


### Anti-Dystrophin antibody ab15277

★★★★★ [23 Abreviews](#) [386 References](#) [4 图像](#)

#### 概述

产品名称	Anti-Dystrophin抗体
描述	兔多克隆抗体to Dystrophin
宿主	Rabbit
经测试应用	适用于: IHC-Fr, IHC-P
种属反应性	与反应: Mouse, Human 预测可用于: Rat, Dog, Pig 
免疫原	Synthetic peptide within Human Dystrophin aa 3650 to the C-terminus (C terminal). The exact sequence is proprietary. Database link: <a href="#">P11532</a>
常规说明	<p><b>This product is FOR RESEARCH USE ONLY. For commercial use, please contact <a href="mailto:partnerships@abcam.com">partnerships@abcam.com</a>.</b></p> <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&amp;As</p>

#### 性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
存储溶液	pH: 7.60 Preservative: 0.1% Sodium azide Constituents: PBS, 1% BSA
纯度	Immunogen affinity purified
克隆	多克隆
同种型	IgG

## 应用

### The Abpromise guarantee

**Abpromise™**承诺保证使用ab15277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	★★★★★ (6)	1/400. Use with acetone-fixed tissues.
IHC-P	★★★★★ (14)	1/100.

## 靶标

### 功能

Anchors the extracellular matrix to the cytoskeleton via F-actin. Ligand for dystroglycan. Component of the dystrophin-associated glycoprotein complex which accumulates at the neuromuscular junction (NMJ) and at a variety of synapses in the peripheral and central nervous systems and has a structural function in stabilizing the sarcolemma. Also implicated in signaling events and synaptic transmission.

### 组织特异性

Expressed in muscle fibers accumulating in the costameres of myoplasm at the sarcolemma. Expressed in brain, muscle, kidney, lung and testis. Isoform 5 is expressed in heart, brain, liver, testis and hepatoma cells. Most tissues contain transcripts of multiple isoforms, however only isoform 5 is detected in heart and liver.

### 疾病相关

Defects in DMD are the cause of Duchenne muscular dystrophy (DMD) [MIM:310200]. DMD is the most common form of muscular dystrophy; a sex-linked recessive disorder. It typically presents in boys aged 3 to 7 year as proximal muscle weakness causing waddling gait, toe-walking, lordosis, frequent falls, and difficulty in standing up and climbing up stairs. The pelvic girdle is affected first, then the shoulder girdle. Progression is steady and most patients are confined to a wheelchair by age of 10 or 12. Flexion contractures and scoliosis ultimately occur. About 50% of patients have a lower IQ than their genetic expectations would suggest. There is no treatment.

Defects in DMD are the cause of Becker muscular dystrophy (BMD) [MIM:300376]. BMD resembles DMD in hereditary and clinical features but is later in onset and more benign.

Defects in DMD are a cause of cardiomyopathy dilated X-linked type 3B (CMD3B) [MIM:302045]; also known as X-linked dilated cardiomyopathy (XLCM). 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.

### 序列相似性

Contains 2 CH (calponin-homology) domains.

Contains 22 spectrin repeats.

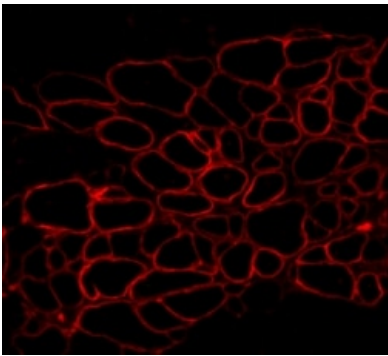
Contains 1 WW domain.

Contains 1 ZZ-type zinc finger.

### 细胞定位

Cell membrane > sarcolemma. Cytoplasm > cytoskeleton.

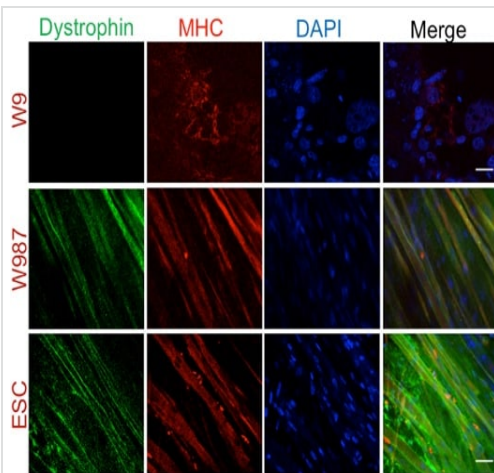
## 图片



Immunohistochemistry (Frozen sections) - Anti-Dystrophin antibody (ab15277)

Muscle stem cells (from normal mouse) were injected into the gastric muscle of an MDX mouse. Dystrophin staining: primary antibody ab15277 and secondary antibody is donkey anti-rabbit Alexa 594.

