

Rabbit Anti-WASP antibody

SL13681R

Product Name:	WASP
Chinese Name:	湿疹血小板减少伴免疫缺陷综合征相关蛋白抗体
Alias:	Eczema thrombocytopenia; IMD2; SCNX; THC; THC1; Thrombocytopenia 1 (X linked); U42471; Was; WASp; WASP_HUMAN; Wiskott Aldrich syndrome (eczema thrombocytopenia); Wiskott Aldrich syndrome; Wiskott Aldrich syndrome protein; Wiskott-Aldrich syndrome protein.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Cow, Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100- 500IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	53kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	lmg/ml
immunogen:	KLH conjugated synthetic peptide derived from human WASP:101-200/502
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	The Wiskott-Aldrich syndrome (WAS) is a disorder that results from a monogenic defect that has been mapped to the short arm of the X chromosome. WAS is characterized by thrombocytopenia, eczema, defects in cell-mediated and humoral immunity and a propensity for lymphoproliferative disease. The gene that is mutated in

the syndrome encodes a proline-rich protein of unknown function designated WAS protein (WASP). A clue to WASP function came from the observation that T cells from affected males had an irregular cellular morphology and a disarrayed cytoskeleton suggesting the involvement of WASP in cytoskeletal organization. Close examination of the WASP sequence revealed a putative Cdc42/Rac interacting domain, homologous with those found in PAK65 and ACK. Subsequent investigation has shown WASP to be a true downstream effector of Cdc42.

Function:

Effector protein for Rho-type GTPases, providing a link with the Arp2/3 complex that regulates the structure and dynamics of the actin cytoskeleton. Important for efficient actin polymerization. Possible regulator of lymphocyte and platelet function.

Subunit:

Interacts with NCK1 (via SH3 domains). Interacts with CDC42, RAC, NCK, HCK, FYN, SRC kinase FGR, BTK, ABL1, PSTPIP1, WIP, and to the p85 subunit of PLC-gamma. Binds the Arp2/3 complex. Interacts (via C-terminus) with ALDOA. Interacts with E.coli effector protein EspF(U).

Subcellular Location:

Cytoplasm; cytoskeleton.

Tissue Specificity:

Expressed predominantly in the thymus. Also found, to a much lesser extent, in the spleen

Post-translational modifications:

Phosphorylated at Tyr-291 by FYN and HCK, inducing WAS effector activity after TCR engagement. Phosphorylation at Tyr-291 enhances WAS activity in promoting actin polymerization and filopodia formation.

DISEASE:

Defects in WAS are the cause of Wiskott-Aldrich syndrome (WAS) [MIM:301000]; also known as eczema-thrombocytopenia-immunodeficiency syndrome. WAS is an X-linked recessive immunodeficiency characterized by eczema,

thrombocytopenia, recurrent infections, and bloody diarrhea. Death usually occurs before age 10. Defects in WAS are the cause of thrombocytopenia type 1 (THC1) [MIM:313900].

Thrombocytopenia is defined by a decrease in the number of platelets in circulating blood, resulting in the potential for increased bleeding and decreased ability for clotting. Defects in WAS are a cause of neutropenia severe congenital X-linked (XLN) [MIM:300299]. XLN is an immunodeficiency syndrome characterized by recurrent major bacterial infections, severe congenital neutropenia, and monocytopenia.

Similarity:

Contains 1 CRIB domain.

Contains 1 WH1 domain. Contains 1 WH2 domain.
SWISS: P42768
Gene ID: 7454
Database links:
Entrez Gene: 7454 Human
Entrez Gene: 22376 Mouse
<u>Omim: 300392</u> Human
Entrez Gene: 22376 Mouse Omim: 300392 Human SwissProt: P42768 Human SwissProt: P70315 Mouse Unigene: 2157 Human
SwissProt: P70315 Mouse
Unigene: 2157 Human
Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
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