abcam

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

SpecificityThis 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 notesThe 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.

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

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

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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-Ill 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

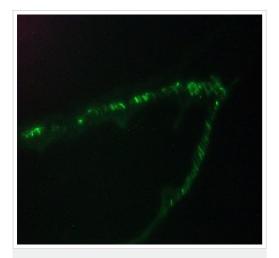






Immunohistochemistry (Frozen sections) - Anti-Desmin antibody [RD301] (ab8976) Fluorescence microscopy of (a) cultured BHK-21 cells, (b) human skeletal muscle and (c) humanmyocardium stained with the monoclonal desmin antibody ab8976.

Immunofluorescence staining of a 7 days old zebrafish embryo



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

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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