abcam

Product datasheet

Anti-Dysferlin antibody [JAI-1-49-3] ab124684

Recombinant RabMAb

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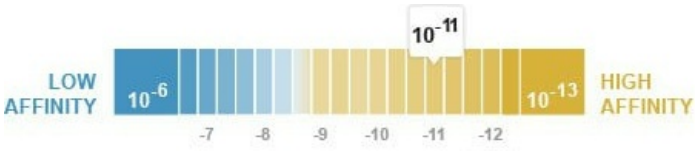
Overview

Product name	Anti-Dysferlin antibody [JAI-1-49-3]
Description	Rabbit monoclonal [JAI-1-49-3] to Dysferlin
Host species	Rabbit
Tested applications	**Suitable for:** ICC/IF, IHC-Fr, WB, IHC-P **Unsuitable for:** Flow Cyt
Species reactivity	**Reacts with:** Mouse, Human
Immunogen	Synthetic peptide within Human Dysferlin aa 100-200. The exact sequence is proprietary. Database link: O75923
Positive control	WB: Human and mouse skeletal muscle tissue lysates. IHC-P: Human skeletal muscle tissue. IHC-Fr: Human and mouse skeletal muscle tissues. ICC/IF: A673 cells.
General notes	This antibody was made in collaboration with the Jain Foundation whose goal is to hasten EVERY avenue that may lead to the cure for LGMD2B/Miyoshi. This product is a recombinant monoclonal antibody, which offers several advantages including: - High batch-to-batch consistency and reproducibility - Improved sensitivity and specificity - Long-term security of supply - Animal-free production For more information see here. Our RabMAb® technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to RabMAb® patents. **We are constantly working hard to ensure we provide our customers with best in class antibodies. As a result of this work we are pleased to now offer this antibody in purified format. We are in the process of updating our datasheets. The purified format is designated 'PUR' on our product labels. If you have any questions regarding this update, please contact our Scientific Support team.** Rat: We have preliminary internal testing data to indicate this antibody may not react with this species. Please contact us for more information.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C. Avoid freeze / thaw cycle.
Dissociation constant (K_D)	$K_D = 7.20 \times 10^{-11}$ M



Learn more about K_D

Storage buffer	pH: 7.20 Preservative: 0.01% Sodium azide Constituents: 40% Glycerol (glycerin, glycerine), 0.05% BSA, 59% PBS
Purity	Protein A purified
Clonality	Monoclonal
Clone number	JAI-1-49-3
Isotype	IgG

Applications

The Abpromise guarantee Our Abpromise guarantee covers the use of ab124684 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
ICC/IF		1/200 - 1/400.
IHC-Fr	★★★★☆ (1)	1/200.
WB	★★★★☆ (3)	1/1000. Detects a band of approximately 280 kDa (predicted molecular weight: 237 kDa). **For unpurified use at 1/1000 - 1/10000.**
IHC-P		1/500. Perform heat mediated antigen retrieval before commencing with IHC staining protocol. See IHC antigen retrieval protocols. **For unpurified use at 1/50 - 1/100.**

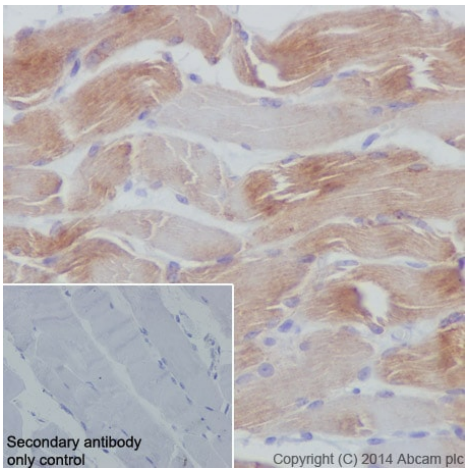
Application notes Is unsuitable for Flow Cyt.

Target

Function	Key calcium ion sensor involved in the Ca(2+)-triggered synaptic vesicle-plasma membrane fusion. Plays a role in the sarcolemma repair mechanism of both skeletal muscle and cardiomyocytes that permits rapid resealing of membranes disrupted by mechanical stress.
Tissue specificity	Expressed in skeletal muscle, myoblast, myotube and in the syncytiotrophoblast (STB) of the placenta (at protein level). Highly expressed in skeletal muscle. Also found in heart, brain, spleen, intestine, placenta and at lower levels in liver, lung, kidney and pancreas.

