

MTND4L Rabbit pAb

Catalog # AP98344

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	P03901
Other Accession	P03901
Host	Rabbit
Clonality	Polyclonal
Calculated MW	10741
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human MTND4L
Epitope Specificity	1-50/98
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 complex I subunit 4L family.
DISEASE	Defects in MT-ND4L are a cause of Leber hereditary optic neuropathy (LHON) [MIM:535000]. LHON is a maternally inherited disease resulting in acute or subacute loss of central vision, due to optic nerve dysfunction. Cardiac conduction defects and neurological defects have also been described in some patients. LHON results from primary mitochondrial DNA mutations affecting the respiratory chain complexes.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	NADH:ubiquinone oxidoreductase (complex I) is an extremely complicated multiprotein complex located in the inner mitochondrial membrane. Human complex I is important for energy metabolism because its main function is to transport electrons from NADH to ubiquinone, which is accompanied by translocation of protons from the mitochondrial matrix to the intermembrane space. Human complex I appears to consist of 41 subunits. A small number of complex I subunits are the products of mitochondrial genes (subunits 1-7), while the remainder are nuclear encoded and imported from the cytoplasm. NADH dehydrogenase subunit 4L (ND4L) is most likely a component of the hydrophobic protein fragment of Complex I. Mutations in the gene encoding for ND4 are implicated in Leber hereditary optic neuropathy, a rare condition that can cause loss of central vision.

Additional Information

Gene ID	4539
Other Names	MTND4L; NU4LM_HUMAN; MT-ND4L; NADH dehydrogenase subunit 4L (ND4L); 7.1.1.2; NADH4L; ND4L; mitochondrially encoded NADH:ubiquinone oxidoreductase core subunit 4L; NADH dehydrogenase 4L; mitochondrially encoded NADH dehydrogenase 4L; complex I ND4L subunit; NADH-ubiquinone oxidoreductase chain 4L

Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:500 0-10000
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	MT-ND4L (HGNC:7460)
Synonyms	MTND4L, NADH4L, ND4L
Function	Core subunit of the mitochondrial membrane respiratory chain NADH dehydrogenase (Complex I) which catalyzes electron transfer from NADH through the respiratory chain, using ubiquinone as an electron acceptor (PubMed: 28844695). Part of the enzyme membrane arm which is embedded in the lipid bilayer and involved in proton translocation (PubMed: 28844695).
Cellular Location	Mitochondrion inner membrane {ECO:0000250 UniProtKB:P03902}; Multi-pass membrane protein

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