abcam

Product datasheet

Anti-FHL1 antibody ab95199

2 图像

概述

产**品名称** Anti-FHL1抗体

描述 兔多克隆抗体to FHL1

宿主 Rabbit

种属反应性 与反应: Human

预测可用于: Sheep, Rabbit, Cow, Dog, Pig, Macaque monkey, Gorilla, Orangutan

A

免疫原 Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.

阳性对照 Recombinant Human FHL1 protein (<u>ab114384</u>) can be used as a positive control in WB. 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

scientific support team who will be happy to help.

纯**度** Immunogen affinity purified

克隆 多克隆

1

同种型 lgG

应用

The Abpromise guarantee

Abpromise™承诺保证使用ab95199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		Use a concentration of 1 µg/ml. Detects a band of approximately 36 kDa (predicted molecular weight: 36 kDa).
IHC-P		Use a concentration of 1 µg/ml. Perform heat mediated antigen retrieval before commencing with IHC staining protocol.

靶标

功能

组织特异性

疾病相关

May have an involvement in muscle development or hypertrophy.

Isoform 1 is highly expressed in skeletal muscle and to a lesser extent in heart, placenta, ovary, prostate, testis, small intestine, colon and spleen. Expression is barely detectable in brain, lung, liver, kidney, pancreas, thymus and peripheral blood leukocytes. Isoform 2 is expressed in brain, skeletal muscle and to a lesser extent in heart, colon, prostate and small intestine. Isoform 3 is expressed in testis, heart and skeletal muscle.

Defects in FHL1 are the cause of X-linked dominant scapuloperoneal myopathy (SPM) [MIM:300695]. Scapuloperoneal syndrome (SPS) was initially described more than 120 years ago by Jules Broussard as 'une forme hereditaire d'atrophie musculaire progressive' beginning in the lower legs and affecting the shoulder region earlier and more severely than distal arm. The etiology of this condition remains unclear.

Defects in FHL1 are the cause of X-linked myopathy with postural muscle atrophy (XMPMA) [MIM:300696]. Myopathies are inherited muscle disorders characterized by weakness and atrophy of voluntary skeletal muscle, and several types of myopathy also show involvement of cardiac muscle. XMPMA is a distinct form of adult-onset X-linked recessive myopathy with several features in common with other myopathies, but the presentation of a pseudoathletic phenotype, scapuloperoneal weakness, and bent spine is unique and might render the clinical phenotype distinguishable from other myopathies.

Defects in FHL1 are the cause of X-linked severe early-onset reducing body myopathy (RBM) [MIM:300717]. RBM is a rare muscle disorder causing progressive muscular weakness and characteristic intracytoplasmic inclusions in myofibers. Clinical presentations of RBM have ranged from early onset fatal to childhood onset to adult onset cases.

Defects in FHL1 are the cause of X-linked childhood-onset reducing body myopathy (CO-RBM) [MIM:300718]. This disorder is allelic to severe early-onset reducing body myopathy (RBM) [MIM:300717].

Contains 3 LIM zinc-binding domains.

Elevated levels during postnatal muscle growth.

Cytoplasm; Cytoplasm. Nucleus and Nucleus. Cytoplasm > cytosol. Predominantly nuclear in

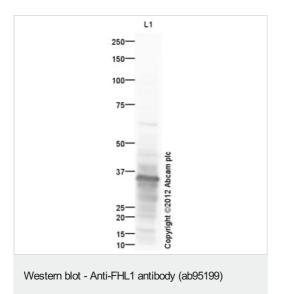
myoblasts but is cytosolic in differentiated myotubes.

2

序列相似性

发展阶段

细胞定位



Anti-FHL1 antibody (ab95199) at 1 μg/ml + Human skeletal muscle tissue lysate - total protein (**ab29330**) at 10 μg

Secondary

Goat Anti-Rabbit lgG H&L (HRP) preadsorbed (ab97080) at 1/5000 dilution

Developed using the ECL technique.

Performed under reducing conditions.

Predicted band size: 36 kDa Observed band size: 36 kDa

Exposure time: 30 seconds



Immunohistochemistry (Formalin/PFA-fixed paraffinembedded sections) - Anti-FHL1 antibody (ab95199)

IHC image of FHL1 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 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 ab95199, 1µg/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.

For other IHC staining systems (automated and non-automated) customers should optimize variable parameters such as antigen retrieval conditions, primary antibody concentration and antibody incubation times.

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