

Product datasheet

Anti-Dystrophin antibody ab15277

★★★★☆ 23 Abreviews 317 References 4 Images

Overview

| | |
|----------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Product name | Anti-Dystrophin antibody |
| Description | Rabbit polyclonal to Dystrophin |
| Host species | Rabbit |
| Tested applications | Suitable for: IHC-Fr, IHC-P |
| Species reactivity | Reacts with: Mouse, Human Predicted to work with: Rat, Dog, Pig  |
| Immunogen | Synthetic peptide within Human Dystrophin aa 3650 to the C-terminus (C terminal). The exact sequence is proprietary. Database link: P11532 |
| General notes | This product is FOR RESEARCH USE ONLY. For commercial use, please contact partnerships@abcam.com. 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. 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. Avoid freeze / thaw cycles. |
| Storage buffer | pH: 7.60 Preservative: 0.1% Sodium azide Constituents: PBS, 1% BSA |
| Purity | Immunogen affinity purified |
| Clonality | Polyclonal |
| Isotype | IgG |

Applications

The Abpromise guarantee Our [Abpromise guarantee](#) covers the use of ab15277 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 | ★★★★★ (6) | 1/400. Use with acetone-fixed tissues. |
| IHC-P | ★★★★☆ (14) | 1/100. |

Target

Function

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.

Tissue specificity

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.

Involvement in disease

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.

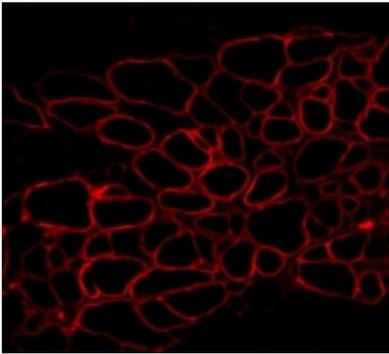
Sequence similarities

Contains 2 CH (calponin-homology) domains.
Contains 22 spectrin repeats.
Contains 1 WW domain.
Contains 1 ZZ-type zinc finger.

Cellular localization

Cell membrane > sarcolemma. Cytoplasm > cytoskeleton.

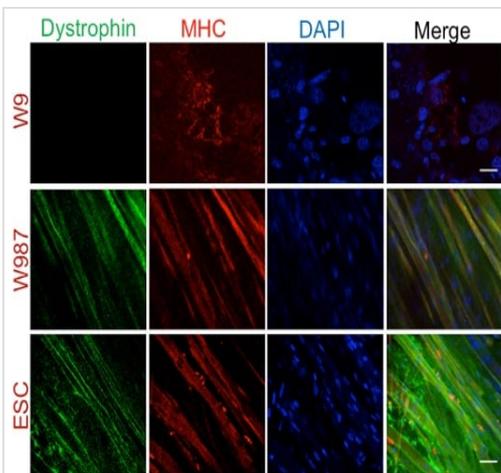
Images



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.



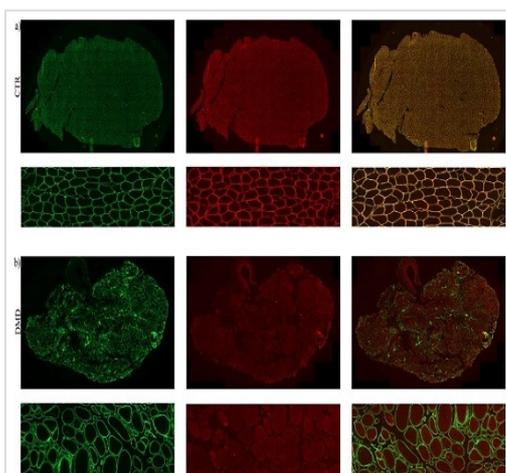
Immunohistochemistry (Frozen sections) - Anti-Dystrophin antibody (ab15277)

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 μ l of phosphate buffered saline (PBS), loaded into an insulin syringe (BD), and injected into the left tibialis anterior (TA) muscle of anesthetized mice. 7.5×10^5 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)

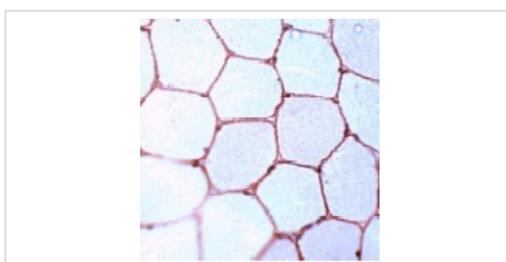
Sardone V. PLoS One. 2018 Mar 26;13(3):e0194540. doi: 10.1371/journal.pone.0194540. eCollection 2018. Reproduced under the Creative Commons license <http://creativecommons.org/licenses/by/4.0/>

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 Definers 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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