

RNF135 Rabbit pAb

Catalog # AP97360

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	Q8IUD6
Other Accession	Q8IUD6
Predicted	Rat, Human, Mouse, Dog
Host	Rabbit
Clonality	Polyclonal
Calculated MW	47888
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human RNF135
Epitope Specificity	288-360/432
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 1 B30.2/SPRY domain.Contains 1 RING-type zinc finger.
SUBUNIT	Interacts with DDX58. Interacts with PCBP2.
Post-translational modifications	Defects in RNF135 are the cause of macrocephalymacrosomia facial dysmorphism syndrome (MMFD) [MIM:614192]. MMFD is an autosomal dominant disorder characterized by the association of macrothrombocytopathy and progressive sensorineural hearing loss without renal dysfunction.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The RING-type zinc finger motif is present in a number of viral and eukaryotic proteins and is made of a conserved cysteine-rich domain that is able to bind two zinc atoms. Proteins that contain this conserved domain are generally involved in the ubiquitination pathway of protein degradation. RNF135 (RING finger protein 135), also known as L13, is a 432 amino acid protein that contains one RING-type zinc finger and one SPRY domain. Via its RING-type zinc finger, RNF135 may play a role in transcriptional regulation and protein degradation events. Defects in the gene encoding RNF135 are the cause of RNF135-related overgrowth syndrome which is characterized by learning disabilities, facial dysmorphism and increased weight and height. Multiple isoforms of RNF135 exist due to alternative splicing events.

Additional Information

Gene ID	84282
Other Names	L13; MMFD; REUL; Riplet; 0610037N03Rik; 2410006N06Rik; U 2-3-0; RN135_HUMAN; RNF135; RIG-I E3 ubiquitin ligase (REUL); RING finger protein 135; RING finger protein leading to RIG-I activation (Riplet); RING-type E3 ubiquitin transferase RNF135; 2.3.2.27; RN135_MOUSE

Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	RNF135 (HGNC:21158)
Function	E2-dependent E3 ubiquitin-protein ligase that functions as a RIGI coreceptor in the sensing of viral RNAs in cell cytoplasm and the activation of the antiviral innate immune response (PubMed: 19017631 , PubMed: 19484123 , PubMed: 21147464 , PubMed: 23950712 , PubMed: 28469175 , PubMed: 31006531). Together with the UBE2D3, UBE2N and UB2V1 E2 ligases, catalyzes the 'Lys-63'-linked polyubiquitination of RIGI oligomerized on viral RNAs, an essential step in the activation of the RIG-I signaling pathway (PubMed: 19017631 , PubMed: 21147464 , PubMed: 28469175 , PubMed: 31006531). Through a ubiquitin-independent parallel mechanism, which consists in bridging RIGI filaments forming on longer viral RNAs, further activates the RIG-I signaling pathway (PubMed: 31006531). This second mechanism that synergizes with the ubiquitin-dependent one would thereby allow an RNA length-dependent regulation of the RIG-I signaling pathway (Probable). Associated with the E2 ligase UBE2N, also constitutively synthesizes unanchored 'Lys-63'-linked polyubiquitin chains that may also activate the RIG-I signaling pathway (PubMed: 28469175 , PubMed: 31006531).
Cellular Location	Cytoplasm. Cytoplasm, Stress granule
Tissue Location	Expressed in skeletal muscle, spleen, kidney, placenta, prostate, stomach, thyroid and tongue. Also weakly expressed in heart, thymus, liver and lung.

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