

Product datasheet

info@arigobio.com

ARG40530 anti-NPHP1 / Nephrocystin 1 antibody

Package: 100 μl Store at: -20°C

Summary

Product Description Rabbit Polyclonal antibody recognizes NPHP1 / Nephrocystin 1

Tested Reactivity Hu, Ms, Rat

Tested Application ICC/IF, IHC-P, WB

Host Rabbit

Clonality Polyclonal

Isotype IgG

Target Name NPHP1 / Nephrocystin 1

Species Human

Immunogen Recombinant fusion protein corresponding to aa. 345-614 of Human NPHP1 / Nephrocystin 1

(NP_001121651.1).

Conjugation Un-conjugated

Alternate Names JBTS4; SLSN1; Juvenile nephronophthisis 1 protein; Nephrocystin-1; NPH1

Application Instructions

Application table	Application	Dilution
	ICC/IF	1:50 - 1:200
	IHC-P	1:50 - 1:200
	WB	1:500 - 1:2000
Application Note	* The dilutions indicate recommended starting dilutions and the optimal dilutions or concentrations should be determined by the scientist.	
Positive Control	Mouse lung	
Observed Size	80 kDa	

Properties

Form Liquid

Purification Affinity purified.

Buffer PBS (pH 7.3), 0.02% Sodium azide and 50% Glycerol.

Preservative 0.02% Sodium azide

Stabilizer 50% Glycerol

Storage instruction For continuous use, store undiluted antibody at 2-8°C for up to a week. For long-term storage, aliquot

and store at -20°C. Storage in frost free freezers is not recommended. Avoid repeated freeze/thaw cycles. Suggest spin the vial prior to opening. The antibody solution should be gently mixed before use.

Bioinformation

Gene Symbol

NPHP1

Gene Full Name

nephronophthisis 1 (juvenile)

Background

This gene encodes a protein with src homology domain 3 (SH3) patterns. This protein interacts with Crkassociated substrate, and it appears to function in the control of cell division, as well as in cell-cell and cell-matrix adhesion signaling, likely as part of a multifunctional complex localized in actin- and microtubule-based structures. Mutations in this gene cause familial juvenile nephronophthisis type 1, a kidney disorder involving both tubules and glomeruli. Defects in this gene are also associated with Senior-Loken syndrome type 1, also referred to as juvenile nephronophthisis with Leber amaurosis, which is characterized by kidney and eye disease, and with Joubert syndrome type 4, which is characterized by cerebellar ataxia, oculomotor apraxia, psychomotor delay and neonatal breathing abnormalities, sometimes including retinal dystrophy and renal disease. Multiple transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Jul 2008]

Function

Together with BCAR1 it may play a role in the control of epithelial cell polarity. Involved in the organization of apical junctions in kidney cells together with NPHP4 and RPGRIP1L/NPHP8 (By similarity). Does not seem to be strictly required for ciliogenesis (By similarity). Seems to help to recruit PTK2B/PYK2 to cell matrix adhesions, thereby initiating phosphorylation of PTK2B/PYK2 and PTK2B/PYK2-dependent signaling. May play a role in the regulation of intraflagellar transport (IFT) during cilia assembly. Required for normal retina development. In connecting photoreceptor cilia influences the movement of some IFT proteins such as IFT88 and WDR19. Involved in spermatogenesis (By similarity). [UniProt]

Calculated Mw

83 kDa

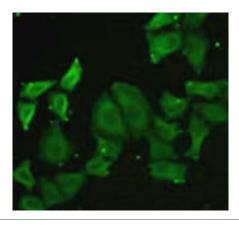
PTM

Phosphorylation by CK2 is required for the interaction with PACS1 and the targeting to the base region of cilia. [UniProt]

Cellular Localization

Cell junction, adherens junction, tight junction. Cell projection, cilium. Cytoplasm, cytoskeleton, cilium axoneme. Note=Colocalizes with E-cadherin and BCAR1 at or near the cell-cell adherens junctions. Localized to respiratory cilia axoneme. Localized to the transition zone of respiratory cilia, photoreceptor-connecting cilia and renal monocilia. [UniProt]

Images

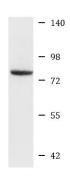


ARG40530 anti-NPHP1 / Nephrocystin 1 antibody ICC/IF image

Immunofluorescence: MCF7 cells stained with ARG40530 anti-NPHP1 / Nephrocystin 1 antibody.

ARG40530 anti-NPHP1 / Nephrocystin 1 antibody WB image

Western blot: 25 μg of Mouse lung lysate stained with ARG40530 anti-NPHP1 / Nephrocystin 1 antibody at 1:1000 dilution.



Mouse lung