abcam

Product datasheet

Anti-Dysferlin antibody ab85802

2 图像

概述

免疫原

产品名称 Anti-Dysferlin抗体

描述 兔多克隆抗体to Dysferlin

宿主 Rabbit

种属反应性 与反应: Human

预测可用于: Orangutan

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

conjugated to keyhole limpet haemocyanin.

(Peptide available as ab97417)

阳性对照 This antibody gave a positive signal in Human skeletal muscle tissue lysate.

常规说明

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

性能

形式 Liquid

存放说明 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.

存储溶液 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.

纯**度** Immunogen affinity purified

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克隆 多克隆

同种型 lgG

应用

The Abpromise guarantee Abpromise™承诺保证使用ab85802于以下的经测试应用

"应用说明"部分下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
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).

靶标

功能 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.

组织特异性 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.

疾病相关 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.

序列相似性 Belongs to the ferlin family.

Contains 5 C2 domains.

发展阶段 Expression in limb tissue from 5-6 weeks embryos; persists throughout development.

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

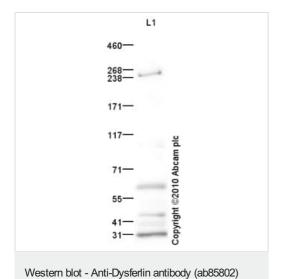
细胞定位

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.

图片



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

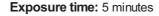
Developed using the ECL technique.

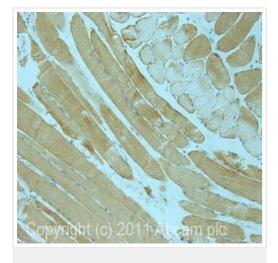
Performed under reducing conditions.

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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