

CEP290 / NPHP6 Antibody (internal region)
Peptide-affinity purified goat antibody
Catalog # AF2719a**Specification**

CEP290 / NPHP6 Antibody (internal region) - Product Information

Application	E
Primary Accession	O15078
Other Accession	NP_079390.3 , 80184
Predicted	Human
Host	Goat
Clonality	Polyclonal
Concentration	0.5 mg/ml
Isotype	IgG
Calculated MW	290386

CEP290 / NPHP6 Antibody (internal region) - Additional Information**Gene ID** 80184**Other Names**

Centrosomal protein of 290 kDa, Cep290, Bardet-Biedl syndrome 14 protein, Cancer/testis antigen 87, CT87, Nephrocystin-6, Tumor antigen se2-2, CEP290, BBS14, KIAA0373, NPHP6

Format

0.5 mg/ml in Tris saline, 0.02% sodium azide, pH7.3 with 0.5% bovine serum albumin

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions

CEP290 / NPHP6 Antibody (internal region) is for research use only and not for use in diagnostic or therapeutic procedures.

CEP290 / NPHP6 Antibody (internal region) - Protein Information**Name** CEP290**Synonyms** BBS14, KIAA0373, NPHP6**Function**

Involved in early and late steps in cilia formation. Its association with CCP110 is required for inhibition of primary cilia formation by CCP110 (PubMed:18694559). May play a role in early ciliogenesis in the disappearance of centriolar satellites and in the transition of primary ciliary vesicles (PCVs) to capped ciliary vesicles (CCVs). Required for the centrosomal

recruitment of RAB8A and for the targeting of centriole satellite proteins to centrosomes such as of PCM1 (PubMed:24421332). Required for the correct localization of ciliary and phototransduction proteins in retinal photoreceptor cells; may play a role in ciliary transport processes (By similarity). Required for efficient recruitment of RAB8A to primary cilium (PubMed:17705300). In the ciliary transition zone is part of the tectonic-like complex which is required for tissue-specific ciliogenesis and may regulate ciliary membrane composition (By similarity). Involved in regulation of the BBSome complex integrity, specifically for presence of BBS2, BBS5 and BBS8/TTC8 in the complex, and in ciliary targeting of selected BBSome cargos. May play a role in controlling entry of the BBSome complex to cilia possibly implicating IQCB1/NPHP5 (PubMed:25552655). Activates ATF4-mediated transcription (PubMed:16682973).

Cellular Location

Cytoplasm, cytoskeleton, microtubule organizing center, centrosome. Cytoplasm, cytoskeleton, microtubule organizing center, centrosome, centriolar satellite. Nucleus {ECO:0000250|UniProtKB:Q6A078} Cell projection, cilium. Cytoplasm, cytoskeleton, cilium basal body {ECO:0000250|UniProtKB:Q6A078} Cytoplasm, cytoskeleton, microtubule organizing center, centrosome, centriole. Cytoplasmic vesicle. Note=Displaced from centriolar satellites in response to cellular stress, such as ultraviolet light (UV) radiation or heat shock (PubMed:24121310). Found in the connecting cilium of photoreceptor cells, base of cilium in kidney intramedullary collecting duct cells (By similarity). Localizes at the transition zone, a region between the basal body and the ciliary axoneme (PubMed:23943788). Localization at the ciliary transition zone as well as at centriolar satellites is BBSome-dependent (PubMed:23943788) {ECO:0000250|UniProtKB:Q6A078, ECO:0000269|PubMed:23943788, ECO:0000269|PubMed:24121310}

Tissue Location

Ubiquitous. Expressed strongly in placenta and weakly in brain.

CEP290 / NPHP6 Antibody (internal region) - Protocols

Provided below are standard protocols that you may find useful for product applications.

- [Western Blot](#)
- [Blocking Peptides](#)
- [Dot Blot](#)
- [Immunohistochemistry](#)
- [Immunofluorescence](#)
- [Immunoprecipitation](#)
- [Flow Cytometry](#)
- [Cell Culture](#)

CEP290 / NPHP6 Antibody (internal region) - Images

CEP290 / NPHP6 Antibody (internal region) - References

Hypomorphic CEP290/NPHP6 mutations result in anosmia caused by the selective loss of G proteins in cilia of olfactory sensory neurons. McEwen DP, Koeneke RK, Khanna H, Jenkins PM, Lopez I, Swaroop A, Martens JR. Proc Natl Acad Sci U S A. 2007 Oct 2;104(40):15917-22. Epub 2007 Sep 26. PMID: 17898177