

Anti-Desmin antibody [RD301] ab8976

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製品の概要

製品名	Anti-Desmin antibody [RD301]
製品の詳細	Mouse monoclonal [RD301] to Desmin
由来種	Mouse
特異性	This antibody reacts exclusively with desmin, which is expressed in smooth and striated muscle cells and their tumors e.g. rhabdomyosarcoma and leiomyosarcoma.
アプリケーション	適用あり: IHC-Fr
種交差性	交差種: Human, Zebrafish
免疫原	Tissue, cells or virus corresponding to Chicken Desmin. Cytoskeletal desmin extract of chicken gizzard.
特記事項	<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>

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
バッファー	Preservative: 0.09% Sodium azide Constituent: PBS
精製度	Protein A purified
ポリ/モノ	モノクローナル
クローン名	RD301
ミエローマ	Sp2/0-Ag14
アイソタイプ	IgG2b
軽鎖の種類	unknown

アプリケーション

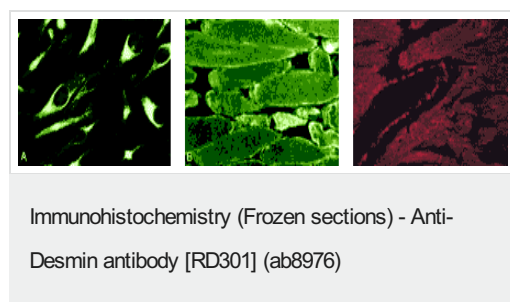
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アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
IHC-Fr		1/100 - 1/2000. with avidin-biotinylated horseradish peroxidase complex (ABC) as detection reagent.

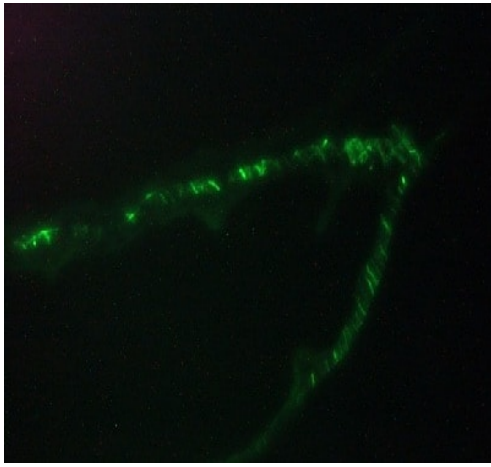
ターゲット情報

機能	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.
関連疾患	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.
配列類似性	Belongs to the intermediate filament family.
細胞内局在	Cytoplasm.

画像



Fluorescence microscopy of (a) cultured BHK-21 cells, (b) human skeletal muscle and (c) human myocardium 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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