

MT-ATP6 Rabbit pAb

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Catalog # AP56875

Product Information

Application	IHC-P, IHC-F, IF
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Msx3
Epitope Specificity	21-120/226
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Mitochondrion inner membrane.
DISEASE	Defects in MT-ATP6 are the cause of neurogenic muscle weakness, ataxia, and retinitis pigmentosa (NARP) [MIM:551500]. 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. 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. Defects in MT-ATP6 are a cause of mitochondrial infantile bilateral striatal necrosis (MIBSN) [MIM:500003]. Bilateral striatal necrosis is a neurological disorder resembling Leigh syndrome.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Other Names	ATP synthase subunit a, F-ATPase protein 6, MT-ATP6, ATP6, ATPASE6, MTATP6
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Dilution

IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500

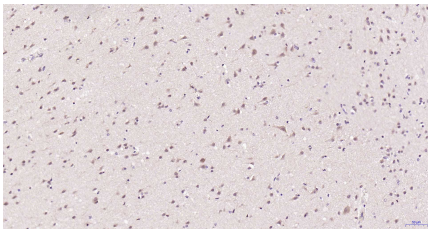
Storage

Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Background

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Images



Paraformaldehyde-fixed, paraffin embedded Human Cerebrum; Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15 min; Antibody incubation with MT-ATP6 Polyclonal Antibody, Unconjugated (AP56875) at 1:200 overnight at 4°C, followed by conjugation to the SP Kit (Rabbit, SP-0023) and DAB (C-0010) staining.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.