

Anti-Desmin antibody [RD301] ab8976

★★★★★ [1 Abreviews](#) [14 References](#) [2 图像](#)

概述

产品名称	Anti-Desmin抗体[RD301]
描述	小鼠单克隆抗体[RD301] to Desmin
宿主	Mouse
特异性	This antibody reacts exclusively with desmin, which is expressed in smooth and striated muscle cells and their tumors e.g. rhabdomyosarcoma and leiomyosarcoma.
经测试应用	适用于: IHC-Fr
种属反应性	与反应: Human, Zebrafish
免疫原	Tissue, cells or virus corresponding to Chicken Desmin. Cytoskeletal desmin extract of chicken gizzard.
常规说明	<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>

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: 0.09% Sodium azide Constituent: PBS
纯度	Protein A purified
克隆	单克隆
克隆编号	RD301
骨髓瘤	Sp2/0-Ag14
同种型	IgG2b
轻链类型	unknown

应用

The Abpromise guarantee **Abpromise™**承诺保证使用ab8976于以下的经测试应用

“应用说明”部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

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

靶标

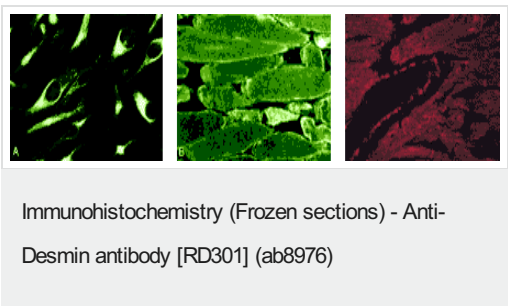
功能 Desmin are class-III intermediate filaments found in muscle cells. In adult striated muscle they form a fibrous network connecting myofibrils to each other and to the plasma membrane from the periphery of the Z-line structures.

疾病相关 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 grindler and distal phenotypes with variable cardiac or respiratory involvement. Facial weakness, dysphagia and gynecomastia 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.

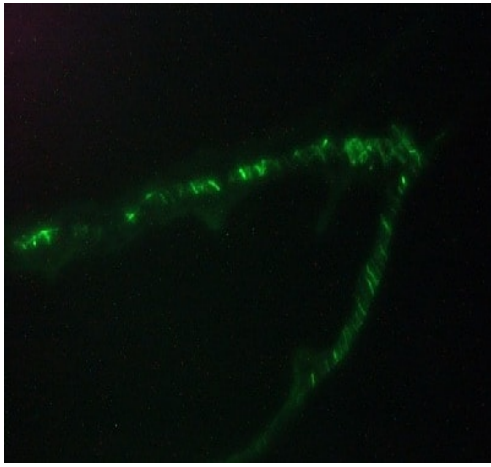
序列相似性 Belongs to the intermediate filament family.

细胞定位 Cytoplasm.

图片



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)

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

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <https://www.abcam.cn/abpromise> or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors