

## Product datasheet

# Anti-FHL1 antibody ab49241

[4 References](#) [3 Images](#)

### Overview

<b>Product name</b>	Anti-FHL1 antibody
<b>Description</b>	Rabbit polyclonal to FHL1
<b>Host species</b>	Rabbit
<b>Tested applications</b>	<b>Suitable for:</b> WB, IHC-P, ICC/IF
<b>Species reactivity</b>	<b>Reacts with:</b> Human <b>Predicted to work with:</b> Mouse, Rat, Sheep, Rabbit, Horse, Guinea pig, Cow, Cat, Dog, Pig
<b>Immunogen</b>	A region within synthetic peptide: VAKKCAGCKN PITGFGKGSS VVAYEGQSWH DYCFHCKKCS VNLANKRFVF, corresponding to C terminal amino acids 217-266 of Human FHL1 <a href="#">Run BLAST with ExPASy</a> <a href="#">Run BLAST with NCBI</a>
<b>Positive control</b>	WB: Fetal muscle lysate. IHC-P: Human kidney epithelial cells.

### Properties

<b>Form</b>	Liquid
<b>Storage instructions</b>	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
<b>Storage buffer</b>	Preservative: None Constituents: 2% Sucrose, PBS
<b>Purity</b>	Protein A purified
<b>Clonality</b>	Polyclonal
<b>Isotype</b>	IgG

### Applications

Our [Abpromise guarantee](#) covers the use of **ab49241** in the following tested applications.

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

Application	Abreviews	Notes
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Application	Abreviews	Notes
WB		Use a concentration of 0.1 µg/ml. Detects a band of approximately 30 kDa (predicted molecular weight: 32 kDa). Good results were obtained when blocked with 5% non-fat dry milk in 0.05% PBS-T.
IHC-P		Use a concentration of 4 - 8 µg/ml.
ICC/IF		Use a concentration of 1 µg/ml.

## Target

### Function

May have an involvement in muscle development or hypertrophy.

### Tissue specificity

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.

### Involvement in disease

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].

### Sequence similarities

Contains 3 LIM zinc-binding domains.

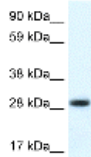
### Developmental stage

Elevated levels during postnatal muscle growth.

### Cellular localization

Cytoplasm; Cytoplasm. Nucleus and Nucleus. Cytoplasm > cytosol. Predominantly nuclear in myoblasts but is cytosolic in differentiated myotubes.

## Images



Western blot - Anti-FHL1 antibody (ab49241)

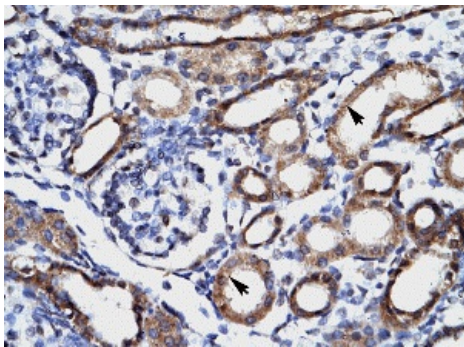
Anti-FHL1 antibody (ab49241) at 0.1  $\mu\text{g/ml}$  +  
Fetal muscle lysate at 10  $\mu\text{g}$

#### Secondary

HRP conjugated anti-Rabbit IgG at 1/50000  
dilution

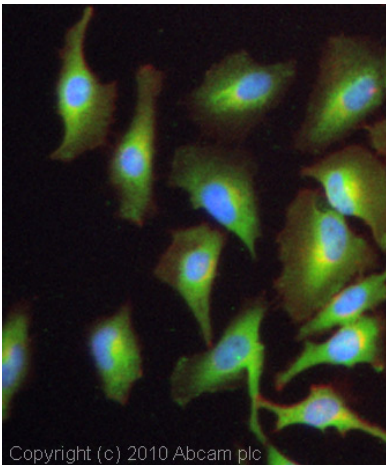
**Predicted band size:** 32 kDa

**Observed band size:** 30 kDa



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-FHL1 antibody (ab49241)

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) analysis of human kidney tissue labelling FHL1 with ab49241 at 4-8 $\mu\text{g/ml}$ . Arrows indicate positively labelled epithelial renal tubule cells. Magnification: 400X.



Immunocytochemistry/ Immunofluorescence - Anti-FHL1 antibody (ab49241)

ICC/IF image of ab49241 stained HeLa cells. The cells were 4% formaldehyde fixed (10 min) and then incubated in 1%BSA / 10% normal goat serum / 0.3M glycine in 0.1% PBS-Tween for 1h to permeabilise the cells and block non-specific protein-protein interactions. The cells were then incubated with the antibody (ab49241, 1  $\mu\text{g/ml}$ ) overnight at +4°C. The secondary antibody (green) was Alexa Fluor® 488 goat anti-rabbit IgG (H+L) used at a 1/1000 dilution for 1h. Alexa Fluor® 594 WGA was used to label plasma membranes (red) at a 1/200 dilution for 1h. DAPI was used to stain the cell nuclei (blue) at a concentration of 1.43 $\mu\text{M}$ .

**Please note:** All products are "FOR RESEARCH USE ONLY AND ARE NOT INTENDED FOR DIAGNOSTIC OR THERAPEUTIC USE"

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