


Anti-Dysferlin antibody ab15108

★★★★☆ [4 Abreviews](#) [5 References](#) [2 Images](#)

Overview

Product name	Anti-Dysferlin antibody
Description	Rabbit polyclonal to Dysferlin
Host species	Rabbit
Tested applications	Suitable for: WB, IHC-P
Species reactivity	Reacts with: Human Predicted to work with: Mouse, Dog 
Immunogen	Synthetic peptide within Human Dysferlin aa 1950-2050 (C terminal). The exact sequence is proprietary. Database link: O75923
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 ab15108 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
WB	★★★★★ (2)	Use a concentration of 1 µg/ml. Predicted molecular weight: 231 kDa.
IHC-P		1/50.

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.

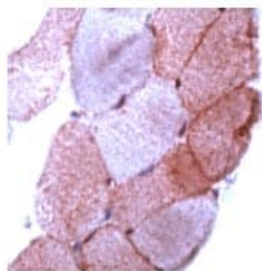
Domain

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 AHNK, AHNK2 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 (ab15108)

ab15108 staining Dysferlin in human skeletal muscle by Immunohistochemistry (FFPE-sections).



Western blot - Anti-Dysferlin antibody (ab15108)

Anti-Dysferlin antibody (ab15108) at 1 µg/ml + Human skeletal muscle tissue lysate - total protein (**ab29330**) at 10 µg

Secondary

Goat polyclonal to Rabbit IgG - H&L - Pre-Adsorbed (HRP) at 1/3000 dilution

Predicted band size: 231 kDa

Observed band size: 231 kDa

Additional bands at: 240 kDa (possible post-translational modification)

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