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Product datasheet

Anti-Dysferlin antibody ab15108

4 Abreviews 5 References 2 图像

概述

产品名称	Anti-Dysferlin抗体
描述	**兔多克隆抗体**to Dysferlin
宿主	Rabbit
经测试应用	**适用于:** WB, IHC-P
种属反应性	**与反应:** Human **预测可用于:** Mouse, Dog
免疫原	Synthetic peptide within Human Dysferlin aa 1950-2050 (C terminal). The exact sequence is proprietary. Database link: **O75923**
常规说明	**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

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
存储溶液	pH: 7.60 Preservative: 0.1% Sodium azide Constituents: PBS, 1% BSA
纯度	Immunogen affinity purified
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab15108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
WB	★★☆☆☆ (2)	Use a concentration of 1 µg/ml. Predicted molecular weight: 231 kDa.
IHC-P		1/50.

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.

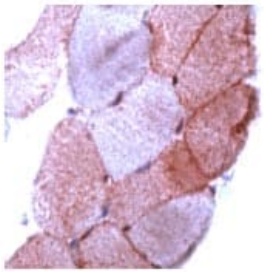
结构域

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

细胞定位

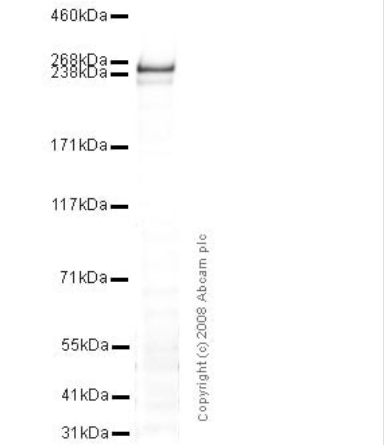
Cell membrane > sarcolemma. Cytoplasmic vesicle membrane. Colocalizes, during muscle differentiation, with BIN1 in the T-tubule system of myotubules and at the site of contact between two myotubes or a myoblast and a myotube. Wounding of myotubes led to its focal enrichment to the site of injury and 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



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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