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phospho-FLNC (Ser2233) Rabbit pAb

Catalog # AP99270

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q14315
Other Accession	Q14315
Reactivity	Mouse
Predicted	Rat, Pig, Human, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	291022
Physical State	Liquid
Immunogen	KLH conjugated synthesised phosphopeptide derived from human FLNC around the phosphorylation site of Ser2233
Epitope Specificity	LG(p-S)FG
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
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	ABP-280; ABP280A; ABPA; ABPL; ARVC15; CMH26; FLN2; MFM5; MPD4; RCM5; 1110055E19Rik; ABP-L; FLN-C; FLNC_HUMAN; FLNC; ABP-280-like protein; Actin-binding-like protein; Filamin-2; Gamma-filamin; FLNC_MOUSE; filamin C; filamin C, gamma (actin binding protein 280); filamin C, gamma; actin binding protein 280; gamma filamin; FLNC | Filamin C (phospho-S2233); p-Filamin C; phospho-Filamin C; p-FLNC; FLNC | Filamin C (phospho-Ser2233)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.