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Polycystin 1 Rabbit pAb

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Catalog # AP52240

Product Information

Primary Accession	P98161
Reactivity	Human
Predicted	Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	462529
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Polycystin 1
Epitope Specificity	131-230/4303
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Membrane; Multi-pass membrane protein. Cell projection, cilium. Note=PKD1 localization to the plasma and ciliary membranes requires PKD2, is independent of PKD2 channel activity, and involves stimulation of PKD1 autoproteolytic cleavage at the GPS domain.
DISEASE	Defects in PKD1 are the cause of polycystic kidney disease autosomal dominant type 1 (ADPKD1) [MIM:173900]. ADPKD is characterized by progressive formation and enlargement of cysts in both kidneys, typically leading to end-stage renal disease in adult life. Cysts also occurs in the liver and other organs. Its prevalence is estimated at about 1/1000.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a member of the polycystin protein family. The encoded glycoprotein contains a large N-terminal extracellular region, multiple transmembrane domains and a cytoplasmic C-tail. It is an integral membrane protein that functions as a regulator of calcium permeable cation channels and intracellular calcium homoeostasis. It is also involved in cell-cell/matrix interactions and may modulate G-protein-coupled signal-transduction pathways. It plays a role in renal tubular development, and mutations in this gene cause autosomal dominant polycystic kidney disease type 1 (ADPKD1). ADPKD1 is characterized by the growth of fluid-filled cysts that replace normal renal tissue and result in end-stage renal failure. Splice variants encoding different isoforms have been noted for this gene. Also, six pseudogenes, closely linked in a known duplicated region on chromosome 16p, have been described. [provided by RefSeq].

Additional Information

Gene ID	5310

Other Names	Polycystin-1, PC1, Autosomal dominant polycystic kidney disease 1 protein, PKD1 (HGNC:9008)
Dilution	Flow-Cyt=1 μg/Test
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	PKD1 (HGNC:9008)
Function	Component of a heteromeric calcium-permeable ion channel formed by PKD1 and PKD2 that is activated by interaction between PKD1 and a Wnt family member, such as WNT3A and WNT9B (PubMed:27214281). Both PKD1 and PKD2 are required for channel activity (PubMed:27214281). Involved in renal tubulogenesis (PubMed:12482949). Involved in fluid- flow mechanosensation by the primary cilium in renal epithelium (By similarity). Acts as a regulator of cilium length, together with PKD2 (By similarity). The dynamic control of cilium length is essential in the regulation of mechanotransductive signaling (By similarity). The cilium length response creates a negative feedback loop whereby fluid shear-mediated deflection of the primary cilium, which decreases intracellular cAMP, leads to cilium shortening and thus decreases flow- induced signaling (By similarity). May be an ion-channel regulator. Involved in adhesive protein-protein and protein-carbohydrate interactions. Likely to be involved with polycystin-1-interacting protein 1 in the detection, sequestration and exocytosis of senescent mitochondria (PubMed:37681898).
Cellular Location	Cell membrane; Multi-pass membrane protein. Cell projection, cilium {ECO:0000250|UniProtKB:O08852}. Endoplasmic reticulum {ECO:0000250|UniProtKB:O08852}. Golgi apparatus {ECO:0000250|UniProtKB:O08852}. Vesicle Secreted, extracellular exosome Note=PKD1 localization to the plasma and ciliary membranes requires PKD2, is independent of PKD2 channel activity, and involves stimulation of PKD1 autoproteolytic cleavage at the GPS region of the GAIN-B domain. PKD1:PKD2 interaction is required to reach the Golgi apparatus from endoplasmic reticulum and then traffic to the cilia (By similarity). Ciliary localization of PKD1 requires BBS1 and ARL6/BBS3 (By similarity). Cell surface localization requires GANAB (PubMed:27259053). Detected on migrasomes and on extracellular exosomes in urine (PubMed:37681898). {ECO:0000250|UniProtKB:O08852, ECO:0000269|PubMed:27259053, ECO:0000269|PubMed:37681898}

Background

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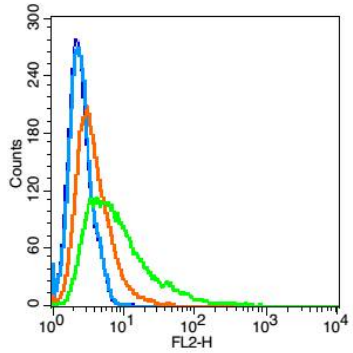
References

Gluecksmann-Kuis M.A.,et al.Cell 81:289-298(1995).
Hughes J.,et al.Nat. Genet. 10:151-160(1995).
Martin J.,et al.Nature 432:988-994(2004).
Ward C.J.,et al.Cell 77:881-894(1994).

Ward C.J.,et al.Cell 78:725-725(1994).

Images



Blank control(blue): Hela(fixed with 2% paraformaldehyde(10 min)).
Primary Antibody:Rabbit Anti-Polycystin 1 antibody(AP52240), Dilution: 1 µg in 100 µL 1X PBS containing 0.5% BSA;
Isotype Control Antibody: Rabbit IgG(orange) ,used under the same conditions); Secondary Antibody: Goat anti-rabbit IgG-PE(white blue), Dilution: 1:200 in 1 X PBS containing 0.5% BSA.