This image was kindly supplied as part of the review submitted by Jessica Tebbets.



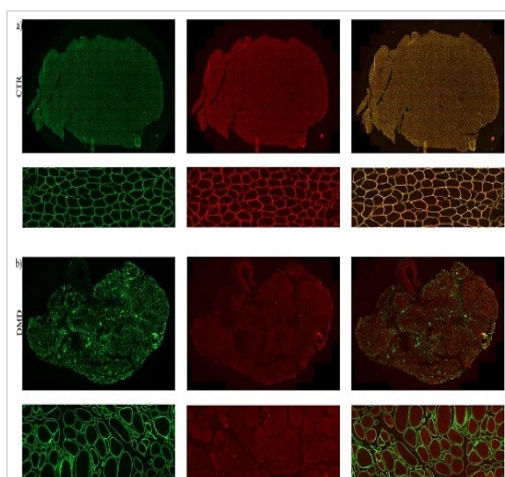
Immunohistochemistry (Frozen sections) - Anti-Dystrophin antibody (ab15277)

Zhao C. et al PLoS One. 2014 Apr 29;9(4):e96279. doi: 10.1371/journal.pone.0096279. eCollection 2014.

Immunofluorescence staining of dystrophin in W9, W987, and ESC. Myosin heavy chain (MHC) identified mouse muscle cells after differentiation. DAPI was used to stain nuclei.

Seventy-two hours before engraftment, 8 week-old *mdx/SCID* mice received 14 Gy of irradiation localized to the hind limb muscles. On the day of engraftment, SM/C-2.6-positive myogenic cells were purified by fluorescence-activated cell sorting (FACS), using a BD Aria II FACS machine and the same labeling protocol as described above for FC analysis, resuspended in 30  $\mu$ l of phosphate buffered saline (PBS), loaded into an insulin syringe (BD), and injected into the left tibialis anterior (TA) muscle of anesthetized mice.  $7.5 \times 10^5$  differentiated and sorted W987 cells were injected. Control mice were injected with PBS alone. Three weeks following engraftment, TA muscles were harvested, fixed in 0.5% paraformaldehyde for 4 hours, dehydrated in 20% sucrose overnight and frozen in optimal cutting temperature (OCT) using liquid nitrogen cooled methyl-butane. Tissue blocks imbedded in OCT were cryosectioned and processed for immunocytochemical analysis using rabbit anti-dystrophin. Secondary antibodies used were donkey anti-rabbit conjugated to Alexafluor 594 and donkey anti-rat conjugated to Alexafluor 488 (Life Technologies). Nuclei were visualized using NucBlue Fixed Cell Stain (Life Technologies).

Gene-corrected *mdx* iPSC W987, non-gene-corrected unexcised *mdx* iPSC W9 and wild-type ESC controls.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Dystrophin antibody (ab15277)

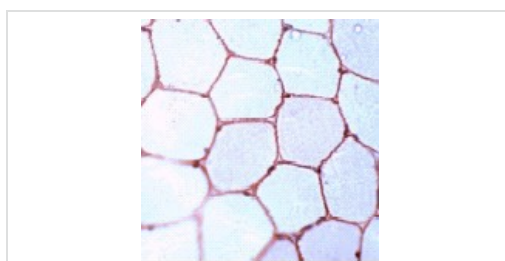
Sardone V. PLoS One. 2018 Mar 26;13(3):e0194540. doi: 10.1371/journal.pone.0194540. eCollection 2018. Reproduced under the Creative Commons license <http://creativecommons.org/licenses/by/4.0/>

Dystrophin quantification in a population of myofibres identified in entire muscle sections performing the double labelling anti-dystrophin ab15277 (red; 1/200 dilution) and anti-spectrin (green; 1/20 dilution).

All the labellings were performed at RT. Human Muscle sections were incubated with the primary antibody combination (anti-dystrophin ab15277 and anti-spectrin) for 1 hour. After three washes with PBS sections were incubated with Alexa Fluor 488 conjugated anti-mouse IgG (1:100, Thermo Fisher Scientific, Hemel Hempstead, UK) and anti-rabbit biotinylated IgG (1:200; GE Healthcare, Amersham PI, UK) for 30 minutes. PBS washes were performed and sections were incubated with Alexa Fluor 594 streptavidin conjugate (1:1000, Thermo Fisher Scientific, Hemel Hempstead, UK).

Representative images of entire muscle sections stained and acquired by the Axio Scan slide scanner and processed with Definens algorithm derived from a control (a) and from a DMD patient (b).

DMD: Duchenne Muscular Dystrophy.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Dystrophin antibody (ab15277)

Immunohistochemical staining of human skeletal muscle with ab15277

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