SANTA CRUZ BIOTECHNOLOGY, INC.

ATPAF2 siRNA (h): sc-93957

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 α 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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2. Wang, Z.G., et al. 2001. Atp11p and Atp12p are assembly factors for the F_1-ATPase in human mitochondria. J. Biol. Chem. 276: 30773-30778.
3. Ackerman, S.H. 2002. Atp11p and Atp12p are chaperones for F_1-ATPase biogenesis in mitochondria. Biochim. Biophys. Acta 1555: 101-105.
4. Bi, W., et al. 2002. Genes in a refined Smith-Magenis syndrome critical deletion interval on chromosome 17p11.2 and the syntenic region of the mouse. Genome Res. 12: 713-728.
5. Picková, A., et al. 2003. Differential expression of ATPAF1 and ATPAF2 genes encoding F_1-ATPase assembly proteins in mouse tissues. FEBS Lett. 551: 42-46.
6. Hinton, A., et al. 2004. The molecular chaperone, Atp12p, from Homo sapiens. *In vitro* studies with purified wild type and mutant (E240K) proteins. J. Biol. Chem. 279: 9016-9022.

CHROMOSOMAL LOCATION

Genetic locus: ATPAF2 (human) mapping to 17p11.2.

PRODUCT

ATPAF2 siRNA (h) is a pool of 3 target-specific 19-25 nt siRNAs designed to knock down gene expression. Each vial contains 3.3 nmol of lyophilized siRNA, sufficient for a 10 µM solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see ATPAF2 shRNA Plasmid (h): sc-93957-SH and ATPAF2 shRNA (h) Lentiviral Particles: sc-93957-V as alternate gene silencing products.

For independent verification of ATPAF2 (h) gene silencing results, we also provide the individual siRNA duplex components. Each is available as 3.3 nmol of lyophilized siRNA. These include: sc-93957A, sc-93957B and sc-93957C.

STORAGE AND RESUSPENSION

Store lyophilized siRNA duplex at -20° C with desiccant. Stable for at least one year from the date of shipment. Once resuspended, store at -20° C, avoid contact with RNAses and repeated freeze thaw cycles.

Resuspend lyophilized siRNA duplex in 330 µl of the RNAse-free water provided. Resuspension of the siRNA duplex in 330 µl of RNAse-free water makes a 10 µM solution in a 10 µM Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

ATPAF2 siRNA (h) is recommended for the inhibition of ATPAF2 expression in human cells.

SUPPORT REAGENTS

For optimal siRNA transfection efficiency, Santa Cruz Biotechnology's siRNA Transfection Reagent: sc-29528 (0.3 ml), siRNA Transfection Medium: sc-36868 (20 ml) and siRNA Dilution Buffer: sc-29527 (1.5 ml) are recommended. Control siRNAs or Fluorescein Conjugated Control siRNAs are available as 10 µM in 66 µl. Each contain a scrambled sequence that will not lead to the specific degradation of any known cellular mRNA. Fluorescein Conjugated Control siRNAs include: sc-36869, sc-44239, sc-44240 and sc-44241. Control siRNAs include: sc-37007, sc-44230, sc-44231, sc-44232, sc-44233, sc-44234, sc-44235, sc-44236, sc-44237 and sc-44238.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor ATPAF2 gene expression knockdown using RT-PCR Primer: ATPAF2 (h)-PR: sc-93957-PR (20 µl). Annealing temperature for the primers should be 55-60° C and the extension temperature should be 68-72° C.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.