FLNC Rabbit pAb

CatalogNo: YN3834

Key Features

Host Species
- Rabbit

Reactivity
- Human,Mouse

Applications
- IHC,IF

MW
- 300kD (Calculated)

Isotype
- IgG

Recommended Dilution Ratios

IHC 1:50-200
IF 1:50-200

Storage

Storage*	-15°C to -25°C/1 year(Do not lower than -25°C)
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Basic Information

Clonality	Polyclonal

Immunogen Information

Immunogen	Synthesized peptide derived from human FLNC AA range: 335-385
Specificity	This antibody detects endogenous levels of FLNC at Human/Mouse

Target Information

Gene name	FLNC ABPL FLN2

Protein Name FLNC

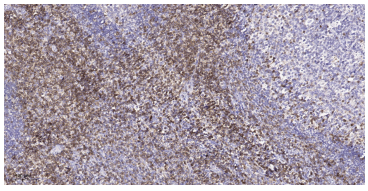
Organism	Gene ID	UniProt ID
Human	2318;	Q14315;
Mouse	68794;	Q8VHX6;

Cellular Localization Cytoplasm . Membrane ; Peripheral membrane protein . Cytoplasm, cytoskeleton . Cytoplasm, myofibril, sarcomere, Z line . 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 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.

Function developmental stage:Expressed in both differentiating and adult muscles.,Disease:Defects in FLNC are the cause of autosomal dominant filaminopathy [MIM:609524, 601419]. 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.,Domain:Comprised of a NH2-terminal actin-binding domain, 24 internally homologous repeats and two hinge regions. Repeat 24 and the second hinge domain are important for dimer formation.,Domain:The filamin 20 repeat mediates interaction with XIRP1.,Domain:The intradomain insert is specific to FLNC and mediates the targeting to developing and mature Z-disks.,Function: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.,miscellaneous:Silenced in MKN28 and MKN74 gastric cancer cell lines due to aberrant methylation of the gene.,similarity:Belongs to the filamin family.,similarity:Contains 1 actin-binding domain.,similarity:Contains 2 CH (calponin-homology) domains.,similarity:Contains 24 filamin repeats.,subcellular location:A small amount localizes at membranes. In striated muscle cells, it predominantly localizes in myofibrillar Z-disks, while a minor fraction localizes with subsarcolemme.,subunit:Homodimer. Interacts with KY (By similarity). Interacts with FLNB, KCND2, ITGB1A, INPPL1, MYOT, MYOZ1 and MYOZ3. Interacts with sarcoglycans SGCD and SGCG.,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.,

Validation Data



Immunohistochemical analysis of paraffin-embedded human tonsil. 1, Tris-EDTA,pH9.0 was used for antigen retrieval. 2 Antibody was diluted at 1:200(4° overnight.3,Secondary antibody was diluted at 1:200(room temperature, 45min).

Contact information

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