SunLong Biotech Co.,LTD
Tel: 0086-571-56623320 Fax:0086-571-56623318
E-mail:sales@sunlongbiotech.com
www.sunlongbiotech.com

[Marker Primary Antibody]

Rabbit Anti-RSPH4A/HRP Conjugated antibody

Catalog Number: SL11472R-HRP

FOR RESEARCH USE ONLY

[DATASHEET]

Host:	Rabbit
Isotype:	IgG
Clonality:	Polyclonal
Target:	Anti-RSPH4A/HRP
Immunogen:	KLH conjugated synthetic peptide derived from human RSPH4A/RSHL3 (435-482aa)
Purification:	affinity purified by Protein A
Concentration:	1mg/ml
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Applications:	WB=1:500-2000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Reactivity:	Human,Mouse(predicted:Rat,Chicken,Dog,Pig,Cow,Horse,Sheep)
Background:	background: 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. **Function:** Probable component of the axonemal radial spoke head. Radial spokes are regularly spaced along cilia, sperm and flagella axonemes. They consist of a thin stalk which is attached to a subfiber of the outer doublet microtubule, and a bulbous head which is attached to the stalk and appears to

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	interact with the projections from the central pair of microtubules. **Subcellular Location:** Cytoplasm; cytoskeleton; cilium axoneme. Radial spoke. **Tissue Specificity:** 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. 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. **Similarity:** Belongs to the flagellar radial spoke RSP4/6 family. **Database links:** **Entrez Gene:** 345895 Human Omim: 612647 Human **SwissProt:** Q5TD94 Human **Unigene:** 160380 Human Important **Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. KartagenerSyndrome: composed of the following triad，bronchiectasis, sinusitis or nasal polyps, and situs inversus (mainly dextrocardia)。If only situs inversus and bronchiectasis are present，then it is incomplete Kartagener syndrome. It is often combined with other congenital malformations. Its etiology is due to congenital deficiency of the axonemal arms of cilia throughout the body, causing loss of ciliary motility, mucociliary transport dysfunction, and persistent infection due to retention of secretions and bacteria. It occurs mostly in school-age children and adolescents, and there is a family history. The main symptoms are cough, sputum production, and hemoptysis that worsen with age, more obvious in the morning, and

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	susceptibility to colds and pneumonia. Common signs are cyanosis and clubbing of the fingers.

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