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NDUFV1 Rabbit pAb

Catalog # AP98503

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P49821
Other Accession	P49821
Reactivity	Mouse
Predicted	Rat, Pig, Human, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	50817
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human NDUFV1
Epitope Specificity	41-140/464
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 51 kDa subunit family.
SUBUNIT	Complex I is composed of 45 different subunits. This is a component of the flavoprotein-sulfur (FP) fragment of the enzyme.
DISEASE	Defects in NDUFV1 are a cause of Leigh syndrome (LS) [MIM:256000]. LS is a severe neurological disorder characterized by bilaterally symmetrical necrotic lesions in subcortical brain regions.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The mitochondrial respiratory chain provides energy to cells via oxidative phosphorylation and consists of four membrane-bound electron-transporting protein complexes (I-IV) and an ATP synthase (complex V). This gene encodes a 51 kDa subunit of the NADH:ubiquinone oxidoreductase complex I; a large complex with at least 45 nuclear and mitochondrial encoded subunits that liberates electrons from NADH and channels them to ubiquinone. This subunit carries the NADH-binding site as well as flavin mononucleotide (FMN)- and Fe-S-biding sites. Defects in complex I are a common cause of mitochondrial dysfunction; a syndrome that occurs in approximately 1 in 10,000 live births. Mitochondrial complex I deficiency is linked to myopathies, encephalomyopathies, and neurodegenerative disorders such as Parkinson's disease and Leigh syndrome. Alternative splicing results in multiple transcript variants encoding distinct isoforms.[provided by RefSeq, Oct 2009]

Additional Information

Gene ID	4723
Other Names	CI-51K; CI51KD; MC1DN4; UQOR1; CI-51kD; NDUV1_BOVIN; NDUFV1; Complex I-51kD (CI-51kD); NADH dehydrogenase flavoprotein 1; NADH-ubiquinone oxidoreductase 51 kDa subunit; 7.1.1.2; NDUV1_HUMAN; NDUV1_MOUSE;

	Q5XIH3_RAT; NADH:ubiquinone oxidoreductase core subunit V1; NADH dehydrogenase (ubiquinone) flavoprotein 1 (51kD); NADH dehydrogenase (ubiquinone) flavoprotein 1, 51kDa; complex I 51kDa subunit; NADH dehydrogenase [ubiquinone] flavoprotein 1, mitochondrial
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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	NDUFV1 (HGNC:7716)
Synonyms	UQOR1
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 peripheral arm of the enzyme, where the electrons from NADH are accepted by flavin mononucleotide (FMN) and then passed along a chain of iron-sulfur clusters by electron tunnelling to the final acceptor ubiquinone (PubMed:28844695). Contains FMN, which is the initial electron acceptor as well as one iron-sulfur cluster (PubMed:28844695).
Cellular Location	Mitochondrion inner membrane {ECO:0000250|UniProtKB:P25708}; Peripheral membrane protein {ECO:0000250|UniProtKB:P25708}; Matrix side {ECO:0000250|UniProtKB:P25708}

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