

Product datasheet

Anti-RAG2 antibody [4D5] ab96486

画像数 1

製品の概要

製品名	Anti-RAG2 antibody [4D5]
製品の詳細	Mouse monoclonal [4D5] to RAG2
由来種	Mouse
アプリケーション	適用あり: WB, ELISA
種交差性	交差種: Human
免疫原	Purified recombinant fragment of Human RAG2 (amino acids 350-527) expressed in E. coli.
ポジティブ・コントロール	RAG2-hlgGfc transfected HEK293 cell lysate

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
バッファー	Preservative: 0.03% Sodium Azide Constituents: Ascites
精製度	Ascites
ポリ/モノ	モノクローナル
クローン名	4D5
アイソタイプ	IgG1

アプリケーション

Our [Abpromise guarantee](#) covers the use of **ab96486** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

アプリケーション	Abreviews	特記事項
WB		1/500 - 1/2000. Predicted molecular weight: 59 kDa.
ELISA		1/10000.

機能

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 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 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.

組織特異性

Cells of the B- and T-lymphocyte lineages.

関連疾患

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 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 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.

配列類似性

Belongs to the RAG2 family.

Contains 1 PHD-type zinc finger.

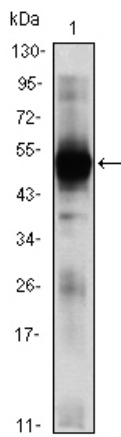
ドメイン

The atypical PHD-type zinc finger recognizes and binds histone H3 trimethylated on 'Lys-4' (H3K4me3). The presence Tyr-445 instead of a carboxylate in classical PHD-type zinc fingers results in an enhanced binding to H3K4me3 in presence of dimethylated on 'Arg-2' (H3R2me2) rather than inhibited. The atypical PHD-type zinc finger also binds various phosphoinositides, such as phosphatidylinositol-3,4-bisphosphate binding (PtdIns(3,4)P2), phosphatidylinositol-3,5-bisphosphate binding (PtdIns(3,5)P2), phosphatidylinositol-4,5-bisphosphate (PtdIns(4,5)P2) and phosphatidylinositol-3,4,5-trisphosphate binding (PtdIns(3,4,5)P3).

細胞内局在

Nucleus.

画像



Western blot - RAG2 antibody [4D5] (ab96486)

Anti-RAG2 antibody [4D5] (ab96486) at 1/500 dilution + RAG2-hlgGfc transfected HEK293 cell lysate

Predicted band size: 59 kDa

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