



MCCA mouse mAb

Catalog No	BYmab-11767
lsotype	lgG
Reactivity	Human; Mouse;Rat
Applications	WB
Gene Name	MCCC1 MCCA
Protein Name	MCCA
Immunogen	Synthesized peptide derived from human MCCA AA range: 620-670
Specificity	This antibody detects endogenous levels of MCCA at Human/Mouse/Rat
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	
Cell Pathway	Mitochondrion matrix .
Tissue Specificity	
Function	catalytic activity:ATP + 3-methylcrotonoyl-CoA + HCO(3)(-) = ADP + phosphate + 3-methylglutaconyl-CoA.,cofactor:Biotin.,disease:Defects in MCCC1 are the cause of methylcrotonoyl-CoA carboxylase deficiency type 1 (MCC1 deficiency) [MIM:210200]. MCC1 deficiency is 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.,pathway:Amino-acid degradation; L-leucine degradation; HMG-CoA from 3-isovaleryl-CoA: step 2/3.,similarity:Contains 1 ATP-grasp domain.,similarity:Contains 1 biotin carboxylation domain.,similarity:Contains 1 biotinyl-binding domain.,subunit:Probably a dodecamer composed of

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Background	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],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

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