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NALP3/CIAS1 Rabbit pAb

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Catalog # AP94237

Product Information

Application	WB
Primary Accession	Q8R4B8
Reactivity	Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	118275
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from mouse NALP3/CIAS1
Epitope Specificity	921-1020/1033
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.
SIMILARITY	Belongs to the NLRP family.Contains 1 DAPIN domain.Contains 9 LRR (leucine-rich) repeats.Contains 1 NACHT domain.
DISEASE	Defects in NLRP3 are the cause of familial cold autoinflammatory syndrome type 1 (FCAS1) [MIM:120100]; also known as familial cold urticaria. FCAS are rare autosomal dominant systemic inflammatory diseases characterized by episodes of rash, arthralgia, fever and conjunctivitis after generalized exposure to cold. Defects in NLRP3 are a cause of Muckle-Wells syndrome (MWS) [MIM:191900]; also known as urticaria-deafness-amyloidosis syndrome. MWS is a hereditary periodic fever syndrome characterized by fever, chronic recurrent urticaria, arthralgias, progressive sensorineural deafness, and reactive renal amyloidosis. The disease may be severe if generalized amyloidosis occurs. Defects in NLRP3 are the cause of chronic infantile neurologic cutaneous and articular syndrome (CINCA) [MIM:607115]; also known as neonatal onset multisystem inflammatory disease (NOMID). CINCA is a rare congenital inflammatory disorder characterized by a triad of neonatal onset of cutaneous symptoms, chronic meningitis and joint manifestations with recurrent fever and inflammation.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	May function as an inducer of apoptosis. Interacts selectively with ASC and this complex may function as an upstream activator of NF-kappa-B signaling. Inhibits TNF-alpha induced activation and nuclear translocation of RELA/NF-KB p65. Also inhibits transcriptional activity of RELA. Activates caspase-1 in response to a number of triggers including bacterial or viral infection which leads to processing and release of IL1B and IL18. Subcellular Location : Cytoplasm.

Additional Information

Gene ID	216799
Other Names	NACHT, LRR and PYD domains-containing protein 3, 3.6.4.-, Cold autoinflammatory syndrome 1 protein homolog, Cryopyrin, Mast cell maturation-associated-inducible protein 1, PYRIN-containing APAF1-like protein 1, Nlrp3 {ECO:0000303|PubMed:17907925, ECO:0000312|MGI:MGI:2653833}
Target/Specificity	Expressed in blood leukocytes. Strongly expressed in polymorphonuclear cells and osteoblasts. Undetectable or expressed at a lower magnitude in B- and T-lymphoblasts, respectively. High level of expression detected in chondrocytes. Detected in non-keratinizing epithelia of oropharynx, esophagus and ectocervix and in the urothelial layer of the bladder.
Dilution	WB=1:500-2000
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

