SANTA CRUZ BIOTECHNOLOGY, INC.

# MCCA (D-17): sc-46162

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*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 (D-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region 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.

Blocking peptide available for competition studies, sc-46162 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

MCCA (D-17) is recommended for detection of MCCA of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (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 (D-17) is also recommended for detection of MCCA in additional species, including equine, canine, porcine and avian.

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.

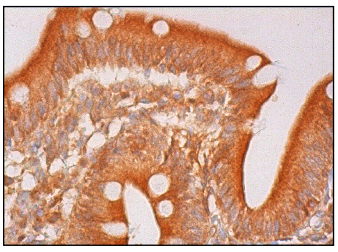
Molecular Weight of MCCA: 75 kDa.

Positive Controls: 3T3-L1 cell lysate: sc-2243, mouse liver extract: sc-2256 or rat liver extract: sc-2395.

## 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) 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. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

## DATA



MCCA (D-17): sc-46162. Immunoperoxidase staining of formalin fixed, paraffin-embedded human small intestine tissue showing cytoplasmic staining of glandular cells.

## 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.