



# MCCA rabbit pAb

<b>Catalog No</b>	YP-Ab-11767
<b>Isotype</b>	IgG
<b>Reactivity</b>	Human; Mouse;Rat
<b>Applications</b>	WB;IHC
<b>Gene Name</b>	MCCC1 MCCA
<b>Protein Name</b>	MCCA
<b>Immunogen</b>	Synthesized peptide derived from human MCCA AA range: 620-670
<b>Specificity</b>	This antibody detects endogenous levels of MCCA at Human/Mouse/Rat
<b>Formulation</b>	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
<b>Source</b>	Polyclonal, Rabbit,IgG
<b>Purification</b>	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
<b>Dilution</b>	WB 1:500-2000;IHC-p 1:50-300
<b>Concentration</b>	1 mg/ml
<b>Purity</b>	≥90%
<b>Storage Stability</b>	-20°C/1 year
<b>Synonyms</b>	
<b>Observed Band</b>	
<b>Cell Pathway</b>	Mitochondrion matrix .
<b>Tissue Specificity</b>	
<b>Function</b>	<p>catalytic activity:ATP + 3-methylcrotonoyl-CoA + HCO<sub>3</sub><sup>(-)</sup> = 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</p>
<b>Background</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],

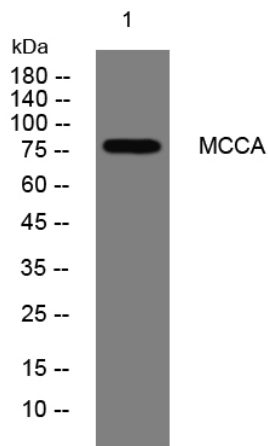
**matters needing attention**

Avoid repeated freezing and thawing!

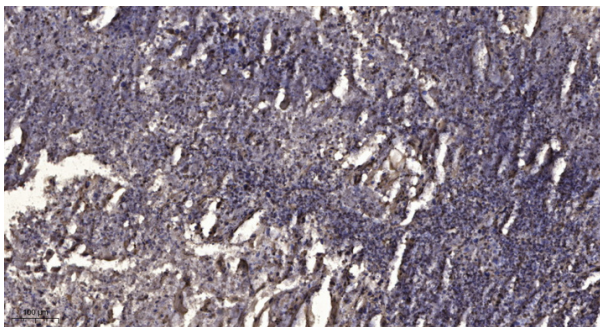
**Usage suggestions**

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

**Products Images**



Western blot analysis of lysates from SH-SY5Y cells, primary antibody was diluted at 1:1000, 4° over night



Immunohistochemical analysis of paraffin-embedded human Squamous cell carcinoma of lung. 1, Antibody was diluted at 1:200(4° overnight). 2, Tris-EDTA,pH9.0 was used for antigen retrieval. 3,Secondary antibody was diluted at 1:200(room temperature, 45min).