

RNF74 Rabbit pAb

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Catalog # AP58559

Product Information

Application	WB
Primary Accession	P15918
Reactivity	Mouse
Predicted	Rat, Pig, Human, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	119097
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human RAG1/RNF74
Epitope Specificity	351-450/1043
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	Nucleus.
DISEASE	Defects in RAG1 are a cause of combined cellular and humoral immune defects with granulomas (CHIDG) [MIM:233650]. CHIDG is an immunodeficiency disease with granulomas in the skin, mucous membranes, and internal organs. Other characteristics include hypogammaglobulinemia, a diminished number of T and B-cells, and sparse thymic tissue on ultrasonography. Defects in RAG1 are a cause of severe combined immunodeficiency autosomal recessive T-cell-negative/B-cell-negative/NK-cell-positive (T(-)B(-)NK(+)) SCID [MIM:601457]. A form of severe combined immunodeficiency (SCID), a genetically and clinically heterogeneous group of rare congenital disorders characterized by impairment of both humoral and cell-mediated immunity, leukopenia, and low or absent antibody levels. Patients present in infancy recurrent, persistent infections by opportunistic organisms. The common characteristic of all types of SCID is absence of T-cell-mediated cellular immunity due to a defect in T-cell development. Defects in RAG1 are a cause of Omenn syndrome (OS) [MIM:603554]. OS is a severe immunodeficiency characterized by the presence of activated, anergic, oligoclonal T-cells, hypereosinophilia, and high IgE levels. Defects in RAG1 are the cause of alpha/beta T-cell lymphopenia with gamma/delta T-cell expansion severe cytomegalovirus infection and autoimmunity (T-CMVA) [MIM:609889]. An immunological disorder characterized by oligoclonal expansion of TCR gamma/delta T-cells, TCR alpha/beta T-cell lymphopenia, severe, disseminated cytomegalovirus infection and autoimmune cytopenia.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Catalytic component of the RAG complex, a multiprotein complex that mediates the DNA cleavage phase during V(D)J recombination. V(D)J recombination assembles a diverse repertoire of immunoglobulin and T-cell receptor genes in developing B and T lymphocytes through rearrangement of

different V (variable), in some cases D (diversity), and J (joining) gene segments. In the RAG complex, RAG1 mediates the DNA-binding to the conserved recombination signal sequences (RSS) and catalyzes the DNA cleavage activities by introducing a double-strand break between the RSS and the adjacent coding segment. RAG2 is not a catalytic component but is required for all known catalytic activities. DNA cleavage occurs in 2 steps: a first nick is introduced in the top strand immediately upstream of the heptamer, generating a 3'-hydroxyl group that can attack the phosphodiester bond on the opposite strand in a direct transesterification reaction, thereby creating 4 DNA ends: 2 hairpin coding ends and 2 blunt, 5'-phosphorylated ends. The chromatin structure plays an essential role in the V(D)J recombination reactions and the presence of histone H3 trimethylated at 'Lys-4' (H3K4me3) stimulates both the nicking and hairpinning steps. The RAG complex also plays a role in pre-B cell allelic exclusion, a process leading to expression of a single immunoglobulin heavy chain allele to enforce clonality and monospecific recognition by the B-cell antigen receptor (BCR) expressed on individual B lymphocytes. The introduction of DNA breaks by the RAG complex on one immunoglobulin allele induces ATM-dependent repositioning of the other allele to pericentromeric heterochromatin, preventing accessibility to the RAG complex and recombination of the second allele. In addition to its endonuclease activity, RAG1 also acts as a E3 ubiquitin-protein ligase that mediates monoubiquitination of histone H3. Histone H3 monoubiquitination is required for the joining step of V(D)J recombination. Mediates polyubiquitination of KPNA1.