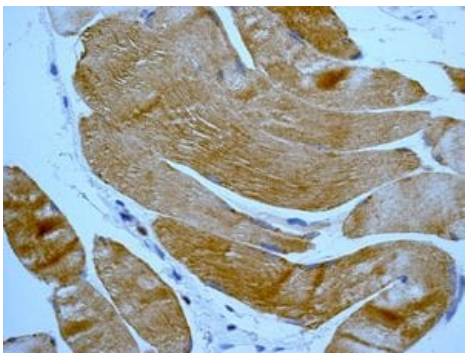
Involvement in disease	Defects in DYSF are the cause of limb-girdle muscular dystrophy type 2B (LGMD2B) [MIM:253601]. LGMD2B is an autosomal recessive degenerative myopathy characterized by weakness and atrophy starting in the proximal pelvifemoral muscles, with onset in the late teens or later, massive elevation of serum creatine kinase levels and slow progression. Scapular muscle involvement is minor and not present at onset. Upper limb girdle involvement follows some years after the onset in lower limbs. Defects in DYSF are the cause of Miyoshi muscular dystrophy type (MMD1) [MIM:254130]. MMD1 is a late-onset muscular dystrophy involving the distal lower limb musculature. It is characterized by weakness that initially affects the gastrocnemius muscle during early adulthood. Otherwise the phenotype overlaps with LGMD2B, especially in age at onset and creatine kinase elevation. Defects in DYSF are the cause of distal myopathy with anterior tibial onset (DMAT) [MIM:606768]. Onset of the disorder is between 14 and 28 years of age and the anterior tibial muscles are the first muscle group to be involved. Inheritance is autosomal recessive.
Sequence similarities	Belongs to the ferlin family. Contains 5 C2 domains.
Developmental stage	Expression in limb tissue from 5-6 weeks embryos; persists throughout development.
Domain	The C2 domain 1 associates with lipid membranes in a calcium-dependent manner.
Cellular localization	Cell membrane > sarcolemma. Cytoplasmic vesicle membrane. Colocalizes, during muscle differentiation, with BIN1 in the T-tubule system of myotubules and at the site of contact between two myotubes or a myoblast and a myotube. Wounding of myotubes led to its focal enrichment to the site of injury and to its relocalization in a Ca(2+)-dependent manner toward the plasma membrane. Colocalizes with AHNAK, AHNAK2 and PARVB at the sarcolemma of skeletal muscle. Detected on the apical plasma membrane of the syncytiotrophoblast. Reaches the plasmma membrane through a caveolin-independent mechanism. Retained by caveolin at the plasmma membrane (By similarity). Colocalizes, during muscle differentiation, with CACNA1S in the T-tubule system of myotubules (By similarity). Accumulates and colocalizes with fusion vesicles at the sarcolemma disruption sites.

Images



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Dysferlin antibody [JAI-1-49-3] (ab124684)

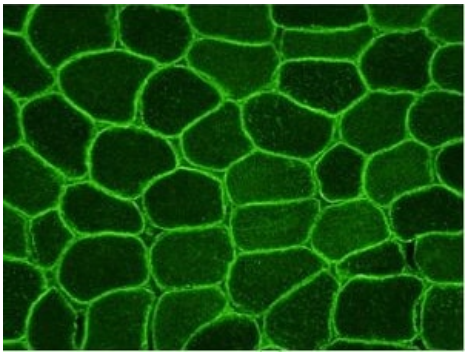
Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) analysis of human skeletal muscle tissue labelling Dysferlin with purified ab124684 at 1/500. Heat mediated antigen retrieval was performed using Tris/EDTA buffer pH 9. ab97051, a HRP-conjugated goat anti-rabbit IgG (H+L) was used as the secondary antibody (1/500). Negative control using PBS instead of primary antibody. Counterstained with hematoxylin.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Dysferlin antibody [JAI-1-49-3] (ab124684)

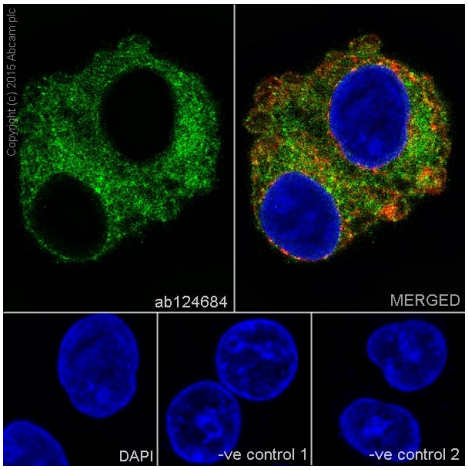
Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) analysis of human skeletal muscle tissue labelling Dysferlin with unpurified ab124684 at a dilution of 1/50.

