**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. MCCAs function as a heterodimer and catalyzes the carboxylation of 3-methylcrotonyl-CoA to form 3-methylglutaconyl-CoA. MCCAs have 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 (MCU), 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**


**CHROMOSOMAL LOCALIZATION**

Genetic locus: MCCC1 (human) mapping to 3q27.1; Mcc1 (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 µg IgG2a 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 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-365754 HRP), 200 µg/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 µg/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 µg/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 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 (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-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Hard-set Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

**DATA**

**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 for detailed protocols and support products.