

# MCCA (D-11): sc-376647

## BACKGROUND

3-methylcrotonyl-CoA: carboxylase (MCC) is an enzyme crucial in the breakdown of the branched chain amino acid leucine. Methylcrotonyl-CoA carboxylase  $\alpha$  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-11) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 465-503 within an internal region of MCCA of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG<sub>2a</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-376647 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

## APPLICATIONS

MCCA (D-11) is recommended for detection of MCCA of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2  $\mu$ g per 100-500  $\mu$ 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 (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.

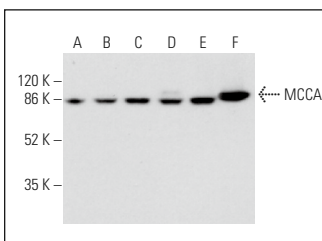
Molecular Weight of MCCA: 75 kDa.

Positive Controls: C2C12 whole cell lysate: sc-364188, A-673 cell lysate: sc-2414 or HeLa whole cell lysate: sc-2200.

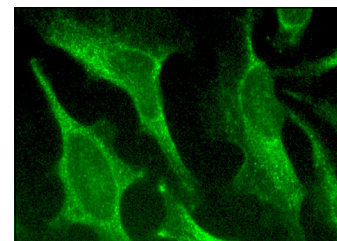
## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  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. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgG $\kappa$  BP-FITC: sc-516140 or m-IgG $\kappa$  BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

## DATA



MCCA (D-11): sc-376647. Western blot analysis of MCCA expression in HeLa (A), A-673 (B), BC<sub>3</sub>H1 (C), SJRH30 (D), C2C12 (E) and NIH/3T3 (F) whole cell lysates.



MCCA (D-11): sc-376647. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization.

## 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](http://www.scbt.com) for detailed protocols and support products.