

MCCA (B-7): sc-365754

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-methylglutacetyl-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

- Bartlett, K., et al. 1984. Isolated biotin-resistant 3-methylcrotonyl CoA carboxylase deficiency presenting with life-threatening hypoglycaemia. *J. Inher. Metab. Dis.* 7: 182.
- 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.

CHROMOSOMAL LOCATION

Genetic locus: MCCC1 (human) mapping to 3q27.1; Mccc1 (mouse) mapping to 3 B.

SOURCE

MCCA (B-7) is a mouse monoclonal antibody raised against amino acids 436-725 mapping at the C-terminus of MCCA of human origin.

PRODUCT

Each vial contains 200 μ g IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

MCCA (B-7) is available conjugated to agarose (sc-365754 AC), 500 μ g/0.25 ml agarose in 1 ml, for IP; to HRP (sc-365754 HRP), 200 μ g/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-365754 PE), fluorescein (sc-365754 FITC), Alexa Fluor[®] 488 (sc-365754 AF488), Alexa Fluor[®] 546 (sc-365754 AF546), Alexa Fluor[®] 594 (sc-365754 AF594) or Alexa Fluor[®] 647 (sc-365754 AF647), 200 μ g/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor[®] 680 (sc-365754 AF680) or Alexa Fluor[®] 790 (sc-365754 AF790), 200 μ g/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

MCCA (B-7) 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 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 (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.

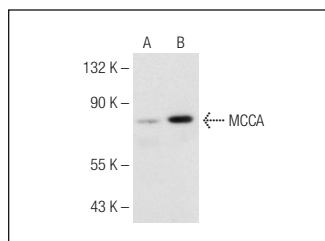
Molecular Weight of MCCA: 75 kDa.

Positive Controls: 3T3-L1 cell lysate: sc-2243, Hep G2 cell lysate: sc-2227 or A-431 whole cell lysate: sc-2201.

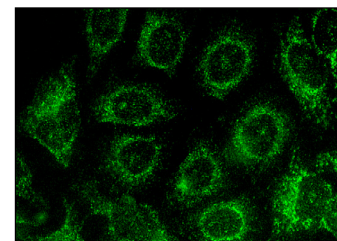
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ 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 κ BP-FITC: sc-516140 or m-IgG κ 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 (B-7): sc-365754. Western blot analysis of MCCA expression in Hep G2 (A) and A-431 (B) whole cell lysates.



MCCA (B-7): sc-365754. Immunofluorescence staining of methanol-fixed HeLa cells showing cytoplasmic localization.

SELECT PRODUCT CITATIONS

- Son, S.M., et al. 2020. Leucine regulates autophagy via acetylation of the mTORC1 component raptor. *Nat. Commun.* 11: 3148.

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.