

## Product datasheet

# Anti-MT-ATP6 antibody [1G7-1G2] - N-terminal ab219825

[2 Images](#)

### Overview

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<b>Product name</b>	Anti-MT-ATP6 antibody [1G7-1G2] - N-terminal
<b>Description</b>	Mouse monoclonal [1G7-1G2] to MT-ATP6 - N-terminal
<b>Tested applications</b>	<b>Suitable for:</b> WB
<b>Species reactivity</b>	<b>Reacts with:</b> Human
<b>Immunogen</b>	Synthetic peptide corresponding to Human MT-ATP6 (N terminal). Database link: <a href="#">P00846</a>
<b>Positive control</b>	WB: Mitochondria from cultured normal control human dermal fibroblasts neonatal (HDFn); Whole cell extract of cultured normal control human dermal fibroblasts neonatal (HDFn).

### Properties

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<b>Form</b>	Liquid
<b>Storage instructions</b>	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
<b>Storage buffer</b>	pH: 7.20 Preservative: 0.02% Sodium azide Constituents: 0.36% HEPES, 0.87% Sodium chloride
<b>Purity</b>	Protein L purified
<b>Purification notes</b>	Purified from hybridoma cell culture supernatant by Protein L affinity chromatography from fetal bovine serum containing medium (Protein L does not bind bovine IgG).
<b>Clonality</b>	Monoclonal
<b>Clone number</b>	1G7-1G2
<b>Isotype</b>	IgG2b
<b>Light chain type</b>	kappa

### Applications

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Our [Abpromise guarantee](#) covers the use of **ab219825** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		Use a concentration of 0.5 - 5 µg/ml. Detects a band of approximately 22 kDa (predicted molecular weight: 25 kDa).

## Target

### 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.

### Involvement in 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.

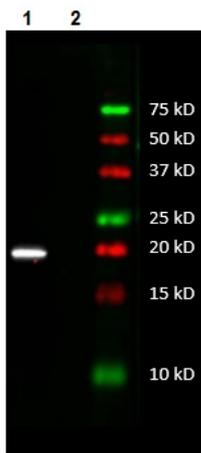
### Sequence similarities

Belongs to the ATPase A chain family.

### Cellular localization

Mitochondrion inner membrane.

## Images



Western blot - Anti-MT-ATP6 antibody [1G7-1G2] - N-terminal (ab219825)

**All lanes :** Anti-MT-ATP6 antibody [1G7-1G2]

- N-terminal (ab219825) at 0.5 µg/ml

**Lane 1 :** Mitochondria from cultured normal control human dermal fibroblasts neonatal (HDFn)

**Lane 2 :** Mitochondria from HDFn cells depleted of mtDNA by long-term proliferation in the presence of ethidium bromide

Lysates/proteins at 10 µg per lane.

### Secondary

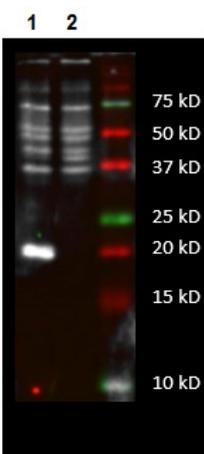
HRP-labeled Goat-anti-mouse IgG

Developed using the ECL technique

**Predicted band size :** 25 kDa

**Observed band size :** 22 kDa

Mitochondrial proteins solubilized in 2% SDS were separated by SDS-PAGE and then transferred to PVDF membranes in CAPS buffer.



Western blot - Anti-MT-ATP6 antibody [1G7-1G2] - N-terminal (ab219825)

**All lanes :** Anti-MT-ATP6 antibody [1G7-1G2]

- N-terminal (ab219825) at 5 µg/ml

**Lane 1 :** Whole cell extract of cultured normal control human dermal fibroblasts neonatal (HDFn)

**Lane 2 :** Whole cell extract HDFn-Rho0 cells depleted of mtDNA by long-term culture in the presence of ethidium bromide

Lysates/proteins at 15 µg per lane.

### Secondary

HRP-labeled Goat-anti-mouse IgG

Developed using the ECL technique

**Predicted band size :** 25 kDa

**Observed band size :** 22 kDa

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