

Rabbit Anti-CCDC40 antibody

SL8091R

CCDC40
卷曲螺旋结构域蛋白40抗体
CCD40 HUMAN; ccdc 40; CCDC40; Coiled coil domain containing 40; coiled-coil
domain containing 40; Coiled-coil domain-containing protein 40.
Rabbit
Polyclonal
Human, Mouse, Rat, Dog, Pig, Cow, Horse, Sheep,
ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500 (Paraffin sections
need antigen repair)
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.
130kDa
cytoplasmic
Lyophilized or Liquid
1mg/ml
KLH conjugated synthetic peptide derived from human CCDC40:851-950/1142
IgG
affinity purified by Protein A
0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
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 0.01M PBS or diluent of
antibody the antibody is stable for at least two weeks at 2-4 °C.
<u>PubMed</u>
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.

Function:

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.

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.

Similarity:

Belongs to the CCDC40 family.

SWISS: Q4G0X9

Gene ID: 55036

Database links:

Entrez Gene: 55036Human

Omim: 613799Human

SwissProt: Q4G0X9Human

Unigene: 202542Human

Important Note:

This product as supplied is intended for research use only, not for use in human,
therapeutic or diagnostic applications.

