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CCDC40 Rabbit pAb

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Catalog # AP58867

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q4G0X9
Predicted	Rat, Pig, Dog, Human, Mouse, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	130113
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CCDC40
Epitope Specificity	851-950/1142
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	Cytoplasm. Cell projection, cilium. Note=Localizes to cytoplasm and motile cilium.
DISEASE	Defects in CCDC40 are the cause of primary ciliary dyskinesia type 15 (CILD15) [MIM:613808]. A disorder characterized by abnormalities of motile cilia. Respiratory infections leading to chronic inflammation and bronchiectasis are recurrent, due to defects in the respiratory cilia; reduced fertility is often observed in male patients due to abnormalities of sperm tails. Half of the patients exhibit randomization of left-right body asymmetry and situs inversus, due to dysfunction of monocilia at the embryonic node. Primary ciliary dyskinesia associated with situs inversus is referred to as Kartagener syndrome.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Required for assembly of dynein regulatory complex (DRC) and inner dynein arm complexes, which are responsible for ciliary beat regulation, thereby playing a central role in motility in cilia and flagella. Not required for outer dynein arm complexes assembly. Required for axonemal recruitment of CCDC39. Involvement in disease:Defects in CCDC40 are the cause of primary ciliary dyskinesia type 15 (CILD15) . A disorder characterized by abnormalities of motile cilia. Respiratory infections leading to chronic inflammation and bronchiectasis are recurrent, due to defects in the respiratory cilia; reduced fertility is often observed in male patients due to abnormalities of sperm tails. Half of the patients exhibit randomization of left-right body asymmetry and situs inversus, due to dysfunction of monocilia at the embryonic node. Primary ciliary dyskinesia associated with situs inversus is referred to as Kartagener syndrome.

Additional Information

Gene ID	55036
Other Names	Coiled-coil domain-containing protein 40, CCDC40 (HGNC:26090)
Dilution	IHC-P=1:100-500,IHC-F=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	CCDC40 (HGNC:26090)
Function	Required for assembly of dynein regulatory complex (DRC) and inner dynein arm (IDA) complexes, which are responsible for ciliary beat regulation, thereby playing a central role in motility in cilia and flagella (PubMed:21131974). Probably acts together with CCDC39 to form a molecular ruler that determines the 96 nanometer (nm) repeat length and arrangements of components in cilia and flagella (By similarity). Not required for outer dynein arm complexes assembly. Required for axonemal recruitment of CCDC39 (PubMed:21131974).
Cellular Location	Cytoplasm {ECO:0000250|UniProtKB:Q8BI79}. Cell projection, cilium. Note=Localizes to cytoplasm and motile cilium. {ECO:0000250|UniProtKB:Q8BI79}

Background

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