

NDUFS1 Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP58106

Product Information

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

Primary Accession
Reactivity
Rat, Pig, Dog
Host
Clonality
Polyclonal
Calculated MW
Physical State
P28331
Rat, Pig, Dog
Rabbit
Polyclonal
T9468
Liquid

Immunogen KLH conjugated synthetic peptide derived from human Ndufs1

Epitope Specificity 51-150/727 **Isotype** IgG

Purity affinity purified by Protein A

SUBCELLULAR LOCATION

SIMILARITY

Buffer

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

Mitochondrion inner membrane.

Defects in NDUFS1 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 Ndufs1 belongs to the complex I 75 kDa subunit family. Mammalian complex

I is composed of 45 different subunits. It locates at the mitochondrial inner membrane. Ndufs1 has NADH dehydrogenase activity and oxidoreductase activity. It transfers electrons from NADH to the respiratory chain. The immediate electron acceptor for the enzyme is believed to be ubiquinone. This protein is the largest subunit of complex I and it is a component of the iron-sulfur (IP) fragment of the enzyme. It may form part of the active site crevice where NADH is oxidized. Mutations in Ndufs1 gene are associated

with complex I deficiency.

Additional Information

Gene ID 4719

Other Names NADH-ubiquinone oxidoreductase 75 kDa subunit, mitochondrial, 7.1.1.2,

Complex I-75kD, CI-75kD, NDUFS1

Dilution WB=1:500-2000,IHC-P=1:100-500,IHC-F=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 NDUFS1

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:30879903, PubMed:31557978). Essential for catalysing the entry and efficient transfer of electrons within complex I (PubMed:31557978). Plays a key role in the assembly and stability of complex I and participates in the association of complex I with ubiquinol-cytochrome reductase complex

(Complex III) to form supercomplexes (PubMed: 30879903,

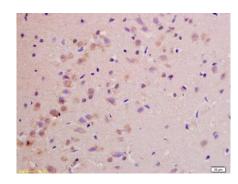
PubMed:31557978).

Cellular Location Mitochondrion inner membrane; Peripheral membrane protein

{ECO:0000250|UniProtKB:P15690}; Matrix side

{ECO:0000250 | UniProtKB:P15690}

Images



Tissue/cell: rat brain tissue; 4% Paraformaldehyde-fixed and paraffin-embedded;

Antigen retrieval: citrate buffer (0.01M, pH 6.0), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum, C-0005) at 37°C for 20 min;

Incubation: Anti-NDUFS1/NADH dehydrogenase (ubiquinone) FeS protein 1(75kD) Polyclonal Antibody, Unconjugated(AP58106) 1:200, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining

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