

产品名称: MT-ATP8 Rabbit Polyclonal Antibody
产品货号: APRab14201



产品概述 (Summary)

产品名称 (Production Name)	MT-ATP8 Rabbit Polyclonal Antibody
描述 (Description)	Rabbit polyclonal Antibody
宿主 (Host)	Rabbit
应用 (Application)	IHC,ELISA
种属反应性 (Reactivity)	Human,Rat,Mouse

产品性能 (Performance)

偶联物 (Conjugation)	Unconjugated
修饰 (Modification)	Unmodified
同种型 (Isotype)	IgG
克隆 (Clonality)	Polyclonal
形式 (Form)	Liquid
存放说明 (Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液 (Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化方式 (Purification)	Affinity purification

免疫原信息 (Immunogen)

基因名 (Gene Name)	MT-ATP8
别名 (Alternative Names)	ATP synthase protein 8 (A6L;F-ATPase subunit 8)
基因 ID (Gene ID)	4509.0
蛋白 ID (SwissProt ID)	P03928.Synthesized peptide derived from human MT-ATP8 AA range: 30-110

产品应用 (Application)

稀释比 (Dilution Ratio)	IHC 1:50-1:200,ELISA 1:5000-1:20000
蛋白分子量 (Molecular Weight)	

研究背景 (Background)

disease:Defects in MT-ATP6 are a cause of infantile bilateral striatal necrosis [MIM:500003]. Bilateral striatal necrosis is a

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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 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.,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

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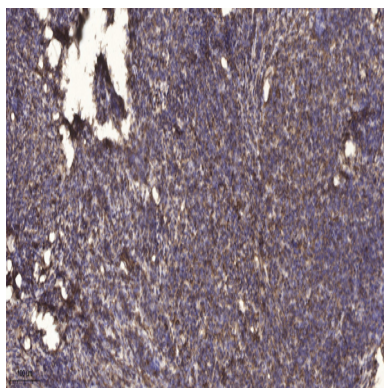
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研究领域 (Research Area)

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图片 (Image Data)



Immunohistochemical analysis of paraffin-embedded human cervical carcinoma. 1, Antibody was diluted at 1:200 (4° overnight) . 2, Tris-EDTA,pH9.0 was used for antigen retrieval. 3,Secondary antibody was diluted at 1:200 (room temperature, 45min) .

注意事项 (Note)

For research use only .