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RSPH4A Rabbit pAb

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Catalog # AP54516

Product Information

Application	WB
Primary Accession	Q5TD94
Reactivity	Human, Mouse
Predicted	Rat, Pig, Dog, Chicken, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	80733
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human RSPH4A/RSHL3
Epitope Specificity	435-482/716
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; cytoskeleton; cilium axoneme. Radial spoke.
DISEASE	Defects in RSPH4A are the cause of primary ciliary dyskinesia type 11 (CILD11) [MIM:612649]. CILD is an autosomal recessive disorder characterized by axonemal 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 situs inversus, due to dysfunction of monocilia at the embryonic node and randomization of left-right body asymmetry. 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	RSHL3 is predicted to be a component of the radial spoke head based on homology with proteins in the biflagellate alga Chlamydomonas reinhardtii and other ciliates. RSHL3 (radial spoke head-like protein 3), also known as radial spoke head protein 4 homolog A, is a 716 amino acid protein that belongs to the flagellar radial spoke RSP4/6 family. Mutations in the RSHL3 gene cause primary ciliary dyskinesia 1, a disease arising from dysmotility of motile cilia and sperm. Existing as three alternatively spliced isoforms, the RSHL3 gene contains 6 exons, is conserved in chimpanzee, dog, cow, mouse, rat, chicken, zebrafish, fruit fly and P.falciparum, and maps to human chromosome 6q22.1.

Additional Information

Gene ID	345895
Other Names	Radial spoke head protein 4 homolog A, Radial spoke head-like protein 3,

	RSPH4A, RSHL3
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.

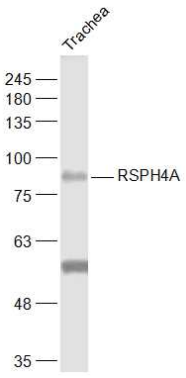
Protein Information

Name	RSPH4A
Synonyms	RSHL3
Function	Component of the axonemal radial spoke head which plays an important role in ciliary motility (PubMed:19200523). Essential for triplet radial spokes (RS1, RS2 and RS3) head assembly in the motile cilia (By similarity).
Cellular Location	Cytoplasm, cytoskeleton, cilium axoneme. Cell projection, cilium
Tissue Location	Expressed in trachea, lungs, and testes (PubMed:23993197). Very strong expression is detected in nasal brushings (PubMed:19200523).

Background

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Images



Sample:
Trachea (Mouse) Lysate at 40 ug
Primary: Anti-RSPH4A (AP54516) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 81 kD
Observed band size: 81 kD

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.