



# SZABO SCANDIC

Part of Europa Biosite

## Produktinformation



Forschungsprodukte & Biochemikalien



Zellkultur & Verbrauchsmaterial



Diagnostik & molekulare Diagnostik



Laborgeräte & Service

Weitere Information auf den folgenden Seiten!  
See the following pages for more information!



### Lieferung & Zahlungsart

siehe unsere [Liefer- und Versandbedingungen](#)

### Zuschläge

- Mindermengenzuschlag
- Trockeneiszuschlag
- Gefahrgutzuschlag
- Expressversand

### SZABO-SCANDIC HandelsgmbH

Quellenstraße 110, A-1100 Wien

T. +43(0)1 489 3961-0

F. +43(0)1 489 3961-7

[mail@szabo-scandic.com](mailto:mail@szabo-scandic.com)

[www.szabo-scandic.com](http://www.szabo-scandic.com)

[linkedin.com/company/szaboscandic](https://www.linkedin.com/company/szaboscandic) 

# MCCA (m): 293T Lysate: sc-121556

## BACKGROUND

3-methylcrotonyl-CoA:carboxylase (MCC) is an enzyme crucial in the breakdown of the branched chain amino acid leucine. Methylcrotonoyl-CoA carboxylase  $\alpha$  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 3q26-q28 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. Inher. 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.
8. Rybak, J.N., et al. 2005. *In vivo* protein biotinylation for identification of organ-specific antigens accessible from the vasculature. *Nat. Methods* 2: 291-298.

## CHROMOSOMAL LOCATION

Genetic locus: *Mccc1* (mouse) mapping to 3 B.

## PRODUCT

MCCA (m): 293T Lysate represents a lysate of mouse MCCA transfected 293T cells and is provided as 100  $\mu$ g protein in 200  $\mu$ l SDS-PAGE buffer.

## STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

## APPLICATIONS

MCCA (m): 293T Lysate is suitable as a Western Blotting positive control for mouse reactive MCCA antibodies. Recommended use: 10-20  $\mu$ l per lane.

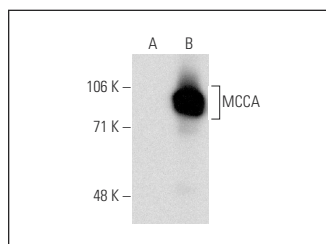
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

MCCA (A-4): sc-271427 is recommended as a positive control antibody for Western Blot analysis of enhanced mouse MCCA expression in MCCA transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

## RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:  
 1) Western Blotting: use m-IgG $\kappa$  BP-HRP: sc-516102 or m-IgG $\kappa$  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.

## DATA



MCCA (A-4): sc-271427. Western blot analysis of MCCA expression in non-transfected: sc-117752 (A) and mouse MCCA transfected: sc-121556 (B) 293T whole cell lysates.

## RESEARCH USE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.