Mitochondrial ATPase is a multisubunit enzyme that catalyzes ATP synthesis during oxidative phosphorylation. It consists of a globular, membrane-extrinsic F₁ catalytic unit, and an H⁺-translocating, membrane-spanning F₀ unit. ATPAF2 (ATP synthase mitochondrial F₁ complex assembly factor 2), also known as ATP12, is a 289 amino acid protein that plays a role in the assembly of the F₁ unit. Localized to the mitochondria, ATPAF2 binds specifically to the F₁αε subunit and prevents it from forming nonproductive homooligomers during enzyme assembly. Defects in the gene encoding ATPAF2 have shown to cause complex V mitochondrial respiratory chain ATPAF2 subunit deficiency (ATPAF2 deficiency), also known as ATP synthase deficiency or ATPase deficiency. ATPAF2 deficiency is an early presenting disease in which lactic acidosis, dysmorphic features and methyl glutaconic aciduria can be major clues in the diagnosis. Dysmorphic features include a large mouth, prominent nasal bridge, micrognathia, rocker-bottom feet and flexion contractures of the limbs associated with camptodactyly.

REFERENCES


CHROMOSOMAL LOCATION

Genetic locus: ATPAF2 (human) mapping to 17p11.2; Atfap2 (mouse) mapping to 11 B2.

SOURCE

ATPAF2 (H-76) is a rabbit polyclonal antibody raised against amino acids 78-153 mapping within an internal region of ATPAF2 of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

ATPAF2 (H-76) is recommended for detection of ATPAF2 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation (1-2 µg per 100-500 µg of total protein [1 ml of cell lysate]), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

ATPAF2 (H-76) is also recommended for detection of ATPAF2 in additional species, including equine, canine, bovine and avian.

Suitable for use as control antibody for ATPAF2 siRNA (h): sc-93957, ATPAF2 siRNA (m): sc-141371, ATPAF2 shRNA Plasmid (h): sc-93957-SH, ATPAF2 shRNA Plasmid (m): sc-141371-SH, ATPAF2 shRNA (h) Lentiviral Particles: sc-93957-V and ATPAF2 shRNA (m) Lentiviral Particles: sc-141371-V.

Molecular Weight of ATPAF2: 33 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:6000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:50-1:500), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 4) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2035 (0.5 µl agarose/2.0 µl). 5) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our website at www.scbt.com or our catalog for detailed protocols and support products.