


Product datasheet

Anti-Dysferlin antibody ab85802

2 图像

概述

产品名称	Anti-Dysferlin抗体
描述	兔多克隆抗体to Dysferlin
经测试应用	适用于: IHC-P, WB
种属反应性	与反应: Human 预测可用于: Orangutan 
免疫原	Synthetic peptide conjugated to KLH derived from within residues 2050 to the C-terminus of Human Dysferlin.参阅Abcam的专有抗源政策(Peptide available as ab97417 .)
阳性对照	This antibody gave a positive signal in Human skeletal muscle tissue lysate.

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	Preservative: 0.02% Sodium Azide Constituents: 1% BSA, PBS, pH 7.4
纯度	Immunogen affinity purified
克隆	多克隆
同种型	IgG

应用

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.

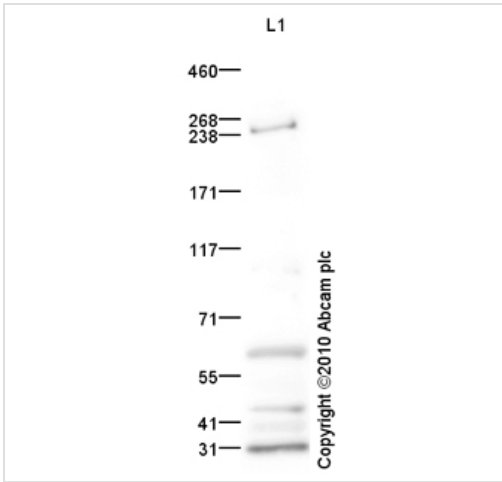
应用	Ab评论	说明
IHC-P		Use a concentration of 5 µg/ml. Perform heat mediated antigen retrieval before commencing with IHC staining protocol.

应用	Ab评论	说明
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	<p>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.</p> <p>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.</p> <p>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.</p>
序列相似性	<p>Belongs to the ferlin family.</p> <p>Contains 5 C2 domains.</p>
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 plasma membrane through a caveolin-independent mechanism. Retained by caveolin at the plasma 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



Western blot - Dysferlin antibody (ab85802)

Anti-Dysferlin antibody (ab85802) 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
Developed using the ECL technique

Performed under reducing conditions.

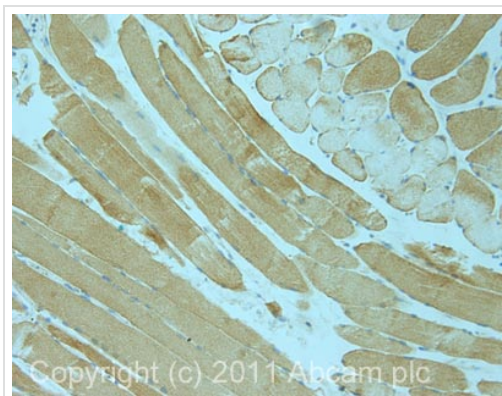
Predicted band size : 237 kDa

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.

Exposure time : 5 minutes



Immunohistochemistry (Formalin/PFA-fixed paraffin-
embedded sections) - Dysferlin antibody (ab85802)

IHC image of Dysferlin staining in human
skeletal muscle formalin fixed paraffin
embedded tissue section, performed on a
Leica Bond™ system using the standard
protocol F. The section was pre-treated 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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