

Product datasheet

Anti-FHL1 antibody ab83571

画像数 1

製品の概要

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| 製品名 | Anti-FHL1 antibody |
| 製品の詳細 | Rabbit polyclonal to FHL1 |
| アプリケーション | 適用あり: WB, ELISA |
| 種交差性 | 交差種: Human 交差が予測される動物種: Mouse, Rat, Sheep, Rabbit, Horse, Guinea pig, Cow, Cat, Dog, Pig |
| 免疫原 | Synthetic peptide from within the N terminal amino acids 51-100: SKEVHYKNRF WHDTCFRCAK CLHPLANETF VAKDNKILCN KCTTREDSPK, of Human FHL1 (NP_001440) Run BLAST with ExPASy Run BLAST with NCBI |
| ポジティブ・コントロール | Human fetal brain lysate |

製品の特性

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| 製品の状態 | Liquid |
| 保存方法 | Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles. |
| バッファー | Preservative: None Constituents: 2% Sucrose, PBS |
| 精製度 | Immunogen affinity purified |
| ポリ/モノ | ポリクローナル |
| アイソタイプ | IgG |

アプリケーション

Our [Abpromise guarantee](#) covers the use of **ab83571** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

| アプリ ケーショ ン | Abreviews | 特記事項 |
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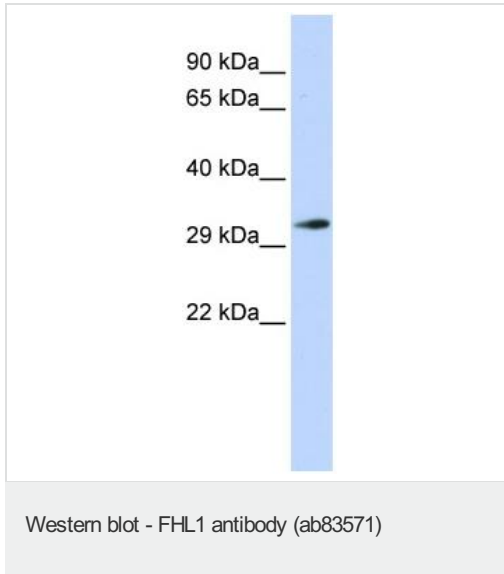
WB Use a concentration of 1 µg/ml. Detects a band of approximately 32 kDa (predicted molecular weight: 36 kDa). Good results were obtained when blocked with 5% non-fat dry milk in 0.05% PBS-T.

ELISA Use at an assay dependent concentration.
ELISA titre using peptide based assay: 1:62500.

ターゲット情報

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| 機能 | May have an involvement in muscle development or hypertrophy. |
| 組織特異性 | 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. |
| 関連疾患 | <p>Defects in FHL1 are the cause of X-linked dominant scapulooperoneal myopathy (SPM) [MIM:300695]. Scapulooperoneal 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.</p> <p>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, scapulooperoneal weakness, and bent spine is unique and might render the clinical phenotype distinguishable from other myopathies.</p> <p>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.</p> <p>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].</p> |
| 配列類似性 | Contains 3 LIM zinc-binding domains. |
| 発生段階 | Elevated levels during postnatal muscle growth. |
| 細胞内局在 | Cytoplasm; Cytoplasm. Nucleus and Nucleus. Cytoplasm > cytosol. Predominantly nuclear in myoblasts but is cytosolic in differentiated myotubes. |

画像



Anti-FHL1 antibody (ab83571) at 1 µg/ml (in 5% skim milk/ PBS) + human fetal brain lysate at 10 µg

Secondary

HRP conjugated anti-Rabbit IgG at 1/50000 dilution

Predicted band size : 36 kDa

Observed band size : 32 kDa

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