

<b>Product name:</b>	MT-ATP8 Rabbit Polyclonal Antibody
<b>Cat number:</b>	ABN14201
<b>Conjugate:</b>	Unconjugated
<b>Size:</b>	100µL
<b>Clone:</b>	Polyclonal
<b>Concentration:</b>	1mg/ml
<b>Host:</b>	Rabbit
<b>Isotype:</b>	IgG
<b>Immunogen:</b>	Synthesized peptide derived from human MT-ATP8 AA range: 30-110
<b>Reactivity:</b>	Human,Rat,Mouse
<b>Applications:</b>	IHC 1:50-1:200,ELISA 1:5000-1:20000
<b>Purification:</b>	Affinity purification
<b>Form:</b>	Liquid
<b>Buffer:</b>	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
<b>Storage:</b>	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

**Background:**

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 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].