

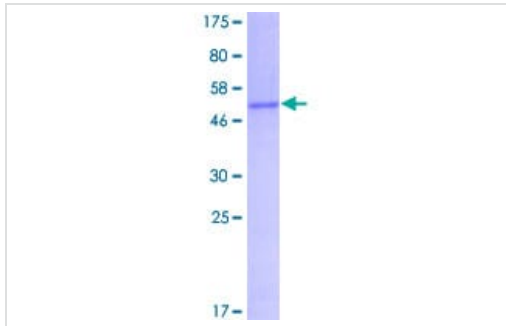
Recombinant Human FHL1 protein ab114384

1 图像

描述	
产品名称	重组人FHL1蛋白
表达系统	Wheat germ
Accession	<u>Q13642-1</u>
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human
序列	MAEKFDCHYCRDPLQGKKYVQKDGHHCCLKCFDKFCANTCVE CRKPIGAD SKEVHYKNRFWHDTCFRCAKCLHPLANETFVAKDNKILCNKC TTREDSPK CKGCFKAIVAGDQNVEYKGTVWHKDCFTCSNCKQVIGTGSFF PKGEDFYC VTCHETKFAKHCVKCNKAITSGGITYQDQPWHADCFVCVTCS KKLAGQRF TAVEDQYYCVDYKYNFVAKKCAGCKNPITGFGKGSSVVAYEG QSWHDYCF HCKKCSVNLANKRFVFHQEQVYCPDCAKKL
预测分子量	57 kDa including tags
氨基酸	1 to 280
技术指标	
Our <u>Abpromise guarantee</u> covers the use of ab114384 in the following tested applications.	
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.	
应用	ELISA Western blot SDS-PAGE
形式	Liquid
制备和贮存	

稳定性和存储	<p>Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.</p> <p>pH: 8.00</p> <p>Constituents: 0.3% Glutathione, 0.79% Tris HCl</p>
常规信息	
功能	<p>May have an involvement in muscle development or hypertrophy.</p>
组织特异性	<p>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.</p>
疾病相关	<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>
序列相似性	<p>Contains 3 LIM zinc-binding domains.</p>
发展阶段	<p>Elevated levels during postnatal muscle growth.</p>
细胞定位	<p>Cytoplasm; Cytoplasm. Nucleus and Nucleus. Cytoplasm > cytosol. Predominantly nuclear in myoblasts but is cytosolic in differentiated myotubes.</p>

图片



SDS-PAGE - Recombinant Human FHL1 protein
(ab114384)

ab114384 analysed on a 12.5% SDS-PAGE Stained with
Coomassie Blue.

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

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <https://www.abcam.cn/abpromise> or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors