

Product datasheet

Anti-RPGRIP1L antibody ab91467

1 图像

概述

产品名称	Anti-RPGRIP1L 抗体
描述	兔多克隆抗体 to RPGRIP1L
经测试应用	适用于: WB, ELISA
种属反应性	与反应: Human
免疫原	Synthetic peptide selected from the N-terminal region of Human RPGRIP1L conjugated to KLH (NP_056087.2).
阳性对照	293 cell line lysates.

性能

形式	Liquid
存放说明	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
存储溶液	Preservative: 0.09% Sodium Azide Constituents: PBS
纯度	Immunogen affinity purified
纯化说明	ab91467 is purified through a protein A column, followed by peptide affinity purification.
克隆	多克隆
同种型	IgG

应用

Our [Abpromise guarantee](#) covers the use of **ab91467** in the following tested applications.

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

应用	Ab评论	说明
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WB

ELISA

应用说明	ELISA: 1/1000. WB: 1/100 - 1/500. Predicted molecular weight: 151 kDa.
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Not yet tested in other applications.

Optimal dilutions/concentrations should be determined by the end user.

靶标

功能

Negatively regulates signaling through the G-protein coupled thromboxane A2 receptor (TBXA2R). May be involved in mechanisms like programmed cell death, craniofacial development, patterning of the limbs, and formation of the left-right axis (By similarity). Involved in the organization of apical junctions in kidney cells together with NPHP1 and NPHP4 (By similarity). Does not seem to be strictly required for ciliogenesis.

组织特异性

Ubiquitously expressed with relatively high level of expression in hypothalamus and islet. During early development, expressed in multiple organs including brain, eye, forelimb and kidney.

疾病相关

Ciliary dysfunction leads to a broad spectrum of disorders, collectively termed ciliopathies. Overlapping clinical features include retinal degeneration, renal cystic disease, skeletal abnormalities, fibrosis of various organ, and a complex range of anatomical and functional defects of the central and peripheral nervous system. The ciliopathy range of diseases includes Meckel-Gruber syndrome, Bardet-Biedl syndrome, Joubert syndrome, nephronophthisis, Senior-Loken syndrome, and Jeune asphyxiating thoracic dystrophy among others. Single-locus allelism is insufficient to explain the variable penetrance and expressivity of such disorders, leading to the suggestion that variations across multiple sites of the ciliary proteome, including RPGRIP1L, influence the clinical outcome.

Joubert syndrome 7

Meckel syndrome 5

COACH syndrome

序列相似性

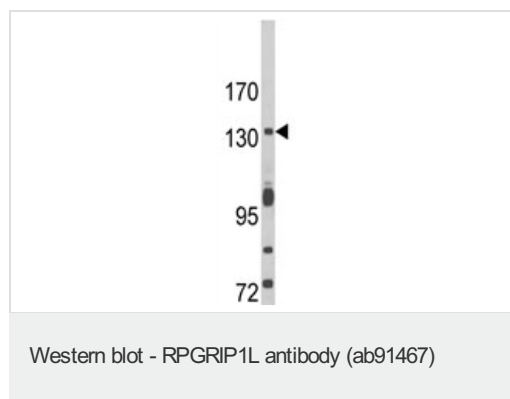
Belongs to the RPGRIP1 family.

Contains 2 C2 domains.

细胞定位

Cytoplasm. Cytoplasm > cytoskeleton > cilium basal body. Cytoplasm > cytoskeleton > cilium axoneme. Cytoplasm > cytoskeleton > microtubule organizing center > centrosome. Cell junction > tight junction. In cultured renal cells, it localizes diffusely in the cytoplasm but, as cells approach confluence, it accumulates to basolateral tight junctions.

图片



Anti-RPGRIP1L antibody (ab91467) at 1/100 dilution + 293 cell line lysates at 35 µg

Predicted band size : 151 kDa

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