

Mimitin Rabbit pAb

Catalog # AP98791

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	Q8N183
Other Accession	Q8N183
Host	Rabbit
Clonality	Polyclonal
Calculated MW	19856
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Mimitin
Epitope Specificity	71-169/169
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 NDUFA12 subunit family.
SUBUNIT	Mitochondrion.
DISEASE	Defects in NDUFAF2 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 disorders, 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:ubiquinone oxidoreductase (complex I) catalyzes the transfer of electrons from NADH to ubiquinone (coenzyme Q) in the first step of the mitochondrial respiratory chain, resulting in the translocation of protons across the inner mitochondrial membrane. This gene encodes a complex I assembly factor. Mutations in this gene cause progressive encephalopathy resulting from mitochondrial complex I deficiency.

Additional Information

Gene ID	91942
Other Names	B17.2L; MC1DN10; MMTN; NDUFA12L; mimitin; 1810058I14Rik; RGD1560158; NDUF2_HUMAN; NDUFAF2; B17.2-like (B17.2L); Myc-induced mitochondrial protein (MMTN); NDUFA12-like protein; NDUF2_MOUSE; NADH:ubiquinone oxidoreductase complex assembly factor 2; NDUFA12-like; NADH dehydrogenase (ubiquinone) 1 alpha subcomplex, assembly factor 2; NADH dehydrogenase (ubiquinone) complex I, assembly factor 2; Myc-induced mitochondrial protein
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-

500,ELISA=1:5000-10000

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 NDUFAF2

Synonyms NDUFA12L

Function Acts as a molecular chaperone for mitochondrial complex I assembly (PubMed:[16200211](#), PubMed:[19384974](#)). Complex I functions in the transfer of electrons from NADH to the respiratory chain. The immediate electron acceptor for the enzyme is believed to be ubiquinone (PubMed:[16200211](#), PubMed:[27626371](#)). Is involved in the initial steps of cilia formation, including removal of CP110 from the mother centrioles, docking of membrane vesicles to the mother centrioles, and establishment of the transition zone (PubMed:[38949024](#)).

Cellular Location Mitochondrion.

Tissue Location Highly expressed in ESCC cells. Also expressed in heart, skeletal muscle, liver, and in fibroblasts

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