



NDUV1 Polyclonal Antibody

Catalog No	BYab-05804
Isotype	IgG
Reactivity	Human;Mouse
Applications	WB;ELISA
Gene Name	NDUFV1 UQOR1
Protein Name	NADH dehydrogenase [ubiquinone] flavoprotein 1, mitochondrial (EC 1.6.5.3) (EC 1.6.99.3) (Complex I-51kD) (CI-51kD) (NADH dehydrogenase flavoprotein 1) (NADH-ubiquinone oxidoreductase 51 kDa subunit)
Immunogen	Synthesized peptide derived from human protein . at AA range: 170-250
Specificity	NDUV1 Polyclonal Antibody detects endogenous levels of protein.
Formulation	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000 ELISA 1:5000-20000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	51kD
Cell Pathway	Mitochondrion inner membrane ; Peripheral membrane protein ; Matrix side .
Tissue Specificity	Brain,Eye,Kidney,Pituitary,
Function	catalytic activity:NADH + acceptor = NAD(+) + reduced acceptor.,catalytic activity:NADH + ubiquinone = NAD(+) + ubiquinol.,cofactor: Binds 1 4Fe-4S cluster .,cofactor: Binds 1 FMN .,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.,disease: Defects in NDUFV1 are a cause of mitochondrial complex I deficiency [MIM:252010]. Complex I (NADH-ubiquinone oxidoreductase), the largest complex of the mitochondrial respiratory chain, contains more than 40 subunits. It is embedded in the inner mitochondrial membrane and is partly protruding in the matrix. Complex I deficiency is the most common cause of mitochondrial disorders. It represents largely one-third of all cases of respiratory

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chain deficiency and is responsible for a variety of clinical symptom

Background

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-binding 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 trans

matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images

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