

Product name:	NDUFS7 Rabbit Polyclonal Antibody
Cat number:	ABN14519
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human NDUFS7. AA range:164-213
Reactivity:	Human,Mouse,Rat
Applications:	IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:20000-1:40000
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

This gene encodes a protein that is a subunit of one of the complexes that forms the mitochondrial respiratory chain. This protein is one of over 40 subunits found in complex I, the nicotinamide adenine dinucleotide (NADH):ubiquinone oxidoreductase. This complex functions in the transfer of electrons from NADH to the respiratory chain, and ubiquinone is believed to be the immediate electron acceptor for the enzyme. Mutations in this gene cause Leigh syndrome due to mitochondrial complex I deficiency, a severe neurological disorder that results in bilaterally symmetrical necrotic lesions in subcortical brain regions. [provided by RefSeq, Jul 2008],catalytic activity:NADH + acceptor = NAD(+) + reduced acceptor.,catalytic activity:NADH + ubiquinone = NAD(+) + ubiquinol.,cofactor: Binds 1 4Fe-4S cluster .,disease: Defects in NDUFS7 are a cause of complex I mitochondrial respiratory chain 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 chain deficiency and is responsible for a variety of clinical symptoms, ranging from neurological disorders to cardiomyopathy, liver failure, and myopathy.,disease: Defects in NDUFS7 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.,function: Core subunit of the mitochondrial membrane respiratory chain NADH dehydrogenase (Complex I) that is believed to belong to the minimal assembly required for catalysis. 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.,similarity: Belongs to the complex I 20 kDa subunit family.,subunit: Complex I is composed of 45 different subunits This is a component of the iron-sulfur (IP) fragment of the enzyme.,