




Anti-Desmin antibody ab80503

5 References **1 图像**

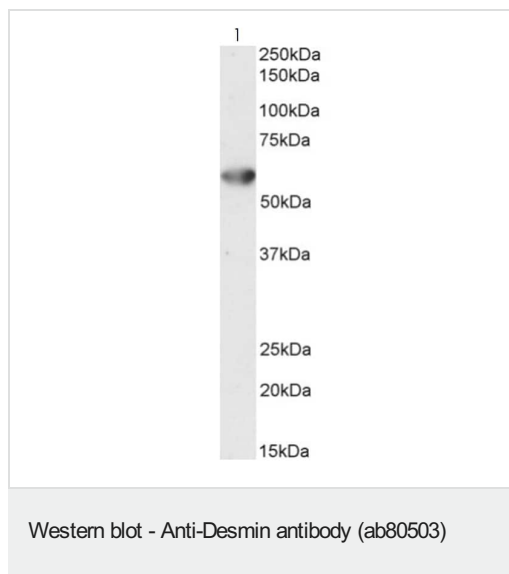
概述

产品名称	Anti-Desmin抗体
描述	山羊多克隆抗体to Desmin
宿主	Goat
经测试应用	适用于: WB
种属反应性	与反应: Mouse 预测可用于: Cow, Dog 
免疫原	Synthetic peptide: C-RDGEVVSEATQQHE , corresponding to C terminal amino acids 454 - 468 of Human Desmin (NP_001918.3)  Run BLAST with  Run BLAST with
阳性对照	WB: Mouse skeletal muscle.
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. Avoid repeated freeze / thaw cycles.
存储溶液	pH: 7.30 Preservative: 0.02% Sodium azide Constituents: 0.5% BSA, Tris buffered saline
纯度	Ammonium Sulphate Precipitation
纯化说明	ab80503 is purified from goat serum by ammonium sulphate precipitation followed by antigen affinity chromatography using the immunizing peptide
克隆	多克隆

同种型	IgG	
应用		
The Abpromise guarantee Abpromise™ 承诺保证使用ab80503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
WB		Use a concentration of 0.1 - 0.3 µg/ml. Predicted molecular weight: 54 kDa. 1 hour primary incubation is recommended for this product.
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 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.	
序列相似性	Belongs to the intermediate filament family.	
细胞定位	Cytoplasm.	
图片		



Anti-Desmin antibody (ab80503) at 0.5 µg/ml + Mouse skeletal muscle lysate at 35 µg

Predicted band size: 54 kDa

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