SANTA CRUZ BIOTECHNOLOGY, INC.

HCS (N-19): sc-23732

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Holocarboxylase synthetase (HCS, HLCS) catalyzes the postranslational covalent attachment of biotin to biotin-dependent carboxylases. These carboxylases include pyruvate carboxylase, propionyl coenzyme A (CoA) carboxylase, beta-methylcrotonoyl CoA carboxylase, and acetyl CoA carboxylase. HCS is an obligate participant in biotin-mediated regulation of its own expression and of biotin-dependent carboxylase mRNA levels in human cells. In addition, biotin regulates the genetic expression of HCS and mitochondrial carboxylases. Multiple carboxylase deficiency (MCD) is a rare metabolic life-threatening disease accountable for early onset biotin-responsive multiple carboxylase deficiency. MCD presents with lactic acidosis, tachypnea, temperature instability, and shock in neonates. Interstingly, MCD can be detected prenataly and, if diagnosed, prenatally administered biotin is effectively taken up by the fetus and prevents functional deficiency of the carboxylases in an affected newborn. The HCS gene maps to human chromosome 21q22.1 and enocodes a cytoplasmic and mitochondrial protein.

REFERENCES

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3. Suormala, T., et al. 1998. Late-onset holocarboxylase synthetase-deficiency: pre- and post-natal diagnosis and evaluation of effectiveness of antenatal biotin therapy. Eur. J. Pediatr. 157: 570-575.
4. Hwu, W.L., et al. 2000. Late-onset holocarboxylase synthetase deficiency with homologous R508W mutation. J. Formos. Med. Assoc. 99: 174-177.
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7. Solorzano-Vargas, R.S., et al. 2002. Holocarboxylase synthetase is an obligate participant in biotin-mediated regulation of its own expression and of biotin-dependent carboxylases mRNA levels in human cells. Proc. Natl. Acad. Sci. USA 99: 5325-5330.
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CHROMOSOMAL LOCATION

Genetic locus: HLCS (human) mapping to 21q22.13; Hlcs (mouse) mapping to 16.

SOURCE

HCS (N-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of HCS 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-23732 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

HCS (N-19) is recommended for detection of HCS of 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).

Suitable for use as control antibody for HCS siRNA (h): sc-72198.

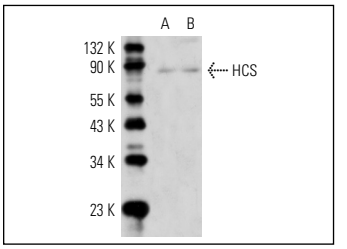
Molecular Weight of HCS: 86 kDa.

Positive Controls: JAR cell lysate: sc-2276 or JEG-3 whole cell lysates.

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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.

DATA



HCS (N-19): sc-23732. Western blot analysis of HCS expression in JAR (**A**) and JEG-3 (**B**) whole cell lysates.

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.