

MCCA (M-280): sc-66988

BACKGROUND

3-methylcrotonyl-CoA:carboxylase (MCC) is an enzyme crucial in the breakdown of the branched chain amino acid leucine. Methylcrotonyl-CoA carboxylase α chain (MCCA), also designated 3-methylcrotonyl-CoA carboxylase 1, is located in the mitochondrial matrix. MCCA functions as a heterodimer and catalyzes the carboxylation of 3-methylcrotonyl-CoA to form 3-methylglutacetyl-CoA. MCCA has a Biotin cofactor. The gene encoding for the 725 amino acid MCCA protein maps to chromosome 3q26-q28 and consists of 19 exons. Defects in this gene are associated with 3-methylcrotonylglycinuria (MCGI), an autosomal recessive disorder characterized by muscular hypotonia and atrophy. Human MCC deficiency, also inherited recessively, is characterized by 3-methylcrotonyl-CoA accumulation. Symptoms may be highly variable, ranging from completely asymptomatic to metabolic acidosis and death in infancy.

REFERENCES

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- Chandler, C.S., et al. 1986. Multiple Biotin-containing proteins in 3T3-L1 cells. *Biochem. J.* 237: 123-130.
- Holzinger, A., et al. 2001. Cloning of the human MCCA and MCCB genes and mutations therein reveal the molecular cause of 3-methylcrotonyl-CoA:carboxylase deficiency. *Hum. Mol. Genet.* 10: 1299-1306.
- Baumgartner, M.R., et al. 2001. The molecular basis of human 3-methylcrotonyl-CoA carboxylase deficiency. *J. Clin. Invest.* 107: 495-504.
- Gallardo, M.E., et al. 2001. The molecular basis of 3-methylcrotonylglycinuria, a disorder of leucine catabolism. *Am. J. Hum. Genet.* 68: 334-346.
- Baumgartner, M.R., et al. 2004. Isolated 3-methylcrotonyl-CoA carboxylase deficiency: evidence for an allele-specific dominant negative effect and responsiveness to Biotin therapy. *Am. J. Hum. Genet.* 75: 790-800.
- Rodriguez, J.M., et al. 2004. Fungal metabolic model for 3-methylcrotonyl-CoA carboxylase deficiency. *J. Biol. Chem.* 279: 4578-4587.

CHROMOSOMAL LOCATION

Genetic locus: *Mccc1* (mouse) mapping to 3 B.

SOURCE

MCCA (M-280) is a rabbit polyclonal antibody raised against amino acids 431-710 mapping near the C-terminus of MCCA of mouse origin.

PRODUCT

Each vial contains 200 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

MCCA (M-280) is recommended for detection of MCCA of mouse and rat origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g 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 MCCA siRNA (m): sc-45693, MCCA shRNA Plasmid (m): sc-45693-SH and MCCA shRNA (m) Lentiviral Particles: sc-45693-V.

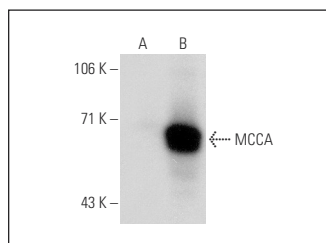
Molecular Weight of MCCA: 75 kDa.

Positive Controls: MCCA (m): 293T Lysate: sc-121556, 3T3-L1 cell lysate: sc-2243 or mouse liver extract: sc-2256.

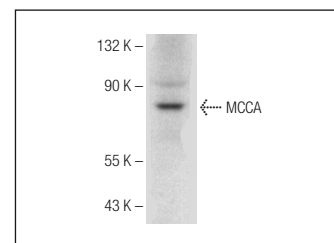
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



MCCA (M-280): sc-66988. Western blot analysis of MCCA expression in non-transfected: sc-117752 (A) and mouse MCCA transfected: sc-121556 (B) 293T whole cell lysates.



MCCA (M-280): sc-66988. Western blot analysis of MCCA expression in A-431 whole cell lysate.

RESEARCH USE

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

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Try **MCCA (B-7): sc-365754** or **MCCA (D-11): sc-376647**, our highly recommended monoclonal alternatives to MCCA (M-280).