

MCCB (C-20): sc-50738

BACKGROUND

Methylcrotonoyl-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. The MCCC2 gene maps to chromosome 5q13.2.

REFERENCES

- Bannwart, C., Wermuth, B., Baumgartner, R., Suormala, T. and Weismann, U.N. 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.
- Baumgartner, M.R., Almashanu, S., Suormala, T., Obie, C., Cole, R.N., Packman, S., Baumgartner, E.R. and Valle, D. 2001. The molecular basis of human 3-methylcrotonyl-CoA carboxylase deficiency. *J. Clin. Invest.* 107: 495-504.
- Gallardo, M.E., Desviat, L.R., Rodríguez, J.M., Esparza-Gordillo, J., Perez-Cerdá, C., Perez, B., Rodríguez-Pombo, P., Criado, O., Sanz, R., Morton, D.H., Gibson, K.M., Le, T.P., Ribes, A., de Córdoba, S.R., Ugarte, M. and Peñalva, M.A. 2001. The molecular basis of 3-methylcrotonylglycinuria, a disorder of leucine catabolism. *Am. J. Hum. Genet.* 68: 334-346.
- Holzinger, A., Röschinger, W., Lagler, F., Mayerhofer, P.U., Lichtner, P., Kattenfeld, T., Thuy, L.P., Nyhan, W.L., Koch, H.G., Muntau, A.C. and Roscher, A.A. 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.
- Desviat, L.R., Perez-Cerdá, C., Perez, B., Esparza-Gordillo, J., Rodríguez-Pombo, P., Peñalva, M.A., Rodríguez De Córdoba, S. and Ugarte, M. 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 (C-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of MCCB of human origin.

PRODUCT

Each vial contains 200 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-50738 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

MCCB (C-20) is recommended for detection of MCCB of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

MCCB (C-20) is also recommended for detection of MCCB in additional species, including equine, canine and avian.

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.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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 or our catalog for detailed protocols and support products.



Try **MCCB (B-1): sc-390836**, our highly recommended monoclonal alternative to MCCB (C-20).