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Rabbit Anti-FHL1 antibody

SL4827R

Product Name:	FHL1
Chinese Name:	骨骼肌蛋白FHL1抗体
Alias:	bA535K18.1; FHL 1; FHL 1B; FHL-1; FHL1; FHL1 protein; FHL1_HUMAN; FHL1A; FHL1B; FLH1A; Four and a half LIM domains 1; Four and a half LIM domains protein 1; Four and a half Lin11 Isl 1 and Mec 3 domains 1; KYO T; LIM protein SLIMMER; MGC111107; RAM14-1; RBP associated molecule 14-1; Skeletal muscle LIM protein 1; Skeletal muscle LIM-protein 1; SLIM 1; SLIM; SLIM-1; SLIM1; SLIMMER; XMPMA.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	35kDa
Cellular localization:	The nucleuscytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human FHL1:151-250/323
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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 °C.
PubMed:	PubMed
Product Detail:	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]

Function:
May have an involvement in muscle development or hypertrophy.

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.

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.

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].

Similarity:
Contains 3 LIM zinc-binding domains.

SWISS:
Q13642

Gene ID:
2273

Database links:

Entrez Gene: 509056Cow

Entrez Gene: 2273Human

Entrez Gene: 14199Mouse

Entrez Gene: 25177Rat

Omim: 300163Human

SwissProt: Q13642Human

SwissProt: P97447Mouse

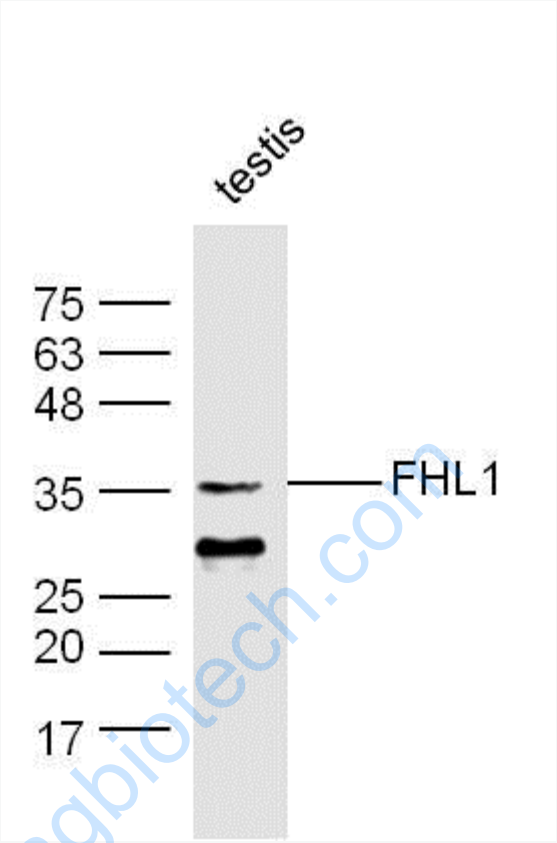
SwissProt: Q9WUH4Rat

Unigene: 435369Human

Unigene: 3126Mouse

Unigene: 54261Rat

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

Picture:	 Sample: testis (Mouse) Lysate at 40 ug Primary: Anti-FHL1 (SL4827R) at 1/300 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 35 kD Observed band size: 35 kD