

Product datasheet

Anti-FHL1 antibody ab229485

5 Images

Overview

Product name	Anti-FHL1 antibody
Description	Rabbit polyclonal to FHL1
Host species	Rabbit
Tested applications	Suitable for: WB, IHC-P
Species reactivity	Reacts with: Mouse, Human Predicted to work with: Rat, Sheep, Cow, Pig 
Immunogen	Recombinant fragment within Human FHL1 (internal sequence). The exact sequence is proprietary. Database link: Q13642
Positive control	IHC-P: Mouse hind brain, muscle and heart tissues; U-87 xenograft tissue. WB: Jurkat whole cell lysate.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
Storage buffer	pH: 7.00 Preservative: 0.01% Thimerosal (merthiolate) Constituents: PBS, 1% BSA, 20% Glycerol
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

Our [Abpromise guarantee](#) covers the use of **ab229485** 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
WB		1/500 - 1/3000. Predicted molecular weight: 36 kDa.
IHC-P		1/100 - 1/1000.

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.

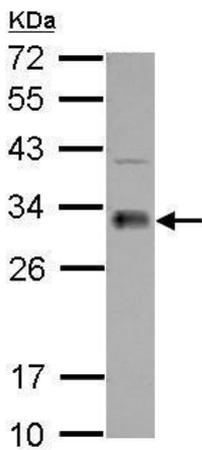
Developmental stage

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

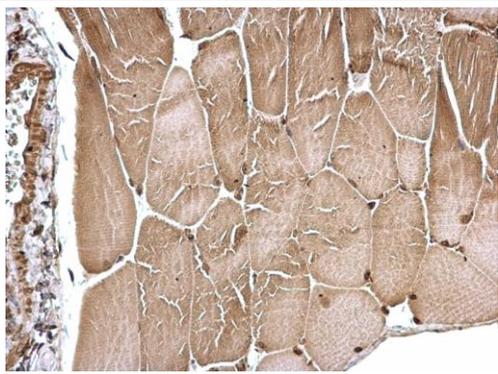


Western blot - Anti-FHL1 antibody (ab229485)

Anti-FHL1 antibody (ab229485) at 1/2000 dilution + Jurkat (human T cell leukemia cell line from peripheral blood) whole cell lysate at 30 μ g

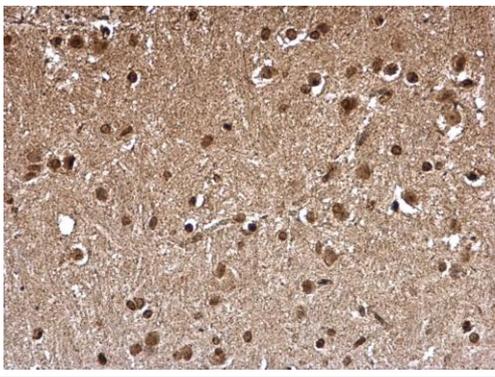
Predicted band size: 36 kDa

12% SDS-PAGE gel.



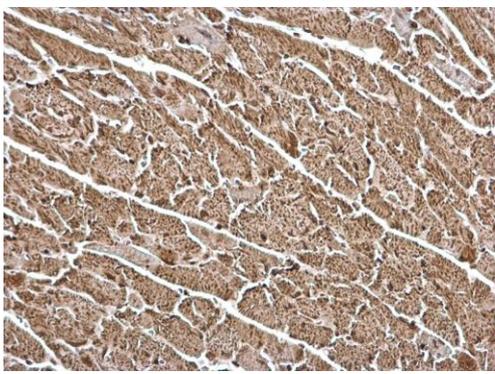
Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-FHL1 antibody (ab229485)

Paraffin-embedded mouse muscle tissue stained for FHL1 using ab229485 at 1/500 dilution in immunohistochemical analysis.



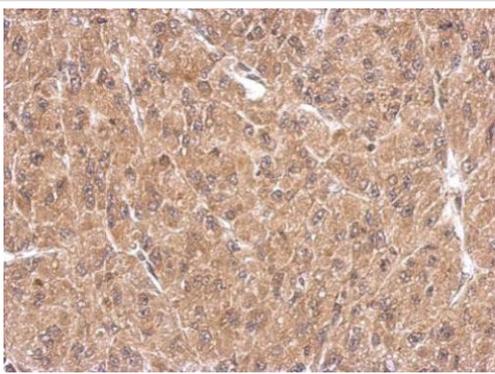
Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-FHL1 antibody (ab229485)

Paraffin-embedded mouse hind brain tissue stained for FHL1 using ab229485 at 1/500 dilution in immunohistochemical analysis.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-FHL1 antibody (ab229485)

Paraffin-embedded mouse heart tissue stained for FHL1 using ab229485 at 1/500 dilution in immunohistochemical analysis.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-FHL1 antibody (ab229485)

Paraffin-embedded U-87 (human) xenograft tissue stained for FHL1 using ab229485 at 1/500 dilution in immunohistochemical analysis.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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