SANTA CRUZ BIOTECHNOLOGY, INC.

ETHE1 (h2): 293T Lysate: sc-110987

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 NFκB 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.

CHROMOSOMAL LOCATION

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

PRODUCT

ETHE1 (h2): 293T Lysate represents a lysate of human ETHE1 transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

APPLICATIONS

ETHE1 (h2): 293T Lysate is suitable as a Western Blotting positive control for human reactive ETHE1 antibodies. Recommended use: 10-20 µl per lane.

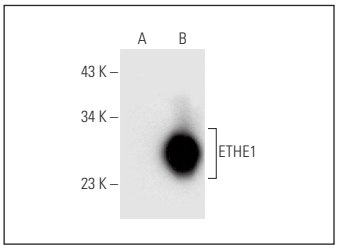
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

ETHE1 (B-12): sc-393869 is recommended as a positive control antibody for Western Blot analysis of enhanced human ETHE1 expression in ETHE1 transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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.

DATA



ETHE1 (B-12): sc-393869. Western blot analysis of ETHE1 expression in non-transfected: sc-117752 (**A**) and human ETHE1 transfected: sc-110987 (**B**) 293T whole cell lysates.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. 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 for detailed protocols and support products.