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

CSRP3 Rabbit pAb

Catalog # AP97619

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	P50461
Other Accession	P50461
Predicted	Rat, Pig, Human, Mouse, Chicken, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	20969
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CSRP3
Epitope Specificity	101-194/194
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 2 LIM zinc-binding domains.
DISEASE	Defects in CSRP3 are the cause of cardiomyopathy dilated type 1M (CMD1M) [MIM:607482]. Dilated cardiomyopathy is a disorder characterized by ventricular dilation and impaired systolic function, resulting in congestive heart failure and arrhythmia. Patients are at risk of premature death.Defects in CSRP3 are the cause of cardiomyopathy familial hypertrophic type 12 (CMH12) [MIM:612124]. Familial hypertrophic cardiomyopathy is a hereditary heart disorder characterized by ventricular hypertrophy, which is usually asymmetric and often involves the interventricular septum. The symptoms include dyspnea, syncope, collapse, palpitations, and chest pain. They can be readily provoked by exercise. The disorder has inter- and intrafamilial variability ranging from benign to malignant forms with high risk of cardiac failure and sudden cardiac death.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a member of the CSRP family of LIM domain proteins, which may be involved in regulatory processes important for development and cellular differentiation. The LIM/double zinc-finger motif found in this protein is found in a group of proteins with critical functions in gene regulation, cell growth, and somatic differentiation. Mutations in this gene are thought to cause heritable forms of hypertrophic cardiomyopathy (HCM) and dilated cardiomyopathy (DCM) in humans. Alternatively spliced transcript variants with different 5' UTR, but encoding the same protein, have been found for this gene. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	8048
Other Names	CLP; CMD1M; CMH12; CRP3; LMO4; MLP; MMLP; CSRP3_BOVIN; CSRP3;

	Cysteine-rich protein 3 (CRP3); CSRP3_HUMAN; Cardiac LIM protein; LIM domain protein, cardiac; Muscle LIM protein; CSRP3_MOUSE; CSRP3_RAT; cysteine and glycine rich protein 3
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	CSRP3
Synonyms	CLP, MLP
Function	Positive regulator of myogenesis. Acts as a cofactor for myogenic bHLH transcription factors such as MYOD1, and probably MYOG and MYF6. Enhances the DNA-binding activity of the MYOD1:TCF3 isoform E47 complex and may promote formation of a functional MYOD1:TCF3 isoform E47:MEF2A complex involved in myogenesis (By similarity). Plays a crucial and specific role in the organization of cytosolic structures in cardiomyocytes. Could play a role in mechanical stretch sensing. May be a scaffold protein that promotes the assembly of interacting proteins at Z-line structures. It is essential for calcineurin anchorage to the Z line. Required for stress-induced calcineurin-NFAT activation (By similarity). The role in regulation of cytoskeleton dynamics by association with CFL2 is reported conflictingly: Shown to enhance CFL2-mediated F-actin depolymerization dependent on the CSRP3:CFL2 molecular ratio, and also shown to reduce the ability of CLF1 and CFL2 to enhance actin depolymerization (PubMed:19752190, PubMed:24934443). Proposed to contribute to the maintenance of muscle cell integrity through an actin-based mechanism. Can directly bind to actin filaments, cross-link actin filaments into bundles without polarity selectivity and protect them from dilution- and cofilin- mediated depolymerization; the function seems to involve its self- association (PubMed:24934443). In vitro can inhibit PKC/PRKCA activity (PubMed:27353086). Proposed to be involved in cardiac stress signaling by down-regulating excessive PKC/PRKCA signaling (By similarity).
Cellular Location	Nucleus {ECO:0000250|UniProtKB:P50463}. Cytoplasm. Cytoplasm, cytoskeleton Cytoplasm, myofibril, sarcomere, Z line Cytoplasm, myofibril, sarcomere Note=Nucleocytoplasmic shuttling protein. Mainly cytoplasmic. In the Z line, found associated with GLRX3 (By similarity) {ECO:0000250|UniProtKB:P50462, ECO:0000250|UniProtKB:P50463}
Tissue Location	Cardiac and slow-twitch skeletal muscles. Isoform 2 is expressed in striated muscle. Isoform 2 is specifically expressed at higher levels in patients with neuromuscular diseases, such as limb- girdle muscular dystrophy 2A (LGMD2A), Duchenne muscular dystrophy (DMD) and dermatomyositis (PubMed:24860983)

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