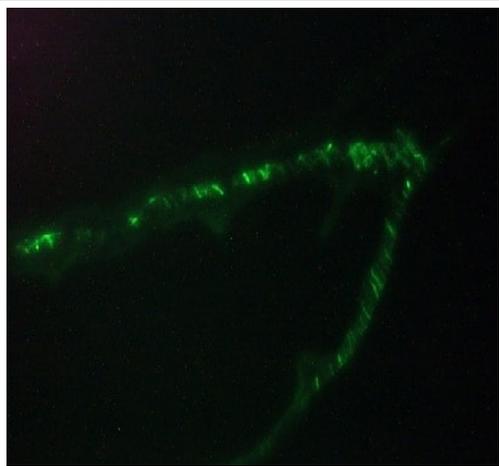
Images



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

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

Immunofluorescence staining of a 7 days old zebrafish embryo



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

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