

MT-ATP8 rabbit pAb antibody

Catalog No :	Source:	Concentration :	Mol.Wt. (Da):
A17884	Rabbit	1 mg/ml	

Applications IHC, ELISA

Reactivity Human

Dilution IHC 1:50-200, ELISA(peptide)1:5000-20000

Rev. 1. 3



INSTRUCTIONS

~~Storage 20°C/1 year~~

Specificity This antibody detects endogenous levels of human MT-ATP8

Source / Purification The antibody was affinity-purified from mouse ascites by affinity-chromatography using specific immunogen.

Immunogen Synthesized peptide derived from human MT-ATP8

Uniprot No P03928

Alternative names ATP synthase protein 8 (A6L) (F-ATPase subunit 8)

Form Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Clonality Polyclonal

Isotype IgG

Conjugation

Background disease:Defects in MT-ATP6 are a cause of infantile bilateral striatal necrosis [MIM:500003]. Bilateral striatal necrosis is a neurological disorder resembling Leigh syndrome.,disease:Defects in MT-ATP6 are a cause of Leber hereditary optic neuropathy (LHON) [MIM:535000]. LHON is a maternally inherited disease resulting in acute or subacute loss of central vision, due to optic nerve dysfunction. Cardiac conduction defects and neurological defects have also been described in some patients. LHON results from primary mitochondrial DNA mutations affecting the respiratory chain complexes.,disease:Defects in MT-ATP6 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 MT-ATP6 are the cause of neurogenic muscle weakness, ataxia, and retinitis pigmentosa (NARP) [MIM:551500].,disease:Defects in MT-CO3 are a cause of cytochrome c oxidase deficiency (COX deficiency) [MIM:220110]; also called mitochondrial complex IV deficiency. COX deficiency is a clinically heterogeneous disorder. The clinical features are ranging from isolated myopathy to severe multisystem disease, with onset from infancy to adulthood.,disease:Defects in MT-CO3 are a cause of Leber hereditary optic neuropathy (LHON) [MIM:535000]. LHON is a maternally inherited disease resulting in acute or subacute loss of central vision, due to optic nerve dysfunction. Cardiac conduction defects and neurological defects have also been described in some patients. LHON results from primary mitochondrial DNA mutations affecting the respiratory chain complexes.,disease:Defects in MT-CO3 are associated with recurrent myoglobinuria [MIM:550500]. Myoglobinuria consists of excretion of myoglobin in the urine.,disease:Defects in MT-CO3 are found in mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes (MELAS) syndrome, a genetically heterogeneous disorder, characterized by episodic vomiting, seizures, and recurrent cerebral insults resembling strokes and causing hemiparesis, hemianopsia, or cortical blindness.,function:Mitochondrial membrane ATP synthase (F(1)F(0) ATP synthase or Complex V) produces ATP from ADP in the presence of a proton gradient across the membrane which is generated by electron transport complexes of the respiratory chain. F-type ATPases consist of two structural domains, F(1) - containing the extramembraneous catalytic core and F(0) - containing the membrane proton channel, linked together by a central stalk and a peripheral stalk. During catalysis, ATP synthesis in the catalytic domain of F(1) is coupled via a rotary mechanism of the central stalk subunits to proton translocation. Key component of the proton channel; it may play a direct role in the translocation of protons across the membrane.,function:Mitochondrial membrane ATP synthase (F(1)F(0) ATP synthase or Complex V) produces ATP from ADP in the presence of a proton gradient across the membrane which is generated by electron transport

Product Images:**Application Key:**

WB-Western IP-Immunoprecipitation IHC-Immunohistochemistry CHIP-Chromatin Immunoprecipitation
IF-Immunofluorescence F-Flow Cytometry E-P-ELISA-Peptide

Species Cross-Reactivity Key:

H-Human M-Mouse R-Rat Hm-Hamster Mk-Monkey Vir-Virus Mi-Mink C-Chicken Dm-D. melanogaster
X-Xenopus Z-Zebrafish B-Bovine Dg-Dog Pg-Pig Sc-S. cerevisiae Ce-C. elegans Hr-Horse All-All
Species Expected

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