

## Product datasheet

# Anti-FHL1 antibody ab83571

### 画像数 1

#### 製品の概要

製品名	Anti-FHL1 antibody
製品の詳細	Rabbit polyclonal to FHL1
由来種	Rabbit
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) <a href="#">Run BLAST with ExPASy</a> <a href="#">Run BLAST with NCBI</a>
ポジティブ・コントロール	Human fetal brain lysate

#### 製品の特性

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: 0.09% Sodium azide 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.

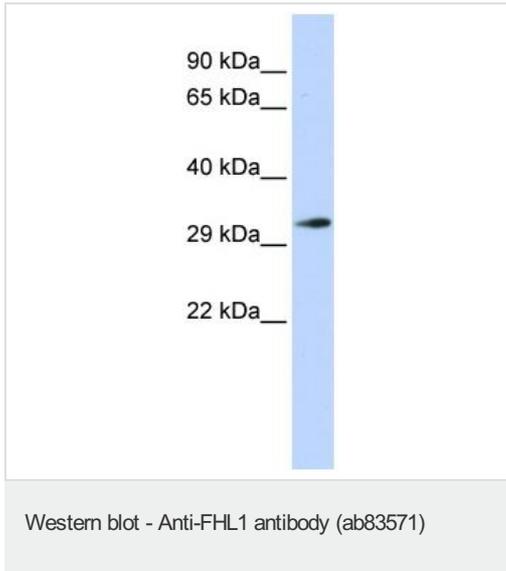
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

アプリ ケーショ ン	Abreviews	特記事項
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

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 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.</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, scapuloperoneal 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

[why is the actual band size different from the predicted?](#)

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