



## Rabbit Anti-MCCC1 antibody

SL18718R

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| <b>Product Name:</b>          | MCCC1                                                                                                                                                                                                                                                                                                                                                                                                                      |
| <b>Chinese Name:</b>          | MCCC1蛋白抗体                                                                                                                                                                                                                                                                                                                                                                                                                  |
| <b>Alias:</b>                 | 3-methylcrotonyl-CoA carboxylase 1; 3-methylcrotonyl-CoA carboxylase biotin-containing subunit; 3-methylcrotonyl-CoA carboxylase, alpha; 3-methylcrotonyl-CoA:carbon dioxide ligase subunit alpha; DKFZp686B20267; FLJ25545; MCC-B; MCCA; MCCase subunit alpha; methylcrotonoyl-CoA carboxylase 1 (alpha); methylcrotonoyl-CoA carboxylase subunit alpha, mitochondrial; methylcrotonoyl-Coenzyme A carboxylase 1 (alpha). |
| <b>Organism Species:</b>      | Rabbit                                                                                                                                                                                                                                                                                                                                                                                                                     |
| <b>Clonality:</b>             | Polyclonal                                                                                                                                                                                                                                                                                                                                                                                                                 |
| <b>React Species:</b>         | Human,Mouse,Rat,Chicken,Dog,Cow,Rabbit,Sheep,                                                                                                                                                                                                                                                                                                                                                                              |
| <b>Applications:</b>          | ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500 (Paraffin sections need antigen repair)<br>not yet tested in other applications.<br>optimal dilutions/concentrations should be determined by the end user.                                                                                                                                                                                         |
| <b>Molecular weight:</b>      | 76kDa                                                                                                                                                                                                                                                                                                                                                                                                                      |
| <b>Cellular localization:</b> | cytoplasmic                                                                                                                                                                                                                                                                                                                                                                                                                |
| <b>Form:</b>                  | Lyophilized or Liquid                                                                                                                                                                                                                                                                                                                                                                                                      |
| <b>Concentration:</b>         | 1mg/ml                                                                                                                                                                                                                                                                                                                                                                                                                     |
| <b>immunogen:</b>             | KLH conjugated synthetic peptide derived from human MCCC1:181-280/725                                                                                                                                                                                                                                                                                                                                                      |
| <b>Lsotype:</b>               | IgG                                                                                                                                                                                                                                                                                                                                                                                                                        |
| <b>Purification:</b>          | affinity purified by Protein A                                                                                                                                                                                                                                                                                                                                                                                             |
| <b>Storage Buffer:</b>        | 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.                                                                                                                                                                                                                                                                                                                                                           |
| <b>Storage:</b>               | Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.                                                                                              |
| <b>PubMed:</b>                | <a href="#">PubMed</a>                                                                                                                                                                                                                                                                                                                                                                                                     |
| <b>Product Detail:</b>        | This gene encodes the large subunit of 3-methylcrotonyl-CoA carboxylase. This enzyme functions as a heterodimer and catalyzes the carboxylation of 3-methylcrotonyl-CoA to                                                                                                                                                                                                                                                 |

form 3-methylglutaconyl-CoA. Mutations in this gene are associated with 3-Methylcrotonylglycinuria, an autosomal recessive disorder of leucine catabolism. [provided by RefSeq, Jul 2008]

**Function:**

Biotin-attachment subunit of the 3-methylcrotonyl-CoA carboxylase, an enzyme that catalyzes the conversion of 3-methylcrotonyl-CoA to 3-methylglutaconyl-CoA, a critical step for leucine and isovaleric acid catabolism.

**Subunit:**

Probably a dodecamer composed of six biotin-containing alpha subunits (MCCC1) and six beta (MCCC2) subunits.

**Subcellular Location:**

Mitochondrion matrix

**DISEASE:**

Methylcrotonoyl-CoA carboxylase 1 deficiency (MCC1D) [MIM:210200]: An autosomal recessive disorder of leucine catabolism. The phenotype is variable, ranging from neonatal onset with severe neurological involvement to asymptomatic adults. There is a characteristic organic aciduria with massive excretion of 3-hydroxyisovaleric acid and 3-methylcrotonylglycine, usually in combination with a severe secondary carnitine deficiency. Note: The disease is caused by mutations affecting the gene represented in this entry.

**Similarity:**

Contains 1 ATP-grasp domain.

Contains 1 biotin carboxylation domain.

Contains 1 biotinyl-binding domain.

**SWISS:**

Q96RQ3

**Gene ID:**

56922

**Database links:**

[Entrez Gene: 56922](#) Human

[Omim: 609010](#) Human

[SwissProt: Q96RQ3](#) Human

[Unigene: 47649](#) Human

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|  | <p><b>Important Note:</b></p> |
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This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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