

CRLF1 Rabbit pAb

Catalog # AP97540

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	O75462
Other Accession	O75462
Host	Rabbit
Clonality	Polyclonal
Calculated MW	46302
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CRLF1
Epitope Specificity	201-300/422
Isotype	IgG
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 type I cytokine receptor family. Type 3 subfamily. Contains 2 fibronectin type-III domains. Contains 1 Ig-like C2-type (immunoglobulin-like) domain.
SUBUNIT	Forms covalently linked di- and tetramers. Forms a heteromeric complex with cardiotrophin-like cytokine (CLC); the CRLF1/CLC complex is a ligand for the ciliary neurotrophic factor receptor (CNTFR).
DISEASE	Defects in CRLF1 are the cause of cold-induced sweating syndrome type 1 (CISS1) [MIM:272430]. Cold-induced sweating syndrome (CISS) is an autosomal recessive disorder characterized by profuse sweating induced by cool surroundings (temperatures of 7 to 18 degrees Celsius). Additional abnormalities include a high-arched palate, nasal voice, depressed nasal bridge, inability to fully extend the elbows and kyphoscoliosis. Defects in CRLF1 are the cause of Crisponi syndrome (CRISPS) [MIM:601378]. Crisponi syndrome is a rare autosomal recessive disorder characterized by congenital muscular contractions of facial muscles, with trismus in response to stimuli, dysmorphic features, bilateral camptodactyly, major feeding and respiratory difficulties, and access of hyperthermia leading to death in the first months of life.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	CRLF1 is a 422 amino acid secreted protein that interacts with cells expressing ciliary neurotrophic factor receptors. A cytokine receptor subunit belonging to the type I cytokine receptor family and type 3 subfamily, CRLF1 is thought to play a role in fetal nervous system development and immunity. CRLF1 is highly expressed in stomach, placenta, heart, ovary, thyroid, bone marrow, appendix, lymph node, spleen, thymus and fetal lung, and promotes neuronal cell survival. CRLF3 is a 442 amino acid protein that contains one fibronectin type-III domain. CRLF3 is expressed in lesion actinic keratosis (AK) and skin and squamous cell carcinoma (SCC), and is thought to negatively regulate the G0/G1 phase of the cell cycle.

Additional Information

Gene ID	9244
Other Names	CRLF1_HUMAN; CRLF1; Cytokine-like factor 1 (CLF-1); ZcytoR5; CRLF1_MOUSE; Cytokine receptor-like molecule 3 (CRLM-3); Novel cytokine receptor 6 (NR6); CrIm3; cytokine receptor like factor 1; cold-induced sweating syndrome
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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	CRLF1 (HGNC:2364)
Function	Functions as secretory protein carrier chaperone that promotes cellular release (PubMed: 10966616). Also functions as a cytokine subunit within the CRLF1-CLCF1 heterodimer, which engages the CNTF receptor complex (CNTFR, IL6ST/gp130, LIFR) (PubMed: 10966616). As part of the CRLF-CLCF1 complex, binds to CNTFR, induces dimerization of the IL6ST/gp130 and LIFR, which activates JAK tyrosine kinases (JAK1 or JAK2 and to lesser extent TYK2) bound to their intracellular domains (PubMed: 11294841). These kinases subsequently phosphorylate IL6ST/gp130 and LIFR (PubMed: 11294841). The tyrosine phosphorylated signaling receptors serve in turn as docking sites for recruitment and activation of signal transducer and activators of transcription (STAT3 and to lesser extent STAT1) (PubMed: 10966616 , PubMed: 11294841). Moreover, CLCF1/CLF heterodimer induces tyrosine phosphorylation of PTPN11, that associates with IL6ST/gp130-LIFR, which in turn recruits PIK3 protein kinase followed by AKT protein phosphorylation (PubMed: 11294841). Participates in the survival of embryonic motor neurons (PubMed: 10966616). May also play a regulatory role in the immune system (PubMed: 9686600).
Cellular Location	Secreted
Tissue Location	Highest levels of expression observed in spleen, thymus, lymph node, appendix, bone marrow, stomach, placenta, heart, thyroid and ovary. Strongly expressed also in fetal lung

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