

NDUFAF6 Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP57384

Product Information

Application IHC-P, IHC-F, IF, ICC, E

Primary Accession

Reactivity
Rat

Host
Clonality
Polyclonal
Calculated MW
Physical State

Q330K2
Rat
Rabbit
Polyclonal
S8176
Liquid

Immunogen KLH conjugated synthetic peptide derived from human NDUFAF6

Epitope Specificity 201-300/333

Isotype IgG

Purity affinity purified by Protein A

Buffer 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.

SUBCELLULAR LOCATION Isoform 1: Mitochondrion inner membrane. Note: Peripherally localized on

the matrix face of the mitochondrial inner membrane. Ref.7 Isoform 2:

Cytoplasm. Nucleus

SIMILARITY Belongs to the NDUFAF6 family.

DISEASE 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. Note: The disease is caused by mutations

affecting the gene represented in this entry.

Important Note This product as supplied is intended for research use only, not for use in

human, therapeutic or diagnostic applications.

Background Descriptions This gene encodes a protein that localizes to mitochondria and contains a

predicted phytoene synthase domain. The encoded protein plays an important role in the assembly of complex I (NADH-ubiquinone

oxidoreductase) of the mitochondrial respiratory chain through regulation of subunit ND1 biogenesis. Mutations in this gene are associated with complex I

enzymatic deficiency. [provided by RefSeq, Nov 2011]

Additional Information

Gene ID 137682

Other Names NADH dehydrogenase (ubiquinone) complex I, assembly factor 6, Putative

phytoene synthase, NDUFAF6, C8orf38

Dilution IHC-P=1:100-500,IHC-F=1:100-500,ICC=1:100-500,IF=1:100-500,ELISA=1:5000-

10000

Format 0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce

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 NDUFAF6

Synonyms C8orf38

Function Involved in the assembly of mitochondrial NADH:ubiquinone oxidoreductase

complex (complex I) at early stages. May play a role in the biogenesis of

complex I subunit MT-ND1.

Cellular Location [Isoform 1]: Mitochondrion inner membrane. Note=Peripherally localized on

the matrix face of the mitochondrial inner membrane

Tissue Location Widely expressed. A lower expression is observed in lung and kidney

compared to heart, muscle and liver (PubMed:27466185) In the kidney,

expression is high in the basal zone of the proximal tubular cells

(PubMed:27466185).

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