

ATP6 Rabbit Polyclonal Antibody

Catalog# UZ-PA01127

Product information

产品货号	UZ-PA01127
别名	ATP6;ATPASE6;MTATP6
产品名称	ATP6 Rabbit Polyclonal Antibody
类别	抗体产品
基因名称	MT-ATP6
推荐应用	WB,ELISA
反应种属	Human,Rat,Mouse
存储缓冲液	Liquid in PBS containing 50% glycerol, and 0.02% New type preservative N.
Human Gene ID	4508
Human Swissprot No.	P00846
免疫原	Synthesized peptide derived from human protein . at AA range: 60-140
稀释度	WB 1:500-2000, ELISA 1:5000-20000
预测分子量	24kDa
运输及保存条件	-20°C/1 year
宿主	Rabbit
同种型	Rabbit,IgG

ATP6 Rabbit Polyclonal Antibody

Catalog# UZ-PA01127 (cont.)

Product information

背景介绍

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 an 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 (F1)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 (F1)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. Part of the complex F(0) domain. Minor subunit located with subunit a in the membrane.,function:Subunits I, II and III form the functional core of the enzyme complex.,similarity:Belongs to the ATPase A chain family.,similarity:Belongs to the ATPase protein 8 family.,similarity:Belongs to the cytochrome c oxidase subunit 3 family.,subunit:F-type ATPases have 2 components, CF(1) - the catalytic core - and CF(0) - the membrane proton channel.,subunit:F-type ATPases have 2 components, CF(1) - the catalytic core - and CF(0) - the membrane proton channel. CF(1) has five subunits: alpha(3), beta(3), gamma(1), delta(1), epsilon(1). CF(0) has three main subunits: a, b and c.,

Mitochondrion inner membrane; Multi-pass membrane protein.

The antibody was affinity-purified from rabbit serum by affinity-chromatography using epitope-specific immunogen.

Polyclonal

细胞定位

纯化

Clonality