SANTA CRUZ BIOTECHNOLOGY, INC.

ATPAF2 (L-14): sc-107447

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Mitochondrial ATPase is a multisubunit enzyme that catalyzes ATP synthesis during oxidative phosphorylation. It consists of a globular, membrane-extrinsic F_1 catalytic unit, and an H^+-translocating, membrane-spanning F_0 unit. ATPAF2 (ATP synthase mitochondrial F_1 complex assembly factor 2), also known as ATP12, is a 289 amino acid protein that plays a role in the assembly of the F_1 unit. Localized to the mitochondria, ATPAF2 binds specifically to the $F_1\alpha$ 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

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CHROMOSOMAL LOCATION

Genetic locus: ATPAF2 (human) mapping to 17p11.2; Atpaf2 (mouse) mapping to 11 B2.

SOURCE

ATPAF2 (L-14) is an affinity purified goat polyclonal antibody raised against a peptide 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.

Blocking peptide available for competition studies, sc-107447 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

ATPAF2 (L-14) 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), 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 (L-14) 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 donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (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 web site at www.scbt.com or our catalog for detailed protocols and support products.