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MT-ND6 Rabbit pAb

Catalog # AP98496

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P03923
Other Accession	P03923
Reactivity	Rat, Human
Predicted	Mouse, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	18622
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human MTND6
Epitope Specificity	51-150/174
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 6 family.
DISEASE	Defects in MT-ND6 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. Defects in MT-ND6 are a cause of Leber hereditary optic neuropathy with dystonia (LDYT) [MIM:500001]; also called familial dystonia with visual failure and striatal lucencies. LDYT is part of a spectrum of Leber hereditary optic neuropathy. It is characterized by the association of optic atrophy and central vision loss with dystonia. Defects in MT-ND6 are a cause of mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes syndrome (MELAS) [MIM:540000]. MELAS is a genetically heterogenious disorder, characterized by episodic vomiting, seizures, and recurrent cerebral insults resembling strokes and causing hemiparesis, hemianopsia, or cortical blindness. Defects in MT-ND6 are a cause of mitochondrial complex I deficiency (MT-C1D) [MIM:252010]. A disorder of the mitochondrial respiratory chain that causes a wide range of clinical manifestations from lethal neonatal disease to adult-onset neurodegenerative disorders. Phenotypes include macrocephaly with progressive leukodystrophy, non-specific encephalopathy, cardiomyopathy, myopathy, liver disease, Leigh syndrome, Leber hereditary optic neuropathy, and some forms of Parkinson disease.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	NADH Dehydrogenase subunit 6 (MTND6) is 1 of the 7 mitochondrial DNA (mtDNA) encoded subunits (MTND1, MTND2, MTND3, MTND4L, MTND4, MTND5, MTND6) included among the approximately 41 polypeptides of respiratory Complex I. Complex I accepts electrons from NADH, transfers them to ubiquinone (Coenzyme Q10), and uses the energy released to pump protons across the mitochondria inner membrane. MTND6 has been

proposed to be a component of the iron-protein fragment.

Additional Information

Gene ID	4541
Other Names	MTND6; NU6M_HUMAN; MT-ND6; NADH dehydrogenase subunit 6; 7.1.1.2; NADH6; ND6; NU6M_PIG; mitochondrially encoded NADH:ubiquinone oxidoreductase core subunit 6; NADH dehydrogenase 6; mitochondrially encoded NADH dehydrogenase 6; complex I ND6 subunit; NADH-ubiquinone oxidoreductase chain 6
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,Flow-Cyt=1ug/Test
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-ND6
Synonyms	MTND6, NADH6, ND6
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:14595656, PubMed:8644732). Essential for the catalytic activity and assembly of complex I (PubMed:14595656, PubMed:8644732).
Cellular Location	Mitochondrion inner membrane {ECO:0000250|UniProtKB:P03924}; Multi-pass membrane protein

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