SANTA CRUZ BIOTECHNOLOGY, INC.

MCCB (B-1): sc-390836

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Methylcrotonoly-CoA carboxylase β chain, or MCCB, is the non-biotin containing subunit of the MCC enzyme. The deduced 563-amino acid polypeptide contains an N-terminal mitochondrial targeting sequence. MCCB is a putative dodecamer composed of six biotin-containing α subunits and six β subunits. MCCB plays a role in leucine catabolism, and catalyzes the conversion of 3-methylcrotonoyl-CoA to 3-methylglutaconyl-CoA, using ATP as an energy source. Defects in the MCCC2 gene which encodes MCCB causes 3-methylcrotonylglycinuria type II (MCGII), a recessive disease characterized by muscular hypotonia and atrophy, probably of spinal origin.

REFERENCES

1. Bannwart, C., et al. 1993. Isolated biotin-resistant deficiency of 3-methylcrotonyl-CoA carboxylase presenting as a clinically severe form in a newborn with fatal outcome. J. Inherit. Metab. Dis. 15: 863-868.
2. Baumgartner, M.R., et al. 2001. The molecular basis of human 3-methylcrotonyl-CoA carboxylase deficiency. J. Clin. Invest. 107: 495-504.
3. Gallardo, M.E., et al. 2001. The molecular basis of 3-methylcrotonylglycinuria, a disorder of leucine catabolism. Am. J. Hum. Genet. 68: 334-346.
4. 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.
5. Desviat, L.R., et al. 2003. Functional analysis of MCCA and MCCB mutations causing methylcrotonylglycinuria. Mol. Genet. Metab. 80: 315-320.

CHROMOSOMAL LOCATION

Genetic locus: MCCC2 (human) mapping to 5q13.2; Mccc2 (mouse) mapping to 13 D1.

SOURCE

MCCB (B-1) is a mouse monoclonal antibody raised against amino acids 241-540 mapping near the C-terminus of MCCB of human origin.

PRODUCT

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

MCCB (B-1) is available conjugated to agarose (sc-390836 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-390836 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-390836 PE), fluorescein (sc-390836 FITC), Alexa Fluor® 488 (sc-390836 AF488), Alexa Fluor® 546 (sc-390836 AF546), Alexa Fluor® 594 (sc-390836 AF594) or Alexa Fluor® 647 (sc-390836 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-390836 AF680) or Alexa Fluor® 790 (sc-390836 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

MCCB (B-1) is recommended for detection of MCCB 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), 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).

Suitable for use as control antibody for MCCB siRNA (h): sc-60998, MCCB siRNA (m): sc-60999, MCCB shRNA Plasmid (h): sc-60998-SH, MCCB shRNA Plasmid (m): sc-60999-SH, MCCB shRNA (h) Lentiviral Particles: sc-60998-V and MCCB shRNA (m) Lentiviral Particles: sc-60999-V.

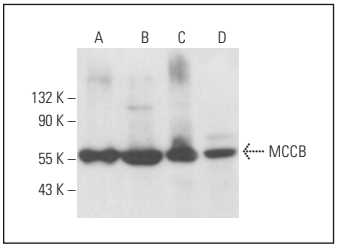
Molecular Weight of MCCB: 61 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, RT-4 whole cell lysate: sc-364257 or HeLa whole cell lysate: sc-2200.

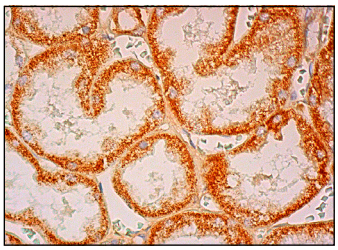
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-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. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



MCCB (B-1): sc-390836. Western blot analysis of MCCB expression in Hep G2 (**A**), RT-4 (**B**) and HeLa (**C**) whole cell lysates and rat liver tissue extract (**D**).



MCCB (B-1): sc-390836. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in tubules.

SELECT PRODUCT CITATIONS

1. Park, S., et al. 2022. Transcription factors TEAD2 and E2A globally repress acetyl-CoA synthesis to promote tumorigenesis. Mol. Cell 82: 4246-4261.e11.

RESEARCH USE

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