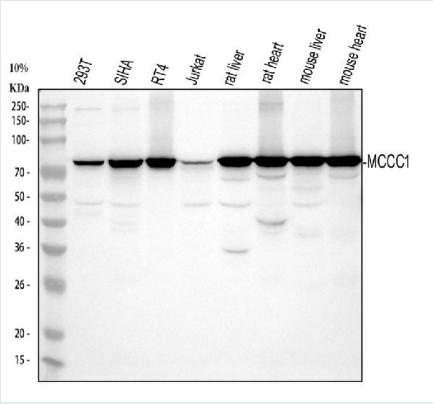
MCCC1 Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	MCCC1, Polyclonal Antibody
Full Product Name	Anti-MCCC1 Antibody Picoband
Product Gene Name	anti-MCCC1 antibody
Product Synonym Names	BAG family molecular chaperone regulator 5; BAG family molecular chaperone regulator 5; BAG family molecular chaperone regulator 5; BAG-5; Bcl-2-associated athanogene 5; BAG5; KIAA0873
Clonality	Polyclonal
Isotype	Rabbit IgG
Host	Rabbit
Species Reactivity	Human, Mouse, Rat
Form/Format	Lyophilized. Each vial contains 4 mg Trehalose, 0.9 mg NaCl, 0.2 mg Na2HPO4.
Purity/Purification	Immunogen affinity purified.
Specificity	Expressed in all tissues examined with lower levels in brain and testis.
Preparation and Storage	At -20 degree C for one year from date of receipt. After reconstitution, at 4 degree C for one month. It can also be aliquotted and stored frozen at -20 degree C for six months.Avoid repeated freezing and thawing.
Molecular Weight	80,473 Da
Chromosome Location	Chromosome: 3; NC_000003.11 (182733006..182817373, complement). Location: 3q27
OMIM	210200
3D Structure	ModBase 3D Structure for Q96RQ3
Immunogen	E coli-derived human MCCC1 recombinant protein (Position: Q104-E715).
Subcellular Localization	Perinuclear region
Reconstitution	Adding 0.2 ml of distilled water will yield a concentration of 500ug/ml.
Cross Reactivity	No cross-reactivity with other proteins.
Protein Function	Inhibits both auto-ubiquitination of PARK2 and ubiquitination of target proteins by PARK2 (By similarity). May function as a nucleotide exchange factor for HSP/HSP70, promoting ADP release, and activating Hsp70-mediated refolding.
Product Categories/Family for anti-MCCC1 antibody	Primary Antibodies; Interleukins
ISO Certification	Manufactured in an ISO 13485:2003 and EN ISO 13485:2012 Certified Laboratory.
Applications Tested/Suitable for anti-MCCC1 antibody	Western Blot (WB), Immunohistochemistry (IHC), Immunocytochemistry (ICC), Immunofluorescence (IF), ELISA (EIA)
Application Notes for anti-MCCC1 antibody	WB: 0.25-0.5ug/ml, Human, Rat IHC: 2-5ug/ml, Human, Mouse, Rat ICC/IF: 5ug/ml, Human ELISA: 0.1-0.5ug/ml

Related Product Information for anti-MCCC1 antibody	The brand Picoband indicates this is a premium antibody that guarantees superior quality, high affinity, and strong signals with minimal background in Western blot applications. Only our best-performing antibodies are designated as Picoband, ensuring unmatched performance.
Product References and Citations for anti-MCCC1 antibody	1. Baumgartner, M. R., Almashanu, S., Suormala, T., Obie, C., Cole, R. N., Packman, S., Baumgartner, E. R., Valle, D. The molecular basis of human 3-methylcrotonyl-CoA carboxylase deficiency. J. Clin. Invest. 107: 495-504, 2001.2. Baumgartner, M. R., Dantas, M. F., Suormala, T., Almashanu, S., Giunta, C., Friebel, D., Gebhardt, B., Fowler, B., Hoffmann, G. F., Baumgartner, E. R., Valle, D. 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, 2004.3. Gallardo, M. E., Desviat, L. R., Rodriguez, J. M., Esparza-Gordillo, J., Perez-Cerda, C., Perez, B., Rodriguez-Pombo, P., Criado, O., Sanz, R., Morton, D. H., Gibson, K. M., Le, T. P., Ribes, A., Rodriguez de Cordoba, S., Ugarte, M., Penalva, M. A. The molecular basis of 3-methylcrotonylglycinuria, a disorder of leucine catabolism. Am. J. Hum. Genet. 68: 334-346, 2001.
Other Notes	Small volumes of anti-MCCC1 antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.
Searchable Terms for MCCC1 purchase	MBS1758815 is an antibody targeting MCCC1. Also known as Anti-MCCC1 Antibody Picoband. Reactive in Human, Mouse, Rat. Host: Rabbit.