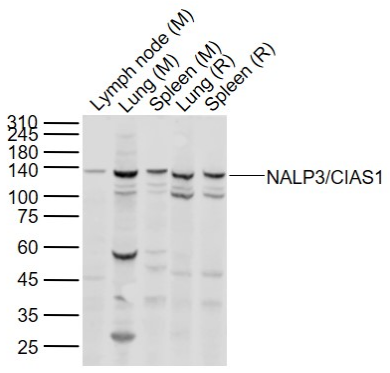
Name	Nlrp3 {ECO:0000303|PubMed:17907925, ECO:0000312|MGI:MGI:2653833}
Function	Sensor component of the NLRP3 inflammasome, which mediates inflammasome activation in response to defects in membrane integrity, leading to secretion of inflammatory cytokines IL1B and IL18 and pyroptosis (PubMed:19362020, PubMed:23582325, PubMed:26642356, PubMed:26814970, PubMed:27374331, PubMed:27929086, PubMed:28656979, PubMed:28847925, PubMed:30518920, PubMed:36178239). In response to pathogens and other damage-associated signals that affect the integrity of membranes, initiates the formation of the inflammasome polymeric complex composed of NLRP3, CASP1 and PYCARD/ASC (PubMed:16407889, PubMed:18403674, PubMed:19362020, PubMed:26642356, PubMed:26814970, PubMed:27374331, PubMed:28847925). Recruitment of pro-caspase-1 (proCASP1) to the NLRP3 inflammasome promotes caspase-1 (CASP1) activation, which subsequently cleaves and activates inflammatory cytokines IL1B and IL18 and gasdermin-D (GSDMD), promoting cytokine secretion and pyroptosis (PubMed:16546100, PubMed:17008311, PubMed:26642356, PubMed:26814970, PubMed:27374331, PubMed:28847925). Activation of NLRP3 inflammasome is also required for HMGB1 secretion; stimulating inflammatory responses (PubMed:22801494). Under resting conditions, ADP-bound NLRP3 is autoinhibited (By similarity). NLRP3 activation stimuli include extracellular ATP, nigericin, reactive oxygen species, crystals of monosodium urate or cholesterol, amyloid- beta fibers, environmental or industrial particles and nanoparticles, such as asbestos, silica, aluminum salts, cytosolic dsRNA, etc (PubMed:16407888, PubMed:16407889, PubMed:16407890, PubMed:18403674, PubMed:19362020, PubMed:37001519). Almost all stimuli trigger intracellular K(+) efflux (PubMed:23809161). These stimuli lead to membrane perturbation and activation of NLRP3 (By similarity). Upon activation, NLRP3 is transported to microtubule organizing center (MTOC), where it is unlocked by NEK7, leading to its relocalization to dispersed trans-Golgi network (dTGN) vesicle membranes and formation of an active inflammasome complex (PubMed:26814970, PubMed:34615873, PubMed:34861190). Associates with dTGN vesicle membranes by binding to

	phosphatidylinositol 4-phosphate (PtdIns4P) (PubMed:30487600). Shows ATPase activity (PubMed:34861190).
Cellular Location	Cytoplasm, cytosol. Inflammasome. Cytoplasm, cytoskeleton, microtubule organizing center. Golgi apparatus membrane. Endoplasmic reticulum. Mitochondrion. Secreted Nucleus. Note=In macrophages, under resting conditions, mainly located in the cytosol and on membranes of various organelles, such as endoplasmic reticulum, mitochondria and Golgi: forms an inactive double-ring cage that is primarily localized on membranes (PubMed:23502856, PubMed:28716882, PubMed:34861190). Upon activation, NLRP3 is transported to microtubule organizing center (MTOC), where it is unlocked by NEK7, leading to its relocalization to dispersed trans-Golgi network (dTGN) vesicle membranes for the formation of an active inflammasome complex (PubMed:34861190) Recruited to dTGN vesicle membranes by binding to phosphatidylinositol 4-phosphate (PtdIns4P) (PubMed:30487600). After the induction of pyroptosis, inflammasome specks are released into the extracellular space where they can further promote IL1B processing and where they can be engulfed by macrophages. Phagocytosis induces lysosomal damage and inflammasome activation in the recipient cells (PubMed:24952504, PubMed:24952505). In the Th2 subset of CD4(+) helper T-cells, mainly located in the nucleus (PubMed:26098997). Nuclear localization depends upon KPNA2 (PubMed:26098997). In the Th1 subset of CD4(+) helper T- cells, mainly cytoplasmic (PubMed:26098997)
Tissue Location	Expressed with high levels in peripheral blood leukocytes, including Th2 lymphocytes and macrophages (PubMed:15302403, PubMed:16546100, PubMed:26098997, PubMed:28847925). Expressed at low levels in resting osteoblasts (at protein level) (PubMed:17907925)

Background

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

Images



Sample: Lane 1: Lymph node (Mouse) Lysate at 40 ug Lane 2: Lung (Mouse) Lysate at 40 ug Lane 3: Spleen (Mouse) Lysate at 40 ug Lane 4: Lung (Rat) Lysate at 40 ug Lane 5: Spleen (Rat) Lysate at 40 ug Primary: Anti-NALP3/CIAS1 (AP94237) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 114-118 kD Observed band size: 120 kD