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RAG2 Recombinant Rabbit mAb

Catalog # AP96414

Product Information

Application	WB
Reactivity	Human, Mouse
Host	Rabbit
Clonality	Recombinant
Physical State	Liquid
Isotype	IgG/Kappa
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the RAG2 family.Contains 1 PHD-type zinc finger.
SUBUNIT	Component of the RAG complex composed of core components RAG1 and RAG2, and associated component HMGB1 or HMGB2.
DISEASE	Defects in RAG2 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 RAG2 are a cause of severe combinedimmunodeficiency 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 RAG2 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.
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 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 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

Other Names	RAG-2; RAG2_HUMAN; RAG2; RAG2_MOUSE; RAG2_RABIT; recombination activating 2; recombination activating gene 2
Dilution	WB=1:500-2000
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.
Name	RAG2 Recombinant Rabbit mAb
Function	Core 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. DNA cleavage by the RAG complex 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 the RAG complex, RAG2 is not the catalytic component but is required for all known catalytic activities mediated by RAG1. It probably acts as a sensor of chromatin state that recruits the RAG complex to H3K4me3.

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