

CCDC39 Rabbit pAb

Catalog # AP96345

Product Information

Application	WB
Primary Accession	Q9UFE4
Other Accession	Q9UFE4
Reactivity	Human
Predicted	Rat, Mouse, Rabbit, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	109901
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CCDC39
Epitope Specificity	651-750/941
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the CCDC39 family.
DISEASE	Defects in CCDC39 are the cause of primary ciliary dyskinesia type 14 (CILD14) [MIM:613807]. 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. Tissue specificity: Mainly expressed in nasal brushings and, to a lesser extent, in lungs and testis. Involvement in disease: Defects in CCDC39 are the cause of primary ciliary dyskinesia type 14 (CILD14). 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	339829
Other Names	CFAP59; CILD14; FAP59; 4921507O14Rik; D3Erttd789e; b2b1304Clo; b2b1735Clo; b2b2025.1Clo; prh; CCD39_CANLF; CCDC39; CCD39_HUMAN; CCD39_MOUSE; coiled-coil domain 39 molecular ruler complex subunit; coiled-coil domain containing 39
Dilution	WB=1:500-2000
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	CCDC39 (HGNC:25244)
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: 21131972). Probably acts together with CCDC40 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 (PubMed: 21131972).
Cellular Location	Cytoplasm, cytoskeleton, cilium axoneme. Note=CCDC40 is required for localization to axonemes.
Tissue Location	Mainly expressed in nasal brushings and, to a lesser extent, in lungs and testis.

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