Additional Information

Gene ID	5896
Other Names	V(D)J recombination-activating protein 1, RAG-1, RING finger protein 74, Endonuclease RAG1, 3.1.-., E3 ubiquitin-protein ligase RAG1, 2.3.2.27, RING-type E3 ubiquitin transferase RAG1, RAG1, RNF74
Dilution	WB=1:500-2000,Flow-Cyt=1 µg/Test
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	RAG1
Synonyms	RNF74
Function	Catalytic component of the RAG complex, a multiprotein complex that mediates the DNA cleavage phase during V(D)J recombination. V(D)J recombination assembles a diverse repertoire of immunoglobulin and T-cell receptor genes in developing B and T- lymphocytes through rearrangement of different V (variable), in some cases D (diversity), and J (joining) gene segments. In the RAG complex, RAG1 mediates the DNA-binding to the conserved recombination signal sequences (RSS) and catalyzes the DNA cleavage activities by introducing a double-strand break between the RSS and the adjacent coding segment. RAG2 is not a catalytic component but is required for all known catalytic activities. DNA cleavage occurs in 2 steps: a first nick is introduced in the top strand immediately upstream of the

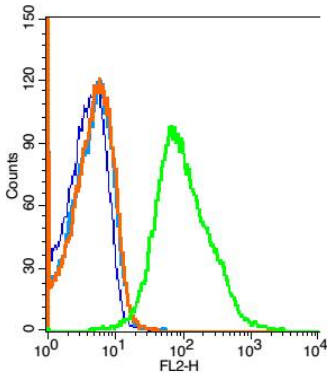
heptamer, generating a 3'-hydroxyl group that can attack the phosphodiester bond on the opposite strand in a direct transesterification reaction, thereby creating 4 DNA ends: 2 hairpin coding ends and 2 blunt, 5'-phosphorylated ends. The chromatin structure plays an essential role in the V(D)J recombination reactions and the presence of histone H3 trimethylated at 'Lys-4' (H3K4me3) stimulates both the nicking and haipinning steps. The RAG complex also plays a role in pre-B cell allelic exclusion, a process leading to expression of a single immunoglobulin heavy chain allele to enforce clonality and monospecific recognition by the B-cell antigen receptor (BCR) expressed on individual B-lymphocytes. The introduction of DNA breaks by the RAG complex on one immunoglobulin allele induces ATM- dependent repositioning of the other allele to pericentromeric heterochromatin, preventing accessibility to the RAG complex and recombination of the second allele. In addition to its endonuclease activity, RAG1 also acts as an E3 ubiquitin-protein ligase that mediates monoubiquitination of histone H3. Histone H3 monoubiquitination is required for the joining step of V(D)J recombination. Mediates polyubiquitination of KPNA1 (By similarity).

Cellular Location	Nucleus {ECO:0000255 PROSITE-ProRule:PRU00820}.
Tissue Location	Maturing lymphoid cells.

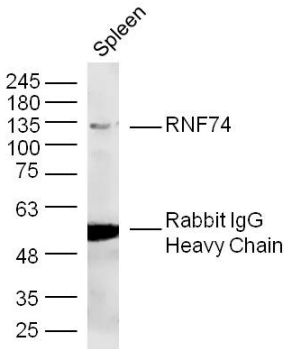
Background

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

Images

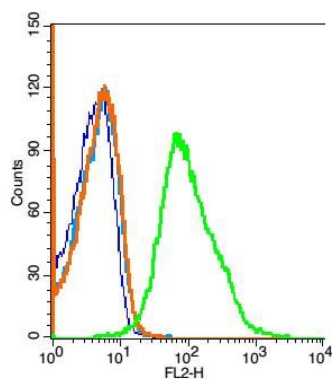


Blank control: Mouse Spleen Cells(fixed with 2% paraformaldehyde (10 min) , then permeabilized with 90% ice-cold methanol for 30 min on ice). Primary Antibody:Rabbit Anti- RNF74 antibody(AP58559), Dilution: 1ug in 100 uL 1X PBS containing 0.5% BSA; Isotype Control Antibody: Rabbit IgG(orange) ,used under the same conditions); Secondary Antibody: Goat anti-rabbit IgG-PE(white blue), Dilution: 1:200 in 1 X PBS containing 0.5% BSA.



Sample: Spleen (Mouse) Lysate at 40 ug
 Primary: Anti-RNF74 (AP58559) at 1/300 dilution
 Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
 Predicted band size: 115 kD
 Observed band size: 130 kD

Blank control: Mouse Spleen Cells(fixed with 2% paraformaldehyde (10 min) , then permeabilized with 90% ice-cold methanol for 30 min on ice).



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Secondary Antibody: Goat anti-rabbit IgG-PE(white
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Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.