10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

# FLNC Rabbit pAb

FLNC Rabbit pAb
Catalog # AP94041

## Product Information

| | |
|---|---|
| **Application** | WB, IHC-P, IHC-F, IF |
| **Primary Accession** | Q14315 |
| **Reactivity** | Human |
| **Host** | Rabbit |
| **Clonality** | Polyclonal |
| **Calculated MW** | 291022 |
| **Physical State** | Liquid |
| **Immunogen** | KLH conjugated synthetic peptide derived from human Filamin 2 |
| **Epitope Specificity** | 251-350/2725 |
| **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** | 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. |
| **SIMILARITY** | elongs to the filamin family.Contains 1 actin-binding domain.Contains 2 CH (calponin-homology) domains.Contains 24 filamin repeats. |
| **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. |
| **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. |
| **Important Note** | This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. |
| **Background Descriptions** | 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.

## Additional Information

| | |
|---|---|
| **Gene ID** | 2318 |
| **Other Names** | Filamin-C, FLN-C, FLNc, ABP-280-like protein, ABP-L, Actin-binding-like protein, Filamin-2, Gamma-filamin, FLNC, ABPL, FLN2 |
| **Target/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. |
| **Dilution** | WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500 |
| **Format** | 0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce |
| **Storage** | Store at -20 °C 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 °C. |

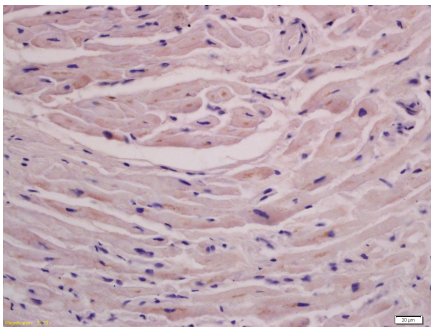
## Protein Information

| | |
|---|---|
| **Name** | FLNC |
| **Synonyms** | ABPL, FLN2 |
| **Function** | Muscle-specific filamin, which plays a central role in sarcomere assembly and organization (PubMed:34405687). Critical for normal myogenesis, it probably functions as a large actin-cross-linking protein with structural functions at the Z lines in muscle cells. May be involved in reorganizing the actin cytoskeleton in response to signaling events (By similarity). |
| **Cellular 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. Targeting to developing and mature Z lines is mediated by the intradomain insert |
| **Tissue Location** | 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. |

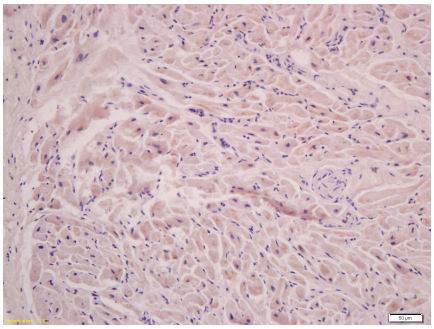
## Background

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

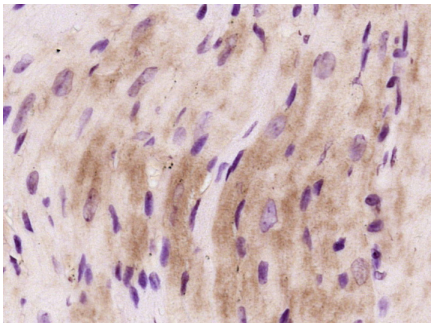
## Images



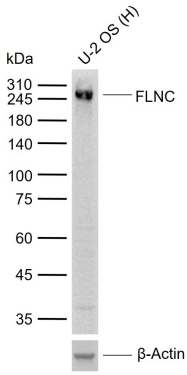
Tissue/cell: rat cardiac muscle; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer ( 0.01M, pH 6.0 ), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min; Incubation: Anti-FLNC Polyclonal Antibody, Unconjugated(AP94041) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining



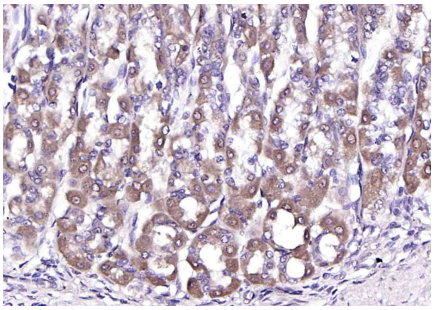
Tissue/cell: rat cardiac muscle; 4% Paraformaldehyde-fixed and paraffin-embedded; Antigen retrieval: citrate buffer ( 0.01M, pH 6.0 ), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37℃ for 20 min; Incubation: Anti-FLNC Polyclonal Antibody, Unconjugated(AP94041) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining



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 (AP94041) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.




Sample: Lane 1: Human U-2 OS cell lysates Primary: Anti-FLNC (AP94041) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 300 kDa Observed band size: 280 kDa



Paraformaldehyde-fixed, paraffin embedded (rat stomach); 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; Incubation with (FLNC) Polyclonal Antibody, Unconjugated (AP94041) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.