Western Blot (WB) of anti-MCCC1 antibody
Figure 1. Western blot analysis of MCCC1 using anti-MCCC1 antibody (MBS1758815).Electrophoresis was performed on a 5-20% SDS-PAGE gel at 70V (Stacking gel)/90V (Resolving gel) for 2-3 hours. The sample well of each lane was loaded with 30 ug of sample under reducing conditions.Lane 1: human SIHA whole cell lysates,Lane 2: human RT4 whole cell lysates,Lane 3: rat liver tissue lysates.After electrophoresis, proteins were transferred to a nitrocellulose membrane at 150 mA for 50-90 minutes. Blocked the membrane with 5% non-fat milk/TBS for 1.5 hour at RT. The membrane was incubated with rabbit anti-MCCC1 antigen affinity purified polyclonal antibody (#MBS1758815) at 0.5ug/mL overnight at 4 degree C, then washed with TBS-0.1%Tween 3 times with 5 minutes each and probed with a goat anti-rabbit IgG-HRP secondary antibody at a dilution of 1:5000 for 1.5 hour at RT. The signal is developed using an Enhanced Chemiluminescent detection (ECL) kit (#MBS176460) with Tanon 5200 system. A specific band was detected for MCCC1 at approximately 75 kDa. The expected band size for MCCC1 is at 80 kDa.

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for MCCC1. It may not necessarily be applicable to this product.

NCBI GI #	116805327
NCBI GeneID	56922
NCBI Accession #	NP_064551.3
UniProt Primary Accession #	Q96RQ3
UniProt Secondary Accession #	Q59ES4; Q9H959; Q9NS97
UniProt Related Accession #	Q96RQ3
GenBank Nucleotide #	NM_020166.3

NCBI Official Full Name	methylcrotonoyl-CoA carboxylase subunit alpha, mitochondrial
NCBI Official Synonym Full Names	methylcrotonoyl-CoA carboxylase 1 (alpha)
NCBI Official Symbol	MCCC1
NCBI Official Synonym Symbols	MCCA; MCC-B
UniProt Protein Name	Methylcrotonoyl-CoA carboxylase subunit alpha, mitochondrial
UniProt Synonym Protein Names	3-methylcrotonyl-CoA carboxylase 1; 3-methylcrotonyl-CoA carboxylase biotin-containing subunit; 3-methylcrotonyl-CoA:carbon dioxide ligase subunit alpha
UniProt Gene Name	MCCC1
UniProt Synonym Gene Names	MCCA; MCCase subunit alpha
UniProt Entry Name	MCCA_HUMAN
NCBI Protein Information	methylcrotonoyl-CoA carboxylase subunit alpha, mitochondrial; MCCase subunit alpha; 3-methylcrotonyl-CoA carboxylase 1; methylcrotonoyl-Coenzyme A carboxylase 1 (alpha); 3-methylcrotonyl-CoA:carbon dioxide ligase subunit alpha; 3-methylcrotonyl-CoA carbox
NCBI Summary for MCCC1	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]
UniProt Comments for MCCC1	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. Ref.9 Catalytic activity: ATP + 3-methylcrotonoyl-CoA + HCO3- = ADP + phosphate + 3-methylglutaconyl-CoA. Ref.9 Cofactor: Biotin. Pathway: Amino-acid degradation; L-leucine degradation; (S)-3-hydroxy-3-methylglutaryl-CoA from 3-isovaleryl-CoA: step 2/3. Subunit structure: Probably a dodecamer composed of six biotin-containing alpha subunits (MCCC1) and six beta (MCCC2) subunits. Subcellular location: Mitochondrion matrix Ref.8. Involvement in 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. Ref.1 Ref.3 Ref.4 Ref.12 Sequence similarities: Contains 1 ATP-grasp domain.Contains 1 biotin carboxylation domain.Contains 1 biotinyl-binding domain. Sequence caution: The sequence BAD92974.1 differs from that shown. Reason: Erroneous initiation.
Research Articles on MCCC1	**1.** 3-methylcrotonyl-CoA carboxylase inhibition has a role in increased excretion of 3-hydroxyisovaleric acid in valproate-treated patients
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