

ETHE1 siRNA (h): sc-97755

BACKGROUND

ETHE1 (ethylmalonic encephalopathy 1), also known as HSCO (hepatoma subtracted clone one protein), is a 254 amino acid protein belonging to the metallo- β -lactamase superfamily and glyoxalase II family. Localizing to the cytoplasm, nucleus and mitochondrion matrix, ETHE1 is ubiquitously expressed and may function in sulfide catabolism. ETHE1 binds two zinc ions per subunit and interacts directly with RELA, preventing its localization to the nucleus thus leading to suppressed p53-induced apoptosis. The gene encoding ETHE1 maps to human chromosome 19q13.31. Mutations to this gene result in ethylmalonic encephalopathy, an infantile metabolic disorder characterized by high levels of ethylmalonic acid, neurodevelopmental delay and regression, recurrent petechiae, acrocyanosis, and death within the first decade of life.

REFERENCES

1. Higashitsuji, H., et al. 2002. A novel protein overexpressed in hepatoma accelerates export of NF κ B from the nucleus and inhibits p53-dependent apoptosis. *Cancer Cell* 2: 335-346.
2. Mootha, V.K., et al. 2003. Identification of a gene causing human cytochrome c oxidase deficiency by integrative genomics. *Proc. Natl. Acad. Sci. USA* 100: 605-610.
3. Tiranti, V., et al. 2004. Ethylmalonic encephalopathy is caused by mutations in ETHE1, a gene encoding a mitochondrial matrix protein. *Am. J. Hum. Genet.* 74: 239-252.
4. Higashitsuji, H., et al. 2007. Enhanced deacetylation of p53 by the anti-apoptotic protein HSCO in association with histone deacetylase 1. *J. Biol. Chem.* 282: 13716-13725.
5. Mineri, R., et al. 2008. Identification of new mutations in the ETHE1 gene in a cohort of 14 patients presenting with ethylmalonic encephalopathy. *J. Med. Genet.* 45: 473-478.
6. Tiranti, V., et al. 2009. Loss of ETHE1, a mitochondrial dioxygenase, causes fatal sulfide toxicity in ethylmalonic encephalopathy. *Nat. Med.* 15: 200-205.
7. Ismail, E.A., et al. 2009. Ethylmalonic encephalopathy. Another patient from Kuwait. *Neurosciences.* 14: 78-80.

CHROMOSOMAL LOCATION

Genetic locus: ETHE1 (human) mapping to 19q13.31.

PRODUCT

ETHE1 siRNA (h) is a pool of 2 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 μ M solution once resuspended using protocol below. Suitable for 50-100 transfections. Also see ETHE1 shRNA Plasmid (h): sc-97755-SH and ETHE1 shRNA (h) Lentiviral Particles: sc-97755-V as alternate gene silencing products.

For independent verification of ETHE1 (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-97755A and sc-97755B.

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 μ l of the RNase-free water provided. Resuspension of the siRNA duplex in 330 μ l of RNase-free water makes a 10 μ M solution in a 10 μ M Tris-HCl, pH 8.0, 20 mM NaCl, 1 mM EDTA buffered solution.

APPLICATIONS

ETHE1 siRNA (h) is recommended for the inhibition of ETHE1 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 μ M in 66 μ l. 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.

GENE EXPRESSION MONITORING

ETHE1 (B-12): sc-393869 is recommended as a control antibody for monitoring of ETHE1 gene expression knockdown by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) or immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker[™] Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850.

RT-PCR REAGENTS

Semi-quantitative RT-PCR may be performed to monitor ETHE1 gene expression knockdown using RT-PCR Primer: ETHE1 (h)-PR: sc-97755-PR (20 μ l). 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.