SANTA CRUZ BIOTECHNOLOGY, INC.

# MCCA (H-290): sc-66960

SANTA CRUZ BIOTECHNOLOGY
The Power to Question

## BACKGROUND

3-methylcrotonyl-CoA:carboxylase (MCC) is an enzyme crucial in the breakdown of the branched chain amino acid leucine. Methylcrotonoyl-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-methylglutaconyl-CoA. MCCA has a biotin cofactor. The gene encoding for the 725 amino acid MCCA protein maps to chromosome 3q27.1 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


1. Bartlett, K., et al. 1984. Isolated biotin-resistant 3-methylcrotonyl CoA carboxylase deficiency presenting with life-threatening hypoglycaemia. J. Inherit. Metab. Dis. 7: 182.
2. Chandler, C.S., et al. 1986. Multiple Biotin-containing proteins in 3T3-L1 cells. Biochem. J. 237: 123-130.
3. 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.
4. Baumgartner, M.R., et al. 2001. The molecular basis of human 3-methylcrotonyl-CoA carboxylase deficiency. J. Clin. Invest. 107: 495-504.
5. Gallardo, M.E., et al. 2001. The molecular basis of 3-methylcrotonylglycinuria, a disorder of leucine catabolism. Am. J. Hum. Genet. 68: 334-346.
6. 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.
7. 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 (human) mapping to 3q27.1; Mccc1 (mouse) mapping to 3 B.

## SOURCE

MCCA (H-290) is a rabbit polyclonal antibody raised against amino acids 436-725 mapping at the C-terminus of MCCA 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.

## STORAGE

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

## APPLICATIONS

MCCA (H-290) is recommended for detection of MCCA of human and, to a lesser extent, mouse and rat 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).

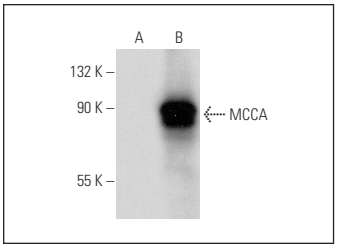
MCCA (H-290) is also recommended for detection of MCCA in additional species, including equine.

Suitable for use as control antibody for MCCA siRNA (h): sc-45692, MCCA siRNA (m): sc-45693, MCCA shRNA Plasmid (h): sc-45692-SH, MCCA shRNA Plasmid (m): sc-45693-SH, MCCA shRNA (h) Lentiviral Particles: sc-45692-V and MCCA shRNA (m) Lentiviral Particles: sc-45693-V.

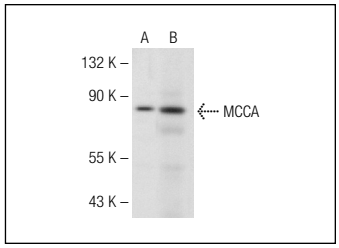
Molecular Weight of MCCA: 75 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227 or A-431 whole cell lysate: sc-2201.

## DATA




MCCA (H-290): sc-66960. Western blot analysis of MCCA expression in non-transfected: sc-117752 (**A**) and mouse MCCA transfected: sc-121556 (**B**) 293T whole cell lysates.




MCCA (H-290): sc-66960. Western blot analysis of MCCA expression in A-431 whole cell lysate (**A**) and mouse liver tissue extract (**B**).

## 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.

MONOS Satisfaction Guaranteed

TTry **MCCA (B-7): sc-365754** or **MCCA (D-11): sc-376647**, our highly recommended monoclonal alternatives to MCCA (H-290).