Product datasheet

Anti-Dystrophin antibody ab15277

Overview

Product name: Anti-Dystrophin antibody
Description: Rabbit polyclonal to Dystrophin
Host species: Rabbit
Tested applications: Suitable for: ICC/IF, IHC-Fr, WB, IHC-P, IHC - Wholemount
Species reactivity: Reacts with: Mouse, Rat, Dog, Human
Predicted to work with: Pig

Immunogen: Synthetic peptide within Human Dystrophin aa 3650 to the C-terminus (C terminal). The exact sequence is proprietary.
Database link: P11532
Positive control: IHC-P: Mouse triceps muscle and huma skeletal muscle ICC/IF: Human muscle fibers and dog muscle cells

Properties

Form: Liquid
Storage instructions: Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
Storage buffer: pH: 7.6
Preservative: 0.1% Sodium azide
Constituents: PBS, 1% BSA
Purity: Immunogen affinity purified
Clonality: Polyclonal
Isotype: IgG

Applications

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.

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<td>Use at an assay dependent concentration.</td>
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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
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 Tebets.

Immunofluorescence staining of dystrophin in W9, W987, and ESC. Myosin heavy chain (MHC) identified 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 µl 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.
Representative images of longitudinal sections of triceps muscle from 12-week old wild type, FRG1 and FRG1/FHL1 mice co-stained for dystrophin to outline the muscle fiber membrane and DAPI to detect nuclei. Boxed region indicates area shown in high magnification image inset. Scale bars = 100 μm.

For the determination of nuclei number per mm myofiber length, 10 μm longitudinal cryosections from triceps muscle were fixed in 4% paraformaldehyde, washed in PBS and permeabilised/blockd (10% horse serum, 1% BSA and 0.1% triton X-100 in PBS) for 1 hour at room temperature. Sections were stained overnight with rabbit anti-dystrophin (Abcam Cat# ab15277, RRID:AB_301813, 1:400), washed in PBS and incubated with goat anti-rabbit Alexa Fluor 594 and DAPI (1:100) for 1 hour at room temperature. Sections were washed in PBS and coverslips mounted using fluoromount mounting media.

Images of Dystrophin/DAPI stained longitudinal sections were captured using an Olympus BX-51 microscope using dotSlide Software.

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. 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.
Immunohistochemical staining of human skeletal muscle with ab15277

ab15277 staining dog muscle cells by ICC/IF. Cells were methanol fixed and incubated with ab15277, diluted 1/200, for 12 hours at 4°C. A FITC conjugated goat anti-rabbit antibody, diluted 1/200, was used as the secondary.

Immunofluorescence analysis of Human muscle fibers, staining Dystrophin with ab15277.

Cells were fixed with formaldehyde and blocked with 10% serum for 20 minutes at 23°C. Samples were incubated with primary antibody (1/100 in diluent) for 12 hours at 2°C. An AlexaFluor®488-conjugated goat anti-rabbit polyclonal IgG (1/200) was used as the secondary antibody.

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