Perform heat mediated antigen retrieval before commencing with IHC staining protocol.



Immunohistochemistry (Frozen sections) - Anti-Dysferlin antibody [JAI-1-49-3] (ab124684)

Immunohistochemistry (Frozen sections) analysis of unfixed frozen human skeletal muscle tissue (control) labelling Dysferlin with unpurified ab124684 at a dilution of 1/200.

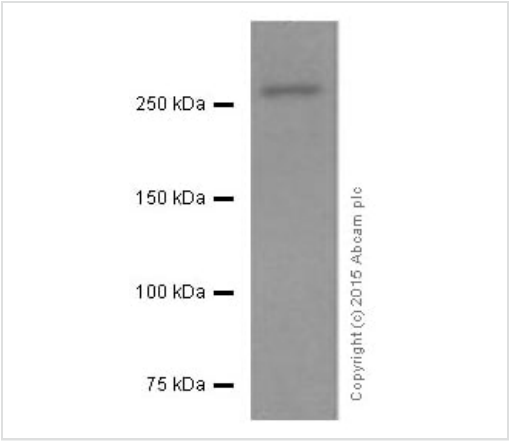


Immunocytochemistry/ Immunofluorescence - Anti-Dysferlin antibody [JAI-1-49-3] (ab124684)

Immunocytochemistry/Immunofluorescence analysis of A673 cells labelling Dysferlin with purified ab124684 at 1/300. Cells were fixed with 4% paraformaldehyde and permeabilized with 0.1% Triton X-100. ab150077, an Alexa Fluor® 488-conjugated goat anti-rabbit IgG (1/500) was used as the secondary antibody. DAPI (blue) was used as the nuclear counterstain. ab7291, a mouse anti-tubulin (1/1000) and ab150120, an Alexa Fluor® 594-conjugated goat anti-mouse IgG (1/500) were also used.

Control 1: primary antibody (1/300) and secondary antibody, ab150120, an Alexa Fluor® 594-conjugated goat anti-mouse IgG (1/500).

Control 2: ab7291 (1/1000) and secondary antibody, ab150077, an Alexa Fluor® 488-conjugated goat anti-rabbit IgG (1/500).



Western blot - Anti-Dysferlin antibody [JAI-1-49-3] (ab124684)

Anti-Dysferlin antibody [JAI-1-49-3] (ab124684) at 1/1000 dilution (purified) + Mouse muscle tissue lysate at 10 µg

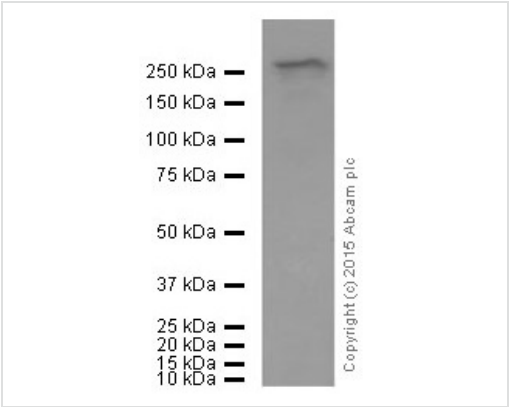
Secondary

Peroxidase-conjugated goat anti-rabbit IgG, (H+L) at 1/1000 dilution

Predicted band size: 237 kDa

Observed band size: 280 kDa

Blocking and dilution buffer: 5% NFDM/TBST.



