abcam

Product datasheet

Anti-Dysferlin antibody ab85802

2 Images

Overview

Species reactivity

Product name Anti-Dysferlin antibody

Description Rabbit polyclonal to Dysferlin

Host species Rabbit

Tested applications Suitable for: IHC-P, WB

Reacts with: Human

Predicted to work with: Orangutan

Immunogen Synthetic peptide corresponding to Human Dysferlin aa 2050 to the C-terminus (C terminal)

conjugated to keyhole limpet haemocyanin.

(Peptide available as ab97417)

Positive controlThis antibody gave a positive signal in Human skeletal muscle tissue lysate.

General notesThe 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. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C or -

80°C. Avoid freeze / thaw cycle.

Storage buffer pH: 7.40

Preservative: 0.02% Sodium azide

Constituent: PBS

Batches of this product that have a concentration < 1mg/ml may have BSA added as a stabilising

agent. If you would like information about the formulation of a specific lot, please contact our

scientific support team who will be happy to help.

Purity Immunogen affinity purified

1

Clonality Polyclonal

Isotype IgG

Applications

The Abpromise guarantee

Our Abpromise guarantee covers the use of ab85802 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-P | | Use a concentration of 5 µg/ml. Perform heat mediated antigen retrieval before commencing with IHC staining protocol. |
| WB | | Use a concentration of 1 µg/ml. Detects a band of approximately 248 kDa (predicted molecular weight: 237 kDa). |

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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

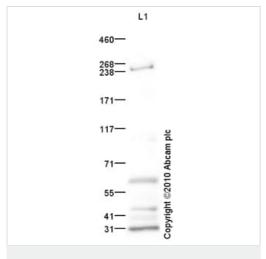
The C2 domain 1 associates with lipid membranes in a calcium-dependent manner.

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



Western blot - Anti-Dysferlin antibody (ab85802)

Anti-Dysferlin antibody (ab85802) at 1 μ g/ml + Human skeletal muscle tissue lysate - total protein (**ab29330**) at 10 μ g

Secondary

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

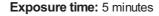
Developed using the ECL technique.

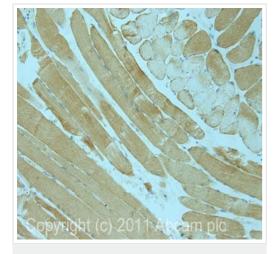
Performed under reducing conditions.

Predicted band size: 237 kDa Observed band size: 248 kDa

Additional bands at: 31 kDa, 44 kDa, 61 kDa. We are unsure as

to the identity of these extra bands.





Immunohistochemistry (Formalin/PFA-fixed paraffinembedded sections) - Anti-Dysferlin antibody (ab85802)

IHC image of Dysferlin staining in human skeletal muscle formalin fixed paraffin embedded tissue section, performed on a Leica Bond TM system using the standard protocol F. The section was pretreated using heat mediated antigen retrieval with sodium citrate buffer (pH6, epitope retrieval solution 1) for 20 mins. The section was then incubated with ab85802, 5µg/ml, for 15 mins at room temperature and detected using an HRP conjugated compact polymer system. DAB was used as the chromogen. The section was then counterstained with haematoxylin and mounted with DPX.

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