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MEFV Rabbit pAb

Catalog # AP98125

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	O15553
Other Accession	O15553
Predicted	Human
Host	Rabbit
Clonality	Polyclonal
Calculated MW	86444
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human MEFV
Epitope Specificity	501-600/781
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 1 B box-type zinc finger. Contains 1 B30.2/SPRY domain. Contains 1 DAPIN domain.
Post-translational modifications	Cleaved by CASP1 Probable. The N-terminal cleavage product localizes to the nucleus as a filamentous network and to the cytoplasm, interacts more strongly with RELA and NFKBIA than the full-length protein, enhances the nuclear localization of RELA and induces NFKBIA proteolysis. The C-terminal cleavage product localizes to the cytoplasm.
DISEASE	Defects in MEFV are the cause of familial Mediterranean fever autosomal recessive (ARFMF) [MIM:249100]. ARFMF is an inherited disorder characterized by recurrent episodic fever, serosal inflammation and pain in the abdomen, chest or joints. ARFMF is frequently complicated by amyloidosis, which leads to renal failure and can be prophylactically treated with colchicine. ARFMF primarily affects ancestral ethnic groups living around the Mediterranean basin: North African Jews, Armenians, Arabs and Turks. The disease is also distributed in other populations including Greeks, Cypriots, Italians and Spanish, although at a lower prevalence. Defects in MEFV are the cause of familial Mediterranean fever autosomal dominant (ADFMF) [MIM:134610]. ADFMF is characterized by periodic fever, serosal inflammation and pain in the abdomen, chest or joints as seen also in the autosomal recessive form of the disease. It is associated with renal amyloidosis and characterized by colchicine unresponsiveness.
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 protein, also known as pyrin or marenostrin, that is an important modulator of innate immunity. Mutations in this gene are associated with Mediterranean fever, a hereditary periodic fever syndrome. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	4210
Other Names	FMF; MEF; PAAND; TRIM20; pyrin; MEFV_HUMAN; MEFV; Marenostrin; MEFV_MOUSE; MEFV innate immunity regulator, pyrin; Mediterranean fever; MEFV, pyrin innate immunity regulator
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	MEFV {ECO:0000303|PubMed:11115844, ECO:0000312|HGNC:HGNC:6998}
Function	Involved in the regulation of innate immunity and the inflammatory response in response to IFNG/IFN-gamma (PubMed:10807793, PubMed:11468188, PubMed:16037825, PubMed:16785446, PubMed:17431422, PubMed:17964261, PubMed:18577712, PubMed:19109554, PubMed:19584923, PubMed:26347139, PubMed:27030597, PubMed:28835462). Organizes autophagic machinery by serving as a platform for the assembly of ULK1, Beclin 1/BECN1, ATG16L1, and ATG8 family members and recognizes specific autophagy targets, thus coordinating target recognition with assembly of the autophagic apparatus and initiation of autophagy (PubMed:16785446, PubMed:17431422, PubMed:26347139). Acts as an autophagy receptor for the degradation of several inflammasome components, including CASP1, NLRP1 and NLRP3, hence preventing excessive IL1B- and IL18-mediated inflammation (PubMed:16785446, PubMed:17431422, PubMed:26347139). However, it can also have a positive effect in the inflammatory pathway, acting as an innate immune sensor that triggers PYCARD/ASC specks formation, caspase-1 activation, and IL1B and IL18 production (PubMed:16037825, PubMed:27030597, PubMed:28835462). Together with AIM2, also acts as a mediator of pyroptosis, necroptosis and apoptosis (PANoptosis), an integral part of host defense against pathogens, in response to bacterial infection (By similarity). It is required for PSTPIP1-induced PYCARD/ASC oligomerization and inflammasome formation (PubMed:10807793, PubMed:11468188, PubMed:17964261, PubMed:18577712, PubMed:19109554, PubMed:19584923). Recruits PSTPIP1 to inflammasomes, and is required for PSTPIP1 oligomerization (PubMed:10807793, PubMed:11468188, PubMed:17964261, PubMed:18577712, PubMed:19109554, PubMed:19584923).
Cellular Location	[Isoform 1]: Cytoplasm, cytoskeleton. Cell projection, ruffle. Cell projection, lamellipodium. Nucleus. Cytoplasm. Cytoplasmic vesicle, autophagosome. Note=Associated with microtubules and with the filamentous actin of perinuclear filaments and peripheral lamellar ruffles (PubMed:11468188). In pre- apoptotic cells, colocalizes with PYCARD/ASC in large specks (inflammasomes) (PubMed:11468188). In migrating monocytes, strongly polarized at the leading edge of the cell where it colocalizes with polymerizing actin and PYCARD/ASC (PubMed:11468188)
Tissue Location	Expressed in peripheral blood leukocytes, particularly in mature granulocytes and to a lesser extent in monocytes but not in lymphocytes. Detected in spleen, lung and muscle, probably as a result of leukocyte infiltration in these tissues. Not expressed in thymus, prostate, testis, ovary, small intestine,

colon, heart, brain, placenta, liver, kidney, pancreas. Expression detected in several myeloid leukemic, colon cancer, and prostate cancer cell lines

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