Western blot - Anti-Dysferlin antibody [JAI-1-49-3] (ab124684)

Anti-Dysferlin antibody [JAI-1-49-3] (ab124684) at 1/1000 dilution (purified) + Human skeletal muscle tissue lysate at 20 µg

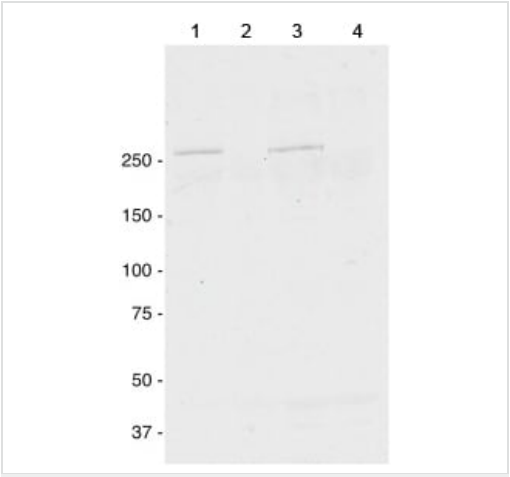
Secondary

Peroxidase-conjugated goat anti-rabbit IgG, (H+L) at 1/1000 dilution

Predicted band size: 237 kDa

Observed band size: 280 kDa

Blocking and dilution buffer: 5% NFDM/TBST.



Western blot - Anti-Dysferlin antibody [JAI-1-49-3] (ab124684)

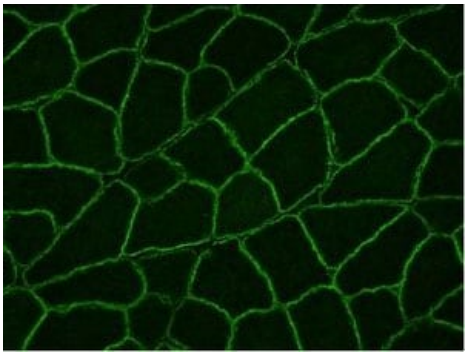
All lanes : Anti-Dysferlin antibody [JAI-1-49-3] (ab124684) at 1/1000 dilution (unpurified)

Lane 1 : Human skeletal muscle tissue lysate (control)
Lane 2 : Human skeletal muscle tissue lysate (LGMD2B)
Lane 3 : Mouse skeletal muscle tissue lysate (wild-type mice)
Lane 4 : Mouse skeletal muscle tissue lysate (Dysf-/- transgenic mouse)

Lysates/proteins at 10 µg per lane.

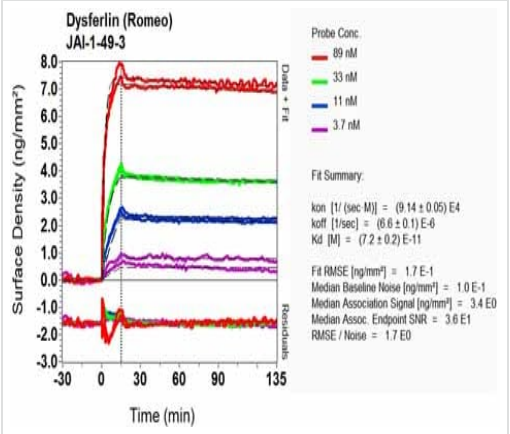
Secondary
All lanes : HRP-conjugated goat anti-rabbit IgG at 1/2000 dilution

Predicted band size: 237 kDa
Observed band size: 280 kDa



Immunohistochemistry (Frozen sections) - Anti-Dysferlin antibody [JAI-1-49-3] (ab124684)

Immunohistochemistry (Frozen sections) analysis of unfixed frozen mouse skeletal muscle tissue (wild type) labelling Dysferlin with unpurified ab124684 at a dilution of 1/200.

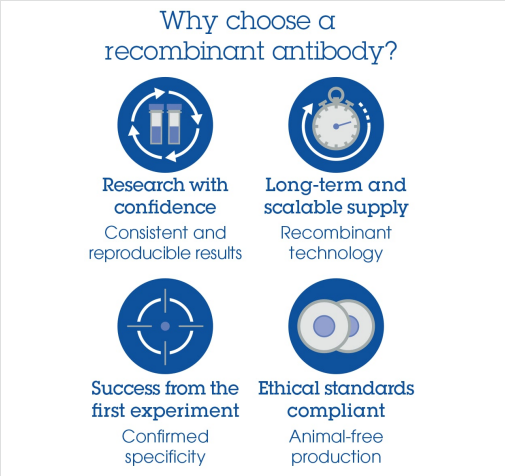


OI-RD Scanning - Anti-Dysferlin antibody [JAI-1-49-3] (ab124684)

Equilibrium disassociation constant (K_D)

Learn more about K_D

Click here to learn more about K_D



Anti-Dysferlin antibody [JAI-1-49-3] (ab124684)

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