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FHL1 Polyclonal Antibody
Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP58248

Specification

FHL1 Polyclonal Antibody - Product Information

Application	**WB, IHC-P, IHC-F, IF, ICC, E**
Primary Accession	Q13642
Reactivity	**Rat, Pig, Dog, Bovine**
Host	**Rabbit**
Clonality	**Polyclonal**
Calculated MW	**35 KDa**
Physical State	**Liquid**
Immunogen	**KLH conjugated synthetic peptide derived from human FHL1**
Epitope Specificity	**151-250/323**
Isotype	**IgG**
Purity affinity purified by Protein A	
Buffer	**0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.**
SUBCELLULAR LOCATION	**Isoform 1: Cytoplasm. Isoform 3: Cytoplasm. Nucleus. Isoform 2: Nucleus. Cytoplasm, cytosol. Note=Predominantly nuclear in myoblasts but is cytosolic in differentiated myotubes.**
SIMILARITY	**Contains 3 LIM zinc-binding domains.**
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].

Important Note

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Background Descriptions

This gene encodes a member of the four-and-a-half-LIM-only protein family. Family members contain two highly conserved, tandemly arranged, zinc finger domains with four highly conserved cysteines binding a zinc atom in each zinc finger. Expression of these family members occurs in a cell- and tissue-specific mode and these proteins are involved in many cellular processes. Mutations in this gene have been found in patients with Emery-Dreifuss muscular dystrophy. Multiple alternately spliced transcript variants which encode different protein isoforms have been described.[provided by RefSeq, Nov 2009]

FHL1 Polyclonal Antibody - Additional Information

Gene ID 2273

Other Names

Four and a half LIM domains protein 1, FHL-1, Skeletal muscle LIM-protein 1, SLIM, SLIM-1, FHL1, SLIM1

Target/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.

Dilution

WB~~1:1000<br \>IHC-P~~N/A<br \>IHC-F~~N/A<br \>IF~~1:50~200<br \>ICC~~N/A<br \>E~~N/A

Format

0.01M TBS(pH7.4), 0.09% (W/V) sodium azide and 50% Glyce

Storage

Store at -20 ℃ for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ℃.

FHL1 Polyclonal Antibody - Protein Information

Name FHL1

Synonyms SLIM1

Function
May have an involvement in muscle development or hypertrophy.

Cellular Location
[Isoform 1]: Cytoplasm. [Isoform 2]: Nucleus. Cytoplasm, cytosol. Note=Predominantly nuclear in myoblasts but is cytosolic in differentiated myotubes

Tissue Location
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

FHL1 Polyclonal Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

FHL1 Polyclonal Antibody - Images

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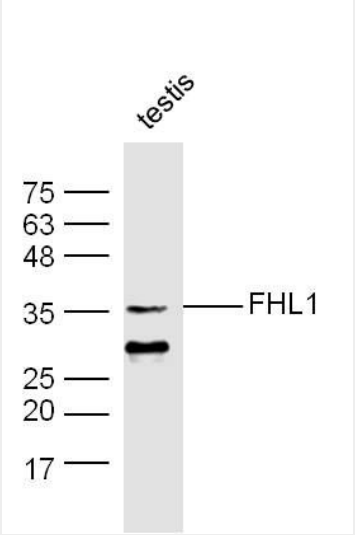

Sample:
testis (Mouse) Lysate at 40 ug
Primary: Anti-FHL1 (bs-4827R) at 1/300 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 35 kD
Observed band size: 35 kD