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Rabbit Anti-FLNC antibody

SL13182R

Product Name:	FLNC
Chinese Name:	细丝蛋白2抗体
Alias:	ABP 280; ABP280; ABP L; ABPL; Actin binding like protein; Actin binding protein 280; Filamin 2; Filamin2; Filamin-2; Filamin C; Filamin C gamma; FLJ10186; FLN 2; FLN2; FLNC; Gamma actin binding protein; Gamma filamin; Protein FLNc; FLNC_HUMAN; Filamin-C; FLN-C; ABP-280-like protein; ABP-L; Actin-binding-like protein.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Sheep,Chimpanzee
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:	300kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Filamin 2:251-350/2725
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:	Filamins are Actin-binding proteins which contain an N-terminal Actin-binding domain, a membrane glycoprotein domain and a C-terminal self-association domain. Filamins help reshape the cytoskeleton by forming flexible cross-links between two Actin

	filaments, which maintain membrane integrity during force application. Filamins also participate in signal transduction pathways associated with cell motility, adhesion, differentiation and survival, and force transduction. The filamin family is comprised of Filamin 1, Filamin 2 and Filamin 3. Filamin 2, also designated Filamin C, is a skeletal- and cardiac-muscle specific form of Filamin, which binds ?-sarcoglycan and ?-sarcoglycan, but not ?-sarcoglycan or ∫-sarcoglycan. Muscular dystrophy, an inherited group of disorders resulting in progressive weakness of muscles in the body, is associated with irregular subcellular localization of Filamin 2 caused by a deficiency in KY, a protein that interacts with Filamin 2. **Function:** FLNC is a muscle-specific filamin, which plays a central role in muscle cells, probably by functioning as a large actin-cross-linking protein. May be involved in reorganizing the actin cytoskeleton in response to signaling events, and may also display structural functions at the Z-disks in muscle cells. Defects in FLNC are the cause of autosomal dominant filaminopathy. Myofibrillar myopathy (MFM) is a neuromuscular disorder, usually with an adult onset, characterized by focal myofibrillar destruction and pathological cytoplasmic protein aggregations. Autosomal dominant filaminopathy is a form of MFM characterized by morphological features of MFM and clinical features of a limb-girdle myopathy. A heterozygous nonsense mutation which segregates with the disease, has been identified in the FLNC gene. **Subunit:** Homodimer. Interacts with KY. Interacts with IGFN1. Interacts with FLNB, KCND2, ITGB1A, INPPL1, MYOT, MYOZ1 and MYOZ3. Interacts with sarcoglycans SGCD and SGCG. Interacts (via filament repeats 17-18, 20-21 and 24) with USP25 (isoform USP25m only). Interacts with FBLIM1. **Subcellular Location:** Cytoplasm. Membrane; Peripheral membrane protein. Cytoplasm, cytoskeleton. Cytoplasm, myofibril, sarcomere, Z line. Note=A small amount localizes at membranes. In striated muscle cells, it predominantly localizes in myofibrillar Z lines, while a minor fraction localizes with subsarcolemme. **Tissue Specificity:** Highly expressed in striated muscles. Weakly expressed in thyroid, fetal brain, fetal lung, retina, spinal cord and bone marrow. Not expressed in testis, pancreas, adrenal gland, placenta, liver and kidney. **Post-translational modifications:** Ubiquitinated by FBXL22, leading to proteasomal degradation. **DISEASE:** Defects in FLNC are the cause of myopathy myofibrillar type 5 (MFM5) [MIM:609524]. A neuromuscular disorder, usually with an adult onset, characterized by focal myofibrillar destruction and pathological cytoplasmic protein aggregations, and

clinical features of a limb-girdle myopathy.
Defects in FLNC are the cause of myopathy distal type 4 (MPD4) [MIM:614065]. MPD4 is a slowly progressive muscular disorder characterized by distal muscle weakness and atrophy affecting the upper and lower limbs. Onset occurs around the third to fourth decades of life, and patients remain ambulatory even after long disease duration. Muscle biopsy shows non-specific changes with no evidence of rods, necrosis, or inflammation.

Similarity:
elongs to the filamin family.
Contains 1 actin-binding domain.
Contains 2 CH (calponin-homology) domains.
Contains 24 filamin repeats.

