SANTA CRUZ BIOTECHNOLOGY, INC.

MCCA (A-4): sc-271427

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 (mouse) mapping to 3 B.

SOURCE

MCCA (A-4) is a mouse monoclonal antibody raised against amino acids 431-710 mapping near the C-terminus of MCCA of mouse origin.

PRODUCT

Each vial contains 200 µg IgG_{2a} kappa light chain 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 (A-4) is recommended for detection of MCCA of mouse and rat origin by Western Blotting (starting dilution 1:100, 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 MCCA siRNA (m): sc-45693, MCCA shRNA Plasmid (m): sc-45693-SH and MCCA shRNA (m) Lentiviral Particles: sc-45693-V.

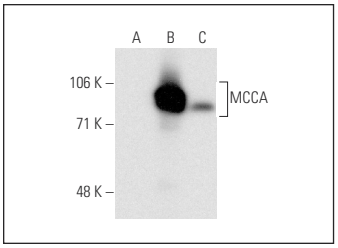
Molecular Weight of MCCA: 75 kDa.

Positive Controls: MCCA (m): 293T Lysate: sc-121556, C2C12 whole cell lysate: sc-364188 or 3T3-L1 cell lysate: sc-2243.

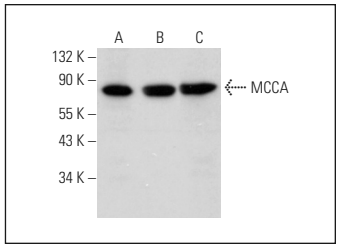
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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-IgGκ BP-FITC: sc-516140 or m-IgGκ 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 (A-4): sc-271427. Western blot analysis of MCCA expression in non-transfected 293T: sc-117752 (**A**), mouse MCCA transfected 293T: sc-121556 (**B**) and 3T3-L1 (**C**) whole cell lysates.



MCCA (A-4): sc-271427. Western blot analysis of MCCA expression in C3H/10T1/2 (**A**), BC_3H1 (**B**) and C2C12 (**C**) whole cell lysates.

SELECT PRODUCT CITATIONS

1. Anderson, K.A., et al. 2017. SIRT4 is a lysine deacylase that controls leucine metabolism and Insulin secretion. Cell Metab. 25: 838-855.e15.
2. Zaganjor, E., et al. 2021. SIRT4 is an early regulator of branched-chain amino acid catabolism that promotes adipogenesis. Cell Rep. 36: 109345.

RESEARCH USE

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