SWISS:
Q14315

Gene ID:
2318

Database links:

Entrez Gene: 2318Human

Entrez Gene: 68794Mouse

Entrez Gene: 362332Rat

SwissProt: Q14315Human

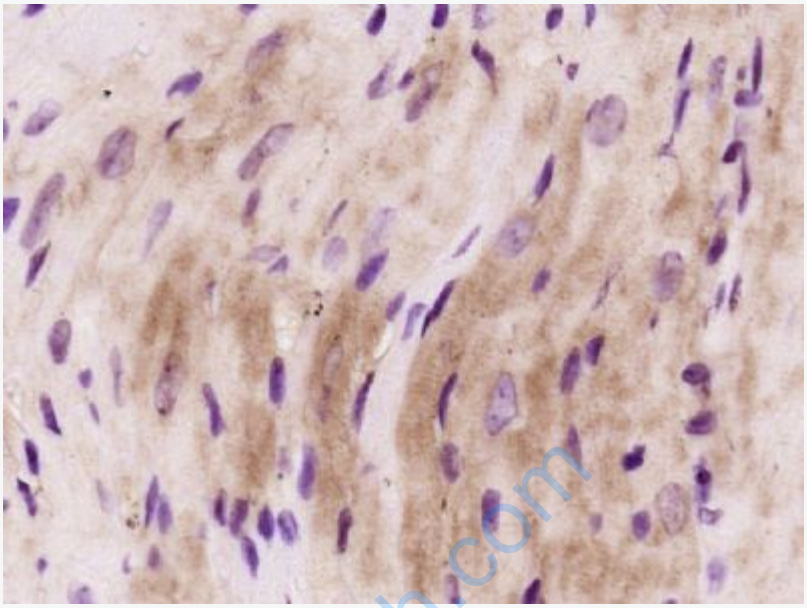
SwissProt: Q8VHX6Mouse

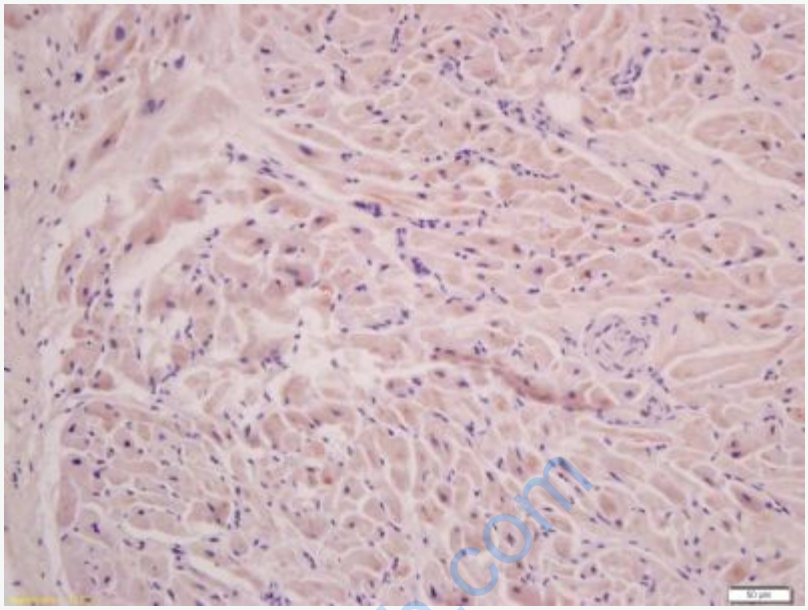
Unigene: 58414Human

Unigene: 39046Mouse

Unigene: 22352Rat

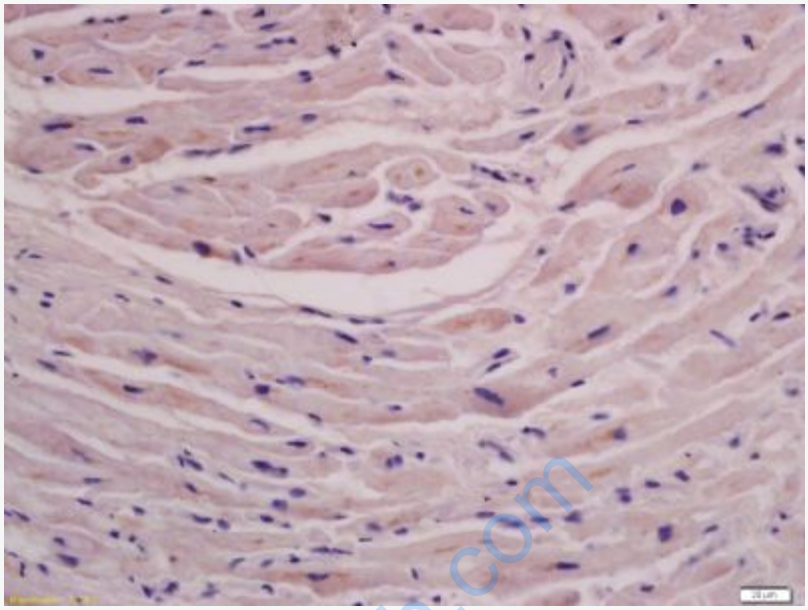
Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Picture:	 Paraformaldehyde-fixed, paraffin embedded (Mouse heart); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (FLNC) Polyclonal Antibody, Unconjugated (SL13182R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.



Tissue/cell: rat cardiac muscle; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min;

Incubation: Anti-FLNC Polyclonal Antibody, Unconjugated(SL13182R) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining



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